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In memory of Ann Judith Maitin and in honor of Dr. David L. Maitin for their love, support, and encouragement.
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Preface

Physical medicine and rehabilitation (PM&R) is a unique field. We have no anatomic region to claim as our terrain, thus we tend to treat the patient as a whole. In PM&R we emphasize functional improvement, and strive to maximize independence, quality of life, mobility, and function.

This text offers insight into the complex specialty of physical medicine and rehabilitation. The field is diverse and dynamic. The book presents a practical approach to the PM&R patient, discusses common pathologies seen, and reviews complications that may present. We emphasize the clinical aspects of PM&R relying on evidence-based medicine. Diagnostic and therapeutic procedures performed by the physiatrist are reviewed, such as electrodiagnostics, musculoskeletal ultrasound, and spine and joint injections.

Current Diagnosis and Treatment: Physical Medicine & Rehabilitation lays down a foundation of anatomy, kinesiology, and biomechanics before progressing to therapeutic exercise and modalities for treatment of pain and dysfunction. The text is completed with a review of primary care issues of concern for the disabled population that we serve.

Practicing physicians, residents and medical students will benefit from the use of this text. Students rotating on physical medicine and rehabilitation services will find that this is a useful introduction to the specialty. Residents will find it helpful to review with this book prior to SAE exams and specialty board exams. Physiatrists who are recertifying or wish to review topics in PM&R will find this text useful. Physicians from other specialties will find the book serves as an efficient means for learning about the field and about particular rehabilitation issues with which their patients may be dealing.

We would like to acknowledge Harriet Lebowitz and Brian Belval for their vision, patience, and invaluable assistance in bringing this project to fruition. We also thank Donna Frassetto for her meticulous editing of the text. We are appreciative of all of the Temple and Moss faculty and alumni, as well as other colleagues, for their tireless efforts in completing their contributions to the book.

I thank my family, Pamela, Maxwell, and Asher, for their support, assistance, and patience during the completion of this book. They have stood by me throughout this prolonged journey.
Ian B. Maitin, MD, MBA
The field of physical medicine and rehabilitation (PM&R) emerged in the 1930s to focus on musculoskeletal and neurologic issues of patients, and further evolved after World War II as veterans returned to the United States with disabling injuries. Restoration of functional ability was established as a fundamental goal of this new field. In 1947, the American Board of Medical Specialties granted PM&R the title of an independent specialty, also known as physiatry.

PM&R focuses on the prevention, diagnosis, and treatment of disorders related to the nerves, muscles, and bones that may produce temporary or permanent impairment or disability. PM&R is often called the “quality of life profession,” because its goal is to enhance patient performance and improve function. The focus is on quality of life—medically, socially, emotionally, and vocationally—after an injury or disease. The approach to the patient is team based, with the physician, rehabilitation nurse, physical therapist, occupational therapist, speech language pathologist, rehabilitation psychologist, prosthetist/orthotist, social worker, therapeutic recreational therapist, and vocational counselor working in conjunction as a treatment team.

The team considers the patient’s progress, future needs, and discharge
planning on a weekly basis. The role of the physiatrist is to act as a medical leader of the team and guide the medical and therapy treatment. The interdisciplinary team promotes regular structured communication among all the members of the team to establish and accomplish the treatment goals. The goal of an inpatient rehabilitation facility is the return of the patient to a safe and functional environment, preferably his or her home or a community-based facility.

Hospital for Special Surgery. What is physiatry? Available at: http://www.hss.edu/what-is-physiatry.asp.

CLINICAL SKILLS

Patient History

The PM&R history is a medicolegal document that follows the format used by other medical disciplines with the addition of key elements that are unique to physiatry. It serves as a tool of communication for members of the rehabilitation team, as well as nonrehabilitation health care professionals, the patient’s health insurance providers, and at times the facilities responsible for ongoing care postdischarge from an acute inpatient rehabilitation unit. Depending on the setting of patient care, the PM&R history may vary from a focused outpatient physiatric evaluation to a comprehensive inpatient assessment. Some patients, especially those being admitted to an acute inpatient rehabilitation unit, may have complex medical problems requiring input and confirmation of the history from the rehabilitation team members. Gathering a complete patient history can require several days as it often depends on input from the physiatrist, other members of the rehabilitation team, and the patient’s family members or caretakers.

A. Chief Complaint

The chief complaint of the rehabilitation patient is the primary concern that led
the patient to seek medical and rehabilitation care. The chief complaint is purely subjective and when possible should be documented in the patient’s own words. In many cases, patients who have sustained stroke, traumatic brain injury or other diseases or injuries causing cognitive alterations will not be able to state a chief complaint. In these cases, it is acceptable for the physician gathering the history to specify the reason for admission as the chief complaint. The chief complaint for a patient admitted to an inpatient rehabilitation service is often associated with ambulation, activities of daily living, communication, or cognition. In the outpatient setting, the patient may have several reasons for seeking physiatric treatment. It is imperative to have the patient rank the complaints in order of most problematic to least bothersome, and to separate those problems that are unrelated to the chief complaint.

B. History of Present Illness

The history of present illness is a detailed account of the chief complaint for which the patient is seeking rehabilitation treatment. It examines information related to the chief complaint, including location, onset, quality, quantity, modifying factors, duration, and associated symptoms and signs. The history of present illness, when skillfully navigated by the physician, can be a valuable encounter between the patient and physician as it serves to establish the physician–patient relationship through the process of gathering information. As part of the history of present illness, details regarding current functional impairments, bowel and bladder impairments, and skin issues relating to the chief complaint should be solicited.


C. Past Medical and Surgical History

Details of the patient’s past medical and surgical history allow the rehabilitation team and the leading physician to formulate an appropriate rehabilitation plan of care that includes necessary precautions that should be in place given the patient’s previous history. This information can alter the patient’s rehabilitation course. When interviewing a patient with possible cognitive impairments, knowledgeable family members, friends, and caretakers should also be interviewed. The interviewer should ask about the patient’s history of
cardiopulmonary disease and associated surgical treatments to ensure that the rehabilitation program does not exceed the patient’s cardiopulmonary limitations. Functional limitations from pulmonary or cardiac etiologies should be noted, as should the modifiable risk factors for cardiac disease, such as smoking, hypertension, and obesity. Similarly, a history of musculoskeletal and rheumatologic disorders and related procedures should be sought. The functional impact of any premorbid disorders should be noted as the patient’s baseline. The patient’s history of neurologic ailments should also be solicited as this can help paint a picture of the premorbid functional level.


D. Family History

It is important to ask about a family history of cardiac disease, cancer, stroke, arthritis, diabetes, neurologic disease, hypertension, psychiatric disorders, and substance abuse. Because rehabilitation patients frequently experience pain and require treatment with appropriate medications, it is important to determine any patient or family history of alcohol or drug abuse.


E. Medications

Documentation of all prescription and over-the-counter medications and supplements is an important element of the history as inaccurate medications can adversely impact the patient’s wellbeing and safety. In 2005, the Joint Commission established medication reconciliation—the process of comparing a patient’s medication orders to all of the medications the patient has been taking—as its National Patient Safety Goal number eight in an effort to minimize polypharmacy-related errors (omissions, duplications, inaccurate dosages, and drug interactions) and promote systematic implementation of medication reconciliation procedures across patient care settings, particularly those
involving transitions from one type or level of care to another.


F. Allergies

The patient’s allergies to medications (including but not limited to major classes of antibiotics), intravenous dye, latex, and various foods should be obtained and carefully documented. The patient or the person providing the history should be questioned in detail regarding the past consequences of exposure to the particular allergen.

G. Social History

A social history describes the personal, vocational, and recreational aspects of the patient’s life that bear clinical significance. Information about the patient’s occupation, activities of daily living, social support, stresses, financial situation, insurance coverage, and recreational habits is included. Complete functional information is also obtained, such as the use of assistive devices, need for assistance, and ability to ambulate distances.

Particular importance should be given to the patient’s environment and living arrangements; for example, whether the patient lives in a house or an apartment, the number of stories in the house or floor on which the apartment is located, whether it is necessary to negotiate stairs to obtain access to the home, and how many steps there are. Relevant information includes whether the stairs have a handrail, and on which side; whether there is elevator access; and home wheelchair accessibility. The location of the bedroom and bathroom should be noted, along with the presence or absence of grab bars in the shower. Much of this information is unique to the field of PM&R because a patient’s functional status after discharge depends on his or her ability to negotiate the physical environment of the home. Prior to discharge, the occupational therapist may visit the home to assess the types of equipment or modifications to the home that will be necessary for a safe discharge. In all cases it is important to inquire about the
patient’s support system, including family, friends, and caretakers, and the extent of assistance that can be provided upon discharge. The need for a home health aide or nursing staff to fill any voids in the care of the patient can then be identified.

Documentation of the patient’s recreational habits, including history of smoking, alcohol, and drug use, is imperative. This information should be sought in an open-ended and nonjudgmental manner. Similarly patients should be asked about their sexual history and any unsafe practices in the past. Data should be gathered regarding the patient’s hobbies and recreational pursuits. Level of education and occupation should also be documented. If the patient’s injuries prevent full return to his or her previous occupation, the need for vocational rehabilitation should be identified. Environmental modifications and assistive devices often make it possible for patients to return to their jobs.

**H. Review of Systems**

The end of the physiatric interview should include a complete symptom checklist addressing all of the vital physiologic systems (Table 1–1). The review of systems should generally begin with an open-ended question such as, “Are you having any other problems that we have not discussed?” The physician can then pose a series of questions about specific health-related problems, prompting the patient to elaborate on areas that are problematic for him or her. Each system should be approached in a systematic fashion. Patients who give a positive response throughout the review of systems, indicating problems in every health-related area, may be engaging in symptom amplification in an attempt to gain attention and emotional support.

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**Table 1–1** Sample findings: review of systems.
### Physical Examination

The physiatric examination is an extension of a thorough general, neurologic, and musculoskeletal physical examination. As in any physical examination an initial assessment and documentation of the patient’s vital signs (temperature, heart rate, blood pressure, and respiratory rate) is customary. An assessment of the cardiac, pulmonary, and abdominal systems is a necessary component of the physiatric examination. Specific areas that constitute a primary focus of the physiatrist are described in detail below.

<table>
<thead>
<tr>
<th>System</th>
<th>Sample Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>General</td>
<td>Fevers, chills, fatigue, appetite, unintentional weight loss</td>
</tr>
<tr>
<td>HEENT</td>
<td>Sinus congestion, nasal bleeding, visual changes, hearing loss, ringing ears, sore throat, headaches</td>
</tr>
<tr>
<td>Lungs</td>
<td>Shortness of breath, sputum, chest pain</td>
</tr>
<tr>
<td>Heart</td>
<td>Palpitations, shortness of breath, chest pain</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Appetite, nausea, vomiting, diarrhea, constipation, bleeding, incontinence</td>
</tr>
<tr>
<td>Genitourinary</td>
<td>Painful urination, frequency, incontinence, blood in urine</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Joint pain, back pain, stiffness, muscle pain, weakness</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Dizziness, numbness, loss of balance, speech or swallow problems</td>
</tr>
</tbody>
</table>

HEENT, head, eye, ear, nose, throat.
A. Cognitive Function

1. Mental status—The patient’s mental status is evaluated with questions aimed at determining but not limited to the patient’s orientation, attention, recall, visuospatial abilities, and language. The patient’s responses during the mental status examination can also provide insight into his or her language ability, medical deficits, and coherence of thinking. During this time the patient’s speech and language pattern can be noted and documented.

2. Consciousness—It is essential to document the patient’s level of consciousness. Consciousness is the state of being aware of one’s surroundings. A lethargic patient shows general slowing of movements and speech but can be easily aroused. Obtundation describes a dulled or blunted state in which the patient is difficult to arouse and once aroused is still confused. Stupor is a state of semiconsciousness in which the patient can be temporarily aroused by stimuli such as pain or noise. In stupor, eye movements become purposeful when the stimulus is applied, wincing may be noted, or papillary constriction may occur. The patient has few or no voluntary motor responses. Delirium is a common condition noted in the inpatient setting. It is characterized by acute or subacute onset and a fluctuant or reversible course. Often a state of restlessness and insomnia ensues, followed by obtundation, emotional lability, and visual illusions. Symptoms may worsen at nighttime especially in the elderly, a manifestation referred to as sundowning.

The Glasgow Coma Scale—an objective method of documenting level of consciousness that assesses eye opening, motor response, and verbal response—is used to evaluate patients, particularly those with traumatic brain injury (see Chapter 13). Coma is the state of unresponsiveness in which the patient’s eyes are closed and in which there is an absence of sleep–wake cycles and no interaction of the patient with the environment. Comatose patients cannot be aroused and have no awareness of self or their surroundings. Those in a vegetative state lack awareness of self or the environment, but have intact sleep–wake cycles. In a minimally conscious state, patients have intact sleep–wake cycles and show evidence of inconsistent but reproducible awareness of self or the environment.

3. **Orientation**—Orientation is characterized by the awareness of one’s person, place, and time. This can be assessed during the mental status examination by asking the patient to state his or her name, specify the present location, and give the date (including year and day of the week). Orientation is typically lost in the following order: time, place, and finally, person.

4. **Memory**—The patient’s memory can be tested by asking him or her to recount information pertaining to recent and remote events. Details about illness, dates of hospitalization, and day-to-day recall can serve to test recent memory. When testing memory, especially in a patient who has been hospitalized for a prolonged period, it is best to test objective facts using questions such as, “Who won the World Series?”, or “Who is the president, now and previously?” Remote memory can be tested by asking the patient to relate personal details such as his or her date of birth, marriage date, and names of children. Additionally, the patient may be given a list of at least three words, and then asked to recall the given words after 5 and 10 minutes. In patients with obvious impairment, prompting may be necessary (ie, by giving the patient multiple choices, with one choice being the correct word).

5. **Mood and affect**—Patient mood and affect should be observed and documented. Mood refers to an inner state that is persistent. Affect refers to a feeling or emotion—often momentary—that is experienced in response to an external occurrence or thought. Mood alterations are common findings in patients with brain injuries. The examiner should assess for anxiety, depressed mood, fear, suspicion, irritability, aggression, lability, apathy, or indifference. Open-ended questions addressing the patient’s feelings and spirits can be helpful in assessing mood. Patients with alterations of affect are often described as having a flat, dull, or monotonous affect.

6. **Abstract thinking**—The patient should be asked to interpret abstract statements such as, “a stitch in time saves nine,” “a rolling stone gathers no moss,” or “people who live in glass houses shouldn’t throw stones.” Keep in mind that cultural and language barriers may prevent adequate testing of abstract thinking.

7. **Judgment and insight**—Insight is determined by evaluating the patient’s recognition of his or her medical problems. Judgment can be tested by asking open-ended questions such as, “Why are there laws?”, or “What would you do if you found a stamped, addressed envelope on the street?”
8. **Attention and concentration**—Attention is demonstrated when the patient is alerted by a significant stimulus and sustains interest in it. Concentration refers to the ability to maintain ongoing mental effort despite distractive stimuli. A patient who is inattentive ignores the examiner’s questions or loses interest in them quickly. A patient with impaired concentration is easily distracted by noises, sights, and thoughts while answering questions.

9. **Apraxia**—Apraxia is the inability to perform previously learned motor tasks correctly despite intact comprehension, complete cooperation, and intact motor and sensory function. In testing for apraxia, patients are usually asked to carry out a series of general activities or tasks that their injuries or illness should not have rendered them unable to physically perform. Patients with ideomotor apraxia are unable to carry out motor responses upon verbal command; however, these acts can be carried out spontaneously. For instance, a patient may be unable to brush his or her hair on command but can do so spontaneously. Ideational apraxia is an abnormality in the conception and sequencing of the movement patterns. Patients can be tested for this form of apraxia by asking them to demonstrate how to use a key, comb, or fork.


**B. Communication**

Language is a fundamental basic of human intelligence and key part of social interaction. All aspects of the patient’s language ability should be examined, including naming, spontaneous production of speech, comprehension, repetition, reading, and writing.

1. **Aphasias**—Aphasias are abnormalities of language functions that are not due to defects of vision, hearing, or motor dysfunction. They can be divided into three categories: fluent, nonfluent, and anomic. (Speech disorders are described in detail in Chapter 38.) Anomia, a deficit of naming, is a common finding in aphasic patients. When asked to name an object, patients often compensate for
their deficit by describing the object with circumlocution. Patients with semantic paraphasia are able to identify the object; however, they offer an incorrect but related word in the same category. For example, a fork may be identified as a spoon. In phonemic paraphasia, the word approximates the correct answer but is phonetically incorrect; thus, a pencil may be described by the patient as a “pentinl.” Aphasias should be distinguished from dysarthria (described below), which is indicative of a motor problem.

A patient’s comprehension can be tested by using general yes–no questions such as, “Do elephants fly?”, or “Does the sun rise from the west?” Comprehension can also be assessed by giving simple commands to patients such as “close your eyes.” Commands should be within the patient’s functional limits. Repetition can be tested by asking patients to repeat single words, short sentences, or a string of words. Reading can be tested by asking the patient to read out loud. If patients have a visual impairment, be sure they are using corrective lenses or eyeglasses and that the written print is big enough for them to read. If the patients have sufficient motor control of their dominant hand, writing may be assessed for word order and grammar. Alexia refers to inability to read or comprehend the written word. Agraphia is a deficiency in the ability to write.

2. **Dysarthria**—Dysarthria is a speech disorder in which the mechanism of speech articulation is deficient. However, the content of the speech itself is unaffected and there is no abnormality of the cortical language mechanism. Thus, the patient has intact comprehension of both the written and spoken word. Dysarthria can be characterized by difficulty in phonation, articulation, resonance, or respiration aspects of speech.

3. **Dysphonia**—Dysphonia is dysfunction in the production of sound. Respiratory movement paresis and pulmonary diseases can cause phonation problems. Dysphonia is often accompanied by hypophonia, which is a decrease in the voice volume due to restricted movement of the breathing musculature. Patients usually speak in whispers and are unable to shout. Indirect laryngoscopy can be utilized to examine the vocal cords for paresis. Vocal cords usually separate in inspiration; however, when they are paralyzed an inspiratory stridor can result. Bilateral vocal cord paresis causes patients to speak in whispers. If only one of the vocal cords is weak, the voice can become hoarse and raspy.
C. Cranial Nerve Function

1. Cranial nerve I (olfactory nerve)—Function of the olfactory nerve is tested by asking the patient to smell a general nonirritating aroma such as a mint or coffee. If the patient can identify the smell, then normal functioning of the olfactory nerve can be assumed. Each nostril is tested separately, with the patient’s eyes closed to avoid visual identification of the source of the scent.

2. Cranial nerve II (optic nerve)—Optic nerve function is assessed by funduscopic examination, measurement of visual acuity, and testing the visual fields of each eye separately. An ophthalmoscope should be used in a dark room to dilate the pupils and examine the fundus. Visual acuity should be tested by using the Snellen eye chart placed about 6 m (20 ft) away from the patient. The acuity is defined as a fraction, with 20/20 being normal. As the visual acuity worsens, the denominator increases. Patients with a known visual impairment should be tested wearing their corrective lenses or eyeglasses. Visual fields can be assessed by confrontation. The optic nerve is the afferent pathway in the papillary light reflex, whereas the oculomotor nerve (cranial nerve III) is the efferent pathway.

3. Cranial nerves III, IV and VI (oculomotor, trochlear, and abducens nerves)—The oculomotor, trochlear, and abducens nerves are tested jointly as all are responsible for ocular motility. The oculomotor nerve innervates all extraocular muscles except the superior oblique, which is innervated by the trochlear nerve, and the lateral rectus, which is innervated by the abducens nerve. The oculomotor nerve is also responsible for innervation of the levator palpebrae muscle (eyelid elevator), pupilloconstrictor muscle (pupil constrictor), and ciliary muscle (responsible for lens thickness). The medial rectus muscle is responsible for adduction, and the primary action of the lateral rectus is abduction. Eye elevation occurs through the action of the superior rectus and inferior oblique muscles. Eye depression is achieved by the actions of the inferior rectus and superior oblique muscles. The superior oblique is responsible for downward gaze, especially during adduction of the eye.
The extraocular muscles are tested by asking the patient to track the movement of the examiner’s finger or a target without moving the head. The object is moved in a full range, including horizontally and vertically. The examiner should observe the range of movement and note any paresis, nystagmus, or other abnormalities. Shape and size of the pupils and their reaction to light and accommodation should also be tested. As previously noted, the optic nerve (cranial nerve II) is the afferent pathway in the papillary light reflex, whereas the oculomotor nerve is the efferent pathway. A normal pupillary light reflex should produce constriction in both pupils when a light stimulus is introduced to either eye separately.

4. Cranial nerve V (trigeminal nerve)—The trigeminal nerve is responsible for sensory function of the face and motor function of the mastication muscles. The ophthalmic division innervates the forehead, the maxillary division innervates the cheek, and the mandibular division innervates the jaw. The trigeminal nerve is tested by assessing response to facial touch and temperature sensation. The examiner places a cool object such as a tuning form on both sides of the patient’s face in all three innervated areas and asks if the tuning fork feels the same on both sides of the face. The trigeminal nerve is also responsible for the afferent portion of the corneal reflex via the ophthalmic division. This can be assessed by lightly sweeping a wisp of cotton across the lateral surface of the eye. The patient should blink bilaterally through the efferent pathway of the reflex via the facial nerve (cranial nerve VII). The motor function of the trigeminal nerve can be tested by assessing the function of the mastication muscles. The patient is asked to open and close the jaw, while the examiner assesses for symmetry. The patient can then be asked to clench the jaw shut while the examiner attempts to open it. If jaw strength is normal, the examiner should not be able to open the patient’s jaw.

5. Cranial nerve VII (facial nerve)—The facial nerve is responsible for the function of the facial muscles and sensation of taste from the anterior two thirds of the tongue. The facial nerve can be tested by observing the patient’s face for symmetry. The patient is asked to wrinkle the forehead, close the eyes tightly, smile, show the teeth, and puff out the cheeks while the examiner looks for asymmetry. The examiner then tries to force the eyes open or compress the cheeks while the patient is puffing them out. In patients with a peripheral lesion of the facial nerve, the entire side of the face is weak and the patient cannot fully close the eye; with a central lesion, the forehead should be spared, as well as some ability to close the eye. Taste sensation can be tested by applying cotton-tipped swabs dipped in a sweet, sour, salty, or bitter solution on the patient’s
protruded tongue and asking the patient to identify the taste.

6. Cranial nerve VIII (vestibulocochlear nerve)—The vestibulocochlear nerve carries sensory information from the vestibule and cochlea of the ear to the brain. The vestibular fibers provide information about the position and rotation of the head, which is needed for balance. The cochlear fibers carry the stimuli of hearing. To test the cochlear division of the nerve, the examiner rubs his or her thumb and index finger near each of the patient’s ears while the patient’s eyes are closed. The patient is asked to indicate on which side the noise was heard. The vestibular fibers should be tested in patients with vertigo or equilibrium problems. The Dix Hallpike test helps to distinguish benign positional vertigo from that caused by a lesion of the central nervous system.

7. Cranial nerves IX and X (glossopharyngeal and vagus nerves)—Both the glossopharyngeal and the vagus nerves can be tested by assessing the patient’s gag reflex. To do so, the examiner stimulates the posterior pharyngeal wall with a sterile blunt object such as a tongue blade and observes for the gag reflex. The afferent pathway of this reflex is mediated by the glossopharyngeal nerve and the efferent pathway by the vagus nerve. The vagus nerve also innervates the muscles of the palate, the pharynx, and the larynx. The glossopharyngeal nerve carries taste sensation from the posterior one third of the tongue and sensation from the pharynx, as well as from the middle ear. Any hoarseness of voice should be noted as this can be indicative of a lesion of the recurrent laryngeal nerve, a branch of the vagus nerve. The examiner should also inspect for position and symmetry of the palate, and for elevation of the uvula both at rest and as the patient phonates “aaah.”

8. Cranial nerve XI (spinal accessory nerve)—The spinal accessory nerve is a motor nerve that innervates the trapezius and sternocleidomastoid muscles. It is tested by checking the strength of the shoulder shrug (trapezius) and head rotation (sternocleidomastoid) against resistance. The ipsilateral sternocleidomastoid muscle turns the head to the contralateral side.

9. Cranial nerve XII (hypoglossal nerve)—This motor nerve is examined by inspecting the patient’s ability to protrude the tongue and to maintain strength when the tongue is protruded into the inner surface of the cheeks. The examiner observes for any deviations of the tongue. In patients with a peripheral lesion of the hypoglossal nerve, the tongue points to the side of the lesion, whereas it deviates contralaterally in those with a central lesion.
D. Sensory Function

The sensory examination tests the primary sensory modalities as well as discriminatory sensations. The primary sensory modalities include light touch, pain, and temperature, and deep sensation involving position (proprioception) and vibration. To assess light touch, the examiner can use a cotton-tipped applicator to lightly stroke the skin. Pain and temperature can be assessed using a sanitary safety pin or another sharp clean object. The examiner should alternate between sharp and blunt object examination, asking the patient to state how each object feels to touch. The use of warm–cold stimuli helps to determine the patient’s perception of temperature.

Proprioception and vibration sensation travels via the dorsal columns. To test proprioception, the examiner vertically moves the patient’s toes or the fingers. Each digit should be held on the sides during the upward or the downward motion, and the patient should be asked in which direction the toe or finger is being moved. Vibration sensation can be checked by using a 128-Hz tuning fork placed on a bony prominence of the limbs such as the malleoli or the olecranon. The patient should be asked if he or she can feel the vibration, and to indicate when the vibration can no longer be sensed.

The discriminatory sensory modalities are tested by assessing two-point discrimination, graphesthesia, and stereognosis. Two-point discrimination is most commonly evaluated with the patient’s eyes closed. Two points of stimulus are introduced at a normal distance of separation, depending on the area of the body being tested. The patient should be able to identify the stimuli as two separate stimuli. To test graphesthesia, the patient is asked to close the eyes and
then identify a number, letter, or symbol that is traced onto his or her palm. Similarly, stereognosis is evaluated by asking the patient to close the eyes and then identify a small object placed in his or her hand. The object should be something common and easily identifiable, such as a key or a coin.

**E. Motor Function**

Motor function is mediated by both upper and lower motor neurons. Upper motor neurons originate in the cerebral cortex and brainstem and project to the lower motor neurons in the brainstem and spinal cord. Lower motor neurons project from the brainstem and spinal cord to skeletal muscles. Lesions of the upper or the lower motor neurons can produce weakness. Signs of upper motor neuron lesions include increased muscle tone, hyperreflexia, and positive Babinski and Hoffman signs. Signs of lower motor neuron lesion include decreased reflexes, muscular atrophy, and fasciculations.

1. **Muscle mass**—Muscles should be carefully inspected to determine whether the bulk of muscle mass is adequate. Decreased muscle mass or atrophy can result from a lower motor neuron lesion. Any muscle atrophy, fasciculation, or twitching movements of the muscle should be noted. Muscle mass should be compared side to side. Muscles of the dominant upper limb are often more prominent and should not be confused with decreased muscle mass on the nondominant side.

2. **Coordination**—Four areas of the nervous system must function cohesively for normal motor movement to occur. These are: (1) the motor system, for muscle strength; (2) the sensory system, for position sense (proprioception); (3) the vestibular system, for balance and coordination of eye, head, and body movements; and (4) the cerebellar system, which allows rhythmic movements and steady posture. The cerebellum is divided into the midline, anterior lobe, and lateral hemisphere. Midline lesions usually produce truncal ataxia, in which the patient cannot sit or stand unsupported. To test for this, the patient is asked to sit at the edge of the bed with arms folded so they cannot be utilized for support. Anterior lobe lesions can result in gait ataxia. In contrast to midline lesions, the patient is still able to sit or stand unsupported but is predominantly unstable when walking. Lateral hemisphere lesions can result in limb ataxia. In this situation, the affected limb has difficulty in rapidly correcting and changing direction. Tests that measure limb coordination include the finger-to-nose test and the heel-to-shin test. The finger-to-nose test measures the patient’s ability to perform point-to-point movements. The patient is first asked to extend the index
finger and touch the nose and with the same finger, and then to touch the examiner’s finger.


3. Tone—Tone is resistance of a muscle to passive movement at a joint. When normal, the limb being tested should be able to be moved easily without any resistance to varying direction and speed. Decreased tone can be described as flaccidity or hypotonia and is seen with lower motor neuron disorders and often following acute cerebrovascular insults. Increased tone can manifest as rigidity or spasticity. Rigidity is an increase in resistance of a limb to passive movement which is constant and independent of velocity. Spasticity is an increase in resistance to passive muscle stretch which is velocity dependent. Rigidity is seen in diseases of the basal ganglia. Spasticity is noted in patients with corticospinal tract lesions. Clonus is a cyclic alteration of muscle contraction of the agonist and antagonist muscles in response to a sustained stretch. Clonus is assessed by a quick jerk of the muscle and is usually tested at the ankle.

Muscle tone can be tested using the Modified Ashworth scale or the pendulum test. It is important to instruct the patient to relax prior to either test. The Modified Ashworth Scale is a reliable six-point ordinal scale used in measuring muscle tone that assigns a grade of 0, 1, 1+, 2, 3, or 4, with each grade representing a description of muscle tone. A grade of 0 indicates no increase in muscle tone. A grade of 1 indicates slight increase of muscle tone, manifested by a catch and release or by minimal resistance at the end range of motion when the affected parts are moved in flexion or extension. A grade of 1+ indicates slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the range of motion. A grade of 2 indicates more marked increase in muscle tone through most of the range of motion, but the affected part is easily moved. A grade of 3 indicates considerable increase in muscle tone, with passive movement being difficult. The highest grade, 4, indicates that the affected part is rigid in flexion or extension. In the pendulum test, the patient is first asked to assume the supine position, and then to fully extend the knee and allow it to drop and swing in the motion of a pendulum. Normally, the limb will swing freely for several cycles, whereas a hypertonic limb will immediately return to the starting position.
4. **Involuntary movements**—The presence of involuntary movements should also be evaluated. Tremors are the most common types of involuntary movements. They are relatively rhythmic oscillatory movements, which can be subdivided into three groups: resting tremors, intentional tremors, and postural tremors. As the name implies, resting tremors are most common at rest and may decrease or disappear with voluntary movements. Postural tremors occur when the affected body part is actively maintaining a posture. Intentional tremors are absent at rest and often become pronounced with activity, especially when the target nears. Hemiballismus refers to repetitive, violent swinging movements usually caused by a deficit in the subthalamic nucleus. Chorea are brief, rapid, jerky, nonrepetitive movements. Tics are brief repetitive, coordinated movements occurring at regular intervals such as shoulder shrugging, grimacing, or repetitive winking. Athetosis consists of twisting and writhing movements and is commonly seen in cerebral palsy. Dystonia is a sustained posturing that can affect small or large muscle groups. An example is torticollis, which is caused by sustained posturing of the neck muscles pulling the head to one side.

5. **Reflexes**—Three groups of reflexes are tested: muscle stretch, superficial, and primitive.

**A. Muscle stretch reflexes**—The muscle stretch reflex, or deep tendon reflex, is a muscle contraction in response to stretching within the muscle. Normal muscle stretch reflexes can be elicited by tapping over the muscle tendon with a reflex hammer, resulting in contraction of the muscle whose tendon is stretched (Table 1–2). The patient is positioned into the midrange of the arch of joint motion and instructed to relax in order to elicit a response. The response levels of the deep tendon reflexes are graded from 0 to 4+ (Table 1–3). A grade of 0 indicates no response; 1+ indicates depressed or suppressed reflex; 2+ indicates a normal response; 3+ indicates a response more brisk than usual; and 4+ indicates that the reflex is hyperactive with the presence of clonus. Clonus is a repetitive, sustained cyclic reflex of agonist and antagonist response elicited by manually stretching the tendon.
Table 1–2 Muscle stretch reflexes.

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Innervation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biceps</td>
<td>C5, C6</td>
</tr>
<tr>
<td>Brachioradialis</td>
<td>C5, C6</td>
</tr>
<tr>
<td>Triceps</td>
<td>C7, C8</td>
</tr>
<tr>
<td>Patellar</td>
<td>L3, L4</td>
</tr>
<tr>
<td>Medial hamstrings</td>
<td>L5, S1</td>
</tr>
<tr>
<td>Achilles</td>
<td>S1, S2</td>
</tr>
</tbody>
</table>

Table 1–3 Muscle stretch reflex grading.

<table>
<thead>
<tr>
<th>Grade</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No response</td>
</tr>
<tr>
<td>1+</td>
<td>Diminished response</td>
</tr>
<tr>
<td>2+</td>
<td>Normal response</td>
</tr>
<tr>
<td>3+</td>
<td>Brisk response</td>
</tr>
<tr>
<td>4+</td>
<td>Hyperactive with clonus</td>
</tr>
</tbody>
</table>

B. SUPERFICIAL REFLEXES—Superficial reflexes are motor responses to scraping of the skin. These reflexes are graded as present or absent, with prominently irregular responses graded absent as well. The plantar reflex is the most common superficial reflex and is elicited by applying a stimulus on the sole of the foot from the lateral border to up and across the ball of the foot. Flexion of the big toe or no response is normal; an abnormal response consists of dorsiflexion of the big toe with fanning of the other toes. This response suggests dysfunction of the corticospinal tract and is known as the Babinski sign. Other noteworthy signs are the Chaddock sign (dorsiflexion of the big toe when a stimulus is applied from the lateral ankle to the lateral foot) and the Stransky sign (which occurs when the little toe is flipped outward and results in an
upturned great toe).

C. PRIMITIVE REFLEXES—Primitive reflexes are an abnormal finding in older children and adults and represent a reversion to a more infantile level of reflex activity, suggesting significant neurologic abnormalities. The rooting and sucking reflexes are elicited, respectively, by stroking the patient’s cheek or stimulating the area around the mouth. In response, the patient turns toward that side and makes sucking motions with the mouth. The grasp reflex is elicited by placing a finger onto the patient’s open palm, resulting in a tightened grip when the examiner attempts to remove the finger. The palmomental response—a sudden contraction of the mentalis, or chin, muscle when the palm of the hand is quickly scratched—suggests unilateral damage of the pre-frontal area of the brain. Finally, the snout reflex is pursing of the lip as a response to a tap right above or below the mouth.


F. Musculoskeletal System

Examination of the musculoskeletal system can be one of the most complex aspects of the general physical examination. The extent of the examination may vary depending on the problems being assessed. Throughout the examination, attention should be directed to both function and structure. The examination should be symmetric. The musculoskeletal examination incorporates inspection, palpation, assessment of joint stability, range of motion, manual muscle testing, and special tests (eg, Hawkins, Neer [for the shoulder], Patrick [for the hip], and Lachman [for the knee]; refer to Chapter 30).

1. Inspection and palpation—Inspection of the musculo-skeletal system begins with observation of the patient during the history portion of the evaluation. The examiner inspects the limbs for symmetry, circumference, and shape, and the spine for common deformities such as scoliosis, kyphosis, and lordosis. In patients with amputation, the residual limb should be examined for level of
amputation, length, and contour. The examiner also assesses the surrounding tissues, noting any skin changes, subcutaneous nodules, masses, edema, scarring, and muscle atrophy. The joints should be evaluated for swelling, warmth, tenderness, redness, and abnormal positioning. Following inspection, palpation is performed to support the initial impressions from inspection. Depending on the clinical situation, joints and muscles should be further assessed for tight muscle bands, tone, crepitus, and fractures.

2. Joint stability—Evaluation of joint stability should include assessment of bony consistency, capsular and cartilaginous integrity, and the strength of ligaments and muscles. Before assessing the involved joint in a patient with compromised function, the examiner should evaluate the noninvolved side as an aid to understanding the patient’s biomechanics. Assessment of joint stability should start with identifying pain, guarding, or resistance in the involved joint. The next step should be an evaluation of joint play to assess end feel, capsular patterns, and joint mobility.

Joint play or capsular patterns assess the integrity of the capsule in a position in which there is minimal bony contact with maximum capsular laxity. To achieve maximum range of motion of the involved joint, it is important that the examiner assess the joint through passive range of motion. The end feel of a joint is determined by the structure of the joint (ie, the tendons, adjacent bones, and muscle attachments that keep the joint in place) and can range from hard to soft. The type of structure that limits the range of motion will usually determine the type of end feel noted. With hip flexion, the end feel is soft due to significant muscle bulk surrounding the hip joint. With passive extension of the hip at full range of motion, the end feel is firm due to tension in the anterior capsule of the hip and tension on the hip flexor muscles. However, firmness that occurs before the end point of range can be a sign of increased tone or capsular tightening, or both. A hard end feel is normally seen with elbow extension because the bones join to lock the elbow in place at the end of that motion. In an arthritic joint a hard end feel can occur before full range is achieved. An empty end feel is recorded when no real end feel is reached, such as when the test is aborted due to pain and tenderness.

Hypomobile joints increase the risk for muscle strains, tendonitis, and nerve entrapments. Hypermobile joints increase the risk for joint sprains and degenerative joint disease. Inflammation of the synovium can cause a hypermobile joint and reduce the strength of the capsule. Associated muscle weakness will further increase the risk of trauma and joint instability. Radiographic imaging can be helpful in cases of suspected joint instability; for instance,
flexion–extension spinal films can help assess vertebral column instability.

3. **Range of motion**—Joint flexibility is the range of motion (ROM) tolerated at a joint. Examining ROM establishes the existing mobility present in the joint being evaluated, which should then be compared with the unaffected joint. Goals and a treatment plan to increase or decrease the ROM can then be developed. ROM testing can also aid in diagnosing and determining the patient’s joint function. This provides information regarding limitations if joint disease is suspected. Hypermobility or hypomobility of joints affect the patient’s ability to perform activities of daily living. An example of joint hypomobility hindering a person’s daily living activities is an inability to climb stairs due to a 70-degree restriction in knee flexion. Additionally, using joint ROM, the examiner can reassess the patient’s status after treatment and compare it with the baseline at the time of initial treatment.

Factors affecting ROM include age, sex, joint structure, and muscles. Normally, the younger the subject, the greater is the ROM. Depending on age and specific joint action, males have a more limited range than females. Some individuals have hypermobile or hypomobile joints owing to genetics or posture. Certain muscles associated with the joint may become stretched or contracted, thereby affecting the joint motion. Passive ROM is the amount of joint motion possible without assistance from the subject. It is usually greater than active ROM because the integrity of the soft tissue structure does not determine the limits of the motion. Integrity of the joint can be assessed through the completion of this test. Active ROM is performed by the patient through all planes of motion without assistance from the examiner. Active ROM wholly evaluates coordination of movement and functional ability and provides limited information regarding joint motion.

ROM testing should be completed before strength testing. Using an instrument called a goniometer, the examiner measures joint ROM in the three cardinal planes of motion: sagittal, coronal, and frontal. The sagittal plane separates the body into left and right halves. The frontal (coronal) plane divides the body into anterior and posterior halves, and the transverse plane divides the body into superior and inferior parts. Measurement of each arc of motion should begin at 0 degrees and proceed toward 180 degrees. Most joints in the anatomic position are at 0 degrees of motion. As joint motion occurs, the amount of joint motion is positively recorded in degrees. For example, in shoulder forward flexion, the normal range for flexion in the 180-degree system is 0–180 degrees, and for extension, 0–60 degrees.

Many practitioners suggest obtaining several measurements and recording a
mean value to increase accuracy. Measurement inaccuracy in the limbs can be as high as 10–30% and without value in the spine if built on visual assessment alone. In joint deformity, the starting position is the starting point of joint motion. Spinal ROM is more difficult to measure, and its reliability has been debated. Radiographs provide the most accurate method of measuring spinal motion.

4. **Muscle strength**—Manual muscle testing is a procedure for evaluating the function and strength of individual muscles and muscle groups based on the effective performance of a movement in relation to the forces of gravity and manual resistance. When performing strength testing, a particular muscle or muscle group is first isolated, then an external force is applied. Manual muscle testing specifically measures the ability to voluntarily contract a muscle or muscle group at a specific joint.

Factors influencing manual muscle test results (and strength) include the effect of gravity, manual force used by the examiner, extent of the injury, cognitive and emotional factors of both the patient and the examiner, number of motor units firing, cross-sectional area of the muscle, line of pull of the muscle fibers, number of joints crossed, sensory receptors, attachments to bone, patient age, fatigue, fear, and misunderstanding. Muscles can be difficult to assess in isolation. For example, the biceps, brachialis, and brachioradialis all assist with elbow flexion. Pain can result in breakaway weakness due to pain inhibition of function and should be documented as such. The presence of substitution should be noted with weak muscles or when movement is uncoordinated. In females, there is an increase in strength until age 20, followed by a plateau and then decline after age 30. Males have an increase in strength until age of 20 and then plateau until somewhat older than 30 years before declining. Type 1 muscle fibers, which are fatigue resistant, sometimes require extended stress on testing to uncover a weakness. An example of a muscle with primarily type 1 muscle fibers is the quadriceps. Type 2 muscle fibers, which fatigue quickly, require less stress on muscle testing to uncover deficits. An example of a muscle with type 2 fibers is the sternocleidomastoid. Patients who cannot actively control muscle tension (eg, those with spasticity from central nervous system disease) should not be evaluated using the standard manual muscle testing methods.

When performing manual muscle testing, the examiner should evaluate for asymmetry of muscle groups. All tests must be performed bilaterally, and the unaffected side should be tested first. Most examiners use the Medical Research Council scale, which grades results using a range from 0 to 5. Grade 0 indicates that no contractile activity can be felt in the gravity-eliminated position. Grade 1
indicates the muscle can be palpated while the patient is performing the action in the gravity-eliminated position. Grade 2 indicates that the patient has all or partial ROM in the gravity-eliminated position. Grade 3 indicates the patient can tolerate no resistance but can perform the movements through the full ROM. Grade 4 indicates the patient can hold the position against moderate to strong resistance and has full ROM. Grade 5 indicates the patient can hold the position against maximum resistance and through complete ROM. Tables 1–4 and 1–5 summarize joint movement ranges and innervation for all major upper and lower extremity muscle groups, respectively. The use of a dynamometer can add a degree of objectivity to measurements for pinch and grip.

Table 1–4 Upper extremity testing.
<table>
<thead>
<tr>
<th>Motion (degrees of ROM)</th>
<th>Muscle</th>
<th>Innervation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder flexion (180)</td>
<td>Deltoïd, anterior portion Coracobrachialis</td>
<td>Axillary C5, C6 Musculocutaneous C6, C7</td>
</tr>
<tr>
<td>Shoulder extension (60)</td>
<td>Deltoïd, posterior portion Latissimus dorsi Teres major</td>
<td>Axillary C5, C6 Thoracodorsal C6, C7, C8 Inferior subscapular C5, C6, C7</td>
</tr>
<tr>
<td>Shoulder abduction (180)</td>
<td>Deltoïd, middle portion Supraspinatus</td>
<td>Axillary C5, C6 Suprascapular C5, C6</td>
</tr>
<tr>
<td>Shoulder adduction (30)</td>
<td>Pectoralis major Latissimus dorsi</td>
<td>Medial/Lateral pectoral C5–T1 Thoracodorsal C6, C7, C8</td>
</tr>
<tr>
<td>Shoulder internal rotation (70)</td>
<td>Subscapularis Pectoralis major Latissimus dorsi Teres major</td>
<td>Superior/Inferior subscapular C5, C6 Medial/Lateral pectoral C5–T1 Thoracodorsal C6, C7, C8 Inferior subscapular C5, C6, C7</td>
</tr>
<tr>
<td>Shoulder external rotation (90)</td>
<td>Infraspinatus Teres minor</td>
<td>Suprascapular C5, C6 Axillary C5, C6</td>
</tr>
<tr>
<td>Shoulder shrug</td>
<td>Trapezius Levator scapulae</td>
<td>Spinal accessory (CN XI) C3, C4, dorsal scapular C5</td>
</tr>
<tr>
<td>Elbow flexion (150)</td>
<td>Biceps brachii Brachialis Brachioradialis</td>
<td>Musculocutaneous C5, C6 Musculocutaneous C5, C6 Radial C5, C6</td>
</tr>
<tr>
<td>Elbow extension (10)</td>
<td>Triceps brachii</td>
<td>Radial C6, C7, C8</td>
</tr>
<tr>
<td>Forearm pronation (90)</td>
<td>Pronator teres Pronator quadratus</td>
<td>Median C6, C7 Anterior interosseous C8, T1</td>
</tr>
<tr>
<td>Forearm supination (90)</td>
<td>Supinator Biceps brachii</td>
<td>Posterior interosseous C5, C6, C7 Musculocutaneous C5, C6</td>
</tr>
<tr>
<td>Wrist flexion (80)</td>
<td>Flexor carpi radialis Flexor carpi ulnaris</td>
<td>Median C6, C7, C8 Ulnar C7, C8, T1</td>
</tr>
<tr>
<td>Wrist extension (70)</td>
<td>Extensor carpi radialis longus Extensor carpi radialis brevis Extensor carpi ulnaris</td>
<td>Radial C6, C7 Radial C6, C7 Posterior interosseous C7, C8</td>
</tr>
<tr>
<td>MCP flexion (90)</td>
<td>Lumbricales Interossei</td>
<td>Median, ulnar C8, T1 Ulnar C8, T1</td>
</tr>
<tr>
<td>PIP flexion (100)</td>
<td>Flexor digitorum superficialis Flexor digitorum profundus</td>
<td>Median C7–T1 Ulnar C8, T1</td>
</tr>
<tr>
<td>DIP flexion (80)</td>
<td>Flexor digitorum profundus</td>
<td>Median, ulnar C7, C8, T1</td>
</tr>
<tr>
<td>MCP extension (20)</td>
<td>Extensor digitorum Extensor indicis Extensor digiti minimi</td>
<td>Posterior interosseous C7, C8 Posterior interosseous C7, C8 Posterior interosseous C7, C8</td>
</tr>
<tr>
<td>Digit abduction (20)</td>
<td>Dorsal interossei Abductor digiti minimi</td>
<td>Ulnar C8, T1 Ulnar C8, T1</td>
</tr>
<tr>
<td>Digit adduction (touch adjacent finger)</td>
<td>Palmar interossei</td>
<td>Ulnar C8, T1</td>
</tr>
<tr>
<td>Thumb opposition</td>
<td>Opponens pollicis Flexor pollicis brevis Abductor pollicis brevis</td>
<td>Median C8, T1 Median, ulnar C8, T1 Median, C8, T1</td>
</tr>
<tr>
<td>Thumb flexion (90)</td>
<td>Flexor pollicis brevis Flexor pollicis longus</td>
<td>Median, ulnar C8, T1 Anterior interosseous C7, C8, T1</td>
</tr>
</tbody>
</table>
Table 1–5 Lower extremity testing.

<table>
<thead>
<tr>
<th>Movement</th>
<th>Muscle</th>
<th>Nerve(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thumb extension (15)</td>
<td>Extensor pollicis brevis</td>
<td>Posterior interosseous C7, C8</td>
</tr>
<tr>
<td></td>
<td>Extensor pollicis longus</td>
<td>Posterior interosseous C7, C8</td>
</tr>
<tr>
<td>Thumb abduction</td>
<td>Abductor pollicis longus</td>
<td>Posterior interosseous C7, C8</td>
</tr>
<tr>
<td></td>
<td>Abductor pollicis brevis</td>
<td>Median C8, T1</td>
</tr>
<tr>
<td>Thumb adduction</td>
<td>Adductor pollicis</td>
<td>Ulnar C8, T1</td>
</tr>
</tbody>
</table>

ROM, range of motion; CN, cranial nerve; MCP, metacarpophalangeal; PIP, proximal interphalangeal; DIP, distal interphalangeal.
<table>
<thead>
<tr>
<th>Motion (degrees of ROM)</th>
<th>Muscle</th>
<th>Innervation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip flexion (100)</td>
<td>Iliacus, Psoas, Tensor fascia lata, Rectus femoris, Pectineus, Adductor longus, brevis, anterior portion of magnus</td>
<td>Femoral, L2–L4, Lumbar plexus, L1–L4, Superior gluteal, L4, L5, S1, Femoral, L2–L4, Femoral or obturator, L2, L3, Obturator, L2–L4</td>
</tr>
<tr>
<td>Hip extension (30)</td>
<td>Gluteus maximus</td>
<td>Inferior gluteal, L5, S1, S2</td>
</tr>
<tr>
<td>Hip abduction (40)</td>
<td>Gluteus medius, minimus, tensor fascia lata</td>
<td>Superior gluteal, L4, L5, S1</td>
</tr>
<tr>
<td>Hip adduction (20)</td>
<td>Adductor brevis, longus, Adductor magnus, anterior portion of magnus</td>
<td>Obturator, L2–L4, Obturator, L3, L4, Femoral or obturator, L2, L3</td>
</tr>
<tr>
<td>Hip internal rotation (40)</td>
<td>Tensor fascia lata, Pectineus, Gluteus minimus, anterior portion</td>
<td>Superior gluteal, L4, L5, S1, Femoral or obturator, L2, L3, Superior gluteal, L4, L5, S1</td>
</tr>
<tr>
<td>Hip external rotation (60)</td>
<td>Piriformis, Gluteus maximus, Superior gemelli or obturator internus, Inferior gemelli or quadratus femoris</td>
<td>Nerve to piriformis, S1, S2, Inferior gluteal, L5, S1, S2, Nerve to obturator internus, L5, S1, S2, Nerve to quadratus femoris, L4, L5, S1</td>
</tr>
<tr>
<td>Knee flexion (135)</td>
<td>Semitendinosus, semimembranosus, Biceps femoris</td>
<td>Tibial portion of sciatic, L5, S1, Tibial portion of sciatic, L5, S1, S2</td>
</tr>
<tr>
<td>Knee extension (10)</td>
<td>Quadriceps femoris</td>
<td>Femoral, L2–L4</td>
</tr>
<tr>
<td>Ankle dorsiflexion (20)</td>
<td>Tibialis anterior, extensor digitorum longus, extensor hallucis longus</td>
<td>Deep peroneal, L4, L5, S1</td>
</tr>
<tr>
<td>Ankle plantar flexion (40)</td>
<td>Gastrocnemius, soleus</td>
<td>Tibial, S1, S2</td>
</tr>
<tr>
<td>Ankle inversion (30)</td>
<td>Tibialis anterior, Tibialis posterior, Flexor digitorum longus, Flexor hallucis longus</td>
<td>Deep peroneal, L4, L5, S1, Tibial, L5, S1, Tibial, L5, S1, S2</td>
</tr>
<tr>
<td>Ankle eversion (20)</td>
<td>Extensor digitorum longus, Peroneus longus brevis</td>
<td>Deep peroneal, L4, L5, S1, Superficial peroneal, L4, L5, S1</td>
</tr>
<tr>
<td>First digit IP extension (0)</td>
<td>Extensor hallucis longus</td>
<td>Deep peroneal, L4, L5, S1</td>
</tr>
<tr>
<td>Second to fifth digit PIP extension (0)</td>
<td>Extensor digitorum longus, Extensor digitorum brevis</td>
<td>Deep peroneal, L4, L5, S1</td>
</tr>
<tr>
<td>First digit IP flexion (60)</td>
<td>Flexor hallucis longus, Flexor hallucis brevis</td>
<td>Tibial, L5, S1, S2, Medial plantar, L5, S1</td>
</tr>
<tr>
<td>Second to fifth digit PIP flexion (35)</td>
<td>Flexor digitorum longus, Flexor digitorum brevis</td>
<td>Tibial, L5, S1, Medial plantar, L5, S1</td>
</tr>
</tbody>
</table>

IP: interphalangeal; PIP: proximal interphalangeal.
Dynamic screening tests of strength can also be performed. Examples include:

- Deep knee bend (squat and rise)—proximal lower limb screening.
- Walking on heels and toes—distal lower extremity screening.
- Asking the patient to hold two of the examiner’s fingers while the examiner attempts to free the fingers by pulling in all directions—upper extremity screening.
- Observing the patient change positions from supine to sitting with the hips and knees bent—abdominal strength screening.

The iliopsoas is tested when the hips and knees are extended. Gait abnormalities can be made obvious by asking a patient to increase his or her walking speed and to walk sideways and backward.

The study of functional anatomy—human anatomy as it relates to function—is an undertaking vital to physiatrists and any medical professionals who care for patients with disabilities or injuries of the musculoskeletal system. This chapter provides an overview of functional anatomy organized by common clinical problems framed from the perspective of anatomic study. There are many excellent resources for further study, which is essential for mastery of this topic. Some of these are listed within this chapter.

THE UPPER LIMB

CLINICAL PROBLEM: SCAPULAR WINGING (Figure 2–1)
The trapezius muscle (Figure 2–1) performs the following functions:

- It stabilizes the scapula (Figure 2–2) proximally in the coronal plane via retraction toward the rib cage and spinous processes.
- It rotates the scapula upward in the coronal plane to maximize upward and outward reach of the arm.
- It rotates the scapula to optimize the length–tension relationship for the other shoulder abductor muscles, including the deltoids and the rotator cuff.
allowing for the most efficient and forceful contraction.

Figure 2–2 Scapula. (Reproduced with permission from Morton DA: The Big Picture: Gross Anatomy. McGraw-Hill, 2011.)

The serratus anterior muscle (Figure 2–1) stabilizes the scapula in the sagittal plane, rotates the scapula upward in the sagittal plane, and protracts the scapula. This produces a combination of lateral excursion in the coronal plane, internal rotation, and anterior translation in the sagittal plane. Weakness of the serratus produces scapular dyskinesis in the sagittal plane, most notably during flexion of the arm.

Management Considerations

The arm can be abducted 180 degrees in the coronal plane. Of this motion, 120 degrees occurs at the glenohumeral joint and 60 degrees occurs at the scapulothoracic joint. Weakness of the trapezius causes impairment in arm abduction and produces a form of scapular dyskinesis. Scapular dyskinesis, or alteration in the normal scapulohumeral rhythm, results in a counter-rotation of the scapula downward, due to the direction of deltoid pull and the unopposed force of gravity on the upper limb. Humeral abduction in the coronal plane can
be limited to as little as 60 degrees of abduction due to the limitation of scapulothoracic joint motion. Shoulder abduction in the upright position causes the scapula to wing laterally in a patient with trapezius weakness.

A patient with serratus weakness may not have noticeable winging when standing upright with the arms at the sides. However, when posterior force is exerted through the humerus onto the scapula, medial winging of the entire vertebral border of the scapula is noted. (This is a more effective way to assess for winging than having the patient place the arms on a wall or having him or her perform a pushup. Patients with serratus weakness may have difficulty performing these maneuvers correctly because of concomitant weakness of other necessary musculature.)

**CLINICAL PROBLEM: ROTATOR CUFF PATHOLOGY**

**Pertinent Anatomy**

The rotator cuff is composed of the supraspinatus, infraspinatus, teres minor, and subscapularis muscles (Figure 2–3). The first three muscles originate from the posterior scapula (Figure 2–2) and insert on the greater tuberosity of the humerus, functioning to externally rotate the humerus. The subscapularis originates on the anterior fossa of the scapula and inserts on the lesser tuberosity of the humerus.
Management Considerations

Collectively, the rotator cuff muscles confer dynamic stability to the glenohumeral joint by holding the humeral head centered within the glenoid fossa. The rotator cuff musculature resists humeral translation and subluxation during the midranges of humeral motion in the coronal or transverse planes. The
subscapularis is the only internal rotator of the humerus that also functions to hold the humeral head in the glenoid dynamically during internal rotation. Thus weakness of the subscapularis may have a more profound impact on strength of internal rotation than weakness in the anterior deltoid or pectoralis muscles. Weakness of the whole rotator cuff produces limited overhead motion as the middle deltoid contraction vector causes anterior–superior subluxation of the humeral head and clearly impairs further glenohumeral joint motion. The rotator cuff muscles function as dynamic stabilizers of the glenohumeral joint by pulling the humeral head into the glenoid fossa throughout flexion, extension, abduction, and adduction of the arm.

**CLINICAL PROBLEM: ACROMIOCLAVICULAR JOINT SEPARATION**

▶ **Pertinent Anatomy**

One of the major joints of the shoulder, the acromioclavicular joint (Figure 2–4) is supported by the acromioclavicular ligament and the coracoacromial ligament. The superior aspect of the acromioclavicular ligament runs horizontally over the joint and provides stability in this direction. The coracoacromial ligament extends to the acromion from a wide base on the coracoid process. The coracoclavicular ligament, while not technically a part of the acromioclavicular joint, is composed of two parts, the trapezoid and conoid ligaments, and attaches the scapula to the clavicle.
Management Considerations

Injury to the shoulder, such as forcibly landing on the shoulder, can produce damage to the acromioclavicular joint and is termed a shoulder separation. Such an injury impairs scapular motion and thus flexion and abduction of the arm. The degree of damage to the acromioclavicular and coracoclavicular ligaments and degree of resulting displacement of the clavicle relative to the acromion are the criteria for the classification of acromioclavicular separations. Type I injuries involve sprained but intact coracoclavicular and acromioclavicular ligaments. Type II injuries are characterized by a complete disruption of the acromioclavicular ligament with a sprained but intact coracoclavicular ligament.
In a type III injury, both the coracoclavicular and acromioclavicular ligaments are completely disrupted. Type IV injuries are characterized by posterior displacement of the clavicle relative to the acromion with buttonholing through the trapezius muscle. In type V injuries, the clavicle is widely displaced superiorly relative to the acromion as a result of disruption of muscle attachments. Type VI injuries are rare and are characterized by displacement of the distal clavicle below the acromion or coracoid process.

**CLINICAL PROBLEM: GLENOHUMERAL DISLOCATION**

► **Pertinent Anatomy**

Static stabilizers of the glenohumeral joint include the following structures (Figure 2–4):

- Glenohumeral joint capsule
- Superior glenohumeral ligament
- Middle glenohumeral ligament
- Inferior glenohumeral ligament
- Coracohumeral ligament
- Coracoacromial ligament

► **Management Considerations**

With removal of the glenohumeral joint capsule the force required to dislocate the glenohumeral joint is decreased by 20%, indicating that this structure has an important role as a static stabilizer. The superior glenohumeral ligament provides posterior and inferior stability when the humerus is in adduction and external rotation. The middle glenohumeral ligament restrains anterior glenohumeral joint instability when the humerus is in 45 degrees of abduction. The inferior glenohumeral ligament protects from anterior and posterior glenohumeral joint instability. Severing the posterior band of this ligament results in posterior subluxation; however, dislocation will not occur until the rotator interval is disrupted.

**CLINICAL PROBLEM: ANTERIOR INTEROSSEOUS NERVE**
INJURY

Pertinent Anatomy

The flexor digitorum superficialis and the flexor digitorum profundus are anterior forearm muscles (Figure 2–5) that flex the fingers at the proximal interphalangeal joint and the distal interphalangeal joint, respectively. The deep anterior forearm muscle, the pronator quadratus, runs from the distal ulna to the radius with fibers running perpendicular to these bones. The flexor pollicis longus, also of the deep compartment of the anterior forearm, inserts onto the distal phalanx of the thumb and is the only muscle that flexes the interphalangeal joint. All of the aforementioned forearm muscles are innervated by the anterior interosseous branch of the median nerve with the exception of the flexor digitorum superficialis, which is innervated by the median nerve proper, and the medial half of the flexor digitorum profundus, innervated by the ulnar nerve.
Figure 2–5 Flexor forearm. (Reproduced with permission from Morton DA: The Big Picture: Gross Anatomy. McGraw-Hill, 2011.)
Management Considerations

With an injury to the anterior interosseous branch of the median nerve weakness of flexor pollicis longus, flexor digitorum profundus to the index and long finger, and weakness of pronator quadratus is observed. Weakness of the pronator quadratus is difficult to clinically detect as the pronator teres is a strong co-agonist. Flexion of the distal joint of the thumb and first digit is readily observed; the “okay” sign may be used to assess for weakness in the muscles supplied by this nerve. (When asked to join the thumb and index finger together in a round “okay” sign, the patient has flattening of the interphalangeal joint of the thumb due to weakness of the flexor pollicis longus and flattening of the distal interphalangeal joint of the index finger due to weakness of the flexor digitorum profundus.)

CLINICAL PROBLEM: ELBOW PAIN IN A THROWING ATHLETE

Pertinent Anatomy

The humeroulnar, humeroradial, and proximal radioulnar joints form a hinge joint at the elbow. The head of the radius articulates with the capitulum of the humerus, and the ulna articulates with the trochlea. The proximal radius also articulates with the proximal ulna, forming the radioulnar joint. The trochlea is shaped such that it extends more distally than the capitulum, producing the valgus carrying angle of the forearm. A single synovial joint capsule, which receives sensory innervation mainly from the musculocutaneous and radial nerves, is present at the elbow surrounding all three joints.

The elbow joint is protected anteriorly and posteriorly by the muscles that cross the joint and medially and laterally by ligaments. The ulnar collateral ligament is made up of three bands: anterior, posterior, and transverse. The anterior band originates at the medial epicondyle of the humerus and inserts on the coronoid process of the ulna. The posterior band also originates at the medial epicondyle and inserts onto the olecranon process of the ulna. The transverse fibers originate and insert on the same bone, the ulna, originating at the olecranon and inserting onto the coronoid process. On the lateral aspect of the elbow lies the radial collateral ligament, which originates at the lateral epicondyle and attaches to the annular ligament. The annular ligament wraps around the radius and attaches at both ends to the coronoid process of the ulna.
Management Considerations

The bony congruencies of the elbow contribute significantly to the stability of the joint. This contribution is especially strong at less than 20 degrees of extension and greater than 120 degrees of flexion. Between near-flexion and near-extension, ligaments provide the stability of the joint. Given the high valgus stresses of the throwing motion the ulnar collateral ligament is a particularly important source of stability. The anterior band of the ulnar collateral ligament functions primarily between full extension and 85 degrees of flexion and is the most important stabilizer of the ulnar collateral ligament complex in the throwing athlete. The posterior band of the ulnar collateral ligament becomes taut at 55 degrees of flexion and remains so through full flexion. The transverse ligament, which originates and inserts on the same bone, the ulna, provides no support during movement.

Dynamic stability is provided during the throwing motion by the flexor–pronator group of muscles (see Figure 2–5). The flexor carpi ulnaris, lying over the ulnar collateral ligament, is the primary dynamic stabilizer. An additional contribution is provided by the flexor digitorum superficialis, particularly when the elbow is in extension.

CLINICAL PROBLEM: ULNAR NEUROPATHY

Pertinent Anatomy

The ulnar nerve is subject to possible entrapment at the following anatomic sites:

- Arcade of Struthers, located 8 cm above the elbow; typically only a site of compression after ulnar nerve transposition.
- Medial head of the triceps.
- Medial intermuscular septum.
- Retrocondylar groove.
- Cubital tunnel: The flexor carpi ulnaris aponeurosis running between the two heads of the flexor carpi ulnaris in an arcade-like fashion from the medial epicondyle of the humerus to the olecranon process of the ulna. The floor of the cubital tunnel is formed by the medial collateral ligament of the elbow.
- Anconeus epitrochlearis, a rare anomalous muscle that crosses ulnar nerve posterior to the cubital tunnel.
• Arcuate ligament or aponeurosis of the flexor carpi ulnaris, which connects the two heads of the flexor carpi ulnaris.
• Deep flexor-pronator aponeurosis, located distal to the medial epicondyle.

Management Considerations

The two most common sites of ulnar nerve compression near the elbow are the retrocondylar groove and the true cubital tunnel between the two heads of the flexor carpi ulnaris. Although the ulnar nerve innervates the flexor carpi ulnaris and the medial half of the flexor digitorum profundus distal to the elbow, the above mentioned ulnar neuropathies at the elbow tend to spare these muscle groups. Electrodiagnostic evidence of denervation in these muscles may or may not be seen. Signs of denervation are consistently seen in the more distally innervated intrinsic muscles of the hand, including the abductor digiti minimi, all of the dorsal and palmar inter-ossei, the lumbricals to the fourth and fifth digits as well as the adductor pollicis.

Clinical Problem: Carpal Tunnel Syndrome

Pertinent Anatomy

There are eight carpal bones that sit just distal to the distal radioulnar joint and dorsal to the flexor retinaculum or palmar aponeurosis. The carpal bones, starting from the radial side of the proximal wrist, are the scaphoid, lunate, triquetrum, and pisiform. The second more distal row of carpal bones from radial to ulnar are the trapezium, trapezoid, capitate, and hamate.

Pertinent surface anatomy in this region includes the anatomic snuff box on the radial aspect of the wrist, and the pisiform and hamate on the ulnar aspect. The floor of the anatomic snuff box comprises the prominences of the scaphoid and trapezium palpated laterally along the palmar surface of the wrist. The borders are the radial styloid proximally and the first metacarpal distally.

Management Considerations

The four tendons of the flexor digitorum profundus, four tendons of the flexor digitorum superficialis, flexor pollicis longus, and the median nerve run through the carpal tunnel. The muscles are located proximally in the forearm and thus
receive innervation from the median nerve or anterior inter-osseous nerve proximal to the carpal tunnel, and are unaffected in carpal tunnel syndrome.

**CLINICAL PROBLEM: WRIST PAIN**

**Pertinent Anatomy**

On the dorsal side of the wrist (Figure 2–6), tendons can grouped into six compartments, arranged from the most radial or lateral aspect of the wrist to the medial or ulnar aspect of the wrist.
Figure 2–6 Extensor forearm. (Reproduced with permission from Morton DA: The Big Picture: Gross Anatomy. McGraw-Hill, 2011.)
Management Considerations

Lister’s tubercle, now known as the dorsal tubercle of the radius, can be used to help localize the compartments on palpation or radiographic imaging as the first two compartments are lateral to Lister’s tubercle and the next four are medial. The first compartment is composed of the tendons of the abductor pollicis longus and the extensor pollicis brevis. The second compartment is composed of the tendons of the extensor carpi radialis longus and the extensor carpi radialis brevis. The third compartment, located just lateral to Lister’s tubercle, is composed of the extensor pollicis longus tendon. The fourth and largest of the compartments is composed of the tendon of the extensor indicis proprius followed by the tendons of the extensor digitorum for digits 2 through 5. All four of these compartments are located just dorsal to the distal radius. The fifth compartment contains the tendon of the extensor digiti minimi and resides just dorsal to the radioulnar joint. The sixth and final compartment sits just dorsal to the ulna and houses the tendon of the extensor carpi ulnaris. All of the muscles in the extensor compartments are innervated by the radial nerve or the posterior interosseous branch of the radial nerve in the forearm.

CLINICAL PROBLEM: Ulnar Collateral Ligament Rupture

Pertinent Anatomy

The metacarpophalangeal joint of the thumb is protected from valgus stress, particular when the thumb is abducted, by the ulnar collateral ligament of the thumb. The ligament originates from the thumb metacarpal and its insertion on the proximal phalanx.

Management Considerations

Acute excessive abduction stress at the metacarpophalangeal joint of the thumb can produce an acute rupture of the ulnar collateral ligament of the thumb. This is known as skier’s thumb and typically occurs in a fall against a ski poll or the ground. A chronic ulnar collateral ligament injury, called gamekeeper’s thumb, can result from repetitive lateral stress applied to an abducted metacarpophalangeal joint. Chronic laxity of the ulnar collateral ligament can result in weakness of the pincer grasp and degenerative joint disease.
THE LOWER LIMB

CLINICAL PROBLEM: FEMOROACETABULAR IMPINGEMENT

Pertinent Anatomy

The hemipelvis is a composite of three bones, the ilium, ischium, and pubis, parts of which join together to form the acetabulum. The acetabulum contains a cartilaginous ring called the acetabular labrum, which increases the articular surface area. The hip joint is also supported by three ligaments: the ischiofemoral ligament, the pubofemoral ligament, and the most powerful of the three, the iliofemoral ligament, also referred to as the Y-ligament of Bigelow. Posteriorly, the pelvis articulates with the bony sacrum, a butterfly-shaped bone at the sacroiliac joints. Anteriorly the two halves of the pelvis meet at the pubic symphysis.

The ilium is the largest portion of the bony pelvis and serves as the origin for the gluteus muscles, the iliacus, and rectus femoris. The ischium houses the origins of the ham-string muscles as well as the adductor magnus. The pubic rami serve as the origin for the adductor muscles.
The iliofemoral ligament not only holds the femoral head tightly in its place in the acetabulum, but it is most crucial for preventing the trunk from falling backward as the pelvis tilts forward in a standing posture. It maintains this stability without any active muscle contraction or energy expenditure.

▶ **Management Considerations**

Femoral–acetabular impingement is a clinical entity representing the premature collision between the femur and acetabulum that diminishes optimal range of motion and can lead to injury. In a cam-type impingement, the femoral head takes on an aspherical shape that causes its lateral portion to collide with the acetabulum prior to complete range of motion. In a pincer-type impingement, which is an acetabular problem, the acetabulum provides excessive coverage around the spherical portion of the femoral head, thereby restricting range of motion. Most patients have a mix of these two types of impingement, and femoral—acetabular impingement is thought by some authorities to predispose a patient to hip osteoarthritis. Osteoarthritis more commonly results from wear-and-tear degeneration at the joint capsule of the hip, resulting in joint space narrowing, osteophyte formation, and subchondral cysts or sclerosis. This often also results in decreased range of motion in the hip, affecting first internal rotation and then external rotation of the hip.

**CLINICAL PROBLEM: LATERAL HIP PAIN & BUTTOCK PAIN**

▶ **Pertinent Anatomy**

The greater trochanter of the femur is the insertion site of the gluteus minimus and medius tendons. The following muscles can also function to rotate the hip (Figure 2–7):

- External rotators: piriformis, obturators, gemelli
- Internal rotators: gluteus medius and minimus, tensor fasciae latae
- Gluteus maximus: a major hip extensor that also extends the knee in closed chain with foot planted on the ground
The piriformis is a pear-shaped muscle, deep and parallel to the gluteus medius, that originates on the anterior sacrum and inserts on the greater trochanter. The piriformis functions to abduct and externally rotate the hip from 0 to 90 degrees of flexion, and then functions to adduct and internally rotate the
hip in greater than 90 degrees of flexion.

Management Considerations

These attachments permit these muscles to perform hip abduction but, more importantly, the force of hip abduction prevents the pelvis and lower limb from dropping down during the contralateral leg’s swing phase of gait. Structurally, the sciatic nerve runs deep to the piriformis, and in some cases the common fibular portion of the nerve may pierce the piriformis and then rejoin the tibial portion of the nerve to descend into the leg.

CLINICAL PROBLEM: PATELLOFEMORAL SYNDROME

Pertinent Anatomy

The anterior thigh muscles (Figure 2–8) insert on the patella and then, via the patellar tendon, onto the tibial tuberosity, functioning to extend the knee. The rectus femoris, vastus lateralis, vastus intermedius, and vastus medialis form the quadriceps muscle. The vastus group originates on the femur, whereas the rectus femoris originates on the anterior inferior iliac spine just above the acetabulum, permitting it to function as both a hip flexor and a knee extensor.
The medial muscle group of the thigh, the adductor group, comprises the adductor longus, adductor brevis, pectineus, gracilis, and adductor magnus muscles. All of the adductors originate on the pubis and insert on the femur, permitting adduction of the femur and flexion of the hip. The adductor magnus has an additional origin on the ischial tuberosity, permitting it to extend the hip. Depending on their specific origin and insertion, these muscles may also function to rotate the femur internally or externally.

Management Considerations

Multiple mechanical and muscular factors contribute to the dynamic stability of the knee joint. The angle between the rectus femoris and patellar tendon is called the Q angle, and when this measurement exceeds normal ranges the patient may be predisposed to patellar problems. A normal range for the Q angle measured in a standing position is between 18 and 22 degrees. Women tend to have Q angles at the higher end of this range.

CLINICAL PROBLEM: INTERNAL KNEE INJURY

Pertinent Anatomy: Ligaments

The knee joint (Figure 2–9) relies on ligamentous structures and a joint capsule to provide stability. The medial collateral ligament originates from the medial condyle of the femur and inserts onto the medial condyle of the tibia. The lateral collateral ligament originates from the lateral condyle of the tibia and inserts onto the fibular head. The anterior cruciate ligament originates from the medial wall of the lateral condyle of the femur. Two bundles of fibers run in an anteromedial direction to insert onto the tibia. The posterior cruciate ligament originates from the medial femoral condyle in the intercondylar notch and, in contrast to the anterior cruciate ligament, runs in a posterior direction to insert onto the tibial plateau.
Management Considerations

The medial and lateral collateral ligaments resist, respectively, valgus and varus stress at the knee. The ligaments are most commonly injured when the foot is planted and the lower limb is impacted from the lateral direction (in medial collateral ligament injuries) or the medial direction (in lateral collateral ligament injuries). Both ligaments resist extremes of axial rotation of the knee and can be
injured in that fashion as well. The anterior cruciate ligament primarily resists anterior translation of the tibia with respect to the femur and can be injured, sometimes with the medial collateral ligament, by a valgus force at the knee with the foot planted. The anterior cruciate ligament can also be also injured in “noncontact” situations when a strong quadriceps contraction pulls the tibia in an anterior direction just after the foot is planted to decelerate an individual’s forward motion. The posterior cruciate ligament, in contrast to the anterior cruciate ligament, resists posterior translation of the tibia with respect to the femur and can be injured in extremes of knee flexion such as a fall onto an already flexed knee.

**Pertinent Anatomy: Menisci**

The medial and lateral menisci are C-shaped fibrocartilaginous structures that rest above the articular surfaces of the tibia, providing a seat for the convex femoral condyles. They are anchored to the intercondylar region of the tibia at either end of the “C” at their anterior and posterior horns, and are connected anteriorly by a transverse ligament. The menisci function to reduce the compressive stress across the tibiofemoral joint and pivot freely during movement.

**Management Considerations**

The joint capsule of the knee is connected to both the medical and lateral menisci; however, the capsular attachment to the lateral meniscus is interrupted by the popliteus tendon. The medial meniscus may be more likely to be injured owing to reduced mobility of the structure relative to the lateral meniscus. Both structures are commonly injured when a force, valgus or varus, affects a flexed knee. Meniscal injury can cause pain but also locking and catching during knee flexion and extension. This can occur when torn meniscal tissue is displaced into more central regions of the joint. The displaced tissue impairs proper motion of the knee, leading to a sensation of locking and catching. Tears of this type are be referred to as “bucket handle” tears. In an older population, without history of acute injury, meniscal injuries are commonly degenerative in nature and can be seen on imaging in conjunction with other signs of osteoarthritis.

**CLINICAL PROBLEM: HAMSTRING INJURY**
Pertinent Anatomy

The posterior thigh muscles, collectively known as the hamstrings, share a common origin on the ischial tuberosity, except for the short head of the biceps femoris. The four hamstring muscles are the semimembranosus, semitendinosus, long head of the biceps femoris, and short head of the biceps femoris. The first three of these muscles extend the hip, and all of these muscles function to flex the knee in an open chain and extend the knee in a closed chain. The medial hamstring group, semimembranosus and semitendinosus, insert on the tibia.

Management Considerations

Clearly the hamstring muscles are powerful knee flexors, as well as hip extensors, and are important during running and gait. Injury to this muscle group can cause significant activity limitation. In swing phase the hamstrings act eccentrically to slow knee extension, and this is possibly the most common time of injury. Hamstring strains occur most often in the biceps femoris muscle and at the myotendinous junction.

CLINICAL PROBLEM: ANKLE SPRAIN

Pertinent Anatomy

The ankle (Figure 2–10) comprises three joints, the talocrural, inferior tibiofibular, and subtalar joints, and allows for movement in dorsiflexion, plantar flexion, inversion, and eversion. The inferior tibiofibular joint encompasses the distal aspects of the tibia and fibula. The inferior tibiofibular ligament supports the tibiofibular joint and allows for a modest amount of rotational movement. This ligament, or syndesmosis, is injured in so-called high ankle sprains. The subtalar joint is the articulation between the talus and calcaneus and allows the foot and ankle to accommodate to uneven terrain. The sinus tarsi divides the subtalar joint into anterior and posterior sections. The most prominent articulation at the ankle is the talocrural joint, which allows for the most noticeable movements at the ankle and those movements with the greatest range of motion, plantar flexion and dorsiflexion. The talocrural joint is formed by the superior surface of the talus and the inferior surface of the tibia.
The ankle joint is more stable in dorsiflexion; consequently, the ankle is most commonly injured in plantar flexion. When a force acts on the preceding joints in quantities exceeding the strength of the supporting structures, injury can occur. The term ankle sprain refers to ligamentous injury around the ankle. The lateral ankle ligaments, the most commonly injured structures, are the anterior talofibular ligament (ATFL), the calcaneofibular ligament (CFL), and the posterior talofibular ligament (PTFL). These ligaments originate from the distal
fibula and insert onto the anterior talus, calcaneus, and posterior talus, respectively. It is easy to imagine, given the insertion of the ATFL on the anterior talus, that plantar flexion will stress the ATFL causing it to become taut. In an inversion injury the distance between the talus and the fibula becomes even greater and the ATFL, therefore, is at increased risk for injury. The ATFL is the first ankle ligament to become injured and isolated injuries to the CFL and PTFL are rare. The medial ligament of the ankle, the deltoid ligament, is much less commonly injured given its relative strength and size. The deltoid ligament originates from the medial malleolus of the distal tibia and inserts anteriorly on the navicular and talus, inferiorly on the calcaneus, and posteriorly on the talus. As can be understood from the preceding description, and observed from Figure 2–10, the ankle ligaments both originate from the long bones of the distal lower leg and insert in an expected fashion based on the bony anatomy of the ankle.

Vascular Diseases

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Vascular disease includes any condition that affects the circulatory system, encompassing diseases of arteries, veins, and lymph vessels as well as blood disorders that affect circulation. It describes a broad group of clinical conditions ranging from the chronic to the acute and life threatening. The increasing prevalence of peripheral artery disease (PAD) and carotid artery disease among Americans may be related to a rise in the prevalence of diabetes mellitus, just as the increased incidence of varicose veins is linked to rising rates of obesity.

Vascular disease is a nearly pandemic condition that has the potential to cause loss of limb or even loss of life. It manifests as insufficient tissue perfusion that may be acutely compounded by either emboli or thrombi to an existing atherosclerotic condition. Many people live daily with vascular disease; however, in scenarios such as acute limb ischemia, this pandemic disease can be life threatening, requiring emergency intervention to minimize morbidity and mortality.

DIABETIC WOUNDS

ESSENTIALS OF DIAGNOSIS

- The lifetime risk of developing lower extremity ulceration in patients with diabetes mellitus approaches 25%.
Comprehensive annual evaluation of the diabetic foot is recommended.
Visual inspection should be performed at every routine visit to the physician.
Although multifactorial in origin, diabetic ulceration is linked primarily to poor blood glucose control.

General Considerations

Diabetic wounds are associated with substantial morbidity and mortality. Diabetes mellitus is a particularly important risk factor in the development of chronic wounds because it is associated with neuropathy, vasculopathy, and immunopathy. Chronic ulceration affects the lower extremities in 1.3% of adults in the United States. However, among diabetic patients the lifetime risk of developing lower extremity ulceration approaches 25%. Two thirds of nontraumatic amputations performed in the United States are secondary to primary diabetic foot ulcers and their complications.

These statistics illustrate the importance of evaluation and prevention of diabetic-related skin infections and the necessity of prompt medical–surgical treatment when infections develop. In 2012, the Infectious Disease Society of America updated its guidelines for the diagnosis and management of diabetic foot infections. The current American Diabetes Association guidelines, which largely agree with those of other organizations, recommend comprehensive annual evaluation of the diabetic foot. This evaluation should include inspection of the foot for the presence of erythema, warmth, and callous, bony, or joint-mobility abnormalities, as well as skin integrity, with time taken to fully evaluate between the toes and under pressure-sensitive metatarsal heads. As part of this evaluation, patients should be tested for loss of protective sensation using tactile, vibratory, and reflex testing, and screened for PAD by asking about claudication symptoms, assessing the pedal pulses, and, in those older than 50 years of age or in any patient having other risk factors of PAD, by assessing the ankle–brachial index (ABI). A visual inspection should also be performed at every routine visit to the physician.

When a patient has a diabetic wound the clinical evaluation should determine the extent and severity of infection, identify the underlying factors that predispose to and promote infection, and assess the microbial cause. The clinical history should note details related to the injury (cause, duration, associated symptoms, and prior treatments, if any). Clinical examination should note the
location of the lesion, the presence and extent of infection (local or systemic),
the extent of the wound itself (eg, involving superficial skin only or infecting
deeper subcutaneous tissues, muscles, bone), and whether bone is visible to the
eye or palpable upon probing. Clinical examination should also include a
neurologic and vascular evaluation. Laboratory evaluation should include tests to
identify systemic or metabolic inflammation (eg, complete blood count, basic
metabolic panel, erythrocyte sedimentation rate [ESR], C-reactive protein
[CRP]), as well as a glucose level, to assess glycemic control. Procalcitonin may
be a helpful inflammatory marker, but its utility has yet to be proven.

Pathogenesis

At the biochemical and cellular level, diabetes mellitus involves a complex array
of metabolic and vascular factors that shift the balance between nerve fiber
repair and nerve fiber damage toward the latter process. The result is a
manifestation of polyneuropathy that preferentially affects nerves of the distal
extremity. In patients with type 2 diabetes, the vascular factors, which cause
ischemia secondary to thickening of the endothelium, are particularly important.

The polyneuropathy involves sensory, motor, and autonomic nerves. Sensory
neuropathy diminishes the protective perception of pain that notifies the
individual when tissue injury has occurred. Motor nerves to the intrinsic muscles
of the foot are affected in approximately half of all diabetic patients, resulting in
claw deformities that transfer pressure to the plantar metatarsal heads. The
increased tissue pressure may lead to skin erosion and ulceration, and, in the
case of the insensate individual, may go unnoticed. Autonomic neuropathy
causes the skin to become dry and susceptible to skin fissures, tearing, and
infection as a result of loss of sweat and oil gland function. Additionally,
diabetes is frequently associated with severe PAD, which affects the smaller
distal arteries, causing atherosclerotic changes. The combination of PAD and
diabetes contributes to higher rates of nonhealing ulcers and limb loss in diabetic
patients compared with nondiabetic patients.

More than 100 known cytologic factors contribute to impaired wound
healing in patients with diabetes. These include decreased or impaired growth
factor production, angiogenic response, macrophage function, collagen
accumulation, epidermal barrier function, quantity of granulation tissue,
keratinocyte and fibroblast migration and proliferation, number of epidermal
nerves, bone healing, and abnormal balance between the accumulation of
extracellular matrix components and their remodeling by matrix
Clinical Findings

Diabetic ulcers (Figure 3–1) can develop as a result of trauma, skin cracks, fissures, or other defects in the skin of the foot or the paronychia. Infection can be localized to the superficial skin at the site of a preexisting lesion or involve the skin or deeper structures beyond the local site, potentially spreading to joints, bones, or the systemic circulation.

Figure 3–1 Diabetic ulcer (Reproduced with permission from Apelqvist J, Bakker K, van Houtum WH, Nабuurs-Franssen MH and Schaper NC. International consensus and practical guidelines on the management and the prevention of the diabetic foot. Diabetes/Metabolism Research and Reviews. 2000;16:S84-92.)

Patients with diabetic foot infections often have universal symptoms of infection (fever, chills, hypotension, and tachycardia) or inflammation (erythema, warmth, swelling, and tenderness), or pustulant material within an ulcer or sinus tract. However, these symptoms are not always noted, as diabetic patients with sensory neuropathy may not experience tenderness, and patients with comorbid PAD may not have excess warmth secondary to ischemia. In such cases, infections may progress to include deeper or systemic systems before a
patient seeks medical attention. Other nonspecific symptoms, including nonpurulent drainage, friable or discolored granulation tissue, and undermining of wound edges, may also be present.

Osteomyelitis can occur in a diabetic foot wound with or without evidence of soft tissue infection. Clinical features associated with the presence of underlying osteomyelitis include grossly visible bone or the ability to probe to bone, ulcer size larger than 2 cm², ulcer duration greater than 2 weeks, and ESR greater than 70 mm/h.

Grading of neuropathy and wounds is equally important in clinical practice. The International Working Group on the Diabetic Foot developed a neuropathy classification system to predict foot ulceration that classifies patients into risk groups (Table 3–1). A commonly used classification system for grading diabetic wounds is shown in Table 3–2.

### Table 3–1 International Working Group classification of the diabetic foot.

<table>
<thead>
<tr>
<th>Group</th>
<th>Clinical Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No evidence of neuropathy</td>
</tr>
<tr>
<td>1</td>
<td>Neuropathy present by no evidence of foot deformity or peripheral vascular disease</td>
</tr>
<tr>
<td>2</td>
<td>Neuropathy with evidence of deformity of foot or peripheral vascular disease</td>
</tr>
<tr>
<td>3</td>
<td>History of foot ulceration or lower extremity amputation</td>
</tr>
</tbody>
</table>

### Table 3–2 Wagner classification for grading diabetic wounds.
Differential Diagnosis

In most cases, the history of the infection or wound is sufficient to isolate the source to diabetes. However, other infections or processes can present with generalized inflammatory changes in the skin of the distal extremities and therefore mimic a diabetic infection. Some of these include trauma, crystal-associated arthritis, Charcot arthropathy, fracture, thrombosis, and venous stasis. Other skin ulcerations that manifest similarly to a diabetic ulceration include, but are not limited to, chronic ulcers from venous disease, pressure ulcers, arterial ischemic skin alterations, and localized burn trauma.

Complications

Many complications may occur when diabetic infections ulcerate. The most topically important are all manner of infections that if unnoticed, undiagnosed, or untreated can progress to osteomyelitis, potentially requiring aggressive therapy, prolonged hospitalization, vascular intervention, or amputation. Along with the multitude of other impaired functions linked to diabetic wounds, the simple act of ulceration causes the breakdown of the body’s natural barrier mechanism—the skin—allowing bacteria ready access. Most diabetic ulcers, if chronic, are colonized and not necessarily infected. When ulcers do become infected the wounds are therefore polymicrobial.

Superficial diabetic ulcers in most individuals are likely caused by aerobic
gram-positive cocci (Staphylococcus aureus, Streptococcus agalactiae, Streptococcus pyogenes, and coagulase-negative staphylococci). Ulcers that are deep, chronically infected, or previously treated with antibiotics are more likely to be polymicrobial. Such wounds may involve the previously mentioned species as well as enterococci, Enterobacteriaceae, Pseudomonas aeruginosa, and anaerobes. Wounds with extensive local inflammation, necrosis, malodorous drainage, or gangrene along with signs of systemic toxicity should be presumed to have anaerobic organisms (Clostridium species, Bacteroides species, and anaerobic streptococci), in addition to the aforementioned organisms.

Considering the predilection of specific organisms to infect diabetic ulcers, further information should be obtained relating to the patient’s the risk of infection with specific organisms. Methicillin-resistant S aureus (MRSA) is a common pathogen, particularly in those who have had previous MRSA infections or known colonization. Other risk factors for MRSA infections are prior antibiotic use, previous hospitalization, and residence in a long-term care facility. P aeruginosa is a prevalent organism in diabetic wounds of patients from warm climates. Macerated ulcers, foot soaking, and other exposure to water or moist environments also increases the risk of P aeruginosa involvement. In the absence of these conditions, it is not a commonly isolated pathogen.

Resistant enteric gram-negative rods (extended spectrum β-lactamase [ESBL] organisms) are increasing in prevalence in diabetic wound infections. Although these bacteria are still uncommon, there is greater risk in patients who have had prolonged hospital stays, prolonged catheterization, prior antibiotic use, or residence in a long-term care facility.

### Treatment

#### A. Preventive Care

The most important tenet of diabetic wound treatment occurs before an individual develops a wound. Preventive foot care should be routinely discussed with patients who are at high risk for wound development, especially those with existing neuropathy. In conjunction with screening, nutritional support, and medical preventative guidance to control glucose and vascular risk factors, the high-risk patient should be instructed to avoid smoking, walking barefoot, using heating pads, and stepping into the bath without first checking water temperature. Additional guidelines include trimming toenails to the shape of the toe, with the removal of sharp edges; daily foot inspection (by the patient or
someone else, if vision or ability is an issue); ensuring proper shoe fit and sock use; and daily bathing of feet in lukewarm water with mild soap, and then blotting dry with a soft towel.

**B. Measures to Promote Wound Healing**

If prevention fails and medical therapy is required for a diabetic wound, multiple treatment strategies can be initiated, all of which have been shown to have positive effects on the healing of diabetic wounds. These include antibiotic treatments, various methods of debridement, assorted topical modalities, wound dressing materials, wound closure techniques, mechanical offloading, and adjunctive therapies.

1. **Antibiotic therapy**—Antibiotics can be used to treat wounds that are clinically infected. Options range from oral to parenteral coverage, depending on the severity of the wound and the likely bacteria present. The choice of agent should always ensure coverage of species such as *S. aureus* and streptococci (group A and B). Cultures should be obtained to confirm pathogens and, if necessary, alter the antibiotic course. The preferred clinical specimens for reliable culture include aspirate from an abscess or curettage from the ulcer base. Superficial swabs are neither reliable nor sufficient for prediction.

2. **Debridement**—Chronic wounds characteristically have decreased angiogenesis and accumulate devitalized tissue, hyperkeratotic tissue, exudate, and bacterial overgrowth on the surface of the wound, creating a biofilm. Debridement can aide in restoring an optimal wound healing environment and can be done in various ways.

   Irrigation with warm, isotonic saline can decrease bacterial load and remove loose material. It should be done with low pressure (< 15 lb psi) as higher pressure could cause local tissue damage by dissecting loose connective tissue and increasing edema.

   Sharp, excisional surgical debridement uses a scalpel to remove devitalized tissue and accumulated debris. It functions to decrease bacterial load and stimulates wound epithelialization. Surgical debridement is the most appropriate choice for removing large areas of necrotic tissue and is indicated when there is excessive bone or joint involvement, gangrenous tissue, or any evidence of sepsis.

   Enzymatic debridement uses topical proteolytic enzymes such as collagenase, fibrinolysin, and deoxyribonuclease that work synergistically with endogenous enzymes to debride the wound. Autolytic debridement is
accomplished by covering a wound so that the endogenous proteolytic enzymes digest the necrotic tissue while the cover prevents external infection. This method should not be used if the wound is infected initially as the topical enzymes will not function as a disinfectant.

Biologic debridement is an alternative method of debridement that utilizes the sterilized larvae of the Australian sheep blowfly (Lucilia cuprina) or the green bottle fly (Lucilia sericata). The larvae (maggots) produce enzymes that degrade necrotic tissue but do not harm healthy tissue. They are kept from migrating by a mesh cover and can remain in place for 48–72 hours before the dressing is changed. The largest disadvantage to this method is the negative perceptions about its use by both patients and staff.

3. Topical therapies—Growth factors and antimicrobials are other alternatives that have been used in diabetic wound care. Platelet-derived growth factor promotes cellular proliferation and angiogenesis. It is indicated for noninfected diabetic ulcers that extend into the subcutaneous tissue and have an adequate vascular supply. Granulocyte–macrophage colony-stimulating factor (GM-CSF) has been used in various chronic wounds to promote healing. Cadexomer iodine is an antimicrobial that reduces bacterial load because it is bactericidal to all gram-negative and gram-positive bacteria. Silver sulfadiazine is a topical antiseptic cream that decreases the incidence of sepsis in cutaneous wounds. It also serves as an antimicrobial, as silver is toxic to bacteria.

4. Wound dressings—Appropriate wound dressings promote ulcer healing by facilitating endogenous mechanisms, absorbing excess exudate, and protecting the wound from the external environment. Wounds that are too wet or too dry heal more slowly; excess fluid causes wound maceration while excess desiccation slows cellular migration to the site. Additionally, as wounds heal the requirement of the wound site changes, so the dressing type should change along with it.

Dressings are classified into three categories—open, semi-open, or semi-occlusive—according to their water-retaining abilities, because maintaining a moist environment is the primary goal in dressing therapy. Open dressings usually consist of gauze, which should never be applied dry. Gauze is inexpensive but requires frequent dressing changes. Semi-open dressings include fine mesh gauze impregnated with some form of ointment; Xeroform and Adaptic dressings are examples. Semi-open dressings are inexpensive but fail to provide good control of exudate or maintain a moisture-rich environment. Semi-occlusive dressings are available with a wide variety of occlusive properties,
absorptive capacities, conformability, and bacteriostatic activity. Examples include films, foams, alginates, hydrocolloids, and hydrogels.

An absorptive dressing (eg, alginate, foam, or hydrofiber) should be used for ulcers with heavy exudate. Alginates are derived from brown seaweed and form a gel on contact. Foams provide thermal insulation, high absorbency, and a moist environment; they can be easily cut to shape, do not shed fibers, and are used for exudative wounds with sloughing skin. Desiccated ulcers lack wound fluids, which stimulate epithelialization; thus, a dressing that can provide a moist environment without causing maceration is needed. Saline-moistened gauze, transparent films, hydrocolloids, and hydrogels are commonly used dressings that meet this need. Saline-moistened gauze provides the correct environment but requires frequent dressing changes as it quickly dries out. Additionally, care is needed when changing the dressing to avoid mechanically debriding healthy tissue.

C. Adjuvant Therapies

A variety of adjuvant therapies may be employed to aid diabetic wound healing, including negative pressure wound therapy, hyperbaric oxygen therapy, and electrical stimulation.

1. **Negative pressure wound therapy**—This therapy, also known as vacuum-assisted closure therapy, relies on subatmospheric pressurized closure of a wound. It has been shown to increase wound perfusion, reduce edema, and reduce the local bacterial burden while increasing the formation of granulation tissue.

2. **Hyperbaric oxygen therapy**—Hyperbaric oxygen increases mobilization of endothelial progenitor cells, which have been shown to aid healing by initiating angiogenesis in a hypoxic wound. The technique has shown utility in preventing amputation and improving wound healing. However, the therapy cannot be targeted to the wound site, and systemic exposure has been associated with serious adverse events, including seizures and pneumothorax.

3. **Electrical stimulation**—Direct electrical current applied to a wound promotes migration and proliferation of fibroblasts. Electrical stimulation has been shown to enhance healing during the initial stages of wound closure.

D. Pressure Modulation and Mechanical Offloading

Pressure modulation is a key intervention in the treatment of diabetic ulcerations,
especially in the lower extremities. Thus, the concept of mechanical offloading is of pivotal importance when considering therapeutic options for the diabetic foot.

1. **Biomechanical considerations**—The biomechanical causes of foot ulceration are pressures that are applied to the foot during stance phase. The ground reaction forces generated in response to weight bearing contain vertical, anteroposterior, and mediolateral components, with the vertical aspect much greater than the other two. (For additional discussion of this topic, see Chapter 4.) The vertical force created during a fast walk phase peaks at approximately 1.5 times body weight. While running, this force increases to approximately 5 times body weight. Vertical force damages tissue by compressing and deforming it. The anteroposterior and mediolateral forces jointly create a shear that damages healthy tissue by stretching it. It is the combination of focal pressures and repetitive stress applied to the plantar aspects of the diabetic foot that results in foot wounds.

2. **Pressure disbursement methods**—Pressure disbursement is most successful when the pressure forces are spread over a wide area or, as possible, eliminated. Common offloading methods include bed rest, wheelchairs, crutches, and the use of orthotics. Although bed rest, wheelchairs, and crutches are commonly prescribed they may precipitate other problems, hurting more than they help. They require the patient to limit full use of the lower extremities, which can lead to muscle loss, contraction, decreased endurance, and other immobility-related problems. They also shift pressure to other body areas, which can cause additional pressure-related injuries. Crutches exert pressure under the arms and place greater pressures on the contralateral high-risk diabetic limb; use of beds and wheelchairs increases pressure on the coccyx and ischial spines. Often patients do not have the endurance or strength to use crutches, or their homes cannot accommodate the bulkiness of a wheelchair. For all these reasons, correct use and compliance with these options has been shown to be quite poor. (These and other issues relating to use of specific assistive devices are discussed in more detail in Chapter 41.)

3. **Orthotic options for the diabetic foot**—Prescribed orthotics to the lower limb have shown much better results in wound healing; they include half-shoes, healing sandals, bespoke (custom-made) therapeutic shoes, removable cast walkers (RCWs), total contact casts (TCCs), Scotchcast boots, and instant total contact casts (iTCCs).

   Half-shoes were developed to reduce postoperative pressure on the forefoot.
The shoe consists of a wedged sole that ends just proximal to the metatarsal heads, eliminating propulsive gait and decreasing ground forces applied on the forefoot. They are inexpensive and easy to apply.

Healing sandals are specially designed to limit plantar progression of the metatarsal heads during propulsive gait by limiting dorsiflexion of the metatarsophalangeal joints through the application of a rigid rocker to the sole. They are lightweight, stable, and reusable. However, they are expensive and the rocker sole requires a significant amount of time and experience to produce.

Bespoke therapeutic shoes are a commonly prescribed customized shoe built with measurements taken from the patient. Although they have not shown utility in helping wounds heal, they are very effective in preventing ulcerations in high-risk feet. Because they are custom made, they can be quite expensive and patients may not find them to be aesthetically pleasing.

RCWs are castlike devices that are removable to allow for self-inspection of the wound and application of topical therapies that require frequent administration. RCWs are indicated for infected ulcers as well as superficial tissue infections. They limit propulsion by keeping the ankle at 90 degrees, thereby helping to decrease forefoot plantar pressure. Patients report better ease in sleeping and bathing as they can take the devices off during these activities; however, they can also remove the device inappropriately. Removability is the best feature of the RCW; it is also paradoxically its worst. The ability to remove the RCW eliminates “forced compliance” in use of the device, which makes TCC the gold standard in achieving pressure distribution.

TCC systems employ a plaster-of-Paris, well-molded, minimally padded cast that maintains contact with the entire plantar aspect of the foot and the lower leg. Intimate fit to the plantar surface of the foot increases the weight-bearing surface area. TCCs are indicated in the treatment of non-infected and nonischemic plantar diabetic wounds, with reported healing rates that range from 72% to 100% over a 5- to 7-week course. They have been shown to reduce pressure at the site of ulceration by 84–92%, reduce inflammatory and reactive components that detract from the reparative process, and reduce or control edema that impedes healing. Because the cast is a solid entity, it prevents exposure to foreign objects and pathogens. Most importantly, however, it ensures patient compliance because the cast is immobile and nonremovable.

The TCC system is not without disadvantages, some of which may preclude prescription of this modality. Application of the cast is time-consuming and requires a trained cast technician. Improper application—the most significant detractor—can cause skin irritation and, in extreme cases, new ulceration.
Because the TCC (unlike the RCW) cannot be removed, the wound cannot be assessed frequently. TCCs are therefore contraindicated in patients with soft tissue infections or osteomyelitis. Patients have also reported difficulty sleeping and bathing because of the precarious immobility of the 90-degree ankle, and the necessity of keeping the cast dry.

Scotchcast boots are an alternative to plaster-of-Paris boots created with much lighter, stronger, and more durable fiberglass polymers. The Scotchcast boot acts much like a TCC and shows similar healing rates, although it is more expensive to create. Small studies have reported fewer complications with the fiberglass material than the plaster-of-Paris TCCs.

Construction of an iTCC device involves simply taking an RCW as a base and wrapping it with bandage, Elastoplast, or casting tape. The ability to remove and reapply the device—but not without effort—addresses both the compliance issues associated with RCWs as well as the limitations of the standard TCC with regard to wound management. The first randomized controlled study comparing the iTCC with the TCC found no differences in healing rates or complications between the two and showed that the cost of materials and personnel was much lower with the iTCC. A parallel study comparing healing rates of the iTCC with the RCW showed that the iTCC was superior.

**Prognosis**

Diabetes is the underlying cause of most nontraumatic lower extremity amputations in developed countries, and infection is the precipitating event for nearly 90% of these amputations. Research has shown that with increasing infection severity there is a clear trend toward increased risk for amputation, increased risk for more proximal anatomic amputation, and increased need for lower-extremity-related hospitalizations. The same research revealed a similar trend toward increasing risk for experiencing other diabetic foot-related complications, such as neuropathy or vascular disease. This suggests that persons with noninfected or mild wounds are highly unlikely to require hospitalization, develop osteomyelitis, or undergo amputation, whereas those with moderate to severe wounds require much more diligent, thorough, and expansive treatment strategies that all too often result in amputation.

Armstrong DG, Lavery LA, Wu S, Boulton AJ: Evaluation of removable and irremovable cast walkers in the healing of diabetic foot wounds: A


PERIPHERAL ARTERY DISEASE
**General Considerations**

PAD describes the condition of impaired blood flow to the extremities, most commonly caused by atherosclerosis. Depending on the degree of arterial stenosis and collateral circulation, PAD can lead to varying clinical presentations. However, in a significant segment of the PAD population, possibly up to 75%, presentation is asymptomatic. PAD prevalence increases significantly with age and is more common in non-Hispanic blacks. Men and women develop PAD at similar rates, but it may be slightly more common in men. The most important risk factors that have been repeatedly associated with PAD are smoking, diabetes mellitus, hypertension, and hyperlipidemia. PAD is also more prevalent in patients with chronic renal insufficiency, hyperhomocysteinemia, hyperviscosity, and hypercoagulable states. A self-reported history of cardiovascular disease, including coronary artery disease, congestive heart failure, and cerebrovascular accidents, has been strongly correlated with PAD.

**Clinical Findings**

**A. Symptoms and Signs**

PAD is often asymptomatic, but when blood flow limitation becomes significant the first symptom is usually intermittent claudication. Intermittent claudication produces leg pain, numbness, or fatigue during exercise and subsides with rest. The symptoms can occur at different levels of the lower extremity, depending on
where the arterial stenosis is located. The most common site of PAD is the superficial femoral artery, leading to intermittent claudication of the calf muscles. PAD can also occur in the iliac, common femoral, and tibioperoneal arteries, causing symptoms in the buttocks, thighs, and feet, respectively.

Individuals with intermittent claudication have dysfunctional oxidative metabolism in the affected muscles. This dysfunction leads to skeletal muscle injury, with loss of muscle fibers and atrophy. As a result, these patients have a lower tolerance for physical activity than those without PAD and an associated decline in the ability to perform activities of daily living. Patients often develop collateral circulation or alterations in their gait pattern suggestive of disease stabilization, but thorough evaluation reveals symptoms that are usually slowly progressive, leading to a decline in pain-free walking distance. About 25% of patients with intermittent claudication progress to rest pain or critical limb ischemia, manifested by ulceration and gangrene. Patients with rest pain frequently describe hanging their legs over the edge of the bed or sleeping in a chair to increase distal perfusion and relieve symptoms. Fontaine first classified this progression from asymptomatic PAD to critical limb ischemia (Table 3–3).

### Table 3–3 Fontaine classification of peripheral arterial disease.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Clinical Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>IIa</td>
<td>Mild claudication</td>
</tr>
<tr>
<td>IIb</td>
<td>Moderate to severe claudication</td>
</tr>
<tr>
<td>III</td>
<td>Ischemic rest pain</td>
</tr>
<tr>
<td>IV</td>
<td>Ulceration or gangrene</td>
</tr>
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</table>

PAD is associated with a three- to six-fold increased risk of cardiovascular mortality. Patients with lower ABI values (indicating more severe PAD) and those with abnormally elevated ABI values (indicating arterial calcifications) have a higher risk of cardiovascular events. Therefore, physicians should pay close attention to any complaints of chest pain, angina, and related symptoms in these patients.

On physical examination, multiple PAD-related findings may be noted.
Auscultation of major arteries, including the aorta, both carotids, and both femorals, may reveal a bruit, but the absence of a bruit does not rule out stenosis. PAD may also cause decreased skin temperature, hair loss, color changes, and hypertrophied toenails. Peripheral pulse evaluation may reveal decreased or absent pulses that could signify arterial stenosis proximal to the point of palpation. Buerger’s test for PAD is positive if pallor develops after limb elevation with subsequent hyperemia after lowering the limb. In acute arterial occlusion, patients may present with the classic “5 Ps”—pain, pallor, pulselessness, paresthesia, and paralysis. These patients require immediate referral to a vascular specialist.

B. Diagnostic Studies

Patients with history or physical examination findings indicative of PAD should be assessed with noninvasive testing. Such testing most often involves measuring the ABI. The ABI is calculated by obtaining blood pressure readings at the dorsalis pedis and posterior tibial arteries and subsequently dividing the higher of those two pressures by the higher blood pressure reading obtained from the patient’s two arms. An ABI of less than 0.9 is typically considered hemodynamically significant and diagnostic of PAD. Patients with diabetes, chronic renal insufficiency, or other diseases that cause vascular calcifications may have a falsely elevated ABI secondary to noncompressible arteries; the ABI in these patients is often greater than 1.4. In such patients, the toe–brachial index can be measured in place of the ABI.

Other noninvasive diagnostic tests that can be ordered include pulse volume recordings, transcutaneous oximetry, arterial continuous wave Doppler, duplex ultrasound, magnetic resonance angiography (MRA), and computed tomographic angiography (CTA). Imaging studies such as duplex ultrasound, MRA, and CTA are usually reserved for patients with critical limb ischemia who will undergo revascularization. The gold standard diagnostic test for PAD is angiography, but this test is invasive and, thus, similarly saved for definitive cases of revascularization.

Differential Diagnosis

As previously described, the patient history usually provides the first important information in diagnosing PAD. Other conditions can present with similar complaints, including (but not limited to) chronic compartment syndrome, venous claudication, nerve root compression, spinal stenosis, and osteoarthritis.
Treatment

A. Lifestyle Modification

Cardiovascular risk factor modification is the mainstay of treatment for patients with PAD. This strategy involves smoking cessation, blood pressure control, blood glucose control in diabetic patients, weight loss, and reduction of low-density lipoprotein cholesterol. Additionally, antiplatelet drug therapy should be initiated in all patients with PAD and either symptoms or signs of cardiovascular disease.

Additional goals of PAD treatment are to decrease symptoms during walking, increase functional performance, and improve quality of life. A supervised exercise program has proven beneficial in achieving these goals, with a treadmill-training program shown to be more effective than strength training. The program should involve at least 3 days per week of exercise, beginning with sessions lasting 30 minutes and progressing to 50–60 minutes in duration. The patient should walk at a speed that induces claudication within approximately 3–5 minutes, and should continue walking until symptoms reach moderate intensity. At that point, the patient should rest until symptoms have completely resolved, and then repeat the process for the duration of the session. The grade or speed of the treadmill should be increased when the patient can walk 8–10 minutes without developing moderate claudication. Studies have shown that a 24-week program is more effective than a 12-week program. If the patient cannot tolerate a treadmill program, arm ergometer training also causes a beneficial training response.

Multiple mechanisms of action may contribute to the success of an exercise program in patients with PAD. First, exercise improves endothelial cell function, with increased vasodilation in response to nitric oxide. Additionally, exercise potentially enhances mitochondrial oxidative energy production in place of glycolytic metabolism. Lastly, an exercise program decreases the presence of inflammatory markers, including interleukin-6, CRP, and adhesion molecules.

B. Pharmacotherapy

In addition to exercise therapy, a number of medications have been tested as therapy for intermittent claudication. Two of these medications, cilostazol and naftidrofuryl, have shown clear benefit for symptomatic improvement. Cilostazol is a phosphodiesterase III inhibitor that causes vasodilation and decreased platelet aggregation. In clinical studies, it has been shown to increase
pain-free walking distance, absolute walking distance, and quality of life in patients with intermittent claudication. Naftidrofuryl is a 5-hydroxytryptamine receptor antagonist that also increases intracellular ATP production. It has a moderate effect on improving pain-free walking distance, treadmill performance, and quality of life in the first 6 months of therapy. These medications can be used for symptom improvement in patients with intermittent claudication, but lifestyle modifications with cardiovascular risk factor reduction should always be used simultaneously. In addition to these two medications, carnitine and propionyl-L-carnitine, which are thought to improve skeletal muscle metabolism, have shown some benefit in improvement of treadmill walking distance and quality of life, but larger studies are needed to determine their clinical effects.

Other medications that have been studied with insufficient evidence include pentoxifylline, prostanoids, and buflomedil. Pentoxifylline is a nonselective phosphodiesterase inhibitor that decreases blood viscosity while improving erythrocyte flexibility, microcirculatory flow, and tissue oxygenation. Studies involving pentoxifylline have generally been poor quality, and no clear benefit for treating intermittent claudication has been shown. The prostanoids prostaglandin E\(_1\) (PGE\(_1\)) and prostacyclin (PGI\(_2\)) are thought to work through many mechanisms of action, including direct vasodilation, inhibition of neutrophils, reduction of vascular smooth muscle proliferation, and inhibition of platelets, among others. Despite their many beneficial effects, they have also not been proven to have clear efficacy in intermittent claudication. Buflomedil inhibits platelet aggregation, improves erythrocyte flexibility, and reduces plasma fibrinogen, with a resultant decrease in blood viscosity. This medication has been widely used for many years for intermittent claudication, but insufficient evidence is available to support its use. Additionally, occasional lethal neurologic and cardiovascular adverse effects have been reported in accidental or intentional overdoses.

### C. Management of Critical Limb Ischemia

Patients with more proximal symptomatic PAD or critical limb ischemia (Figure 3–2) will most likely require more invasive therapy consisting of revascularization. The primary goals of treatment of critical limb ischemia should be pain relief, ulcer healing, and avoidance of limb loss. No medications have been shown to benefit symptoms or healing in critical limb ischemia. Some evidence supporting the use of hyperbaric oxygen therapy or spinal cord stimulation is available, but these therapies may be best suited for patients who cannot undergo revascularization. The revascularization process can entail either
surgical or endovascular procedures, which can be further subdivided into angioplasty and stent placement. A vascular specialist should perform these procedures, the details of which are beyond the scope of this chapter. However, studies have shown that supervised exercise training after or in conjunction with revascularization is superior to revascularization alone. Therefore, the importance of exercise therapy in all stages of PAD cannot be overemphasized.
Patients with acute arterial occlusion require immediate referral to a vascular specialist and anticoagulation with intravenous heparin. Emergent revascularization must be achieved within 6 hours of onset to prevent irreversible injury to the ischemic tissue. In cases of irreversible tissue damage, amputation is the final treatment option. Amputation can also be considered when patients have severe pain or infection with PAD that is not amenable to revascularization procedures. This outcome is a rare one for patients with PAD and is estimated to be less than 3% over a 5-year period.
Prognosis

The annual rate of cardiovascular events, including myocardial infarction, ischemic stroke, and vascular death, for patients with PAD is approximately 5–7%. These events are also the most common causes of death among patients with PAD (myocardial infarction, 40–60%; ischemic stroke, 10–20%, and vascular death, 10%). One study reported a 2.5–3.5-fold increased risk of mortality and cardiovascular events in patients with proximal PAD as compared with distal PAD. Additionally, the ABI has been shown to be correlated to mortality in patients with PAD, with every 0.10 decrease in ABI corresponding to a 10% increase in relative risk for cardiovascular events. In patients with chronic critical limb ischemia, the 1-year mortality rate is approximately 20%. In most people with PAD, leg symptoms remain stable. About 10–15% of patients improve, and 15–20% get worse. The outlook is better for people who are able to remain tobacco-free, stay on a healthy diet, keep their blood cholesterol under control, and exercise regularly.


Thromboangiitis obliterans, also known as Buerger’s disease, is a nonatherosclerotic, segmental, inflammatory disease that affects small and medium-sized arteries and veins of the extremities. It is a vasculitis characterized by an inflammatory thrombus with noninvolvement of the vessel wall. The disease is most prevalent in the Middle East and Asia. In the United States the incidence of thromboangiitis obliterans is about 12 per 100,000; it is more common in males, with onset most often occurring between the ages of 40 and 45 years. Overall prevalence has declined in the United States in conjunction with smoking cessation efforts. Smoking is essential for the initiation and progression of thromboangiitis obliterans. Additional risk factors include cigar smokers, marijuana use, smokeless tobacco use, and chronic periodontal infection.

Clinical Findings

A. Symptoms and Signs

Patients with thromboangiitis obliterans are typically young smokers with ischemic symptoms (claudication of feet and hands more than of legs or arms).
Superficial thrombophlebitis often occurs prior to symptoms of ischemia and consists of tender nodules that follow a venous distribution. Generally, at least two extremities are involved. Distal arteries and veins are affected first followed by involvement of more proximal vessels. Ulcerations and rest pain can be presenting symptoms in patients with more advanced disease. Raynaud’s phenomenon may be present in 40% of patients.

Patients should have a comprehensive neurovascular examination. A detailed history should be obtained, including questioning about use of all tobacco products. Referral to a vascular specialist is essential so that appropriate clinical testing and imaging can be performed. ABI and wrist–brachial indexes should always be obtained in patients with signs of ischemia. An Allen’s test may be performed; this involves compressing one of the arteries of the hand after blood has been forced out by clenching it into a fist. Failure of the blood to diffuse to the hand when opened indicates that the artery not compressed is occluded. A positive Allen’s test in a young smoker with digit ischemia can suggest thromboangiitis obliterans; 70% of patients may have sensory deficits on neurologic examination.

**B. Diagnostic Studies**

There are no specific laboratory tests to diagnose thromboangiitis obliterans. In addition to routine laboratory studies, the workup should include ESR, CRP, antinuclear antibodies, anti-centromere antibody, anti-SCL-70, cold agglutinins, and cryoglobulins. Laboratory studies are done to rule out other disease states than can have a similar presentation. Imaging and biopsy are rarely needed for diagnosis. When arteriography is performed findings include segmental occlusion, distal involvement of small to medium-sized vessels, and corkscrew collaterals (Figure 3–3). There is no evidence of atherosclerosis.
Treatment

Treatment focuses on smoking cessation; this is the only definitive therapy for thromboangiitis obliterans. Noncompliance causes significant progression and can result in amputation. Nicotine replacement should not be offered as it may cause disease progression. Pharmacologic and group therapy should be part of a comprehensive smoking cessation plan. Surgical revascularization is rarely performed owing to distal vascular involvement. Other treatments with variable results include iloprost infusion, sympathectomy, calcium channel blockers, and intermittent pneumatic compression.
LYMPHEDEMA

ESSENTIALS OF DIAGNOSIS

- Lymphedema results when the amount of lymph exceeds transport capacity.
- The most common cause in the United States is malignancy and its treatment.
- Patients usually present with unilateral swelling of a limb.
- Radionucleotide lymphoscintigraphy is the gold standard for evaluation of the lymphatic system.

General Considerations

Lymphedema is a progressive, insidious condition resulting from the collection of protein-rich fluid in the interstitium due to a disruption in lymphatic flow. The lymphatic system functions to transport interstitial fluid that is collected by lymph capillaries to the blood. Lymphedema results when the amount of lymph exceeds transport capacity, resulting in fluid accumulation in the interstitial
Pathogenesis

The transportation of lymph occurs in a low-pressure system that is not closed, unlike the high-pressure and closed cardiovascular system. Movement occurs through contraction and relaxation of smooth muscle along with compression from adjacent skeletal muscle contraction and arterial pulsation. One-way valves prevent backflow during transport. The thoracic duct collects the lymph from most of the body and drains into systemic circulation at the left brachiocephalic vein. The areas not collected by the thoracic duct include the right upper body, right head and neck, and occasionally the left lower lung; these areas enter venous circulation through the right lymphatic duct.

Lymphedema is categorized into primary and secondary types. Primary lymphedema refers to a congenital or inherited dysfunction of the lymphatic system; it can also be further subdivided based on age at presentation. Congenital lymphedema has an autosomal-dominant inheritance pattern and occurs from birth to 2 years of age. Lymphedema praecox, also autosomal dominant, typically presents at puberty; it usually occurs in females and is the most common cause of primary lymphedema. Lymphedema tarda is the least common of the congenital lymphedemas. It is seen in individuals older than 35 years of age and is thought to be caused by incompetent valves.

Secondary lymphedema is caused by injury to the lymphatic system, resulting from either disease or iatrogenic causes. It is associated with malignancy, infection, obesity, and trauma. The most common cause of secondary lymphedema worldwide (affecting 120 million people) is filariasis, which results from infection with the parasitic nematode *Wuchereria bancrofti*. Lymphatic filariasis occurs when adult worms occupy the lymphatic system and obstruct lymphatic flow. Chronic infections can lead to elephantiasis, and the related lymphedema is permanent. In the United States, secondary lymphedema is associated with malignancy and its treatment. Secondary lymphedema can occur from direct infiltration of lymphatic vessels and channels by a malignant growth or as a result of treatments such as lymphadenectomy and radiation therapy.

Overwhelmingly, breast cancer is the most common malignancy associated with lymphedema, and subsequently the most studied. This is largely due to treatments and surgery associated with breast cancer, such as axillary node dissection and radiation therapy. Upper extremity edema develops in 10–40% of
women undergoing axillary node dissection. The risk of lymphedema with axillary node dissection increases significantly with the addition of radiation therapy compared with axillary node dissection alone; in one study, rates of lymphedema were doubled with the addition of radiation therapy. Although radiation therapy is often necessary, axillary node dissection can sometimes be avoided. The advent of sentinel lymph node biopsy has decreased the number of unnecessary axillary node dissections. Sentinel node biopsy decreases the risk of lymphedema by more than half when compared with axillary node dissection. (For further discussion of lymphedema in cancer patients, see Chapter 35.)

Additional causes of secondary lymphedema include obesity and infection. In patients with morbid obesity lymphatic return is impaired, which results in significant edema. Infectious causes of lymphedema are not limited to filariasis. It has been reported that recurrent cellulitis and streptococcal lymphangitis can also cause lymphedema.

Clinical Findings

Patients with lymphedema usually present with unilateral swelling of a limb, which can involve both the upper and lower extremities (Figure 3–4). The edema is initially pitting and progresses from distal to proximal. The pitting edema eventually develops into nonpitting edema after fibrosis of the subcutaneous fat. Patients complain of tightness or heaviness of the affected extremity. Over time the dermis continues to thicken and elephantiasis nostra verrucosa (ENV) can develop. ENV is characterized by hyperkeratosis and papillomatosis of the epidermis, with underlying woody fibrosis of the dermis and subcutaneous tissue.
Figure 3–4 A, B, C: Lymphedema.

Fissuring and skin breakdown can also occur with chronic edema, causing recurrent infections. A positive Stemmer sign—the inability to pinch the dorsal aspect of the skin between the first and second toes—may be seen on physical examination. If edema is extensive, range of motion in the affected joint can be significantly diminished. This can cause difficulty with activities of daily living and have a negative impact on patient quality of life.

Measurements should be recorded in patients with lymphedema to aid in diagnosis, help in determining the effectiveness of treatment, and monitor for progression of disease. The most common measurements are circumferential measurements of the extremities. A difference of more than 2 cm between extremities is deemed significant. Water displacement, another method of measuring for lymphedema, has been shown to effectively diagnose early lymphedema in postoperative breast cancer patients. Volume measurements are taken preoperatively to obtain a baseline. A difference of 200 mL in volume measurement postoperatively is considered clinically significant edema. Diagnostic imaging should begin with radiographs followed by Doppler ultrasound. Radiographic imaging will allow a fracture to be ruled out as a cause of edema, while Doppler ultrasound evaluates the deep and superficial vasculature. Radionuclide lymphoscintigraphy is the gold standard for the evaluation of the lymphatic system. It is a safe, repeatable diagnostic modality that identifies lymphatic dysfunction.

Treatment

In the treatment of lymphedema, a standardized staging system is used to ensure accurate communication among the multidisciplinary group of providers treating patients. The most commonly used staging scale was put forth by the International Society of Lymphology (Table 3–4).

Table 3–4 International Society of Lymphology staging scale for lymphedema.
Regardless of the etiology of lymphedema (primary or secondary), treatment begins with complete decongestive therapy (CDT). Early diagnosis and treatment is beneficial and can prevent progression to the chronic phase. CDT is a two-phase system. The first phase involves maintaining excellent hygiene with regard to skin and nail care in hopes of reducing infection. Other components include manual lymphatic drainage (MLD) and limb compression for 24 hours. MLD is a massage-like modality that moves fluid from distal to proximal with the goal of augmenting superficial lymph drainage. Therapy should occur at least 5 days per week, and weekly limb measurements should be obtained to monitor for progression. The goal of the second phase of CDT is to preserve the gains made in phase 1. Compression garments should be worn during the day, and self-compression bandages may be worn at night. Measurements should be taken every 6 months.

To be beneficial the components of CDT require strict compliance. Compression bandages should be worn 24 hours a day during phase 1 of CDT. The goal of compression bandaging is to diminish ultrafiltration by applying external compression. The bandages consist of many layers of padding and short (low) stretch bandaging. The principle for short-stretch bandaging is that it is not active at rest. However, the pressure applied during patient movement (muscle contraction) is believed to cause mechanical stimulation of the smooth muscles

<table>
<thead>
<tr>
<th>Stage</th>
<th>Clinical Description</th>
</tr>
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<tbody>
<tr>
<td>0</td>
<td>Latent stage. There is damage to the lymphatic vasculature, but the transport capacity is still sufficient. No clinical edema.</td>
</tr>
<tr>
<td>1</td>
<td>Spontaneously reversible. Edema will subside with 24 hours of limb elevation. Edema may pit.</td>
</tr>
<tr>
<td>2</td>
<td>Spontaneously irreversible. Fibrosis begins in this stage. Edema will not resolve with 24 hours of limb elevation.</td>
</tr>
<tr>
<td>3</td>
<td>Lymph static elephantiasis. Tissue is fibrotic. Edema is irreversible.</td>
</tr>
</tbody>
</table>
in lymphatic vessels. Compression garments are used during phase 2 of CDT; these are fitted, elastic, and have two-way stretch. They function by creating greater pressures distally than proximally, which promotes fluid mobilization. The amount of pressure exerted is variable and can be as much as 50 mm Hg. Compression garments should be custom fitted and replaced every 3–6 months.

Treatment of lymphedema can be challenging, and much emphasis has therefore been placed on prevention, which can be divided into primary and secondary components. With regard to primary prevention, it is recommended that sentinel node biopsy be performed over axillary node dissection whenever possible. Other techniques include limiting the extent of lymph node dissection and advanced radiation therapy techniques. Prophylactic exercise has been shown to reduce the incidence of lymphedema after anterior lymph node dissection. However, weighted and resistance exercises are not recommended during the acute postsurgical phase.

Secondary prevention focuses on patient education. Patients should be taught how to properly measure and monitor their skin for changes, and should be instructed to practice strict skin hygiene to prevent infection and avoid medical procedures in the affected limb. Ideal body weight should be maintained, as obesity is a known risk factor for lymphedema. Exercise (aerobic and resistive) has been shown to reduce the symptoms of lymphedema and reduce exacerbations, in addition to increasing strength. The National Lymphedema Network recommends that compression garments be worn during exercise.

Treatment of lymphedema is typically nonsurgical; however, some indications for surgical therapy include failed medical therapy and recurrent cellulitis. Two types of procedures are differentiated: physiologic and reductive. Physiologic procedures increase the capacity of the lymph system by creating new channels. Reductive procedures involve removing fibrotic and fatty tissue. More investigation is needed regarding the role of surgical intervention in the treatment of lymphedema.


Harris SR, Hugi MR, Olivotto IA, et al.: Clinical practice guidelines for the


SUPERFICIAL THROMBOPHLEBITIS
ESSENTIALS OF DIAGNOSIS

- Pain, erythema, and tenderness to palpation overlying superficial veins.
- Lower extremity is affected more often than upper extremity.
- Commonly occurs in patients with varicose veins.
- Deep vein thrombosis must be ruled out as a differential diagnosis.

General Considerations

Superficial thrombophlebitis (STP) is a condition that most commonly manifests as an area overlying superficial veins in a lower extremity that becomes erythematous, painful, tender, and warm to touch; often a palpable mass is present. There are two underlying mechanisms in its development: inflammation and clot formation. STP is estimated to affect approximately 3–11% of the general population and more commonly involves the greater saphenous vein rather than small saphenous vein. It is also more commonly found in individuals with varicose veins.

Clinical Findings

The diagnosis of STP can often be made clinically, based on the previously described findings. However, venous duplex ultrasound should be performed if the area involved is in the proximal one third of the medial thigh, clinical extension is noted, and the leg is more swollen than expected. If STP is present Doppler findings would include vein wall thickening, subcutaneous edema, and possible luminal thrombosis. A biopsy specimen can be obtained (although rarely needed), which may show evidence of acute vasculitis of medium- to large-sized veins in the upper subcutaneous and lower dermis regions. In addition, evidence of inflammatory cells, wall thickening, connective tissue formation, and eventual vessel recanalization if thrombus has been present may be seen histologically.
Differential Diagnosis

Although commonly seen in patients with concomitant varicose veins, STP has a broad list of other causes of which physicians must be aware. They include but are not limited to hypercoagulable states (as seen in pregnancy, oral contraceptive use, malignancy), other inherited or acquired thrombophilic states (eg, protein C or S deficiency), vessel wall injury resulting from needle puncture or substance infusion, thromboangiitis obliterans (Buerger’s disease), infection, and deep vein thrombosis (DVT). If clinically indicated, a workup should be performed to rule out these more serious causes. In individuals with bilateral STP, involvement of proximal greater saphenous vein and coexisting risk factors, DVT should be suspected. Finally, erythema that extends beyond the vein border in a patient who has a fever should prompt consideration of suppurative thrombophlebitis.

Treatment

The treatment of STP is often conservative, with a majority of cases spontaneously resolving in 2–3 weeks. Elevation of the affected extremity, cool or warm compresses for pain relief, continued ambulation, initiation of compressive stockings (barring any contraindication), and nonsteroidal anti-inflammatory drugs are all accepted forms of conservative treatment. It is not uncommon for STP to reoccur in the same vessel but different segments over time. The American College of Chest Physicians recommends treatment with therapeutic anticoagulation when the segment of vein involved is greater than or equal to 5 cm in length, within 5 cm or less of the deep venous system, and medical risk factors are present. Treatment duration is for a total of 4 weeks. Surgical vein ligation or excision is often the treatment of last resort.


Leon L, Giannoukas A, Dood D, et al.: Clinical significance of superficial
DEEP VEIN THROMBOSIS

DVT is discussed in detail in Chapter 5 (see Venous Thromboembolism [Deep Vein Thrombosis]).

CHRONIC VENOUS INSUFFICIENCY

ESSENTIALS OF DIAGNOSIS

- Lower extremity venous dysfunction resulting from venous hypertension.
- Aching, swelling, or heaviness of the lower extremity that is worse with activity.
- Symptoms improved with rest and leg elevation.

General Considerations

Chronic venous insufficiency (CVI) should be thought of as occurring on a continuum of chronic venous disease. More specifically CVI is the term given to later stage findings in chronic venous disease, equating to a Clinical–Etiology–
Anatomy–Pathophysiology (CEAP) classification of 4–6, as depicted in Table 3–5. CVI has been defined as venous dysfunction resulting in edema, skin discoloration, and ulceration due to a decreased ability to pump oxygen-depleted blood back to the heart. Vessels commonly involved include the greater saphenous vein, small saphenous vein, and perforator veins connecting the superficial and deep venous systems of the lower extremities. According to the Vascular Disease Foundation, venous insufficiency is 10 times more common than PAD in the United States, with more than 6 million people having CVI and a half million having venous ulcers.

Table 3–5 CEAP classification of chronic venous disorders.
<table>
<thead>
<tr>
<th>Clinical classification</th>
<th>C0</th>
<th>No visible or palpable signs of venous disease</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>C1</td>
<td>Telangiectasias or reticular veins</td>
</tr>
<tr>
<td></td>
<td>C2</td>
<td>Varicose veins</td>
</tr>
<tr>
<td></td>
<td>C3</td>
<td>Edema</td>
</tr>
<tr>
<td></td>
<td>C4a</td>
<td>Pigmentation or eczema</td>
</tr>
<tr>
<td></td>
<td>C4b</td>
<td>Lipodermatosclerosis or atrophie blanche</td>
</tr>
<tr>
<td></td>
<td>C5</td>
<td>Healed venous ulcer</td>
</tr>
<tr>
<td></td>
<td>C6</td>
<td>Active venous ulcer</td>
</tr>
<tr>
<td></td>
<td>S</td>
<td>Symptomatic, including ache, pain, tightness, skin irritation, heaviness, and muscle cramps, and other complaints attributable to venous dysfunction A: asymptomatic</td>
</tr>
<tr>
<td>Etiologic classification</td>
<td>Ec</td>
<td>Congenital</td>
</tr>
<tr>
<td></td>
<td>Ep</td>
<td>Primary</td>
</tr>
<tr>
<td></td>
<td>Es</td>
<td>Secondary (post-thrombotic)</td>
</tr>
<tr>
<td></td>
<td>En</td>
<td>No venous cause identified</td>
</tr>
<tr>
<td>Anatomic classification</td>
<td>As</td>
<td>Superficial veins</td>
</tr>
<tr>
<td></td>
<td>Ao</td>
<td>Perforator veins</td>
</tr>
<tr>
<td></td>
<td>Ad</td>
<td>Deep veins</td>
</tr>
<tr>
<td></td>
<td>An</td>
<td>No venous location identified</td>
</tr>
<tr>
<td>Pathophysiologic classification</td>
<td>Pr</td>
<td>Reflux</td>
</tr>
<tr>
<td></td>
<td>Po</td>
<td>Obstruction</td>
</tr>
<tr>
<td></td>
<td>Pr,o</td>
<td>Reflux and obstruction</td>
</tr>
<tr>
<td></td>
<td>Pn</td>
<td>No venous pathophysiology identifiable</td>
</tr>
</tbody>
</table>

Primary and secondary causes of CVI are differentiated. Primary causes include intrinsic differences in vein morphology, wall weakness, and valve incompetence. Secondary causes include DVT, STP, and atriovenous fistulas. May–Thurner’s syndrome, occlusion of the left iliac vein by an overlying right common iliac artery, has also been described as potential cause of CVI. Whether from primary or secondary causes, the mechanism creating the changes seen in CVI results from elevated venous hypertension.

The cascades of biochemical changes resulting from venous hypertension are numerous. Both microcirculatory and macrocirculatory systems are affected, with evidence of leukocyte adhesion to endothelium, and increased platelets, fibrin, and erythrocyte aggregation in areas of elevated pressures, causing inflammatory and hypoxic changes to the vessel and capillaries. The histologic changes that result disrupt the flow of blood from the superficial to deep venous system, ultimately causing reversal of flow and a decreased ability to return blood that is pooling in the lower extremity.

Clinical Findings

Commonly described symptoms of CVI include aching, burning, cramps, swelling, heaviness, and restless legs. Symptoms are often exacerbated with heat or activity and improved with rest and leg elevation. Signs of CVI include lower extremity edema, hyperpigmentation of skin around the ankles (caused by extravasation of blood cells), visible or palpable vein aneurysms, lipodermatosclerosis (Figure 3–5; an inflammation of fat beneath the skin), skin ulceration (Figure 3–6), and decreased ankle mobility. Clinicians must rule out the presence of PAD, as well as peripheral neuropathy, as these conditions can also produce ulcerative changes to the lower extremities. Many tests, including plethysmography, contrast venography, intravascular ultrasound, computed tomography, and magnetic resonance venography, have been used to confirm the presence of CVI; however, venous duplex ultrasound has become the test of choice.
Figure 3–5 Lipodermatosclerosis.
Treatment

A. Compression Therapy

The cornerstone of treatment in CVI is compression, which may be applied in various ways. These methods include elastic dressings, rigid compression, and intermittent pneumatic pressure. Most of the studied beneficial results of compression were achieved using compression stockings that provided 35–45 mm Hg of pressure. CVI patients who may benefit from compression include those with significant edema, weeping areas, lipodermatosclerosis, and venous ulceration. Contraindications to compression therapy include moderate to severe PAD, cellulitis, and DVT. Patients with an ABI of 0.6–0.9 can be started on compression therapy, but cautiously. Major concerns when using this therapy include skin breakdown over bony surfaces and compression neuropathies. For individuals who cannot tolerate this therapy, intermittent pneumatic compression can be performed for 4 hours daily. In addition, leg elevation should be promoted by positioning the affected leg above the level of the heart for 30 minutes, three to four times daily.

B. Pharmacotherapy
There are no Food and Drug Administration (FDA)–approved medications for CVI; however, some venoactive drugs have been shown to improve venous tone and capillary permeability. The Society for Vascular Surgery gives grade 2b recommendations for the use of diosmin, hesperidin, micronized purified flavonoid fraction (MPFF), or horse chestnut seed extract for pain and swelling. In addition, grade 2b recommendations have been given for the use of pentoxifylline or MPFF, along with compression, to assist in venous ulcer healing. Aspirin has also been shown to increase the rate of healing and decrease ulcer relapse rates.

Currently there is no evidence that the use of hydrocolloid dressings beneath compression stockings produces any healing benefit over compression alone. Moreover, there is no role for diuretics in the treatment of edema in patients with CVI. Antibiotics are not useful, unless there is clinical suspicion of infection; in such cases, antibiotics that are effective against MRSA or *Pseudomonas*, or both, should be utilized until results of cultures are obtained.

**C. Exercise**

The effects of exercise have also been examined in patients with CVI. Patients with severe CVI have decreased ankle joint range of motion, as well as decreased calf muscle pump function. Exercise in the form of lower extremity flexibility and strengthening—more specifically, focusing on ankle strength and power training—have been shown to improve calf blood flow hemodynamics, ankle joint mobility, and calf strength.

**D. Surgical Treatment**

Invasive treatment options should be considered if patients do not respond to conservative measures. A vascular specialist should make the decision as to timing of and procedure selection, if required. For this reason it is important that CVI patients establish care with one of these specialists early in the disease process. Superficial vein incompetence, perforator vein incompetence, deep vein incompetence, and deep vein obstruction are the specific anatomic areas that either surgical or endovascular intervention will target. Surgical interventions include vein ligation, division, or stripping and various vein–vein bypass surgeries. Additionally, surgical debridement of an ulcer can be performed if clinically indicated. Endovascular procedures accomplish venous ablation through multiple techniques, including radiofrequency, laser, thermal, and sclerotherapy, as well as stenting of deeper veins.
VARICOSE VEINS

ESSENTIALS OF DIAGNOSIS

- Abnormally dilated, elongated, and tortuous subcutaneous veins greater than 3 mm in diameter.
- Affects the superficial venous system most notably, particularly the great and small saphenous veins.
- Although linked with valvular dysfunction, varicosities may form in the absence of such dysfunction as a result of local venous incompetence alone.
General Considerations

Varicose veins are abnormally dilated, elongated, and tortuous subcutaneous veins that form as a progression of dilation from normal vein diameter, through small enlargements known as telangiectasias or spider veins, eventually crossing the threshold required to define a vein as varicose (3 mm in diameter). Among adults in the United States the prevalence of varicose veins is approximately 23%. Varicose veins are twice as common in women as in men; it is estimated that in the United States 22 million women, as compared with 11 million men, are affected. Risk factors include female sex, prolonged standing, increased height, congenital valvular dysfunction, venous hypertension of obesity, and multiple pregnancies. Inheritance is suggested by autosomal-dominant lineage with incomplete penetrance. In patients who do not have primary venous disease, varicose veins can develop as part of the post-thrombotic syndrome after DVT or result from congenital venous malformations.

Varicose veins occur in the lower limbs because of upright position and increased hydrostatic pressure. Varicosities are most often seen in superficial veins, because they are subject to greater resistance to flow. Gravity, in addition to hydrostatic fluid column pressure, disallows spontaneous venous blood return from the legs upward to the heart in an upright individual. The lower limb musculature solves this dilemma by providing pump-like action for antigravity movement. The returning blood column cannot be continually driven toward the heart by compression alone; thus, veins have built-in oneway valves inhibiting retrograde flow, which, in combination with muscle activity, allow for stepwise blood progression through the venous vasculature.

Pathogenesis

In recent years there has been a paradigm shift away from theories proposing that mechanical dysfunction, including valvular incompetence and venous or arteriovenous malformation, were the primary causes of varicose veins. Hypotheses about causation now emphasize complex molecular and histopathologic alterations in the vessel wall and the extracellular membrane, which are now known to be consistent in all cases of varicose veins. Increased venous pressure causes stretching of the endothelium, which can in turn promote expression of cytokines and adhesion molecules, activation of extracellular signal-related kinases, and free radical production in venous varicosities. Although valvular incompetence and localized venous hypertension are found in
most patients with varicose veins, the histopathologic changes are uniformly present. This is the leading impetus for the paradigm shift regarding pathogenesis. Nonetheless, because the well-known mechanopathologic forces are present in the majority of patients with varicose veins, it stands to reason that valvular incompetence and venous hypertension likely modulate disease severity.

**Clinical Findings**

Varicose veins vary widely in clinical presentation. A patient may be completely asymptomatic with very large varicosities noted clinically or may present with very small varicose veins and significant clinical symptoms. In the classic presentation, the enlarged veins may lead to discomfort, skin changes, and emotional distress secondary to unaesthetic cosmetic appearance, as well as impairment in activities of daily living. Varicose veins are classified using the CEAP (Clinical–Etiology–Anatomy–Pathophysiology) system previously described for CVI (see Table 3–5, earlier).

Symptoms and signs typically include swelling, dermatic changes (termed *stasis dermatitis* or *venous eczema*), restlessness, limb heaviness and fatigue, nocturnal leg cramps, atrophic blanche (whitened, irregular scar-like patches at stasis areas), and direct tenderness. Often patients also complain of varied sensations ascribed to varicosities, including aching, throbbing, burning, and tingling. In severe cases, patients may present after the development of stasis ulcers (*Figure 3–7*).
The clinical workup prior to any intervention should include a complete pulse examination to rule out arterial association and a complete duplex ultrasound examination to evaluate both the superficial and deep venous systems.

**Complications**

As most varicose vein cases are benign the complication rate for varicose veins overall is quite small, and life-threatening complications are rare. However, in relation to the associated clinical findings, a variety of complications may
develop secondary to venous stasis, localized venous hypertension, and limb edema. Pain, emotional distress, and deleterious effect on activities of daily living, including ambulation and ability to perform workplace functions, have been noted. In addition, skin changes can progress to dermatitis, cellulitis, lipodermatosclerosis (hardening of the subcutaneous fat layer), venous ulcerations (see Figure 3–7), and, in severe cases (more often in obese patients), fat necrosis.

### Treatment

#### A. Conservative Measures

Conservative treatment options for varicose veins include graduated compression stockings, medications, leg elevation, and lifestyle changes, including weight loss and regular exercise. The mainstay of first-line therapy is variable-gradient compression stockings, which have been shown to decrease discomfort and correct swelling, as well as to enhance cellular nutritional exchange and improve microcirculation. Compression forces of 20–30 mm Hg are implemented to help with pain and edema control. Patients with venous stasis ulcers are fitted with 30–40 mm Hg stockings. Compression stockings have practical limitations that may limit compliance. The elderly may not be able to don or doff the stockings effectively, morbidly obese patients may have trouble fitting into the stockings initially, and patients with prestocking skin damage may have trouble with skin management.

Medications have been shown to benefit some patients with varicosities, but the study size was too limited to allow conclusive recommendations for therapeutic use. Escin, a purified horse chestnut seed extract, is used in Europe and acts by inhibiting platelet aggregation, which in turn limits edema and capillary permeability and may improve lymphatic drainage. Purified flavinoids (eg, Diosmin) reduce symptoms of pain and heaviness in patients with varicose veins, and can be used as an active adjunct to compression stockings in ulcer healing.

#### B. Invasive Procedures

Invasive treatment options are available if symptoms persist, if size or location of varicose veins prove functionally or cosmetically limiting, or if the patient prefers or desires a procedural approach. Sclerotherapy and vein stripping are two commonly performed treatment measures.
In sclerotherapy, a sclerosing agent is injected into the problematic vein to induce endothelial damage; the ensuing thrombosis and fibrosis eventually close off the vessel from future blood flow. Three classes of drugs are used for this purpose: osmotics, alcohols, and detergents. The most commonly used sclerosant worldwide is Polidocanol, a detergent that is approved by the U.S. FDA for use in sclerotherapy. Complications of sclerotherapy include allergic reactions to the sclerosants, hyperpigmentation caused by extruded hemosiderin, matting (which occurs when very fine capillaries develop at the site of injection and become inflamed, causing cellulitis), and, rarely, small ulcerations at the injection site. Sclerotherapy is often used in refractory or recurrent cases. A Cochrane Review concluded that sclerotherapy was preferable to surgery in the short term (< 1 year) with regard to treatment success, complications, and cost, but that surgery was better in the long term (at > 5 years) using the same measurement variables. Multiple treatments are often required, and complete resolution of varicose veins using sclerotherapy alone is unlikely, especially as diameter increases.

The surgical option for correction of varicose veins is termed *stripping* and involves the removed of the vein, in its entirety, from the leg, thereby correcting the varicosity immediately. When first described, the procedure called for venous stripping of the entire saphenous vein, from ankle to groin. However, this approach was found to increase the risk of neurapraxic injury to the vein, which can be permanent. Subsequently, the approach was adjusted to recommend removal of that portion of the saphenous vein between the groin and the calf. Recurrence of varicose veins after stripping is a major drawback estimated to occur in 10–50% of patients as collateral vessels form. Other complications of stripping, secondary to the procedure’s invasiveness, include wound infections and delayed healing. Often stripping is combined with microphlebectomy, a less invasive surgical approach in which multiple small incisions are made and portions of the vein removed, thereby halting flow throughout the vein as a whole. Microphlebectomy can also be used as monotherapy, and has the added benefit that it can be performed in an outpatient setting if the targeted veins are small enough and do not require suturing.

Surgical vein procedures have in most cases been supplanted by less invasive endovenous percutaneous procedures, which produce equivalent results that are associated with less risk and reduced rehabilitation needs. Endovenous laser ablation acts by means of local direct thermocoagulation of the vein, causing cessation of flow. It is more useful in telangiectasias (< 1.0 mm) but can be used for varicosities on the smaller end of the spectrum. Endovenous radiofrequency ablation acts in a similar fashion, but uses high-intensity signals to produce
thermic energy. These approaches are especially helpful in patients with varicosities of the ankle and foot, which are difficult to treat using sclerotherapy and have higher rates of ulcer formation. Other benefits of this treatment option include lack of complications related to small capillary formation and matting.

**Prognosis**

The prognosis for patients who receive appropriate treatment is good, because even if treatment strategies call for surgical or invasive cessation of blood flow through the affected vein, the overall venous return is relatively unaffected.


RAYSNAUD’S SYNDROME

ESSENTIALS OF DIAGNOSIS

- Manifests in two forms (primary and secondary). The primary form (Raynaud’s disease) is a vasospastic disorder for which there is no identifiable underlying cause. The secondary form (Raynaud’s phenomenon) connotes vasospasm that occurs secondary to another underlying condition or disease.
- Characterized by episodic attacks of vasospasm in response to cold or an emotional trigger.
- Diagnosis is clinical.
- The classical appearance is intense pallor of distal extremity followed in sequence by cyanosis and then hyperemia on rewarming.

General Considerations

Initially described in 1862, Raynaud’s syndrome (RS) remains a poorly understood clinical condition. It is more common in females than males and presents most commonly between ages 20 and 40 years. The syndrome can occur as an isolated entity, historically referred to as Raynaud’s disease or primary RS, or in association with other conditions, often referred to as...
Raynaud’s phenomenon or secondary RS. Secondary RS is associated most frequently with connective tissue diseases, including scleroderma, systemic lupus erythematosus, rheumatoid arthritis, and Sjögren’s syndrome, but can also be associated with atherosclerosis, arteritis, cancer, collagen vascular disease, thoracic outlet syndrome, embolic occlusion, occupational disease, and certain medications.

Attacks usually last 30–60 minutes and can vary in frequency, pain, and impairment. In some patients attacks occur only seasonally in the cold of winter, producing mild symptoms; others experience multiple episodes per day that are associated with severe pain and distal extremity ulceration. Overall, incidence of ischemic ulceration is quite rare in RS but is seen more often in severe secondary RS. Episodes of RS usually affect distal extremities bilaterally, whereas primary RS occasionally has unilateral preference.

The exact pathogenesis of distal extremity vasospasm remains unknown. Theories include exaggeration of the thermoregulatory response as well as abnormalities in peripheral adrenoreceptor activity or expression, blood viscosity, or endothelial function.

**Clinical Findings**

The clinical presentation of RS, primary or secondary, is characterized by cold- or stress-induced skin color changes that follow, classically, a described three-phase pattern. Initially the digits blanch in comparative isolation to the proximal extremities. Second, the distal digits become cyanotic and turn shades of blue. The third phase is described as a progression through hyperemia and rubor as the digits warm and revascularize. However, most patients do not progress through the three phases as classically described, noting only pallor and cyanosis. The fingers and hands are most often affected. In certain patients, the toes and feet may be involved. Affected less commonly are the tips of the tongue and nose.

As witnessed events are rare because of the transient and randomly episodic nature of RS, the diagnosis remains primarily clinical and is based mostly on patient descriptions of cold or emotionally induced skin changes. Practitioners often request that patients provide self-taken photographic evidence for verification and assessment. To rule out arterial obstruction as the cause of symptoms, upper extremity pulse volume recordings should be performed. Photoplethysmographic waveform recording is a modality that allows the examiner to differentiate between vasospastic and obstructive causes. Laser Doppler and digital thermography have been used primarily in research studies.
but have not found practical clinical use. Other tools, such as “cold challenge tests,” with ice water immersion and monitored temperature recovery, are highly sensitive but lack specificity. In addition, secondary to associated disorders of RS, routine screening for antinuclear antibody and rheumatoid factor are commonly done.

Complications

The most common complication is uncontrolled pain in the distal extremities. Complications, such as digital ulcers, are extremely rare in primary RS. With secondary RS, digital ulcers, although more common than in primary RS, remain rare. Most complications associated with secondary RS are related to the underlying conditions causing the secondary symptoms.

Treatment

The primary goals in the treatment of RS are to decrease the frequency and intensity of attacks, and to decrease the duration of attack episodes, It bears mentioning that should secondary RS (ie, Raynaud’s phenomenon) be present, the underlying disease cause should also be treated. Connective tissue disorders associated with secondary RS are frequently managed with immunosuppressive or immunomodulating agents. These agents independently have not been shown to treat RS symptoms, so concurrent therapy with vasodilatory agents is often appropriate.

A. Conservative Measures

Conservative treatment is the mainstay of RS therapy initially, with the primary goal being prevention of episodes by maintaining warmth, limiting cold exposure, and minimizing avoidable vasoconstriction. Dressing warmly, wearing hats and mittens (rather than gloves), employing heat conservation techniques, avoiding unnecessary cold exposure, and possibly moving to a warmer climate may substantially improve symptoms. RS patients should avoid medications, including nicotine, that induce vasoconstriction and thereby initiate an attack. For patients who do not respond to conservative measures, whose symptoms limit their ability to carry out activities of daily life, or those at risk for skin ischemia, additional pharmacologic, surgical, and behavioral treatments are available.
B. Pharmacotherapy

Several pharmacologic and invasive therapies for the treatment of RS symptoms have been studied; however, no specific treatments are currently approved by the U.S. FDA specifically for RS because no medical therapy has yet proved to be universally beneficial. As the cause of RS remains unknown (and therefore treatment cannot be targeted), nonconservative treatment has been directed at minimizing symptoms by inducing general vasodilation. Several vasodilatory medications have proved beneficial for this purpose in RS treatment.

Among this group, calcium channel blockers are the most studied agents and the first-line choice in pharmacologic treatment. Dihydropyridines, such as nifedipine or amlodipine, have been shown to be more effective than nondihydropyridines at reducing both frequency and severity of vasospastic attacks because the dihydropyridine class is more potent to vasculature. However, this vasopotency also ensures that dihydropyridines are associated with more side effects than the nondihydropyridine class. Diltiazem, a nondihydropyridine, has some therapeutic benefit, but it is minimal when compared with nifedipine, currently considered to be the most effective agent. Amlodipine may hold more potential in RS treatment than nifedipine as its half-life is longer and use is less often associated with side effects.

$\alpha_1$ Blockers, such as prazosin, have been shown to decrease the RS burden by one to two fewer attacks per day, as well as decrease the duration of attack episodes, and these drugs have modest benefit in secondary RS. Long-acting formulations of doxazosin and terazosin may also be of benefit in reducing the frequency and severity of attacks but have yet to be systematically studied.

Medications that interact with the renin–angiotensin system have undergone trials as possible agents for treatment of RS symptoms. Angiotensin-converting enzyme inhibitors, such as captopril, improve blood flow in the distal extremities during RS attacks but have not shown efficacy in decreasing the frequency or severity of episodic attacks. Angiotensin II receptor blockers, such as losartan, decrease both the frequency and severity of attacks and, therefore, have greater utility in RS. Promising data suggest that losartan is more efficacious than nifedipine, in terms of both therapeutic benefit and side effect profile, and studies are ongoing.

The phosphodiesterase inhibitor class has recently been shown to be beneficial in the treatment of secondary RS. Inhibition of phosphodiesterase leads to a direct increase in cyclic-GMP, which results in vasodilation. The strongest evidence thus far is for sildenafil, which was associated with a decrease in the frequency and intensity of secondary RS attacks. Additionally,
sildenafil proved therapeutic in ulcer healing—a unique characteristic that nifedipine, for example, lacks. No evidence yet supports sildenafil in primary RS therapy. Because long-acting formulations are preferred in RS therapy, tadalafil and vardenafil are being studied to determine their utility in treatment. Currently, it appears that these agents are best used in conjunction with other therapies rather than as monotherapy. The phosphodiesterase inhibitor class, overall, has growing clinical relevance.

Nitrates have utility in the treatment of RS; however, because of the frequency of side effects they are not first-line therapies. Symptomatically they can be used to alleviate periacute ischemic attacks by applying a topical formulation on the affected digits. Oral and intravenous preparations can be used also. Topical formulations have not been shown to decrease episode frequency.

Endothelin is a potent vasoconstrictor and bosentan is an endothelin receptor antagonist that have been shown to decrease the incidence of new digital ulcers in scleroderma patients. Bosentan did not decrease healing time of ulceration or pain, and it has known side effects on liver function, which limit its utility.

Prostacyclins are vasodilators that have therapeutic benefit in the treatment of ischemia from fixed occlusive disease. Intravenous iloprost, a prostacyclin, has been shown to reduce the frequency, intensity, and duration of RS attacks; it has also shown efficacy in decreasing healing time for ischemic ulcers. However, the oral form of the drug has shown no benefit.

**C. Invasive Modalities**

Should pharmacotherapy fail, invasive and surgical therapy modalities are the next progressive step in treatment. As these techniques are relatively unstudied and results to date have been variable, they are most often implemented as last-resort therapies in severely refractive patients.

The use of percutaneous sympathetic nerve blocks has been shown to be effective in patients with refractory RS. Several agents used for this purpose have been studied. Injection of mepivacaine or bupivacaine has been shown to help relieve symptoms and facilitate ulcer healing. Botulinum toxin, injected into the digits targeting the digital neurovascular bundles controlling local vasospastic activity, has also been shown in preliminary studies to produce symptomatic relief in patients with refractory RS.

Open cervicothoracic sympathectomy, a surgical procedure initially proposed to denervate the sympathetic response to the distal extremities, has been largely replaced by thorascopic sympathectomy in appropriate candidates with secondary RS. Thorascopic sympathectomy is not indicated in primary RS, as
recurrence rates are high post-procedure. However, the procedure can produce significant alleviation of symptoms in some patients with severely refractory secondary RS complicated by ulceration or pain.

Distal vascular adventitial stripping of the hand and digital arteries has been shown to aid in ulcer healing and improvement of ischemic pain. However, this procedure has not been adequately studied and currently is used only in patients at significant risk of tissue necrosis or loss from ischemia.

Transcutaneous electrical nerve stimulation has also been used in some patients to induce vasodilation. Implanted spinal cord stimulators are being studied and appear to reduce pain and promote ulcer healing in patients with severe symptoms of secondary RS. Studies are ongoing.

D. Alternative Therapies

In addition to the conservative, pharmacologic, and invasive or surgical treatment options for RS outlined above, alternative therapy modalities have been studied. These include temperature biofeedback, acupuncture, and laser therapy. Temperature biofeedback, a technique in which patients are taught methods of skin temperature self-regulation, has resulted in variable outcomes. When patients are able to learn the techniques, the results appear promising; however, several studies have noted problems with regard to inadequate learning of the techniques. Acupuncture is another modality that may have relevance; a small study has shown improvement in frequency and severity of episodic attacks. The mechanism suggested is local nerve simulation and vasodilation from needle-induced chemokine release.

Prognosis

In general, primary RS (Raynaud’s disease) is milder and easier to treat, usually responding to conservative measures, whereas the vastly more variable secondary RS (Raynaud’s phenomenon) is more difficult to control.


Successful diagnosis and treatment of musculoskeletal dys-function requires an understanding of both basic anatomy and the dynamic relationship of anatomic structures to one another known as functional anatomy. Biomechanics is the study of the physical actions of forces or mechanics applied to a biologic system and the implications this has on anatomic and functional relationships. The forces can be divided into static and dynamic types. Static biomechanics involve a physiologic system in which the forces result in a state of equilibrium with a net of zero change in the system velocity. In this case the body can remain at rest or in motion; however, a constant velocity persists due to the balance of forces. Dynamic forces result in net acceleration of the physiologic system due to the unbalanced application of forces.

Kinetics is the study of motion; this is a general term with implications across many sciences. Human kinetics is the study of motion of the human body with a focus on the forces that produce motion. The structure and stability of each extremity reflect the forces imparted and, ultimately, the functional demands placed on the limb. The functional demands of the upper limb are vastly different than those of the lower limb, and the forces imparted are therefore different. The amount of movement versus amount of stability necessary for function at a joint governs its size, shape, and infrastructure. This difference is clearly seen when the hip joint is compared with the glenohumeral joint. Kinematics is a branch of science that describes the motion of points, objects, or systems without consideration of the cause of this motion, and can
also be described as the geometry of motion. This includes consideration of time, displacement, velocity, acceleration, and space factors of a system’s motion.

Two types of motion occur about most joints to varying degrees based on function: translation (linear displacement) and rotation (angular displacement). These two movements occur within the three orthogonal planes: the coronal, sagittal, and transverse. Although complex motion occurs across many planes simultaneously, the motion can be subdivided into the aforementioned planes with the use of image capture and computer-aided motion analysis. When analyzing motion that is occurring in any one plane, the structure that serves as or creates that axis of rotation, perpendicular to the major plane of motion, must be determined. For example, motion in the coronal plane occurs about an axis of rotation that is in the sagittal plane. If one understands the structures that determine the axis of rotation, these can then be examined for pathologic disease processes or function.

Kinesiology requires an understanding of functional anatomy and combines the sciences of kinetics and kinematics as well as biomechanics, anatomy, and physiology in the study of movement. It is through a fundamental understanding of kinesiology and implementation of its principles that one can recognize normal and abnormal function and use this information to optimize patient recovery, performance, and avoidance of injury.

Having a framework within which to interpret abnormal function is vital to the success of a rehabilitation physician. One such system has been proposed by Dr. Gerald Herbison of Thomas Jefferson University Hospital in Philadelphia. Aberrant motion is examined and then classified as being due to pain, paralysis or paresis, or contracture. Pain alone, through increased afferent input, can inhibit muscle contraction. Without muscle contraction, there is decreased joint stability and joint motion. Paralysis or weakness clearly can affect joint motion through the lack of either concentric or eccentric muscle contraction, but also due to improper joint positioning for optimal motion, and possible joint instability due to the lack of necessary agonist–antagonist muscle function across a joint. Finally, joint contracture, physically impeded motion, can occur with either normal or abnormal soft tissue physiologic findings. If these factors are considered in the analysis of motion, the cause of the abnormality, and ultimately a diagnosis, can be determined within a clinical context.

FUNCTIONAL MACHINERY

Lever Arm
Within the human body, the musculoskeletal system uses three main types of mechanics to produce motion: levers, wheel-axles, and pulleys. A lever uses a rigid bar that turns about an axis of rotation, or fulcrum. In the human body bones function as the levers, joints compose the axes of rotation, and muscles produce force that cause motion. All lever systems contain these three components; however, the arrangement of these components may differ across different types of joints involved in different types of movements (Figure 4–1). The force or effort is due to contraction of a muscle and is usually represented by the insertion of the muscle on a bone. The resistance can be the center of gravity of the lever or the location of the application of some external resistance. In a type 1 lever arm (FIGURE 4–2A), the fulcrum is located between the resistance and the force; an example of this is elbow extension with the arm in an overhead position. The triceps inserts on the olecranon, the elbow joint is distal to this point, and the resistance force is the center of gravity of the forearm. In type 2 lever arms (FIGURE 4–2B), the resistance is located between the fulcrum and the force. There are few physiologic examples of this type of lever arm, but a common example is a heel raise. In a heel raise the axis of rotation is located distally at the metatarsophalangeal joint, the resistance is the weight of the body between the axis and the applied force of contraction of the gastrocnemius–soleus complex proximally at the calcaneus. Finally, in a type 3 lever arm (FIGURE 4–2C), the force is located between the fulcrum and the resistance. This is the most common type of lever arm. Examples of a type 3 lever arm are the brachialis muscle and the iliopsoas muscles. Here the axis of rotation is proximal, the resistance force at the center of mass of the limb is distal, and the contracting muscle force is at some point between the two.
Figure 4–1 Lever arms. **A.** Long lever of the latissimus dorsi and the effect of its pull on the position of the scapula. **B.** Short lever of the lower trapezius fibers and the effect of its pull on the position of the scapula.
Figure 4–2 Lever arms. A. Type 1 lever arm. B. Type 2 lever arm. The resistance is the center of mass, and this falls between the force of the contraction of the muscles acting in plantar flexion and the fulcrum, which is the metatarsal-phalangeal joint. C. Type 3 lever arm. The resistance is the weight of the forearm and any weight carried in the hand. The applied force is due to contraction of the elbow flexor complex where the insertion of these muscles on the proximal portion of the radius and ulna are distal to the axis of rotation of the elbow joint. This is the most common physiologic lever arm.

The mechanical advantage (Figure 4–3) of a system can be calculated by first calculating the workload of the system and then calculating the differential between the workload of the internal moment arm (muscle force) and the external moment arm (load), where the length of the force arm (df) is divided by the length of the resistance arm (dr) over which a force is applied. The workload can then be calculated as $(F \times df) / (R \times dr)$, where $F$ and $R$ are the magnitude of the force of the applied force and the resistance, respectively. A mechanical advantage ratio greater than 1 favors the internal moment arm. In a type 2 system (Figure 4–4), panels A and B), the internal moment arm is always larger than the external moment arm, thereby creating a condition in which less force may be needed to overcome a very large external force. This is why it is difficult to test the gastrocnemius–soleus system with the patient either sitting or lying prone. The external force applied by the examiner will be far less than that produced during weight bearing; therefore, even if weakness is present, it is unlikely to be noted due to the mechanical advantage maintained by the plantar flexors as a type 2 lever arm. Type 2 lever arms always have a mechanical advantage ratio greater than 1, making these systems very efficient (see Clinical Correlation 4–1). In a type 3 system (Figure 4–3, panels C and D), the most common type physiologically, the internal lever arm is always shorter than the external lever arm, and a greater internal force is always necessary to overcome the external force. This type of system produces relatively little mechanical advantage, with the ratio being less than 1.
Figure 4–3 Mechanical advantage. Relationship of length of moment arm (distance A–D) and muscle contraction (represented by size of green arrow). There is increasing distance from insertion point A through D, which results in an increase in the length of the lever arm of the contracting muscle. The force of muscle contraction produces angular displacement of the distal limb; this force is called torque. The force of torque is $t = F \times d$, where $F$ is the magnitude of muscle contraction (in Newtons) and $d$ is the length of the lever arm. There is an inverse relationship between muscle contraction ($F$) and lever arm length ($d$), whereby to produce the same amount of angular displacement or torque, with increased distance, less muscle contraction force is required to balance the externally applied resistance. With muscle contraction force held constant, mechanical advantage refers to the amount of work required to move an external resistance. Following the same principle, with increased lever arm or, in this case, moment arm, less work is required to displace the same external load that is proportional to the length of the moment arm. Therefore, systems with longer lever arms (insertion point D) will have a mechanical advantage (yellow-shaded area) over those with shorter lever arms (point A), if the magnitude of force of muscle contraction (green arrows) is proportionally smaller compared to the length of the moment arm. For instance, at insertion point A, the force of muscle contraction must be equal to the 10 lb of external load, whereas at point D, the force of muscle contraction is one quarter the force at A, due to the increased length of the moment arm. The mechanical advantage occurs due to the
decreased stress and work incurred by the contracting muscle with longer moment arms.

**Figure 4–4** Comparison of the mechanical advantage of different types of lever arms (A–D). **A.** The relationship of the vector of the force produced by muscle contraction versus resistance of externally applied force and the fulcrum for rotation or angular displacement in a type 2 lever arm. **B.** The mechanical advantage (shaded area) of this system, a type 2 lever arm system, due to the larger moment arm (distance of arrow from fulcrum) of the contracting muscle’s long lever arm compared to the lever arm of the external force. This type of
system has a mechanical advantage ratio > 1. This is produced by the moment arm of the muscle force \((F \times df)/(R \times dr)\). Thus, with a force equal to resistance, there is a mechanical advantage > 1 due to the longer moment arm from the muscle contraction based on the orientation of its insertion. During a single-leg stance, in which the external load that must be overcome by the active muscles is usually approximately 1.5–2 times body weight, this muscle group is capable of producing forces of contraction likely greater than 200–400 kg. Clearly, an externally applied force magnitudes smaller, produced by the force of one upper limb during muscle testing, will not be able to overcome this muscle unless significant weakness is present. C. The relationship of the vector of the force produced by muscle contraction versus resistance of externally applied force and the fulcrum for rotation or angular displacement in a type 3 lever arm. D. The mechanical advantage of this system, a type 3 lever arm system, is due to the larger moment arm of the external resistance’s long lever arm compared to the lever arm of the muscle force. There is a mechanical advantage ratio < 1, as demonstrated by \((F \times df)/(R \times dr)\). To overcome the mechanical advantage of the external force in this type of system, the magnitude increase of muscle force of contraction must be proportional to the length advantage of the external load. This is shown by the proportionally larger muscle force arrow (demonstrated by the grey force arrow).

**CLINICAL CORRELATION 4–1: Weakness of the Ankle Plantar Flexors**

The ankle plantar flexors exemplify a type 2 lever system. As such, the muscle and the muscle lever arm always maintain a mechanical advantage over the external force and load lever arm. This is why it is difficult to determine the presence of weakness in the ankle plantar flexors when the examiner grasps the foot with his or her hands and applies an opposite force. The most accurate means of assessing weakness in this muscle group is to engage the muscle in a more physiologic activity, such as heel raises, where a much larger force than the weight of the person can be added.

**Wheel & Axle**

The wheel-and-axle relationship has many physiologic correlates and has
implications for angular acceleration or torque, and magnitude of movement. In most systems, the wheel radius is larger than the axle radius. This relationship holds for the force arm; therefore, the wheel will have a mechanical advantage over the axle. For this reason, a smaller force may be applied to the wheel to move a relatively larger resistance applied to the axle. If the force is reversed and so applied to the axle, the wheel will spin faster than the axis and travel a greater distance because of the same mechanical advantage relationship.

An example of such a system is the humerus and forearm. The humerus serves as an axis of rotation. The humeral external rotators contract a small amount but produce up to 90 degrees of external rotation. The distance that the forearm and hand will travel is far greater than the length of muscle shortening between the muscle origin and insertion at the axis of rotation (axle). Similarly, with internal rotation, as in the acceleration phase of pitching, joint displacement of the glenohumeral joint would likely be deleterious in light of the ultimate speed with which the ball leaves a pitcher’s hand. However, as described, due to the distance relationship, the speed of the humeral internal rotation is reduced by a magnitude proportional to the length of the wheel radius.

### Pulley

Although less numerous than the other mechanical systems, pulley systems exist in the human body and serve to change the effective direction of an applied force. This decreases work and makes movement of an object easier. Pulleys can be singular or combined in a system in which the mechanical advantage of one pulley is equal to 1, and the addition of each new pulley increases the mechanical advantage by a factor of 1. A physiologic example of a pulley system is seen in the peroneus longus and brevis muscles, which arise from the proximal and middle third of the fibula and insert on the base of the first and fifth metatarsals, respectively. Over their course, they run posterior to the distal fibula. Without traversing this course, the insertion of the peroneus longus muscle would be on the base of the first metatarsal and the muscle would then function more completely as an ankle dorsiflexor. The distal fibula functions to create a pulley system, in which a change in the force vector is created by the peroneus muscle complex. In this case the forces applied to the respective metatarsal heads now serve to plantar flex and evert the foot (Figure 4–5).
**Figure 4–5** Force vectors of the peroneus muscle complex. **A.** The peroneus complex force vector would result in dorsiflexion and eversion, absent the pulley effect created by the lateral malleolus. **B.** The peroneus complex force vector produces plantar flexion and eversion due to the pulley created by its course posterior to the lateral malleolus.


OPEN CHAIN & CLOSED CHAIN KINEMATICS: AN INTERACTION OF MUSCLE & JOINT

Muscle action describes motion in an open chain system or a closed chain system. In an open chain system, the distal link is free to move while the more proximal links are fixed. In a closed chain system the distal link is fixed and the proximal end is free to move. Steidler translated this system to human motion in the 1940s, proposing that each link in the chain is a limb and the interaction of two links is similar to a joint. Most physiologic motion involves a closed chain system, because the forces applied to the distal limb are usually weight bearing. Weight bearing imparts an axial load through a joint which, in combination with the ground reactive force, creates a fixed limb. In contrast, in an open chain system, the joint forces at the distal segment are usually perpendicular, producing angular acceleration at the joint and not compression. Once the distal limb is engaged in a closed system, the motion at any one joint in the limb will affect all other joints in the system; this is in direct opposition to open chain kinematics, where all joint motion is independent of motion at the other joints in the limb. Most muscles, especially during the gait cycle, function in a closed chain system and in a manner that may be counterintuitive to their normal open chain activity. For example, the gluteus maximus normally functions as an external rotator and an extensor of the hip; however, from initial contact through the loading phase of gait it is eccentrically active and functions as a knee extensor. During this period, the function of the muscle is to counteract the ground reaction force, which falls behind the knee and produces a knee flexion moment (Figure 4–6). If there is fatigue or weakness of the quadriceps, this may lead to knee buckling; however, the function of this muscle can help to reduce this effect.
Figure 4–6 Function of the gluteus maximus during gait. Gluteus maximus function increases from initial contact (left image) to a maximal electromyographic function at midstance (right image). The gluteus maximus initially functions to oppose the ground reaction force (GRF), which at contact is anterior to the hip joint and would produce hip flexion. With increasing weight bearing, the GRF continues to move and at midstance is posterior to both the hip and the knee. This force at the knee would produce excessive knee flexion, which is opposed by the contraction of the gluteus maximus in the closed kinetic chain. In this type of system, because the distal limb is fixed, the force of contraction produced by the gluteus muscle causes motion at the proximal end. The net effect is knee extension.

Wheelchair transfers and propulsion in a tetraplegic patient provide an example of adaptive function in a closed system. In a tetraplegic patient with injury at the level of C6, normal triceps function should be lacking. However, most C6 tetraplegic patients are capable of producing elbow extension. The mechanism for this motion does not involve active triceps extension; rather, it involves substitution of muscles having intact innervation that can produce elbow extension in a closed chain system. In the transverse plane, a tetraplegic patient may lean forward excessively to activate the pectoralis and anterior deltoid muscles. In this plane of motion, both of these muscle groups can
produce passive elbow extension via adduction of the proximal humerus with the
distal limb fixed on a push rim. The end result is elbow extension. In the sagittal
plane, the serratus anterior and anterior deltoid will function as elbow extensors,
again by drawing the proximal humerus forward (relative flexion) while the
distal limb is fixed, producing net elbow extension. Finally, in the coronal plane,
activation of the latissimus dorsi may be able to adduct the proximal humerus
enough to produce elbow extension (Figure 4–7).
**Figure 4–7** Functional substitution. **A.** In the sagittal plane the anterior deltoid functions as an elbow extensor. **B.** In the transverse plane the pectoralis major functions as an elbow extensor. **C.** In the coronal plane the latissimus dorsi functions as an elbow extensor.

**KINESIOLOGY OF THE SHOULD JOINT**

Inman in 1954 described in detail overhead motion, and the contribution of the individual joints to this motion. In this description, he used the term *scapulohumeral rhythm* to describe the relationship between motion at the scapulothoracic joint and the glenohumeral joint. There is a predictable relationship of motion between these two joints through the full range of overhead motion, whereby for every 2 degrees of glenohumeral motion, there is 1 degree of scapulothoracic motion. Therefore, maximal overhead motion is attained via 120 degrees of glenohumeral motion and 60 degrees of scapulothoracic motion. From this finding it is evident that the motions of these two joints are interdependent; however, scapulothoracic joint motion is also dependent on maximal motion of the clavicle at the sternoclavicular and acromioclavicular joints. Further, the motion at the scapulothoracic joint is vital to maintaining the muscle forces necessary for producing overhead motion.

The motion of the scapula along the thoracic wall is made possible due to the contribution of the two joints involving the clavicle, the sternoclavicular and acromioclavicular joints. The clavicle is capable of elevation, depression, protraction, and retraction at the sternoclavicular joint. Elevation and depression of the scapula mimic motion at the scapula; maximal elevation has been measured at 30–45 degrees and maximal depression at 10 degrees in the coronal plane. This motion generally occurs by 90 degrees of arm elevation. Protraction and retraction at the sternoclavicular joint have been measured to be approximately 15–30 degrees. The clavicle is also capable of posterior rotation in the sternoclavicular joint up to 35 degrees (where the inferior aspect of the clavicle moves via rotation anteriorly and superiorly). The sum total of joint motion at the sternoclavicular joint is at least 30 degrees of elevation and approximately 30–40 degrees of posterior rotation. This motion directly contributes to scapulothoracic motion. The clavicular motion at the acromioclavicular joint occurs both early (between 0 and 30 degrees) and late (after 130 degrees of arm elevation), where a total of approximately 20–40 degrees of additional scapular elevation occurs. Posterior clavicular rotation allows for the amount of acromioclavicular joint motion that occurs.
Shoulder Joint Motion

At the glenohumeral joint, motion in the sagittal plane includes flexion and extension; in the coronal plane glenohumeral motion includes abduction and adduction. Internal and external glenohumeral rotation can occur in multiple planes. At the scapulothoracic joint, the description of motion can be confusing owing to the number of terms across disciplines describing similar motion. Active motion of the scapula toward the midline is called retraction. Active motion away from the midline is called protraction. This is different than passive movement of the scapula away from the midline, which will be referred to here as lateral excursion. The scapula can move in a cephalad direction along the thoracic wall, called elevation, or move caudally, called depression. Finally, the scapula can rotate up or down in the coronal, sagittal, or scapular plane. During scapular upward rotation in the coronal plane, the inferior angle moves more laterally, and the superior angle more medially, while there is net elevation of the glenoid. Similarly, during the same motion in the sagittal plane, the inferior angle moves more anteriorly, and the superior angle more posteriorly, and there is a net elevation of the glenoid.

Shoulder Stability

The stability of the shoulder joint, namely the glenohumeral joint, is influenced by several factors. The glenoid surface area is approximately one third that of the humeral head and even with the increased area conferred by the labrum is still smaller than the humeral head. Instability, if it does occur, usually happens in the anterior plane, followed less frequently by inferior instability, and least frequently it occurs in the posterior direction.

The static position of the scapula maximizes bony articulation with the humerus due to the positioning of the scapula approximately 30 degrees anterior to the coronal plane coupled with retroversion of the humeral head along the humeral shaft of similar angulation. In addition, there is coupling of the posterior tilt of the glenoid fossa, together with the posteriorly tilting humeral head, providing a relationship that also counteracts the tendency toward anterior instability. An approximately 5-degree superior tilt of the glenoid in the coronal plane statically also counteracts downward displacement. Lateral stability at the glenohumeral joint is provided by the superior glenohumeral ligament and coracohumeral ligaments. During arm elevation, other structures are responsible for stability, especially the interior structures. In the middle range of abduction,
support is through the subscapularis muscle, the middle glenohumeral ligament, and the superior band of the inferior glenohumeral ligament. In the upper ranges of elevation, the axillary pouch of the inferior glenohumeral ligament provides the majority of stabilization.

▶ Forces at the Shoulder

At any point in time and in any position of the arm, three forces act on the glenohumeral joint: the weight of the limb, the force generated by the active muscle pull during abduction above the center of rotation (deltoid and supraspinatus), and a combination of friction and pressure at the glenohumeral joint and the muscles opposing the force of abduction below the center of rotation. The abducting muscle forces examined across the full range of overhead motion produce a sine wave, with maximal force encountered at about 90 degrees of elevation, in which the magnitude is approximately eight times the weight of the extremity. However, the resultant force (friction plus depressor muscle pull) is larger in magnitude than the abductor force, approximately ten times the weight of the extremity. This discrepancy in force is necessary to prevent excessive humeral translation or subluxation, which could lead to clinical signs of impingement. The force of the glenohumeral muscles is also counterbalanced by the scapulothoracic muscles, specifically the trapezius and serratus. Motion via the deltoid and supraspinatus can also produce unwanted motion at the scapula. Specifically, unopposed motion of this muscle group without concomitant muscle activity of the scapulothoracic muscles would result in instability of the scapula, and abnormal shoulder range of motion.

▶ Muscle Activity Across the Shoulder

A. Humeral Movements

The deltoid and supraspinatus muscles function in shoulder abduction, and while both contribute to the full range of overhead motion, individual contribution differs depending on arm position. The deltoid muscle is most active after 90 degrees of overhead motion. Comparison of amplitude of electromyographic activity has shown that the muscle is slightly more active during flexion than abduction. The pectoralis major also contributes to overhead motion; however, not all fibers are active, and it is not at all active during abduction motion. In flexion, the clavicular head activity is maximal near 90 degrees, and again
around 120 degrees, where it is likely working in concert with the anterior portion of the deltoid muscle. The supraspinatus muscle is also maximally active near 80–90 degrees of overhead motion, with maximal flexion activity occurring before abduction.

The rotator cuff muscles—specifically, the supraspinatus, infraspinatus, and teres minor—are usually thought of in the context of their role in humeral external rotation. However, the infraspinatus and teres minor along with the subscapularis muscle act to depress the humerus during overhead arm motion. Humeral compression and depression ensure that the larger humeral head can slide and rotate along the smaller glenoid surface. The sliding and rotatory motion of these muscles is the result of the downward force vector produced, which opposes the upward force vector produced by the humeral abductor group. This maintains smooth rotation and prevents superior subluxation and excessive translation of the humeral head within the glenoid. Over the full range of overhead motion, activity of the supraspinatus muscle increases linearly. Activity curves for abduction and flexion of the teres minor are similar, and occur around 100–120 degrees of motion. The subscapularis shows maximal activity around 90 degrees and maintains a plateau of activity through 130–140 degrees of motion. Analysis of the combined activity of this group demonstrates two peaks of electromyographic activity, at 80–90 degrees and again at 110–120 degrees. The first peak likely represents the function of the group to provide a downward force vector in opposition to the force produced by the deltoid and supraspinatus, and the second peak, the activity of the muscles in providing adequate rotatory and sliding of the humeral head to achieve full range of motion.

B. Scapulothoracic Movements

The scapulothoracic muscle group is composed of the rhomboids, levator scapula, latissimus dorsi, trapezius, pectoralis minor, and serratus anterior muscles. The scapulothoracic muscles provide scapular stabilization in response to the pull of the muscles not originating from the scapula (humeral abductors and depressors), provide static positioning of the scapula, rotate the scapula in response to functional demands, and maintain optimal muscle-tension relationships of the scapulohumeral muscles during overhead motion.

Activation of various scapulothoracic muscles during overhead motion produces the following actions at the scapula: elevation–depression, protraction–retraction, and rotation up–down in the coronal–sagittal plane. Knowledge of the function of the individual muscles is necessary before one can understand the complex motion of the muscle group. Further, one must consider the origin–
insertion of the muscles and the vectors of their muscle pull in relation to the various axes of motion about the scapular joints. The axis of rotation for upward and downward rotation occurs along the scapular spine approximately one third of the distance from the vertebral border, where the scapular spine in the static position is at about the level of T3 or T4, and the axis of rotation for internal and external rotation occurs about the acromioclavicular joint. The rhomboids originate from the midline of the C7–T5 spinous processes and insert medially at the vertebral border of the scapula. Therefore, while the insertion of the muscle is below the axis of rotation, the majority of the force vector produced by these muscles is cephalad and medial, and the pull is from above the axis of rotation. The rhomboids cause scapular retraction, elevation, and downward rotation in the coronal plane. The latissimus dorsi has both short fibers originating from the T6 area and long fibers from the thoracodorsal fascia that insert on the inferior angle of the scapula. The insertion of these fibers is medial and below the axis of rotation of the scapula. The resultant motion produced by the latissimus acts to depress, retract, and rotate the scapula downward in the coronal plane. The levator scapula originates from the C1–C4 vertebrae and inserts at the superior medial border of the scapula. The levator elevates and retracts the scapula. However, due to the orientation of its fiber pull, superiorly and medially, and the insertion above and medial to the axis of rotation, the levator causes downward scapular rotation. The trapezius has an upper, middle, and lower division. All of the fibers of the trapezius will retract the scapula. The upper fibers insert along the clavicle and acromion, at a point that is lateral and above the axis of scapular rotation. The upper trapezius produces scapular elevation and upward rotation in the coronal plane. The middle trapezius fibers insert along the lateral portion of the scapular spine, where the insertion is again lateral and above the axis of rotation, thereby producing scapular retraction and upward rotation. Finally, the lower trapezius fibers insert medially on the spine of the scapula, where the force of pull is below and medial to the axis of rotation. This produces scapular depression and yet continues to rotate the glenoid upward in the coronal plane. The serratus anterior arises from the costicartilaginous portion of the first eight ribs and runs posteromedially along the thoracic wall to insert along the ventral portion of the scapula along the vertebral border. The serratus produces protraction of the scapula and also rotates the glenoid up in the sagittal plane.

During all overhead motion there is also relative motion of the scapula, such that its resultant end position compared with the static position produces a net motion, with increases in the posterior tilting, upward rotation, and external rotation of the scapula. It is important to remember that the humerus must externally rotate during overhead motion to avoid abutting the tubercle structures
under the acromion; the magnitude of external rotation is anywhere from 10 to 50 degrees. The scapula, to accommodate this motion and to maintain optimal glenohumeral positioning, rotates externally up to 2 degrees, rotates upward in either the coronal or sagittal plane up to 40 degrees, and tilts posteriorly up to 20 degrees. This activity is produced by the coordinated muscle activity of the scapulothoracic musculature in response to forces generated at the humerus and scapula by the scapulothoracic musculature (see Clinical Correlation 4–2).

**KINESIOLOGY OF THE ELBOW JOINT**

The elbow joint has multiple functions based on activity; it serves as a fulcrum during lifting, as a weight-bearing joint during transfers, and in coordination with the shoulder joint and wrist joint is involved in performing dexterous movements with the hand and fingers. Pathologic changes in the elbow joint thus have important implications related to these aforementioned functions and can precipitate overuse of the wrist and shoulder joints.

**Elbow Joint Motion**

The elbow joint is capable of flexion and extension as well as pronation and supination. Normal range of motion for flexion is up to 140–160 degrees, and between 170 and 180 degrees in extension. In pronation and supination 70–80 degrees of motion is possible. The orientation of the humeroulnar joint produces a slight valgus position of the forearm, known as the carrying angle, formed by the angle of intersection of axial lines through the humerus and ulna. In women, a normal carrying angle is between 10 and 15 degrees, and in men, between 5 and 10 degrees.

Joint stability is provided by the joint capsule, radial and ulnar collateral ligaments, and by the bony articulations about the elbow joint. With valgus strain, the majority of stability is provided by the flexor muscles and the medial, or ulnar, collateral ligament. This ligament has three portions, with the anterior being the strongest; however, with increasing flexion the posterior and oblique fibers contribute increasingly to joint stability. With varus strain, stability is provided by the radial collateral ligament. In addition, the elbow joint is most stable in extension owing to the articulation of the ulna and posterior humerus. With full extension, the olecranon of the ulna slides into the deep recess of the olecranon fossa on the posterior humerus, providing additional elbow stability.
Loss of overhead motion can result from innumerable causes, including trauma, degenerative joint changes, and overuse. However, if this result is considered in the context of pain, paralysis, and contracture, the evaluation, analysis, and subsequent treatment is clarified. In the evaluation of decreased overhead motion, history and clinical context are paramount in forming a differential diagnosis. As noted in the text discussion, the scapular rotator group should also be considered as a potential cause of weakness. Suppose a construction worker were to develop long thoracic nerve palsy. This would be very detrimental, impairing his ability to continue work. Weakness of the serratus anterior muscle would affect all overhead motion. On examination, there would be weakness of the rotator cuff musculature, biceps, coracobrachialis, and pectoralis muscles. Technically this would be termed pseudo-weakness, caused not by intrinsic muscle pathology, but rather by destabilization of the origin point of the various muscles, inability to maintain optimal scapular position, and loss of the optimal length–tension relationship during muscle activation. It would be very easy to mistake this serratus weakness for other causes, such as rotator cuff impingement, rotator cuff tear, and even, potentially, a radiculopathy.

The mechanism whereby serratus weakness can present as extremity weakness reflects the role of this muscle in scapular rotation. Recall that the serratus rotates the scapula upward in the sagittal plane. The serratus, therefore, functions with all motion that occurs in the sagittal plane. Both elbow and shoulder flexion, as well as internal and external rotation, can be tested in the sagittal plane. The biceps, brachialis, and rotator cuff muscles all originate on the scapula and insert at points along the humerus. With activation of these muscles, two forces are produced at the glenohumeral joint: the force due to their muscle pull, and that due to the weight of the arm. The net of these forces across the glenohumeral joint causes the scapula to rotate downward. If serratus weakness results from denervation, these forces will be unopposed by serratus rotation in the opposite direction. Despite normal physiologic findings, the muscles will appear weak. However, if the thoracoscapular muscle group is considered in the evaluation of all patients with extremity muscle weakness, missing this as a source of weakness can be avoided (Figure 4–8).
Weakness of the serratus anterior muscle can cause destabilization of the scapula and will therefore affect all proximal upper limb muscles due to their scapular attachment. Muscle testing may reveal weakness of shoulder or elbow flexion due to weakness of the serratus muscle. During applied extension force at either the elbow or shoulder, the scapula is rotated downward in the sagittal plane or derotated. This may produce quasi-weakness of the tested muscle due to a “destabilizing effect.”

**Forces at the Elbow**

The muscles that cross the elbow joint and insert onto the proximal aspect of the radius and ulna function in a type 2 lever arm system (see **FIGURE 4–2B**). As with all type 2 levers, the muscles have relatively short lever arms, and because of this the active muscle must generate a very large amount of force to overcome the applied load on the distal forearm or hand. A muscle that inserts in close proximity to the joint, and thus the axis of rotation, is able to generate a very large amount of rotational torque, and thus rotational excursion, in the form of larger joint range of motion.

Maximal force of elbow flexion is generated between 90 and 100 degrees. From the fully extended position to about 30 degrees of flexion, only about 30% of the maximal flexion force can be generated. The function of the joint as a
weight-bearing joint is apparent when examining the intraarticular pressure during heavy lifting, which can be in excess of three times body weight. Maximal extension forces are achieved between 0 and 30 degrees of flexion. Interestingly, maximal isometric force of extension is about 40% less than maximal flexion forces.

**KINESIOLOGY OF THE WRIST & HAND**

The act of making a fist or bringing the thumb and index finger together exhibits the complex interaction of muscle and joints in the wrist and hand. Grasp is the act of finger flexion used to maintain the fixed position of an object in the hand, most commonly against the palm in power grasp, or between the fingers in precision grasp.

Power grasp is driven by the long finger flexors, specifically by the flexor digitorum profundus. The flexor digitorum superficialis is also involved, but only during resisted finger flexion. The majority of power is derived from the fourth and fifth digits, as well as the thumb, and control of the object in the palm is aided by the first, second, and third digits. There are three types of power grip and analysis of these demonstrates the complexity of grip control. Cylindrical grip occurs when the fingers are flexed and the thumb is maintained in a flexed and adducted position in contact with the second and third fingers. The major muscle activity is in the flexor digitorum profundus, flexor pollicis longus, and the adductor pollicis. With greater grip demands, the flexor digitorum superficialis and the interossei become activated. Spherical grip is used for opening bottles. It is very similar to cylindrical grip in terms of patterns of flexor muscle activity; however, there is greater activity of the interossei because of the abducted position of the fingers at the metacarpophalangeal joints. The hook power grip, unlike the cylindrical and spherical grips, involves no direct thumb muscle contribution. This type of grip is induced by proximal interphalangeal joint flexion and is activated primarily via the flexor digitorum superficialis and profundus muscles.

The wrist functions to transmit grip forces, balance muscle forces during gripping, and position the fingers during grasp. Wrist position is vital to grip integrity due to its role in maintenance of an optimal length–tension relationship of the long flexor muscles.
The wrist is capable of flexion and extension, ulnar and radial deviation, and minimal amounts of pronation and supination. Maximal extension range is from 45 to 75 degrees and flexion range is from 65 to 80 degrees. Ulnar deviation range of motion is greater than radial deviation, with ulnar range between 30 and 45 degrees and radial range between 15 and 25 degrees. Despite the common conception of wrist motion in one degree of freedom, wrist motion actually occurs as part of a complex of motion. An example is the so-called dart-thrower motion, when the axis is on an oblique angle and the most common pattern of motion seen is dorsal radial extension to palmar ulnar flexion.

**Forces at the Wrist**

The forces at the wrist joint are due to the combination of carpal bone motion and the influence of carpal bone motion on the moment arm of the long wrist flexors and extensors. With radial deviation of the wrist, the scaphoid flexes; this motion increases the anterior–posterior diameter of the wrist. The flexion motion causes the distal portion of the scaphoid to move in a volar direction relative to its resting position in a nondeviated hand. The distal pole acts as a pulley to increase the moment arm of the flexor carpi radialis. With ulnar deviation, the scaphoid moves into an extension position. This motion produces a decrease in the anterior–posterior direction of the wrist. The scaphoid is displaced in a dorsal direction and creates a convex pulley on the dorsal wrist surface. The result is an increase in the moment arm of the extensor carpi radialis.

**KINESIOLOGY OF THE HIP**

The hip or femoroacetabular joint is the major weight-bearing joint in the human body and its bony structure derives its shape from the forces it bears. The acetabulum is located at the juncture of the three bones comprising the pelvis: the ilium, ischium, and pubis. It is oriented in an inferior and anterolateral direction to articulate with the femoral head. Increasing the surface area of articulation with the femoral head is the acetabular labrum, which is a fibrocartilaginous lip that surrounds the external rim of the acetabulum. The orientation of the acetabulum and femoral head provides a very stable articulation. The stability and large area of articulation are consistent with the substantial loads that must be tolerated across this joint (see Clinical Correlation 4–3).
Wolff’s law is a physiologic principle that states that the shape and density of bone are a result of the forces borne by that bone, whereby adaptation occurs via bone remodeling as a result of forces applied through both weight bearing and muscular pull. At birth, the angle between the femoral shaft and femoral neck is between 150 and 160 degrees and the amount of femoral torsion, or the angle between the transverse axes of the head and neck (fem-oral anteversion) compared with that of the transverse axis between the femoral condyles, is about 40 degrees. However, by adulthood, the angulation of the femoral head and neck is about 120–130 degrees and the femoral anteversion is usually less than 15 degrees. The changes in these angles develop over time in response to the pull of various muscle groups across the hip joint, for instance, the hip abductor group (gluteus medius, minimus, and tensor fascia lata) and the external rotators (gluteus maximus, piriformis, gemelli and quadratus). The effect of a lack of muscle pull is seen in children with spina bifida. In these children, one of the most common locations for neurologic injury due to spinal dysraphism occurs at the fourth lumbar segment. Below this level there is commonly muscle denervation, affecting much of the hip girdle musculature. The lack of muscle pull from the hip abductor group and the hip external rotators results in hip angulation closer to that seen early in development, as well as hypoplasia of the acetabulum due to lack of muscle pull and weight-bearing forces across the joint. Thus, these patients are at an increased risk for hip dislocation (Figure 4–9).
Normal femoral neck angulation in adulthood is between 120 and 130 degrees. If muscular force pull is inadequate, abnormally large angulation will occur, similar to that found closer to birth ($150–160$ degrees. Hypoplasia of the acetabulum may also result, with shallow depth and abnormal contours.

**Hip Joint Motion & Stability**

Stability of the hip derives from the bony architecture, cartilage, ligamentous structures, hip capsule, and the muscles crossing the hip joint. The acetabular articular cartilage is thickest at the superior aspect, consistent with the compressive forces produced during upright weight bearing. The hip capsule is one of the strongest and thickest joint capsules in the body. The ligamentum teres is a weak synovial attachment that runs from the acetabulum and transverse acetabular ligament to the fovea capitis on the femoral head. Extending from the anterior inferior iliac spine and the rim of the acetabulum to the intertrochanteric line is the iliofemoral ligament (also known as the Y ligament of Bigelow). The latter name reflects the fact that from its origin point, the ligament splits into two segments. The transverse segment runs parallel with the femoral neck and inserts close to and medial to the greater trochanter; the descending portion inserts
proximally on the femoral shaft on the medial side. The major function of the iliofemoral ligament is to resist excessive hip extension. Additional anterior stability is provided by the pubofemoral ligament, which arises from the pubic ramus. Its fibers blend laterally with the iliofemoral ligament. Finally, additional support to resist excessive extension and posterior translation is provided by the ischiofemoral or capsulofemoral ligament. This ligament runs from the ischium, posterior to the acetabulum, and then inserts along the intertrochanteric line.

Maximal hip flexion is approximately 130–140 degrees, and maximal extension is 30–45 degrees. Further motion is possible, however, due to mobility of the pelvis (pelvic tilting and innominate rotation) as well as lumbar spine flexion and extension. Hip external rotation usually averages about 45 degrees, and internal rotation is usually slight less, at 30–40 degrees. Hip internal rotation is usually the first motion that is reduced secondary to arthritic change. Hip abduction occurs to about 45 degrees, and adduction across the midline occurs to between 15 and 30 degrees.

 Forces at the Hip

Estimates of hip forces vary, owing to differences in methods of measurement, activity, level of exertion, and comorbidities with deviation from normal gait patterns. Most estimates agree that during single-leg stance, the forces across the hip joint are twice the normal body weight for a given patient. With double-leg stance, the force at each hip joint is equal to body weight. However, estimates of the contribution of muscle activity across the hip, in addition to the activity being performed during measurement, demonstrate hip compression forces of at least six to seven times body weight. For example, measurement of the force exerted through the plant leg in a high jumper have been estimated to be greater than 1000 lb; in a man weighing less than 175 lb, this amount of force will produce an equal and opposite reactive force that must be transmitted through the hip.

 Muscles Acting on the Hip

The muscles acting across the hip joint are multiple, and their action is best analyzed in functional groups. However, it should be noted that these muscles are among the strongest in the body, and it is the force of contraction of these muscles that can produce acetabular forces far greater than the compression due
to body weight in isolation.

Hip flexion is accomplished by the psoas and iliacus muscles arising from the lumbar spine and iliac fossa, respectively. These muscles insert on the lesser trochanter and produce external rotation in addition to hip flexion. Other muscles that produce hip flexion, but to a lesser degree, due to the relatively short lever arms for flexion, include the tensor fascia lata, rectus femoris, and gracilis.

Hip extension is produced by the gluteus maximus and the hamstring complex. The hamstrings originate on the bony pelvis and cross both the hip and the knee joint. Because of this, these muscle produce not only hip flexion in an open chain joint system, but also knee extension in a closed chain system. The gluteus maximus is also a very strong external rotator of the hip.

Hip adduction is accomplished by the adductors (brevis, longus, and magnus), the pectineus, and the sartorius, with the majority of force contributed by the adductor complex. All of the hip adductors function in hip flexion, up to approximately 40 degrees. After this point, however, sufficient tension in the muscle develops to produce resistance to flexion; consequently, beyond this point, activation of the muscle will aid hip extension.

Hip abduction and internal rotation is accomplished via the lateral hip musculature. These muscles originate from the medial outer and iliac fossa and the anterior superior iliac spine and include the gluteus medius, gluteus minimus, and tensor fascia lata. The gluteus medius and minimus take an anterior, inferior, and lateral approach and insert on the inner and anterior aspect of the greater trochanter. It has been speculated that these muscles are active in hip internal rotation only with concomitant hip flexion; however, electromyographic analysis has shown them to be active even with a neutral hip position. Of interest, muscle testing of this group is difficult in any position other than the seated position with hip flexion. In the lateral recumbent position, weakness of this muscle group can easily be substituted for using either the hip flexors or hip external rotators if the patient is allowed to rotate externally into a more supine position (see Clinical Correlation 4–4).

External hip rotation is produced by the piriformis, gemelli, quadratus femoris, and obturator externus. These muscles all arise from the bony pelvis and insert posteromedially on the greater trochanter. The external rotators can be thought of in a similar fashion to the shoulder rotator cuff. These muscles not only produce external rotation, but are responsible for smoother rotation and compression of the femoral head within the acetabulum during all far end range of motion.
Muscle activity is responsible for tremendous joint compressive forces, far in excess of the combined force of gravity and body weight, and thus will alter gait mechanics. A Trendelenburg gait occurs due to weakness of the gluteus medius and minimus, and produces hip drop on the side contralateral to the involved muscle group. To compensate, the patient usually lurches with his or her upper body or side bent toward the ipsilateral side in order to pull the hip upward, or prevent further hip drop and gait instability. However, an uncompensated Trendelenburg gait may occur, usually in patients with degenerative hip disease. In such cases, the diseased hip is painful. To reduce pain symptoms, the patient develops a gait that minimizes gluteus medius and minimus contraction by lurching to the involved side, thus avoiding additional compressive forces across the joint. The force that must be produced by the hip abductor complex during gait is at least 2.5 times normal body weight. Therefore, during normal single-leg stance the force across the hip is 3.5 times normal body weight. By adopting an uncompensated gait, the patient reduces this force to near body weight, thereby reducing compression and pain (Figure 4–10).
Hip pain is due to the sum of two major forces: body weight and the force of muscle contraction, in this case, of the gluteus minimus and medius (hip abductors). During gait, the hip abductors contract to prevent hip drop and produce compression of the femoral head into the acetabulum. In an arthritic patient, this can produce pain. An uncompensated Trendelenburg gait occurs when a patient with arthritis utilizes a gait that minimizes the contraction of the gluteus medius by leaning the majority of the upper body over the arthritic side. In doing so, the patient minimizes the lever arm of the external load across the hip. With a single-leg stance, the torque produced by the hip abductors ($T_{abd}$) must be equal to the torque of body weight ($T_{bw}$) across the center of mass. This is represented by the equation $T_{abd} = T_{bw}$, where $T_{abd} = F_{abd} \times d_{abd}$ (and $d_{abd} = 1$), and where $T_{bw} = F_{bw} \times d_{bw}$ (and $d_{bw} = 2$). Substituting then for force, $F_{abd} = 2 \times bw$. The second force, the weight of the body, is the total body weight (tbw) minus the weight of one limb that is below the center of mass, or approximately one sixth of tbw. Therefore, with a single-leg stance, the force at the acetabulum, the sum of muscle contraction/compression and body weight, is 2.55 tbw. Because tbw is fixed, the patient can minimize these forces by decreasing the force of hip abductor contraction. This is done by shifting the upper trunk over the painful side, in effect shortening the lever arm of the resistance the contracting muscle must oppose. If enough shift occurs, the $T_{bw}$
can be reduced; $F_{bw} \times dbw$, where $dbw$ may be $1 \leq dbw \leq 2$. The use of a cane also significantly reduces the $Tabd$. In a 100-kg male, for example, the application of even a small amount of force (eg, 10 kg of downward force) can significantly reduce both activity of the hip abductors and hip pain caused by femoroacetabular compression, owing to the long lever arm of the force applied. With a cane, the $Tabd = T_{bw}$; however, because the force produced using the cane is in the same direction as the force of hip abduction, the equation should be $T_{cane} + T_{abd} = T_{bw}$, where $((F_{cane} \times dcane) + (F_{abd} \times dabd)) = F_{bw} \times dbw$. Solving for $F_{abd}$; $((F_{abd} \times 1) + (10 \, kg \times 6)) = (100 \, kg \times 2)$; $F_{abd} = 200 \, kg - 60 \, kg = 140 \, kg$. The application of only 10 kg of force thereby reduces the force of abduction to 140 kg (remember that normally this is at least 200 kg). Cane use therefore can result in 30–45% reduction in the force of contraction of the hip abductors.

**KINESIOLOGY OF THE KNEE**

The knee joint is composed of the femorotibial and patellofemoral joints. It is crucial for the smooth coordination of movement and for transmission of joint and reactive forces during walking, running, and jumping. Because of this the knee joint is one of the most common locations for development of degenerative changes and pain.

**Knee Joint Motion**

Major joint motion occurs in flexion and extension; however, internal–external rotation as well as varus–valgus movements are possible. In addition to these major joint motions, all of these actions have components of compression–distraction and mediolateral and anteroposterior translation. Normal knee motion in the sagittal plane ranges from slight hyperextension of $−4$ to $0$ degrees to $130–160$ degrees of flexion. Translation motion is also greatest during knee flexion.

In the fully extended position the knee is stabilized by the joint capsule, ligaments, and muscles. However, maximal anterior translation of the tibia occurs around 30 degrees of flexion due to the laxity of the anterior cruciate ligament relative to its tension in full extension. Similarly, maximal posterior translation of the tibia occurs with 90 degrees of knee flexion, due to relative laxity of the posterior cruciate ligament. This is why, when testing for ligamentous injury, the knee is positioned at 30 degrees of flexion for Lachman’s test and at 90 degrees of flexion for the posterior drawer test.
Motion in the frontal plane consists of varus and valgus angulation and depends on the degree of knee flexion. Maximal angulation occurs around 30 degrees of flexion and is greater with varus than with valgus motion due to the laxity of the lateral collateral ligament during flexion and relative tightness of the medial collateral with the same amount of angulation. Varus and valgus movement averages approximately 3–15 mm in either direction.

Transverse plane motion of the knee involves internal and external rotation of the tibia on the femur. During full extension, there is minimal transverse plane motion. Internal rotation increases progressively during flexion and reaches a nadir between 100 and 120 degrees. During gait, external rotation occurs as the knee moves from flexion back into extension. This results from the differential size of the femoral condyles, with the medial condyle having a greater radius than the lateral condyle. This “screw-home” mechanism is responsible for increased stability of the knee during terminal swing and initial contact phases of gait.

Forces at the Knee

Forces at the knee are dependent on activity, foot position, knee angulation, location of the ground reactive forces, and body position, among other factors. Anterior and posterior translatory forces at the knee are counteracted by the capsular and ligamentous structure. The anterior cruciate ligament (ACL) runs from the posteromedial portion of the lateral femoral condyle and inserts on the anterior portion of the intercondylar groove of the knee. The ACL is the primary deterrent to excessive anterior translation of the tibia on the femur. In addition, the medial meniscus provides resistance to excessive translation, especially if the ACL is compromised. The ACL is maximally tight in full extension; therefore, this is the most stable position of the knee for resisting anterior translation, and it is also the position from which the ACL is most likely to be damaged when a high-magnitude posterior translatory force is applied to the femur. The posterior cruciate ligament (PCL) runs from the antero-lateral portion of the medial femoral condyle and inserts relatively posteriorly on the intercondylar groove of the knee. Similar to the ACL, the PCL is maximally tight in full extension and in this position will resist excessive posterior translation of the tibia on the femur. Internal rotatory forces about the tibia are resisted by the medial collateral ligament and the anterior cruciate ligaments. External rotatory forces are restrained by the posterior joint capsule and the posterior cruciate ligament.


Immobility is the enemy of function. Much of physiatric treatment revolves around movement and its antithesis, immobility. This concept applies equally to generalized immobility (progressive functional decline in senior citizens as a result of cumulative effects of pain, fear of falling, and muscle weakness), forced immobility (bed rest during hospitalization or experienced by astronauts in microgravity), and immobilization of discrete body parts (range-of-motion [ROM] restriction caused by spasticity, contracture, or splinting or casting of fractures).

Rehabilitation marshals the body’s ability to change, adapt, and grow in response to stimuli. This is as true for neuroplasticity in a stroke patient as it is for strengthening of the rotator cuff muscles in a patient with chronic tendinopathy. Immobility is not a null state in which bodily functions remain in physiologic equilibrium. Like movement and exercise, immobility is also a condition that stimulates physiologic adaptation, leading to rapid changes in cardiovascular, pulmonary, musculoskeletal, and neurologic function that ultimately decrease our ability to interact with the world around us. The cause and effects of immobility in patients should always be examined and reexamined. These factors have great bearing on an individual’s function, independence, safety, and emotional well-being.

**BED REST & IMMOBILIZATION**

Throughout the history of medicine, enforced immobilization or bed rest has been a staple of treatment for both illness and injury. Conditions that historically
have been or currently are treated with enforced rest include acute low back pain, heart attack, tuberculosis, fracture, and critical illness (eg, sepsis).

Actual and theoretical justifications for bed rest include reducing metabolic demands of the body, thus “conserving” these resources for recovery. Reduction of oxygen consumption by muscles—both skeletal and cardiac—decreases oxygen demand by all tissues, resulting in less mechanical ventilation, lower Fio\textsubscript{2}, and less risk of ventilator-induced lung injury. Other perceived benefits include decreased cardiac stress, improved central nervous system perfusion, bone healing, and pain control (eg, in the immobilization of an injured appendage for comfort). Additional perceived benefits apply to safety: maintenance of intravenous access and artificial airways, fall prevention, and prevention of occupational injuries to nursing staff.

Specialists in physical medicine and rehabilitation work in the borderland between mobility and immobility. Patients with spinal cord injury, neuropathy, stroke, or peripheral nerve injury with sensory impairment do not experience the same discomfort that would prompt a neurologically intact person to shift position or treat a wound or laceration. Physiatrists also treat patients who are protectively immobilized in splints or casts for fractures, ligament tears, or tendon injury. They often encounter and treat the effects of self-imposed or functional immobility of joints due to stiffness and pain.

**MUSCULOSKELETAL CONSEQUENCES OF INACTIVITY**

1. **Weakness**

Muscle mass drops by 1.5–2% *per day* in the first 2–3 weeks of bed rest. A muscle completely at rest therefore loses 10–15% of its strength each week, resulting in a loss of roughly 50% after 3–5 weeks of complete immobilization. The lower limb and truncal antigravity muscles (hip extensors, knee extensors, ankle plantar flexors, and paraspinals) are preferentially affected.

Decrease in muscle mass is driven by atrophy. Individual muscle fibers decrease in size, resulting in loss of tension-generating ability and proportional loss of torque. In one study of nine healthy male volunteers exposed to absolute, horizontal bed rest, the cross-sectional area of the gastrocnemius–soleus (ankle plantar flexor) complex decreased by 12% as measured on magnetic resonance imaging. The same muscles lost 26% of strength as measured by dynamometer. Notably, the ankle dorsiflexors (not an antigravity muscle in normal function) did not show significant decrease of area or strength in this study.

Neurogenic muscle immobility resulting from central or peripheral nerve
injury has even more dire consequences. Peripheral nerve injury causes flaccid paralysis, and the affected muscles can lose up to 95% of their bulk. If the denervation is irreversible (ie, neurotmesis), the muscle fibers are replaced by fatty atrophy and connective tissue. In spastic paralysis caused by upper motor neuron injury (eg, stroke or spinal cord injury), the antigravity muscles can lose 30–40% of their bulk.

### Prevention

Contraction of a muscle to 20% of maximal tension for several seconds a day will prevent loss of strength. However, this does little to counteract the multitude of other physiologic effects of bed rest.

### Treatment

Disuse myopathy may be reversed with exercise. However, the recovery of strength occurs at a rate of only 6% per week at submaximal (65–75% max) exercise. Thus, for every day of hospital bed rest, the patient must pay back 2–3 days of exercise to return to baseline muscle strength. This figure does not account for the recovery time from other, nonmuscular physiologic effects of bed rest, nor from the illness that resulted in hospitalization in the first place.

### 2. Contracture

A contracture is a fixed joint deformity resulting from immobilization of the joint. The immobilization itself may be caused by splinting or casting of a fracture, or by other disruptors of dynamic tension, such as spasticity from an upper motor neuron lesion (active opposition) or denervation with flaccid paralysis (lower motor neuron lesion, which results in no opposition). Other factors include bed positioning or intrinsic muscle and soft tissue changes caused by burns or connective tissue disease, such as scleroderma.

### Risk Factors

Groups at highest risk include those with joint disease or paralysis of a muscle group, the frail and elderly, the cognitively impaired, and the very passive, including those with mental illness and catatonia. Muscles at highest risk of
contracture during immobilization are those that cross two joints, such as the hamstrings, back muscles, tensor fascia lata, rectus femoris, gastrocnemius, and, in the upper limb, the biceps brachii.

Pathogenesis

Ligaments are often viewed as relatively fixed, stable structures. In reality, they are dynamic, complex structures that actively respond to the presence or absence of mechanical forces. Thus, joint immobilization can lead to physiologic changes of both the ligamentous insertion and the ligament body itself.

Collagen fibers in areas of movement develop loose alveolar connective tissue, and frequent stretching maintains length. In the absence of movement, the collagen develops into a dense mesh of interconnected sheets and will shorten if not stretched frequently. This can progress to intraarticular fibrofatty infiltration and persistent adhesions.

A series of animal studies investigated the results of 8 weeks’ immobilization in a total body cast. At the end of the trial, knee ligament stiffness decreased to 69% of normal, maximum load at failure decreased to 61% of normal, and energy absorption decreased to 68% of normal. Of note, at 1 year followup, the ligaments had still not returned to their premorbid condition.

Clinical Findings

Contractures caused by immobility inevitably lead to more severe and prolonged impairment unless aggressive and persistent intervention is undertaken. Contractures interfere with positioning in bed and in wheelchairs, and may bring pressure-sensitive areas into contact with surfaces that increase the risk of deep tissue injury. For example, an immobilized patient with hip flexion contractures may be unable to maintain a supine or semisupine position in bed, have a reduced turning schedule, and develop trochanteric pressure wounds. Contractures also interfere with activities of daily living and the administration of nursing care and hygiene. Left untreated, spastic contracture promotes moisture, fungal infection, maceration and breakdown of the skin, and potentially cellulitis, sepsis, and death.

In the realm of active movement, contractures interfere with normal biomechanics and may prevent or impede safe transfers and ambulation. Even if they allow safe ambulation, lower limb contractures may shorten step length,
destabilize stance, and increase overall energy consumption, thus decreasing activity tolerance. Upper limb contractures may prevent effective use of assistive devices or generate mechanical disadvantage in functional tasks or tool use that confounds accommodative or adaptive techniques.

**Prevention & Treatment**

Prevention is essential. It is easier to prevent a contracture than to correct one, though it does require time, labor, and attention to detail. A consulting physiatrist is in a position to assist in the prevention of contractures in the acute care setting by recommending and implementing active and passive ROM exercise programs, static and dynamic bracing (eg, resting splints, functional bracing), serial casting if trained personnel are available, bed and chair positioning, and referral for surgical release when appropriate.

If a joint must be immobilized it should be done in the stretched position if possible, to decrease muscle atrophy, degree of contracture, and loss of tensile strength. If immobilization in stretch is impossible, an alternative is to immobilize in the neutral position to balance length and tension of opposing muscles. Early active immobilization after stabilization is beneficial, if permitted by the nature of the surgical repair.

Established contractures should be assessed for underlying etiology (bony deformity versus spastic paralysis), hard versus soft end point, degree of pain or discomfort both at rest and when stretched, and degree of impact on function. Contractures may be treated with deep heating to 40–45° C, passive ROM, and terminal stretch for 25–30 seconds. Chemoneurolysis with botulinum toxin or phenol, serial casting, splinting, or use of other devices (eg, hinged, locking knee brace; dynamic splint; or continuous passive motion device) may be prescribed when appropriate. Care must be taken with frail or chronically immobilized patients (eg, the elderly or those with spinal cord injury) to prevent insufficiency fractures. **Table 5–1** lists contraindications to aggressive ROM programs for contracture.

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**Table 5–1** Contraindications to aggressive range-of-motion programs for contracture.
CARDIOVASCULAR CONSEQUENCES

Like the musculoskeletal system, the heart and blood vessels will actively adapt to prolonged bed rest, as anyone who has attempted to ambulate a patient in the intensive care unit can attest. Studies have shown that heart rate increases by 7–10 beats per minute after 7–14 days of bed rest. Concomitantly, the elevated heart rate decreases systolic ejection and diastolic filling time, rendering the heart less able to meet increased metabolic demands. Table 5–2 summarizes these and other cardiovascular changes associated with prolonged bed rest.

### Table 5–2 Cardiovascular complications of immobility and bed rest.

<table>
<thead>
<tr>
<th>Increased heart rate</th>
<th>Decreased cardiac output</th>
<th>Decreased stroke volume</th>
<th>Impaired left ventricular function</th>
<th>Decreased cardiac reserve</th>
<th>Postural hypotension</th>
</tr>
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</table>

Orthostatic hypotension can occur after 3 weeks of bed rest, or even shorter periods in elderly individuals. The mechanism is multifactorial and involves pooling of blood in the venous circulation of the lower limbs, decreased circulating blood volume, loss of sympathetic tone, and increased heart rate with decreased ventricular diastolic filling.

In a classic 1968 study by Saltin and colleagues, 24 young, healthy male
volunteers were subject to 20 days of strict bed rest. The experiment produced a 27% decrease in maximal oxygen uptake, a 25% decrease in stroke volume, and a 20% increase in heart rate. These effects may be exacerbated in the frail and elderly, and in those with preexisting cardiovascular impairment, as well as causing more significant functional consequences in these groups.

**GASTROINTESTINAL CONSEQUENCES**

Bed rest removes the normal benefits that gravity and activity exert on the digestive system. Esophageal and gastric transit times are prolonged in the esophagus and stomach, up to 66% slower in the supine position. This effect extends to the colon, where stasis leads to excessive reabsorption of water, constipation, and fecal impaction. Furthermore, in the seated (upright) position, stool exerts pressure on the anal sphincter creating the urge to defecate. In the supine position the effect is absent. Bed rest in hospital settings, particularly in cases of trauma, is often accompanied by opioid pain medications, which slow gut motility. This is potentiated by NPO status and lack of dietary fiber and oral hydration.

The loss of normal positioning also allows acidic gastric secretions to collect in the upper stomach and exert pressure against the lower esophageal sphincter. Coupled with the decreased gastric bicarbonate secretion occurring in bed rest, which lowers stomach pH, this may increase symptoms of gastroesophageal reflux and cause or exacerbate preexisting dysphagia.

**RENAL & GENITOURINARY CONSEQUENCES**

The urinary system also depends partly on gravity for normal function. The renal calices drain into the ureters by gravity alone. Supine bed rest causes areas of stasis that, coupled with hypercalciuria as a result of bone demineralization, can increase the risk of renal calculi. Drainage of urine through the ureters is also abetted by gravity, though ureteral peristalsis will fill the bladder even in supine position.

The urge to urinate, created by pressure on the urinary sphincter, bladder wall, and bladder neck, is decreased in the supine position as abdominal organs no longer press downward on the bladder. This may lead to loss of bladder wall tone and elasticity, urinary retention, stasis, and infection. Stasis and urinary tract infection, particularly with urease-splitting organisms that alkalinize the urine, also contribute to the formation of renal stones.

**PULMONARY CONSEQUENCES**
PULMONARY CONSEQUENCES

The mechanism of atelectasis in supine bed rest is thought to occur because of upward (cephalad) shifting of the diaphragm along with downward (dorsal) shifting of the heart in the supine position. Left lower lobe atelectasis may be observed on radiographs within 48 hours of initiating bed rest. Table 5–3 summarizes these and other pulmonary complications of bed rest.

Table 5–3 Pulmonary complications of immobility and bed rest.

<table>
<thead>
<tr>
<th>Decreased ventilation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventilatory muscle deconditioning</td>
</tr>
<tr>
<td>Decreased respiratory capacity</td>
</tr>
<tr>
<td>Compensatory increase in respiratory rate</td>
</tr>
<tr>
<td>Alteration in ventilation/perfusion ratio (less ventilation, increased perfusion) → arteriovenous shunting</td>
</tr>
<tr>
<td>Impaired cough</td>
</tr>
<tr>
<td>Atelectasis</td>
</tr>
<tr>
<td>Hypostatic pneumonia</td>
</tr>
</tbody>
</table>

MENTAL HEALTH CONSEQUENCES

Immobility in the hospital setting leads to sensory deprivation, mental deterioration, delirium, behavioral disturbances, and dependency. These effects are accentuated in cognitively impaired patients, who are present in large numbers in acute and subacute rehabilitation settings. Consider the effects of prolonged intensive care unit and hospital stays on patients with stroke, traumatic brain injury, or spinal cord injury, and in elderly patients with early or not-so-early stage dementia. Although isolation itself does not cause cognitive decline, isolation combined with lack of physical exercise does. Other complications of sensory deprivation include intellectual regression, depression, short attention span, and poor motivation.

Immobility and confinement to health care facilities foster dependency. Patients conform to expected roles. The result of overprotective caregivers is often increasingly passive patients. Additionally, vastly different levels of functional independence may be noted in patients based on whether the patient
interaction is with the physical or occupational therapist or with nursing staff.


PRESSURE ULCERS

General Considerations

Pressure ulcers are areas of ischemic injury that develop when soft tissue is compressed under bony prominences throughout the body. The term pressure ulcer is preferred over other sometimes obsolete nomenclature such as decubitus ulcer, bedsore, pressure sore, and dermal ulcer. Individuals at highest risk include the elderly, those with impaired mobility, and those with sensory impairment (eg, spinal cord injury, peripheral neuropathy, stroke, or other neurologic injury). Unintentional weight loss, protein–energy malnutrition, and low body mass index are additional risk factors for developing pressure ulcers.

Pathogenesis

Pressure ulcers are caused by the combination of extreme or prolonged pressure, shear forces, friction, and micro-climate effects. According to the National
Pressure Ulcer Advisory Panel (NPUAP), pressure, a unit of force per unit area, occurs perpendicularly, whereas shear is the force per unit area exerted parallel to the plane of interest. The resistance of parallel motion between the body and underlying surface is known as friction. Microclimate refers to skin temperature and moisture conditions of the skin and contributes to the development of pressure ulcers. Common areas for skin breakdown from these factors include the sacrum, back, buttocks, heels, occiput, and elbows. Most pressure ulcers occur in the lower body; an estimated 36% occur at the sacrum and 30% on the heels.

**Prevention**

A successful preventative program first identifies individuals at risk. The Braden Scale for Predicting Pressure Sore Risk evaluates six areas—sensory perception, moisture, activity, mobility, nutrition, and friction and shear—to stratify risk on a 23-point scale. In addition to identifying high-risk patients, preventative strategies include reducing pressure, friction, and shear forces, and optimizing nutrition. Many studies have evaluated efficacy of pressure-reducing surfaces, including dynamic and static support mattresses. Dynamic support surfaces mechanically alternate pressure forces, whereas static surfaces or overlays provide a constant level of pressure relief. A retrospective study by Reddey and colleagues determined that support surfaces, in particular mattress overlays, are superior to standard mattresses; however, the study found no significant difference between dynamic and static support surfaces. Because dry sacral skin is a contributing factor to pressure ulcer development, the study recommended that standard preventative measures should include moisturizing sacral skin. Although repositioning techniques are often the first-line prevention measure in the clinical setting (eg, turning the patient every 2 hours), there is no definitive evidence to support this recommendation. From a nutritional standpoint it is helpful to obtain an assessment from a registered dietician, particularly in the acute care setting. NPUAP guidelines recommend that patients receive a diet that provides 30–35 kcal per kilogram and 1.25–1.5 g of protein per kilogram of body weight per day. Additionally, ascorbic acid may contribute to wound healing.

**Clinical Findings**

In 1975 Darrell Shea developed a staging system for pressure ulcers that became
the foundation for current staging criteria. Pressure ulcer staging has undergone multiple revisions since 1975; however, Shea’s concept of tissue depth has remained. The most recent updates to the pressure ulcer classification system were made in 2007 by NPUAP. Current NPUAP staging guidelines, with accompanying illustrations, are outlined in Table 5–4.

Table 5–4 Staging of pressure ulcers.
<table>
<thead>
<tr>
<th>Schematic Illustration</th>
<th>NPUAP Stage</th>
<th>Photographic Illustration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal: No skin discoloration or breakdown.</td>
<td><img src="image1.png" alt="" /></td>
<td></td>
</tr>
<tr>
<td>Suspected deep tissue injury: Skin discoloration with intact skin.</td>
<td><img src="image2.png" alt="" /></td>
<td></td>
</tr>
<tr>
<td>Stage 1: Intact skin with nonblanchable erythema usually over a bony prominence.</td>
<td><img src="image3.png" alt="" /></td>
<td></td>
</tr>
</tbody>
</table>
Stage II: Partial-thickness loss of dermis, an open ulcer with a pink wound bed, or blister.

Stage III: Full-thickness tissue loss with no visible bone, tendons, or muscle. Undermining or tunneling may be present.

Stage IV: Full-thickness tissue loss with exposed bone, tendon, or muscle. Slough, eschar, tunneling, and undermining are typically present.
Complications

Pressure ulcers have been linked to pain, depression, local infection, osteomyelitis, anemia, sepsis, gas gangrene, necrotizing fasciitis, and increased hospital length of stay and mortality. Infection should be suspected in cases of delayed wound healing. Infections are typically polymicrobial, including gram-positive and gram-negative bacteria as well as anaerobes.

Treatment

Pressure ulcer treatment includes wound assessment, pressure relief, management of friction or shear, management of bacterial colonization and infection, education, and quality improvement. It is important to keep the wound moist, clean, and noninfected. This is accomplished by means of local wound care, which includes dressing changes and debridement. Commercially available
dressing types are too numerous to list, but common dressing properties include exudate absorption, hydration, debridement, and antimicrobial activity. Currently, no specific dressing type has been found to be superior to others. Chemical, enzymatic, or surgical debridement may be necessary to keep the wound base clean. The ultimate goals of treatment are to manage moisture levels and bacterial balance in order to promote wound healing.


HETEROTOPIC OSSIFICATION

General Considerations

Heterotopic ossification (HO) is the condition in which mature lamellar bone forms within the soft tissue, typically around a joint. The term derives from heteros (Greek: “other”), topos (Greek: “place”), and os (Latin “bone”). HO may be hereditary or acquired (eg, traumatic or neurogenic etiology). Fractures, especially of the hip, are most commonly associated with HO. The reported
incidence of HO after total hip arthroplasty ranges from 5% to 90%, and after total knee arthroplasty, from 3% to 39%. Traumatic etiologies of HO also include dislocation and burns. Neurogenic HO is commonly seen in spinal cord injury and brain injury. The reported incidence in spinal cord injury ranges from 10% to 53%, and in traumatic brain injury, from 11% to 76%. These ranges vary widely owing to the largely asymptomatic nature of early HO. Hereditary conditions associated with HO include fibrodysplasia ossificans progressiva, progressive osseous heteroplasia, and Albright’s hereditary osteo-dystrophy.

### Pathogenesis

The exact pathophysiologic mechanism of HO remains unclear. It has been proposed that HO develops as a postinjury inflammatory response. Within the soft tissues, inflammatory mediators such as prostaglandins are upregulated and act to differentiate mesenchymal stem cells into osteoblasts. As in normal bone formation, osteoblasts utilize alkaline phosphatase and tropocollagen to ultimately mineralize the bony matrix. Thus, histologic and radiographic findings mimic those of normal bone.

### Clinical Findings

#### A. Symptoms and Signs

The individual clinical signs of HO are nonspecific. Signs may include, but are not limited to, pain, edema, decreased ROM, and fever. In the brain injury population, HO preferentially affects hips, elbows, shoulders, knees, and thighs in descending order of frequency. In patients with spinal cord injury, HO develops below the neurologic level of injury and involves the hips more frequently than the knees. Risk factors for HO include pressure ulcers, urinary tract infections, renal stones, venous thromboembolism, spasticity, and trauma. In nonhereditary HO, symptoms typically appear from 3 to 12 weeks after the precipitating event. If left untreated, HO may lead or contribute to the development of joint ankylosis, loss of ROM, and pain. This can result in additional impairment with ambulation and activities of daily living, and increase the burden of care for both nursing staff and family members. HO also increases the risk of pressure ulcers and venous thromboembolism due to the resultant immobilization.
B. Diagnostic Studies

If HO is suspected for reasons of rehabilitation diagnosis, loss of function, or physical signs such as pain with ROM and manipulation of joints, the clinician should check serum alkaline phosphatase as an initial screening measure. Although alkaline phosphatase is not highly specific for the condition, in the presence of HO levels can be elevated up to 3.5 times above the normal reference range. Prostaglandin levels may also be measured; specifically, 24-hour urinary prostaglandin E₂ (PGE₂) levels may be monitored to detect active HO. If urinary excretion of PGE₂ increases sharply, then imaging is indicated. For final confirmation the nuclear medicine triple phase bone scan is the gold standard for diagnosis. Although plain film radiographs can be used, visible HO on X-ray may lag behind positive bone scan findings by as much as 2 weeks. If an initial bone scan is negative and clinical suspicion remains high, the test should be repeated at a later date.

Treatment

Early treatment of HO is essential to avoid major complications later on. A comprehensive rehabilitation program focusing on ROM, training in activities of daily living, and psychological recovery is an integral part of nonpharmacologic treatment. During the acute phase of HO, passive ROM exercises are initiated, and active ROM and aggressive strengthening exercises are avoided to rest the affected joint. Primary pharmacologic treatment of HO consists of nonsteroidal antiinflammatory medications (NSAIDs), cyclooxygenase (COX) inhibitors, and sodium bisphosphonates. It has been proposed that NSAIDs block the inflammatory cascade, which is thought to contribute to HO formation. Indomethacin or ibuprofen are typically the NSAIDs of choice. There is limited but growing evidence that supports the use of bisphosphonates in the treatment and prevention of HO. Bisphosphonates inhibit the formation of hydroxyapatite crystals, a necessary step for bone formation.

In patients who have significant physical or functional impairment as a result of HO, surgical excision may be considered; however, excision is associated with a high risk of recurrence. Surgical intervention should be deferred until the HO has matured: 6 months postinjury when associated with trauma, 12 months after spinal cord injury, and 18 months after brain injury. This recommendation is based on the supposition that mature HO is less likely to recur after surgical removal.


VENOUS THROMBOEMBOLISM (DEEP VEIN THROMBOSIS)

General Considerations

The American College of Chest Physicians (ACCP) publishes comprehensive and summary guidelines for treatment of venous thromboembolism (VTE). An overview of the rehabilitation-relevant portion of the guidelines is presented here. (For prophylactic pharmacotherapy used in the prevention of VTE, refer to Chapter 10.)

Clinical Findings

Patients should be assessed clinically for risk factors and symptoms. First-line diagnostic testing in low-risk patients should be done with a moderate or high-sensitivity D-dimer assay or a compression ultrasound of the suspected limb. If the D-dimer result is negative, no further testing need be conducted. If positive, it should be followed up with ultrasound studies of the proximal veins. If compressive ultrasound is negative in a patient with suspected VTE, it should be followed with either a moderate to highly sensitive D-dimer assay or a followup ultrasound in 1 week. Obviously, outside factors may affect the choice of diagnostic testing. For example, ultrasound evaluation would be impractical if not useless in a patient with lymphedema, morbid obesity, or a casted limb.
Conversely, a known premorbid condition that elevates d-dimer levels would render that assay equally unhelpful. Alternative testing with venography, or computed tomography or magnetic resonance venography, may be required.

## Treatment

### A. Medical Management

For acute deep vein thrombosis (DVT) of the lower limb, with initiation of vitamin K antagonist therapy (eg, warfarin), the patient should be bridged with therapeutically dosed parenteral anticoagulants (eg, enoxaparin or unfractionated heparin). If clinical suspicion of DVT is high, then bridging agents should be initiated while awaiting test results. If moderate, bridging agents should be started if test results are to be delayed for more than 4 hours. If clinical suspicion is low, bridging agents may be delayed pending test results, as long as results are expected within 24 hours.

If the patient has acute but isolated DVT in the distal leg, without severe symptoms or risk factors, the thrombosis may be monitored with serial ultrasound for 2 weeks. This is also the suggested approach for patients with high risk of bleeding. If patients have severe symptoms or risk factors for extension, anticoagulation should be initiated as for acute proximal DVT. Enoxaparin or fondaparinux are the preferred agents, although alternative agents may be used if these are unavailable, cost-prohibitive, or contraindicated due to renal impairment. Table 5–5 summarizes information about these and other agents that may be used in the treatment of DVT. Bridging therapy should be started at the same time as warfarin and continued for at least 5 days and until the international normalized ratio (INR) is greater than 2.0 for at least 24 hours. For acute DVT of the lower limb, anticoagulant therapy alone is recommended over systemic thrombolysis, intraarterial thrombolysis, or surgical thrombectomy. Even if these more aggressive options are pursued, the anticoagulation regimen should be the same as in patients who do not undergo the intervention.

<p>| Table 5–5 | Pharmacotherapy for deep vein thrombosis. |</p>
<table>
<thead>
<tr>
<th>Generic Drug (Proprietary Brand)</th>
<th>Delivery</th>
<th>Mechanism</th>
<th>Contraindications</th>
<th>Monitoring</th>
</tr>
</thead>
<tbody>
<tr>
<td>Argatroban (Acova)</td>
<td>Intravenous</td>
<td>Direct thrombin inhibitor</td>
<td>Hepatic impairment</td>
<td>aPTT</td>
</tr>
<tr>
<td>Bivalirudin (Angiomax)</td>
<td>Subcutaneous injection</td>
<td>Direct, reversible thrombin inhibitor</td>
<td>Renal impairment</td>
<td>Check baseline creatinine</td>
</tr>
<tr>
<td>Dalteparin (Fragmin)</td>
<td>Subcutaneous injection</td>
<td>Binds to and accelerates antithrombin III → inhibiting thrombin and factor Xa</td>
<td>HIT, epidural or spinal anesthesia, renal or hepatic impairment</td>
<td>CBC, platelets, anti-Xa levels if renally impaired</td>
</tr>
<tr>
<td>Enoxaparin (Lovenox)</td>
<td>Subcutaneous injection</td>
<td>Binds to and accelerates antithrombin III → inhibiting thrombin and factor Xa</td>
<td>HIT, epidural or spinal anesthesia, renal or hepatic impairment</td>
<td>CBC, platelets, anti-Xa levels if renally impaired</td>
</tr>
<tr>
<td>Fondaparinux (Arixtra)</td>
<td>Subcutaneous injection</td>
<td>Selectively binds to antithrombin III, neutralizing factor Xa and inhibiting thrombin</td>
<td>Epidural or spinal anesthesia, renal impairment, thrombocytopenia with antiplatelet antibodies</td>
<td>Check baseline creatinine, periodic blood count with platelets, anti-Xa levels</td>
</tr>
<tr>
<td>Unfractionated Heparin</td>
<td>Intravenous infusion, subcutaneous injection</td>
<td>Binds to and accelerates antithrombin III → inhibiting thrombin and factor Xa</td>
<td>HIT, epidural or spinal anesthesia, renal or hepatic impairment</td>
<td>aPTT</td>
</tr>
<tr>
<td>Dabigatran etexilate (Pradaxa)</td>
<td>Oral</td>
<td>Direct, reversible thrombin inhibitor (free and clot-bound)</td>
<td>Bleeding, age &gt; 75 y</td>
<td>Check baseline creatinine</td>
</tr>
<tr>
<td>Rivaroxaban (Xarelto)</td>
<td>Oral</td>
<td>Selectively blocks active site of factor Xa (free and clot-bound)</td>
<td>Hepatic or renal impairment, epidural or spinal anesthesia</td>
<td></td>
</tr>
<tr>
<td>Warfarin (Coumadin)</td>
<td>Oral</td>
<td>Inhibits vitamin K-dependent coagulation factor production (factor II, VII, and IX), protein C and S</td>
<td>Hepatic or renal impairment; spinal or epidural anesthesia or puncture, blood dyscrasias, noncompliant patients, protein C or S deficiency</td>
<td>PT/INR</td>
</tr>
</tbody>
</table>

aPTT, activated partial thromboplastin time; HIT, heparin-induced thrombocytopenia; CBC, complete blood count; PT/INR, prothrombin time/international normalized ratio.
Inferior vena caval (IVC) filters are not recommended in patients receiving appropriate anticoagulation therapy. IVC filters are recommended if there is a contraindication to anticoagulation (such as bleeding); however, if the risk of bleeding resolves the standard method for anticoagulation should be implemented, as described above. The presence of a long-term IVC filter without other risk factors does not in itself indicate the need for anticoagulation.

Patients with asymptomatic acute DVT in the lower limb should receive anticoagulation therapy as for those with symptomatic DVT. Compression stockings are also recommended and should be worn for 2 years to prevent post-thrombotic syndrome.

B. Mobilization

Early ambulation rather than bed rest is recommended and encouraged in patients with acute lower limb DVT, unless pain and swelling are too severe to permit ambulation. In these cases, (passive) compression stockings should be used. This strategy may be at odds with conventional wisdom at the clinician’s institution, owing to the mistaken notion that a thrombus is more likely to become dislodged with activity. An informal poll conducted in one university hospital in 2009 revealed discrepancies in the allowable activity levels of patients with acute DVT, which the clinician may find replicated across many institutions. In the survey, nurses expected orders for bed rest without mechanical compression devices. Physical therapists expected orders for bed rest until anticoagulated, followed by ambulation. Physicians recommended ambulation with the addition of other measures (standard anticoagulation, antiplatelets, or compression devices).

Multiple clinical trials have found that early ambulation in cases of acute DVT does not statistically increase the risk of pulmonary embolus. A large-scale clinical trial in 2005 by Trujillo-Santos and colleagues examined the occurrence of pulmonary embolus in patients with acute DVT, comparing strict bed rest to ambulation during the first 15 days of anticoagulant treatment. No statistically significant difference in the occurrence of fatal pulmonary embolus or major bleeding was noted between the two groups. However, the bed rest group did demonstrate a higher risk of minor bleeding and mortality, whereas the ambulation group enjoyed earlier resolution of pain and edema and a lower rate of post-thrombotic syndrome. Thus, one may feel confident in allowing one’s patients with acute DVT to ambulate early, with the additional benefits of decreasing morbidity and mortality.
C. Special Patient Populations

1. Neurosurgical patients—The ACCP guidelines suggest that mechanical antithromboembolism stockings, ideally with intermittent pneumatic compression, be used preferentially over pharmacologic prophylaxis or no prophylaxis in patients following craniotomy or spinal surgery. If a patient is at very high risk for VTE, the surgeon should be consulted regarding when pharmacologic prophylaxis will be considered safe and appropriate.

Patients with major trauma with neurologic injury, including traumatic brain injury and spinal cord injury, should be preferentially treated with pharmacologic prophylaxis (unfractionated or low-molecular-weight heparin) or mechanical prophylaxis with intermittent pneumatic compression devices. If there are no lower extremity injuries that would prevent application of mechanical devices, then both pharmacologic and mechanical prevention should be used concomitantly. If pharmacologic agents are contraindicated, then mechanical measures should be used until the risk of bleeding decreases, at which time pharmacologic prophylaxis should be started. IVC filters alone should not be used for primary prevention of VTE.

2. Orthopedic patients—In the rehabilitation setting, there are legions of patients who have undergone total joint replacements (hips or knees) or surgery to fix hip fractures. Unless contraindicated, these patients should be preferentially treated with low-molecular-weight heparin for a minimum of 10–14 days postoperatively, with an optimum duration of up to 35 days postoperatively in the rehabilitation and outpatient setting. Alternative agents (eg, warfarin, unfractionated heparin, fondaparinux, apixaban, dabigatran, rivaroxaban, and aspirin) are available but are less favored owing to a higher risk of bleeding, higher risk of VTE, or lack of long-term safety data. If mechanical measures alone are indicated, because of increased bleeding risk, then portable units with memory and reporting functions to ensure compliance are recommended, with a minimum wear time of 18 hours daily.


47S.
Spasticity is a commonly encountered condition that can have a devastating impact on affected patients. Lance described it as “a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes with exaggerated tendon reflexes, resulting from excitability of the stretch reflex.” It can also be defined as the constant and unwanted contractions of one or more muscle groups as a result of an injury or insult to the brain or spinal cord. The condition can be mild, with patients experiencing only minor discomfort or inconvenience, or major, with the spasticity leading to immobility and the development of contractures and pressure sores. However, clinicians must use caution when applying such descriptors of severity to muscle overactivity so as not to misrepresent their clinical impact to the patient. For example, relatively slight resistance to passive motion, evaluated as “mild” by a physician, may have a quite significant functional impact for a patient, who might describe the same phenomenon as “severe.” Even mild degrees of spasticity can impair the ability to perform basic activities of daily living, including hygiene, dressing, and toileting. In addition, spasms associated with spasticity can cause pain, interrupt sleep, negatively influence mood, and impair mobility.

Although appropriate and timely treatment is needed to obtain optimal results, there is some evidence that spasticity is undertreated. Delayed or inadequate treatment can lead to maladaptive remodeling of the affected body part, leading to shortening of muscles and contracture of tendons or soft muscle and, ultimately, to a permanent physical deformity.
**CLINICAL FINDINGS**

▶ **Symptoms & Signs**

Pathologic changes in the central nervous system often create a constellation of symptoms or signs that encompass both positive and negative components. Weakness and loss of dexterity, the most commonly encountered negative phenomena, are relatively easy to define. Other negative signs include atrophy, fatigability, and loss of selective motor control. The positive components are more complex, with diverse pathophysiologic mechanisms. Observable phenomena include increased resistance to passive stretch, muscle–tendon hyperreflexia, clonus, co-contraction of synergistic muscle groups, and spontaneous flexor–extensor spasms. Spasticity is only one of these features, namely, a velocity-dependent increase in resistance to passive range of motion (ROM). Collectively, all of the positive signs can be called “muscle overactivity,” with the qualification that abnormal pathology extends beyond the muscle itself. Frequently, and perhaps unfortunately, the term spasticity is often applied to the entire collection of signs. Given this common practice, spasticity and muscle overactivity are used somewhat interchangeably for remainder of this chapter.

The causes of spasticity are heterogeneous. This syndrome is usually seen in conditions that involve damage to the portion of the brain or spinal cord that controls voluntary movement. Spasticity can be associated with spinal cord injuries, multiple sclerosis, cerebral palsy, stroke, and traumatic or anoxic brain injury, as well as metabolic or degenerative diseases such as adrenoleukodystrophy, amyotrophic lateral sclerosis, hereditary spastic paraparesis, stiff-person syndrome, and phenylketonuria. Although spasticity is a common condition, its incidence and prevalence are difficult to determine because of its association with a wide variety of disease processes. By combining several sources, it is reasonable to estimate that upward of 2 million
people in the United States experience spasticity. The estimated prevalence of spasticity for the most common etiologies is shown in Table 6–1.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Prevalencea</th>
<th>Reasonable Proportion Experiencing Spasticity</th>
<th>Estimated Number of Spastic Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral palsy</td>
<td>750,000</td>
<td>50%</td>
<td>375,000</td>
</tr>
<tr>
<td>Multiple sclerosis</td>
<td>400,000</td>
<td>60%</td>
<td>240,000</td>
</tr>
<tr>
<td>Cerebrovascular accident</td>
<td>7,000,000</td>
<td>20%</td>
<td>1,400,000</td>
</tr>
<tr>
<td>Traumatic brain injury</td>
<td>1,500,000</td>
<td>33%</td>
<td>500,000</td>
</tr>
<tr>
<td>Spinal cord injury</td>
<td>200,000</td>
<td>50%</td>
<td>100,000</td>
</tr>
<tr>
<td>Total, all conditions</td>
<td></td>
<td></td>
<td>2,615,000</td>
</tr>
</tbody>
</table>


**Patterns of Spasticity**

The clinical presentation of spasticity is quite diverse owing to the range of central nervous system disease that may produce muscle overactivity. One approach used to differentiate spasticity focuses on the number of muscle groups...
involved. Diffuse patterns of spasticity can involve all four limbs (quadriplegic), both lower extremities (paraplegic or diplegic), or the upper and lower extremities on the same side of the body (hemiplegic). Combination of these patterns can also be seen (eg, paraplegic and hemiplegic) in the same patient. The axial musculature of the cervical, thoracic, and lumbar spine can similarly be involved. Alternatively, a more localized appearance that involves only a few muscles or muscle groups can be detected. Mixtures of focal and diffuse patterns can also be recorded in the same patient.

Flexion (the movement of a limb to decrease the angle of a joint), extension (the opposite movement) or combined flexion–extension synergies can be observed. Motor synergies are stereotyped movements of the entire limb that reflect loss of independent joint control and that limit a person’s ability to coordinate his or her joints in flexible and adaptable patterns, thereby precluding performance of many functional motor tasks. In the upper extremity, the flexion synergy is often characterized by simultaneous shoulder abduction and elbow flexion. Conversely, upper extremity extension synergy is characterized by simultaneous shoulder adduction and elbow extension. In the lower limb, flexion synergy consists of hip flexion, abduction, and external rotation, along with knee flexion, ankle dorsiflexion, and inversion. Lower limb extension synergy consists of hip extension, adduction, internal rotation, knee extension, ankle plantar flexion, inversion, and toe plantar flexion. These synergistic patterns can appear as both a static positioning of an affected limb and a nonspecific movement pattern. Consider the following example: A man is asked to feed himself; in doing so, he abducts his shoulder and flexes his elbow but does not pick up his spoon. When the same man is asked to open a door, he also abducts his shoulder and flexes his elbow.


Measurement of Spasticity

Spasticity can be graded according to severity (mild, moderate, or severe) using both objective and subjective measures. However, as previously noted, clinicians must be careful when applying severity descriptors to muscle overactivity so as not to misrepresent their clinical impact to the patient. From a technical
standpoint, spasticity levels can be affected by many factors, including temperature, emotional status, time of day, level of pain, body position, and the amount of prior stretching. Given this potential variability, interpretation of serial measurements can be problematic.

A. Clinical Rating Scales

Hypertonia can be evaluated clinically using a number of well-established rating scales. The most commonly used are the Ashworth, Modified Ashworth, and Tardieu scales (Table 6–2), which are generally considered to have fair to good interrater and intrarater reliability. One criticism of the Ashworth and Modified Ashworth scales is their inability to distinguish between the rheologic properties of the soft tissues and the neural contributions to hypertonia. The Tardieu scale attempts to address this difficulty by measuring two angles: R1, the angle at which resistance is first encountered during a quick muscle stretch, and R2, the final angle, which reflects the maximum range of movement during a slow muscle stretch. The difference between the two is claimed to represent the true amount of spasticity, or spasticity angle.

Table 6–2  Commonly used spasticity rating scales.
B. Neurophysiologic and Other Tests

More sophisticated measures of hypertonia include neurophysiologic tests that attempt to quantify the muscle response to stretch (surface electromyographic activity, H-reflex response, the H-reflex standardized to the M-wave max, or the F-wave response) or instrumented measurements of stiffness and torque with accelerometers. The pendulum test is a biochemical method of assessing spasticity in a limb by extending the limb and then letting it swing freely against gravity. The oscillating pattern observed is mathematically assessed to obtain data such as time delay and muscle stretch reflex threshold, which can identify subtle changes in spasticity. One drawback to the use of the pendulum test is great variability when testing the same individual multiple times. This makes it less reliable when measuring treatment outcomes. Another issue is that force velocity and force length do not have a linear relationship. Therefore, positioning of the limb, muscle length, relaxation, and multiple other factors affect test reliability. The Wartenberg pendulum score is calculated during the gravity-induced pendulum-like movement of the lower limb as the ratio of joint angles measured by goniometry or computerized video motion analysis.

C. Subjective Measures

<table>
<thead>
<tr>
<th>Score</th>
<th>Ashworth Scale</th>
<th>Modified Ashworth Scale</th>
<th>Tardieu Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increased tone</td>
<td>No increased tone</td>
<td>No resistance to passive ROM</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in tone causing a “catch” when the limb is moved in flexion or extension</td>
<td>Slight increase in tone causing a catch and release or minimal resistance at the end range of the joint in flexion or extension</td>
<td>Slight resistance to passive ROM</td>
</tr>
<tr>
<td>1+</td>
<td>—</td>
<td>Slight increase in tone, with a catch, followed by minimal resistance throughout remainder (less than half) of ROM</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>Increased tone, with no difficulty moving limb into flexion</td>
<td>Increased tone throughout most ROM; affected part is still easy to move</td>
<td>Catch followed by a release</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in tone. Passive movement is difficult</td>
<td>Considerable increase in tone; passive movement is difficult</td>
<td>Fatigable clonus (&lt;10 sec)</td>
</tr>
<tr>
<td>4</td>
<td>Limb is rigid in flexion or extension</td>
<td>Affected part is rigid in flexion or extension</td>
<td>Infatigable clonus (&gt;10 sec)</td>
</tr>
</tbody>
</table>

ROM, range of motion.
Subjective measures include patient assessment of spasm intensity and logs of spasm frequency. There is an inconsistent correlation between subjective report and objective measures of spasticity.


**TREATMENT**

Spasticity can have beneficial or deleterious effects, and both may be noted in the same patient. Advantageous effects potentially include assistance with mobility, maintenance of posture, improvement of vascular circulation, preservation of muscle mass and bone mineral density, prevention of venous thrombosis, and assistance in reflexive bowel and bladder function. Conversely, spasticity can interfere with positioning, mobility, comfort, and hygiene. Spasticity has also been linked to increased metabolic demands, which can be problematic in the nutritionally compromised patient. Spontaneous spasms can interfere with sleep or duration of wheelchair use. Spasms can also lead to skin breakdown because of the shearing effect or impaired healing of surgical wounds due to tension along suture lines.

The relationship of spasticity to pain is complex. Spasticity can limit the range of motion about a joint, causing musculoskeletal pain. In this scenario, reduction of spasticity may decrease the pain associated with biomechanical limitations. However, central nervous system disease can also produce neuropathic pain, for which modulation of spasticity may not be effective in
reducing symptoms. Clinicians must therefore consider all aspects of a patient’s spasticity before embarking on a treatment plan. Rather than complete elimination of spasticity, a more realistic goal may be titration to maximize the risk–benefit ratio.

Rehabilitation Techniques

Therapeutic interventions are essential to the management of spasticity, both in isolation and in combination with other treatment modalities described in this chapter. A multitude of different techniques have been reported to modulate muscle overactivity, including range of motion exercises, stretching, therapeutic exercise, constraint-induced therapies, neurodevelopmental techniques, positioning, splinting, neuroprosthetics, serial casting, and functional electrical stimulation. Ideally, therapy will attempt to maximize the beneficial aspects of muscle overactivity and mitigate the detrimental aspects.

A. Stretching

Stretching is one of the primary strategies used by physical and occupational therapists for spasticity management. Static or low-velocity stretching has been the hallmark of this technique as applied manually by the treating therapist. Traditionally, a minimum of 30 seconds is recommended to achieve therapeutic benefit, with some data suggesting that longer duration stretches are more beneficial. Mechanical stretching devices such as dynamometers (Cybex, Kin-Com, Biodex, etc) are increasingly utilized. These devices allow for longer therapy sessions with programmed adjustability based on intelligent design biofeedback. Cost can be a limiting factor in the use of these devices, but this must be balanced against the operational benefit of freeing up clinician time. Several studies have shown positive effects of stretching, such as decreased motor activity on electromyography (EMG), greater range of motion, and decreased stiffness, but there is limited evidence of the long-term benefits of this intervention.

B. Casting

Serial casting involves the sequential application of casting material, either plaster or fiberglass, in a circumferential manner around the spastic joint. The area to be casted (eg, foot and ankle) is covered with a stocking, bony prominences are carefully padded, and the cast is applied at a comfortable end
range of motion. Casts are changed weekly at an enhanced joint angle. Serial casting is discontinued when no incremental increase in range of motion is seen on two sequential castings. Patients are then transitioned to a bivalved cast as a long-term maintenance strategy. The purported mechanism of spasticity reduction for this intervention is that casts minimize changes in muscle length and tension, thus reducing the excitatory input from muscle spindles. This diminished input, in turn, decreases the activity of the spinal reflex arc. Alternatively, casting might reduce sensory input with similarly diminished reflex activity. Potential complications of serial casting include skin breakdown, venous thrombosis, compartment syndrome, and regional decreases in bone mineral density.

**C. Splinting**

Splinting is a useful, noninvasive method for preventing contractures associated with spastic conditions and maintaining range of motion. A variety of prefabricated and constructed splints are available. Examples include resting hand splints, wrist cock-up splints, elbow extension splints, and posterior foot splints. Dynamic splints have a self-adjusting elastic component such as spring wire or rubber band and can serve both as passive assistance for movement of weaker muscles and as a resistant component for muscles with increased tone. Proprietary dynamic splinting devices (eg, Saeboflex, Dynasplints) are commonly used. Saeboflex was designed to allow training of grasp-and-release activities when a hemiplegic patient has upper extremity flexor hypertonicity with limited extensor activity. Dynasplints consist of padded cuffs and struts that are hinged at the joint axis. They allow tension and force to be adjusted across a given joint.

**D. Strength Training**

Perhaps the most commonly utilized rehabilitation modality is strength training. This technique is particularly important in the spastic patient given the potential for coexistence of muscle overactivity and weakness. Additionally, several approaches described later in this chapter have the potential to increase weakness.

Findings from several studies support the idea that the capacity for strength can be increased in individuals with central nervous system disease without exacerbation of spasticity. The parameters of strength training protocols are almost limitless, with no particular approach being preferential in the spastic patient. Although there is minimal evidence to suggest that strength training
reduces spasticity, the potential for comorbid weakness in this patient population mandates consideration of these techniques following other interventions.

E. Nerve Stimulation

Both functional electrical stimulation (FES) and transcutaneous electrical nerve stimulation (TENS) can be used to reduce pain and spasticity, improve muscle tone, and facilitate function in patients with upper motor neuron injury. In FES, an electrical current is applied to intact nerves of the body in order to generate a muscle contraction. Several studies have reported benefits after use of FES (improved range of motion, decreased spasticity, and improved foot drop) and TENS (improved motor function and spasticity) in post-stroke patients.


► Oral Pharmacotherapy

Five oral medications are frequently used to manage spasticity: baclofen, diazepam, dantrolene, clonidine, and tizanidine. Among other compounds being investigated for this indication are the cannabinoids. Currently available medications are best suited for patients with global or multisite muscle overactivity. (Additional discussion of agents commonly prescribed for spasticity appears in Chapter 10.)
A. Advantages of Therapy

The use of oral agents is noninvasive, does not require specialized technical expertise, and is generally considered a “first-line” approach. Most oral medications available for use in patients with spasticity are generic and thus inexpensive. In general, these pharmaceuticals are neither controlled nor scheduled medications. Other advantages include their utility as a breakthrough strategy (ie, they offer one of the only methods by which patients can exert a degree of self-control in an effort to manage irregular spasticity patterns), as well as for secondary indications such as pain, insomnia, epilepsy, and mood disorders.

B. Limitations of Therapy

Oral medications are poor choices for patients with focal or regional presentations of spasticity. The most significant issue relating to their use may be the lack of tolerability by patient populations that are prone to spasticity, particularly those with acquired cerebral injury. Potential adverse effects of individual medications are reviewed below, but clinicians should be aware that sedation, drowsiness, lethargy, and impairment in cognitive processing are concerns with many of these preparations. For the most part, oral spasticity medications have short half-lives, which necessitates multiple administrations per day. This can prove challenging for patients and caregivers.

C. Diazepam

Historically, the drug with the longest record in treating spasticity is diazepam, with an almost half century of clinical use. This agent is the prototypical molecule for the benzodiazepine class of medications. Benzodiazepines stimulate γ-aminobutyric acid A (GABA_A) receptors located primarily in the brainstem and spinal cord, resulting in enhanced presynaptic inhibition. All benzodiazepines are considered central nervous system depressants, with anxiolytic, hypnotic, antispastic, and antiepileptic properties. Half-life and sedative–hypnotic properties (ie, whether a particular medication is more sedative or more hypnotic) are the primary features used to distinguish among the various agents. Diazepam is a relatively long-acting GABA agent in contrast to shorter-acting benzodiazepines, such as oxazepam and lorazepam. Clinical efficacy for benzodiazepines has been reported in patients with multiple sclerosis and spinal cord injury. Pediatric use has also been described.

Central nervous system effects often limit the utility of benzodiazepines in
the treatment of spasticity, with sedation being reported as the most functionally disabling side effect. At higher doses, somnolence can progress to respiratory depression, coma, or death. The use of benzodiazepines can also lead to physiologic dependence, with the potential for a withdrawal syndrome if the medication is abruptly discontinued or tapered too rapidly.

D. Baclofen

Baclofen is perhaps the most commonly used oral medication in the treatment of spasticity. This agent, which is structurally similar to GABA, exerts an agonist effect on presynaptic GABA$_B$ receptors within the brainstem, dorsal horn of the spinal cord, and other sites in the central nervous system. The beneficial effects on spasticity are primarily mediated through depression of monosynaptic and polysynaptic spinal reflexes by blocking the release of neurotransmitters. However, the drug’s action on supraspinal receptors may lead to significant side effects. Baclofen has been demonstrated to be most effective in patients with spasticity of spinal origin, including spinal cord injury, and multiple sclerosis.

When administered orally, baclofen is absorbed through a relatively small section of the upper portion of the small intestine. The mean half-life of oral baclofen is 3.5 hours. The drug is primarily excreted unchanged by the kidneys, with a lesser amount (6–15%) being metabolized by the liver. Side effects include sedation, confusion, dizziness, and nausea. Overdosage may lead to respiratory suppression, hypotension, bradycardia, and unresponsiveness. Abrupt withdrawal may lead to seizures, mental status changes, or a clinical picture consistent with neuroleptic malignant syndrome, which, if untreated, may lead to death. Significant adverse effects have been demonstrated in both an elderly stroke population and a mixed population of patients with acquired brain injury. Baclofen appears to lower seizure threshold, regardless of whether the dose is increased or decreased. Thus, this medication should be used cautiously in patients with a history of seizures.

In light of the drug’s limited gastrointestinal absorption and short plasma half-life, patients must often self-administer baclofen several times per day. Efforts are ongoing to circumvent this problem using prodrug technology. Baclofen is the racemic mixture of mirror-image isomers. The R isomer is considered the more pharmacologically active molecule. R-baclofen has been reformulated with a carrier molecule that affords enhanced absorption through the gastrointestinal tract. The net result is reduction in spasticity using only twice-daily dosing. A small clinical trial has demonstrated reduced spasticity in individuals with spinal cord injury, and there is an ongoing clinical trial
involving use of the modified drug in individuals with multiple sclerosis.

**E. Tizanidine**

Another commonly used oral medication is tizanidine. This molecule is a selective $\alpha_2$-adrenergic receptor agonist that is structurally similar to clonidine. Tizanidine is active at both spinal and supraspinal levels. At the spinal level, it reduces presynaptic interneuronal activity by inhibiting the release of excitatory amino acids and substance P. In the brain, tizanidine inhibits firing of neurons within the locus ceruleus. This effect downregulates the facilitory influence of the cerebrospinal spinal tract. The effect on spinal substance P and the supraspinal effects are the proposed mechanisms of the potential analgesic effects for tizanidine.

Tizanidine is generally well tolerated; the most frequently reported side effects are dry mouth, drowsiness, dizziness, and fatigue. Comparative studies indicate that its efficacy is similar to that of baclofen and diazepam, with less patient discontinuance due to adverse events. The most common adverse effects are sedation, dizziness, and dry mouth. Approximately 5% of patients who take the medication on a long-term basis have elevated hepatic enzymes on serum testing. This abnormality typically resolves with either drug discontinuance or dosing reduction. Routine surveillance of hepatic enzymes is recommended.

**F. Clonidine**

Like tizanidine, clonidine is an $\alpha_2$-adrenergic receptor agonist. Its antispastic effect is attributed to presynaptic inhibition of sensory afferents. This same mechanism is also postulated to result in the drug’s antinociceptive properties. In contrast, its antihypertensive effects are thought to be due to central sympatholytic properties. Clonidine is highly lipophilic, which allows for multiple routes of administration (ie, oral, transdermal, intravenous, rectal, epidural, and intrathecal).

**G. Dantrolene**

Dantrolene is a peripherally acting antispasticity agent that decreases release of calcium from the sarcoplasmic reticulum of skeletal muscle cells. This results in an uncoupling of muscle contraction from electrical neuronal impulses. Dantrolene is specific for skeletal muscle and thus preferentially affects reflexive contractions (spasticity) rather than voluntary muscle movement. Hepatotoxicity is a significant limitation; approximately 1% of patients treated with this agent
develop significant liver abnormalities. This risk is elevated at dosages greater than 400 mg/day (the manufacturer’s maximum recommended dosage). Dantrolene has demonstrated efficacy in a number of patient populations, including those with spinal cord injury, cerebral palsy, and multiple sclerosis. The peripheral nature of this compound makes it particularly attractive in the treatment of spasticity after brain injury. Pediatric use has also been described.

**H. Investigatory Agents**

Researchers have shown considerable interest in modulation of spasticity via the endocannabinoid system. Type 1 cannabinoid receptors (CB1) are expressed primarily in the presynaptic membrane of the neurons and use a negative feedback mechanism to regulate transmitter release at GABAergic, glutamatergic, and dopaminergic synapses. Agonists at CB1 have produced antispastic effects in experimental animal studies. The most important cannabinoids for pharmacologic use are Δ-9-tetrahydrocannabinol (THC) and cannabidiol (CBD). Sativex, commercially available outside the United States, is a plant extract that contains THC and CBD in defined quantities. It is administered as an oromucosal spray. It is approved in many countries for treatment of spasticity in patients with multiple sclerosis for whom conventional antispastic therapy has not been effective. There are ongoing clinical trials of this preparation in the United States.


Botulinum Toxin Therapy

Perhaps no technique has affected the management of spasticity more than the introduction of focal neurolysis with botulinum toxin, a protein and neurotoxin produced by the anaerobic bacterium *Clostridium botulinum*. Type A and B toxins are currently utilized for medical purposes. Both consist of so-called heavy chains (100 kDa) joined by a single disulfide bond to a light chain (50 kDa). The light chain is the toxic portion of the molecule. Botulinum toxin blocks neurotransmitter release at peripheral cholinergic nerve terminals, resulting in decoupling at the neuromuscular junction. Blockage is achieved in four distinct steps: (1) binding to receptors on the presynaptic membrane, (2) endocytotic uptake into nerve terminals, (3) translocation across the endosomal membrane, and (4) inhibition of neurotransmitter endocytosis. The molecular targets for this last step are different for each serotype. Type A toxin cleaves a 25-kDa synaptosomal-associated protein (SNAP-25) to prevent binding of synaptic vesicles to the presynaptic membrane, whereas type B toxin cleaves synaptobrevin, also known as vesicle-associated membrane protein (VAMP).

Botulinum toxin injections are used in the management of spasticity to reduce the force created by the contraction of the overactive muscle or muscle groups. A reduction in muscle tension can lead to improvement in both passive and active range of motion, as well as permitting a more successful outcome with stretching techniques, thus facilitating use of the bracing, splinting, and casting techniques discussed previously. Additionally, patients may experience improved motor control following injection of the toxin. Given these results, it is of paramount importance to couple toxin injections with a rehabilitation protocol whether the goal is improved passive or active function.

A. Preparations

Among the type A serotypes are different formulations with different standard doses. All botulinum toxin type A products have the same 150-kDa molecule at their core. Abobotulinumtoxin A surrounds this molecule with a 350-kDa to 750-kDa shell of complexing proteins, whereas onabotulinumtoxin A uses a 750-kDa protein complex for this purpose. Incobotulinumtoxin A has no complexing proteins. The clinical significance of these complexing proteins is not clear at present, and it would appear that they dissociate from the core molecule quickly upon dilution with saline. It is vitally important to recognize that the biologic activity units are unique to each toxin preparation and cannot be converted (or compared) to each other. Table 6–3 lists botulinum neurotoxins commercially

<table>
<thead>
<tr>
<th>Table 6–3</th>
<th>Botulinum neurotoxins commercially available</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Preparations</td>
<td></td>
</tr>
<tr>
<td>Abobotulinumtoxin A</td>
<td>350-kDa to 750-kDa complexing proteins</td>
</tr>
<tr>
<td>Onabotulinumtoxin A</td>
<td>750-kDa protein complex</td>
</tr>
<tr>
<td>Incobotulinumtoxin A</td>
<td>No complexing proteins</td>
</tr>
</tbody>
</table>
available in the United States. Only abobotulinumtoxin A has received a formal Food and Drug Administration (FDA) indication for use in adult patients with upper extremity spasticity although there are ongoing clinical trials involving other preparations and indications.

Table 6–3 Botulinum toxins commercially available in the United States.

<table>
<thead>
<tr>
<th>Serotype</th>
<th>Brand Name</th>
<th>FDA Recommended Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Botox</td>
<td>Onabotulinumtoxin A</td>
</tr>
<tr>
<td>A</td>
<td>Dysport</td>
<td>Abobotulinumtoxin A</td>
</tr>
<tr>
<td>A</td>
<td>Xeomin</td>
<td>Incobotulinum A</td>
</tr>
<tr>
<td>B</td>
<td>Myobloc</td>
<td>Rimabotulinumtoxin B</td>
</tr>
</tbody>
</table>

B. Administration

For botulinum toxin to be effective, it must injected directly into the overactive contracting muscle. Various techniques are available to facilitate muscle localization. Some clinicians use clinical judgment along with visual inspection and palpation to guide toxin injection. Others rely on EMG for guidance. This latter technique involves insertion of a needle with an insulated outer core and an electrically conductive inner core that is only exposed at the needle tip. The needle tip functions as a probe through which EMG signals can be recorded. Once excessive EMG activity is detected, the toxin is injected. Electrical stimulation can be applied through this type of needle, as well. The most common technique involves nerve stimulation capable of delivering 0.1–0.5-millisecond duration pulses once or twice per second. The stimulator should have a rheostat to control current flow. An ammeter (which measures electrical current) facilitates precise localization as motor points will respond to much lower amperage compared with surrounding muscle or soft tissue. More recently, ultrasound has been used by some clinicians to facilitate muscle localization. One advantage of this method is the ability to identify vascular structures using the Doppler mode of ultrasound machine. The choice of localization technique should be based on local expertise and resources. A recent consensus statement noted that there is insufficient evidence to support or refute the use of one
localization technique over another.

The selection of muscles to be injected by botulinum toxins is typically based on the spastic presentation in the particular patient. Although presentations of excessive muscle contraction vary, several common synergies are observed in both the upper and lower extremities. In the upper extremity, these include the adducted and internally rotated shoulder, flexed elbow, flexed wrist, pronated forearm, thumb in palm deformity, and clenched hand. In the lower extremity, common representations include the flexed hip, adducted thighs, extended or flexed knee, hyperextended first toe, and equinovarus foot. Also common is a synergy in which the heel is turned toward the midline, the foot is plantar flexed, the inner border of the foot is supinated, and the anterior foot is displaced medial to the vertical axis. Toe curling or clawing can coexist with the equinovarus deformity. Although clinical observation and physical examination are most often used to determine which muscle should be injected, laboratory evaluation of gait and motor control using dynamic multichannel electromyography along with kinetic and kinematic data can enhance the selection process by identifying the relative contributions of the potential muscles or muscle groups.

Several factors influence the dosing of botulinum toxin, including the patient’s prior response to injections, amount of tone, residual function of the spastic muscle, and potential impact of excessive tonal reduction. Readers are referred elsewhere for further information about dosing and muscle selection strategies.

C. Adverse Reactions

Adverse effects of botulinum toxin therapy can be grouped into three broad categories. First, diffusion of the toxin away from the intended sites of action can lead to unwanted inhibition of transmission at neighboring nerve endings. Although localized diffusion is undesirable, it is rarely a serious problem. More worrisome is distal diffusion to muscles controlling respiration and swallowing. In 2008, the FDA reported that botulinum toxins have “been linked in some cases to adverse reactions, including respiratory failure and death, following treatment of a variety of conditions using a wide range of doses.” Several of these cases involved pediatric use, where weight-based dosing strategies must be considered. Administration of botulinum toxins with other agents that affect neuromuscular function, such as aminoglycosides, may increase the effect of botulinum toxin. Second, sustained blockade of transmission can produce effects similar to anatomic denervation, including muscle atrophy. The third undesirable effect is immunoresistance to botulinum toxins. Resistance results from the
development of circulating antibodies that bind to the heavy chain and prevent its association with nerve membranes, thus preventing internalization of the functional active light chain. Auxiliary proteins in the toxin complex theoretically could promote the immune response to the toxin. Bruising or bleeding at the injection site is a consequence not of the toxin, but rather of the injection procedure. Clinicians should be sensitive to this consequence in patients who are using antiplatelet or anticoagulant therapy. Other adverse events include headaches, flulike syndromes, blurred vision, dry mouth, and fatigue.

D. Special Considerations

As previously noted, the use of botulinum toxin should always be coupled in some way with a rehabilitation protocol. This can be facilitated by a formal referral to physical or occupational therapy or by instructions to the patient or caregiver regarding a home exercise program. For patients in whom only passive function is the treatment goal, slow or static stretching techniques should be applied either manually, through range-of-motion devices, or by the application of an orthotic device. For patients whose goals include active function, therapeutic exercise such as constraint-induced movement therapy, unloaded cycling, body-weight–supported treadmill training, and strengthening exercises are appropriate. With regard to other treatment modalities, a growing body of literature suggests that botulinum toxin effects are enhanced when combined with electrical stimulation.

Botulinum toxin therapy is an effective treatment for muscle overactivity, but the duration of effect is limited. The duration of neurolytic blockade varies with the type of neurotoxin and the type of nerve terminal. At the human neuromuscular junction the effect typically lasts from 2 to 4 months; in the autonomic nervous system, it may last for more than 1 year.


Intrathecal Baclofen Therapy

Intrathecal baclofen (ITB) therapy is a powerful technique for management of spastic hypertonia. ITB infusion exerts its therapeutic effect by delivering baclofen directly into the cerebrospinal fluid, thus affording enhanced
distribution of this agent to target neurons in the spinal cord. Before proceeding with this therapy, appropriate candidates need to be counseled that this treatment modality involves an invasive procedure and represents a long-term commitment. In general, patients can be considered candidates for ITB therapy when:

- Spasticity is poorly controlled despite maximal therapy with other modalities.
- Spasticity is poorly controlled because of limited patient tolerance of other modalities.
- Adjustable spasticity reduction afforded by a programmable variable flow pump would be advantageous.

The FDA-approved indications for ITB therapy include spasticity of spinal (traumatic spinal cord injury and multiple sclerosis) and cerebral origin (acquired brain injury, cerebral palsy, and stroke). Efficacy of this therapy has also been shown for degenerative conditions of the brain and spinal cord (ie, amyotrophic lateral sclerosis, hereditary spastic paraparesis).

A. Initiation of Therapy

1. Intrathecal trial of medication—An intrathecal trial is performed to expose the patient to the potential agents that will be administered on a more permanent basis upon pump implantation. The typical procedure for an ITB trial is to perform a lumbar puncture and inject a bolus dose of a baclofen solution into the cerebrospinal fluid. The most commonly used initial screening dose is 50 mcg of baclofen. Clinical effects from a screening bolus are seen within 1–3 hours postinjection, and peak effects are typically observed 4–6 hours postinjection. The effects of the screening bolus are always temporary, routinely lasting 6–8 hours; however, prolonged effects of a single test bolus have been reported. Screening boluses can be repeated if the initial injection is unsuccessful. Sequential assessments every few hours are warranted to evaluate the onset, peak, and resolution of ITB effects. Intrathecal trials can potentially differentiate range-of-motion deficits due to severe spasticity, which are potentially reversible without surgery, from fixed contractures.

2. Pump implantation—Once a positive trial response is observed, a patient may proceed to pump implantation. The initial dosage of ITB is often determined by the patient’s response to the test dose. A reasonable starting dose is 100–200% of the bolus dose divided over a 24-hour period. If a patient has
demonstrated prolonged or excessive hypotonia during the screening phase, it may be prudent to start at 50% of the bolus dose divided over a 24-hour period. It is imperative that the implanting physician communicate closely with the physician responsible for the trial when determining the appropriate starting dose. The patient should continue all oral antispasticity medications until a weaning schedule is prescribed.

3. Dosage modification—Dose adjustments can commence immediately after pump implantation. In general, it is reasonable to wait 24 hours between each dosing adjustment to allow for the full effects of the drug to be observed. During the titration phase of ITB therapy, patients are usually weaned from oral antispasticity medications. The amount of each intrathecal adjustment varies based on patient tolerability. Nonambulatory patients may tolerate dose adjustments of 20% of total daily dose, whereas others, especially ambulatory patients, will require lower titration increments (5–10%). Adverse effects that may be seen during this phase of therapy include excessive hypotonia, changes in bowel and bladder status, and increased thromboembolic risk. The frequency and size of dosing adjustments should be individualized based on the patient’s response to prior changes.

4. Postimplantation rehabilitation—If ITB is anticipated to affect the patient’s active functional status, a rehabilitation program after implantation is appropriate. Postimplant rehabilitation may also be needed to ensure appropriate caregiver training. Disciplines that may be involved in the rehabilitation process include physiatry, physical therapy, occupational therapy, and rehabilitation nursing. Issues that may require attention include incisional care, medical management (eg, spinal headache, pain assessment, medication adjustment, dosing changes), mobility, self-care ability, and bowel and bladder function. Patients, especially ambulatory individuals, should be thoroughly counseled on the need for postimplant rehabilitation in order to maximize the benefits of ITB therapy.

B. Maintenance Therapy
Following the titration phase of ITB therapy, the patient enters the maintenance phase of therapy. Aspects of this phase include refilling the pump reservoir with new medication, troubleshooting any infusion system malfunction, and replacing the pump for battery replenishment. Reservoir refills are a sterile, office-based procedure that occur every few weeks to few months. Standard baclofen
solutions are stable in the pump reservoir for up to 6 months. The pump has a low residual reservoir volume, which is the lowest volume that supports stable flow. The refill interval is the time required for the pump to dispense the volume of solution from a full reservoir to the low reservoir volume. The refill interval reflects the baclofen concentration and daily dose. Pump refills are scheduled to have sufficient residual reservoir volume prior to the alarm date to avoid “low reservoir syndrome” and associated symptoms of ITB withdrawal. Pump refills are typically accomplished by palpating for the pump externally and using a template to guide a needle into the reservoir chamber. Fluoroscopy or ultrasound can be used to assist in port localization and needle placement.

For patients with chronic, nonprogressive neurologic conditions, ITB dosing should be relatively stable during the maintenance phase of therapy. Individuals with progressive diseases such as amyotrophic lateral sclerosis or multiple sclerosis may require increasing dosages over time. Patients with previously well-controlled hypertonia on a stable dosing regimen who present with increased spasticity should be examined carefully. Although pharmacologic tolerance to ITB might necessitate increased dosing, this phenomena should not be assumed until a thorough clinical evaluation has been performed. Comorbidities of neurologic disease can serve as noxious stimuli that act as “triggers” for increased spasticity (eg, urinary tract infection, bladder distention, urolithiasis). If no cause for increased spasticity is discovered, the search for a system malfunction should be promptly undertaken, as described below.

C. Potential Complications

1. Pump- and catheter-related problems—Potential causes for loss of effectiveness of ITB therapy include programming errors and mechanical problems involving the pump or catheter (eg, kinks, holes, occlusions). Some of these problems are readily detectable; discovery of others is more challenging. In general, programming and refilling errors tend to be easily identified and corrected. Malfunctions involving the pump mechanism are rare but when present are also rather simply confirmed. Catheter problems are relatively frequent and may vary as to presentation and ease of diagnostic identification. Prompt identification of any delivery system problems is imperative given the potential serious effects of ITB overdose and withdrawal.

Two initial techniques for investigating pump malfunction are pump “interrogation” and checking of the pump residual reservoir volume. Interrogation of the pump’s dosing parameters should match the prescribed dosing. The presence of an audible alarm (as well as an electronic alarm during
pump interrogation) or discovery of an unexpected “extra” residual volume in the reservoir suggests a pump-related malfunction. Alarm conditions generally occur in either a low battery or low reservoir situation. A low battery alarm will sound when the battery has reached significant discharge. A low reservoir alarm will indicate that the pump has delivered nearly all of the contents of the reservoir and that the patient needs an immediate refill. An increase in reservoir residual volume might indicate an abnormality of the pump rotor. The presence of a permanent rotor stall or low battery condition should prompt urgent replacement of the pump.

Once mechanical pump abnormalities and programming or solution difficulties have been eliminated from consideration, attention turns to identifying potential catheter problems. Most often this involves imaging techniques. Plain radiography is a typical first step. If the films are normal, a catheter access port aspiration can be undertaken. This procedure involves accessing a port that is in direct continuity with the catheter (the catheter access port [CAP]). Because the distal end of the catheter lies within the subarachnoid space, the investigator should be able to readily withdraw cerebrospinal fluid through the catheter. Aspiration of at least 2–3 mL is necessary for determination of a “normal” aspiration since the volume of the catheter is typically less than 0.25 mL. Failure to aspirate fluid can suggest catheter disruption or occlusion, keeping in mind that some neurologic patients may produce minimal cerebrospinal fluid in the region of the catheter tip. Once the catheter has been cleared of the drug solution and cerebrospinal fluid has been obtained, contrast medium can be injected and visualized fluoroscopically or with computed tomography. Extravasation of dye from the catheter can be used to diagnose catheter breaks, catheter tip loculations, and migration of the catheter into the subdural or epidural spaces.

2. Withdrawal syndrome—Abrupt reduction of ITB delivery can result in a withdrawal syndrome that can have serious, if not fatal, consequences. The severity of ITB withdrawal syndrome is not consistently related with dosing levels. Perhaps the most common symptomatic presentation is return of the patient’s “baseline” degree of hypertonia. Additional characteristics of this syndrome can include pruritus, seizures, hallucinations, and autonomic dysreflexia. Some patients demonstrate a life-threatening syndrome exemplified by exaggerated or rebound spasticity (ie, greater than baseline degree of hypertonia), fever, hemodynamic instability, and altered mental status. If not treated aggressively, this syndrome can progress over 24–72 hours to include rhabdomyolysis, multiorgan system failure, and, rarely, death. Following
recognition of ITB withdrawal, initial treatment includes supportive care, careful observation, and replacement of baclofen via enteral route or preferably through restoration of intrathecal delivery. Adjunctive pharmacotherapy can also include administration of benzodiazepines or cyproheptadine.

3. **Overdose**—In contrast to withdrawal, which can occur despite vigilant attention, ITB overdose is generally due to human miscalculation during dosing adjustments or concentration changes. Symptoms of overdose include profound hypotonia or flaccidity, hyporeflexia, respiratory depression, apnea, seizures, coma, autonomic instability, hallucinations, hypothermia, and cardiac rhythm abnormalities. Initial management includes maintenance of airway, respiration, and circulation. Secondary measures include reduction or temporary interruption of intrathecal delivery by pump reprogramming. Optional measures for ITB overdose include cerebrospinal fluid drainage via CAP aspiration or lumbar puncture, and administration of an “antidote.” While not true antidotes, both physostigmine and flumazenil have been reported to reduce central side effects, such as somnolence and respiratory depression. Physostigmine is the more commonly utilized agent but may produce adverse effects such as bradycardia, seizures, and increased respiratory secretions.


Orthopedic Management

Orthopedic interventions for correction of spastic deformities have been described for over 170 years and may represent the oldest form of spasticity management. Their primary role is to address the biomechanical aspects of spasticity that have not been adequately addressed by other measures. As with all treatment plans, the potential objectives of orthopedic management must always be well defined for both patients and caregivers. Goals include correction of musculoskeletal misalignments, deformities, and contractures. It is crucial before undertaking any orthopedic interventions that patients and caregivers understand the difference between contracture, which is a fixed, static shortening of the musculotendinous unit, and spasticity, which is dynamic abnormality of the central nervous system. Orthopedic interventions should be undertaken in individuals who demonstrate biomechanical musculoskeletal abnormalities with associated functional impairments (eg, gait abnormalities, problems in carrying out activities of daily living) or pain. “Asymptomatic” deviations of alignment should be monitored for progression or treated with more conservative measures such as stretching, bracing, physical therapy, and so on, before proceeding to surgical interventions.

Orthopedic interventions for the spastic patient generally fall into four main categories: musculotendinous lengthening, tendon transfer, osteotomy, and arthrodesis. It is not uncommon for multiple procedures to be undertaken simultaneously.

A. Muscle Lengthening Techniques

Lengthening a muscle or tendon can be accomplished using any of the following techniques. One can lengthen through the tendon in a Z-type fashion, sometimes referred to as the half-section splitting-and-gliding method. In this procedure, a vertical or longitudinal cut is made in the center of the tendon equal to the length the tendon will be extended. A transverse cut is then made at each end of the incision on the opposite sides of the tendon, producing an elongated Z pattern. Sutures can then reunite the ends of the tendon. This technique likely provides a
greater gain in the range of motion but may be associated with overlengthening of the tendon and shortening of the muscle. A second and perhaps more favored technique is an intramuscular or aponeurotic lengthening release through the tendinous portion at the musculotendinous junction. Repair is not necessary, and the chances of overlengthening the muscle are minimized; however, the chance of recurrence is higher. Lastly, a resection can release the tendon at its insertion, with resuturing to proximal soft tissues or bone. Selective surgical lengthening techniques that minimize the subsequent weakness of the muscle tendon unit are favored. Muscle lengthening has the added benefit of altering activity at the Golgi receptors within. This results in a temporary decrease in spasticity as well as achieving a functional muscle lengthening. The specific goal of the surgery and a patient’s current function help to dictate which procedure is chosen.

B. Tendon Transfer

Tendon transfers attempts to utilize increased muscle tone in a beneficial manner by moving a muscle tendon unit with spasticity to an antagonist group that possesses excessive weakness. For instance, a patient who demonstrates excessive knee extension would undergo a procedure in which the rectus femoris is transferred to the hamstrings. Similarly, correction of a varus foot deformity could be achieved by using a split transfer of either the spastic posterior tibialis or the anterior tibialis to the lateral foot.

C. Osteotomy

Osteotomies, which are incisions or transections of a bone, are useful in many orthopedic procedures to correct misalignment and have added benefit in the spastic patient. This approach relies on the concept of “lever-arm dysfunction,” which was initially described by Gage and colleagues. This problem, discussed in detail in Chapter 4, is defined as a disruption in the moment generation of a muscle joint complex because of an ineffective lever arm. The result of lever arm dysfunction is functional weakness and decreased power production. Gage described five distinct types of lever-arm deformities: (1) short lever-arm, (2) flexible lever-arm, (3) malrotated lever-arm, (4) an abnormal pivot or action point, and (5) positional lever-arm dysfunction. Osteotomies not only attempt to correct the bony malalignment but also to position the surrounding muscles in a more efficient biomechanical position. Perhaps, the most commonly cited example of this approach is a derotational osteotomy of the proximal femur. Increased femoral anteversion often occurs in children with spastic cerebral palsy because of excessive hip adductor spasticity. The osteotomy provides a
more normal alignment of the hip joint and enables muscles that cross the hip joint to contract along a more appropriate force line.

**D. Arthrodesis**

The most permanent option in orthopedic management of the spastic patient is arthrodesis (ie, fusion of a joint). For some neurologically based deformities, dynamic correction of certain deformities is not likely to be successful. Common examples include triple arthrodesis (fusion of the talocalcaneal, talonavicular, and calcaneocuboid joints) to correct bony hindfoot deformities, arthrodesis of the first metatarsal phalangeal joint for hallux valgus or dorsal bunions, and arthrodesis of the interphalanges for valgus or flexion deformity of the great toe.

**E. Special Considerations**

Orthopedic interventions can be undertaken in both children and adults, although the prevalence of such undertakings is much higher in the pediatric age group. As outlined earlier, spasticity is present in several neurologic conditions, including cerebral palsy, brain injury, spinal cord injury, spinal cord disease, stroke, and multiple sclerosis. Among these conditions, the majority of reports describe orthopedic inventions in individuals with cerebral palsy. Perhaps an order of magnitude less are reports outlining orthopedic management of the abnormal distal lower extremities in patients with stroke or brain injury. There are also a number of less commonly cited examples. Muscle releases of the elbow flexors and the shoulder adductors–internal rotators can improve range of motion and decrease pain in patients with stroke or brain injury. Clearly, there is considerable opportunity for continued expansion of orthopedic interventions for all spastic patients regardless of age or diagnosis.


Keenan MA: The management of spastic equinovarus deformity following


Neurosurgical Management

The primary aim of neurosurgical procedures for spasticity reduction is to interrupt the stretch reflex or increase the inhibitory influence on motor pathways in the spinal cord. These interventions can be grouped into peripheral procedures (rhizotomy, neurectomy, etc) and central procedures (cordotomy, myelotomy, or neuromodulation techniques). The development of other therapies described earlier in this chapter has resulted in decreased use of neurosurgical interventions for spasticity. The primary neuromodulation technique currently employed is ITB therapy (see earlier discussion). When attempting neurosurgical interventions for spasticity management, it is of paramount importance to recognize that the upper motor neuron syndrome presents with both positive and negative symptoms. Careful clinical evaluation and patient selection are crucial in these procedures to maximize spasticity reduction without creating new deficits or exacerbating preexisting deficits.

The afferent limb of the stretch reflex includes the skeletal muscle spindle and its afferent fibers. These neural structures travel proximally in the peripheral nerve and ultimately into the posterior spinal root of the spinal cord, where they synapse with the motor neurons arising from the anterior horn. Posterior rhizotomy interrupts the afferent limb of the stretch reflex at the posterior spinal nerve root or rootlet level. This procedure arose from Sherrington’s classic findings that demonstrated reduction in hypertonicity following sectioning of
posterior nerve roots in decerebrate cats. In the 1970s, Fasano and colleagues noted that certain posterior rootlets show an abnormal response to electrical stimulation in spastic patients. Rootlets that responded to a train of electrical stimuli with the expected brief muscular contraction were spared, whereas others, which produced an abnormal, continuous, or prolonged contraction that sometimes spread to other muscle groups, were divided. These researchers identified children with spastic cerebral palsy not including dystonia or significant preexisting weakness as the best candidates for selective posterior rhizotomy. In the early 1980s, using a similar approach, Laitinen and colleagues had encouraging results in a group of adults with spasticity from spinal cord injury or multiple sclerosis. Currently intraoperative EMG assessment and aggressive postoperative rehabilitation are considered the standard care in this procedure. Potential complications associated with selective posterior rhizotomy include increased weakness; sensory loss; inadequate relief of spasticity; sexual, bowel, or bladder dysfunction; spinal deformity; infection; urinary tract infection or cystitis; hemorrhage; and cerebrospinal fluid leakage.

Alternatively, the efferent limb of the spinal reflex arc (the motor neuron that arises from the anterior horn and runs into the anterior spinal nerve root and distally into the appropriate peripheral nerve) can also be interrupted surgically. Various sites have been dissected, including anterior rhizotomy and peripheral neurectomy. In general, these approaches have been abandoned because of the resulting flaccid paralysis and atrophy along with the possibility of dysesthesias (in the case of peripheral neurectomy involving mixed sensory and motor nerves). In the upper extremities, neurectomy of the musculocutaneous nerve has been used successfully for relief of spastic elbow flexion with microsurgical technique with nerve stimulation for selective ablation of individual nerve fascicles. In the lower extremities, peripheral neurectomies may be performed at a number of sites, including the obturator nerve, tibial nerve, posterior tibial nerve, and sciatic nerve. Some series report a fairly high rate of recurrence with these procedures.

Central neurolytic procedures are performed infrequently because of the greater risk of complications. McCarty and Kiefer described cordectomy in 1949. This procedure was accompanied by complete motor and sensory loss, worsened bladder function, and is no longer performed for the treatment of spasticity. Bischof introduced longitudinal myelotomy in 1951 and later modified the procedure from a lateral to a dorsal longitudinal myelotomy to avoid damaging the descending motor pathways. This procedure has been advocated for complete or near-complete spinal diseases with severe, intractable,
bilateral lower extremity spasticity. Lastly, Sindou described a modification of selective posterior rhizotomy in which afferent fibers are dissected as they enter the posterolateral sulcus (also known as the dorsal root entry zone [DREZ]). The DREZotomy procedure destroys nociceptive and myotactic fibers, sparing the dorsal column-medial lemniscus pathway. The result is disruption of the spinal reflex arc.


Neurogenic Bowel & Bladder

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NEUROGENIC BOWEL

ESSENTIALS OF DIAGNOSIS

► Inability to control the bowels.
► Difficulty evacuating the bowels.
► Increased bowel incontinence or constipation.

General Considerations

The term neurogenic bowel is used to describe alterations in the functioning of the gastrointestinal (GI) system of patients after a neurologic injury. These alterations are of concern because of issues of bowel control and, ultimately, the fear and embarrassment that accompany them. The greatest change in GI function after a spinal cord injury (SCI) involves bowel evacuation. In contrast to the challenge facing individuals with bladder incontinence (discussed later in this chapter), continence with a neurogenic bowel is achievable for most individuals, especially when the patient and family are highly motivated and disciplined.
A. Epidemiology
Fecal incontinence and fecal impaction occur in 0.3–5% of the general population, and this rate may be as high as 10–50% in older or hospitalized patients. Studies have reported that quality of life is affected by fecal incontinence in 62% of patients with SCI. In addition, of the population sampled 39% reported constipation, 36% reported hemorrhoids, and 31% reported abdominal distention 1 year post-SCI.

B. Neuroanatomy and Function of the Bowel
The colon is responsible for fecal formation, storage, and defecation. The colon and anorectal mechanisms receive parasympathetic, sympathetic, and somatic innervations. Between the smooth muscle layers and under the mucosa of the colon is the intrinsic enteric nervous system (ENS), composed of Auerbach’s (myenteric) plexus and Meissner’s (submucosal) plexus. Auerbach’s plexus is involved primarily in motor control and Meissner’s plexus, primarily in sensory control. Although in neurogenic bowel there is loss of direct somatic sensory or motor control, with or without sympathetic and parasympathetic innervation, the ENS usually remains intact.

1. Autonomic nervous system control—GI tract motility is controlled via the parasympathetic nervous system through its ability to enhance the motility of the colon. Stimulation is received from the vagus nerve and the splanchnic (pelvic) nerves. The vagus nerve innervates the ascending to mid transverse colon; the splanchnic nerves originating from S2 to S4 innervate the descending colon and rectal region.

2. Somatic nervous system control—The somatic nervous system controls external anal sphincter tone, helping to promote continence.

3. Enteric system control—The ENS typically is thought of as the key to proper functioning of the entire GI tract. Meissner’s and Auerbach’s plexuses contain an estimated 10–100 million neurons. Segment-to-segment coordination within the GI tract is largely regulated by the ENS; because of this, the ENS is sometimes called the “third part” of the autonomic nervous system. It has its own nerve–blood barrier, similar to that of the central nervous system (CNS).

    Although the ENS can function autonomously, it usually communicates with the CNS through the parasympathetic and sympathetic nervous systems. The ENS is composed of efferent neurons, afferent neurons, and interneurons, which
together allow it to carry reflexes and act as an integrating center, regardless of whether CNS input is available.

4. Anal region control—In neurologically intact individuals, the internal anal sphincter relaxes with the filling of the rectum. It is composed of smooth muscle and is under the influence of the sympathetic nervous system (T11–L2). The external anal sphincter helps to maintain continence by increasing its tone. It is made up of a circular band of striated skeletal muscle and is controlled by the pudendal nerve (S2–S4).

Within a neurologically intact individual, the combined actions of the internal and external anal sphincters allow for the storage of stool. Once a certain volume is attained, rectosigmoid distention causes reflex internal anal sphincter relaxation. Volitional cortical activity sends a signal to the pontine defecation center, which in turn allows contraction of the levator ani muscles. This activity opens the proximal canal and relaxes the external anal sphincter and puborectalis muscles. During this time, reflexive rectal propulsive contractions also help with the evacuation of stool.

Clinical Findings

General symptoms and signs in individuals with a neurogenic bowel include fecal incontinence, difficulty with evacuation, associated neurologic bladder symptoms, and associated symptoms of autonomic dysreflexia in patients with spinal cord lesions at the level of T6 and above.

A spinal cord lesion that occurs above the conus medullaris is considered an upper motor neuron (UMN) lesion and typically manifests as underactive propulsive peristalsis, overactive segmental peristalsis, or bowel distention. A lesion occurring at the level of the conus medullaris, cauda equina, or inferior splanchnic nerve is considered a lower motor neuron (LMN) lesion and typically manifests as colonic slowing. This results in constipation, fecal incontinence, and difficulty with the act of voiding.

The history and physical examination provide essential information when evaluating patients who may be experiencing bowel dysfunction. A complete American Spinal Injury Association (ASIA) examination is always recommended during this process. (For additional information, see Chapter 12.)

A. Upper Motor Neuron Lesions

Patients who have a UMN lesion (above the conus medullaris) manifest the
following symptoms: increased transit time, hyperreflexia leading to spasms that cause frequent defecation, low rectal capacity (because the nerves react to only a smaller volume of stool), and reduced or no control of the external sphincter.

Spinal cortical sensory pathway deficits lead to a decreased ability to sense the urge to defecate. Most people with UMN neurogenic bowel, however, have a vague feeling of discomfort when excessive rectal or colonic distention is present. It is thought that these sensations may be mediated by autonomic nervous system fibers that bypass the zone of SCI by way of the paraspinal sympathetic chain or via the vagal parasympathetic afferents.

B. Lower Motor Neuron Lesions

In patients with an LMN lesion, the following symptoms are generally noticed: increased rectal capacity, loose lower bowels, reduced rectal contractions, and reduced or no control of the external sphincter.

Typically, patients with LMN neurogenic bowel have parasympathetic and sympathetic innervation deficits. In the case of an isolated pudendal nerve insult, the colonic transit times are usually normal and fecal incontinence is predominant. The decreased tone of the distal colon is a result of loss of parasympathetic supply.

Differential Diagnosis

A multitude of medical diagnoses may cause a patient to manifest a neurogenic bowel. Among the most common are SCI, dementia, spina bifida, multiple sclerosis, diabetes mellitus, Brown-Séquard syndrome, central cord syndrome, myelomeningocele, and simple aging. In addition, neurogenic bowel has been noted in patients who had recently undergone pelvic surgery or vaginal delivery.

Complications

Bowel problems occur in 27–62% of SCI patients. Patients who develop a neurogenic bowel are at risk for numerous complications, including fecal impaction, hemorrhoids, ileus gastritis and ulcers, cholelithiasis, pancreatitis, superior mesenteric artery syndrome, electrolyte imbalance, acute abdomen, and cancer.

A. Fecal Impaction
The overall goal of a bowel program is to prevent the complication of fecal impaction. This problem usually presents as diminished or absent passage of stool, and sometimes as overflow diarrhea. Fecal impaction has been known to cause autonomic dysreflexia in patients and, in extreme cases, a perforated viscus. If impaction is suspected, a manual disimpaction should be attempted with anesthetic jelly in order to prevent autonomic dysreflexia. If the impaction is found to be more proximal, enemas or potent oral stimulants may be indicated. In repetitive cases of fecal impaction, especially those that result in episodes of autonomic dysreflexia, an ileostomy or colostomy may be indicated.

B. Hemorrhoids

Hemorrhoids are usually caused by increased rectal pressure and are also often associated with prolonged efforts to remove hard stools. Symptoms include bleeding, pain, and potentially autonomic dysreflexia. Effective treatment strategies include maintaining soft stools and regular bowel movements. Medicated suppositories and topical steroid ointments can be effective in treating symptoms. Care must be taken, however, because suppositories, enemas, and digital stimulation can also exacerbate symptoms.

C. Ileus Gastritis

Dysphagia is sometimes seen in patients with neurogenic bowel. Factors that predispose this patient population to gastroesophageal reflux disease (GERD) include delayed gastric emptying, recumbent positioning, immobilization, and certain drugs. Although the occurrence of GERD is not increased in SCI patients, this patient population may undergo endoscopy less frequently, allowing esophagitis and gastritis to reach a more advanced clinical course prior to diagnosis.

D. Gallbladder Involvement

The reason for the increased incidence of cholelithiasis in patients with neurogenic bowel resulting from SCI is unknown. No correlation has been found with the neurologic level of injury, duration of SCI, age, obesity, or diabetes mellitus, although subsequent development of cholelithiasis may correlate with the severity of the SCI lesion. Because of the loss of normal sensory perception, the signs and symptoms of gallbladder disease can be atypical or even absent. Consequently, the pathologic process may not be discovered until it has reached an advanced stage. Symptoms that have been reported in patients with coexisting
neurogenic bowel and cholelithiasis or cholecystitis include a right upper quadrant mass, fever, tachycardia, and cholestasis. Predisposing factors for cholecystitis include use of narcotic drugs, mechanical ventilation with positive end-expiratory pressure, and hyperalimentation. The need for cholecystectomy remains at the same level as in patients without neurogenic bowel, as long as proper prophylactic measures are taken.

E. Superior Mesenteric Artery (SMA) Syndrome
SMA syndrome is a condition in which the third part of the duodenum is intermittently compressed by the overlying superior mesenteric artery. Factors that predispose patients to this syndrome include rapid weight loss, prolonged supine positioning, and the use of a spinal orthosis. Symptoms of SMA syndrome include pain and vomiting, particularly in the supine position. Treatment consists of upright body positioning, weight gain, and, in severe cases, a duodenojejunostomy.

F. Acute Abdomen
The loss of sensory, motor, and reflex functions in patients with SCI usually masks the signs of acute abdomen such as pain, tenderness, rigidity, and guarding, although pain may still be present. Early clues include nausea, vomiting, malaise, a vague feeling of being unwell, bowel or bladder incontinence, bladder spasms, and diarrhea. On physical examination the patient may be febrile, have abdominal distention, tenderness to palpation, decreased or absent bowel sounds, and possibly rigidity or spasm. Acute abdominal pathology includes a perforated viscus (such as peptic ulcer disease), cholecystitis, appendicitis, and bowel obstruction. Acute abdomen has a 9.5% mortality rate in SCI patients, and proper medical treatment without delay is imperative to reduce mortality in these patients.

G. Colorectal Cancer
Colorectal carcinoma in patients with a neurogenic bowel is discovered at a typically more advanced stage than in the general population. This delay in detection might occur because the most common symptoms of GI cancer—distention, constipation, and pain—could also be attributed to other GI complications of neurogenic bowel. Proper colorectal screening, using fecal occult blood testing and colonoscopy, is important for this patient population, and the appropriate screening guidelines should be followed.
Treatment

The treatment goals for patients with neurogenic bowel are generally similar, regardless of the classification (ie, hyperreflexic or areflexic bowel): effective evacuation without fecal incontinence. Stool consistency is a key factor in finding the balance between effective evacuation and fecal incontinence, which should be managed with diet and medications.

A. Medical Management

1. Acute phase postinjury—During the acute phase (5–21 days) of neurogenic bowel after an SCI, most or all of the reflex activity below the level of the lesion is temporarily lost or depressed as a result of spinal shock. This time period is characterized by less reflex-mediated defecation. Vigilant monitoring is essential as patients are at increased risk for gastric atony and bowel ileus.

2. Chronic phase postinjury—Once the spinal shock has resolved, a bowel program is typically initiated. During this time period, continued caution is advised. Depending on the level of the spinal lesion, the patient may develop a hyperreflexic or hyporeflexic bowel. Hemorrhoids can develop in response to high pressures caused by the passage of hard stools. In addition, with LMN lesions, the chronic passage of large, hard stools can eventually result in rectal prolapse with an overstretched, incompetent sphincter.

3. Basic components of a bowel program—The bowel program encompasses methods, behaviors, techniques, and medications for self-care that are appropriate for the patient, along with processes and procedures for assisted defecation and evacuation, as necessary. This program constitutes the treatment regimen for the patient with a neurogenic bowel. Although adequate rectal evacuation is usually defined as evacuation on a daily or every-other-day basis, some studies suggest that average SCI patients with an areflexic bowel have a bowel evacuation pattern of twice per day, whereas those with a hyperreflexic bowel have an evacuation pattern of three times per week.

A. Diet—The patient should be encouraged to consume a consistent, balanced diet emphasizing sufficient daily fiber (15–30 g) and adequate fluid intake (generally 1–2 L/day).

B. Activity—To promote intestinal motility, patients are encouraged to spend time out of bed and, as their condition improves, to resume an active lifestyle.
C. Medications—Anticholinergic medications, narcotics (which decrease peristalsis), and contact irritants such as castor oil should be avoided in patients with a neurogenic bowel. Medications that can be used to aid evacuation include stool softeners, stool bulking agents, and oral stimulants. Stool softeners (eg, docusate sodium [Colace]) aid in bowel movements by increasing fluid in the GI tract. Agents such as psyllium (Metamucil) increase stool bulk. Oral stimulants (eg, sennosides [Senokot], bisacodyl tablets [Dulcolax]) stimulate peristalsis by action on Auerbach’s plexus. Suppositories help to trigger the anorectal reflex. Some, such as bisacodyl suppository, can also trigger local peristalsis. Evidence indicates that polyethylene glycol–based suppositories (eg, Magic Bullet) are superior to hydrogenated vegetable oil–based suppositories because they dissolve faster and act more rapidly. Mini-enemas are another option and act in a similar fashion to the suppositories.

4. Bowel care and assisted evacuation measures—The process of assisted defecation should occur at a consistent, scheduled time in order to regulate the bowel and, if possible, take advantage of existing physiologic reflexes.

If the gastrocolic reflex is present (increased colonic activity within 15–60 min after a meal), the patient should be instructed to use the commode and evacuating technique within 1 hour after a meal. The position of being upright on a commode takes advantage of gravity in the evacuation process and facilitates the Valsalva maneuver.

If the anorectal–rectocolic reflex is present (reflex relaxation of the internal anal sphincter when the rectal wall is stretched), the schedule for bowel care should include suppositories, enemas, or manual digital stimulation (see below), all of which can activate this reflex, except in patients with LMN lesions.

Oral stimulants should be administered 6–12 hours before the scheduled assisted evacuation to take full advantage of the effect of the medication. The administration time may need to be adjusted, based on individual results, to coincide with the gastrocolic reflex.

5. Techniques used to trigger rectal evacuation—Patients who require assistance with rectal evacuation may benefit from one of more of the following measures.

A. Suppositories and mini-enemas—These options can help in transitioning the patient from stimulant suppositories (eg, Magic Bullet, glycerin suppositories) as the need for local peristalsis diminishes.
B. **Digital stimulation**—In this technique, a lubricated gloved finger is inserted and the anal canal is stretched for 20–30 seconds, causing reflex contraction of the colon (anorectal reflex).

C. **Digital disimpaction and manual extraction**—This technique, which involves manual emptying of the rectal vault, does not rely on the anorectal reflex.

D. **Enemas**—Although useful for assisted evacuation in many patients, enemas may be difficult to perform in those with a neurogenic bowel, especially if the anal sphincter is incompetent (eg, in patients with an areflexic bowel).

E. **Transanal irrigation**—This is a retrograde irrigation technique using a specialized catheter with an inflatable balloon that is inserted into the rectum. The balloon is inflated to prevent leakage of the water used as a washout fluid and, after several minutes, deflated. The catheter is then removed and the bowel allowed to evacuate. Transanal irrigation may be particularly helpful in the control of fecal incontinence but caution is advised. While in one study initial success rates were high (approximately 80% after 3 months), this rate declined over time to 35% after 3 years. Even successful users of this technique may experience side effects, which include abdominal pain, minor rectal bleeding, fatigue, and general discomfort.

**B. Surgical Treatment**

If after a sufficient amount of time and various trials the conservative measures described above are ineffective, then surgical options should be considered.

1. **Electrical stimulation or sacral neuromodulation**—This technique can be tried to assist with evacuation and diminish symptoms of constipation, fecal incontinence, or both, in selected patients with incomplete SCI when conservative measures are unsuccessful.

2. **Antegrade irrigation**—In this procedure, a tract is made from the abdominal wall to the appendix, a catheter is passed through this appendicostomy, and the colon is irrigated in an antegrade fashion. Studies report better results in younger patients with spina bifida. Stenosis of the tract can occur over time.

3. **Colostomy**—This procedure is an option once all other options fail. A left colostomy may be more desirable as the resultant feces is less liquid than that
produced with a right colostomy. This option may decrease bowel care time and improve quality of life for many patients. However, while it may resolve problems of rectal evacuation and rectal fecal incontinence, additional issues may be introduced, including ostomy bag leakage, prolapse of the ostomy, or local skin irritation.

4. **Ileostomy**—An ileostomy procedure is usually performed if the entire colon is atonic. There may be less absorption of water since the feces does not enter the colon. Otherwise this process is similar to the colostomy and can provide a similar increase in quality of life for patients with intractable bowel disease.

### Prognosis

With proper medical treatment and attentiveness to a bowel regimen, patients with a neurogenic bowel can lead healthy and productive lives. Just as in the general population, those who maintain a regular level of activity and fitness have a greater chance of maintaining regular bowel movements. Patients with SCI are prone to many of the diseases that affect the healthy population, such as upper and lower GI tract malignancies. Consequently, physicians should screen all patients over the age of 50 years for colorectal cancers using fecal occult blood testing and colonoscopy.


NEUROGENIC BLADDER

ESSENTIALS OF DIAGNOSIS

► Inability to void.
► Inability to properly empty the bladder even if able to void.
► Increased postvoid residual volume of urine.
► Involuntary incontinence.

General Considerations

The bladder stores urine produced by the kidneys and empties voluntarily. When the bladder does not perform properly, the cause may be structural abnormalities or neurologic dysfunction. The focus in this discussion is on evaluation and management of the latter category.

A. Epidemiology

Neurologic dysfunction of the bladder can derive from injury or disease at the level of the brain–corticopontine mesencephalic nuclei, the spinal cord, the autonomic nervous system, or the sacral nerves to the bladder. Neurogenic bladder can thus occur in association with a wide range of conditions, including cerebrovascular accidents, head injuries, degenerative disorders of the CNS,
multiple sclerosis, SCI, and autonomic dysfunction. Symptoms depend on the location of the lesion and function of the nerves that are damaged (Table 7–1).

**Table 7–1** Comparison of key characteristics in upper motor neuron (UMN) versus lower motor neuron (UMN) neurogenic bladder.
<table>
<thead>
<tr>
<th>Urinary problem</th>
<th>LMN Bladder</th>
<th>UMN Bladder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Causes (^4)</td>
<td>Most common—flaccid bladder, spastic sphincter, or both in spinal shock, the reflex arc is not functioning due to initial trauma. Other causes include conus medullaris syndrome, cauda equina syndrome, syringomyelia, acute cerebrovascular accident (detrusor areflexia)</td>
<td>Most common—spastic bladder, incompetent sphincter, or both in spinal cord injury, there is return of the reflex arc after initial trauma. Other causes include subacute cerebrovascular accident (detrusor hyperreflexia) and multiple sclerosis (detrusor hyperreflexia is most commonly seen)</td>
</tr>
<tr>
<td>Lesion location</td>
<td>Involves sacral micturition center (S2–S4) Exclusively involves peripheral innervation of bladder</td>
<td>Above sacral micturition center (above S2)</td>
</tr>
<tr>
<td>Clinical presentation</td>
<td>Large, areflexic, flaccid bladder Tight, spastic sphincter Failure to empty</td>
<td>Small, overactive, spastic bladder Failure to store</td>
</tr>
<tr>
<td>Treatment</td>
<td>Intermittent catheterization Crédé maneuver (suprapubic pressure) Valsalva maneuver Drugs to induce urination: • Cholinergic receptor stimulators (eg, bethanechol [Urecholine]) • α-Adrenergic receptor blockers (eg, prazosin [Minipress], phenoxymethamine [Dibenzyline], terazosin [Hytrin], doxazosin [Cardura])</td>
<td>Drugs to enhance storage of urine: • Anticholinergic medications (eg, tolterodine [Detrol] and propantheline [Pro-Banthine]) are most commonly used • Direct smooth muscle relaxers (eg, oxybutynin [Ditropan]) • α-Receptor and β-receptor stimulators (eg, imipramine [Tofranil], ephedrine) allow storage</td>
</tr>
</tbody>
</table>

\(^4\)These are the most commonly seen types of bladder pathology in each case. However, several causes can fall into either category or display features of both LMN and UMN pathology at different times throughout the natural course of the disease. For example, patients with multiple sclerosis may have detrusor hyperreflexia, detrusor areflexia, or detrusor sphincter dysfunction; those with cerebrovascular accident may present initially with detrusor areflexia followed by detrusor hyperreflexia.


Overall, the inability to control urination affects 1.6% of men and 8.5% of women between the ages of 15 and 64 years. Commonly occurring disabilities that can present with a neurogenic bladder include stroke, multiple sclerosis,
Parkinson’s disease, and SCI. Up to 15% of stroke patients are affected by urinary retention due to a flaccid bladder, which usually later changes to a spastic bladder. Among patients with multiple sclerosis, 40–90% are affected; for Parkinson’s disease, the range is 37–72%. Among patients with SCI, the percentage affected is dependent on the severity (ie, complete versus incomplete injury) and level of the damage to the spinal cord.

**B. Neuroanatomy and Function of the Urinary Tract and Bladder**

Bladder function is based on principles that involve the storage of urine and the eventual emptying of urine from the bladder. The lower urinary tract consists of the urinary bladder and its outlet, the urethra, and is responsible for the storage and occasional elimination of urine. The lower urinary tract is controlled by a complex set of peripheral autonomic and somatic nerves, which in turn are controlled by neural pathways in the spinal cord and brain. Defects in the lower urinary tract can lead to urinary retention or incontinence, an embarrassing condition that affects over 200 million people worldwide.

In a healthy adult with intact bladder and pelvic floor muscle tone, the bladder can store up to 200–300 mL of urine before a signal is emitted from the stretch receptors in the wall of the bladder. This sensory signal is transmitted to the spinal cord at the level of S2–S4. The spinal cord then relays several signals: first a signal travels up the spinal cord to the brain, signaling bladder fullness; a second reflexive signal then stimulates the sympathetic branches to open the bladder sphincter and the parasympathetic branches to initiate emptying.

**1. Central pathway control**—One of the two main pathways in the control of voiding function is the central pathway, the first portion of which is located in the frontal lobe, within the corticopontine mesencephalic nuclei. This area of the brain functions to inhibit the parasympathetic sacral micturition center and permit the storage of urine in the bladder. Within the pons are the pontine mesencephalic nuclei. It is here that the control of bladder contraction and sphincter relaxation occurs. If there is loss of control from this center, the result is detrusor sphincter dyssynergia. The pelvic and pudendal nuclei are involved in sacral micturition by integrating stimuli from the cephalic centers. This area helps mediate the parasympathetic S2–S4 sacral micturition reflex. The motor cortex to the pudendal nucleus is involved in voluntary control (contraction and inhibition) of the external urethral sphincter.

**2. Peripheral pathway controls**—The parasympathetic efferent nerve fibers originate in the detrusor nucleus in the intermediolateral gray matter of the sacral
cord at S2–S4. They travel through the pelvic nerves to parasympathetic receptors of the detrusor muscles and act to stimulate the cholinergic receptors causing bladder contraction and emptying.

The sympathetic efferent nerve fibers originate in the intermediolateral gray matter from T11 to L2. They travel through hypogastric nerves to α₁- and β₂-adrenergic receptors within the bladder and urethra. Stimulation of α₁-adrenergic receptors at the base of the bladder and prostatic urethra causes smooth muscle contraction. Stimulation of the β₂-adrenergic receptors within the body of bladder causes smooth muscle relaxation. Together, the activation of these two receptors functions in the storage of urine.

The somatic efferent nerve fibers originate at the pudendal nucleus of sacral segments of S2–S4. They travel through the pudendal nerve to innervate striated muscle of the external urethral sphincter. These nerves help in the voluntary contraction of the sphincter and prevent emptying or leakage of urine.

The afferent nerve fibers originate at the detrusor muscle stretch receptors, external urethral sphincters, perineum, and genitalia. These fibers travel through the pelvic and pudendal nerves to the sacral cord. The myelinated Aδ fibers respond to bladder distention, stimulating the parasympathetic emptying of the bladder. Although unmyelinated C fibers are not necessary for normal voiding, there is increased activity within these fibers after SCI.

3. **Receptors of the bladder**—The cholinergic muscarinic receptors are located within the bladder wall, trigone, and bladder neck, and are involved in bladder contractions, ability to increase intravesicular pressure, and subsequent voiding. The α₁-adrenergic receptors are located within the base of the bladder and prostatic urethra. Norepinephrine binds to these receptors to cause contraction of the internal sphincter. The β₂-adrenergic receptors are located within the neck and body of the bladder. Norepinephrine binds to these receptors to cause relaxation and decrease intravesical pressure.

4. **Urethral sphincters**—The internal sphincter is composed of involuntary smooth muscle and contracts to promote urine storage. It has a large number of α-adrenergic receptors and is innervated by the T11–L2 sympathetic hypogastric nerves, under the control of the autonomic system. The external sphincter consists of voluntary skeletal muscle, and helps to prevent leakage or volitional emptying. It is innervated by the S2–S4 pudendal nerves.

5. **Normal bladder storage**—Bladder storage is a function of the sympathetic
nervous system. Sympathetic tone predominates to promote internal sphincter contraction and bladder relaxation. The T11–L2 sympathetic efferent fibers travel through the hypogastric nerves to activate $\alpha_1$- and $\beta_2$-adrenergic receptors, causing the sphincter to contract and the body of the bladder to relax, thus allowing for storage of urine. The $\alpha_1$-adrenergic receptors function to cause contraction of the internal sphincter at the base of the bladder and the prostatic urethra, whereas the $\beta_2$-adrenergic receptors cause relaxation of the body of the bladder.

6. Normal bladder emptying—Bladder emptying is a function of the parasympathetic nervous system. Parasympathetic tone predominates to promote bladder contraction and emptying. The S2–S4 parasympathetic efferent fibers travel through the pelvis to activate cholinergic (muscarinic $M_2$) receptors. Acetylcholine stimulates cholinergic receptors in the bladder wall, trigone, neck, and urethra, causing bladder contraction.

Clinical Findings

Because malfunction of the nerves innervating the bladder may be caused by conditions as varied as trauma to the brain or spinal cord, or congenital abnormalities, a careful and individualized evaluation of patients who present with urinary voiding problems is essential. Table 7–1 summarizes key characteristics that aid in distinguishing clinical presentations of neurogenic bladder.

A. Symptoms & Signs

The patient history is helpful in determining whether voiding symptoms existed prior to the time of injury or dysfunction. A thorough history and careful neurologic examination (including an ASIA examination; see Chapter 12) can help differentiate among neurologic disorders, determine the level of a lesion, and clarify the extent of dysfunction. These features have importance in predicting the type of lower urinary tract dysfunction that might be present. A record of the patient’s fluid intake, output, postvoid residuals (PVRs), and incontinence episodes over a 24-hour period should also be obtained. This information, along with results from the other tests mentioned below, can aid in determining the type of neurogenic bladder that is present.

Symptoms of a neurogenic bladder can be very misleading and do not always
correlate with objective findings. They can be classified into three categories, defined below.

1. **Inability to void**—Symptoms in this category may result from the bladder being unable to contract and therefore empty, or from the bladder being unable to contract sufficiently to overcome the resistance of the urethral sphincter as in dyssynergia.

2. **Inability to empty completely**—Symptoms in this category may result from decreased bladder contractions, or from increased urethral sphincter resistance that prevents the bladder from emptying completely.

3. **Incontinence**—Involuntary leakage of urine may be due to neurogenic factors or to other factors that must be determined or ruled out. Possible nonneurogenic factors that can be treated or modified include urinary tract infections (UTIs) or cystitis (causing frequency and incontinence); medications such as diuretics and anticholinergics (causing increased bladder filling and decreased emptying, which can exacerbate urgency and incontinence); obesity (causing external pressure on the bladder, which can result in incontinence); constipation, impaction, or rectal distention (causing pressure on the bladder); certain foods, caffeine, and alcohol (which can have a diuretic effect), and behavioral factors such as trying to avoid the activity involved in voiding (eg, getting out of bed, asking for assistance), which can also result in overflow urgency and incontinence.

**B. Diagnostic Studies**

Testing for upper and lower urinary tract dysfunction is individualized for each patient and neurologic condition. The anatomic level of the lesion can suggest the most likely pattern of bladder dysfunction (see Table 7–1); however, this should be confirmed with a urodynamic evaluation (UDE).

1. **Upper tract evaluation**—Ultrasound is a low-risk and relatively low-cost test for routine structural evaluation of the upper urinary tract. It can identify chronic obstruction and dilation, scarring, renal masses (obstructive and cystic), and stones but is not sensitive enough to detect acute ureteral obstruction, which is best identified using a noncontrast-enhanced computed tomography (CT) scan.

   A CT scan without contrast is often performed to evaluate for acute obstruction from stones. It is the most sensitive test for detecting small bladder stones in patients who might have a collapsed bladder from an indwelling
Computed tomography–intravenous pyelography (CT-IVP) has supplanted the use of excretory urograms as a diagnostic tool for evaluating patients with upper urinary tract dysfunction. However, this test should not be administered to patients with serum creatinine concentrations greater than 1.5 mg/dL, or those with insulin-dependent diabetes mellitus, to prevent contrast-related nephropathy.

2. Lower tract evaluation—PVR volumes provide a quick and simple way to assess bladder function, specifically, the ability of the bladder to empty. This information is especially helpful when combined with bladder pressure readings obtained from a UDE. PVR volumes obtained at the bedside by means of a bladder scan are preferable to those derived from postvoid catheterization collections in determining residual urine volumes. A PVR volume that is less than 20% of capacity is no longer considered to be indicative of a “balanced” bladder as high intravesicular pressures may coexist with low PVR values.

Cystography may be performed to evaluate for ureteral reflux and can also reveal the outline and shape of the bladder. The only routine indication for cystoscopy is the presence of a long-term indwelling suprapubic or urethral catheter, which increases the risk of bladder tumor development. Screening using this test is recommended every 5 years in high-risk patients, such as smokers, and every 10 years in those with no known risk factors.

Other tests for evaluation of urinary tract dysfunction include urethral pressure profiles and the bethanechol (Urecholine) stimulation test; both tests have limited value and high false-positive and false-negative rates.

3. Urodynamic evaluation—A typical urodynamic test takes about 30 minutes to perform. A comprehensive UDE provides information about the sensation and capacity of the bladder, as well as the presence of involuntary detrusor activity. The tests used to evaluate these functions include the voiding flow rate, cystometry, sphincter EMG, and urethral pressure profilometry. The objective in performing these tests is to attempt to reproduce the patient’s typical micturition cycle and associated symptoms.

Sensations evaluated include the first sensation of bladder filling, which usually occurs at 100 mL, the first urge to void, and a strong desire to void (proprioceptive sensations). The accepted normal bladder capacity is 300–600 mL. The functional bladder capacity is a combination of voided volume and residual urine volume. High PVR volumes (≥ 150 mL) may indicate UTIs. Increased levels are also associated with overflow incontinence.
During the cystometry portion of the UDE, saline is infused at a rate of 40–60 mL/min. The patient is then asked to identify various sensory thresholds, including the first sensation of filling, the first desire to void, and a strong desire to void. When a patient achieves sensations at lower than expected sensory thresholds, the diagnosis of increased bladder sensation is made. Alternatively, when sensation is achieved at a higher than expected sensory threshold, or in some cases not at all, the diagnosis of decreased bladder sensation is made.

During the filling phase, any involuntary bladder contractions should be noted. Although involuntary detrusor contractions are identified in up to 20% of asymptomatic patients during cystometry, their presence should always be considered abnormal, prompting consideration of detrusor overactivity. Once the bladder is filled to 50% of the expected capacity, filling is temporarily stopped to evaluate for an incompetent urethral sphincter closure mechanism; this is frequently seen in patients with stress incontinence.

When the bladder is filled to capacity, the patient is given permission to void. If the patient voids involuntarily before this point, the diagnosis of detrusor overactivity can be made. During the voiding phase, a flow rate is obtained using a flowmeter. At the same time, intravesicular pressure is calculated. These measurements, collected in a pressure-flow study, are used to evaluate for a physiologic bladder obstruction. The diagnosis of bladder obstruction is confirmed by the simultaneous observation of elevated bladder pressures and low urinary flow rates. Used in combination with fluoroscopy, these tests can allow the examiner to determine the level of obstruction.

EMG activity of the urinary sphincter is evaluated to determine if there is synergy between the bladder and sphincter. In many patients with neurogenic bladders, obstruction occurs because of lack of neurally mediated coordination of the sphincter and bladder, a process termed detrusor–sphincter dyssynergia. The examiner must be able to differentiate between true dyssynergia and dysfunctional voiding, a learned behavior or an artifact.

### Differential Diagnosis

The diagnosis of neurogenic bladder is usually made on the basis of the history and physical examination of the patient, but further testing may be needed to differentiate the neurologic etiology. Table 7–1 contrasts key characteristics that aid in differentiating UMN lesions from LMN lesions. Conditions that mimic neurogenic bladder must also be excluded.
A. Central Etiologies: Suprasacral Lesions (UMN)

Patients with lesions that arise above the level of the spinal cord usually present with bladder spasticity. The sacral arc remains intact; however, a loss of inhibition from higher centers results in a spastic bladder and sphincter. The degree of spasticity will vary from patient to patient.

The differential diagnosis of suprasacral lesions that can cause bladder spasticity includes SCI above the level of the sacral segments (conus medullaris) secondary to multiple etiologies, trauma, syringomyelia, myelitis, multiple sclerosis, Parkinson’s disease, brain tumors, dementia, cerebrovascular accidents, inflammatory disorders (eg, encephalitis, meningitis), and iatrogenic factors. Lesions in the internal capsule, secondary to a cerebrovascular accident or Parkinson’s disease, are known to cause both spastic and semiflaccid bladder. When lesions occur about the pontine micturition center, detrusor–striated sphincter dyssynergia is usually absent. Sphincter spasticity and voiding dyssynergia can ultimately lead to detrusor hyperactivity, high voiding pressures, ureteral reflux, or obstruction. This progression, in turn, may eventually lead to a decrease in renal function.

Suprasacral lesions can cause a wide range of symptoms. Besides the obvious incontinence, patients may experience precipitate urge, frequency, residual urine, urinary retention, and recurrent UTIs. It is important to assess for autonomic dysreflexia in patients with an SCI above the level of T6. Overdistention of the bladder, dyssynergic voiding, or simply inserting a catheter can cause the symptoms of autonomic dysreflexia, which includes elevated blood pressure, bradycardia, and diaphoresis.

B. Peripheral Etiologies: Sacral Lesions (LMN)

Major causes of a neurogenic bladder via injury to the sacral spinal roots or LMNs include injury to the detrusor motor nucleus, the afferent feedback pathways, and peripheral nerves.

Injury to the spinal cord at the micturition center (S2–S4), more specifically the detrusor motor nucleus, is the most common cause of a flaccid neurogenic bladder. A flaccid bladder may also occur in SCI patients who are in spinal shock, or stroke patients who are in the acute flaccid hemiplegic stage. Damage at the detrusor motor nucleus may be the result of infection secondary to diseases such as herpes zoster or poliovirus, herniated disks that injure the cauda equina or sacral nerve roots, myelodysplasias, or iatrogenic factors such as surgery or radiation. In these injuries, external sphincter and perineal muscle
tone are diminished, but the patient usually does not manifest urinary incontinence because of the compensatory increase in bladder storage associated with flaccidity.

Injury to the afferent feedback pathways can result from various neuropathies, including diabetes mellitus, tabes dorsalis, pernicious anemia, and posterior spinal cord lesions. There is no direct injury to the detrusor motor nucleus in these injuries; however, there is a loss of sensory input to the detrusor nucleus or a loss of neurotransmission in the dorsal horns of the cord.

Peripheral injuries are the third significant cause of an atonic bladder in a neuropathic patient. Typical causes of this kind of injury include fractures, mishaps from surgery, radiation therapy, chronic infection, interstitial cystitis, and carcinoma in situ. Typically in these injuries, the smooth muscle remains intact, but there is no central reflex to organize muscle activity. A pelvic fracture typically tears the nerves to the external sphincter, whereas surgical complications usually damage only the sensory innervation of the external sphincter. In radiation therapy there is usually a denervation of the detrusor or the sphincter.

C. Other Etiologies: Conditions That Mimic Neurogenic Bladder

Several conditions that can mimic the symptoms of a neurogenic bladder should be considered in the differential diagnosis; they include cystitis, interstitial cystitis, cystocele, chronic urethritis, myogenic damage, and infravesical obstruction.

Complications

Common complications in individuals with a neurogenic bladder include UTIs, stone formation, and incontinence. Serious complications include hydronephrosis, infection, decompensation of the ureterovesical junction, and loss of renal function.

A. Urinary Tract Infections

1. Asymptomatic UTIs—It is important to differentiate colonization (a positive culture with no significant pyuria) from an infection (a positive culture along with pyuria). Asymptomatic UTIs in an SCI patient being managed with an indwelling Foley catheter are generally not treated. There is also no support in the literature for the use of prophylactic antibiotics in the prevention of UTIs. If
the patient desires, vitamin C, cranberry juice, and methenamine salts can be used as acidifying agents. Exceptions are patients undergoing invasive procedures (cystoscopy and UDEs), those in whom vesicoureteral reflux has been noted, or those showing growth of urease-producing organisms (eg, *Escherichia coli*, *Staphylococcus epidermidis*, *Klebsiella*, *Proteus*, *Pseudomonas*, and *Providentia*). Identification of this latter group is important as urea-splitting organisms will ultimately produce struvite calculi (discussed below).

2. **Symptomatic UTIs**—Patients who have a symptomatic UTI (fever, malaise, increased spasticity, or neurogenic pain) warrant treatment. Criteria for treatment with or without symptoms include the presence of more than 100,000 organisms per milliliter in a clean-catch midstream specimen, more than 100 organisms per milliliter during a bladder catheterization, and pyuria. In patients with suspected pyelonephritis, the infection should be treated aggressively with appropriate antibiotics to prevent renal loss. Patients with epididymitis should also be treated with antibiotics, along with bed rest and scrotal elevation. If these patients have an indwelling Foley catheter, it should be either removed or replaced.

3. **UTI prophylaxis**—UTIs in patients with neurogenic bladders can be prevented to some degree by adequately draining the bladder at pressures below 40 cm H₂O, either by intermittent catheterization or by timely surgical relief.

**B. Calculi**

Several factors contribute to renal stone formation in patients with a neurogenic bladder. Bed rest and inactivity can cause demineralization of the skeleton, mobilization of calcium, and subsequent hypercalciuria. In addition, a reduction in fluid intake can contribute to urinary stasis, increasing the concentration of calcium in the urine. Finally, infection with urea-splitting organisms results in alkalization of the urine, reducing the solubility of calcium and phosphate.

Bladder stones often occur with indwelling catheters, are usually soft, and can usually be crushed and washed out through a cystoscope sheath. If they are too large to be removed using this method, a suprapubic cystotomy can be performed.

Ureteral stones can be removed by anterograde or retrograde retrieval methods or via shock wave lithotripsy.

Kidney stones in individuals with a neurogenic bladder are usually the result of an infection. When the infection is left untreated, the stones eventually
become the source of constant infection and, eventually, loss of renal function. Although smaller stones can often be removed using endoscopic procedures, some may require open surgery owing to their size.

**C. Hydronephrosis**

“Back pressure” is applied on the kidneys primarily through two mechanisms. In the first of these, trigonal stretch secondary to residual urine and detrusor hypertonicity eventually causes abnormal pull on the ureterovesical junction, producing increased resistance to the passage of urine. This is referred to as a “functional” obstruction of urine and is usually relieved by indwelling catheter drainage or by intermittent catheterization, combined with the use of anticholinergics.

Eventually, trigonal hypertrophy and detrusor spasticity can lead to decompensation of the ureterovesical junction, causing reflux—a second mechanism that produces back pressure. This progression is usually due to a combination of high intravesical pressure and trabeculation of the bladder wall. The increased stiffness of the ureterovesical junction weakens the valvelike function, slowly eroding its ability to prevent urine reflux. If an indwelling catheter fails to alleviate the problem, antireflux surgery can be considered. Progressive hydronephrosis may require a nephrostomy. If all other methods fail, a urinary diversion may be performed.

**D. Autonomic Dysreflexia**

Autonomic dysreflexia is seen in patients with SCI occurring at the level of T6 and above; however, it is more common in patients with spastic lesions above T1. Symptoms include increased elevations in systolic or diastolic blood pressure (or both), sweating, bradycardia, headache, and piloerection. An overdistended bladder usually precipitates this phenomenon. It is a sympathetically mediated reflex behavior triggered by sacral afferent feedback to the spinal cord. Immediate catheterization is indicated as the first treatment option and can usually prompt resolution of symptoms.

**E. Bladder Tumors**

As previously noted, there is increased risk for bladder tumors in patients with an indwelling catheter, whether urethral or suprapubic. Cystography is indicated in these cases (see earlier discussion).
Treatment

Treatment options for neurogenic bladder can be grouped into three categories: (1) medical measures to manage or alleviate the inability to void or incontinence, (2) surgical procedures to correct structural problems and improve function in patients with chronic incontinence, and (3) neurostimulation techniques. The algorithm shown in Figure 7–1 provides a protocol for the initial treatment of a patient with a neurogenic bladder. Table 7–1 differentiates treatment measures specific to UMN and LMN lesions. Detailed discussion of these and other management strategies follows.
Figure 7–1 An algorithm for the initial treatment of neurogenic bladder of known etiology. PVR, postvoid residual; UA, urinalysis; C&S, culture and sensitivity; UTI, urinary tract infection; UDE, urodynamic evaluation. (Reproduced with permission from the JFK–Johnson Rehabilitation Institute, Edison, NJ.)

A. Medical Management: Inability to Void
1. Flaccid, areflexic, or hypotonic bladder—

A. Catheterization—An indwelling urethral catheter provides continuous drainage of urine from the bladder. Its potential side effects include recurrent UTIs, bladder stones, urethral erosions, and risk of bladder carcinoma. Maintenance needs include oral fluid intake of at least 2 L/day, catheter changes every 3–4 weeks, and placement of the drainage bag below the level of the bladder to prevent reflux of urine back into the bladder.

A suprapubic catheter is a percutaneous indwelling catheter inserted through the suprapubic region that also provides continuous drainage. It is associated with fewer recurrent UTIs, and no urethral erosions, but other side effects are similar to those of an indwelling urethral catheter.

An intermittent catheterization program (ICP) has fewer side effects than use of an indwelling catheter. However, this option is dependent on hand function or assistance from another person. A benefit of this approach is that recurrent UTIs are less frequent than with indwelling or suprapubic catheters. Use of sterile catheters further reduces the frequency of UTIs compared with the clean technique of reusing catheters after cleansing.

B. Pharmacotherapy—Medications to stimulate bladder detrusor contractions, such as bethanechol (Urecholine), can be tried. This therapy should be used with an external collection device or condom catheter, especially if voluntary control is lacking. For this method to be effective, the patient must have decreased sphincter tone and contraction, and acceptable PVR volumes. Otherwise intermittent catheterizations will still need to be performed.

C. Positioning—Patients can be instructed in the use of the Credé or Valsalva maneuver to increase intravesicular pressure through manual suprapubic pressure (Credé maneuver) or increased abdominal pressure (Valsalva maneuver). For this method to be effective, the patient must have decreased urethral sphincter activity, and sphincter–detrusor dyssynergia cannot be present.
PVR volumes should be used to determine if this method has been successful, thus avoiding unnecessary catheterizations. However, patients may still have a problem with ureteral reflux leading to hydronephrosis and hemorrhoid exacerbation.

2. **Spastic, hyperreflexic, hypertonic bladder, or incomplete voiding with increased PVR volumes**—

A. **Pharmacotherapy**—α-Adrenergic receptor blocking agents such as tamsulosin (Flomax) and terazosin (Hytrin) are available that decrease sphincter–detrusor dyssynergia or decrease sphincter resistance sufficiently to allow adequate voiding or improve bladder emptying. They may need to be combined with a cholinergic medication, such as bethanechol, if bladder contraction is not adequate. PVR volumes should be checked to determine if bladder emptying is adequate. In addition, a UDE should be performed to ensure that bladder pressures are not excessive, as this state could lead to ureteral reflux.

B. **Sphincterotomy**—If the medications mentioned above are not effective, transurethral sphincterotomy and the use of an external condom catheter with a leg bag or diaper can be considered. The sphincterotomy is not permanent and needs to be repeated. As with other management options, PVR volumes are needed to assess the degree of bladder emptying. This method is usually not effective in decreasing intravesicular pressure; therefore, hydronephrosis remains a potential complication.

C. **Urethral stenting**—A stainless steel woven mesh urethral stent is another option that can be used to maintain sphincter patency. An external urine collection device or diaper again is needed. Possible side effects include stone encrustation, stent migration, and urethral strictures from tissue growth around the stent.

D. **Other approaches**—If the previously mentioned methods are not appropriate options, or their use proves unsuccessful, the patient may have no treatment option other than an indwelling catheter or intermittent catheterizations. If the patient experiences incontinence between catheterizations or has elevated bladder pressure on UDE as a result of bladder contractions secondary to overactivity or hyperreflexia, use of anticholinergic agents such as tolterodine (Detrol) or oxybutynin (Ditropan) may be needed to decrease the bladder contractions, especially as the bladder is being emptied using the intermittent
catheterization technique.

**B. Medical Management: Incontinence**

1. **Psychological approaches**—Some patients with a neurogenic bladder benefit from behavior modification techniques that use timed voiding or emptying to decrease the incontinence.

2. **Pharmacotherapy**—Anticholinergic medications can help to decrease bladder contractions. Tricyclic antidepressants, which are anticholinergic, can also be helpful in treating neuropathic pain. Antispasticity agents such as tizanidine (Zanaflex) and baclofen may also be beneficial.

3. **Botulinum toxin injection**—Botulinum toxin injection for neurogenic detrusor overactivity can decrease bladder contractions. This procedure is usually well tolerated; however, repeated treatments are usually necessary. Potential side effects include pain, procedure-related UTI, mild hematuria, increased PVRs, and urinary retention.

4. **External collection**—External collection devices, condom catheters for male patients, or diapers can also be used. However, prolonged contact of urine with skin is a concern with these methods.

**C. Surgical Treatment**

Surgical options such as bladder augmentation or auto-augmentation can be explored after more conservative treatment measures have failed.

1. **Bladder augmentation**—This surgical technique uses segments of the patient’s bowel to create a large-capacity bladder with low intravesicular pressure. It is an option when conservative treatments fail and the treatment goal is to achieve intermittent catheterizations without incontinence. Complications include metabolic abnormalities, acidosis, recurrent UTIs, stones, and adenocarcinomas.

2. **Bladder auto-augmentation**—This technique, also called a detrusor myomectomy, surgically increases the size of the bladder. Bladder capacity gradually increases (approximately 25%) over 1–6 months, helping to prevent or decrease incontinence between catheterizations. This method is preferred over bladder augmentation.
3. **Bladder diversion**—This approach is used when physiologic problems interfere with the ability to perform catheterizations. Two types of bladder diversions are performed: ileal conduit procedures and ileovesicostomy.

**A. ILEAL CONDUIT**—In this technique, the ureters are resected from the bladder and attached to a segment of ileum, which is then brought out through a stomacutaneous opening in the abdominal wall. Urine is collected in a bag placed over the stoma.

**B. ILEOVESICOSTOMY**—With an ileovesicostomy, the ileal segment acts as a conduit between the bladder and the skin. This procedure maintains the ureterovesical junction, which avoids the complications of ureteroileal stricture at the anastomosis site. A benefit of this procedure is that it can be reversed at a later date if necessary.

**D. Neurostimulation or Neuromodulation**

Patients must have an intact sacral reflex arc and bladder contractions to derive potential benefit from neurostimulatory techniques. Although the exact neurologic mechanism of action is unknown, sacral nerve stimulation has proved helpful in patients with overactive, spastic bladder and nonobstructive urinary retention. Some studies indicate that the therapeutic effects may be temporary. Stimulation at the level of the pudendal nerve may be beneficial for patients with overactive bladder symptoms that are not responsive to more conservative measures. Treatment considerations include the complications of lead migration, lack of efficacy over time, and the fact that any future magnetic resonance imaging studies will be contraindicated.

**Prognosis**

Complete recovery from a neurogenic bladder is uncommon; thus the goal of treatment should be to manage its effects. The clinician must keep in mind that the most common complication in a patient with neurogenic bladder is renal injury from increased bladder pressure or infection. Consequently, patients with a neurogenic bladder should have close followup to preserve their renal function. The use of baseline creatinine levels and renal ultrasound is beneficial in this effort.

For individuals with mild involvement, therapy complemented by incontinence pads usually provides enough relief for a patient to maintain a
relatively normal lifestyle. When this is not possible, catheterization may be necessary. Patients who are both motivated and physically able to learn intermittent self-catheterization are more likely to achieve a better quality of life, with fewer complications and less disability, than those who are maintained on continuous catheterization.


The use of physical agents and devices to reduce pain and improve function has a long history in the practice in medicine. In recent decades, technology has increased the variety of modalities available to the practitioner. This chapter surveys the most commonly used options, with the goal of aiding the practitioner in choosing among alternatives. Despite widespread use and decades of experience, however, consensus is lacking regarding indications, implementation, and efficacy for many of these approaches.

SUPERFICIAL HEAT THERAPY

Heat may be transferred to a patient from an object, a device, or water using hot packs, heat lamps, paraffin baths, and whirlpools. All of these options are considered superficial heating modalities because they are unable to produce temperature changes of more than a few degrees at depths of 1–2 cm. It is generally accepted that heat increases the extensibility of collagen tissue, decreases joint stiffness, relieves muscle spasms, provides pain relief, and increases both blood flow and metabolism. Superficial heat is an adjunctive treatment and generally not the primary curative intervention. Table 8–1 lists general precautions and contraindications for its use. Permanent brown skin discoloration (erythema ab igne) can result from repeated chronic overuse of superficial heat.

Table 8–1 Precautions and contraindications for use of superficial heat.
Hot Packs

Many different varieties of hot packs are available. Outer layers may be made of rubber, cloth, or soft plastic; the interior may be filled with a gel, water, or another substance with a high heat capacity. A common type used in clinics is a hydro-collator pack, which contains a silicate gel that absorbs water and creates a high heat content. This pack is usually submerged in a self-contained heated water solution. It is removed and wrapped in towels to protect the patient from the pack temperature of 70–80°C. Some hot packs can be heated in a microwave. Other heating devices circulate a fluid through tubes to a pad or cuff. Electrical current can also be used to produce heat in a pad.

A. Physiologic Mechanism
Hot packs warm tissue by conduction. Heat conduction, also called diffusion, is
the direct microscopic exchange of the kinetic energy of particles through the boundary between two systems. Skin tolerance, tissue thermal conductivity, and the body’s responses to increased temperature limit the effective depth of temperature changes to a few centimeters.

**B. Applications**

Hot packs are commonly applied to patients with a variety of conditions, including muscle sprain and strain, tendonitis, bursitis, muscle spasm, contracted muscles or joints, osteoarthritis, rheumatoid arthritis, spasticity, chronic pelvic inflammatory disease, and superficial phlebitis. Hot packs can be used to facilitate hematoma resolution or muscle relaxation prior to traction or stretching.

**C. Prescription**

In writing a prescription for use of hot packs, the patient’s name, age, diagnosis, goal, and precautions are delineated. The prescription should include the area of application and time usage. Time of application can vary from a few to as long as 30 minutes. Frequency of application is typically three times a week. Flexibility can be provided for the therapist by expressing this information as a range; for example, “Hot packs to lumbar sacral area for 10–20 minutes to tolerance.” Extra caution must be used when applying hot packs to insensate individuals as burns have been reported. For this reason, many hospitals use only a very low heat system for this patient group.


**Heat Lamps**

Although inexpensive and versatile, heat lamps are used less frequently in clinics today than in the past. They consist of a bulb (typically 250 W) energized by electricity that emits a light source that can be incandescent or a special infrared. Ordinary incandescent lightbulbs emit large amounts of infrared light, so special tungsten or quartz-type bulbs are rarely necessary. The patient is positioned about 40–50 cm from the light source. Moving the lamp farther away or closer to the target tissue controls the amount of heat administered.
A. Physiologic Mechanism

Heat lamps convert radiant energy to heat. In this case the infrared light generated is converted to heat energy on the target tissue. The distance between the heat lamp and the patient controls the rate of heating and the temperature achieved. The intensity radiating from a point source (energy per unit of area perpendicular to the source) is inversely proportional to the square of the distance from the source.

B. Applications

The applications for heat lamp use are generally the same as those for hot packs. Factors influencing the choice include ease of application to the target tissue (placement can be difficult depending on the patient’s position) and whether the area to be treated is intolerant to pressure. If the patient would benefit most from moist heat, then hot packs will be preferable to a heat lamp.

C. Prescription

In writing a prescription for heat lamp use, the patient’s name, age, and diagnosis are delineated, along with the treatment goal, precautions, area of the body to be treated, and duration. Precautions are included to adjust to tolerance of the patient and to monitor frequently. The typical duration is a few to 20–30 minutes, and frequency is generally three times a week, although daily application for a limited time may be helpful in some patients. An example might be, “Heat lamp to right medial elbow area for 10 minutes to tolerance.” Any integrative modalities should also be listed, along with how they fit into the treatment plan.


Paraffin Baths
Application of superficial heat via paraffin bath utilizes a small container of mineral oil and paraffin wax (1 part mineral oil to 6 or 7 parts wax) that is heated to 52–54°C in a tabletop or standalone unit. The patient immerses a clean limb (usually the hand) into the unit and coats it several times (eg, 7–12), obtaining a thick glove of paraffin wax. The coated limb is then placed in an insulated mitten or towel for up to 30 minutes. The conductive heating provided by this method facilitates active range of motion (ROM), friction massage, and improvements in hand function for patients with rheumatoid arthritis and scleroderma.

A. Physiologic Mechanism

Heat transferred by conduction is obtained by summation of several layers of wax that build up on the affected area, typically the hand. Because paraffin has a low heat capacity and insulates the engulfed tissue, the received temperature of 45–54°C is tolerated by the tissue better than similarly heated water. Heating of the skin to 47°C at first is followed by a decline to a few degrees above baseline. While subcutaneous tissue temperatures can increase 3–5°C, intramuscular and intraarticular temperatures increase only by 1°C. Despite good tolerance by patients, a thermometer inserted in the mixture is recommended for safety. The temperature elevations in the treated tissue decrease after 15–20 minutes.

B. Applications

Paraffin baths have many applications, including subacute or chronic wrist, hand, digit, foot, or ankle injuries; contractures from burns; osteoarthritis; inflammatory arthropathies; and muscle, tendon, and joint contractures.

C. Prescription

The prescription for a paraffin bath should delineate the patient’s name, age, and diagnosis; outline goals; and list precautions, which include specifying “to tolerance of patient” and “monitor frequently.” Duration is 20–30 minutes; frequency can be three times a week for several weeks.

A sample prescription might read, “[Patient Name], 35-year-old male, contracture of digits post-traumatic hand injury. Goals: Increase ROM of digits. Precautions: To tolerance and frequent communication with patient to reference any increased discomfort. Paraffin wax to left hand via dipping technique, wrap for warmth 20–30 minutes, then active assisted and passive ROM to tolerance.”
Whirlpool or Hydrotherapy

In whirlpool therapy, also called hydrotherapy, a tank containing water may be heated, cooled, or agitated to provide the desired treatment effect. Tanks vary in size from 120 to several thousand liters. Large tanks (eg, Hubbard) allow for treatment of multiple body areas simultaneously.

Musculoskeletal injuries, burns, wound cleaning, arthritis, and stiff or frozen joints are the main indications for hydrotherapy. Stiff joints can be heated to facilitate movement, and acute traumatized limbs can be cooled. Immersion in water can decrease stress on bones and joints. Benefits in wound and burn care include removal of gross contaminants, toxic debris, and dilution of bacterial content. Additionally, bandages can be removed more easily following hydrotherapy.

Hydrotherapy uses water as the medium, but alternative media can include pulverized corn cobs or small beads heated and circulated in a chamber by hot jet air (a modality termed fluidotherapy). In this variation, the patient’s limb is placed in a dry, high-temperature (46.1–48.9° C), low-heat-capacity environment. Hand and foot temperatures of 42° C and 39.5° C have been obtained after 20 minutes. Advantages include the ability to perform ROM exercises and massage.

Table 8–2 lists contraindications for whirlpool therapy. The long-term benefits of this approach remain controversial.
A. Physiologic Mechanism

In whirlpool therapy, heat is transferred around tissue by convection. Heating of the body or body parts is facilitated by the agitation of the water (or alternative medium), thus preventing formation of an insulating layer. The temperature of the heated whirlpool water is generally 33–39° C for a patient without evidence of ischemia. If a small area of the body is treated, a greater range of temperatures can usually be tolerated before core body temperature change is noted. Limbs can be heated to a higher temperature (43–45° C) in healthy adults, but in individuals with vascular impairment, temperatures above 32° C should be avoided because of the risk of precipitating an ischemic event. For immersion of larger areas in a patient without vascular impairment, neutral temperatures (33–36° C) are recommended to decrease the likelihood of adverse changes to core body temperature. In healthy people near-total immersion in temperatures up to 40° C (104° F) can be tolerated for short periods; this equates to the maximum usually recommended for personal hot tubs. Exposure time to temperatures higher than that of the normal body must be monitored as the body’s ability to cool itself can be overcome, leading to altered homeostasis.

B. Applications

Whirlpool is well suited for the treatment of wounds and burns. Large tanks such as the Hubbard can accommodate the whole body. The variable agitation, heat, and solvent action can help debride a wound and remove adherent bandages or dead skin. The use of various additives can provide a bactiricidal element to the overall process, aiding in the treatment of infections. However, there is little consensus on which antimicrobials to use or their effectiveness. Among the commonly used antimicrobial agents are sodium hypochlorite, providine–iodine,
chlorhexidine gluconate–isopropanol, and chloramine. Isotonic sodium chloride can be added for treatment of very large wounds to minimize fluid shifts (900 g/100 L) in sensitive individuals.

Concern has been expressed about the possibility that antimicrobial agents used in whirlpool treatments may adversely affect the healing of wounds. Research suggests that overuse of additives may harm the cells involved in tissue repair. For this reason, use of antimicrobial additives should be directed to necrotic, heavily exudative wounds, with monitoring and discontinuation of chemicals as the wound cleans up. Concerns about cross-contamination have led some hydrotherapy providers to use disposable liners for each patient treatment.

Mobilization of joints in patients with arthritis or other musculoskeletal conditions (eg, involving the ankles and wrists, but not limited to them) can be facilitated, particularly after cast removal. Acutely sprained joints can be cooled via whirlpool. Muscle soreness and fatigue in athletes can be addressed with cold-water immersion in a nonagitated tank. (See also Cryotherapy, below).

C. Prescription

In addition to listing the patient’s name, age, diagnosis, goals, and precautions, the hydrotherapy prescription must specify the area to be treated, water temperature, and any additives. Treatment duration is generally 15–20 minutes for each session, although shorter times may be appropriate for some patients. Treatment with additives is continued until the wound starts to improve (eg, a few days to weeks or more).

As an example, a diabetic patient with a leg wound from a burn and contractures of the ankle might benefit from the following prescription: “Whirlpool to left leg wound as tolerated for 15–20 minutes 3 times a week for 2 weeks. Keep temperature below 32° C and use sodium hypochlorite until wound begins to become clean. Please stretch ankle after heating. Discontinue treatment if patient complains of increased pain or worsening of condition and return for evaluation in 2 weeks.”


CRYOTHERAPY

Like therapeutic heat, cryotherapy, the therapeutic application of cold, has utility in treating a wide variety of conditions. Ice and cold are both considered superficial agents. The depth of cold penetration is limited by adipose tissue, and by tissue circulatory responses that attempt to maintain homeostasis, and is generally a few centimeters. Vapor sprays use an evaporative mechanism of action to freeze the outer skin, albeit for a few seconds. Ice packs made of ice, water, and various thin materials are inexpensive and effective options. Cryogel chemical packs are easily transported and stored, and can be instantly activated by punching the pack, which starts an endothermic chemical reaction. Other cryogel packs, which may be soft or hard, are stored in a freezer for later use. Icewater devices with cuffs or tubes can be used to selectively cool specific areas, such as the knee or shoulder, and provide a relatively constant cooling action utilizing circulation from an ice bath reservoir. Large tanks or tubs can be filled with icewater for near-full immersion. Ice massage combines pressure on the tissue in addition to the cold.

The generally accepted effects of cooling are decreases in tissue blood flow (vasoconstriction), tissue metabolism, oxygen utilization, inflammation, muscle spasm, spasticity, nerve conduction, and pain. Decreases in tissue temperature have been shown to decrease muscle force production, muscular power, and proprioception. Table 8–3 lists contraindications for cryotherapy.

Table 8–3 Contraindications for use of cryotherapy.
A. Physiologic Mechanism

The cooling effect of an ice pack begins at the skin, dropping the temperature to 12–13°C in 10 minutes. Subcutaneous tissue temperature decreases 3–5°C in 10 minutes, and deep muscular temperatures falls the least, by a degree or less. With longer periods of cooling (more than 20 minutes, up to 3 hours), forearm muscle temperatures drop 6–16°C, and intraarticular knee temperatures by 5–6°C. Muscle spindles, γ fibers, nerve conduction, and muscle contraction are all influenced by cold. When cooling is provided by means of whirlpool, the agitated water and ice limits formation of an insulation layer.

In muscle injury, the early use of cryotherapy is associated with significantly smaller hematoma formation between the ruptured myofiber stumps, less inflammation, and somewhat accelerated early regeneration. In rheumatoid arthritis, reductions in histamine and intraarticular collagenase have been reported. The initial onset of cooling causes a local reflex and increased sympathetic tone, resulting in vasoconstriction; however, subsequent body response to further cooling may or may not lead to a reactive vasodilation. The dilation of deeper vessels limits the effect of the cold on deeper tissue. Analgesia can be achieved in 7–10 minutes.

B. Applications

Cryotherapy applications include acute soft tissue injuries and trauma, reduction of compartmental pressures, muscle soreness, sprains, strains, spasms, spasticity, fractures, traumatic joint injuries, joint inflammation, joint surgeries, burns, chronic musculoskeletal pain, dental pain, oral surgery, injection sites, and postsurgical sites.
C. Prescription

The prescription of cold modalities must take into account the area to be treated, the tolerance of that area, and the duration of treatment desired. A vaporizing spray will quickly cool the skin; however, the effect lasts for only few seconds. The transient increased pain threshold can be useful in reducing the discomfort of injections of joints or trigger points. Typical application times for cold packs and ice are about 20 minutes. A cryocuff or device can be applied for longer periods (eg, 1–2 hours) in patients after joint surgery, depending on tolerance. Because whirlpool temperatures below 13–15° C are usually uncomfortable, session times for this cooling modality are typically 10–20 minutes.

The patient’s name, age, and diagnosis should be clearly delineated, as well as goals and precautions. A sample prescription might read, “[Patient Name], 44-year-old male, with lumbar sprain. Goal: Reduce spasm and pain. Precautions: To tolerance, frequent monitoring, and discontinue if increased discomfort. Ice to right side lumbar area for 10–20 minutes, 3 times a week for 2 weeks. [Any other modalities to be used should be noted.] Return for evaluation in 2 weeks.”


DEEP HEATING THERAPY

Ultrasound, short-wave diathermy (SWD), and microwave diathermy can attain deep heating of body tissues, which may have benefit for some patients after traumatic injury or surgery. The choice of modality is often based on availability, but subtle differences exist that may influence the practitioner’s decision of which to prescribe.
Therapeutic Ultrasound

Therapeutic ultrasound uses high-frequency (> 20 kHz) sound waves that can be focused, refracted, and reflected. A medium such as a gel or water is required for transmission of sound waves, which have both thermal and nonthermal effects in tissue. Heat production and its effects on tissue are the best known of these effects. Nonthermal effects (eg, cavitation, acoustic streaming, and standing waves) are used in separation technology to obtain blood components, for destruction of blood clots, in scalpel and bone cutting, and to disrupt cell membranes for laboratory evaluation. The nonthermal ability to alter cell membrane permeability and function may have some benefit for wound healing and is used in phonophoresis (see later discussion). More research into these and other nonthermal effects is needed. Generally, thermal effects are generated at higher intensities.

The benefits of therapeutic ultrasound include increased metabolic activity and blood flow, decreased pain, increased tendon extensibility, and the ability to heat tissues at a greater depth than with any other modality. The ideal area to be treated is small, as larger areas require additional windows of treatment. Bone healing in acute and nonunion fractures has been demonstrated. However, despite apparent promotion of the proliferation phase of myoregeneration, this modality has not shown a positive (muscle-healing, enhancing) effect on the final outcome of muscle healing. Contraindications for therapeutic ultrasound are listed in Table 8–4.

Table 8–4 Contraindications for use of therapeutic ultrasound.
A. Physiologic Mechanism

Deep heat is produced when sound wave energy is converted into heat as it passes through body tissues. Travel through tissues is met with little absorption until it reaches structures with high collagen content (bone, periosteum, ligaments, capsules, fascia, tendons, scars, and tissue interfaces). Therapeutic ultrasound typically employs incident waves of either 1 or 3 MHz transmitted as either pulsed or continuous waveforms, depending on the desired physiologic effects. Longer wavelengths tend to penetrate more deeply. Shorter wavelengths interact with tissue at shorter depths. The depth of heating can be up to 8 cm. The total area treated depends on the transducer area and may require several different orientations or windows. Temperature increases of up to 46° C in deep tissues (ie, bone–muscle interface) can occur. Ultrasound-induced temperature rise varies with tissue properties (absorption coefficient, density, perfusion) and device parameters (pulse duration, pulse repetition frequency, beam or scanning configuration). Tissues adjacent to bone are particularly susceptible to significant heat increase via conduction.

B. Applications

Therapeutic ultrasound is commonly used in the treatment of soft tissue injuries,
including muscle, tendon, ligament, and bursa. It may aggravate tissue damage and swelling if used too soon after an injury. Other indications include subacute hematomas, postpartum perineal pain, healing of fractures, degenerative arthritis, contractures of joints or soft tissue, and carpal tunnel syndrome.

**C. Prescription**

After the patient’s name, age, and diagnosis are delineated, the goal should be outlined. The prescription for therapeutic ultrasound should include a frequency setting, pulse ratio, intensity setting, duration, and frequency. More superficial lesions might call for a 3 MHz frequency; for a deeper lesion, 1 MHz would be more appropriate. The pulse ratio is determined by the state of the tissue. An acute tissue state is more energy sensitive, so a larger pulse ratio (lower duty cycle) of 1:4 is best suited. As more chronic tissue states are encountered, a smaller ratio of 1:2 or 1:1 may be appropriate. A continuous mode can also be used for chronic lesions. The intensity (listed as W/cm\(^2\)) is generally confined within 0.5–2.0 W/cm\(^2\), although up to a maximum of 3 W/cm\(^2\) may be administered. When used to facilitate healing the intensity setting should be lower for acute injuries (0.1–0.3) and higher for chronic injuries (0.3–0.8). When used to heat tissue, intensities of 0.8–1.0 W/cm\(^2\) are used for superficial tissues and greater than 1.5 W/cm\(^2\) for deep tissues (eg, in the hip). The duration of treatment is typically 5–10 minutes.

As an example, a prescription for therapeutic ultrasound might state, “[Patient Name], 56-year-old male, frozen left shoulder status–post rotator cuff tear. Goal: Improve ROM and decrease pain. Precautions: To tolerance. Monitor response and stop if increased discomfort. Therapeutic ultrasound to left shoulder 1 MHz, pulse ratio 1:1, 0.8 W/cm\(^2\) for 5–10 minutes, 3 times a week for 3 weeks. Return for evaluation in 3 weeks.”


Wilkin LD, Merrick MA, Kirby TE, Devor ST: Influence of therapeutic

## Short-Wave Diathermy

Radio waves (electromagnetic energy) are converted to heat as they travel through the body. In SWD, various applicators in the form of plates, coils, drums, cables, and mats are arranged around the patient to create a circuit, of which the patient is a part. SWD can be inductive or capacitive. Inductive SWD creates a magnetic field using cables or drums to induce circular electric fields in tissue. This produces high temperatures in water-rich tissues (e.g., muscles, skin). The body acts as a receiver, and eddy currents are induced in the tissues in its field. This can increase tissue temperature 4–6° C above normal. Capacitive applicators utilize electrode plates on either side of the body that produce high temperatures in water-poor tissue (e.g., fat, bone).

The Federal Communications Commission (FCC) limits use to 13.56 MHz (22-m wavelength), 27.12 MHz (11 m), and 40.68 MHz (7.5 m). Most commercial machines operate at a frequency of 27.33 MHz and a wavelength of 11 m. Machines can use continuous or pulsed modes. Contraindications are listed in Table 8–5.

| Table 8–5 | Contraindications to the use of short-wave and microwave diathermy. |
A. Physiologic Mechanism
As the high-frequency waves travel through the body tissues between the condensers or the coils, they are converted into heat. In contrast to ultrasound, SWD can heat large areas of tissue. The degree of heat and depth of penetration depend in part on the absorptive and resistance properties of the tissues that the waves encounter. SWD preferentially heats low impedance tissues such as skeletal muscle, blood, and synovial fluid. Depth of muscle heating is 4–5 cm. Soft tissue treated with SWD maintains tissue temperature increases two to three times longer than with ultrasound.

B. Applications
Indications include chronic prostatitis, refractory pelvic inflammatory disease, myalgia, back spasms, soft tissue injuries, inflammation (joint or tissue), sprains or strains, tendonitis, tenosynovitis, bursitis, rheumatoid arthritis, periostitis, capsulitis, and osteoarthritis. SWD can also be used as an adjunct to stretching of tight tissue and soft tissue mobilization.

C. Prescription
The patient’s name, age, diagnosis, goals, and precautions should be delineated.

<table>
<thead>
<tr>
<th>Superficial heat contraindications (see Table 8–1) plus:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metal</td>
</tr>
<tr>
<td>Jewelry</td>
</tr>
<tr>
<td>Ischemia</td>
</tr>
<tr>
<td>Malignancy</td>
</tr>
<tr>
<td>Implants, pacemakers, stimulators, defibrillators, pumps, etc</td>
</tr>
<tr>
<td>Small clips, intrauterine devices</td>
</tr>
<tr>
<td>Active growth plates</td>
</tr>
<tr>
<td>Infection</td>
</tr>
<tr>
<td>Contact lenses</td>
</tr>
<tr>
<td>Menstruating or pregnant uterus</td>
</tr>
<tr>
<td>Acutely inflamed joints</td>
</tr>
<tr>
<td>Cochlear implants</td>
</tr>
<tr>
<td>Eyes</td>
</tr>
<tr>
<td>Gonads</td>
</tr>
</tbody>
</table>
A prescription for SWD should also include the area to be treated and duration. Dosing is based on the patient’s perception of heating and feedback about pain. Duration of therapy is generally 20–30 minutes. An example might read, “[Patient Name], 47-year old-female, right hip sprain. Goal: Decrease pain and increase ROM. Precautions: To tolerance, communicate frequently with patient, and stop if discomfort increases. SWD to right hip area for 10–15 minutes, 3 times a week for 2 weeks. Active assisted ROM and passive ROM to tolerance after heating. Return for evaluation in 2 weeks.”


 Microwave Diathermy

Similar to SWD, microwave diathermy converts electromagnetic energy (microwaves) into thermal energy. The devices utilize a frequency higher than that of SWD. FCC-approved frequencies are 915 MHz (33-cm wavelength) and 2456 MHz (12 cm). Microwave diathermy units focus their energy as a beam from an antenna and do not use capacitors or inductors. The fraction of power absorbed by the target tissue is dependent on the frequency of the electromagnetic wave, the dielectric constant, and the electrical conductivity of the tissue. This modality was popular decades ago but has largely been replaced by ultrasound and hot packs. Contraindications are the same as those for SWD and are listed in Table 8–5.

A. Physiologic Mechanism

Microwaves do not penetrate tissues as deeply as ultrasound or SWD. Average temperatures obtained are approximately 41° C at a depth of 1–3 cm. Using 915 MHz, subcutaneous fat temperatures may increase to 10–12° C. The 915-MHz applicators are better for heating muscle tissue.

B. Applications
Microwave diathermy may be used to heat superficial muscles and joints such as the shoulder, to speed the resolution of hematomas, and for local hyperthermia in cancer patients.

### C. Prescription

Microwave diathermy can be considered an option if all the precautions and contraindications are satisfied and the tissue depth of heating desired is 1–4 cm. The patient’s name, age, diagnosis, goals, and precautions should be delineated. Duration of treatment is usually 5–10 minutes and frequency is typically three times a week for a few weeks. The patient and provider must wear protective eyewear.


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### ELECTROTHERAPY

Electrical stimulation is used for therapeutic and functional purposes. Transcutaneous electrical nerve stimulation (TENS) can provide relief for patients with musculoskeletal pain using low level electrical current, whereas functional electrical stimulation (FES) has the potential to allow movement and function in a paretic and nonfunctional limb. (FES is sometimes referred to as functional neuromuscular stimulation [FNS] or neuromuscular electrical stimulation [NMES].)

#### Transcutaneous Electrical Nerve Stimulation

A transcutaneous electrical nerve stimulator is a small, portable, battery-powered device that interfaces with a patient by means of 2- or 4-wire connected leads or pads. The primary aim of the device is to aid in pain relief. The TENS unit has several controls that typically include intensity, frequency (pulse rate), pulse width, mode, and time. Intensity control ranges from 0 to 100 mA or 1–10 on a dial. The frequency settings range from 2 to 250 Hz. High frequency is considered to be 50–130 Hz, and low frequency is considered to be 2–10 Hz.

Burst mode uses a combination of a high-frequency setting but at low–frequency intervals (2–3 seconds). The pulse width ranges from 10 to 1000
microseconds. Some machines feature a multimodal or modulation mode in which the TENS units randomly apply different modes and settings in an attempt to avoid neuromodulation (ie, the habituation effect). The duration of the stimulus can be varied along with the frequency. The time of application is also variable. Some of these units may have preset time controls (eg, 15–30 minutes, or continuous operation).

Intense stimulations of high intensity and long duration are less well tolerated; the time of application for this type of use is usually 15–30 minutes. With other modes, time of application is usually 30 minutes or longer, and it is not uncommon for TENS to be applied for several hours at one session.

The efficacy of TENS in published reports is mixed. The multiple variables and heterogeneous designs of investigations has produced calls for larger, more powerful studies to substantiate its use. Table 8–6 lists contraindications for electrical therapy (TENS and FES).

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**Table 8–6** Contraindications for use of electrical therapy.

<table>
<thead>
<tr>
<th>Contraindication</th>
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<tbody>
<tr>
<td>Stimulation over carotid sinus or over/across heart</td>
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<tr>
<td>Pregnancy</td>
</tr>
<tr>
<td>Seizure disorders</td>
</tr>
<tr>
<td>Malignancy</td>
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<tr>
<td>Insensate or atrophic skin</td>
</tr>
<tr>
<td>Allergies to gels or pads</td>
</tr>
<tr>
<td>Inability to report stimulation or voice pain</td>
</tr>
<tr>
<td>Arterial or venous thrombosis, thrombophlebitis</td>
</tr>
<tr>
<td>Transcerebral, ocular, oral, or anterior neck electrode placement</td>
</tr>
</tbody>
</table>

**A. Physiologic Mechanism**

The mechanism of action by which TENS achieves its effects is not known. Most theories focus on the relay of composite nerve signals in the sensory nervous system. The gate control theory posits that the TENS unit activates Aβ (100 Hz) sensory fibers. These fibers then compete with Cδ pain fibers for priority in the brain, thereby reducing the perception of pain. An alternative theory holds that Aδ fibers (2–5 Hz) are activated by the TENS unit, which then causes the release of endogenous opioids in the spinal cord (encephlin),
providing pain relief.

A combination approach postulates the simultaneous activation of both the Aβ and Aδ fibers when high-frequency stimulation at 2- to 3-second intervals (burst mode) is used. The electrodes are subjectively placed for maximum benefit and can be distal or proximal to painful area.

B. Applications

TENS is used for musculoskeletal pain, headache, temporomandibular joint pain, pelvic pain, visceral abdominal pain, dysmenorrhea, neuropathic pain, arthritis, dental pain, and postoperative recovery.

C. Prescription

The significant variability in response to TENS units necessitates that clinicians choose among three different implementation options. The first is a conventional approach in which the intensity is adjusted to a comfortable level, providing current at 10–30 mA, frequency of greater than 100 Hz, pulse duration of less than 200 microseconds, and duration of 30 minutes or a few hours.

The second is an acupuncture-like setting, with frequency of 1–10 Hz, pulse duration of greater than 200 microseconds, and a high stimulus intensity at or near the threshold of tolerance for the patient. This setting, although it can be uncomfortable, may be considered for patients who fail to respond to a conventional approach. Patients rarely tolerate a duration of more than 15–30 minutes.

The third approach uses repeated pulsed bursts of TENS. A train of high-frequency bursts (100 Hz) is cycled at 1–2 bursts per second. Ultimately, the patient response dictates the settings, and patients should be taught how to safely vary the settings on the TENS unit. The duration using this setting is usually 30–60 minutes.

Because of the variability in treatment options and patient responses, the prescription is most often written as a TENS trial, and the clinician works with the patient to adjust the parameters for maximum benefit. Further research may help to define the utility of specific parameters for specific conditions.


Functional Electrical Stimulation

In FES, an electrical device interfaces with a patient’s nerve or muscle tissue via surface or implantable electrodes to cause a functional movement. Immobilized or injured muscles, denervated muscle, and muscles affected by upper motor neuron injury are common targets of FES. Applications can be therapeutic or
functional. Functional uses include standing or ambulatory activities, upper limb performance of activities of daily living, and control of respiration and bladder function by means of a neuroprosthesis (a device or system that uses FES). Commercial systems for use in patients with foot drop and upper extremity problems are available. Therapeutic effects include motor relearning, treatment of hemiplegic shoulder pain, spasticity, injured muscles, cardiovascular conditioning in spinal cord injury, wounds, prevention of muscle atrophy, disuse osteoporosis, fracture healing, and deep vein thrombosis. Research is increasingly focusing on new technologies such as harnessing cortical control signals that may provide an enhanced means of interfacing with a neuroprosthesis to facilitate functional movement.

A. Physiologic Mechanism

IN FES, an electrical current is applied through electrodes to one or more targeted muscles. The muscle size, position of the electrodes, and amount of intervening tissue between muscle and skin dictate the intensity needed from the FES unit to cause a contraction. Because the threshold for eliciting a nerve fiber action potential is 100–1000 times less than for muscle fiber stimulation, FES systems act by stimulating the nerve itself or the motor point of the nerve prior to the neuromuscular junction. Nerve fiber recruitment and resultant force characteristics of muscle contraction are modulated by both stimulus pulse width and stimulus frequency. Limitations of FES include lack of effective control interfaces and rapid muscle fatigue. The recruitment of nerve fibers using FES is the opposite of the body’s normal physiology (which uses small diameter before large diameter). The nerve stimulus threshold is inversely proportional to the diameter of the neuron. Thus, large-diameter nerve fibers, which innervate larger motor units, are recruited preferentially. Fatigue with FES can be a factor because the large-diameter motor fibers innervate fatigable fibers.

Parameters include intensity, pulse duration, waveform type, and frequency. The intensity of the stimulus is slowly increased to achieve a threshold contraction of the muscle desired. This is often referred to as the “ramp up” or “ramp down” and can be variably programmed. Frequency usually ranges between 10 and 100 Hz. The pulse cycle duration is usually about 150–200 microseconds for small muscles and 200–350 microseconds for large muscles. The stimulus on time or off time is altered to achieve the effect desired.

B. Applications

FES is an effective therapy for hemiplegia, spinal cord injury, multiple sclerosis,
peripheral nerve injury, and weak muscles that are difficult for a patient to volitionally contract. FES aids in maintaining muscle mass after immobilization, decreases bone loss in patients with spinal cord injury, and prevents complications from immobility such as deep vein thrombosis, osteoporosis, and loss of cardiovascular conditioning (in spinal cord injury patients with chronic obstructive pulmonary disease).

**C. Prescription**

The patient’s name, age, diagnosis, goals, and precautions should be delineated. For a patient with musculoskeletal injuries, the prescription might then read, “FES to right upper and middle trapezius for 5–10 minutes. Followed by [other modalities/treatments].” For the a patient with a cerebrovascular accident, it might read, “FES to right extensor forearm. Have patient attempt contraction then facilitate with FES.” Session duration is not typically specified as considerable time is needed to obtain the correct placement for maximal response. In addition, the contraction of a muscle can be uncomfortable and tolerance is specific to the individual. Commercially available FES systems are written as a durable medical equipment (DME) item.


**Iontophoresis**

Electrical stimulation of charged chemicals can cause their passage through the
skin. Transdermal administration of drugs via iontophoresis allows for localized delivery. With this method, a small electrical device is connected to the patient via a positive and a negative electrode. Active transport is created by the passage of the electrical current through the electrodes to the charged particles. This form of delivery is usually well tolerated and has the advantage of eliminating the need for injection, which is sometimes painful or traumatic. The most common adverse effects are irritation under the electrode, warmth, itching, and tingling at the application site. Transient erythema and urticaria have been reported. The extent of the penetration is related to tissue thickness, treatment time, and local blood flow. Iontophoresis of ionized drugs provides a 20- to 60-fold increase in skin penetration over topical application. Many ionized drugs are available, including lidocaine, epinephrine, methylprednisolone succinate, dexamethasone phosphate, several antivirals, and various antibiotics. Botulinum toxin has been used in patients with palmar hyperhidrosis with positive results. However, the efficacy of iontophoresis for musculoskeletal pain syndromes is mixed. The contraindications for iontophoresis are similar to those for electrical therapy (see Table 8–6), and include allergy to the drug being used.

A. Physiologic Mechanism
For a drug to be used in iontophoresis it must be able to transfer electrical energy. Like charges will repel each other, so a positive electrode will repel cationic drugs (eg, dexamethasone) and a negative electrode will repel anionic drugs. The cathodic stimulation of cationic drugs requires a current, which is usually measured in milliamperes, ranging from 0.1 to 40 mA and a duration time of 10–40 minutes. The depth of penetration of dexamethasone has been reported to be 3 cm in some patients, although significant variation is seen.

B. Applications
Indications for iontophoresis include carpal tunnel syndrome, bursitis, lateral epicondylitis, achilles tendonitis, heel spurs, plantar fasciitis, hyperhidrosis, local anesthesia for skin biopsies or eyelid surgery, cutaneous cutdowns in dialysis patients, preinjection topical anesthesia, and postherpetic neuralgia.

C. Prescription
The patient’s name, age, diagnosis, goals, and precautions, as well as the area to be treated and the drug to be used, are included in the iontophoresis prescription. As an example, “[Patient Name], 27-year-old male, with lateral epicondylitis.
Goal: Decrease pain. Precautions: To tolerance, monitor for safety and comfort. Iontophoresis with dexamethasone solution, 4 mg/mL, to the left lateral epicondyle area, 3 times a week for 3 weeks. Return for evaluation in 3 weeks.”


**PHONOPHORESIS**

Phonophoresis is the technique of delivering medications (most often pain-relieving drugs) transdermally by means of ultrasonic sound waves that increase cell permeability. Corticosteroids such as hydrocortisone acetate and dexamethasone are most commonly used; however, anesthetics such as lidocaine 1% have been administered using this method. The technique requires that the drug be applied with a coupling medium. The efficacy and optimal parameters of phonophoresis are not well defined and have been questioned. The contraindications for phonophoresis are nearly identical to those for therapeutic ultrasound (see Table 8–4), and include allergy to the drug being used.
A. Physiologic Mechanism
The nonthermal effects of ultrasound on cell permeability are the basis for the results ascribed to phonophoresis. The typical ultrasound setting employed is 1.5 W/cm² at a frequency of 1–2 MHz; the delivery can be either pulsed or continuous. The area being treated may be pretreated with ultrasound, shaved, or covered after treatment to augment the absorption of medication. The amount of medication used per session and frequency of sessions varies and guidelines are not well defined.

B. Applications
Phonophoresis has been used for sarcoid nodules, keloids, shoulder pain, lateral epicondylitis, tenosynovitis, and tendonitis.

C. Prescription
After listing the patient’s name, age, diagnosis, goals, and precautions, a sample prescription might specify, “Phonophoresis with dexamethasone cream, 4 mg/g, to affected area for 5–7 minutes.”


Hoppenrath T, Ciccone CD: Is there evidence that phonophoresis is more effective than ultrasound in treating pain associated with lateral epicondylitis? Phys Ther 2006;86:136–140.

LOW-LEVEL LASER THERAPY
Low-level laser therapy (LLLT) utilizes a small device that emits a monochromatic, coherent light in the red and near-infrared ranges (wavelength 600–1000 nm) with wattages from 5 to 500 mW. These nonwarming light waves are directed through a probe to the desired tissue. Often called “cold lasers,” low-level lasers do not produce sensation nor do they burn the skin. Commonly used devices include gallium–arsenide (GaAs), gallium–arsenide–aluminum (GaAsAl), and helium–neon (HeNe) lasers. GaAs lasers using the 904-nm wavelength are most often used for pain and inflammation because they provide
the deepest tissue penetration. In recent years an inexpensive LED version (using noncoherent light) has become available as a different light source.

Confusion exists about the mechanisms of action of LLLT at the molecular, cellular, and tissue level. Although approved by the Food and Drug Administration as safe for use in the United States since 2002, the efficacy of these devices is mixed. Several studies have shown a response to LLLT in the treatment of various conditions; others have failed to show significant benefit. The wide range of study parameters (eg, wavelength, fluence, irradiance, treatment timing and repetition, pulsing, and polarization) and small sample sizes make it difficult to draw definitive conclusions. Contraindications for LLLT are listed in Table 8–7.

Table 8–7 Contraindications for use of low-level laser therapy.

<table>
<thead>
<tr>
<th>Absolute</th>
</tr>
</thead>
<tbody>
<tr>
<td>Direct irradiation of the eyes</td>
</tr>
<tr>
<td>Pregnancy</td>
</tr>
<tr>
<td>Carcinoma</td>
</tr>
<tr>
<td>Thyroid gland</td>
</tr>
<tr>
<td>Hemorrhage</td>
</tr>
<tr>
<td>Immunosuppressant drug therapy</td>
</tr>
<tr>
<td>Treatment over sympathetic ganglia, vagus nerves, or cardiac region</td>
</tr>
<tr>
<td>(in patients with heart disease)</td>
</tr>
<tr>
<td>Active growth plates</td>
</tr>
<tr>
<td>Relative</td>
</tr>
<tr>
<td>Infected tissue</td>
</tr>
<tr>
<td>Reproductive organs</td>
</tr>
<tr>
<td>Impaired sensation or responsiveness</td>
</tr>
</tbody>
</table>

A. Physiologic Mechanism

In laser therapy, the photons emitted cause a nonthermal photobiomodulation response, altering cell and tissue function. At the cellular level, LLLT may cause photodissociation of nitric oxide from complex transmembrane structures. Increased cellular adenosine triphosphate (ATP) produced by LLLT may
contribute to the positive effects, both by raising cellular energy levels and by upregulating the cyclic adenosine monophosphate (cAMP) molecule (biochemically formed from ATP) that is involved in many signaling pathways. The increased ATP production by the mitochondria and increased oxygen consumption on the cellular level may relax muscles, increase serotonin, increase endorphins, increase antiinflammatory effects (through reduced prostaglandin synthesis), improve blood circulation to the skin (eg, in neuralgia and diabetes), decrease permeability of the nerve cell membrane for Na/K, causing hyperpolarization, increase lymphatic flow, and decrease edema. Despite ongoing research, the biochemical mechanism underlying the therapeutic effects of LLLT is not yet well established.

B. Applications
Among the varied uses for LLLT are musculoskeletal pain (including carpal tunnel syndrome); myofacial pain; trigger points; treatment of low back, neck, shoulder, elbow, or facet joints; radiculopathy or osteoarthritis; patellofemoral disorder; wound healing; oral mucositis or dental pain; as a component of psoriasis treatment; acne treatment or folliculitis; stroke; traumatic brain injury; degenerative central nervous system disease; and spasticity.

C. Prescription
Any prescription for LLLT must include a caution to use laser protective eyewear (for both patient and provider). Although the wide variety of conditions treated and LLLT parameters precludes general prescription ranges, a few examples can illustrate the information typically specified. For instance, mild to moderately severe carpal tunnel syndrome has been treated with infrared laser dosages ranging from 6 to 13.5 J per treatment point, and two to five points per session; lateral humeral epicondylitis with infrared laser dosages of 4 J per treatment point, and one to two points per session; and rotator cuff tendinitis with dosages of 10 J per treatment point, and two to three points per session. Treatment sessions usually number 10–15 over 2–3 weeks.

Alfredo PP, Bjordal JM, Dreyer SH, et al: Efficacy of low level laser therapy


De Luigi J: Complementary and alternative medicine in osteoarthritis. PM R 2012;4:5S.


TRACTION

Traction is the technique of using a pulling force to overcome the static positioning of joints, bones, and soft tissues. The basic concept is simple, and anatomic studies have provided evidence for separation of vertebral bodies and foraminal spaces. However, scientifically rigorous evidence-based studies are lacking and efficacy of traction remains questionable, being short term and limited at best. Despite this, and possibly because of anecdotal reports of success, cervical traction and lumbar traction remain options in the rehabilitation of patients with radiating neck and back pain.

Lumbar spinal traction can be continuous (hours to days), sustained (20–60 minutes), or intermittent (alternating traction and relaxation with cycles of a few
minutes or less). It can be applied manually or with the assistance of a machine. Lumbar traction requires a high force (30–50% of body weight), often 50–150 lb. A harness with a motor or pulleys and weights are traditional mechanisms. Some traction tables are split so that the pull (force) is more effectively placed on the patient rather than on overcoming friction. Inversion traction for the lumbar spine requires the patient to place his or her feet in special boots that essentially hang the patient upside down. The side effects of increased blood pressure, facial petechiae, headaches, vision changes, and joint pain limit its appeal.

In the past, cervical traction units were often connected to a weight (water-filled bag) suspended over a door. Fortunately, the lack of adequate force generation and difficulty positioning the patient contributed to the fazing out of this type of cervical traction from common use. More recently, neck devices utilizing air bladders that distract the neck from the shoulders or trunk have been applied to a supine patient with the neck in 20–30 degrees of flexion. Contraindications for traction are listed in Table 8–8.

Table 8–8 Contraindications for use of traction.

<table>
<thead>
<tr>
<th>General Traction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligamentous instability</td>
</tr>
<tr>
<td>Osteomyelitis</td>
</tr>
<tr>
<td>Discitis</td>
</tr>
<tr>
<td>Primary or metastatic bone tumor</td>
</tr>
<tr>
<td>Spinal cord tumor</td>
</tr>
<tr>
<td>Severe osteoporosis</td>
</tr>
<tr>
<td>Untreated hypertension</td>
</tr>
<tr>
<td>Severe anxiety</td>
</tr>
<tr>
<td>Clinical signs of myelopathy</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Cervical Traction</td>
</tr>
<tr>
<td>Vertebral basilar insufficiency</td>
</tr>
<tr>
<td>Rheumatoid arthritis or connective tissue disorder</td>
</tr>
<tr>
<td>Midline herniated nucleus pulposus</td>
</tr>
<tr>
<td>Acute torticollis</td>
</tr>
</tbody>
</table>
A. Physiologic Mechanism

Anatomic studies have shown enlargement of intervertebral foramina, separation of apophyseal joints, stretching of muscles and ligaments, and reduction in prolapsed discs and nucleus pulposis pressure after the application of traction. Some investigators have concluded that 2–20 mm of elongation of the cervical spine is possible with 25 lb or more of tractive force. The average for the lumbar spine is slightly more than 3 mm at one intervertebral foramen. Normalization of bilateral somatosensory evoked potentials has been reported in lumbosacral radiculopathies.

B. Applications

Relying on its anatomic effects rather than on evidence-based indications, the conditions most relevant for traction seem to be radiculopathies of the cervical or lumbar spine. Given the lack of efficacy and the potential for real harm, careful patient selection is necessary.

C. Prescription

The patient’s name, age, diagnosis, goals, and precautions are listed. Because soft tissue limits the force reaching vertebrae, modalities to relax the muscles may be used to increase tolerance. Intermittent or sustained traction should be delineated and, owing to lack of clinical guidelines, patient comfort may dictate which is used. The duration of treatment is generally 10–30 minutes and frequency is three to five times a week for 3–6 weeks. Lastly and most importantly, guidelines for discontinuation should be outlined. If symptoms become worse or new symptoms develop (eg, pain, dizziness, weakness, autonomic symptoms) traction should be discontinued.

A sample prescription might read, “[Patient Name], 49-year-old male. Diagnosis: Cervical radiculopathy, left side. Goal: Decrease or eliminate arm pain. Precautions: To tolerance, monitor response to traction, avoid cervical extension. Please use superficial heat and massage to cervical paraspinal area to tolerance for 8–10 minutes prior to cervical traction in order to relax the muscles favorably for cervical traction. Intermittent cervical traction with patient supine and head flexed 20–30 degrees. Start with low weight and slowly adjust to tolerance each session or over a number of sessions. Attempt to obtain at least 25 lb of weight. Maximum cervical weight: 45 lb. Duration: 10–30 minutes. Frequency: 3 times a week for 2 weeks. Return for evaluation in 2 weeks.”

Home traction should only be used after a clinic trial of traction has been
successful.


**MASSAGE**

Massage therapy is one of the oldest therapeutic modalities and, given that only a pair of trained hands are necessary, the simplest. Classic or western massage is basically synonymous with Swedish massage. Four techniques are used. *Effleurage* is the focus of pressure via hands gliding across skin. *Petrissage* involves compression of the underlying tissue (skin and muscle) between the fingers and thumb of one hand or between the two hands of the practitioner. *Tapotement* or percussion massage involves rhythmical, repetitive blows of the hands on the target tissue. Deep tissue, friction, or deep friction massage all refer to the use of gradually increased pressure applied to target tissue, usually with the heel of the hand or thumbs. Multiple other types of massages exist. Massage techniques are often integrated with other methods that focus on relaxation, posture, meditation, rolling, kneading, nutrition, and so on.

Western-style massage techniques are often used for musculoskeletal problems, and eastern techniques for visceral or musculoskeletal complaints. The physiatrist may prescribe massage to decrease swelling in the limbs secondary to stroke or lymphedema, or pain from muscle injury. Some integrative health systems incorporate therapeutic massage into complementary/wellness programs or as adjuncts in cancer, pediatric, or psychiatric care, or surgical procedures. *Table 8–9* lists contraindications for massage.

*Table 8–9* Contraindications for use of massage.
**A. Physiologic Mechanism**

Vascular changes from the mechanical pressure of massage are thought to result from the progression of fluid to lower pressure areas ahead of the working hands. Mobilized fluid from around cells enters the lymphatic or venous system where it is precluded from return by valvular mechanisms. Amounts of fluid moved are small and significant effects on the heart have not been reported. Massage has been reported to decrease blood viscosity, hematocrit, and lactate, and to increase circulating fibrinolytic compounds, natural killer cells, myoglobin, glutamic oxaloacetic transaminase, creatinine kinase, and lactic dehydrogenase. Histamine is also released and can assist in local vasodilation. The decrease in spasms and muscle recovery attributed to massage may result from the mobilization of metabolic waste products out of target tissue, and from enhanced blood flow. Stimulation of cutaneous receptors and spindle receptors in superficial skeletal muscle facilitates more distant reflexive effects on the somatic and visceral nervous system.

**B. Applications**

Massage is effective for muscle sprain, strain, fatigue, contracture, spasm, tension relief, anxiety, lymphedema, contractures, stiff joints, fibromyalgia, and
edema caused by weakened limbs. Benefits of massage have also been reported in Parkinson’s disease.

**C. Prescription**

In addition to the patient’s name, age, diagnosis, and precautions, goals should be delineated, such as edema reduction in a particular limb or other area of the body. Adjuncts to massage, such as passive ROM or other modalities, should be clearly stated. If the technique is important, it should be noted. For example, an injured muscle such as the levator scapula may require deep tissue massage because of its location; however, in a patient with fibromyalgia, use of this technique might be very painful. The duration of massage depends on the size of area being treated, chronicity, and use of other treatment modalities. If included as part of a multimodal approach, massage may be applied for 10–15 minutes. Individually, sessions of 30 minutes or more are not unusual. Duration of therapy is typically three times a week for 2–4 weeks, followed by reevaluation.

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Therapeutic exercise encompasses basic physiology and physiologic sciences as well as exercise science. This chapter briefly outlines the use of physical activity, muscular contraction, and physical exertion in order to prevent, treat, and rehabilitate physical conditions as well as improve performance or fitness. Exercise can be used to increase or improve strength, endurance, aerobic capacity and conditioning, flexibility, body mechanics, and proprioception, and can help a patient progress toward functional movements and activities. Although the chapter summarizes essential knowledge, readers are encouraged to explore additional sources for more comprehensive details.

AEROBIC & ANAEROBIC METABOLISM

Adenosine triphosphate (ATP) is the primary energy substrate used by muscle cells (myocytes) to generate contractile force during exercise. When the amount of intramuscular ATP stores is depleted after 10 seconds of exercise, it must be replenished. ATP regeneration occurs through three processes, using aerobic (ie, inclusive of O₂) and anaerobic (exclusive of O₂) avenues. All three ATP generation systems occur simultaneously. Activities that are shorter in duration and have greater intensity rely on anaerobic systems, whereas those of lower intensity and longer duration rely on aerobic systems.
The two anaerobic systems fueling muscle activity are the adenosine triphosphate–phosphocreatine (ATP-PC) system and anaerobic glycolysis. Of the two, the ATP-PC system regenerates ATP more quickly because it involves only a single chemical reaction. In this reaction, the bond between phosphate (P) and creatine (C) in phosphocreatine (PC) is broken, releasing energy and a phosphate group to resynthesize ATP from ADP: PC + ADP → ATP + P. Unfortunately, the PC stores of myocytes can sustain only about 30 seconds of activity. Activities involving short, powerful bursts (eg, the swing of a baseball bat, a 100-m sprint, or powerlifting) primarily rely on the ATP-PC system.

Glycolysis, the second aerobic system, involves a series of reactions whereby simple (glucose) and long-chain (glycogen) carbohydrates are metabolized into pyruvate. During both aerobic and anaerobic glycolysis, ATP and pyruvate are synthesized. However, during anaerobic conditions, pyruvate is preferentially fermented into lactate so that glycolysis, and its ATP generation, can continue. Anaerobic glycolysis is, nevertheless, limited in its ability to generate ATP because the acidification of cytosol by lactate eventually impedes glycolytic enzyme activity. Interestingly, although lactate can impede glycolysis, some lactate is transferred to the systemic circulation. There, lactate may be converted to glycogen in the liver through the Cori cycle. Alternatively, intracellular and extracellular lactate may be oxidized into pyruvate and then metabolized for ATP in a process known as the lactate shuttle. Anaerobic glycolysis produces more ATP over a longer time frame than the ATP-PC system; however, both anaerobic systems generate less than 10% of the body’s potential ATP. Highly intense, short-duration exercises of up to 2 minutes, such as 400-m sprints or repetitive weight lifting, will exhaust both systems.

Aerobic Metabolism

On the other hand, in aerobic conditions, pyruvate is metabolized into acetyl-coenzyme A (CoA) before entering the tricarboxylic acid (TCA) cycle in the mitochondria. Noncarbohydrate energy sources, such as fats and proteins, are also catabolized into the acetyl-CoA substrate, but at slower rates than carbohydrates. The TCA cycle produces ATP as well as most of the energy-rich oxidizing agents used in oxidative phosphorylation through the electron transport chain. The remaining 90% of ATP is generated by means of oxidative phosphorylation, which is the chief source of ATP production after exhausting the ATP-PC and anaerobic glycolytic systems. This occurs in endurance activities lasting more than 3–4 minutes, such as long-distance running over a
distance of more than 1 mile and cross-country skiing events.


MUSCLE STRUCTURE, PHYSIOLOGY, & BIOMECHANICAL PRINCIPLES

The potential energy of ATP must be converted into mechanical force to create movement during exercise. Such a process occurs in muscular tissue with great efficiency. Muscles are composed of fascicles containing myocytes. Contraction of the myocytes relies on the extensive network of myofibrils filling the cytoplasm. A myofibril is either a thick myosin myofilament, or a thin actin myofilament. Both types of myofilament overlap near each other in units called sarcomeres, as shown in Figure 9–1.

▲ Figure 9–1 A skeletal muscle sarcomere is depicted with actin and myosin filaments as well as titin elastic fibers.
The sarcomeres line up end to end (which, in skeletal muscle, accounts for its striated appearance). Muscles contract by means of an overlapping phenomenon involving sliding filaments. For contraction to occur, calcium ions must first be released into the cytoplasm from their storage location in the sarcoplasmic reticulum of the myocyte. Actin is composed of troponin and tropomyosin molecules; in the absence of calcium, tropomyosin blocks the myosin head from interacting with actin. However, when calcium is present it binds to troponin, preventing tropomyosin from interfering with the myosin binding site on actin. This frees troponin to bind myosin. ATP can then be converted to energy and ADP at the myosin head. The newly released energy breaks the bond at the actin–myosin head, allowing the myosin head to bind to another actin site, contracting the sarcomere length, and thus the myocyte and muscle tissue length. Force is generated when the muscle contracts against load.

Muscle Length versus Force

The optimal muscle length for active force generation is known as the natural resting length. At the level of the sarcomere, this is the length at which actin and myosin overlap maximally without collision of opposing actin filaments, resisting further contraction. A muscle is typically near natural resting length at the midrange of joint motion if the muscle acts on that joint. The total tension generated by a muscle is the sum of active muscle tension and passive elastic tension. Active tension is created by actin–myosin head interaction; passive tension is derived mainly from titin fibers that connect the myosin to actin filaments, as seen in Figure 9–2. At lengths shorter than neutral resting length, total tension is simply a factor of active contraction of the muscle. But as the muscle is lengthened, the total tension is the combination of both active and passive tension. Active tension decreases at lengths greater than neutral resting length, whereas passive tension continues to increase. In general, the maximum overall tension within a muscle is at a length slightly longer than its resting length.
Muscle Types

Skeletal muscle fibers do not all contract in the same manner, and researchers have divided fibers into subtypes based on contraction speed and endurance. Each subtype varies in how quickly ATP is consumed, sources of fuel utilized for ATP, speed of muscle contraction, and rate of fatigue. The two categories are type I (slow twitch) and type II (fast twitch). Type I fibers generally have fewer myofibrils in their cytoplasm, which results in less muscle bulk and force generation. Myosin head–actin crosslinking and ATP usage is also two to three times slower in type I fibers compared with type II. However, type I myosin fiber contractions can last up to ten times longer than those of type II. Type I fibers possess significant amounts of mitochondria, oxygen-binding myoglobin, and capillary access. These factors give the muscle fibers a distinctive dark-red color and, more importantly, mean that they excel at oxidative phosphorylation. Type II fibers, which lack the mitochondria and myoglobin content of type I fibers, rely more heavily on ATP-PC and anaerobic glycolysis for fast ATP production. These fibers have copious actin–myosin filaments, creating greater cross-sectional muscle fiber volume, and also force production. It is apt to state that type II fibers are designed for sprinting, whereas type I are geared for endurance activities.

Although this binary distinction between fiber types is useful conceptually, in
reality a continuum of fibers shares characteristics of both fiber types. Additionally, type II fibers are subdivided into three categories, designated IIa, IIx, and IIb. Type IIa fibers, which produce greater force than type I, are relatively fatigue resistant and have high oxidative and glycolytic capacities. Type IIx fibers produce greater force at the cost of slightly less oxidative capacity and fatigue resistance. Finally, type IIb fibers are most similar to the type II fibers discussed earlier, with high glycolytic and force generation abilities but poor oxidative capacity and fatigue resistance.

Muscle Contraction

A *concentric contraction* is a muscle-shortening contraction. When the force generated by the muscle is sufficient to overcome resistance during the contraction, the muscle will shorten. An *eccentric contraction* is a muscle-lengthening contraction. This occurs when the force generated by the muscle is insufficient to overcome the external load and the muscle lengthens despite contraction. This type of contraction can also be used to decelerate and control movements of limbs and joints as well as to control the movement of external objects being lifted or carried. An *isometric contraction* is a contraction of the muscle that involves neither muscle shortening nor lengthening (ie, no joint movement). The tension in the muscle equals the load; thus, no movement is created. The length of the muscle during contraction has an impact on the force it can generate during isometric contraction. The natural resting length of the muscle (described earlier) yields the highest isometric force, and when the length from this neutral point either increases or decreases, the isometric force will decrease.

The muscle–tendon insertion site, angle of the insertion, and distance of insertion from the center of joint rotation all affect muscle leverage, which can be represented by torque. In biomechanics, torque is the product of the distance of a force from the center of joint rotation and the magnitude of the perpendicular force vector. If the insertion angle of the tendon is at 90 degrees (ie, perpendicular to the bone), then torque production is the greatest for a given tension in the muscle. As the angle increases or decreases from 90 degrees, the torque will also decrease. Tendon insertion distance from the center of rotation also affects torque. Insertion distances further from the center of rotation increase the torque produced by a muscle for a given contractile force, but at the expense of total excursion of movement, or angular range. For a given contraction, the muscle will shorten a certain amount, and if tendon insertion is
further from the center of rotation of the joint, the excursion of movement of that joint will be less than if the tendon inserted closer to the center of rotation. Slight anatomic differences in muscle–tendon or fiber insertion points may allow different force or torque production for a given contractile effort and could account for variability in performance capabilities.

Variability in muscle fiber alignment also affects the force generation and contraction speed of a muscle. Alignment is defined as the angle between the muscle fibers and a line drawn between the origin and insertion of the entire muscle, which is typically the plane of force generation. The simplest arrangement exists when fibers run parallel to the force generation plane; these are called fusiform fibers. Compare this with pennate fibers, in which there is a 5- to 30-degree angle between the fibers and the force generation vector. This arrangement gives pennate fibers an advantage over fusiform as it allows for higher densities of fibers in a cross-sectional area. Comparing cars parked in parallel with cars parked diagonally provides an analogy; more cars fit within a given space when parked diagonally rather than parallel parked. With muscles, however, greater force generation comes at the cost of overall muscle contraction speed, as each fiber needs to contract a longer distance to shorten the entire muscle. Although fusiform fibers may have a smaller cross-sectional fiber area, they are capable of contracting significantly faster than pennate fibers. Examples of pennate muscles include the soleus and vastus medialis muscles. Sartorius and biceps brachii muscles have a fusiform arrangement.

Contraction velocity also affects torque. In the early 20th century, Hill proposed a force–velocity relationship showing that muscle torque is greatest during isometric contraction and decreases hyperbolically as contraction velocity increases. This theoretical curve is represented in Figure 9–3, with force expressed as torque.
Figure 9–3 Hill’s proposed force–velocity curve. Eccentric contractions have only shown greater force production compared with isometric contraction in laboratory modeling, as predicted by Hill (dotted line). However, physiologically, maximal eccentric force production is achieved at approximately isometric levels (solid line).

The isometric and concentric portion of this model has been supported experimentally; for example, isokinetic devices have measured muscle-generated torque showing that there is less torque as the velocity of concentric contraction (measured in revolutions per minute) increases. Eccentric contractions have, at least in vivo, been shown to produce significantly greater forces than isometric contractions. In situ, voluntary eccentric muscle contraction force in humans supersedes isometric contraction force only minimally, if at all, and may be limited by a neurologic tension-regulating mechanism to protect the muscle tissue. Additionally, despite diminished contractile force at higher concentric contraction velocities, muscular power (force times velocity) increases.


**NEUROMUSCULAR PRINCIPLES**

A motor unit consists of a group of myocytes and the α motor neuron axon innervating them. The muscle fibers within a motor unit contract simultaneously when an action potential propagates from the α motor neuron’s cell body and down its axon, causing acetylcholine release from the presynaptic axon terminal into the neuromuscular junction. Acetylcholine is a neurotransmitter that binds the postsynaptic nicotinic acetylcholine receptors on the postsynaptic myocyte membrane surface, inducing calcium release from the sarcoplasmic reticulum of the postsynaptic myocyte. The end result is a contraction, as described previously by the sliding filament theory. Different motor units are dispersed throughout the cross-sectional area of a muscle. Each motor unit varies in numbers of fibers and fiber subtype composition. The overall magnitude of a muscle contraction is thus not necessarily proportional to muscle bulk, but rather reflects the number, size, and fiber composition of the motor units that are contracting simultaneously. The number of fibers per motor unit is variable and dependent on the size and function of the muscle. Muscles that perform fine control and small-coordinated movements, such as the lumbricals, contain only a few fibers per motor unit. This is unlike muscles that perform gross movements and activity, such as the gastrocnemius, which have many fibers per motor unit.

**Muscle Activation**

The central nervous system asserts precise control over how many and what
types of motor units are activated during bulk muscle contraction. Not every muscle fiber is activated at once. Histologically, fast-conducting motor neurons innervate motor units with a greater percentage of type II fibers while slower, smaller motor neurons innervate motor units with more type I fibers. Henneman’s size principle states that motor units containing the fewest myocytes and most type I fibers are recruited (or activated via action potential) first for any muscle contraction. As demands and the need for higher force production arise, larger motor units with increasing amounts of type II fibers are recruited. The benefit of recruiting the smaller type I dense motor units first is that a contraction can be sustained for the longest amount of time by relying on oxidative phosphorylation energy production rather than anaerobic processes. Further recruitment of motor units with stronger contractility also enables fine control over the amount of force generated. In addition to gradually selecting motor units with larger sizes to generate force, motor unit action potentials also become more frequent, and contraction frequency increases.

If motor unit recruitment is maximized, the muscle is said to display its maximum voluntary contraction (MVC). Although a muscle may be able to activate all motor units of the muscle at once (a point that is debated) to achieve a MVC, the maximal force produced by the muscle is intimately related to the muscle’s cross-sectional area. The absolute muscle strength per cross-sectional area varies somewhat, depending on the source consulted, but historical values average around 15 N/cm².

## Muscle Fatigue

Muscle fatigue can be defined generally as any reduction in the maximal force-generating capacity. It is an active impairment of motor performance manifested by either an increased sense of effort to maintain desired force output, or an overall involuntary decline in force output. Several factors other than muscle fiber type composition lead to muscle fatigue.

Inadequate central drive involves a decrease in motor unit recruitment for MVC following a fatiguing task. Muscle fiber membrane excitability decreases with continuous stimulation, thus creating a failure of action potential propagation. Contractile force may decline without a change in EMG activity, an occurrence termed excitation–contraction coupling failure. With sustained muscle contraction, metabolic byproducts can accumulate locally, resulting in intramuscular acidosis. This affects the myofibril cross-bridges, leading to a decline in the number of cross-bridges that can be formed as well as the force
exerted by each cross-bridge. At exercise levels below maximum aerobic intensity, but for long durations (> 1 hour), this process will lead to glycogen depletion within the muscle.

Muscle fatigue is reversible, but recovery time is variable depending on the cause of that fatigue. Short-duration, intermittent, submaximal contractions may result in fatigue related to excitation–contraction coupling failure, with a slow recovery of up to 30–60 minutes. In contrast, fatigue from the accumulation of metabolites appears to occur rapidly during maximal or near-maximal contractions but has a recovery time of less than 2 minutes.

Finally, the individual’s current fitness level and psychological motivation level also affect the rate of development of fatigue. Training can reduce the fatigability of a muscle, but the response seems to be task specific. Of note, since muscle endurance training appears to be task specific, clinical endurance tests using measures such as an isokinetic device may have little carryover to functional activities.


**PRINCIPLES OF AEROBIC CONDITIONING**

\( \text{Vo}_2 \) is defined as the total amount of oxygen consumption from arterial blood per unit time. It can be represented by the Fick equation, in which the difference between arterial and venous oxygen content is multiplied by cardiac output:

\[ \text{Vo}_2 = \text{CO}(\text{Ao}_2 - \text{Vo}_2). \]

As aerobic intensity increases, more arterial oxygen is used for oxidative phosphorylation, and \( \text{VO}_2 \) increases. This continues until a maximum is reached, in which there is no further increase in oxygen uptake despite increases in exercise intensity—the \( \text{Vo}_{2\text{max}} \). This value represents the *maximal* amount of oxygen consumption from the blood per unit time and is thought to be the most
accurate measure of an individual’s aerobic capacity for a specific activity. VO$_{2\text{max}}$ is affected by many variables, including type of exercise, effort of the individual, training status of the individual, predominant muscle fiber type used in exercise, and adaptations to aerobic exercise, such as capillary vascularization within muscle.

**Metabolic Equivalent**

A *metabolic equivalent (MET)* is a unit that is used to estimate, generally, the metabolic cost of a physical activity. It is depicted as a multiple of the resting metabolic rate, and thus is a multiple of the VO$_2$ found at rest. One MET equates to approximately 3.5 mL of O$_2$/kg/min, or 250 mL/min in males and 200 mL/min in females of average sizes. For greater accuracy in individuals of varying mass, the MET may also be expressed as VO$_2$ divided by body mass: mL/(min × kg). METs are often used in place of VO$_2$ to easily gauge the relative intensity of a given physical activity.

Both aerobic and anaerobic conditioning rely on the overload principle, which states that increasing *frequency, duration, or intensity* when performing an activity (mode) will stimulate physiologic adaptations furthering an individual’s capacity to perform the same activity. A progressive training program should be structured to increase at least one of these components over time to produce exercise gains.

The specificity principle refers to the specific adaptations that are made to imposed demands on the system (SAID principle), which occurs in both neurologic and biomechanical overload. Strength and endurance gains made during overload training with a specific exercise modality are realized in activities most similar to that modality. For example, optimal training for an activity should include the same muscle groups, joint ranges of motion and positioning, aerobic intensity, and duration as are used in the activity. This has been referred to as functional training, in which motions and actions in training replicate those that will be performed during the sport or competition.

All aerobic physiologic gains made through training are reversible and can be quickly lost as a result of detraining if the individual ceases to perform the specified activity. Losses in VO$_{2\text{max}}$ and muscular capillary density have been reported in trained individuals confined to bed rest for several weeks, and heart size decreases to pretraining levels when endurance training ceases.


AEROBIC TRAINING

**Physiologic Responses**

**A. Initial Responses**

Starting within 24 hours of aerobic training, intravascular albumin synthesis is upregulated and plasma volume increases via osmotic fluid transfer. With continued aerobic training over several more weeks, plasma volume increases by 12–20%. Oxygen transport and thermoregulation through sweating undergo optimization, with increased total hemoglobin content and fluid available for sweating. Plasma volume expansion creates several key anatomic changes. The heart receives greater preload due to increased venous return, increasing stroke volume. Through the Frank–Starling law, myocardial contractility increases under higher left ventricular end-diastolic volume, and the stroke volume at all heart rates is improved. Gradually, the myocardium undergoes eccentric hypertrophy, with the increased left ventricular end-diastolic volume. Whereas aerobic training typically increases heart volume with eccentric hypertrophy, resistance training typically causes concentric hypertrophy, producing a greater left ventricular mass.

**B. Vascular and Pulmonary Responses**

With aerobic conditioning, oxygen consumption by the body becomes increasingly more efficient, and VO$_2$, and VO$_{2}\text{max}$ are significantly increased
during training. Compared with untrained individuals, aerobically trained individuals have a lower heart rate for a given VO$_2$. This is in part due to the increase in cardiac output from improved stroke volume (since CO = HR × SV). As an aerobically trained heart beats faster toward maximal heart rate, stroke volume increases significantly more than in untrained individuals, who primarily rely on heart rate increases for improved cardiac output. The higher VO$_2$ at both submaximal and maximal heart rates has also been attributed to more effective blood flow redistribution and improved muscular ATP generation at lower arterial oxygen saturation levels (PO$_2$).

Several peripheral vascular changes occur with aerobic training that optimize blood flow redistribution and oxygen uptake. At maximal and submaximal exercise, vascular resistance is diminished as a result of nitric oxide–mediated vasodilation. In addition, some studies have shown that muscle fiber type may switch during training to improve oxidative capacity. At maximal exercise, blood shunting away from inactive areas is enhanced. Arteriogenesis and angiogenesis within peripheral muscles improve plasma flow to exercising muscle tissue, increasing oxygen extraction. Splanchnic and renal blood flow also may decrease with aerobic training to provide additional shunting.

Aerobic training decreases the ventilatory equivalent for oxygen (V$_E$/VO$_2$), which represents the minute-ventilation to oxygen consumption ratio. Restated, there is need for less ventilation to accomplish similar amounts of oxygen consumption. Greater amounts of oxygen are extracted from the inspired air; thus, ventilatory frequency and volume decrease. Because less inspiratory work is required, the pulmonary musculature requires less oxygen and does not fatigue as quickly. More oxygen is therefore available to the muscles directly used in exercise. Furthermore, owing to improved respiratory muscle endurance, longer periods of submaximal ventilation are tolerated with generation of greater inspiratory forces.

C. Hormonal Response

Aerobic exercise training has been shown to have numerous hormonal effects in the body. Growth hormone rises more slowly during exercise. Resting growth hormone levels are greater when aerobic training takes place at intensities above the lactate threshold than at the lactate threshold, or even when compared with untrained individuals. Insulin and glucagon sensitivity also increase, and lower plasma concentrations of both are found during exercise than prior to aerobic training. In males, reduced resting prolactin and testosterone levels have been
reported after long-term endurance training. In women undergoing long-term endurance training, follicle-stimulating hormone and luteinizing hormone levels may also show significant variability from normal values. These abnormalities may be responsible for anovulation and amenorrhea, which produce the so-called “female athlete triad” described in women who participate in rigorous sports training. Catecholamine secretion is decreased at rest after continued aerobic training. While exercising, catecholamine levels increase, but the extent of the increase depends on the intensity of the exercise and, possibly, the duration as well. Some studies show that in trained individuals there is a greater capacity to secrete epinephrine and norepinephrine than in untrained individuals.

There is significant evidence that aerobic exercise diminishes the risk of developing type 2 diabetes in adolescents and adults. The greatest benefits are realized in those with preexisting risk factors, including obesity, family history of diabetes, and sedentary lifestyle. In type 2 diabetics, aerobic training has been associated with a reduction in the amount of medications needed for glycemic control because insulin sensitivity is improved.

Over time, aerobic training increases muscular concentrations of citric acid cycle enzymes involved in oxidative phosphorylation, producing higher mitochondrial content, as well as higher glycogen stores. These changes potentiate oxidative ability of myocytes by creating more ATP and will gradually lead to preferential hypertrophy of the slow-twitch type I and IIa fibers compared with fast-twitch IIx and IIb fibers.

Compared with carbohydrates, metabolism of fats is significantly enhanced after aerobic training. The rates of both fat catabolism into triacylglycerols and muscular uptake of triacylglycerols for fatty acid β-oxidation into glucose are accelerated. Systemic glucose availability is also augmented with increased liver gluconeogenic capacity.

Management of Aerobic Training

A. Exercise Intensity

When prescribing an aerobic training program, increases in intensity, frequency, and duration are included to produce gains through the overload principle, as previously mentioned. Of the three training variables, intensity affects physiologic adaptations the most. Aerobic intensity can be monitored using various techniques. Measuring energy expenditure per unit time, or power output, is useful to describe an individual’s aerobic capability but does not
reflect the stress placed on the aerobic system. Given the same amount of energy expenditure, the physiologic stress placed on individuals differs owing to variances of training level as well as genetics. It is more relevant to measure intensity relative to the individual’s maximal aerobic stress level. Such techniques include a subjective rating of exertion such as the Borg scale (see below). More objective methods include percentages of maximal heart rate, METs, lactate threshold, and \( \text{VO}_{2\text{max}} \).

It is possible to gauge exercise intensity by calculating desired percentages of \( \text{VO}_{2\text{max}} \) or maximal heart rate as goals during a workout. This would, however, be challenging, as measuring these data can be technically difficult as well as taxing to the participant. Estimations are more easily made of both values when first planning an exercise program. Traditionally, maximal heart rate (\( \text{HR}_{\text{max}} \)) has been quickly calculated using the following equation based on population studies by Haskell and Fox:

\[
\text{HR}_{\text{max}} = 220 - \text{Age}.
\]

Recently published studies have proposed that a more accurate estimation is represented by a revised equation:

\[
\text{HR}_{\text{max}} = 206.9 - (0.67 \times \text{Age}).
\]

Percentages of calculated, or measured, \( \text{HR}_{\text{max}} \) may also be used as guidelines for intensity goals.

Another technique is the Karvonen method, which uses a percentage of the heart rate reserve to determine a target rate:

\[
\text{HR}_{\text{target}} = \text{HR}_{\text{rest}} + x(\text{HR}_{\text{max}} - \text{HR}_{\text{rest}}),
\]

where \( x \) represents the desired percentage of heart rate reserve, which is usually set at 40–90%, depending on the participants current physical ability.

Alternatively, the individual’s subjective rating of exercise intensity provides an adjunct to the preceding techniques, or it may be used exclusively. The Borg Rating of Relative Perceived Exertion (RPE) scale ranges from 6, which is “very, very light,” to 19, which is “very, very hard.” Ratings between 13 and 17 have been shown to approximate 61–92% of \( \text{HR}_{\text{max}} \) and 51–85% of \( \text{VO}_{2\text{max}} \). The so-called “talk test,” which is the intensity above which carrying on a conversation is uncomfortable, is another often-used subjective method. Aerobic exercise that takes place at this intensity level has been shown to fulfill current exercise guidelines. (See Table 9–1.)
Table 9–1 Classification of physical activity intensity, based on physical activity lasting up to 60 minutes.

<table>
<thead>
<tr>
<th>Intensity</th>
<th>Relative Intensity</th>
<th>Absolute Intensity (METs) in Health Adults (Age in Years)</th>
<th>Strength-Type Exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>V̇O₂peak (%)</td>
<td>Heart Rate Reserve (%)</td>
<td>Maximal Heart Rate (%)</td>
</tr>
<tr>
<td>Very Light</td>
<td>&lt; 25</td>
<td>&lt; 30</td>
<td>&lt; 9</td>
</tr>
<tr>
<td>Light</td>
<td>25–44</td>
<td>30–49</td>
<td>9–10</td>
</tr>
<tr>
<td>Moderate</td>
<td>45–59</td>
<td>50–69</td>
<td>11–12</td>
</tr>
<tr>
<td>Hard</td>
<td>60–84</td>
<td>70–89</td>
<td>13–16</td>
</tr>
<tr>
<td>Very Hard</td>
<td>≥ 85</td>
<td>≥ 90</td>
<td>&gt; 16</td>
</tr>
<tr>
<td>Maximal*</td>
<td>100</td>
<td>100</td>
<td>20</td>
</tr>
</tbody>
</table>

Notes:
- MET: metabolic equivalent; RPE: rating of perceived exertion; V̇O₂peak: maximal oxygen uptake.
- *Based on 8–12 repetitions for persons under age 50 y and 10–15 repetitions for persons aged 50 y and older.
- ‡Maximal values are approximate mean values achieved during maximal exercise by healthy adults. Absolute intensity (METs) values are approximate mean values for men. Mean values for women are approximately 1–2 METs lower than those for men.

Currently there is controversy over the intensity ranges necessary to produce endurance and metabolic benefits. The latest guidelines from the American College of Sports Medicine (ACSM) propose that most adults should aim for moderate to vigorous intensities. Those who are untrained, elderly, or who are beginning a new exercise program are advised to exercise at the lower targeted intensity range (eg, light to moderate) and to slowly progress to higher intensities to continue producing a training response. HRmax values approximately 13 beats/min lower have been reported in participants in aerobic exercise programs that primarily involve upper extremities (eg, swimming) compared with those that primarily involve lower extremities (eg, running). This
difference should be recognized if heart rate is used to gauge intensity in such activities.

B. Lactate Threshold

Although heart rate and VO_{2max} calculations modulate intensity based on cardiopulmonary performance, blood lactate measurements provide information that can guide training intensity based on the oxidative capacity of exercising muscles and the associated vasculature. Multiple studies have shown that exercise at or slightly above the lactate threshold may produce improved responses in endurance athletes. The lactate threshold, also known as the anaerobic threshold, is defined as the point at which oxidative phosphorylation begins to lag behind anaerobic (lactate-generating) forms of energy production. At intensities below this threshold, blood lactate levels are constant. However, at intensities above the lactate threshold, a net excess of lactate is formed and a rise is detected. The lactate threshold can be measured directly by taking blood samples, or calculated using the respiratory quotient from gas exchange during exercise. It is thought that consistently exercising at levels slightly above the lactate threshold overloads the aerobic system, upregulating oxidative enzyme synthesis.

C. Duration of Exercise

The duration of aerobic exercise necessary to produce training benefit is also greatly debated. Previously sedentary individuals may require only 5-minute-long training sessions whereas active individuals would require longer intervals for benefits to be realized. In some large prospective cohort trials, exercise duration necessary to use 1000 kcal per week has been reported to be the minimum requirement for cardio–metabolic benefit. In general, the ACSM has recommended sustaining a targeted heart-rate range for 20–60 minutes. Workout durations of athletes training for specific events should correlate with the duration of the goal event, owing to the specificity of the training response. Although a dose-dependent response between energy expenditure and health benefit has been proposed, significant durations of exercise also increases the risk of suffering injury as a result of overtraining.

Overtraining syndrome is manifested most often by deterioration in physical performance, mood alterations, and high cortisol-to-cortisone ratio. This state occurs in response to an excess of training load, psychological stress, and competitive endeavors when combined with inadequate recovery quality and
duration. There are two forms of overtraining, sympathetic and parasympathetic. The sympathetic form involves increased sympathetic activity at rest, including hyperexcitability and restlessness, which impairs rest quality and consequently, exercise performance. In the more common parasympathetic form, there is insufficient sympathetic response when exercising. Feelings of fatigue and malaise during both exercise and rest periods predominate. Poor exercise performance, sleep, and eating patterns; increased infection susceptibility; and mood disturbances (anger, depression) are commonly seen.

There is no reliable method to predict overtraining, and an athlete must be closely monitored for symptoms of an overtrained state. The frequency, duration, and intensity of exercise at which overtraining occurs differ among individuals. Furthermore, the amount of rest time necessary to recuperate from overtraining syndrome is also specific to the individual.


Robergs RA, Landwehr R: The surprising history of the $HR_{\text{max}} = 220 - \text{Age}$ equation. JEPOnline 2002;5:1–10.


STRENGTH TRAINING

Physiologic Responses

A. Initial Responses

Studies have shown that the earliest training-induced improvements in maximal muscle force occur without any overall increase in muscle bulk, or hypertrophy. In general, untrained individuals cannot likely recruit all motor units voluntarily.

B. Neural Synchronization

After resistance training, an individual is able to develop the ability to more efficiently recruit more motor units for functional activity. By means of this principle, called neural synchronization, more motor units are recruited at the same time. A few factors that change with resistance training, including increased firing frequency of motor units, may contribute to this response. The typical normal maximal firing frequency of motor units is 100–200 Hz; with resistance training, this can increase by up to 40%. The increased firing rate can decrease the time to maximum muscle contraction (reaction time). Similarly, maximal electromyographic (EMG) motor unit amplitude can increase with resistance training (up to 50%, according to some studies). However, a plateau in both the firing frequency and EMG amplitude increases occurs with training. Thus, with continued resistance training, no further gains are appreciated in these neural adaptations.

C. Muscle Hypertrophy

As resistance training continues beyond the initial stages of neural adaptations, muscular structural changes occur. Muscle hypertrophy begins to become the dominant factor in increased muscle strength. Muscle hypertrophy refers to enlargement of the muscle cell; specifically, the contractile elements of the muscle enlarge. The extracellular matrix also expands in order to support the growth. Sarcomeres are added in series or in parallel within the muscle cell.

Three primary factors are responsible for initiating the hypertrophic response to resistance exercise: mechanical tension (mechanical stress and load), muscle damage, and metabolic stress. Mechanical tension produced by both force generation and stretch is essential to muscle growth. The combination of the two seems to have an additive effect in this process. It is thought that the tension associated with resistance training disturbs the integrity of the skeletal muscle, causing molecular and cellular responses in myofibers and satellite cells. The
hypertrophic response to passive tension may favor fast-twitch fibers much more than slow-twitch. Exercise and resistance training often result in local damage to the muscle fibers, which is thought to aid in the hypertrophic response. With nonuniform lengthening of muscle tissue, this can cause a shearing of myofibrils and disruption of muscle membranes. Calcium homeostasis may be disturbed by these membrane disruptions and opening of stretch-activated channels. The response to this myotrauma is similar to the inflammatory response. Neutrophils travel to the area of microtrauma and release cytokines that attract macrophages and lymphocytes. The macrophages remove cellular debris and also release cytokines that activate myoblasts, lymphocytes, and more macrophages. This is thought to lead to the release of various growth factors that regulate satellite cell proliferation and differentiation. Metabolite accumulation may be just as important as high force development in the hypertrophic response to training. Metabolites such as lactate, hydrogen ions, phosphate, creatine, and others build up due to anaerobic glycolysis for ATP production. This alters the hormonal milieu as well as leading to cellular swelling, free-radical production, and increased activity of growth-oriented transcription factors.

Another factor that contributes to muscle hypertrophy is cellular swelling, through increases in protein synthesis and decreases in proteolysis. Hypoxia has been shown to contribute to increases in muscle hypertrophy, especially when combined with exercise. Hypoxia increases lactate accumulation and reduces acute lactate clearance, which can lead to increased cellular swelling. Rises in lactate can also cause elevations in anabolic hormones and cytokines. Reactive hyperemia is also a response to local hypoxia and can allow for the delivery of anabolic hormones and growth factors to satellite cells, aiding in cellular repair and hypertrophy.

**D. Muscle Hyperplasia**

Muscle hyperplasia (an increase in the total number of myocytes) has been postulated to play a role in increased muscle bulk. Satellite cells found in the muscular basement membrane have the potential to proliferate and differentiate into myoblasts and myocytes, as well as provide additional myonuclei to the parent myocyte. Normally, these cells support existing muscle fibers during normal growth, hypertrophy, and in the regeneration process following injury or disease. Research showing satellite cell–induced muscular hyperplasia in humans is, however, lacking. Most data suggest that muscle is a postmitotic tissue and does not undergo significant cell replacement throughout its life span. As an individual ages, particularly beyond 60 years, sarcopenia is common due
to an overall loss of myocytes.

**E. Hormonal Response**

Hormones and cytokines play important roles in the muscle hypertrophic response, serving as regulators of the anabolic process. They are involved in satellite cell proliferation and differentiation and can facilitate satellite cell repair of damaged muscle fibers. Many hormones contribute to the hypertrophic response, including hepatocyte growth factor, interleukin-5 (IL-5), interleukin-6 (IL-6), fibroblast growth factor, leukemia inhibitory factor, insulin, insulin-like growth factor-1 (IGF-1), testosterone, and growth hormone (GH). The most important and widely studied of these are IGF-1, GH, and testosterone.

IGF-1 is thought to provide the main anabolic response during mechanical loading. Receptors for this hormone are found in satellite cells, myofibers, and Schwann cells. Three isoforms of the hormone have been identified, namely, IGF-1Ea, IGF-1Eb, and IGF-1Ec. All three are expressed in muscle tissue. IGF-1Ec, commonly referred to as mechano growth factor (MGF), is activated by mechanical stimulation and is sensitive to muscle damage. IGF-1 helps to induce muscle hypertrophy by increasing the rate of protein synthesis in muscle cells. MGF activates satellite cells and aids in the proliferation and differentiation of these cells, and IGF-1Ea enhances the fusion of satellite cells with muscle fibers and facilitates the transfer of myonuclei to the muscle cell. IGF-1 also increases the intracellular calcium concentration in the cell, leading to activation of other calcium-dependent anabolic pathways.

Testosterone is a steroid that functions in muscle and affects neurotransmitter release, nerve regeneration, and nerve cell body size. Nearly all (98%) of the steroid is protein bound in serum; only 2% is unbound and hence biologically active. The actions of testosterone are magnified with mechanical load, resulting in increased protein synthesis rates and decreased protein breakdown. Testosterone also stimulates the release of GH and promotes satellite cell replication and activation. Resistance training has been shown to upregulate androgen receptor concentration as well as increase testosterone secretion. Acute responses, however, are more limited in women and the elderly, which is why the overall hypertrophic potential in these populations are more limited, despite resistance training.

GH is a polypeptide hormone secreted by the anterior pituitary gland and released in a pulsatile fashion. The greatest nonexercise release occurs during sleep cycles. GH plays a role in the regulation of immune function, bone modeling, and extracellular fluid volume. Training-induced increases in GH
have been correlated with the magnitude of type I and type II muscle fiber hypertrophy. GH is also thought to be involved in the exercise-induced increase in expression of IGF-1 and upregulation of the MGF isoform.

F. Changes to Bone and Collagen

After strength training, tendons and ligaments display increased thickness, weight, and strength. Muscle hypertrophy is also accompanied by an increase in the collagen content of the surrounding connective tissue matrix as well as the muscle connective tissue sheaths. Bone also adapts to resistance training, but much more slowly than muscle tissue; it can take up to 6–12 months for such adaptation to be noted. Bone density increases in a force- and strain-dependent fashion. High power, heavy resistance (load), and higher volumes produce the most changes in bone density. Resistance training has also been shown to increase the thickness of hyaline cartilage on the articular surfaces of the joints, which can facilitate the shock-absorbing function in the joint. The VA\textsubscript{O}\textsubscript{2max} is generally not significantly affected by resistance training, except with the use of a “circuit training” regimen that employs high volumes and short rest periods between exercises.

Management of Strength Training

In order to properly maximize exercise-induced muscle hypertrophy, alteration of training variables is essential, which goes along with the principle of specificity of training. These training variables include intensity, volume, exercise selection, rest intervals, repetition speed, periodization, and exercising to muscle failure.

A. Intensity of Resistance Exercise

Intensity of exercise, or amount of load, is probably the most important exercise variable for stimulating muscle growth. It is often represented as a percentage of a one-repetition maximum (% 1 RM) and usually refers to a number of repetitions of a given weight. A moderate number of repetitions (usually between 6 and 12) seems to be optimal, whereas high repetitions (> 15) are comparatively inferior in producing muscle hypertrophy. Additionally, loads less than 65% of 1 RM are not sufficient to produce substantial hypertrophy. Moderate repetitions rely more heavily on glycolysis, which results in a buildup of metabolites that have a significant impact on anabolic processes. Low repetitions rely
predominantly on the phosphocreatine system, and thus may not produce a large buildup of metabolites. Testosterone and GH release is stimulated more strongly through moderate repetition sets compared with low repetitions, as well. Some investigators have proposed that muscles with a higher percentage of slow-twitch fibers may respond better to higher repetition programs, but research on this theory is fairly sparse.

B. Volume of Resistance Exercise
The volume of resistance exercise generally refers to the combination of total repetitions, sets, and load performed during an exercise session. Higher volume and multiple sets are superior to single-set routines with respect to muscle hypertrophy. Higher volume programs generate more glycolytic activity and thus buildup of metabolites that stimulate the anabolic response. Higher testosterone levels, as well as acute release of GH, are noted with these programs. To optimize the hypertrophic response, the exercise program should incorporate a progressive increase in volume over a periodized cycle, with a brief period of overreaching. Caution should be used, however, as prolonged overreaching can lead to an overtrained state that has catabolic effects on muscle, resulting in chronically decreased concentrations of testosterone and luteinizing hormone, and increased cortisol levels.

C. Single versus Multijoint Exercises
It is well known that varying exercise parameters, including direction, angle of lift or pull, and position of joints with exercise, causes different activation patterns within a muscle. Muscles have different attachment sites, and even fibers within a muscle have different angles and attachments compared with others, so varying position or angle may be necessary to fully exercise certain muscles. Almost every muscle is composed of fibers that are aligned in different directions or different planes, often having a conical shape. Thus, certain motor units or sections of a muscle may be activated with a particular plane of movement or direction, whereas another portion of the muscle may function better within a different plane. A good example is the biceps brachii, where the motor units in the lateral aspect of the muscle are recruited primarily for elbow flexion, the motor units in the medial aspect primarily for supination, and centrally located motor units for nonspecific combinations of flexion and supination.

To achieve the greatest benefit, exercises should consist of stimuli in multiple planes of motion or using multiple angles. A trainer may advocate for
using more free movement or free weight protocols to achieve this goal, as opposed to machine-based weight-training regimens. Many weight machines have fixed angles of movement or fixed directions that may not allow the adaptations of movements to achieve a multiplane approach. In contrast, cable-type machines or free weights may allow for the variability of movement to appropriately train all aspects of the muscle, thus preventing imbalances or other mechanical dysfunctions.

Multijoint exercises recruit large amounts of muscle mass. This has an impact on the anabolic hormonal response. Postexercise hormonal elevations are related to the muscle mass involved in the exercise program. Increases in testosterone and GH levels are greater in response to multijoint exercises than single-joint ones. Multijoint exercises also require significant stabilization of other areas of the body, thus activating many other muscles that otherwise would likely not be stimulated with single-joint movements. These supporting muscles undergo isometric contraction to stabilize movements and allow other muscles to produce power and motion. For example, it is estimated that over 200 muscles are activated during a squat. Consequently, multijoint exercises with free weights or free-movement programs will yield an overall higher volume of exercise in a session because of the need for activation of large numbers of muscles for stabilization of supporting joints and body regions.

On the other hand, single-joint exercises allow the trainer to focus on individual muscles in contrast to multijoint movements. Additionally, caution must be used when prescribing multijoint movement exercises to avoid overuse or overtraining of certain muscles. In multijoint movements, some muscles are favored; overemphasis on these “prime movers” could lead to muscle imbalances. The use of single-joint exercises can selectively target underdeveloped muscles that can help correcting muscle imbalances and mechanical dysfunction.

The use of unstable surfaces has been shown to require extensive activation of other supporting musculature, including the core muscles, to help provide stability to a body region or joint. In a similar fashion to multijoint movement exercises, this increases the overall work of the body and larger muscle activity during an exercise routine, thus yielding a higher volume of exercise within the set or overall session. This could be favorable for an overall fitness-type exercise routine but for a hypertrophy-oriented program can be counterproductive. With increasing instability comes greater activation of surrounding and core musculature, causing a significant decrease in force output to the prime muscle movers. This in turn will have a negative effect on muscle hypertrophy of that
“prime muscle” but may lead to more hypertrophy of the other supporting muscles, especially the abdominals.

**D. Periodization**

Periodization, also known as periodized training, incorporates a systematic variation in exercise intensity, repetitions, sets, or frequency over a specified period of time. This approach is often used to prevent overtraining as well as to keep the exercise regimen interesting to the individual. Periodization can also be used in a training calendar to prepare for competition, with the goal of building up to peak performance at the time of competition. Figure 9–3, included earlier, represents a simple linear periodization strategy, based on Matveyev’s original model, which allows an athlete to build up to a competition stage at which peak performance is seen.

The amount of rest allowed during resistance training sets has different effects on strength capacity and metabolite buildup, which in turn affects anabolic and hypertrophic responses. Rest periods shorter than 30 seconds between sets increase metabolite buildup but impair both strength recovery and muscle performance in subsequent sets. Long rest periods (> 3 minutes) permit full strength recovery, allowing the individual to train with maximum force capacity. Although the mechanical tension is maximized with increased rest intervals, metabolic stress is minimized, blunting the anabolic drive. After resistance training, the majority of strength is recovered within the first minute after a set. Consequently, moderate amounts of rest, lasting between 60 and 90 seconds after a set or exercise is completed, appear to be optimal. This approach enhances the anabolic environment better than longer rest intervals and is characterized by greater induced hypoxia, increased metabolite buildup, and an increased spike in anabolic hormone concentration. An adaptive response to reduced rest intervals has been suggested, which may support a need for periodization.

**E. Muscle Failure**

Muscular failure is defined as the point at which a muscle can no longer produce any further force to concentrically lift a given load. Training to failure is necessary for maximal hypertrophic response and results in increased motor unit recruitment, as well as increased metabolic stress and hormonal response. The number of repetitions performed before failure affects the results. Studies have shown that performing 1–6 repetitions maximum (RM) is best for increasing maximal dynamic strength, whereas 8–12 RM is most effective for muscular
hypertrophic response. Performing 12–15 RM (or more) has little effect on maximal strength or hypertrophy but increases muscular endurance. However, performing the same number of repetitions before failure in an unvarying regimen is associated with an increased risk of overtraining; thus, programs should be varied to avoid overtraining, and for optimal muscular fitness.

**F. Specificity of Exercise**

Many resistance training programs can achieve muscle hypertrophy, although the “principle of specificity” (ie, SAID principle, described earlier) dictates that the most effective program is similar to an individual’s desired activities in the type and nature of muscular activation. Historically, two progressive resistance exercise programs have been used, the DeLorme program and the Oxford technique.

Dr. Thomas DeLorme developed one of the earliest recognized strength training programs while a captain in the U.S. Army. His method involved determining an individual’s 10 repetition maximum (10 RM); that is, the maximum amount of weight that individual could lift ten times before failure. Once this point was identified, the individual would exercise 3–5 days a week, performing 10 repetitions of fractions of the 10 RM weight (10%, then 20%, then 30%, etc), up to a new determined 10 RM, at which point the process began again. The program underwent several modifications, with subsequent training intervals set at 10 repetitions of 50% of 10 RM, then 75% of 10 RM, and finally 100% of 10 RM. Some critics commented that achieving 100% of 10 RM was difficult owing to the number of repetitions and weight required to reach this level. The Oxford technique was developed to address these concerns. This technique starts at 100% of 10 RM, progressing to 75% of 10 RM and then 50% of 10 RM. Some studies have shown that the Oxford technique is less effective at increasing the 10 RM level than DeLorme’s method, but no definitive study has been done.

Evidence suggests that a better transfer of training may occur if lower weights (ie, 50% or 75% of 10 RM) are used but the number of repetitions increases to the point of muscle fatigue. In other words, an individual who lifts at a lower weight might be able to achieve similar results to one lifting maximal weight fewer times so long as he or she increased the number of repetitions to the point of failure (ie, the point at which the weight cannot be lifted one more time). However, the number of repetitions needed to reach failure may be excessive, and true muscle failure may be unobtainable if a low enough weight is used. Furthermore, other factors play a role in contributing to “failure”—not
limited to other local joint or tissue strain or stress, local tissue metabolic effects, and so on—all related to the larger mechanical work performed with low weights and high repetitions. Thus, there is a much better efficiency, and probably even efficacy, in using higher weights with lower repetitions. These principles underlie the development of the popular “pyramid” programs that are used frequently in weight-training gyms.


Toigo M, Boutellier U: New fundamental resistance exercise determinants of
FLEXIBILITY & STRETCHING

Flexibility and stretching exercises are important components of therapeutic exercise programs, helping individuals to maintain a range of motion (or improve range of motion if restricted) that allows desired activities to be performed. Reduction in the range of motion of a joint can affect functional activities, including ambulation, self-care, work, and sports tasks. Even small reductions in a joint’s range of motion can have a biomechanical impact on other tissues, structures, and surrounding joints, requiring compensatory mechanics or motions to accomplish desired tasks. This can lead to secondary biomechanical problems or tissue injuries and pain. Janda noted that muscle tightness can inhibit muscle antagonists by means of proprioceptive factors, in what he described as a crossed syndrome. Maintenance or restoration of a functional range of motion is therefore necessary for maximum strength production. Stretching is a common technique used to improve range of motion.

The benefits of stretching include injury prevention and improved performance with tasks, including sports. Some physiotherapists also believe that stretching can reduce postexercise soreness. It should be noted that there is no optimal degree of flexibility, and ideal joint ranges of motion may differ for each individual, activity, or sport. In fact, athletic performance can be enhanced by a certain amount of “tightness.” This seemingly incongruity may reflect the use of stored muscular elastic energy and a reduced need for muscle-stabilizing activity. It is thought that some athletes perform better because of inherent tightness.

Practitioners need to be familiar with the normal ranges of motion for various joints and limbs before progressing with a stretching program (Table 9–2). Specific measurements of cervical and lumbar ranges of motion are challenging, with the range of motion essentially derived from many segments or even regions of motion. For example, cervical motion also incorporates thoracic motion, and lumbar motion incorporates thoracic, sacral, and even pelvic motion. Published ranges of motion for the cervical and lumbar spine vary, but generally accepted averages, such as those used in disability assessments, are provided in Table 9–3.
Table 9–2 Average normal ranges of motion for joints of the upper and lower extremities.
<table>
<thead>
<tr>
<th>Joint</th>
<th>Degree of Motion</th>
<th>Joint</th>
<th>Degree of Motion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder</td>
<td></td>
<td>Ankle</td>
<td></td>
</tr>
<tr>
<td>Horizontal flexion</td>
<td>141</td>
<td>Plantar flexion</td>
<td>56</td>
</tr>
<tr>
<td>Horizontal extension</td>
<td>45</td>
<td>Dorsiflexion</td>
<td>13</td>
</tr>
<tr>
<td>Neutral abduction</td>
<td>14</td>
<td>Forefoot</td>
<td></td>
</tr>
<tr>
<td>Forward flexion</td>
<td>167</td>
<td>Inversion</td>
<td>37</td>
</tr>
<tr>
<td>Backward extension</td>
<td>62</td>
<td>Eversion</td>
<td>21</td>
</tr>
<tr>
<td>Internal rotation</td>
<td>69</td>
<td>Thumb</td>
<td></td>
</tr>
<tr>
<td>Elbow</td>
<td></td>
<td>Abduction</td>
<td>58</td>
</tr>
<tr>
<td>Flexion</td>
<td>143</td>
<td>IP flexion</td>
<td>81</td>
</tr>
<tr>
<td>Extension</td>
<td>1</td>
<td>MCP flexion</td>
<td>53</td>
</tr>
<tr>
<td>Forearm</td>
<td></td>
<td>CMC flexion</td>
<td>15</td>
</tr>
<tr>
<td>Pronation</td>
<td>76</td>
<td>IP extension</td>
<td>17</td>
</tr>
<tr>
<td>Supination</td>
<td>82</td>
<td>MCP extension</td>
<td>8</td>
</tr>
<tr>
<td>Wrist</td>
<td></td>
<td>CMC flexion</td>
<td>20</td>
</tr>
<tr>
<td>Flexion</td>
<td>76</td>
<td>Fingers</td>
<td></td>
</tr>
<tr>
<td>Extension</td>
<td>75</td>
<td>DIP flexion</td>
<td>80</td>
</tr>
<tr>
<td>Radial deviation</td>
<td>22</td>
<td>PIP flexion</td>
<td>100</td>
</tr>
<tr>
<td>Ulnar deviation</td>
<td>36</td>
<td>MCP flexion</td>
<td>90</td>
</tr>
<tr>
<td>Hip</td>
<td></td>
<td>DIP extension</td>
<td>0</td>
</tr>
<tr>
<td>Flexion</td>
<td>122</td>
<td>PIP extension</td>
<td>0</td>
</tr>
<tr>
<td>Extension</td>
<td>10</td>
<td>MCP extension</td>
<td>45</td>
</tr>
<tr>
<td>Abduction</td>
<td>46</td>
<td>Great Toe</td>
<td></td>
</tr>
<tr>
<td>Adduction</td>
<td>27</td>
<td>IP flexion</td>
<td>60</td>
</tr>
<tr>
<td>Internal rotation</td>
<td>47</td>
<td>IP extension</td>
<td>0</td>
</tr>
<tr>
<td>External rotation</td>
<td>47</td>
<td>MTP flexion</td>
<td>37</td>
</tr>
<tr>
<td>Knee</td>
<td></td>
<td>MTP extension</td>
<td>63</td>
</tr>
<tr>
<td>Flexion</td>
<td>143</td>
<td>Toes</td>
<td></td>
</tr>
<tr>
<td>Hyperextension</td>
<td>10</td>
<td>DIP flexion</td>
<td>55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>PIP flexion</td>
<td>38</td>
</tr>
<tr>
<td></td>
<td></td>
<td>MTP flexion</td>
<td>35</td>
</tr>
<tr>
<td></td>
<td></td>
<td>MTP extension</td>
<td>40</td>
</tr>
</tbody>
</table>

IP; interphalangeal; CMC, metacarpal; DIP, distal interphalangeal; PIP, proximal interphalangeal; MCP, metacarpophalangeal; MTP, metatarsophalangeal.

Table 9–3 Average ranges of motion for joints of the cervical and lumbar spine.

<table>
<thead>
<tr>
<th>Joint</th>
<th>Degree of Motion</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cervical Spine</strong></td>
<td></td>
</tr>
<tr>
<td>Flexion</td>
<td>45</td>
</tr>
<tr>
<td>Extension</td>
<td>45</td>
</tr>
<tr>
<td>Lateral flexion</td>
<td>45</td>
</tr>
<tr>
<td>Rotation</td>
<td>80</td>
</tr>
<tr>
<td><strong>Lumbar Spine</strong></td>
<td></td>
</tr>
<tr>
<td>Flexion</td>
<td>90</td>
</tr>
<tr>
<td>Extension</td>
<td>30</td>
</tr>
<tr>
<td>Lateral flexion</td>
<td>30</td>
</tr>
<tr>
<td>Rotation</td>
<td>30</td>
</tr>
</tbody>
</table>

Methods of Stretching

The methods for increasing muscle length or range of motion can include manual stretching or mechanical stretching. Mechanical stretching is a sustained (or possibly intermittent) external, end-range stretch force, applied with overpressure and by manual contact using a physical device. Example include dynamic braces (eg, Dynasplint) and serial casting. Manual stretching is performed by the individual without using a device. Various types of manual stretching include ballistic, static, dynamic, and proprioceptive neuromuscular facilitation (PNF).

A. Ballistic Stretching

Ballistic stretching incorporates a rapid, forceful, intermittent stretch characterized by the use of quick bouncing movements that create momentum to stretch the tissues. This type of stretching can lead to muscle microtrauma and collagen deposition, producing muscular tightness. More energy is absorbed into muscle and tendon during ballistic stretching, raising the risk for muscle–tendon injury, including avulsion. Thus, this type of stretching is not favored.

B. Static Stretching

In static stretching, a muscle is stretched slowly by holding the muscle at its position of greatest tolerated length. Tension should be felt within the muscle, but pain should be avoided. The optimal duration is debatable, but it is thought
that 15–30 seconds is required to create increased muscular length. However, prolonged stretching for 30 seconds or longer may be detrimental, resulting in decreased strength for up to 1 hour afterward. This is thought to be due to local tissue and muscle ischemia caused by increased pressure within the muscle. Some researchers contend that this may happen with as little as 5 seconds of prolonged maximum stretch.

C. Dynamic Stretching

Dynamic stretching involves active movements of a muscle group that cause a stretch without holding the end position. This type of stretching may have more advantages than static stretching with respect to muscular performance. Dynamic stretching utilizes more proprioceptive input, brings increased blood flow to muscles and tissues, and increases power, flexibility, and range of motion. Its effectiveness is specific to the exercise performed. Therefore, to maximize muscle performance for a desired activity, the stretching motion should be similar to the movement performed during that activity.

As mentioned earlier, muscles are three-dimensional. Just as a resistance routine should exercise a muscle group in different planes, stretching can also involve movements in multiple planes in order to lengthen as many fibers within the muscle as possible. Some investigators have termed this dynamic on–off stretching technique three-dimensional stretching. The stretch is usually held for several seconds, relaxed, and then repeated in the sagittal, frontal, and transverse planes.

D. Proprioceptive Neuromuscular Facilitation

PNF stretching involves autogenic or reciprocal inhibition of a targeted muscle.

Autogenic inhibition relies on the firing of the Golgi-tendon organ (GTO) to cause reflexive muscle relaxation as the muscle is stretched to its end point, activating the GTO. PNF techniques include hold–relax and contract–relax. Using the hold–relax technique, a muscle is lengthened to its end point and then isometrically contracted with a submaximal effort for at least 3 seconds. The muscle is then relaxed and a new muscle end point is determined (ie, “taking up the slack”). This sequence can be repeated a few times for further lengthening. The contract–relax technique is similar, however, the contraction is concentric against resistance (ie, moving the joint through normal range of motion); then, after relaxing, the new end point is determined. The contract–relax technique is thought to be inferior to hold–relax, as activation of the GTO is lost during the concentric movement.
Reciprocal inhibition involves voluntary contraction of the antagonist muscle, leading to reduced activation of the target muscle by means of type Ia-inhibitory interneuron activation. An example would be a hold–relax with opposing muscle contraction technique. This is similar to the hold–relax technique described earlier but is followed by an opposing muscle contraction while the increased range of motion is taken up. The individual can also perform an opposing muscle contraction, once end range is achieved, without first performing the isometric contraction of the target muscle.

Management of Flexibility & Stretching

According to the ACSM, stretching is most effective after the temperature of a muscle is increased through light endurance or strength-training exercise, or both, or following use of heating modalities. A precise temperature has not been elucidated. To promote maximal range of motion gains, stretching should occur daily; however, performing the exercises two to three days per week can also prove effective. Although dynamic, static, or PNF stretching techniques may be used, increasing evidence shows that multiplanar dynamic and PNF stretching is most effective. Stretches should be held for 10–30 seconds and repeated two to four times, with a goal of performing 60 seconds of stretching per muscle group, per session.


Along with new therapeutic interventions that have emerged in the growing field of physical medicine and rehabilitation, pharmacologic agents continue to play an important role in management of the patient’s rehabilitation course. This chapter reviews an array of medications that are often used for conditions frequently managed in physiatric practice. Detailed discussion of each of these conditions—spasticity, traumatic brain injury, pain, and venous thromboembolism (deep vein thrombosis)—is found elsewhere in this book, and readers are referred to those chapters for additional information.

SPASTICITY

In spasticity, a motor neuron disorder (most often involving the upper motor neuron) leads to an abnormal increase in muscle tone secondary to hyperexcitability of the stretch reflex. (See Chapter 6 for additional information.) Current pharmacologic treatment utilizes oral antispasmodic medications and invasive chemodenervation agents. The choice of agents involves consideration of benefits versus complications of spasticity and expectations of the antispasmodic, such as functional benefit or pain relief.

Oral Antispasticity Agents

A variety of oral antispasmodic agents is available; however, only four medications are approved by the U.S. Food and Drug Administration (FDA) for spasticity management: baclofen, diazepam, tizanidine, and dantrolene. These oral agents have a systemic effect, reducing generalized muscle tone; however, all are associated with significant systemic side effects. Because all four
medications involve a degree of hepatic metabolism, caution is advised in patients with liver disease. Table 10–1 contrasts key features of these medications.

### Table 10–1 Antispasticity agents.

<table>
<thead>
<tr>
<th>Medication</th>
<th>Mechanism of Action</th>
<th>Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>Acts on GABA&lt;sub&gt;B&lt;/sub&gt; receptors</td>
<td>Sedation, weakness, lower seizure threshold</td>
</tr>
<tr>
<td>Diazepam</td>
<td>Acts on GABA&lt;sub&gt;A&lt;/sub&gt; receptors</td>
<td>Sedation, memory impairment, weakness, decreased rapid eye movement sleep</td>
</tr>
<tr>
<td>Tizanidine</td>
<td>Centrally acting α&lt;sub&gt;2&lt;/sub&gt;-adrenergic agonist</td>
<td>Sedation, hypotension, dry mouth, bradycardia, dizziness, weakness</td>
</tr>
<tr>
<td>Dantrolene</td>
<td>Inhibits calcium release at sarcoplasmic reticulum</td>
<td>Sedation, weakness, diarrhea, hepatotoxicity, nausea</td>
</tr>
</tbody>
</table>

#### A. Baclofen

Baclofen is a centrally acting antispasmodic agent that binds to the presynaptic and postsynaptic γ-aminobutyric acid B (GABA<sub>B</sub>) receptors as a GABA agonist. Activation of the GABA<sub>B</sub> receptors suppresses excitatory neurotransmitters and reduces γ motor neuron excitability and muscle spindle sensitivity. Side effects include sedation and weakness; therefore, baclofen is started at a low dose and titrated to a tolerable dose. Other potential side effects include lowering of the seizure threshold. The greatest risk with this medication is sudden withdrawal, which can lead to seizures, hallucinations, rebound spasticity, and fever. In addition, patients with renal disease may require a dosage adjustment since baclofen is renally excreted. Overdose of baclofen can be treated with physostigmine.

#### B. Diazepam

Diazepam is a centrally acting antispasmodic agent that binds to the GABA<sub>A</sub> receptors and causes membrane hyperpolarization by opening membrane chloride channels. The most common side effect is sedation. Other side effects
include memory impairment, addiction with withdrawal, cognitive impairment, decreased rapid eye movement (REM) sleep, and muscle weakness. Because of the cognitive side effects, diazepam is usually not administered to patients with traumatic brain injury. Overdose of diazepam can be treated with flumazenil.

C. Tizanidine

Tizanidine is a centrally acting antispasmodic agent that binds to the $\alpha_2$-adrenergic receptors, acting as an agonist. Activation of the $\alpha_2$-adrenergic receptors leads to an increase in presynaptic motor neuron inhibition, suppressing excitatory neurotransmitters and reducing spinal reflexes. Tizanidine has a rapid onset and a very short half-life, which requires frequent dosing. The major side effect is sedation. Other side effects include muscle weakness, hypotension, liver toxicity, dry mouth, bradycardia, and dizziness. Similarly, clonidine can be used as an antispasmodic agent, in addition to its use in hypertension. Clonidine is unique in that it can also be administered as a transdermal patch. Clonidine has greater antihypertensive properties when compared with tizanidine.

D. Dantrolene

Dantrolene is a peripherally acting antispasmodic agent that inhibits the release of calcium from the sarcoplasmic reticulum in striated muscles, rather than affecting neurotransmitters. By interfering with calcium release, dantrolene reduces muscle contraction (particularly the extrafusal muscle fiber and fast-twitch fibers) and muscle spindle sensitivity. It has some minor effects on smooth and cardiac muscle. Sedation is a lesser effect with dantrolene compared with the other oral antispasmodic agents due to its peripheral action. In addition, dantrolene has the potential to cause liver toxicity; however, the literature reflects a low 1.8% occurrence.


Injectable Antispasticity Agents

A. Phenol
Phenol is an alcohol-based chemical neurolytic agent that denatures neural protein and induces axonal destruction. When injected to the nerve, phenol demyelimates the γ fibers and destroys the axons. A 5% concentration is generally used for spasticity management; however, reports have demonstrated effects with concentrations between 2% and 7%. In addition, lower concentrations of phenol produce a transient anesthetic effect. Advantages of phenol are its inexpensive cost and immediate onset of effects. These effects may last up to 6 months. Side effects include dysesthesia, muscle pain, weakness, and systemic reactions, including convulsions and cardiovascular compromise.

**B. Botulinum Toxin**

Botulinum toxin is a neurotoxin produced by the bacterium *Clostridium botulinum*. The neurotoxin, which is subdivided into seven serotypes (A–G), inhibits the release of acetylcholine (ACh) at the neuromuscular junction. The mechanism of action of the toxin involves proteolytic cleavage of the SNARE complex (soluble NSF attachment protein receptor) at the presynaptic axon terminal. The SNARE complex consists of mainly synaptobrevin, SNAP-25, and syntaxin. The toxin cleaves the vesicles containing ACh, thereby preventing the release of ACh to the synaptic cleft. Toxin serotypes A, C, and E proteolytically cleave SNAP-25, whereas serotypes B, D, F, and G cleave synaptobrevin, and serotype C also cleaves syntaxin. This ultimately results in denervation and improvement of spasticity.

Currently, toxin serotypes A and B are being administered for spasticity management. Serotype A formulations include onabotulinumtoxinA (Botox), abobotulinumtoxinA (Dysport), and incobotulinumtoxinA (Xeomin); serotype B is rimabotulinumtoxinB (Myobloc). Dosing of serotypes A and B is not equivalent without a direct formula conversion. Dosage is dependent on the type of botulinum toxin and the patient’s weight, muscle size, level of spasticity, and treatment goal. The response is generally dose-dependent; thus, the more toxin is injected, the greater is the resulting muscle weakness.

The effects of botulinum toxin usually occur within 24–72 hours after injection. The medication peak effect occurs at approximately 4–6 weeks and has a duration of 2–6 months. This 2- to 6-month duration is the timeframe for collateral sprouting of the axon. The side effects and complications of botulinum toxin are weakness, pain at the injection site, infection, flulike syndrome, dysphagia with cervical injections, nerve injury, respiratory failure, and antibody formation.
For additional discussion of the various agents used in the treatment of spasticity, and nonpharmacologic treatment measures, refer to Chapter 6.


**TRAUMATIC BRAIN INJURY**

Traumatic brain injury occurs in approximately 1.7 million people every year with an estimated cost of 60 billion dollars. Individuals who survive the brain injury sustain physical, cognitive, and neurobehavioral deficits. Current pharmacologic agents play an important role in modulating the traumatic brain-injured individual’s neurochemistry in order to improve cognition, attention, and arousal. Chapter 13 explores a range of management issues relating to rehabilitation of patients with traumatic brain injury. The focus here is on pharmacologic agents that aid in postinjury recovery and rehabilitation.

Treatment of patients with traumatic brain injury may include medications administered with the goal of improving the patient’s consciousness and arousal state. Five such agents are described below and contrasted in Table 10–2. There is currently insufficient evidence to support the use of a single pharmacologic agent or combination of agents for this purpose. However, recent evidence has suggested the important role of amantadine for patients emerging from low-response states.

**Table 10–2** Pharmacologic agents for consciousness and arousal.
Neurostimulants such as amphetamine and methylphenidate have demonstrated some benefits with arousal state. These agents increase dopamine and norepinephrine availability, thus stimulating the central nervous system. The dosing of methylphenidate is started low and twice a day. Side effects include tachycardia, palpations, fever, bruising, hypertension, and restlessness; therefore,
heart rate and blood pressure monitoring is vital in these patients after administration. The risk and frequency of seizures with the use of methylphenidate has not been shown to be increased in this patient population.

► **Amantadine**

Dopaminergic agents such as amantadine have produced improvement in consciousness and arousal in patients with traumatic brain injury. Amantadine is indicated for the treatment of parkinsonism and influenza. Because it increases presynaptic and postsynaptic dopamine availability, along with being a weak NMDA receptor antagonist, amantadine has also been used to promote the functional recovery of patients with prolonged disorders of consciousness after traumatic brain injury. The common side effects of amantadine are orthostatic hypotension, insomnia, dizziness, dry mouth, constipation, and headache.

► **Bromocriptine**

Bromocriptine has been used in the treatment of disorders involving reduced states of arousal and consciousness. Bromocriptine acts predominantly at the postsynaptic D2 dopamine receptor along with 5-HT1 and 5-HT2 serotonin receptors. Studies have demonstrated that bromocriptine can help improve processing speed, strategic control, and hemispatial neglect. The side effects include orthostatic hypotension, vomiting, bowel changes, headache, nasal congestion, involuntary movements, and liver enzyme elevation.

► **Carbidopa-Levodopa**

Carbidopa-levodopa is a combination medication (levodopa plus carbidopa) that allows dopamine to cross the blood–brain barrier by preventing peripheral decarboxylation of dopamine. The combined agent influences arousal and consciousness by acting on the dopamine receptors of the frontal lobe, basal ganglia, and white matter tracts. The side effects include orthostatic hypotension, involuntary movements, vomiting, loss of appetite, bowel changes, headache, nasal congestion, and liver enzyme elevation.

► **Modafinil**
Modafinil is clinically indicated for narcolepsy, obstructive sleep apnea, and fatigue related to multiple sclerosis, and may also be used in the treatment of patients with problems of arousal and consciousness. Although the exact mechanism of action is unknown, studies have shown effects on catecholamine, dopamine, serotonin, histamine, adenosine, and monamine oxidase B systems in the hypothalamus, hippocampus, and amygdala. In patients with traumatic brain injury, modafinil has demonstrated improvement in arousal by decreasing daytime sleepiness. The most common side effects are headaches, nausea, vomiting, nervousness, anxiety, and insomnia. The effect on seizure threshold has not been fully established. A relative contraindication to modafinil is recent myocardial infarction or angina. The role of modafinil in emergence from impaired consciousness remains to be investigated.


PAIN

Pain is an unpleasant, subjective experience that is often a significant issue for patients who require physical and rehabilitative care. Pain can be classified as acute or chronic. Acute pain is associated with tissue damage, increased autonomic nervous activity, and resolution with healing of injury. Chronic pain extends beyond the expected period of healing, has no protective function,
degrades health and function, and contributes to depressed mood. Pain can be further separated into nociceptive pain (which arises from a stimulus outside the nervous system and is proportionate to receptor stimulation) or neuropathic pain (which arises from a primary lesion or dysfunction in the nervous system, requires no nociceptive stimulation, and is disproportionate to receptor stimulation), or a mix of nociceptive and neuropathic pain.

Pain can be managed pharmacologically at different levels of the pain pathway (Figure 10–1) using multiple pharmacologic agents. Several of these agents are described here. For additional discussion of pain management, see also Chapter 21.
Nonopioid Analgesics

Acetaminophen is a commonly used pain medication that acts peripherally on pain and does not inhibit nociception or change the perception of pain. Its analgesic mechanism of action is unclear but involves weak inhibition of cyclooxygenase (COX), possibly selective to COX-2. Due to its action on COX, acetaminophen has weak antiinflammatory activity. Acetaminophen is rapidly absorbed in the gastrointestinal tract. The onset of analgesia is approximately 11 minutes, peaking in 30–60 minutes, with a half-life of 1–4 hours and duration of 1–4 hours. Studies of its benefits for pain relief have reported mixed results compared with aspirin or ibuprofen. Although, the safety profile of acetaminophen is superior to that of nonsteroidal antiinflammatory drugs (NSAIDs), in that gastrointestinal and coagulation effects with acetaminophen are minimized, high doses can lead to liver, kidney, and brain damage. The common side effects include nausea, rash, and headache. Chronic use may lead to anemia, thrombocytopenia, and hematologic malignancies.

Nonsteroidal Antiinflammatory Drugs

NSAIDs are frequently used in the treatment of various pain manifestations and are classified as mild analgesics. The pain-reducing mechanism derives from their effect on antiinflammatory and analgesic pathways. The antiinflammatory effects occur through inhibition of the enzyme COX, resulting in reduction of arachidonic acid to prostaglandin H₂. Depending on the NSAID, inhibition may be nonselective (COX-1 and -2) or selective for COX-2. The antiinflammatory action leads to an analgesic effect by interfering with prostaglandin sensitization of nociceptors. Patients are generally more tolerant to NSAIDs than to opioids, experiencing fewer side effects, lack of psychological or physical dependence, and no development of tolerance. Side effects of NSAIDs include gastrointestinal ulceration and bleeding, renal insufficiency, hepatic inflammation, and hypertension. Because of the gastrointestinal side effects, certain NSAIDs are enteric-coated. Taking NSAIDs with meals or
coadministration of an H₂ blocker or proton pump inhibitor has also been shown to be effective in reducing gastrointestinal side effects. Selective COX-2 inhibitor NSAIDs have fewer gastrointestinal side effects; however, the selective COX-2 inhibitors increase the risk of cardiovascular toxicity.

NSAIDs are most often administered orally. Currently, one NSAID, diclofenac, is FDA approved for topical use. Diclofenac can be prescribed as a pill, gel, jelly, solution, or a patch. Topical administration aims to restrict NSAID action to the pain location and to decrease blood concentrations, resulting in fewer systemic side effects. Topical diclofenac has an increased risk for hepatotoxicity; therefore, it is important to monitor liver enzymes before and after administration. Table 10–3 contrasts pharmacologic properties of commonly prescribed NSAIDs.

<table>
<thead>
<tr>
<th>Medication</th>
<th>Half-Life (h)</th>
<th>Peak Effect (h)</th>
<th>Duration (h)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aspirin</td>
<td>4–16</td>
<td>1–2</td>
<td>4–6</td>
<td>Irreversible effect on platelet aggregation</td>
</tr>
<tr>
<td>Diclofenac</td>
<td>6–8</td>
<td>2–4</td>
<td>10–12</td>
<td>Decreased gastrointestinal (GI) toxicity</td>
</tr>
<tr>
<td>Indomethacin</td>
<td>4–5</td>
<td>1–2</td>
<td>4</td>
<td>Greater GI toxicity and central nervous system side effects</td>
</tr>
<tr>
<td>Ibuprofen</td>
<td>2–4</td>
<td>1–2</td>
<td>4–6</td>
<td>Rapid onset</td>
</tr>
<tr>
<td>Naproxen</td>
<td>12–17</td>
<td>2–3</td>
<td>8–12</td>
<td>Longer duration</td>
</tr>
<tr>
<td>Sulindac</td>
<td>14–16</td>
<td>3–4</td>
<td>8–12</td>
<td>Decreased renal toxicity</td>
</tr>
<tr>
<td>Meloxicam</td>
<td>15–20</td>
<td>5–12</td>
<td>8–12</td>
<td>Lesser GI toxicity</td>
</tr>
<tr>
<td>Celecoxib</td>
<td>11–13</td>
<td>2–9</td>
<td>8–12</td>
<td>Selective cyclooxygenase-2 (COX-2) inhibitor Increased cardiovascular toxicity</td>
</tr>
<tr>
<td>Ketorolac</td>
<td>3.5–9</td>
<td>2–3</td>
<td>4–6</td>
<td>Short-term use</td>
</tr>
</tbody>
</table>

Table 10–3 Nonsteroidal antiinflammatory drugs.
Opioid Analgesics

Opioid analgesics are narcotics used to treat moderate to severe pain. Opiates can be divided into three classes: those that are naturally occurring (e.g., morphine), those that are semisynthetic (e.g., oxycodone, hydrocodone), and those that are synthetic (e.g., methadone, fentanyl). The mechanism of action involves binding of opioid receptors (μ, κ, δ, OFQ/N) on neurons, peripheral nerves, and joints. Binding of these receptors leads to pain relief by decreasing pain perception, decreasing pain reaction, and increasing pain tolerance. Opioid receptor binding also results in the wide range of side effects, which include respiratory depression, decreased gastrointestinal motility, nausea, vomiting, pruritus, orthostatic hypotension, and mood alteration. Most opiates are metabolized in the liver and eliminated in the kidney. Continued administration of opiates can lead to the development of tolerance and physical dependence on the opiate. Severe adverse effects of opiates are reversed by naloxone, an opiate antagonist. Table 10–4 contrasts features of several commonly prescribed opioid analgesics.

Table 10–4 Opioid analgesics.
<table>
<thead>
<tr>
<th>Medication</th>
<th>Route and Dosage (mg)</th>
<th>Half-Life (h)</th>
<th>Duration (h)</th>
<th>Peak Effect (h)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine</td>
<td>PO: 20–30 IM: 10</td>
<td>2–3</td>
<td>2–4</td>
<td>0.5–1.5</td>
<td>Multiple routes: PO, IM, IV, rectal, intrathecal</td>
</tr>
<tr>
<td>Morphine CR</td>
<td>PO: 20–30 IM: 10</td>
<td>2–3</td>
<td>8–12</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Oxycodone</td>
<td>PO: 20</td>
<td>2–3</td>
<td>3–4</td>
<td>0.5–1</td>
<td>Fast-acting</td>
</tr>
<tr>
<td>Oxycodone CR</td>
<td>PO: 20</td>
<td>2–3</td>
<td>8–12</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Hydromorphone</td>
<td>PO: 7.5 IM: 1.5</td>
<td>2–3</td>
<td>2–4</td>
<td>0.5–1.5</td>
<td>Fast-acting</td>
</tr>
<tr>
<td>Methadone</td>
<td>IM: 10</td>
<td>12–190</td>
<td>4–12</td>
<td>1–2</td>
<td>Avoid in patients with significant respiratory, hepatic, or renal failure</td>
</tr>
<tr>
<td>Hydrocodone</td>
<td>PO: 30</td>
<td>2–4</td>
<td>3–6</td>
<td>1–2</td>
<td>Combined with acetaminophen or NSAID</td>
</tr>
<tr>
<td>Codeine</td>
<td>PO: 120 IM: 200</td>
<td>2–3</td>
<td>3–6</td>
<td>1–2</td>
<td>Weak, short-acting</td>
</tr>
<tr>
<td>Fentanyl transdermal</td>
<td>25 mcg</td>
<td>16–24</td>
<td>58–72</td>
<td>12</td>
<td>Avoid heat application directly on patch</td>
</tr>
<tr>
<td>Oxymorphone</td>
<td>PR: 10 IM: 1</td>
<td>2–3</td>
<td>2–4</td>
<td>0.5–1.5</td>
<td>Rectal suppository</td>
</tr>
</tbody>
</table>

NSAID, nonsteroidal antiinflammatory drug.

Tramadol is a synthetic dual-mechanism analgesic agent used in the treatment of moderate to severe pain. The dual mechanism of action produces analgesia by enabling the drug to bind to opioid receptors with codeine-like affinity as well as serotonin and norepinephrine receptors; however, the drug is not classified as a controlled substance. Tramadol has been effective in treating nociceptive and neuropathic pain. The onset of action is within 1 hour and peaks within 1.5–2 hours. Tramadol has a half-life of 6 hours and duration of 4–6
hours. Side effects include nausea, dizziness, sedation, pruritus, dry mouth, and sweating. There is a risk of physical dependence, but this is not as severe as with opiates. Caution is advised in patients concurrently taking medications affecting seizure threshold, tricyclic antidepressants, carbamazepine, and monoamine oxidase inhibitors.

Tapentadol is another synthetic dual-mechanism analgesic agent for moderate to severe pain. The dual mechanism of action produces analgesia by acting as an agonist to opioid receptors as well as a norepinephrine reuptake inhibitor. The potency of tapentadol is between that of tramadol and morphine. Its onset of action is within 30 minutes and peaks within 1–1.5 hours. Tapentadol has a half-life of 4 hours and duration of 3–4 hours. The drug has fewer gastrointestinal side effects and produces less tolerance compared with opiates. However, patients can develop nausea, dizziness, and sedation. Patients who concurrently take antiseizure medications and antidepressants have an increased risk of hallucinations and potential risk of serotonin syndrome; thus, cautious use is advised.

**Gabapentinoids**

Gabapentin is a structural analog to GABA that was initially developed for treatment of epilepsy. The drug is now widely used to treat neuropathic pain owing to its combination of antihyperalgesic and antiallodynic, but not antinociceptive, properties. The mechanism of action is unknown and seems to involve a complex synergy of GABA synthesis, calcium channel binding, and non-NMDA receptor antagonism. The onset of drug action is within 1–3 hours, peaking within 2–4 hours. The half-life is 5–7 hours and duration is 8–12 hours. Although the drug begins to act within hours, studies have demonstrated that onset of pain relief generally begins at 1–3 weeks. Side effects include loss of coordination, lightheadedness, sedation, thirst, dizziness, peripheral edema, and weight gain. Renal function is an important factor in dosing as gabapentin is renally eliminated.

Pregabalin has similar effects to gabapentin but it also has anticonvulsant, antihyperalgesic, and anxiolytic properties. It is FDA approved for treatment of neuropathic pain resulting from diabetes, spinal cord injury, and fibromyalgia. Pregabalin is structurally similar to the inhibitory neurotransmitter (GABA) and binds to voltage-gated calcium channels, leading to a reduction of calcium influx and excitatory neurotransmitters; however, it does not directly bind to GABA_A, GABA_B, or benzodiazepine receptors. Pregabalin, like gabapentin, reduces the
hyperexcitability of dorsal horn neurons caused by tissue damage. The onset of action is within 30 minutes to 3 hours and peaks within 1.5 hours. The half-life is 6.3 hours. In marked contrast to gabapentin, onset of pain relief occurs generally within 24–48 hours. Side effects include loss of coordination, lightheadedness, sedation, thirst, dizziness, peripheral edema, and thrombocytopenia. Pregabalin can also potentiate the effects of benzodiazepines due to the increase in GABA. Because its mechanism of action differs from that of gabapentin, both medications can be combined, which can have the benefit of lowering the dosage required and number of side effects. Moreover, pregabalin has been shown to be synergistic with opioids by having opioid-sparing effects and decreasing opioid consumption.


VENOUS THROMBOEMBOLISM

One of the objectives during a patient’s inpatient rehabilitation course is to prevent the occurrence of venous thromboembolism (VTE). The mainstay of VTE prevention is prophylaxis using pneumatic compression devices and pharmacologic agents. The latter are discussed below. For additional treatment measures used in patients with diagnosed VTE or suspected deep vein thrombosis, refer to Chapter 5. Four drugs are commonly prescribed for VTE prevention: unfractionated heparin, low-molecular-weight heparin, warfarin, and rivaroxaban (Table 10–5).

Table 10–5 Agents for venous thromboembolic prophylaxis.
**Unfractionated Heparin**

The use of standard unfractionated heparin for VTE prophylaxis has been well documented since the early 20th century. Heparin acts at multiple sites of the blood coagulation cascade by binding to antithrombin III and then catalyzing the inactivation of factors IIa, Xa, IXa, and XIIa. Heparin has a dual function as an anticoagulant and an antithrombotic. In VTE prophylaxis, the drug is administered subcutaneously. The side effects include bleeding, bruising, hyperkalemia, and heparin-induced thrombocytopenia (for which studies have demonstrated a high hemorrhagic rate of 8–15%). Effective reversal of heparin involves the administration of protamine sulfate.

**Low-Molecular-Weight Heparin**

Low-molecular-weight heparin (LMWH) is derived from standard heparin by enzymatic and chemical methods. Average molecular weight is 4.5 kDa compared with 15 kDa for heparin. The enzymatic and chemical changes result in a higher bioavailability, longer half-life, and less nonspecific binding of plasma proteins. The side effects are generally similar to those for heparin, and

<table>
<thead>
<tr>
<th>Medication</th>
<th>Mechanism of Action</th>
<th>Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unfractionated heparin</td>
<td>Binds to antithrombin III</td>
<td>Hemorrhage, bruising, HITS</td>
</tr>
<tr>
<td>Low-molecular-weight heparin</td>
<td>Binds to antithrombin III</td>
<td>Hemorrhage, bruising, HITS</td>
</tr>
<tr>
<td>(LMWH; enoxaparin, fondaparinux)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rivaroxaban</td>
<td>Inhibits factor Xa</td>
<td>Hemorrhage, bruising</td>
</tr>
<tr>
<td>Warfarin</td>
<td>Inhibits vitamin K</td>
<td>Hemorrhage, bruising, skin</td>
</tr>
<tr>
<td></td>
<td>coagulation factors</td>
<td>necrosis</td>
</tr>
</tbody>
</table>

HITS, heparin-induced thrombocytopenia.
include bleeding, bruising, hyperkalemia, and heparin-induced thrombocytopenia. However, there is less risk of microvascular bleeding compared with heparin. Effective reversal of LMWH involves the administration of recombinant activated factor VII; protamine sulfate is only partially effective in reversal. Common LMWH agents administered for VTE prophylaxis are enoxaparin and fondaparinux. Enoxaparin differs from fondaparinux in having a rapid onset and a long half-life; however, fondaparinux causes more frequent episodes of bleeding than enoxaparin.

### Rivaroxaban

More recently, an oral pharmacologic factor Xa inhibitor, rivaroxaban, has been approved for VTE prophylaxis. Its mechanism of action involves inhibition of both free and bound factor Xa, interrupting the intrinsic and extrinsic pathways of the blood coagulation cascade. Studies have demonstrated that rivaroxaban is as effective and safe as subcutaneous LMWH in preventing VTE in patients after total hip or knee arthroplasty. Currently, there is no effective reversal treatment for complications of rivaroxaban.

### Warfarin

Warfarin is classified as a coumarin. Its mechanism of action involves inhibition of liver synthesis of vitamin K–dependant coagulation factors (II, VII, IX, and X). The drug is absorbed rapidly in the gastrointestinal tract, where it is bound to plasma proteins. Generally, 36–72 hours is required to attain a stable loading dose; however, warfarin dosing is dependent on vitamin K intake, protein levels, liver metabolism, drug interaction, gastrointestinal absorption, genetics, and compliance from the patient. Compared with other VTE prophylactic agents, warfarin has a long half-life (20–60 hours). The international normalized ratio (INR) is used to monitor drug levels in the blood and effectiveness of therapy. Side effects of warfarin include bleeding, bruising, and possible reduction in bone mineral density. An uncommon complication is skin necrosis due to small vessel thrombosis from the drug’s inhibition of proteins C and S. The reversal of warfarin’s effects involves the administration of vitamin K and prothrombin complex concentrate or fresh frozen plasma.

Beyer-Westendorf J, Lutzner J, Donath L, et al: Efficacy and safety of


Gait Analysis

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Philip Noto, DO
Albert Esquenazi, MD

Normal human locomotion has been the focus of intense clinical observational analysis for several decades. Through these efforts, the basic components of the gait cycle, its phases, and subphases have been identified, defined observationally, and linked to their kinematic, kinetic, and muscle behaviors. More in-depth analysis has revealed combinations of subphases that define the operational features of gait—organized components of the gait cycle reflecting functional features that achieve important operating objectives of the locomotion system. These objectives include advancement of the body’s center of mass by means of swing phase and stance phase propulsion mechanisms, foot–floor clearance mechanisms of the swinging limb, and antigravity stability mechanisms that operate during loading of body weight and the subsequent period of single limb support. Common gait deviations have also been analyzed in terms of how they relate to the larger context of the operational features of gait, providing insight into rehabilitative strategies that can best address operational features gone awry.

KEY CONTRIBUTORS TO HUMAN LOCOMOTION

This chapter describes human locomotion as a sequence of repetitive bodily motions organized functionally as a gait cycle. The chapter provides working descriptions of the gait cycle, its phases and sub phases, its kinematics, kinetics, and muscle kinesiology. Major objectives of the gait cycle are delineated including advancing the body’s center of mass, controlling upright stability against gravity and clearing the foot from the floor in order to avoid stumbling.
and falling. The chapter goes on to develop the concept of operational features of gait, namely, clinically observable components of the gait cycle that work together to achieve major gait objectives such as translation of the center of body mass and maintaining upright stability. Based on concepts developed in the paper, the chapter ends with case scenarios of patients with gait dysfunction that will hopefully alert the reader to useful ways of analyzing and treating problems of human locomotion. In analyzing human gait, the examiner must be knowledgeable about three contributors that have key roles in locomotion: ground reaction force, joint moments, and center of mass.

## Ground Reaction Force

A force is a push or pull on an object that results from its interaction with another object. Forces result from interactions. Some forces result from contact interactions; normal, friction, and tension forces are examples of contact forces. Other forces are the result of interactions of the action-at-a-distance type (eg, gravitation and magnetic forces). According to Newton, whenever objects A and B interact with each other, they exert forces upon each other.

When a person stands on the floor, the body exerts a downward force on the floor (at minimum, the force of body weight). In a reciprocal manner, the floor exerts an upward force on the person’s body. Two forces result from this interaction: a force on the floor, and a force on the person’s body. These two forces are called action and reaction forces. Newton’s Third Law of Motion formally describes the relationship between these two forces: for every action, there is an equal and opposite reaction. To illustrate the concept of action–reaction forces, imagine preparing to get off a rowboat on a lake. What happens when we step off the rowboat onto the dock? As we move in the direction of the dock, the boat tends to move (accelerate) in the opposite direction. Acceleration is produced when a force acts on a mass.

What happens when we take a step on the ground? Does the ground move in the opposite direction (like the boat)? Physical analysis of this question involves Newton’s Second Law ($F = ma$), which describes the relationship between force, mass, and acceleration. The greater the mass of an object being accelerated, the greater the amount of force needed to accelerate that object. When we step off the rowboat onto the dock, the rowboat moves because its mass is relatively small. When we step on the ground, however, the mass of the earth is so large that the force exerted by the body against it can only minutely accelerate the earth. On the other hand, the body’s mass is small so that the reaction force of
the earth, termed the *ground reaction force*, can easily accelerate the body. By Newton’s Third Law of Motion, the body’s pushing force on the earth is the same as the earth’s push back force on the body. Schematically, equation 1 below reflects the reaction force exerted by the earth on the person’s small body mass, resulting in a large acceleration. Equation 2 reflects the action force exerted by the person on the earth, resulting in a very small acceleration of the very large mass of the earth.

\[
\begin{align*}
(1) \quad F &= m \times a \\
(2) \quad F &= m \times a
\end{align*}
\]

In summary, a person’s body has mass and, when the body stands on the ground, the body mass exerts a force on that ground. When the body exerts a pushing force on the ground, the ground exerts a push back force of equal magnitude on the body. This push back force is called the *ground reaction force* (GRF). When entities of different masses exert a force on each other, the entity with the smaller mass will have the larger acceleration. The ground reaction force is one of the keys to human locomotion because the smaller mass of the human body is easily accelerated by the ground reaction force during the gait cycle.

---

**Joint Moments**

The fundamental objectives of human locomotion are to move the body’s mass from one place to another and provide antigravity stability so that a person doesn’t fall down. Locomotion mechanisms in the bipedal human are built around two facts: each lower limb is configured with many movable joints and, during walking, movements within and across each limb must be coordinated because both limbs are connected to the superincumbent mass by a pelvis. Translating body mass by means of bipedal locomotion depends on a repetitive sequence of joint motions that simultaneously move the body’s center of mass along an intended line of progression while counteracting the effects of gravity on upright posture and joint motion.

Movable joints in the walking human generate changing joint rotations that require control by muscular, soft tissue, and body weight forces. To understand how joint motion is controlled, we introduce the concept of *joint moment*, or force exerted by means of leverage. In general, a moment $M$ is defined by a force exerted through a lever arm $r$, namely, $M = rF$ (where force $F$ is applied at
a distance $r$ from the axis of rotation). Figure 11–1 illustrates a person standing in equilibrium with a flexed knee. If unopposed by the quadriceps, body weight force ($B$) falling behind the knee would promote knee flexion collapse because of the rotary effect of joint moment $Ba$. The force generated by quadriceps contraction ($Q$) produces a counter joint moment, $Qb$. When $Ba$ equals $Qb$, the system is in equilibrium and no motion occurs, all joint moments being perfectly counterbalanced. (Conversely, net unbalanced moments do result in joint motion, a finding characteristic of normal locomotion.)

\[ \text{Figure 11–1 Forces generated when standing with a flexed knee.} \]

\[ \text{Center of Mass} \]

The center of mass (COM) of any object is a point location where all of that object’s mass is imagined as concentrated. If we think of all bodily segments connected by joints as separate “objects” having their own CoM, we now assert that the CoM of the whole human body is that point in space where all joint
moments created by the configuration of jointed body segments (ie, limb, trunk, and head postures) are in equilibrium. (Joint moments include all forces derived from muscle contraction, soft tissue resistance, and superimposed body weight acting on the lever arms of their respective axes of rotation). As Figure 11–2 illustrates for a standing person, the location of the CoM is not a fixed point because it varies with body segment configuration. Similarly, as the body propels forward during ambulation, the CoM moves vertically and laterally; it does not behave as a fixed point. Viewed in the sagittal plane, the vertical displacement of the CoM moves as a smooth sinusoidal curve (Figure 11–3). This curve’s amplitude in a normal individual is approximately 5 cm (2 in.). The center of gravity also oscillates side to side during ambulation, generating another sinusoidal curve whose amplitude is approximately 6 cm (2.5 in.) (Figure 11–4). The significance of CoM motion for this discussion is that forces acting on the body during locomotion are conceptualized as acting through its CoM. In an upright standing human, the CoM is located just anterior to the S2 segment of the sacrum and, in general, during normal locomotion, it can be assumed to reside approximately there as well.

▲ Figure 11–2 Displacement of center of mass (COM).
**Figure 11–3** Vertical displacement of CoM (Reproduced with permission from Inman VT, Eberhart HD. J Bone Joint Surg Am 1953;35:543.)

**Figure 11–4** Displacement of CoM with gait: horizontal displacement (a), vertical displacement (b), and a composite of both horizontal and vertical
Putting It All Together

When a person stands, his or her body weight is applied as a force against the support surface of the ground and conceptualized as originating from the CoM. Newton’s Third Law states that when two bodies exert force on each other, these forces (termed action and reaction forces) are equal in magnitude, but opposite in direction. By this law, a reaction force is generated by the ground against the standing person’s body weight, the so-called ground reaction force (GRF). The GRF can be depicted as a vector having magnitude and direction. As a person walks and weight is shifted from one limb to the other, a series of GRFs are generated as weight is loaded onto, fully borne by, and then unloaded from each limb respectively and repetitively. GRFs can be measured in the laboratory using force plates situated in the lab’s flooring. Force plate output (normal and tangential shear forces) can then be processed electronically to generate a visible line superimposed over the video of a walking subject that represents the GRF vector in magnitude and direction. The GRF provides clinical information on magnitude and direction of joint moments for the weight-bearing limb as the vector passes anterior, posterior, lateral, or medial to the various joint centers of rotation. Many of the photographs and illustrations in this chapter depict the GRF generated by a subject walking in a gait laboratory.

PHASES & SUBPHASES OF GAIT

Human locomotion is characterized by a sequence of limb and trunk motions that have functional significance for advancing the body’s CoM and providing upright stability. The organized, complex sequence of joint motions inherent in walking has good repeatability and tight variability (ie, the standard deviation of gait parameters is small). Furthermore, bipedal human locomotion is a cyclical behavior, especially involving one or another lower limb maintaining contact with the ground at all times (Table 11–1).

Table 11–1 Characteristics of gait.
<table>
<thead>
<tr>
<th>Term</th>
<th>Definition/Description</th>
<th>Example/Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gait cycle</td>
<td>Behavior that begins with an event involving one extremity and continues until the event is repeated once again with the same extremity.</td>
<td>The distance measured from a point on one foot to the same point on that same foot at the next stance.</td>
</tr>
<tr>
<td>Stride length</td>
<td>One complete gait cycle.</td>
<td></td>
</tr>
<tr>
<td>Step</td>
<td>Activity that begins with an event involving one extremity and continues until the event is repeated with the contralateral extremity.</td>
<td></td>
</tr>
<tr>
<td>Step length</td>
<td>Longitudinal distance between successive heel contacts of opposite feet; varies directly with height and inversely with age.</td>
<td>The distance measured during double support, from the foot that has just completed single support to the foot that has just completed swing. Left step length is measured when the left foot has just completed swing; likewise, right step length is measured when the right foot has just completed swing.</td>
</tr>
</tbody>
</table>

![Diagram](Image)

- **Left Step Length**
- **Right Step Length**

**Lateral Distance Between the Heels (BASE WIDTH)**

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition/Description</th>
<th>Example/Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Free velocity</td>
<td>Rate of ambulation chosen by a person when asked to walk comfortably; varies directly with height and inversely with age.</td>
<td>Normal adult = 80 m/min, 1.3 m/s, 4.3 ft/s, 3 mi/h; 90% of all individuals fall in the range 0.9–1.8 m/s (3.0–5.9 ft/s).</td>
</tr>
<tr>
<td>Cadence</td>
<td>The number of steps per unit time (1 min).</td>
<td>Normal adult = 90–120 steps/min. (See earlier figure.)</td>
</tr>
<tr>
<td>Base of support*</td>
<td>Mean lateral distance between the heels.</td>
<td>90% of individuals fall in the range of 2.5–12.7 cm (1–5 in.).</td>
</tr>
<tr>
<td>Substitution mechanism</td>
<td>Motion used in an attempt to remedy the loss of one or another component of the normal gait pattern.</td>
<td>Circumduction is a mechanism used to achieve clearance when knee flexion or dorsiflexion is inadequate.</td>
</tr>
</tbody>
</table>
The cyclical nature of gait, with its repetitive sequencing of joint motions, has led to eight descriptive functional units or subdivisions of the gait cycle, including two major phases, seven subphases, and one event. (Some authors do away with the term *subphase* entirely and refer to seven phases and one event). The two major phases, swing and stance, incorporate the seven subphases and one event, as discussed below.

### Initial Contact

**Definition:** The instant the foot contacts the ground (*Figure 11–5*).
Commentary: For normal locomotion with a continuum of gait velocities ranging between 0.9 and 1.8 meters per second (m/s), initial contact is characterized by a heel strike. At initial contact, the GRF generates three joint moments: ankle plantar flexion, knee extension, and hip flexion.

Ankle: At initial contact, the ankle joint is about neutral, held there by contraction of dorsiflexor muscles of the foot. The GRF falls posterior to the ankle joint axis, resulting in a plantar-flexion moment that leads to
subsequent plantar flexion of the ankle.

**Knee:** At initial contact, the knee is fully or almost fully extended, the quadriceps muscle is active and acts to absorb the shock of heel strike, and the GRF falling anterior to the knee also contributes to preventing sudden knee flexion collapse resulting from the abrupt impact of heel contact with the floor, especially at higher gait velocities.

**Hip:** The hip at initial contact is flexed 30 degrees. With the GRF falling anterior to the hip and creating a flexion moment, previous muscle activity of the hamstrings that has helped to decelerate the swinging limb now helps to control the hip’s flexion moment at initial contact, taking note that gluteus maximus and adductor magnus contraction are only just beginning to activate.

>Loading Response

**Definition:** Shifting body weight onto the stance limb immediately following initial contact ([Figure 11–6](#), right) to lift of the opposite extremity ([Figure 11–6](#), left).
Commentary: Loading response reflects the shift of body weight from the contralateral support limb to the new support limb. During the loading response subphase, a period of double support exists because both limbs are actually in contact with the ground until the contralateral limb is lifted off.

Ankle: The loading response subphase begins immediately after initial contact and, at the ankle, is marked by gradual plantar flexion of 15 degrees, controlled by activity of the pretibial muscles (tibialis anterior, great and long toe extensors, and peroneus tertius). Heel strike may be heard but, normally, controlled plantar flexion prevents foot slap. The GRF falls posterior to the ankle joint (see Figure 11–6).

Knee: During the loading response, a flexion joint moment occurs at the knee,
the GRF falling posterior to the knee joint. Eccentric contraction of the quadriceps group controls the rate at which the knee flexes. Contraction of the hamstrings quiets down as their role in decelerating the swinging limb comes to an end. At end of the loading response, the knee is flexed approximately 15 degrees.

**Hip:** Hip flexion of 30 degrees remains fairly constant throughout loading response. The GRF, initially anterior to the hip at initial contact, moves closer to the hip’s axis of rotation during the loading response. This reduces the effect of the hip flexion moment that is present at the instant of initial contact. Restraint of the hip flexion moment is provided by the gluteus maximus and adductor magnus. The activity of both of these muscles increases after initial contact while activity of the hamstrings deactivates.

**Midstance**

**Definition:** Lift of the contralateral extremity (Figure 11–7, right) to a position in which the body is directly over the stationary foot (Figure 11–7, left).
Commentary: In normal locomotion, lift of the contralateral extremity occurs concurrently with toe-off, especially the great toe. When the contralateral toes come off the ground, the period of ipsilateral single limb support of body mass begins. During midstance, the foot maintains contact with the ground throughout the entire subphase.

Ankle: At the onset of midstance, the ankle remains slightly plantar flexed (about 5 degrees). As the period of midstance develops, ankle movement to about 5 degrees of dorsiflexion materializes. The GRF moves over the forefoot as the tibia progresses forward (see Figure 11–7). With the GRF
now anterior to the ankle, a dorsiflexion moment takes effect, reflecting forward acceleration of the tibia. This moment is controlled by activity of the soleus. However, gastrocnemius activity also contributes to attenuating the dorsiflexion moment that controls the rate of advancement of the tibia.

**Knee:** Maximum stance phase knee flexion is present at the beginning of midstance. During this subphase the knee undergoes extension by virtue of active quadriceps contraction. By the middle of midstance, the GRF moves just anterior to the knee joint’s axis of rotation, resulting in an extensor joint moment. Therefore, at the end of midstance, there is no longer any need for quadriceps activity as extension stability is provided passively by bony segment alignment and posterior soft tissue structures.

**Hip:** During midstance, the hip progressively extends as the tibia and femur advance forward. In the early part of the subphase, the hamstrings (semitendinosus and semimembranosus) contribute to extension, but, by late midstance, no further muscular effort is required. During the single support of midstance, the weight of the contralateral swing limb causes the pelvis on that side to drop about 4 degrees. The hip abductors, chiefly gluteus medius, are responsible for preventing excessive pelvic tilt.

### Terminal Stance

**Definition:** Subphase immediately following the position in which the body is directly over the stationary foot (Figure 11–8, right) to a point just prior to initial contact of the contralateral limb (Figure 11–8, left).
Figure 11–8 Terminal stance.

**Commentary:** The main force of propulsion advancing the body forward occurs during terminal stance. As the tibia continues to rotate forward in early terminal stance, buildup of calf muscle tension soon raises the heel off the ground. The GRF moves well into the forefoot and the CoM follows its sinusoidal path, descending from its high point at the end of midstance.

**Ankle:** In early terminal stance, the ankle continues to dorsiflex approximately 10 degrees, allowing the tibia to rotate forward. Calf muscle contraction subsequently intensifies and the heel rises. Dorsiflexion motion gives way to plantar flexion such that by the end of terminal stance (ie, the end of single support), the ankle reaches about 5 degrees of plantar flexion. The GRF has moved well anterior to the ankle joint along the forefoot (resulting in a long
lever arm) so that strong contraction of both the soleus and gastrocnemius muscles is able to accomplish heel rise and control the dorsiflexion moment generated by the GRF. Heel rise also initiates dorsiflexion at the metatarsophalangeal joints. When body weight is transferred to the contralateral limb, calf muscle contraction deactivates quickly.

**Knee:** During terminal stance, the knee is in full extension and remains so for the duration of this subphase. No quadriceps activity is needed to control the knee because the GRF vector is anterior to the knee joint. Knee stability is maintained and hyperextension is prevented by the posterior ligamentous and capsular structures.

**Hip:** As the CoM advances beyond the supporting foot and the GRF passes posterior to the hip, passive extension of the hip occurs and hip extensor contraction is not required throughout terminal stance. By the end of terminal stance, the hip has moved into about 10 degrees of hyperextension, a position exerting stretch on the iliacus that commonly initiates hip flexor contraction. The forward propulsion effect of calf muscle contraction in terminal stance followed by hip flexor contraction in preswing combine to advance the body along the line of progression, with the CoM falling forward and downward, driven by momentum.

### Pre-Swing

**Definition:** Interval from initial contact of the contralateral limb ([Figure 11–9](right)) to a point just prior to lift ([Figure 11–9](left)) of the ipsilateral limb from the ground.
**Commentary:** Pre-swing is technically a stance phase entity but it is given a swing appellation because hip flexor contraction coupled with unloading of the limb is an important source of acceleration of the CoM along its sinusoidal trajectory.

**Ankle:** During pre-swing, the ankle plantar flexes to about 20 degrees of plantar flexion. This is a momentum-driven behavior since the plantar flexor muscles have already ceased contraction at this point, their role in active propulsion being complete.

**Knee:** During pre-swing, the knee flexes about 35 degrees, driven passively by actively contracting hip flexors; no knee flexor muscles are active during this subphase. From another perspective, a flexion torque is created by the GRF falling behind the knee, as the tibia advances forward.

**Hip:** Dynamic flexion of the hip is generated during this subphase, primarily by
contraction of the iliacus as well as the rectus femoris.

**Initial Swing**

**Definition:** Interval from lift of the foot off the ground (Figure 11–10, right) to maximum knee flexion (Figure 11–10, left).

**Commentary:** As the hip continues to flex, bringing the thigh segment upward and forward, the knee also flexes, keeping the toes of the plantar-flexed foot away from the floor.

**Ankle:** Active dorsiflexion of the ankle takes place in this subphase but only 10 degrees of plantar flexion is achieved. Therefore, toe clearance in early
swing does not depend on ankle dorsiflexion. Muscles that act to dorsiflex the foot include the tibialis anterior, extensor digitorum, hallucis longus, and peroneus tertius. The antagonist calf muscles are silent.

**Knee:** Momentum generated by contracting hip flexors generates knee flexion torque, bringing the knee to an angle of about 60 degrees by the end of initial swing. The knee flexion moment may have contributions from the biceps femoris, sartorius, and gracilis, especially at higher gait velocities.

**Hip:** At the end of pre-swing, the hip has achieved the neutral position. During the initial swing subphase, the hip moves to 20 degrees of flexion. The iliacus is the prime hip flexor at this point; however, activity of the sartorius, gracilis, and adductor longus may also contribute.

▶ **Midswing**

**Definition:** Subphase immediately following maximum knee flexion (Figure 11–11, right) to a vertical tibia position (Figure 11–11, left).
Commentary: As the swing limb advances and the tibia moves toward a vertical position, clearance of the foot from the floor is facilitated by active ankle dorsiflexion.

Ankle: Dorsiflexion of the ankle to neutral is completed and sustained by the anterior compartment muscles. Development of a vertical tibia continues the need for active foot control by the tibialis anterior, peroneus tertius, and extensor digitorum longus.

Knee: The knee undergoes passive extension in preparation for initial contact. This movement is driven by momentum and facilitates advancement of the swing limb. When the tibia is vertically oriented at the end of midswing, the
knee angle is in approximately 30 degrees of flexion.

**Hip:** The hip reaches 30 degrees of flexion, its maximum value, during midswing by contraction of the iliacus muscle.

### Terminal Swing

**Definition:** Subphase immediately following a vertical tibia position (Figure 11–12, right) to a point just prior to ground contact (Figure 11–12, left).

![Figure 11–12 Terminal swing.](image)

**Commentary:** The foot is the leading edge of limb advancement. As advancement of the swing limb ends, the limb is positioned for the upcoming stance phase by deceleration of hip flexion, knee extension, and continued
dorsiflexion of the ankle.

**Ankle:** The ankle basically holds in neutral through action of the pretibial muscles.

**Knee:** The tibia moves from a vertical position with respect to the ground to an oblique position at the end of terminal stance (see Figure 11–12), placing the knee in roughly full extension. At this point the hamstrings are active to decelerate the rate of knee extension and prevent hyperextension, particularly at higher velocities. The quadriceps group also becomes active to stabilize the joint during the impact of initial contact and in anticipation of the flexor moment that will be encountered in the loading response.

**Hip:** The hip maintains 30 degrees of flexion throughout terminal swing. Momentum moves the limb forward while active contraction of the hamstrings restrains further hip flexion, decelerating forward momentum (as it does for knee extension). In effect, restraint by hamstring muscles, such as the semimembranosus, semitendinosus, and biceps femoris, functions to control the acceleration and position the swinging limb in preparation for initial contact.

The subphases thus far described are defined with respect to the sagittal plane. In the frontal plane, key observations are made regarding stability in single limb support, primarily during midstance (Figure 11–13).
In a gravitational environment, translation of the human center of gravity by means of a bipedal strategy requires management of certain operational objectives or features. These operational features include (1) advancing the CoM continuously along the direction of progression from a swing phase perspective.
and a stance phase perspective; (2) stability of loading of the stance limb during double support and the subsequent single limb support; and (3) foot clearance (of the floor) during swing phase. The individual subphases described previously are combined in specific ways to achieve these operational features of human locomotion on level ground. The operational features discussed here—swing phase advancement, stance phase advancement, weight-bearing support, and foot–floor clearance—promote several “functional” products of locomotion, such as step and stride length, base width, cadence, velocity, and rhythmicity, mentioned later in the chapter.

▶ Swing Phase Advancement

Swing phase advancement refers to the forward propulsion of the CoM, accompanied by a swinging limb (Figure 11–14). Dynamic flexion of the hip is powered by the iliacus during the subphase of pre-swing, often accompanied by rectus femoris activity. Active hip flexion is a strong contributor to forward acceleration of the CoM and thigh advancement. Smooth advance of the swing phase limb begins at initiation of pre-swing (Figure 11–14, right) when the contralateral limb makes initial contact, beginning the loading process for the “contra-” limb simultaneously with the unloading process for the “ipsi-” limb. Hip flexor contraction during pre-swing begins a process of thigh advancement by means of a hip flexion moment, eventually adding leg advancement during mid- and terminal swing. The thigh segment advances forward in pre-swing, even as the anterior portion of the foot retains waning contact with the ground. As a consequence of thigh advancement in pre-swing, during which some contact between foot and ground still remains, the knee begins to flex. Momentum generated by the hip flexors continues to generate a knee flexion moment through the next subphase of initial swing. The knee flexion moment may be supplemented by contraction of the biceps femoris, sartorius, and gracilis. Knee motion reverses in the next subphase of midswing when the knee begins to extend, continuing its extension through the subphase of terminal swing. If one observes the trajectory of the foot from pre-swing through terminal swing and initial contact, one can appreciate swing phase advancement in the form of a stride.
Stance Phase Advancement

Forward progression of the body from a stance phase perspective begins during early loading response as the pretibial muscles activate in order to control plantar flexion of the foot (Figure 11–15). After initial contact, the foot plantar flexes onto the ground in a controlled manner. When the foot is planted on the floor, the pretibial muscles are able to act in a closed kinetic chain to pull the tibia forward. Similarly, contraction of the quadriceps group further up the chain brings the femur forward in tow, advancing the thigh segment with the leg. The segments of leg and thigh, working synchronously, continue to rotate forward. With the foot on the ground, the ankle continues dorsiflexing until the CoM passes anterior to the ankle joint, at which time the calf muscles engage to control the rate of leg advancement until they cause heel rise in terminal stance. Heel rise itself is associated with forward propulsion of the body, namely, calf muscle pushoff against the ground. Heel rise also accelerates the forward fall of the body, the tibia and femur being angled forward in this part of the gait cycle. At the point of heel rise, the CoM is ahead of the metatarsal heads (in the sagittal plane), thus the body’s mass is moving “downhill” from its previous high point at the end of midstance.
Note that in the preceding description, the heel, ankle, and metatarsophalangeal joints provide fulcrum points, transitioning the mass along the line of progression to their respective contact points with the ground. Specifically, they facilitate advancement of the superincumbent mass of the body over the stance phase support limb. Effective execution of these mechanisms assumes that the ankle has the required range of motion for plantar and dorsiflexion. If range of motion in either direction is limited, the position of the GRF relative to proximal joints will be altered. Advancement will then elicit compensatory mechanisms, resulting in deviation from the “normal” gait described here. Likewise, the knee and hip must be able to move through their respective ranges of motion to allow fluid progression. Finally, the configuration of the foot and ankle, especially that part of the foot making initial contact, will have implications for the configuration of more proximal joints, again owing to the position of the GRF.

**Weight-bearing Support**

Two units comprise weight-bearing support: the first is the period when weight is shifted onto the postswing limb entering the stance phase, a period of double support, as both limbs are in contact with the floor (*Figure 11–16*). The second is when only one limb, the stance limb, is solely in contact with the ground, and weight bearing with full single support passes through this limb (*Figure 11–17*). Weight shifts onto the limb entering stance phase during the loading response.
A. Period of Double Support

This period comprises one event (initial contact) and the subphase of loading response. The hip is flexed 30 degrees at initial contact. The GRF falls in front of the hip joint, creating a strong flexion moment. Both the gluteus maximus and the hamstrings are activated in order to control this flexion moment during the loading response. Similarly, the torso is under the same forward momentum during terminal swing, and the abrupt stop at initial contact calls upon the erector spinae group to prevent flexion just above the hip at the intervertebral joints.

For the knee at initial contact, the GRF falls anterior to the joint axis, creating a knee extension moment. Continuing on from the previous terminal swing, activity of the quadriceps and hamstring help stabilize the neutral position of the knee joint. Shortly after initial contact, the GRF moves behind the knee and the knee flexes approximately 15 degrees. The quadriceps fire eccentrically, helping to absorb energy of the somewhat jarring impact of initial contact on the heel (Figure 11–16, right).

As the limb enters stance phase, ankle–foot position is critical for receiving weight of the superincumbent mass (Figure 11–16, middle and left images). At initial contact, the ankle is in neutral. In the sagittal plane, the GRF is posterior to the ankle joint, creating a plantar-flexion moment. The pretibial muscles (tibialis anterior, extensor digitorum longus, and extensor hallucis longus) control this plantar-flexion moment by means of a lengthening contraction during the loading response, when the foot plantar flexes 15 degrees. When the foot becomes fixed on the ground, the relationship between the body and the
stance limb is akin to an inverted pendulum. The stance limb represented by this pendulum will rotate forward, controlled by an interplay of dorsiflexor and plantar-flexor muscles, starting at double support and ending during single limb support.

**B. Period of Single Support**

The period of single limb support begins with contralateral toe-off during the subphase of midstance. Throughout midstance, the hip extends as the tibia and femur advance forward in order to maintain an upright posture of the spine and torso (Figure 11–17, middle three images from right). At this point these actions occur devoid of any need for muscle contraction.

![Figure 11–17 Weight shifting onto the limb entering stance phase.](image)

At the beginning of midstance, the knee is at its point of maximum flexion during the stance phase (about 15 degrees). Because the GRF falls behind the knee, quadriceps action is necessary to prevent collapse and provide stability. By the second half of midstance, knee extension stability is provided passively by the posterior ligamentous structures as the GRF is now anterior to the knee joint center.

At the beginning of midstance, the ankle remains slightly plantar flexed and as the subphase develops, the ankle gradually dorsiflexes. As the tibia rotates forward, the GRF moves along the forefoot (Figure 11–17, middle three images from right). When the GRF is positioned anterior to the ankle joint, a dorsiflexion moment is generated that would lead to instability were it not for the eccentric contraction of the soleus and gastrocnemius. As the soleus slows the forward progression of the tibia, preventing a “fall” into dorsiflexion, it does so in a closed kinetic chain that simultaneously holds back knee flexion,
facilitating knee extension as the femur maintains its forward velocity and ultimately passes over ankle. In midstance, when viewed from a frontal plain, there is an additional element of stability that can be appreciated during single limb support. Because of the closed kinetic chain, the pelvis is apt to rotate on the head of the femur. At this time, the CoM is medial to the axis of rotation of the weight-bearing hip. The medial CoM produces an adduction moment of the joint, allowing the pelvis to rotate around a sagittal axis through the hip joint. Pelvic tilt or pelvic drop to the side of the swing limb requires muscular control. The stabilizing forces here are the hip abductors, particularly the gluteus medius. By providing an eccentric contraction, the tilt of the pelvis is controlled (about 4 degrees) and the pelvis remains in a more or less horizontal position.

Clearance

Advancement of the swing phase limb requires clearance of the foot from the floor, especially the toes. Starting with the end of terminal stance and continuing on through the end of pre-swing (Figure 11–18, three images on right), the toes come off the floor as a result of proximal hip and knee flexion. At the end of pre-swing (Figure 11–18, middle image), the knee is flexed about 30 degrees. The knee continues to flex during initial swing to a maximum of about 60 degrees (Figure 11–18, left image). The hip flexes to 30 degrees. Proximal hip and knee flexions are the main clearance mechanism during early swing. By the end of initial swing, the ankle is still in plantar flexion, giving evidence that hip and knee flexion rather than ankle dorsiflexion are prime factors generating clearance of the swinging foot from the ground during early swing. As the thigh and leg continue to advance forward (Figure 11–19), the ankle continues to dorsiflex, reaching neutral by the end of midswing (Figure 11–19, left image) and remains so until just after initial contact.
Figure 11–18 Clearance resulting from hip and knee flexion.

Figure 11–19 As the swing limb advances dorsiflexion becomes instrumental in clearance.

PATHOLOGIC GAIT

Functional Performance & Common Gait Deviations

Normal locomotion generates several functional “products.” These include the
production and ability to manage (1) a range of step and stride lengths; (2) a range of base widths; (3) a range of surface contours and topographies, including ramp and stair climbing; (4) a range of velocities and cadences; (5) a range of energy-consuming gaits; and (6) changing direction and making turns. A range of normal performance parameters is presented in Table 11–1, cited earlier.

As previously described, there are four major operational features of gait that result in the production of functional gait “products.” These include stance phase advancement, swing phase advancement, weight-bearing stability with support against the effect of gravity on jointed body segments, and foot–floor clearance. Neurological, musculoskeletal, cardiopulmonary, and psychological impairments generate gait deviations that affect the operational features of gait, resulting in performance deficits such as shortened step and stride lengths and a narrowed range of available gait velocities. The next section describes various common gait deviations, detailing how they relate to the operational features of gait and, as a result, how functional performance is affected.

Stance Limb Advancement & Gait Deviations

Mechanisms that contribute to stance limb advancement include active ankle dorsiflexion during early stance, active extension of the knee in midstance, passive hip extension throughout stance, ankle plantar flexion in terminal stance, and knee flexion control in late stance in conjunction with ankle plantar flexion.

Figure 11–20 illustrates the normal sequence of stance phase advancement for a right lower extremity. In contrast, Figure 11–21 reveals a patient who has difficulty with stance limb advancement. Ankle dorsiflexion does not develop after full contact of the foot with the floor during loading response and onward through early terminal stance. Knee hyperextension develops by the end of loading response and persists through most of the subsequent stance phase. Passive hip extension is absent throughout stance and an anterior lean of the trunk is present. Ankle plantar flexion does not develop in terminal stance and there is no pushoff to power translation of the center of gravity. The functional consequences of these gait deviations are a shortened contralateral step length and a slow gait velocity. Restrained motion at the ankle, knee hyperextension, an anterior trunk lean, and a flexed hip, collectively and individually, typically result in a slow gait velocity and a shortened contralateral step length (see Figure 11–21).
Figure 11–20 Normal stance advancement.

Figure 11–21 Impaired stance advancement.

Swing Limb Advancement & Gait Deviations

Mechanisms that contribute to swing limb advancement include active hip
flexion during early swing, contralateral arm swing during early and midswing, passive knee extension (mid- and terminal swing), pelvic rotation about a vertical axis (the pelvis rotating 4 degrees forward and 4 degrees backward during normal gait), and forward falling of the trunk after midstance. **Figure 11–22** illustrates the normal sequence of swing phase advancement for a right lower extremity. In contrast, **Figure 11–23** reveals a patient who has difficulty with swing limb advancement. Active hip flexion during early swing is an important mechanism underlying swing limb advancement. The sequence in **Figure 11–23** reveals a patient whose hip is fused in neutral, preventing active hip flexion. As a consequence, he generates a posterior trunk lean in order to advance the right lower limb and achieve a right step. The compensatory gait deviations that actually enable the posterior lean to materialize include excessive dorsiflexion of the left ankle and excessive flexion of the left knee in terminal stance (see **Figure 11–23**, last two images on right). The combined deviations of posterior trunk lean, excessive left ankle dorsiflexion, and knee flexion in terminal stance enable the patient to compensate for absent hip flexion on the right, resulting in the production of a reasonable right step length. Walking for this patient is arduous, and he walks slowly.

![Figure 11–22 Normal swing advancement.](image)
An important mechanism of swing phase advancement is passive knee extension during mid- and terminal swing, normally generated by momentum consequent to contralateral pushoff and ipsilateral active hip flexion. Figure 11–24 reveals a patient who is just making initial contact with the left lower extremity. A vertical left tibia is indicative of absent knee extension in terminal swing, the latter subphase of gait having dropped out completely for this patient. This patient has an upper motor neuron syndrome and involuntarily active hamstrings restrain knee extension during the operational feature of swing phase advancement, resulting in a shortened left step.
Figure 11–24 Shortened step length owing to loss of knee extension.

Clearance & Gait Deviations

Mechanisms that contribute to clearance of the floor by the foot include hip and knee flexion during pre-swing and initial swing, ankle dorsiflexion in mid- and terminal swing, and tilt of the pelvis to the side of the swing limb controlled by the contralateral hip abductors during single limb support. Figures 11–25 through 11–27 illustrate the normal sequence for clearance of the swing phase.
**Figure 11–25** At the end of pre-swing, the knee flexes about 30 degrees.
▲ Figure 11–26 At the end of initial swing, the knee flexes as much as 60 degrees. The hip is flexed 30 degrees.
△ Figure 11–27 By the end of midswing, the ankle has dorsiflexed to neutral (0 degrees).

In the frontal plane, the pelvis normally tilts about 5 degrees to the side of the swing limb, resulting in a drop of the whole limb toward the floor. Too much pelvic tilt owing to weakness of the contralateral hip abductors of the stance limb can result in a foot drag (impaired clearance) of the swing limb. The deviation of excessive pelvic tilt (also termed positive Trendelenburg) results in a greater drop of the swing limb toward the floor, impaired clearance being a potential consequence if the patient is unable to compensate (Figure 11–28).
**Figure 11–28** Trendelenburg gait. Solid line indicates normal 5-degree pelvic tilt (about a sagittal axis through the pelvis); horizontal arrow points to shaded hip abductors (gluteus medius); solid line from pelvis to medial foot represents the GFR (ground reaction force); dotted line, excessive pelvic tilt; vertical arrow, excessive drop of whole limb toward the floor.

**Figure 11–29** depicts a patient using circumduction to clear the foot from the floor during swing phase on the left. The patient has a “stiff” (extended) knee in swing phase, and knee flexion during pre-swing through midswing is particularly deficient, resulting in a functionally increased leg length and potentially impairing foot–floor clearance. The patient uses the compensatory
motor behavior of circumduction, a gait deviation best viewed behind the patient (see sequential heel positions of left swing). Note how the trajectory of the heel takes a circuitous anterolateral to anteromedial route.

▲ Figure 11–29 A patient using circumduction to clear the foot from the floor during swing phase on the left.

▶ Weight-bearing Stability

Transfer of body weight normally begins during the loading response. Patients with equinovarus deformity of the ankle–foot system make initial contact with the lateral border of the foot (Figure 11–30; compare A and B). Such contact puts pressure on the lateral border of the foot and, especially, on the head of the fifth metatarsal bone. Weight bearing in this type of unstable configuration, especially if painful, is poorly tolerated, and patients attempt to shorten the time they spend in stance phase on such a limb. The contralateral step becomes shortened as the patient hurriedly shortens stance time on the equinovarus limb. Overall gait performance is slowed and dysrhythmic.
**Figure 11–30** A: Equinovarus in terminal swing. **B:** Comparison of terminal swing in the same patient.

Single support begins with contralateral toe-off (Figure 11–31, far left) and terminates with contralateral initial contact (Figure 11–31, far right). Antigravity support of body weight is most critical during the period of single support. **Figure 11–31** illustrates the sequence of lower limb configurations during the period of single support. The amplitude of the GRF for the right lower limb in the photo is approximately constant and represents full body weight. Knee extension beyond midstance is observed, and hip extension is noted to occur throughout the period of single limb support.
Figure 11–31 The sequence of lower limb configurations during the period of single support.

Figure 11–32 reveals a patient who has a problem with single limb support. The patient is observed holding onto the parallel bars during single limb support for the left lower extremity. The middle image in Figure 11–32 reveals that the patient has a flexed hip and a flexed knee in what should be the subphase of terminal stance. These deviations are in striking contrast to the extended hip and knee seen in the rightward comparison figure of normal terminal stance. The magnitude of the patient’s GRF is small, suggesting that a large proportion of body weight is being absorbed by the upper limbs grasping the parallel bars. Excessive dorsiflexion of the patient’s left ankle may be contrasted with plantar-flexion pushoff and heel-off of normal terminal stance seen in the rightmost figure. Because the patient has absent pushoff, a flexed knee, and a flexed hip, the right step length is shortened. A flexed right knee in terminal swing also contributes to the shortened right step length.
Figure 11–32 A patient who has a problem with single limb support.

CLINICAL CASE STUDY

The following case example of a patient with spastic hemiparesis, who has problems with two operational features of gait, illustrates how identifying impairments in the operational features of a patient’s gait cycle helps a clinician understand a patient’s clinical complaints and maladaptive performance. Describing impairments related to advancing the CoM, foot–floor clearance, and upright stability provides important information for conceptualizing treatment of gait dysfunction, especially regarding the use of lower extremity orthotics and assistive devices.

Description

A 55–year–old woman sustained a right hemiparesis as the result of a stroke involving the left internal capsule. When she was seen 6 months after the stroke,
she complained of tripping on the edge of a living room rug and, occasionally, on the lip of a threshold entrance into her bathroom. Clinical examination of her right side revealed full passive range of motion of hip and knee while ankle dorsiflexion was to +10 degrees (mildly reduced range of motion). Pinprick of the foot and proprioception of the great toe and ankle were intact. Hypertonia was present for the ankle plantar flexors (Ashworth 2), knee flexors (Ashworth 3), knee extensors (Ashworth 3), and hip flexors (Ashworth 2). She did not have selective control of lower limb joints individually, and she moved the lower limb forward as a whole unit during swing phase. She was able to bear weight on the limb during stance while using a cane contralaterally. Her gait pattern during early swing is shown in Figure 11–33.
Figure 11–33 Failure of clearance in early midswing secondary to lack of knee flexion.

Discussion

Recall that advancement of the swing phase limb requires clearance of the foot from the floor, especially clearance of the toes. During the early part of swing phase, the toes come off the floor as a result of proximal hip and knee flexion, the distal ankle remaining in the range of plantar flexion during pre-swing, even as ankle dorsiflexor muscles begin to contract. Active continuation of hip flexor contraction and passive knee flexion during initial and midswing lift the toe to clear the floor and advance the limb. The leftmost image in Figure 11–33 depicts the end of the pre-swing phase, when the toe is about to come off the floor. The three middle images reveal a drag of the anterior portion of the shoe along the floor, with clearance coming only in the far right image. (The duration of the drag is approximately 100 ms or 0.1 s.) The images also reveal the cause of the drag: inadequate hip and knee flexion. At the end of pre-swing (far left image), the knee is flexed about 30 degrees, as expected, but the hip is inadequately flexed due to proximal hip retraction. As the patient’s gait proceeds, the subphase of initial swing drops out entirely as the knee does not flex more than 30 degrees, even though up to 60 degrees of knee flexion is normally expected by the end of initial swing. The ankle is plantar flexed, but during the pre-swing through initial swing part of the gait cycle, the ankle is not expected to be fully dorsiflexed to neutral. Hence, the drag of the foot on the floor (ie, the lack of foot floor clearance) is a result of proximal hip and knee flexion inadequacy and not of inadequate dorsiflexion, or “drop foot.” Treatment will have to take these observations into account. (Consider: Given a patient with inadequate hip or knee flexion, or both, in early swing phase, would an ankle–foot orthosis set in neutral alleviate an early swing phase toe drag?) This initial examination of the case focuses on an impairment of clearance, corresponding to the patient’s complaint of toe drag and tripping on low-level elevations (rug edges, raised room thresholds) that ordinarily would be cleared by an advancing swing phase limb.

A second problem area unmasked by observational gait analysis relates to the operational feature of stance phase advancement. The patient’s gait pattern during the operational feature of stance advancement is shown in Figure 11–34.
Shortened step length on the left side because of lack of dorsiflexion on the right side.

At initial contact (Figure 11–34, far left), the whole sole connects with the ground as a unit and the ankle configures in plantar flexion. At the end of the loading response (second image from left), with the contralateral toe about to come off the floor, the right ankle remains in a high degree of plantar flexion, different from the slight degree of about 5 degrees of plantar flexion that characterizes normal locomotion. **FIGURE 11–34C** (end of midstance) and **Figure 11–34D** (end of terminal stance) reveal the persistent plantar flexion attitude of the ankle. The tibia remains oriented posteriorly rather than anteriorly, the heel remains on the ground, and, as a consequence, the left step is very short (Figure 11–34, far right). In this hemiparetic patient, the tibia is restrained by a tight heel cord that does not allow the tibia to advance forward. In addition, weakness of calf muscles prevents heel rise in terminal stance. The absence of calf muscle pushoff eliminates one of the main propulsive drivers of the limb in the subsequent swing phase. Restrained plantar flexion in stance phase combined with absent pushoff results in a retarded advancement of the CoM over the stance phase limb. The functional result is a shortened contralateral step, a slowed gait velocity, and temporal asymmetries of step time, stance time, and swing time that create an awkward, dysrhythmic gait, requiring more time for the patient to get where she is going and frequently making for feelings of self-consciousness.

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Spinal Cord Injury

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Spinal cord injury (SCI) results from trauma or damage to the spinal cord, causing disruption of communication between the brain and spinal cord to end organs and limbs, with resultant sensory, motor, and autonomic dysfunction. The causes of SCI can be divided into traumatic and nontraumatic. Traumatic causes are typically high-velocity events producing neurologic deficits that occur with an acute and recognizable onset. Nontraumatic injuries tend to have a subacute to chronic course, with slower onset of neurologic deficits.

GENERAL CONSIDERATIONS

Epidemiology

Current estimates indicate that between 236,000 and 327,000 people in the United States are living with SCI. There is an estimated annual incidence of SCI, not including those who die at the scene of the accident, of 40 cases per million people in the United States, or approximately 12,000 new cases per year. About 13% of new SCI cases are captured by the National Spinal Cord Injury database, which has a network of SCI model systems that first began collecting data in 1973. There have been 28 SCI model system sites since 1973, and 14 are currently active. From 1973 to 1979, the average age of injury was 28.7 years
and most injuries occurred between the ages of 16 and 30 years. Since 2005, the average age of injury have increased to 41.0 years, with 80.6% of injuries occurring in males. Before to 1980, 81.8% of SCIs occurred among males. The incidence of SCI has been decreasing in Caucasians and increasing in minority populations, particularly African Americans and Hispanics. This likely is due, at least in part, to general population trends in the United States. Epidemiologic data for traumatic SCI in British Columbia, Canada, yield similar results, with a median age of injury of 35 years and an 80% gender preference for males. In mainland China, epidemiologic data covering the period from 2001 to 2007 indicated that patients with spinal trauma accounted for 4.58% of all patients hospitalized due to trauma. Among these patients, 69.02% were male and 30.98% were female. The most common causes of spinal trauma were motor vehicle accident (33.6% of SCIs), high falls (31.25%), and falls without height (23.23%), and these causes combined accounted for about 88% of the spinal traumas documented.


**Etiology**

Data compiled since 2010 show that motor vehicle collisions account for 36.5% of reported traumatic SCIs, falls for 28.5%, violence for 14.3%, sport injuries for 9.2%, and other or unknown causes for 11.4%. In the category of sports injuries, American high school football players have the highest incidence of severe cervical injuries per year of all high school athletes. Neck and cervical spine fractures accounted for 4.1% of the 517,726 American high school football injuries reviewed from the 2005–2006 season.

Since 2010, the most frequent neurologic category at discharge of patients
with SCI is incomplete tetraplegia (40.6%), followed by incomplete paraplegia (18.7%), complete paraplegia (18.0%), and complete tetraplegia (11.6%). Less than 1% of affected individuals experience full neurologic recovery by discharge from hospital. Over the past 15 years the percentage of patients with incomplete tetraplegia has trended upward, with a corresponding decrease in the incidence of complete paraplegia and complete tetraplegia. Currently the causes of death that have the greatest impact on life expectancy of the traumatically spinal cord–injured population are pneumonia and septicemia. The determining factors in statistical life expectancy are age at the time of injury, level of injury, and degree of injury (completeness or incompleteness).


Anatomic Considerations

A. Spinal Column and Cord

The spinal cord resides in the vertebral canal, the walls of which are defined as follows: the anterior wall border is the posterior longitudinal ligament and the posterior wall border, the ligamentum flavum. The spinal cord is suspended within three layers of meningeal tissue, the innermost being the pia mater and the outermost, the dura mater; in between is the arachnoid mater. The spinal cord itself terminates in the adult no further than the L2 vertebral body, although there can be some variation, with termination occurring as high as T12. The vertebral canal itself continues to the sacral hiatus, just superior to the coccyx. In newborns, however, the spinal cord extends as low as the L3 vertebral body.

There are 33 vertebral bodies in the human spine, divided into 7 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 4 bodies that form the coccyx. They are matched with 31 pairs of peripheral nerve roots, 8 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 1 coccygeal. It is important to distinguish upper and lower motor neurons, as pathologic conditions affecting each of these structures have
significantly different clinical presentations. An upper motor neuron (UMN) is a neuron that begins in the cerebral cortex and connects to a motor nucleus in the anterior horn of the spinal cord. A lower motor neuron (LMN) connects with the motor nucleus in the anterior horn and ends as it synapses with skeletal muscle tissue, outside of the spinal cord. The conus medullaris is the distal tip of the spinal cord. The cauda equina ("horse’s tail") is a bundle of nerves containing the five lumbar nerve roots, the five sacral nerve roots, and the coccygeal nerve root. The nerves of the cauda equina consist of LMNs.

**B. Vascular Supply**

The arterial blood supply of the spinal cord consists of the anterior spinal artery and two posterior spinal arteries. They begin in the posterior cranial fossa as branches of the vertebral arteries. Radicular branches of the anterior and posterior spinal arteries feed individual levels. The artery of Adamkiewicz is a large anterior radicular artery that typically originates between spinal cord segments T9 and L2. There are two watershed areas of the spinal cord, which include the cervicothoracic junction (distal cervical cord through T4) and the thoracolumbar junction.

**C. Transmission of Neurologic Signals**

The spinal cord in cross section reveals the central butterfly-shaped gray matter surrounded by white matter that contains ascending and descending spinal cord tracts, each with specific structural architecture made up of columns of various neural tissues. The deficits associated with specific patterns of injury to the spinal cord are related to the course and location of these tracts.

The major tracts discussed in this chapter are depicted in Figure 12–1. The lateral corticospinal tract is composed of descending nerve fibers that are responsible for motor control. Nerve fibers of this tract originate in the cortex of the brain, travel caudally through the midbrain, and decussate at the level of the medulla, with 90% of the fibers crossing to become the lateral corticospinal tract. Of note, a small portion of the descending fibers split to travel both anteriorly and ipsilaterally to become the anterior corticospinal tract and the uncrossed lateral corticospinal tract, respectively. The ascending anterior and lateral spinothalamic tracts convey sensory information to the brain for nociception, temperature, and some simple light touch fibers. These fibers decussate at the ventral white commissure of the spinal cord and ascend a nerve root level before crossing over and continuing their ascension to the cortex of the brain. The dorsal columns (fasciculus gracilis and fasciculus cuneatus) also
contain sensory information. These sensory fibers differ from the spinothalamic sensory fibers in that they relay information regarding vibration, proprioception, and light touch. These fibers cross in the medulla as they continue up to the cortex of the brain.

\[\text{Figure 12–1 Ascending and descending long tracts of the spinal cord} \]


The autonomic system is composed of the sympathetic nervous system and the parasympathetic nervous system. The two systems work in concert to affect many visceral functions, including defecation, heart rate, blood pressure, and respiration, among others. Sympathetic nerves originate from the lateral horn in
The thoracic spinal cord and the intermediolateral cell column at spinal levels T1–L3. The parasympathetic nervous system originates from cranial nerves III, VII, IX, and X; the midbrain; and sacral nerve roots.


**PATHOGENESIS**

The spinal cord is composed of grey matter on the inside surrounded by white matter on the outside. Gray matter at a particular segment provides peripheral output at that level. Subsequently, segmental injury to gray matter results in compromised function at or near that level. White matter includes long ascending and descending tracts. Injury to white matter at any particular level can result in compromised function at all levels below that level.

**Traumatic Spinal Cord Injury**

In traumatic injuries, primary injury to the spinal cord results from compression or laceration of the cord, resulting in disruption of axons and neuronal membranes. This can occur with fractures, dislocations, stab wounds, and gunshot wounds. Secondary injury can occur within 8–24 hours (early) following the primary injury, resulting from a cascade of events that leads to hemorrhage, edema, infarction, and necrosis. Late secondary injury can be seen weeks, months, or years after the initial injury and can result in neuroma formation, wallerian degeneration, arachnoidopathy, and syringomyelia.

In most adults, the spinal cord terminates between the first and second lumbar vertebral bodies. As a result, fractures occurring above L1 frequently result in an SCI characterized by UMN findings. After initial spinal shock (absence of all deep tendon reflexes), increased tone may be seen, manifested by brisk reflexes or involuntary spasms, or both. Fractures at and below L2
frequently result in a cauda equina injury, characterized by LMN symptoms (hypotonia, flaccidity, diminished or absent reflexes, muscle atrophy, flaccid bowel and bladder). Fractures at the level of L1 or L2 frequently affect the conus medullaris, and if nerve root injury also occurs, can present with a mixed UMN and LMN presentation.

Bearing in mind the anatomy of the spinal cord, most SCIs result from cervical- or thoracic-level injuries rather than lumbar- or sacral-level injuries. There are several notable cervical-level fracture–dislocations. Atlantooccipital and atlantoaxial dislocations usually are fatal. Of note, individuals with rheumatoid disease, Down syndrome, and congenital skeletal dysplasias are at significantly increased risk of atlantoaxial instability and subluxation. A burst fracture of C1 is called a Jefferson fracture. Fractures involving the odontoid process, or dens, are classified based on the areas of fracture involvement. In type 1, fractures are limited to the tip of the dens (and surgery is frequently not needed). Type 2 odontoid fractures involve the base of the dens or junction with the vertebral body of C2; this is considered an unstable fracture and generally requires surgical fixation. The rate of nonunion with external fixation alone is upward of 60%. Type 3 odontoid fractures involve the body of C2 and frequently can be managed nonsurgically (90% of fractures heal with use of a halo device for 3 months). Hyperextension of the cervical spine can result in SCI (classically known as central cord syndrome, in which upper extremities are weaker than lower extremities). C4–5 is the most common level at which cervical hyperextension injuries occur. Compression fractures of the cervical spine result from cervical flexion or axial loading. C5 is the level at which cervical compression fracture most commonly occurs. Unilateral facet joint dislocation can occur from a cervical flexion–rotation injury. Bilateral facet joint dislocations occur from cervical flexion injuries. C5–6 is the most common level for both unilateral and bilateral facet joint dislocations.

Injuries to the thoracolumbar spine are most commonly seen with motor vehicle accidents. Other organ injury is seen in up to 50% of individuals with thoracolumbar trauma.

▶ Nontraumatic Spinal Cord Injury

There are many nontraumatic causes of spinal cord injury, including spinal stenosis with myelopathy, spinal neoplasms, spinal vascular disease, infections, degenerative and demyelinating diseases, and toxic and metabolic myelopathies.

Spondylosis and spondylolisthesis of the spine can cause compression of the
spinal cord. This is more frequently seen in the cervical spine but can also affect the thoracolumbar spine. Depending on the location of the stenosis (central or neuroforaminal), one may see more UMN presentations with hyperreflexia and spastic muscle weakness in myelopathy or LMN presentations with hyporeflexia and flaccid muscle weakness in radiculopathy.

Spinal neoplasms can affect the spinal cord either directly (primary spinal cord tumor) or indirectly (spinal column tumor causing compression of the spinal cord). The most common neoplasms of the spinal cord are intradural primary tumors (nerve sheath tumors, eg, schwannomas and neurofibromas; meningiomas; ependymomas; and astrocytomas). However, metastatic disease (from lymphoma, lung, breast, and prostate cancer) to the spinal column that secondarily compresses the spinal cord occurs almost 25 times more commonly than primary spinal cord tumors.

Spinal vascular disease includes spinal arterial infarction (most often as a consequence of profound systemic circulatory impairment, surgical cross-clamping, or disease of the aorta or its major branches), spinal venous infarction (most commonly in hypercoagulable states), spinal vascular embolic disease (most commonly from atheromatous or fibrocartilaginous embolism), and spinal vascular malformations (usually manifesting acutely due to hemorrhage).

Infections affecting the spinal cord can include abscesses (epidural, subdural, or intramedullary), suppurative spinal leptomenigitis (accompanying intracranial meningitis), Pott’s disease (tuberculosis of the spine), tabes dorsalis (neurosyphilis), and viral myelitis or myelopathy (poliovirus, West Nile virus, HIV).

Degenerative or demyelinating diseases include multiple sclerosis, amyotrophic lateral sclerosis (ALS), primary lateral sclerosis, spinal muscular atrophy, Friedreich’s ataxia, and hereditary spastic paraplegia.

Toxic or metabolic myelopathies can be seen in cases of vitamin $B_{12}$ deficiency, postintrathecal injection (with anesthetics, alcohols, hypertonic saline, steroids, or chemotherapeutic agents), and postangiography.


Severe SCI is associated with an immediate rise in blood pressure; however, this stage is brief and is followed by spinal shock, characterized by hypotension, bradycardia, and hypothermia. Hypotension can also result from other types of shock and should be distinguished and treated accordingly. Spinal shock is caused by decreased tone and lack of reflexes below the level of the SCI, and the drop in blood pressure is related to changes in the sympathetic tone of the vasculature. Various signs have been identified to signal that a patient is coming out of spinal shock. This transition can occur anytime from 24 hours to 3 months post-SCI and includes disappearance of the delayed plantar reflex, appearance of cutaneous reflexes (e.g., bulbocavernosus and cremasteric reflexes), return of deep tendon reflexes, and return of reflexive detrusor function.

A. Phases of Spinal Shock

Ditunno and colleagues have proposed that spinal shock be divided into four phases. Phase 1 occurs over day 0–1 and is marked by areflexia or hyporeflexia attributed to a loss of descending nervous stimulation. It has also been postulated...
that the lack of reflexes and flaccidity noted are due to hyperpolarization of the nerves caudal to the SCI.

Phase 2 starts on days 1–3 and is marked by the initial return of reflexes, attributed to supersensitivity of spinal cord tissue below the level of denervation. Proposed mechanisms for this supersensitivity include reduced excitatory neurotransmitter reuptake, increased synthesis and insertion of neurotransmitter receptors into the postsynaptic membrane, decreased degradation of the postsynaptic receptors, and altered synthesis and composition of receptor subunits. The first reflex to return in this phase is the delayed plantar reflex, which has been observed as early as several hours after the injury. Persistence of this reflex beyond 7 days is associated with poor prognosis for ambulation and motor recovery. Additional reflexes tend to return in the following sequence: bulbocavernosus reflex, cremasteric reflex, ankle jerk, Babinski sign, and knee jerk. In general, cutaneous polysynaptic reflexes are recovered before deep tendon reflexes.

Phase 3 spans weeks 1–4 and is characterized by hyperreflexia attributed to new synaptic growth. Considerable variability is seen in the return of reflexes during this phase. Autonomic function also improves, with improvements in vagus-mediated bradyarrhythmias and hypotension as well as the emergence of autonomic dysreflexia.

Phase 4 occurs over months 1–12 and is characterized by further hyperreflexia attributed to soma-supported synaptic growth, particularly of long axons. Cutaneous reflexes become hyperactive. Detrusor tone has been noted on urodynamic studies to improve at about 4–6 weeks after injury, although this may not correlate clinically with improved function of the bladder.

Associated with acute SCI are arrhythmias, most commonly bradycardia, although supraventricular tachycardia and ventricular tachycardia have also been reported. The proposed mechanism for bradycardia is disruption of the autonomic tone causing an excess of parasympathetic tone to be exerted on the myocardium. Hypotension is also commonly seen due to loss of vasoconstrictor tone and peripheral arterials, thus causing pooling of blood in the peripheral vasculature. There is often loss of bowel control and urinary retention due to an atonic and flaccid bladder acutely. Priapism is commonly noted immediately following a complete SCI and typically resolves in 1–2 days. It tends to be of a high flow, arterial, nature and is self-limited. Female patients are often amenorrheic after SCI.
B. Incomplete Spinal Cord Injury Syndromes

1. Central cord syndrome (CCS)—CCS is the most common of the incomplete SCI syndromes and results from cervical cord injury. It comprises 50% of incomplete injuries and 9% of all traumatic SCIs. Individuals with CCS present with greater weakness in the upper limbs than in the lower limbs, and often with bladder dysfunction and variable sensory loss. Most commonly CCS occurs in elderly individuals owing to hyperextension injury of a degenerative cervical spine, often as the result of a fall with a blow to front of head. CCS can occur with or without fracture or dislocation of vertebrae. The suspected mechanism of injury involves compression of the cord anteriorly by degenerative bony and disc components, and posteriorly by an inward bulging of the ligamentum flavum in a narrowed spinal canal. The exact cause of this specific pattern of injury is unclear. There is evidence that it is not related to the center of the spinal cord directly. Patients younger than age 50 tend to have a better prognosis for independent ambulation and return of bowel and bladder function. The pattern of recovery typically begins with recovery of strength and sensation in the lower limbs, followed by bowel and bladder recovery; finally, proximal and then distal upper limb strength and sensation recover.

   Cruciate paralysis is associated with fractures of the C1 and C2 vertebrae, with neurologic compromise at the cervical medullary junction. The pattern of deficits is almost exclusively limited to the upper limbs, with minimal or no lower limb deficits found. Most patients recover completely. There is no widely accepted explanation for this pattern of deficits.

2. Brown–Séquard syndrome (BSS)—BSS refers to hemisection of the spinal cord that produces a characteristic pattern of deficits. Two to four percent of all
traumatic SCIs are in a Brown–Séquard or Brown–Séquard-like pattern of injury. Classically, BBS is associated with knife or gunshot injury but it can result from any damage that causes hemisection of the spinal cord. The classic pattern of deficits is ipsilateral flaccid paralysis at the level of injury, ipsilateral loss of position and vibratory sense below the level of injury (dorsal column), ipsilateral UMN findings below the level of injury (cortical spinal tract), and contralateral loss of pain and temperature below the level of injury (spinal thalamic tract). Injuries that contain some elements of this pattern but not the complete pattern are referred to as Brown–Séquard “-like” or “-plus” syndrome. Of all SCI syndromes, BSS is associated with the greatest chance of recovery, with 75–90% of patients able to ambulate independently and 70% independently performing activities of daily living (ADLs) at discharge from an inpatient rehabilitation facility. Greater than 80% of these patients regain bowel and bladder continence. The recovery of muscle function correlates with pain and temperature sensory deficits in the given area. Typically, ipsilateral proximal extensors recover first and distal flexor muscles, last.

3. **Anterior cord syndrome (ACS)**—ACS is a pattern of injury that affects the anterior two thirds of the spinal cord, sparing the posterior columns. Most often, ACS results from vascular compromise of the spinal cord, but other causes include retropulsed discs or bone fragments. The pattern of deficits involves loss of motor function and pinprick sensation; light touch sensation is preserved, as is proprioception and deep pressure sensation. This condition carries a poor prognosis, with only 10–20% of patients showing some degree of motor recovery.

4. **Posterior cord syndrome (PCS)**—PCS is the least common of the incomplete SCI syndromes. It typically involves loss of dorsal column function, including light touch, proprioception, and deep pressure sensation, with preservation of pain, temperature, and light touch sensation and varying degrees of motor function. On physical examination, Lhermitte’s sign may be present. PCS may be caused by trauma but is also associated with various metabolic and infectious diseases, such as advanced syphilis and vitamin B\textsubscript{12} deficiency. It carries a poor prognosis of recovery.

5. **Epiconus syndrome**—This SCI involves the segment above the conus medullaris, consisting of L4–S1 cord segments, with sparing of the reflex function of the sacral segments. It manifests as a UMN lesion of L4–S1.
6. **Conus medullaris syndrome**—Conus medullaris syndrome involves the terminal end of the spinal cord between the L1 and L2 vertebrae. It is typically a symmetric LMN lesion of the S2–4 nerve roots. Pain is less common than in cauda equina syndrome, although low back pain may be present. Patients present with a flaccid bladder and rectum. Lower limb strength may be preserved or may be flaccid with findings typical of an LMN lesion. Bowel and bladder are typically areflexic with LMN dysfunction.

7. **Cauda equina syndrome**—This syndrome involves the segments below the L1–2 vertebral level, affecting the nerve roots referred to as the cauda equina (see Anatomic Considerations, earlier). This syndrome can be thought of as multiple radiculopathies and is characterized by flaccid paralysis and atrophy of the lower limbs. Although sensation may be preserved, it is often painful. The lesion is usually asymmetric. Cauda equina syndrome carries a better prognosis than UMN lesions and often occurs in combination with conus medullaris syndrome. Bowel and bladder are typically areflexic with LMN dysfunction.


### Evaluation of Spinal Cord Injury

**A. History & Physical Examination**

Diagnosis of SCI incorporates pertinent information from the history and physical examination, with a focus on elements of trauma to the neural axis and
identification of risk factors for SCI. These risk factors include a history of malignancy, epidural abscesses and hemorrhage, and exposure to infections of the spinal cord. Physical examination should include identification of UMN signs in areas innervated caudal to the cranial nerves, as well as changes in sensation, motor strength, and autonomic function in these areas. Physical examination may also be used to assess for findings associated with the underlying cause of the SCI and any concomitant injuries, in the case of traumatic event.

**B. Imaging Studies**

Radiographic and computed tomography (CT) scans allow for quick evaluation of the bony spinal column and can assist in localization of injury and provide information about the stability of the spine. Disruption of the thoracolumbar spine is often stated in terms of the three columns of the spine, as delineated by Denis. Using this approach, the anterior column is viewed as comprising the anterior longitudinal ligament and the anterior two thirds of the vertebral body; the middle column as comprising the posterior one third of the vertebral body and the posterior longitudinal ligament; and the posterior column as comprising the facet joints, ligamentum flavum, and remaining interspinous ligaments. Disruption of two or more of the columns on imaging is considered an unstable spine, requiring stabilization before mobility is permitted.

Radiographic and CT scans can also be used to evaluate for underlying causes of the SCI, such as malignancy of the vertebrae, vascular malformations, infections, or hematomas. Magnetic resonance imaging (MRI) is the ideal modality for viewing the spinal cord, as it is particularly useful for evaluation of soft tissues. It is less sensitive than CT scan to fractures of the vertebrae; however, edema in vertebrae seen on MRI has a high correlation with fracture. MRI is not useful in prognosticating functional outcomes of SCI, but hemorrhagic changes of the spinal cord on MRI correlate with a poor probability of neurologic recovery. Spinal cord injury without radiographic abnormality (SCIWORA) is typically noted in children due to many factors, including large head-to-neck ratio and increased flexibility of the pediatric spine (see later discussion). Consequently, in a high-velocity injury the spine may flex or extend, causing cord damage without injury to vertebrae. SCIWORA in the elderly typically results from a cervical hyperextension force and produces injury to the spinal cord without overt bony fracture or dislocation. As MRI becomes increasingly more sensitive to injuries of the spinal cord, SCIWORA is becoming less common.
C. Neurologic Assessment

Neurologic assessment of a person with traumatic SCI is best done using a standardized neurologic examination as endorsed by the International Standards for Neurological Classification of Spinal Cord Injury (ISNSCI), known as the International Standards (Figure 12–2). The American Spinal Injury Association (ASIA) has developed a web-based training course called InStep, located at www.asialearningcenter.com, to assist in learning how to perform the examination.
The examination consists of two main components, a motor examination and a sensory examination. The latter involves testing for sensation to pinprick and to light touch. Additionally, a rectal examination is performed to test for voluntary anal contraction and sensation to deep anal pressure. Motor strength is a reflection of corticospinal tract integrity, and sensation to pinprick and light touch reflect the functions of the spinothalamic tract and dorsal columns, respectively. Twenty-eight key dermatomes are tested for pinprick and light touch from C2 through S4–5, and 10 key muscle groups are tested for strength from C5 to T1 and from L2 to S1, as outlined in Table 12–1.

**Table 12–1** Key muscle groups for testing in spinal cord injury.
Sensation is graded on a three-point scale, from 0 to 2, where 0 represents “absent,” 1 represents “altered” (either diminished or enhanced), and 2 represents “normal.” The control point for sensation is the face, assuming cranial nerves are intact. Motor strength is graded on a six-point scale, from 0 to 5, where 0 is used to represent no movement; 1 is visible or palpable twitch, 2 is active movement through full range of motion (ROM) with gravity eliminated; 3 is active movement through full available ROM against gravity; 4 is active movement through full available ROM with moderate resistance; and 5 is active movement through full available ROM with full resistance. The total number of motor points is obtained by adding the motor strength scores for the upper as well as lower limbs. Similarly, a score is obtained for sensation by adding up the total scores for each dermatome.

The neurologic examination yields several key levels that assist in classification and functional prognostication. The motor level is defined as the most caudal level with a muscle grade of 3 out of 5 or greater with all key muscle groups above this level having a muscle grade 5 out of 5. The sensory

<table>
<thead>
<tr>
<th>Root Level</th>
<th>Muscle Group</th>
<th>Muscles</th>
</tr>
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<tbody>
<tr>
<td>C5</td>
<td>Elbow flexors</td>
<td>Biceps, brachials</td>
</tr>
<tr>
<td>C6</td>
<td>Wrist extensors</td>
<td>Extensor carpi radialis longus and brevis</td>
</tr>
<tr>
<td>C7</td>
<td>Elbow extensors</td>
<td>Triceps</td>
</tr>
<tr>
<td>C8</td>
<td>Long finger flexors (distal phalanx of middle finger)</td>
<td>Flexor digitorum profundus</td>
</tr>
<tr>
<td>T1</td>
<td>Finger abductors (little finger)</td>
<td>Abductor digiti minimi</td>
</tr>
<tr>
<td>L2</td>
<td>Hip flexors</td>
<td>Iliopsoas</td>
</tr>
<tr>
<td>L3</td>
<td>Knee extensors</td>
<td>Quadriceps</td>
</tr>
<tr>
<td>L4</td>
<td>Ankle dorsiflexors</td>
<td>Tibialis anterior</td>
</tr>
<tr>
<td>L5</td>
<td>Long toe extensor</td>
<td>Extensor hallucis longus</td>
</tr>
<tr>
<td>S1</td>
<td>Ankle plantar flexors</td>
<td>Gastrocnemius, soleus</td>
</tr>
</tbody>
</table>
level is the most caudal level with normal (2 out of 2) sensation to both light touch and pinprick. These levels can be further divided into right and left. The neurologic level of injury (NLI) is the most caudal level at which both motor and sensory function is intact bilaterally. Paraplegia is defined by injury from T1 and below (caudal), with the upper limbs intact. Tetraplegia results from injuries above T1, with all limbs being affected.

The ASIA Impairment Scale (AIS) classifies SCI as complete or incomplete, using categories A through E as detailed in Table 12–2. This classification schema is helpful for communication and prognostication.

### Table 12–2 American Spinal Injury Association (ASIA) impairment scale.

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>A Complete</td>
<td>No motor or sensory function is preserved in sacral segments S4–5</td>
</tr>
<tr>
<td>B Incomplete</td>
<td>Sensory but not motor function is preserved below the NLI (either deep anal pressure or PP or LT at S4–5)</td>
</tr>
<tr>
<td>C Incomplete</td>
<td>Motor function is preserved below the NLI and more than half the key muscles below the NLI have a muscle grade &lt; 3</td>
</tr>
<tr>
<td>D Incomplete</td>
<td>Motor function is preserved below the NLI and at least half (or greater) the key muscles below the NLI have a muscle grade of ≥ 3</td>
</tr>
<tr>
<td>E Normal (full recovery)</td>
<td>Motor and sensory function are intact</td>
</tr>
</tbody>
</table>

NLI, neurologic level of injury; PP, pinprick; LT, light touch.

*Motor function preservation below the NLI can be achieved in one of two ways: (1) intact voluntary anal contraction, or (2) sacral sensory sparing plus motor sparing more than 3 levels below the motor level of injury, which may include non-key muscle groups.*
Developed in 2009, the Autonomic Standard Assessment Form is a means of documenting the autonomic nervous system in the context of SCI. The grading scale is divided into three parts: general autonomic function, which evaluates autonomic control of cardiac function, temperature regulation, and autonomic and somatic control of the bronchopulmonary system; (2) lower urinary tract, bowel, and sexual function; and (3) urodynamic evaluation.


**TREATMENT**

**Acute Management**

The management of an acute SCI begins in the field. Emergency personnel should immobilize all suspected SCIs using a hard cervical collar and secure patients on a hard backboard. Spinal immobilization should be continued if SCI is confirmed until definitive treatment occurs. In traumatic events, a concomitant brain injury should be suspected in a patient with an altered level of consciousness. Other injuries commonly associated with SCI include fractures, intraabdominal and pelvic injuries, and pulmonary contusions and hemorrhage.

As with all traumatic injuries, an immediate assessment of the patient’s airway, respiration, and circulation should occur. Should the patient require intubation due to airway or respiratory complications or to the level of injury itself, the jaw-thrust maneuver should be used to prevent further injury to the spine.

Emergency personnel should be mindful of potential cardiopulmonary compromise associated with SCIs. Hypotension commonly occurs. Hemorrhage must be ruled out as a potential cause of low blood pressure. Next, neurogenic shock (bradycardia, hypotension, and peripheral vasodilation) should be
considered as a culprit. The treatment of neurogenic shock starts with fluid resuscitation, typically an isotonic crystalloid solution, with a goal systolic blood pressure of 90–100 mm Hg. Care providers must be cautious to avoid flash pulmonary edema resulting from overadministration of intravenous fluids. Atropine may be used for significant bradycardia.

The use of corticosteroids in the setting of acute SCI has been the focus of much debate. The National Acute Spinal Cord Injury Study (NASCIS) trials I, II, and III were completed in the United States in an attempt to demonstrate functional gains after the administration of methylprednisolone acutely. In NASCIS I, it was determined that the dose of steroid used was inadequate. However, statistically significant adverse effects of steroid usage were still noted. NASCIS I laid the foundation for the following two trials. In NASCIS II, methylprednisolone (30 mg/kg bolus followed by 5.4 mg/kg per hour over 23 hours) was compared with naloxone (5.4 mg/kg bolus followed by 4.5 mg/kg per hour over 23 hours) and placebo. NASCIS II was a negative study. However, post-hoc analysis revealed that patients treated with steroid within 8 hours of injury had statistically significant motor recovery, which lasted up to the final assessment at 1 year. Of note, this motor recovery was defined by an improvement of only 5 motor points. Lastly, NASCIS III, completed in 1997, compared tirilazad (an antioxidant agent) with methylprednisolone (5.4 mg/kg per hour) given over both 24 hours and 48 hours. All patients received an initial bolus of methylprednisolone. This was also a negative study. As with NASCIS II, post-hoc analysis revealed a 5-point motor recovery at 1 year. However, Functional Independence Measure (FIM) scores remained unchanged at 1 year.

As stated in the clinical practice guidelines for health care professionals, “no clinical evidence exists to definitively recommend the use of any neuroprotective pharmacologic agent, including steroids, in the treatment of acute SCI in order to improve functional recovery.” Until recently, the Congress of Neurological Surgeons (CNS) held that physicians who utilize steroids in the treatment of acute SCI should be aware that the known adverse side effects of these agents outnumber any suggested benefits. In March 2013, the CNS released a new statement, indicating that “Administration of methylprednisolone (MP) for the treatment of acute SCI is not recommended” due to lack of clinical evidence to support its use. Despite the lack of substantiation, the use of methylprednisolone in acute SCI has been common practice in many institutions. This may change, however, in response to the CNS’s definitive statement. If steroids are utilized acutely, the NASCIS trials are used as a guide for dosing and treatment length.
Nonoperative versus Operative Treatment

Once SCI is diagnosed in an individual with an interrupted spinal column, consideration of definitive treatment begins. Two routes for intervention exist: bracing and surgical intervention. The goal of either treatment is a painless, balanced, stable spine with optimum neurologic function and maximum spinal mobility. Nonoperative management is indicated for stable injuries that do not have the potential for progressive deformity or neurologic injury. Many bracing options exist. The type of brace chosen depends on the level of injury requiring relative immobilization. The principle of bracing is to provide a force vector opposite to that of the injury. Some key bracing options are listed in Table 12–3. For more details, refer to Chapter 28.

Table 12–3 Common options in spinal bracing.
Indications for surgical intervention include the need for spinal cord decompression or for spinal stabilization in patients with significant neurologic deficits or with unstable spinal injuries. Examples of injuries that commonly require surgical intervention include fracture–dislocations, flexion–distraction injuries, and burst fractures with neurologic deficits. Relative contraindications to operative intervention include complete SCI for more than 24 hours and a medically unstable patient.

Experimental treatments to decrease or halt acute and late phase spinal cord damage are beyond the scope of this chapter.

<table>
<thead>
<tr>
<th>Level of Injury</th>
<th>Bracing Options</th>
</tr>
</thead>
<tbody>
<tr>
<td>High cervical</td>
<td>Halo: offers flexion, extension, and rotational control of the cervical spine; used in management of unstable injuries.</td>
</tr>
<tr>
<td>Mid cervical</td>
<td>Philadelphia collar and Miami J: indicated for stable injuries only; provide flexion and extension control.</td>
</tr>
<tr>
<td>Low cervical or high thoracic</td>
<td>Stermooccipital mandibular immobilizer (SOMI): indicated for stable cervical spine fractures or for use after cervical spine fusion. Minerva: may be used in management of unstable cervical injuries.</td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td>Thoracolumbar sacral orthosis (TLSO): provides flexion, extension, side bending, and rotational control. Cruciform anterior spinal hyperextension (CASH) and Jewett: primarily provide flexion control; often used in management of compression fractures.</td>
</tr>
</tbody>
</table>

Rehabilitation

The rehabilitation of patients with SCI should begin in the acute care setting and continue in a specialized spinal cord rehabilitation unit once medical and spinal stability are achieved. The rehabilitation approach is interdisciplinary by nature. The team commonly consists of physical therapists, occupational therapists, speech language pathologists, psychologists, case managers, rehabilitation nurses, physiatrists, and consultants.

The goals of rehabilitation are to maintain medical stability, address mobility and ADLs, educate and train the patient and his or her family, plan for discharge (including recommending home modifications, if necessary), and prescribe durable medical equipment. Functional goals for the patient depend on the motor level of injury. Key motor levels of injury and associated potential functional goals are highlighted in Table 12–4. Rehabilitation specialists use numerous techniques and devices to assist their patients in gaining functional independence. A brief survey of notable methods follows.

Table 12–4 Functional goals after spinal cord injury.
Functional electrical stimulation (FES) works with intact LMNs and peripheral nerves to stimulate paralyzed muscles. FES can be used to stimulate and pace the diaphragm, to aid in erection and ejaculation, and to improve overall strength and cardiovascular fitness. FES is used in conjunction with other therapeutic modalities.

Surgical reconstruction of the upper extremity in tetraplegia by means of tendon transfer, tenodesis, contracture release, and other techniques can offer some functional restoration of the limb. These procedures alter the mechanics of the extremity, allowing better participation in activities such as self-care, transfers, and wheelchair propulsion.

Outside of the surgical realm, assistive devices can greatly enhance patients’ ability to perform ADLs. There are several categories of such assistive devices, including those that aid in feeding, dressing, hygiene and grooming, and communication. For example, a universal cuff with a utensil holder allows patients with elbow flexion control the ability to self-feed. Tenodesis orthoses facilitate prehension in individuals with wrist extension control. Items such as toothbrushes and writing tools may be built up to allow for better grasp.

Electronic assistive devices allow for greater independence for patients with high-level SCIs. There are four basic categories of such devices: mobility, communication, manipulation, and environmental control. Examples of such devices include the power wheelchair with sip-and-puff technology, which
allows independence with wheelchair mobility using the mouth as a control unit, and speech recognition software, which enables individuals to control computerized devices by means of the voice. Environmental control units (ECUs) afford some level of independence with everyday activities, such as controlling a light switch, television, and computer. Often these are controlled via a joystick or voice recognition system, depending on the individual’s abilities. By increasing the individual’s relative independence, ECUs decrease the need for constant attendant care, provide a sense of empowerment, and improve quality of life.

Robot-assisted therapy and locomotor training are emerging therapy options to retrain upper and lower limbs after central insult in individuals with incomplete motor injuries. The premise is to perform goal-directed movements in order to improve strength, coordination, and proprioception. Robot-assisted gait therapy has been widely used since the 1990s and relieves therapists of the demanding activities involved in manual locomotor training. Various devices with different functionality exist. One such device, the Lokomat, is a driven gait orthosis and body weight support system that allows patients to walk on a motorized treadmill via a preprogrammed gait pattern. Another option, a powered exoskeleton device, is worn by the patient and provides at least some activation energy for limb advancement.

In all cases, the rehabilitation process of an individual with an SCI is interdisciplinary, comprehensive, and lifelong. As new treatment advances are made, patients are presented with more opportunities to increase their independence and have a better quality of life.


COMPLICATIONS
Cardiac & Circulatory

A. Hemodynamic and Vascular Changes

Acute SCI resulting in a neurologic level of injury (NLI) at T6 and above may cause sympathetic denervation, leading to reduction of blood pressure and systemic vascular resistance. The abrupt loss of sympathetic stimulation causes arterial dilation, decreased venous return to the heart, and subsequent hypotension. This phenomenon can occur at any time within the first several weeks after the initial injury. The hypotension is treated with aggressive intravenous fluids and vasopressors. There is no clear consensus on an ideal target mean arterial pressure; however, mean arterial pressure of 85–90 mm Hg is generally considered a reasonable goal. Abrupt decrease of sympathetic output also can cause severe bradycardia. This may be exacerbated by vagal stimulation caused by endotracheal suctioning. Often the bradycardia resolves within 6–8 weeks after injury; however, it can progress to complete cardiac arrest and sometimes necessitates cardiac pacing. Anticholinergic medications are typically first-line treatments for symptomatic bradycardia following SCI. The bradycardia coupled with hypotension may significantly reduce cardiac output, precipitating a second SCI due to hypoperfusion and infarction of the spinal cord, particularly in watershed areas.

B. Orthostatic Hypotension

As the term implies, orthostatic hypotension is a reduction in systolic blood pressure by 20 mm Hg or diastolic blood pressure by 10 mm Hg that occurs when moving from a supine to an upright position. This blood pressure change, by definition, must occur within 3 minutes of a postural change. In patients with SCI, orthostatic hypotension often is asymptomatic; however, it may cause dizziness, blurred vision, fatigue, or lightheadedness. The phenomenon is caused by two factors. In the first, excessive venous pooling secondary to reduction in lower extremity muscle tone and impaired sympathetic innervation results in decreased venous return to the heart, with a lowered cardiac output. In the second, injury to the spinal cord causes a change in efficacy of the baroreceptors of the heart. Typically, a drop in cardiac volume would prompt the baroreceptors to trigger increased sympathetic tone. With SCI above the level of T6, however, the sympathetic response is blunted. To compound matters, parasympathetic innervation of the heart remains intact, causing unbalanced parasympathetic stimulation.

Orthostatic hypotension often improves over the course of several weeks to
months after the injury. Treatment includes behavioral changes, such as moving more slowly when altering positions and shoulder shrugging, and physical modalities, such as abdominal binders and compressive leg stockings to help minimize venous pooling. Patients can also be trained in reflexive compensation to position changes, using a tilt table. Medical approaches that are used try to increase plasma volume and blood return to the heart. Salt tablets are often used as a first-line agent to treat the hypotension. Midodrine is a sympathomimetic $\alpha_1$ agonist that causes vasoconstriction and an increase in peripheral vascular resistance, improving symptoms of hypotension by increasing venous return to the heart. Typical doses range from 2.5 to 10 mg, two to three times daily. Timing of medication should be adjusted to minimize hypertension while the patient is supine and maximize its therapeutic benefits while upright. Other vasopressors (eg, ephedrine, pseudoephedrine, and phenylpropanolamine) have been used because of their sympathomimetic effect, which increases blood return to the heart. Fludrocortisone has mineralocorticoid effects that cause extension of the plasma volume by increasing renal sodium absorption and water retention; typical doses are 0.1–0.2 mg daily.

**C. Autonomic Dysreflexia**

Autonomic dysreflexia typically occurs in individuals with spinal cord lesions at T6 and above but has been reported at levels as caudal as T10. It is defined as an increase in systolic blood pressure of 40 mm Hg above the patient’s baseline; or a diastolic blood pressure rise of 20 mm Hg above baseline; or a systolic blood pressure of 150 mm Hg, if the baseline is not known, with an associated altered vasomotor tone. This potentially life-threatening medical emergency is further described in [Chapter 36](#).

The condition results from unchecked sympathetic tone and most commonly causes pounding headache, facial flushing, and nasal congestion. Additional findings include sweating, piloerection, and skin blanching or flushing above the level of injury; blurred vision; anxiety; and diaphoresis. Common causes are constricting clothing, bladder distension or infection, and fecal impaction. However, any irritant below the level of injury can trigger autonomic dysreflexia, including intraabdominal pathology, ingrown toenails, and induction of labor. Consequences of autonomic dysreflexia include intracranial hemorrhage, seizure, and death. In more than 90% of incidents, the condition is caused by bladder distention; the next leading cause is bowel distention.

Treatment involves concurrent diagnosis and blood pressure control. If no obvious reversible cause of the autonomic dysreflexia is found, further
investigation is warranted, including imaging of the abdomen and retroperitoneal spaces. Specifics of these and other treatment steps are detailed in Chapter 36. Individuals and their families must be educated to recognize the signs and symptoms of autonomic dysreflexia and its immediate treatment. Individuals can also benefit from prescribed medications that can be taken on an as-needed basis if they have regular episodes of dysreflexia. A medical identification bracelet or wallet card that describes the condition to emergency personnel who may be involved in the individual’s treatment is also helpful. Although patients with autonomic dysreflexia classically present with headaches, silent autonomic dysreflexia (without symptoms) is possible. Sometimes patients are able to identify unique symptoms that develop when they are dysreflexic.

D. Cardiovascular Disease

In the chronic phase of SCI, increased cardiovascular disease is noted, with risk of increased low-density lipoprotein cholesterol, decreased high-density lipoprotein cholesterol, and decreased cardiac conditioning. These effects are similar to the changes seen in sedentary able-bodied individuals and are likely related to decreased physical activity. Often patients report an obese abdominal pannus that is incongruent with their general weight. If the abdominal musculature is paralyzed, it will fail to resist the weight of the abdominal organs, which will push out, mimicking a paunch. Treatment is cosmetic and entails abdominal binders or elasticized clothing.

Treatment of cardiac and circulatory complications often centers on early identification of symptoms, patient education, and physical modalities (including positioning and medical intervention). The earlier discussion outlines specific treatment strategies for bradycardia and orthostatic hypotension. In the chronically spinal cord–injured patient, safe activity and a prudent diet should be encouraged while monitoring laboratory markers of metabolism, such as lipid panels and glucose testing.


Respiratory

Most deaths after SCI result from diseases of the pulmonary system; 72% of these are due to pneumonia, which is the leading cause of death at all time periods postinjury and most prevalent in patients with tetraplegia. In the acute phase of SCI, respiratory problems are the most common complications. These problems include atelectasis, bronchospasm, and pneumonia. Individuals with acute SCI from C1 to C4 experience respiratory complications at a rate of about 84%. Those with a high NLI are likely to require mechanical ventilation acutely. In the acute and chronic phases of SCI, affected individuals experience bronchial hyperreactivity due to impaired sympathetic innervation to the lungs. Sleep-disordered breathing is seen in up to 60% of people with tetraplegia. Smoking and obesity have been associated with worsening pulmonary function tests in the SCI population as in the general population.

Immediately following an SCI, vital capacity, expiratory capacity, and total lung capacity are significantly decreased. Vital capacity may decrease as much as 30% in a C5–6 injury. Over the first year postinjury, volumes partially improve, with an increase in inspiratory and expiratory flow rates and a decrease in functional residual capacity. Improvement is attributed to development of spasticity of intercostal and abdominal musculature as well as resolution of inflammation and edema. After the first year, changes are more gradual.

Individuals with paraplegia and tetraplegia alike have weak expiration and cough, leading to difficulty in clearing secretions. Individuals with an NLI above C5 experience decreased inspiration resulting from loss of innervation to the diaphragm. Individuals with an NLI below C5 have incompletely innervated intercostal muscles, resulting in a decreased ability to take a deep breath. At all levels of injury patients experience impaired secretion clearance because of a weak cough owing to abdominal muscle weakness.

Strategies to minimize respiratory complications in individuals with SCI include prevention, early recognition of compromise, and aggressive treatment of atelectasis and pneumonia. Pneumococcal influenza immunization and secretion mobilization techniques are used as preventive measures against
Pneumonia.

Secretion mobilization strategies include deep breathing and coughing exercises, as well as assisted cough using the “quad cough,” a modified Heimlich-like maneuver. High-frequency percussive ventilation, such as a chest vest or a “respiratory bed,” also aids in secretion management. A mechanical insufflation–exsufflation (MIE) device, also known as a “coughalator,” is a noninvasive modality that simulates a cough. It inflates the lungs at a preset pressure or volume and then rapidly decompresses them at a similar preset setting. This device has at least two advantages over traditional suctioning. First, its use results in less trauma to tissue compared with catheter insertion. Second, it is more likely to clear both lungs, whereas catheter suctioning has been shown preferentially to clear the right lung. Contraindications to MIE therapy are recent trauma to the lungs, recent pneumothorax, and history of recurrent pneumothorax. Despite concerns relating to trauma from this device, damage is rare when used at appropriate pressures (eg, ± 40 mm Hg).

Another way to prevent atelectasis and pneumonia in ventilator-dependent patients is through use of high tidal volume ventilation. Guidelines in common use advocate tidal volumes as high as 20 mL/kg of ideal body weight, compared with traditional intensive care unit protocols, which advocate 8–10 mL/kg, and the adult respiratory distress syndrome (ARDS) protocols, which recommend 6–8 mL/kg of ideal body weight. The advantage of high tidal volume ventilation is that it is more likely to expand peripheral alveoli. When these alveoli are not sufficiently expanded, atelectasis often ensues, predisposing patients to mucus plugging, pleural effusions, and decreased production of surfactants. Rate of respiration for patients on a ventilator must often be decreased to maintain consistent ventilation rates. “Dead space” may be added to the ventilator circuit to compensate for lower PCO₂ values, which can cause respiratory alkalosis. Positive end-expiratory pressure has not been shown to remedy atelectasis. Most community-ventilated patients have intrinsic lung disease and thus may not have the elasticity to tolerate high tidal volumes. In the SCI patient without underlying pulmonary pathology or trauma to the lungs, the inability to breathe results from neurologic damage, not an intrinsic defect in the pulmonary tissue.

An additional factor in preventing pneumonia in the SCI patient is early recognition of dysphagia and use of appropriate aspiration precautions. Pneumonia may manifest in atypical fashion in patients with SCI. Consequently, a low threshold of suspicion for pneumonia must be maintained for SCI patients presenting with altered respiratory status. There should also be a low threshold for inpatient admission of patients who develop community-acquired
pneumonia. Good oral hygiene is thought to be protective of pneumonia, particularly in ventilator-dependent patients.

Electrical stimulation may be useful in maximizing the ventilatory ability of patients while maximizing independence and reducing the risk of pneumonia. Two means of using functional stimulation to this end are diaphragmatic pacing and pacing of the phrenic nerve. For both methods, an intact phrenic nerve is required. This can be confirmed through electrodiagnostic evaluation of the phrenic nerve and diaphragm. A pacing device is surgically implanted directly onto the diaphragm near the points where the phrenic nerve enters the muscle. Leads to the pacer are tunneled subcutaneously through the skin and connected to an external stimulating device. For phrenic nerve pacing, a pacer is implanted onto the phrenic nerve itself through either a cervical or a thoracic approach. The device is totally contained within the body and is triggered by a radiofrequency receiver positioned topically above the pacer, usually over the anterior chest wall. With either device, the diaphragm generally has to be reconditioned over time as the patient is gradually weaned from the ventilator.

Individuals with SCI have a higher incidence and prevalence of sleep apnea compared with the general population and are at high risk of both central and obstructive sleep apnea. Although there is a correlation with the anatomic level of the injury, no clear correlation has been found with the severity or completeness of the injury. However, obesity is correlated with sleep apnea in both the general population and the SCI population. Sleeping in the supine position may also be associated with sleep apnea. Individuals with SCI are more likely to sleep in a supine position and move around less during the night. Long-term compliance with the use of continuous positive airway pressure in individuals with SCI is similar to that in the general population, with the exception of those having decreased hand dexterity (correlated with decreased use).


Vascular

SCI results in increased venous stasis, hypercoagulability, and intimal injury. In combination, this is known as Virchow’s triad and predisposes individuals to the formation of deep vein thrombosis (DVT). Stasis after SCI is caused by peripheral venous dilatation and decreased blood flow from impaired sympathetic output and reduced muscle tone. Blood stasis contributes to endothelial damage. Hypercoagulability after SCI is related to alterations in the coagulation cascade in the setting of neurologic injury. There is an increase in collagen-induced platelet aggregation in SCI patients as compared with healthy adults. The prevalence of DVT in individuals after acute SCI has ranged in studies from about 10% to 100%, with data from the model centers showing that about 9.8% develop DVT during acute inpatient rehabilitation and that the incidence of venous thromboembolism is as high as 47–100% in the first year.

Signs and symptoms that may indicate DVT include swelling of the calf, pitting edema, dilated superficial veins, fever, and lower limb erythema; however, tenderness of the calf may not be present in patients with an insensate limb due to SCI. Side-to-side discrepancies in calf circumference may also be useful in diagnosis of this complication. DVT should be in the differential diagnosis of any fever workup. Doppler ultrasound is widely accepted as a safe, efficient, economic, and reliable means of diagnosing DVT. The d-dimer, a degradation product of cross-linked fibrin clots, has a high sensitivity but low specificity in the diagnosis of DVTs. The gold standard for diagnosis is contrast venography; however, this is rarely performed as it is invasive, painful, and
expensive.

The development of DVT can be significantly reduced with appropriate prophylaxis; thus thromboprophylaxis should be initiated as soon as clinically possible. General discussion of DVT prophylaxis and treatment was included earlier this book, in Chapters 5 (Immobility) and 10 (Pharmacotherapy). The Consortium for Spinal Cord Medicine has developed DVT prophylaxis guidelines for patients with SCI. These recommendations are divided into three categories: motor complete, motor incomplete, and motor complete with other risk factors. For individuals with motor incomplete SCI, compression stockings or compression boots with unfractionated heparin (5000 units three times daily) is recommended. For individuals with motor complete SCI, compression stockings or compression boots and unfractionated heparin or low-molecular-weight heparin (LMWH) is recommended. For individuals with motor complete SCI with other risk factors (eg, lower limb fracture, previous thrombosis, history of cancer, heart failure, obesity, or age > 70 years), compression stockings or compression boots and unfractionated heparin or LMWH are recommended and an inferior vena cava filter should be considered.

The American College of Chest Physicians has also published guidelines regarding DVT prophylaxis. The guidelines do not differentiate between motor incomplete and complete SCI. They recommend prophylaxis with LMWH or vitamin K antagonist, and do not support the solitary use of low-dose unfractionated heparin, graded compression stockings, or inferior vena cava filter for prophylaxis.

Pulmonary embolism refers to embolization of a thrombus to the lungs, potentially occluding blood flow. According to the SCI model system database, pulmonary embolism accounts for 10% of overall mortality in the first year post-traumatic SCI and is the leading cause of sudden death. Placement of inferior vena caval filters is often performed as prophylaxis; however, there is lack of consistent evidence that they protect against pulmonary embolism and they may actually increase the risk of thrombus formation. An additional complication of inferior vena caval filters is their propensity to migrate in individuals with SCI. It has been suggested that use of assisted coughing techniques, such as the “quad cough,” may precipitate the migration of these filters.

Pulmonary embolism is difficult to diagnose based on signs and symptoms and must be included in the differential diagnosis whenever a patient has a change in respiratory status or develops chest pain or fevers of unknown origin. Arterial blood gases are often ordered as part of a pulmonary embolism workup although they are of relatively little value in confirming a diagnosis. A negative
D-dimer test can be helpful in excluding pulmonary embolism but is not diagnostic. Definitive diagnosis is made by spiral CT or pulmonary angiography, the former being the more common imaging technique as it is considered safer and is more readily available. Pulmonary embolism can be suggested by a ventilation/perfusion scan (V/Q scan), the results of which are reported in terms of probability and are not diagnostic. Treatment of a pulmonary embolism is full anticoagulation with either vitamin K agonist, unfractionated heparin dosed to an appropriate activated partial thromboplastin time (aPTT) level, or therapeutic dose of LMWH. Novel new oral anticoagulants increasingly are being used to treat thromboembolic disease. They have the convenience of being orally administered and do not require laboratory monitoring. A potential drawback is that there is no accepted means of reversing their effect in patients with active bleeding or overdose.

Duration of DVT prophylaxis and treatment is controversial. It has been recommended that individuals with motor complete SCI without additional risk factors for DVT undergo prophylaxis for 8 weeks with LMWH. For those with motor complete SCI and additional risk factors, 12 weeks of prophylaxis with LMWH is recommended. Individuals with motor incomplete SCI should receive prophylaxis for up to 8 weeks. An inferior vena cava filter can be inserted for pulmonary embolism prophylaxis if anticoagulation is contraindicated. Treatment of DVT and pulmonary embolism is typically continued for at least 6 months. Left untreated, DVTs have been found to rethrombose at a rate of 29–47%. With treatment, the rethrombosis rate decreases to 4.7–30.3%.

Post-thrombotic or post-phlebitis syndrome, characterized by lower extremity edema and pain, is caused by destruction of the venous valves of the lower extremity, resulting in venous reflux and hypertension. Signs and symptoms of post-thrombotic syndrome also include dull ache, cramping, itchiness, venous ectasia, stasis changes, hyperpigmentation, and lipodermatosclerosis. Affected individuals are at risk for cellulitis and venous ulcers. In addition, the added weight of the affected limb due to retained fluid may make transfers and lower limb management more difficult for the individual and caregivers. Among patients with a history of DVT, 27–47% will develop post-thrombotic syndrome within the first 2 years of DVT onset, which can be associated with a decrease in work ability and an increase in life stress. Post-thrombotic syndrome is generally treated by the use of graded compression stockings (30–40 mm Hg) for 2 years after DVT development to reduce untoward effects. Intermittent compression extremity pumps can be used in severe cases.
Gastrointestinal

A. Swallowing Function

Determination of swallowing ability after SCI is important for two reasons: (1) difficulty with swallowing may lead to a diminished nutritional status, which can compound the catabolic state post-SCI; and (2) individuals with SCI are susceptible to pulmonary complications, and dysphagia may precipitate aspiration pneumonia. Studies have shown an incidence of dysphagia in tetraplegic patients in the range of 17% to 41%.

The swallowing process can be divided into three components: oral phase, pharyngeal phase, and esophageal phase. Dysphagia (impaired swallowing) can be related to altered function of any of these stages. Clinical signs may include coughing after eating, particularly after drinking thin liquids. Dysphagia is often first suspected at the bedside and can be investigated with a video fluoroscopic swallowing study (VFSS, often referred to as a modified barium swallow test) or fiberoptic endoscopic examination of swallowing (FEES). VFSS involves consuming solids and liquids marked with a barium contrast under fluoroscopic observation such that the path of ingested material can be visualized should it penetrate the sinuses or become aspirated. FEES entails direct fiberoptic visualization of the oropharynx to the level of the vocal cords. It does not allow for visualization of a fluid bolus once past the vocal folds.

Risk factors for dysphagia include an anterior approach to cervical procedures, cervical soft tissue edema, and cervical hardware. Patients who
undergo tracheostomies are at increased risk of aspiration, although there is evidence that a Passy–Muir valve may decrease this risk. Individuals who have complete SCI (ASIA category A) have a greater risk of swallowing complications than those who have incomplete injuries. Cervical immobilization devices, including collars, sternococciptal mandibular immobilizer (SOMI) braces, and halo vest braces, have been shown to impair swallowing ability. Aspiration has been linked to nasogastric tubes and older age of the individual. Typically, dysphagia improves over the first several months post-SCI although the exact mechanism for this improvement is unknown.

Treatment for dysphagia may include removing underlying risk factors (eg, tracheostomies or nasogastric tubes) and working with speech language pathologists to engage the individual in exercises to strengthen pharyngeal muscles and pharyngeal coordination, as well as modify eating behaviors to lessen the risk of penetration and aspiration (ie, alternating solids and liquids or controlling volumes of food consumed in one swallow). Alternate means of nutrition and hydration should be considered early in the clinical course of patients who are suspected of not receiving adequate oral intake.

B. Nutrition and Hydration

In addition to swallowing ability, it is important to assess nutritional and hydration status after SCI. Particularly if there is a concomitant traumatic brain injury, individuals may demonstrate functional or delayed swallow without aspiration but have poor arousal, decreased desire for food, or impaired speed of swallowing, resulting in inadequate oral intake. In such cases, alternate means of providing nutrition and hydration should be considered.

C. Gallstones

SCI has been associated with an increased incidence of cholelithiasis. Although studies have consistently demonstrated greater rates of asymptomatic gallstones, rates of acute cholecystitis are not significantly increased in this population. Gallstone formation has been associated with NLI above T10, possibly related to decreased motility of the gallbladder and bile ducts in the absence of sympathetic stimulation. Despite impaired sensation post-SCI, individuals with acute cholecystitis typically present with traditional symptoms of right upper quadrant abdominal pain with radiation to the right shoulder.

D. Bowel Dysfunction
SCI leads to neurogenic bowel dysfunction. Dysfunction may be due to UMN injury, LMN injury, or a mixed injury. UMN bowel dysfunction is caused by injury of the spinal cord above the conus medullaris and is associated with impaired peristalsis and overactivity of the external anal sphincter. UMN bowel dysfunction can lead to severe constipation. LMN neurogenic bowel dysfunction is related to injury at or below the conus medullaris and involves a flaccid sphincter with stool incontinence. Injury to the spinal cord at the level of the distal thoracic spine through the proximal lumbar spine may result in a mixed UMN/LMN pattern of bowel injury with a combination of variable characteristics of both UMN injury and LMN injury.

Neurogenic bowel can result in significant physical and psychological distress, especially in the presence of unplanned defecation. Thus, establishing a bowel program in which stool evacuation can be timed and performed in a controlled fashion is of the utmost importance. A bowel program is devised for individuals after a careful history and physical examination with consideration of type of motor neuron injury and involves specific timing of oral medications and rectal stimulation or disimpaction. There are also surgical options for individuals with severe bowel dysfunction. (See Chapter 7 for extended discussion of neurogenic bowel and its management.)


Vallès M, Mearin F: Pathophysiology of bowel dysfunction in patients with motor incomplete spinal cord injury: Comparison with patients with motor
Genitourinary

The lower urinary tract includes the bladder, internal sphincter, external sphincter, and urethra. The bladder, lined with smooth muscle (the detrusor muscle) consists of the trigone (base), body, and neck. Normally the bladder functions to store urine when not micturating and to empty the body of urine when appropriate. Sympathetic innervation promotes the storage of urine, whereas parasympathetic stimulation causes detrusor contraction. Under normal circumstances the bladder is able to accommodate increasing volumes with only small changes in the intravesicular pressure. Normal voiding requires coordination of multiple components of the pelvis, including relaxation of the striated muscle of the urethra and pelvic floor, the detrusor muscle, and the internal and external sphincters.

SCI can cause either UMN or LMN pathologies affecting genitourinary function, often resulting in neurogenic bladder. The UMN syndrome is characterized by disruption of the descending spinal pathways, resulting in loss of cortical inhibition of sacral reflex arcs. LMN syndrome causes impaired motor output of the bladder and a decrease in detrusor contractility, resulting in a flaccid bladder. During the initial spinal shock phase of acute SCI, the bladder is usually flaccid regardless of the type and location of the injury. Bladder management post-SCI takes into account the type of injury, functional ability of the individual, and lifestyle. (For further discussion of neurogenic bladder, see Chapter 7).

Individuals with SCI are at risk of recurrent urinary tract infections. Often they must be educated about the difference between bacterial colonization and true infection. They are also at increased risk of renal and bladder calculi. Elevated voiding pressures over time can lead to hydronephrosis. Thus, individuals with a neurogenic bladder require ongoing monitoring of upper and lower urinary tract and renal function. In addition, chronic indwelling catheter use is a risk factor for squamous cell carcinoma of the bladder and necessitates annual monitoring using cystoscopy.

Decubitus ulcers are a common yet devastating complication of SCI. They result from ischemia of the skin caused by pressure or shear forces. Pressure-related ulcers occur when direct pressure to the skin over time (generally > 2 hours) is greater than capillary pressure, resulting in ischemia. Typically the ulcers occur over bony prominences, including the ischium, scapula, greater trochanters, lateral malleoli, and heels, in patients with limited mobility. Shear-related injury results from a friction force that causes occlusion of blood vessels and subsequent breakdown of the skin. The coccygeal and sacral areas are typical locations of shear-related injury. (Further discussion of pressure ulcers appears in Chapter 5.)

Decubitus ulcers are estimated to occur in one quarter to one third of individuals with SCI. Factors that place these individuals at increased risk for skin breakdown include: (1) impaired regulation of blood supply to the skin, resulting from to changes in the autonomic innervation of their blood vessels; (2) impaired mobility and paralysis, which makes them less likely to shift weight and move spontaneously; and (3) lack of sensation, which prevents them from feeling the otherwise painful effects of ischemia to the skin.

Additional factors influencing the risk of developing decubitus ulcers include a higher, more complete level and severity of injury; increased age (in particular, age > 50 years); duration of injury; social support; level of education; and medical comorbidities.

Prevention of the skin breakdown is of the utmost importance and is accomplished by minimizing pressure or shearing forces to the skin. In addition, maximizing nutritional status, particularly maintaining adequate protein levels and adequate hydration, may help mitigate decubitus ulcers. Individuals with SCI and their caregivers should be educated about the importance of weight shifting every 2 hours while in bed and every 30 minutes while seated, and encouraged to adhere to a regular routine. Individuals who cannot weight shift independently may require support services or adaptive equipment to aid in weight shifting (ie, power wheelchair with a tilt-in space function). Education
should include training on regular inspection of the skin and recognition of early signs of skin compromise. Early signs of skin breakdown include redness and induration of the skin.

Treatment of a wound begins with investigation of the cause and mitigation of the provocative factors. Sources of increased pressure may include wheelchairs, cushions, and any other surfaces that the individual may rest upon, and seating position and posture should both be examined. Reversing any underlying conditions, such as nutritional deficits or comorbid conditions, is important in treatment of wounds. The wound itself may be treated with a variety of dressings that are beyond the scope of this chapter. The overall goal of wound care is to create the best environment possible for skin regrowth. This includes ensuring the correct moisture levels of the skin as well as minimizing bacterial growth in the wound. Additional treatments may include electrical stimulation to the wounds, or vacuum-assisted closure (VAC) dressings, which apply a negative pressure gradient to the wound and are used to promote blood flow while removing excess drainage from the wound. There is no clearly defined role for hyperbaric oxygen in the treatment of these wounds. Ultimately, wounds may need to be surgically debrided and closed.

Prior to closure of the wound, bacterial growth must be addressed. The individual may require prolonged antibiotic therapy. Wounds may not heal for various reasons, such as an underlying infection or osteomyelitis, Marjolin’s ulcer (a malignancy that may form in chronic wounds or fistula formation), persistent pressure to the region, or malnutrition. Care must be used when placing any padding around the wound as it may decrease pressure over the actual wound while increasing pressure to surrounding areas, thereby increasing the area of compromised skin. In addition, any packing to the wounds must be placed with caution as increased pressure caused by the packing within the wound may prevent adequate blood flow, thereby inhibiting healing.

A. Osteopenia and Fracture

Osteopenia is an often overlooked complication of SCI. Bone wasting occurs most rapidly in the first 3–6 months and can persist for 1–2 years in trabecular bone and even longer in cortical bone. Bone loss is most significant at the distal femur, with the proximal tibia being the second most commonly affected site. Typically, bone above the level of injury is spared. Bone wasting may ultimately result in loss of 40–50% of bone density. The bone loss is attributed to increased osteoclastic activity and bone reabsorption, while osteoblast activity is decreased as is new bone formation.

A more complete injury and less ambulation are risk factors for a more severe disease process. Increased tone is thought to be protective, although research has been limited to small studies with mixed results. Associated with osteopenia is hypercalcemia, particularly in the acute phase of the SCI. The hypercalcemia predisposes individuals to renal and bladder calculi. Also associated with the acute SCI is vitamin D deficiency, which has been found to be prevalent in greater than 90% of individuals with SCI. Osteopenia is a risk factor for fracture, to which SCI patients are more susceptible than the general population. No direct correlation has been found between degree of osteopenia and fracture risk. Fractures most often occur at the knee; the distal femur is the most common site followed by the proximal tibia. The gold standard for diagnosis of osteopenia and osteoporosis is the dual-energy X-ray absorptiometry (DEXA) scan, although currently there is no means of directly correlating DEXA scan results with absolute risk of fracture.

Signs and symptoms of acute fracture in individuals with SCI include increased tone, autonomic dysreflexia, low-grade fevers, fatigue, localized redness, warmth over the fracture site, and bony abnormalities to palpation. Complications of fracture may include DVT, skin breakdown, and impaired mobility and self-care, and may necessitate rehabilitation and functional retraining. Most fractures heal with conservative treatment; however, surgical intervention may be necessary to control rotational deformities.

Treatment interventions have been categorized as pharmacologic and nonpharmacologic. Nonpharmacologic options revolve around the concept of Wolf’s law in that weight bearing over bone will increase its strength and density. Thus, treatment includes maintaining elevated tone, standing, electrical stimulation, and applying ultrasound vibration to bones. There is limited evidence to support efficacy of these modalities, although several studies have demonstrated improvement when these therapies are performed intensely, entailing sessions on most days of the week for at least 1–2 years.
Pharmacologic interventions include repletion of vitamin D (25-hydroxyvitamin) and calcium, if levels are low. Results of calcitonin and bisphosphonate use have been disappointing when studied as treatment for spinal cord–related osteopenia and osteoporosis; however, administration may have a protective effect if given early in the injury, before bones thin and reach steady state.

The patient should be educated about both the risk of fracture and atypical signs and symptoms associated with it, which differ from those in a neurologically intact individual. It is important to decrease fracture risk by protecting against trauma and falls and limiting activities that may result in injury.

There are no clear guidelines or accepted standard of practice regarding monitoring of bone density in patients with SCI. Some experts have advocated for routine DEXA scans, but it is unclear in the absence of ongoing fractures how the information obtained from these scans would change an individual’s treatment. In a patient with fractures, secondary causes of bone wasting such as hypogonadism should be excluded if clinically suspected.

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**Smith EM: Treatments for osteoporosis in people with a disability. PM R 2011;3:143–152.**

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**B. Heterotopic Ossification**

Heterotopic ossification (described earlier in Chapter 5) refers to the formation of bone in nonskeletal tissue, usually between the muscle and joint capsule. The bone growth may bridge the joint entirely. When this bone formation occurs in
muscle alone, it is referred to as myositis ossificans. It is estimated that 20% of individuals with SCI develop heterotopic ossification below the level of injury. The normal bony growth generally occurs in neurologically impaired limbs and is more prevalent in spastic than flaccid limbs. The hip is the most common site, followed by the knee, elbow, and shoulder. In the hip, heterotopic ossification most commonly develops at the anteromedial aspect. Individuals with complete SCI are at higher risk for its development. There is no correlation between NLI and heterotopic ossification, although lumbar injuries tend to produce less bone growth than cervical- and thoracic-level injuries. Soft tissue damage may be a risk factor, as well.

The underlying cause of heterotopic ossification is unclear. Prostaglandins, which normally regulate osteoblasts and osteoclast function, may play a role in its development. Other contributing factors include hypercalcemia, tissue hypoxia, changes in sympathetic nerve activity, prolonged immobilization, and disequilibrium of parathyroid hormone and calcitonin levels. The disease is more common in males than females, and there may be a genetic predisposition. Additionally, presence of a tracheostomy, being a smoker, and having fewer comorbidities are associated with a higher risk of developing this condition.

Heterotopic ossification generally manifests with nonspecific symptoms such as fever and local redness, warmth, swelling, or decreased ROM in the affected joint. Differential diagnosis includes DVT, fracture, septic arthritis, impending pressure ulcer, and cellulitis. Alkaline phosphatase is a nonspecific laboratory marker that may be elevated early in the disease course. An elevated serum creatine phosphokinase (CPK) level may be associated with the early inflammatory phase and severity of heterotopic ossification and can be helpful in treatment planning and evaluation of response to treatment. C-reactive protein is one of the acute phase reactants that may be elevated in the acute stage and can also be used to plan and track response to treatment. Calcium levels are not diagnostic of heterotopic ossification. The gold standard for diagnosis is a triple-phase bone scan. Both MRI and ultrasound imaging modalities have been used to identify heterotopic ossification early in its course. There is generally a 3-week delay in plain radiographic findings. Typically, heterotopic ossification occurs in the first year postinjury, and the bone grows for 6 months to 1 year until it matures. Heterotopic ossification may cause difficulties in positioning of the affected individual because of resulting limitations in the ROM of affected joints. Complications may include pain, pressure ulcer, spasticity, and impingement of peripheral nerves or local vasculature.

Currently, there is no accepted method for prevention of heterotopic
ossification following SCI. Thus, early detection and treatment is paramount. The goal of early treatment is to prevent further heterotopic growth and maximize preservation of joint ROM. Treatment options include gentle ROM, bisphosphonates (eg, disodium etidronate), nonsteroidal antiinflammatory drugs (eg, indomethacin), radiation therapy, and surgical excision.

Typically, disodium etidronate is prescribed for a total of 6 months upon diagnosis of heterotopic ossification. Treatment may consist of high-dose administration for 3 months (20 mg/kg divided into twice-daily dosing) followed by low-dose therapy for 3 months (10 mg/kg per day divided into twice-daily dosing) if initial laboratory findings are normal. When initial laboratory values are elevated, high-dose disodium etidronate is typically administered for the entire 6-month course. Bisphosphonates are used in the treatment of heterotopic ossification to prevent additional formation of bone. It is unclear whether disodium etidronate or other pharmacologic agents actually retard the progression of heterotopic ossification or merely delay the mineralization that occurs during this progression such that mineralization recurs once the treatment course is stopped.

When initial C-reactive protein and CPK levels are elevated, adding indomethacin until levels normalize (if not contraindicated) can help to reduce inflammation and pain. Indomethacin has also been shown to minimize bone formation during the growth phase. Of note, the drug has been shown to reduce formation of heterotopic ossification as a preventative measure, but it is not routinely used because of its potential to interfere with postsurgical bone healing. Unlike indomethacin, disodium etidronate has receive approval from the U.S. Food and Drug Administration (FDA) for this indication.

Radiation therapy may be used in patients with severe bony growth to inhibit heterotopic bone growth. Radiation disrupts the differentiation process of mesenchymal cells into osteoblasts.

During treatment it is important to maintain ROM as much as possible through physical therapy or a home exercise program. If heterotopic ossification significantly impedes function or interferes with the patient’s quality of life, surgical excision may be considered. However, surgery should not be performed until bone maturation has been demonstrated on bone scan (approximately 1 year), as the highly vascular nature of developing bone growth increases the risk for intraoperative bleeding.

CT scans with three-dimensional reconstruction are helpful in planning surgical removal of heterotopic ossification. Recurrence after surgery is thought to be 17–58%, although recurrent disease often is not as severe as the initial


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**Neurologic**

Following SCI, neurologic complications such as post-traumatic myelomalacia, syringomyelia, or tethered cord syndrome may ensue.

**A. Post-traumatic Myelomalacia**

In post-traumatic myelomalacia, scar tissue forms in the area of injury to the spinal cord. This tissue consists of micro cysts, reactive astrocytosis, and thickening of the pia and arachnoid tissues. Myelomalacia may occur at any time from months to decades after the initial injury and is noted in 0.3–2.2% of patients with chronic SCI.

**B. Syringomyelia**

Also known as a syrinx, syringomyelia refers to fluid-filled cystic lesions that develop in the grey matter of the SCI. Syringomyelia may be idiopathic or due to previous injury in the spinal cord or congenital malformation (ie, Chiari malformations). In patients with congenital malformation, symptoms of
Syringomyelia may not manifest until periods of growth (e.g., during adolescence or early adulthood). Syringomyelia develops in about 20–30% of individuals with traumatic SCI and enlarges in about 10–20% of these individuals. Risk factors for a syrinx include a complete injury and kyphotic deformation of the spine. Neurologic decline occurs in about 3–8% of patients with a syrinx and may appear months to decades after the traumatic SCI. Although the exact etiology is not well understood, it is believed that syringomyelia represents a disruption of cerebrospinal fluid flow in the subarachnoid space, possibly as the result of scar formation, ischemia due to pressure or traction, or cytotoxic chemical release during the injury. The syrinx can extend in either a rostral or caudal direction and may be worsened by increases in intraabdominal and thoracic pressures. Affected individuals should be cautioned against using the Valsalva maneuver or excessive intraabdominal pressure when voiding (bowel or bladder).

Presenting symptoms of a syrinx include changes in neuropathic pain, new sensory deficits, new motor deficits, worsening spasticity, bowel or bladder changes, and autonomic dysfunction. The diagnostic test of choice is an MRI scan. Strictly speaking, contrast is not required to make the diagnosis of a syrinx; however, contrast may be helpful for ruling out other conditions in the list of differential diagnoses. Electrodiagnostic evaluation may also play a role in limiting the list of differential diagnoses or ruling out peripheral causes of new weakness or numbness.

Treatment options for patients with syringomyelia are suboptimal and center on symptomatic treatment. If symptoms are the cause of neurologic decline, surgery may be warranted. Surgical techniques have included shunts, cordectomies, and duraplasties; however, all have met with limited success.

C. Tethered Cord Syndrome

In this syndrome, the nerve roots or the cauda equina become fixed to the meninges. This may be have congenital causes or be secondary to trauma, inflammation, or surgery. In acquired tethered cord syndrome, it is often scar tissue that adheres the nerve roots and cauda equina to the meninges. Often patients can be monitored without specific treatment; however, they may develop changes in pain, sensory deficits, weakness, and altered bowel and bladder patterns. Symptoms may also be positional or be worsened by flexion and extension.

Treatment for tethered cord syndrome involves bracing and surgical release of the tethered cord. However, risks of surgery include potential of additional
trauma to the area, with increased risk of recurrence of the tethered cord syndrome. In pediatric patients, tethered cord syndrome presents a unique problem as symptoms may be exacerbated during periods of growth in childhood. If this occurs, the child may require surgical treatment of the tethered cord.


Endocrine

Individuals with SCI are at increased risk of metabolic syndrome, including coronary artery disease, dyslipidemia, and hyperglycemia with insulin resistance. The increased risk has been attributed to the relative decrease in physical activity, decreased lean muscle mass, and greater percentage of adipose tissue compared with uninjured counterparts. SCI patients who views themselves as overweight are more likely to have a lower level of subjective well-being when rating themselves. Interestingly, several studies have suggested that the risk for diabetes mellitus is lower in those with tetraplegia than paraplegia even when accounting for various levels of obesity.

There is evidence that individuals with SCI who are active can reduce their
risk for heart disease. Increased physical activity can result in improved lipid profiles and less atherosclerotic disease. It is reasonable to suggest that SCI individuals be monitored more closely for dyslipidemia and glucose intolerance compared with their uninjured counterparts.


Reproductive & Sexual

Sexual organs rely on the autonomic nervous system for proper functioning. Sexual function in males involves the ability to have an erection and the ability to ejaculate. Erections can be achieved by psychogenic or reflexic means. In men with UMN lesions, the ability to have reflexic erections is generally preserved; a male will have an erection in response to tactile stimulation of his genitalia. In those with LMN lesions, there may be an absence of reflexic erections. Psychogenic erections are generally absent in patients with UMN lesions but may be preserved in those with LMN lesions and may be mediated by a sympathetic reflex arc that exits the spinal cord at the lower thoracic levels. Erections are maintained by parasympathetic stimulation causing blood flow from the internal pudendal artery to the penis. The role of sympathetic
stimulation in maintaining erection is less clear. In men coordination of ejaculation is under sympathetic nervous system control. The sympathetic nervous system triggers peristalsis in the smooth muscle of ampulla, seminal vesicles, and prostate, as well as contractions of the bulbocavernosus, ischiocavernosus, and pelvic musculature to shunt semen to the urethral meatus.

Sexual function in females involves the ability to have engorgement of the genitalia, vaginal lubrication, and peristalsis of the reproductive organs. A mechanism similar to that of the male erection causes engorgement and swelling of the labia and clitoris as well as lubrication of the vagina. Vaginal lubrication and engorgement can occur in response to direct tactile stimulation or psychogenic stimulation. The sympathetic nervous system is responsible for rhythmic contractions of the uterus, fallopian tubes, paraurethra glands, and pelvic floor musculature.

The incidence of male impotence after SCI approximates 75%, and women with SCI are less likely to be able to achieve orgasm compared with their uninjured counterparts.

Treatment of sexual dysfunction in individuals with SCI begins with counseling that addresses both the psychological implications of the change in sexuality and the logistics of intimacy, given the limitations imposed by SCI. Issues to be discussed with the individual and his or her partner include suggestions to empty one’s bladder before sexual intercourse and options for unique positioning during intercourse.

First-line medical treatment for erectile dysfunction in men includes phosphodiesterase 5 (PDE-5) inhibitors such as sildenafil. The role of these medications in the treatment of women with sexual dysfunction is not well defined; however, use of these agents is thought to improve vaginal engorgement and lubrication. Should oral medications fail, vasoactive substances to promote blood flow can be injected directly into the penis. Such medications include papaverine, phentolamine, and prostaglandin E₁. Other options include vacuum suction devices that create negative pressure to promote blood flow to the penis, which is then maintained by a constriction band placed at the base of the penis. More invasive techniques involve implantation of a prosthesis into the penis. This carries risks of skin breakdown and infection. Patients with SCI commonly use a nitroglycerin-based antihypertensive agent to manage autonomic dysreflexia and should be educated to avoid nitroglycerin products if they have recently used PDE-5 medications as the combination can cause severe hypotension.

Fertility is significantly diminished in male SCI patients owing to post-SCI
impaired ejaculation and poor sperm quality. Although sperm of affected individuals tend to have decreased motility, they can be harvested through electroejaculation and vibratory stimulation techniques for use in intrauterine insemination or in vitro fertilization. Fertility is generally unaffected in women with SCI. Women may be amenorrheic in the months after SCI; thereafter, menstruation often occurs irregularly before becoming regular. Women should be cautioned that despite being amenorrheic or irregular, the possibility of pregnancy exists.

The most significant complication of pregnancy in a woman with SCI is autonomic dysreflexia (described earlier in this chapter, and in Chapter 36). The risk is greatest during labor but is present throughout pregnancy, particularly in women with injuries at T6 or above. Hypertension associated with autonomic dysreflexia must be distinguished from that of preeclampsia based on clinical findings. Autonomic dysreflexia is associated with bradycardia and occurs in an episodic fashion, with improvement of blood pressure when the noxious stimulus is removed. During labor, the risk is mitigated by use of epidural anesthetics. Any women at risk for autonomic dysreflexia during labor should receive epidural anesthesia despite potential lack of sensation with delivery.

The pregnant SCI patient must also be vigilant for other complications. She is at increased risk of bladder distention, which is a common cause of autonomic dysreflexia. Because 35% of women with SCI have urinary tract infections during pregnancy, use of antibiotic suppression is recommended. At a minimum, frequent urinary cultures should be obtained. Pregnancy can lead to constipation, and the bowel routine should be monitored for any changes. The growing size and weight of the pregnant woman increase the risk of decubitus ulcers. Thus, extra vigilance must be maintained in ensuring frequent skin checks and weight shifts. Equipment such as wheelchairs may need to be refitted or cushions modified to accommodate increased weight and girth. Pregnant women with SCI must also be vigilant for signs or symptoms of pregnancy-related infections, such as intraamniotic infection, which may present uncharacteristically in those with SCI. A gravid uterus may compress venous return, causing hypotension; this may be further exacerbated by the underlying abnormal regulation of vascular tone. Women should maintain good hydration and alter their positioning if symptoms of orthostasis develop. Impairment of pulmonary function can occur, particularly in the late stages of pregnancy. Those whose vital capacity is reduced because of high-level injuries may require ventilatory support, particularly during labor. Women with SCI are also at risk of unrecognized contractions and a painless unexpected delivery. They must be monitored with
serial cervical examinations after 28 weeks and educated on the signs and symptoms of labor in the setting of SCI. Often, home uterine activity monitoring is recommended.

The risk of congenital malformations or intrauterine fetal death in women with SCI who were injured before becoming pregnant is similar to that in the general obstetric population. Those with congenital SCI should undergo genetic counseling if there is a hereditary basis for their condition.

Condoms have been found to be safe for use in individuals with SCI; however, oral contraceptives containing estrogen are controversial because of the risk for thrombosis. Women should be counseled about smoking cessation, especially if taking an oral contraceptive pill. Barrier contraceptives, such as sponges, cervical caps, and diaphragms, may be difficult for some individuals with SCI to use, as physical limitations and impaired sensation may affect their ability to apply these devices and monitor positioning.


**Pain**

The prevalence of pain after SCI is estimated to be between 25% and 96%, with the wide range of variability likely due to varied study methodology and definitions of pain. Pain after SCI can have many origins but is often related to the local effects of trauma and surgery, injury to peripheral and central nervous systems, and muscle imbalances, requiring novel use of the intact muscles.
A. Classification

Many attempts have been made to develop a classification scheme for pain in SCI in order to standardize reporting of pain and foster more accurate documentation. The current best-known classification scheme is the International Spinal Cord Injury Pain Classification (ISCIP). As shown in Figure 12–3, the ISCIP classification scheme divides pain into several tiers and subtiers. There are four tier 1 types, labeled (1) nociceptive pain, (2) neuropathic pain, (3) other pain, and (4) unknown pain. Each tier 1 pain type is divided into a tier 2 pain subtype, which in turn is divided into a tier 3 primary pain source or pathology.
<table>
<thead>
<tr>
<th>Tier 1: Pain type</th>
<th>Tier 2: Pain subtype</th>
<th>Tier 3: Primary pain source and/or pathology (write or type in)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nociceptive pain</td>
<td>Musculoskeletal pain</td>
<td>e.g., glenohumeral arthritis, lateral epicondylitis, comminuted femur fracture, quadratus lumborum muscle spasm</td>
</tr>
<tr>
<td>Visceral pain</td>
<td></td>
<td>e.g., myocardial infraction, abdominal pain due to bowel impaction, cholecystitis</td>
</tr>
<tr>
<td>Other nociceptive pain</td>
<td></td>
<td>e.g., autonomic dysreflexia headache, migraine headache, surgical skin incision</td>
</tr>
<tr>
<td>Neuropathic pain</td>
<td>At level SCI Pain</td>
<td>e.g., spinal cord compression, nerve root compression, cauda equina compression</td>
</tr>
<tr>
<td>Below level SCI pain</td>
<td></td>
<td>e.g., spinal cord ischemia, spinal cord compression</td>
</tr>
<tr>
<td>Other neuropathic pain</td>
<td></td>
<td>e.g., carpal tunnel syndrome, trigeminal neuralgia, diabetic polyneuropathy</td>
</tr>
<tr>
<td>Other pain</td>
<td></td>
<td>e.g., fibromyalgia, Complex Regional Pain Syndrome type I, interstitial cystitis, irritable bowel syndrome</td>
</tr>
<tr>
<td>Unknown pain</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Figure 12–3** International Spinal Cord Injury Pain Classification (ISCIP).

B. Assessment

Similar to pain assessment in the non-SCI population, consideration should first be given to causes of pain that are reversible or can be treated. In the SCI population, possible causes of pain include peripheral neuropathies such as ulnar neuropathy (producing paresthesias in the arms and hands), angina pectoris or cholecystitis (manifesting as chest tightness or pain), rotator cuff pathology (with pain in the shoulders or upper arms), and heterotopic ossification or orchitis (possibly manifesting with pain in the hip or groin region). Often, however, pain is poorly localized below the NLI. When taking a history, it is important to ascertain changes in patterns, timing, and progression of pain. Physical examination findings are altered due to impaired sensation. For example, an SCI patient with appendicitis may not have tenderness over McBurney’s point, but instead present with general malaise, nausea, fever, or abdominal discomfort. Often laboratory testing and imaging is required to rule out a diagnoses. Fluctuations in pain may be the result of underlying stressors. Frequently pain worsens with secondary illnesses, such as urinary tract infection or upper respiratory infections. Pain may also fluctuate with psychological stress and weather changes or cold temperatures.

C. Management

When treating the SCI patient with pain, one must be aware of the multifactorial nature of the pain. The pattern of the pain should be established with the patient. Secondary causes of pain and underlying triggers should be ruled out. Treatment can incorporate oral medications, physical modalities and psychotherapy, and treatment goals should be established with the patient.

1. Nonpharmacologic measures—Treatment of pain resulting from musculoskeletal causes such as overuse of the shoulders, myofascial pain syndrome, or rotator cuff injury should include evaluation of seating system and postures, as modifications of body mechanics alone may improve underlying pain.

Once secondary causes of pain and underlying triggers are ruled out, the primary goal of treatment is to improve level of function. Patients should be encouraged to use medications as a tool to maximize functionality and not to
arbitrarily lower pain scale ratings. Evidence of the efficacy of nonpharmacologic modalities in the treatment of neuropathic pain is inconsistent. Several small studies reported that use of cognitive behavioral therapy or self-hypnosis may not lower actual pain levels, but these methods may help patients cope and achieve a more functional state with pain. There is evidence showing the efficacy of transcranial electrical stimulation and transcranial magnetic stimulation in the reduction of neuropathic pain, and conflicting evidence of the efficacy of transcutaneous electrical stimulation for pain. Acupuncture has a long history of use in pain management; however, evidence of its efficacy remains inconclusive for neuropathic pain related to SCI. The technique has, however, been proven to be safe and is not associated with autonomic dysreflexia.

2. Pharmacologic measures—Several pharmacologic agents are available for the treatment of pain in the SCI population. NSAIDs have limited efficacy in neuropathic pain but may be useful as general analgesia and in patients with myalgia. First-line nonopioid treatment of neuropathic pain generally includes antiseizure medications and antidepressants; however, opioid medications are often required as well. Of the antiseizure medications, pregabalin is currently the only medication with a primary FDA indication for neuropathic pain related to SCI. Other antiseizure medications (eg, gabapentin, topiramate, and lamotrigine) have been used off label with good efficacy for neuropathic pain. It is understood that gabapentin and pregabalin bind to voltage-gated calcium channels and inhibit neurotransmitter release. They have shown good efficacy versus placebo in neuropathic pain in multiple settings. The most common side effects include dizziness and sedation. Starting at a low dose and gradually titrating up can mitigate these side effects. Both gabapentin and pregabalin are renally excreted, and dosing must be adjusted accordingly. The maximal dose in patients with normal renal function is 3600 mg/day for gabapentin and 600 mg/day for pregabalin.

Other first-line medication options for neuropathic pain are antidepressants that inhibit both norepinephrine and serotonin reuptake. These include the tricyclic antidepressants as well as selective serotonin and norepinephrine reuptake inhibitors (SSNRIs). Both classes of medications are efficacious in neuropathic pain resulting from several causes. Tricyclic antidepressants (amitriptyline, nortriptyline, and desipramine) are typically prescribed but should be used with caution, particularly in the elderly, as all may be sedating. These drugs may have cholinergic side effects, including urinary retention, constipation, and dry mouth, as well as cardiac toxicity and QT prolongation.
Thus, baseline QT intervals should be established in the geriatric population. SSNRIs (venlafaxine and duloxetine) have the advantage of good efficacy in the treatment of depression as well as nerve-related pain. The maximal dose is 225 mg/day for venlafaxine and 60 mg/day for duloxetine. Studies evaluating use of high-dose duloxetine have shown no additional benefit over standard dosing.

Tramadol has also demonstrated efficacy in the treatment of neuropathic pain. It acts as a weak opioid receptor agonist but also inhibits reuptake of serotonin and norepinephrine and is chemically similar to a tricyclic antidepressant. Tramadol is considered to have less abuse potential than opioid analgesics and may be taken on an as-needed basis. It has the side effects of lethargy and confusion, particularly in elderly patients. It may also precipitate serotonin syndrome when used in conjunction with other medications that inhibit serotonin reuptake, and can lower seizure thresholds, which is an important consideration in the dually brain- and spinal-cord injured patient.

Opioid analgesics have efficacy in the treatment of neuropathic pain both as single agents and in conjunction with other nonopioid medications. Combining morphine and gabapentin increases the efficacy of both medications while decreasing overall side effects. Caution is advised when prescribing opioid medications as they may be addictive, may decrease respiratory drive, and can cause urinary retention and constipation.

Other medications that have been used for neuropathic pain include various antiseizure medications, among them, carbamazepine, lamotrigine, oxcarbazepine, topiramate, and valproic acid. Antidepressants such as bupropion and citalopram have also been used, though with limited efficacy. Dronabinol, a cannabinoid medication, is thought to decrease neuropathic pain although this has not been demonstrated in clinical studies. Mexiletine is an antiarrhythmic medication that has traditionally been used in the treatment of neuropathic pain; however, it carries the risk of being proarrhythmogenic and thus its current use is rare.

Localized neuropathic pain may be treated with topical analgesics such as topical lidocaine (thought to affect sodium channels of the cell membrane) and capsaicin (thought to deplete substance P in the cell). Care must be used with capsaicin as it may cause irritation, particularly if one touches the face or eyes with the medication.

Intrathecal clonidine and intrathecal opioids have demonstrated benefits in management of severe pain. Intrathecal medications such as ziconotide, which is based on a marine snail venom, are emerging treatment options for patients with intractable neuropathic pain. Onabotulinum toxin injections have shown efficacy...
in the treatment of neuropathic pain, although this approach is still considered experimental. There is evidence that the toxin, once injected, is absorbed by cells and carried to the central nervous system.


**Spasticity**

Spasticity is a velocity-dependent increase in muscle tone that is indicative of a UMN lesion. It affects about two thirds to three quarters of SCI patients. Spasticity may be beneficial to some patients, and some may learn to trigger spasticity to aid with standing and bed mobility. Spasticity has also been associated with preserved bone density and DVT prevention. However, at times spasticity may interfere with self-care, transfers, comfort, or ability to perform home exercise programs. Spasticity can be exacerbated by certain positions, particularly the supine position, as well as changes in temperature, psychologic
stress, or physiologic stress. Often an increase in spasticity is the first indication of underlying pathology such as an infection or a bowel- or bladder-related issue. Owing to the fluctuation of spasticity within a given time period, in-office diagnosis of spasticity has a limited role, and treatment is largely determined by self-reporting of symptoms by the patient.

Treatment of spasticity is focused on symptom relief. Initial treatment measures focus on managing the environment and using physical modalities such as stretching. If necessary, several pharmacologic treatment options are available in the form of patches (eg, clonidine), oral medications, and injectables. Often muscle relaxants (eg, carisoprodol, cyclobenzaprine) are confused with UMN syndrome antispasticity medications, although there is overlap in the case of benzodiazepines. Intramuscular onabotulinum toxin and nerve ablation with alcohol or phenol are other treatment options. Spasticity also can be treated with intrathecal medications such as baclofen. Chapter 6 discusses this topic in detail.


Psychiatric

The rate of depression in the SCI population is higher than that in the general population. Depression affects about 30% of people living with SCI, and rates of major depression have been estimated at 10–15%. These rates improve slightly after the first year but generally remain stable throughout the lifetime of the person with SCI. These findings correlate with life satisfaction rates, which also remain stable for this population. SCI patients have a higher rate of divorce than the general population, although marriage has been identified as a major source of support. Perceived control, resilience, sense of coherent self-worth, hope, and purpose in life are consistently associated with higher qualities of life and well-
being in people with SCI.

Factors that increase the risk of depression in individuals with SCI include a lack of autonomy, greater severity of disability, fewer years of education, less mobility, less social support, preinjury psychiatric and psychological issues (eg, substance abuse), inadequate coping mechanisms, pain, divorce, unemployment, pressure ulcers, and neurologic deficits. High levels of depression have also been associated with urinary incontinence and indwelling catheters, and a lower rate with individuals who perform intermittent catheterization for bladder management. (The reasons for this association are unclear.) Women with SCI have a greater incidence of depression than their male counterparts.

The most commonly prescribed class of medications for SCI patients with depression is the selective serotonin reuptake inhibitors (SSRIs). Although it has been suggested that SSRIs may exacerbate spasticity, there is no clear and accepted correlation.

The incidence of post-traumatic stress disorder (PTSD) in civilians with SCI is similar to that in the general civilian population. Although the rate of PTSD is relatively low, this disorder has a high correlation with major depressive disorder. When an individual presents with one of these disorders, it is recommended that the other be screened for, as well. Increased physical activity has been postulated to decrease depression in individuals with SCI.

Individuals with SCI have a high rate of suicide, estimated to be about 3–5 times higher than that of the general population. About half have suicidal thoughts. The most common means of committing suicide is by gunshot, although committing suicide by passive self-neglect is believed to be underestimated in surveys.

Risk factors for suicide include previous suicide attempts, increased degree of postinjury dependency, lower self-esteem, poor coping mechanisms, younger age, and paraplegia (versus tetraplegia). Individuals at risk may display expressions of shame, apathy, and hopelessness; have difficulty with attention and concentration; or become restless and agitated. Treatment of those at risk revolves around psychological counseling and utilization of community resources in an effort to create social infrastructure for the patient.


Bombardier CH, Fann JR, Tate DG, et al: An exploration of modifiable risk


SPECIAL MANAGEMENT CONSIDERATIONS

★ Dual Diagnosis: Concomitant Traumatic Brain & Spinal Cord Injury

Traumatic brain injury (TBI) commonly coexists with traumatic SCI; estimates of prevalence range from 25% to 70%, depending on methodologic and study considerations. These patients present a unique clinical challenge. Patients with concurrent TBI and SCI present to rehabilitation at a later stage in their treatment course, although they may have similar initial FIM scores. The dual diagnosis patient may progress more slowly in rehabilitation, requiring additional time to reach the levels of function achieved by patients with an SCI alone. The rehabilitation process for SCI requires the ability to problem solve and learn new ways to achieve daily tasks. Concurrent brain injury may pose a challenge to both the patient and the treatment team as TBI can impair cognition. Additionally, TBI may result in behavioral challenges, greater incidence of
psychopathology, and more severe neuropsychologic impairments. Subsequently, individuals with dual diagnosis may require increased nursing hours of care and have higher daily costs as well as higher costs per FIM change.

Risk factors for concomitant TBI with SCI include alcohol consumption at the time of injury, completeness of the spinal cord injury (AIS A), higher level of neurologic injury, and injuries resulting from motor vehicle crashes and falls.

Special considerations must be made when prescribing medication to patients with dual diagnosis, because centrally acting medications commonly used in the treatment of SCI may have cognitive and sedative effects that could compound the effects of the TBI. Additionally, care must be taken to avoid overstimulating patients who are recovering from TBI. When possible, performing therapies in quieter, less distracting environments can be helpful.


▶ Pediatric Spinal Cord Injury

SCI is relatively rare in children, as compared with adults. However, this life-changing event entails a complex and long-term continuum of care for the pediatric patient. The rehabilitation team must address a range of considerations, including medical complications, growth issues, and the psychological and psychosocial implications of the injury to a growing child or developing teen and his or her family. Understanding the long-term complications and providing interventions for prevention are key components in the care of these patients.

A. Epidemiology and Etiology

Although the vast majority of SCI injuries involve adults, reports have estimated
that up to 20% occur in children. One epidemiologic study from 1997–2000 estimated an incidence rate of 1.99 per 100,000 children. As in the adult population, males in their teenage years are more likely to suffer an SCI than females, at a ratio of 2:1. Sports-related injuries and acts of violence are common mechanisms of injury, with the latter notably higher among African Americans and Hispanics. There is no gender predilection among younger children, especially for those younger than 5 years of age. Motor vehicle crashes remain the leading cause of SCI, followed by falls; other causes are listed in Table 12–5. Traumatic injuries in this age group tend to involve the cervical spine in more than half the cases and result in complete tetraplegia. Nontraumatic medical causes are likely to result in paraplegia and incomplete injuries.

**Table 12–5** Causes of pediatric spinal cord injury and dysfunction.

<table>
<thead>
<tr>
<th>Traumatic</th>
<th>Nontraumatic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Motor vehicle crashes</td>
<td>Transverse myelitis</td>
</tr>
<tr>
<td>Gunshot wounds</td>
<td>Infection, postvaccination</td>
</tr>
<tr>
<td>Stab wounds</td>
<td>Inflammatory or autoimmune processes (eg, systemic lupus erythematosus)</td>
</tr>
<tr>
<td>Nonaccidental trauma (abuse)</td>
<td>Vascular insults</td>
</tr>
<tr>
<td>Sports injuries (American football, ice hockey, wrestling, diving, skiing, snowboarding, rugby, cheerleading)</td>
<td>Tumor</td>
</tr>
<tr>
<td>Falls</td>
<td>Spinal cord anomalies</td>
</tr>
<tr>
<td>Birth or delivery complications</td>
<td>Spinal and cardiac surgery complications</td>
</tr>
<tr>
<td></td>
<td>Vertebral abnormalities causing spinal cord compression (eg, atlantoaxial subluxation in Down syndrome, skeletal dysplasia)</td>
</tr>
<tr>
<td></td>
<td>Metabolic or toxin-related (eg, chemotherapy-related myelopathy)</td>
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</tbody>
</table>
Infants and children up to 8 years of age tend to have high cervical spine injuries. This is due to their relatively larger head, with the fulcrum of movement being higher at the C2–3 (infants) and C3–4 levels injury compared with the C5–6 level in adults. Shoulder and lap belt injuries commonly occur in children weighing less than 60 pounds when the lap belt rises above the pelvic brim. This results in flexion and distraction forces in the midlumbar spine and may cause abdominal wall and intraabdominal injuries and SCI. A Chance fracture, occurring most commonly in the midlumbar region in children, is often referred to as the “seatbelt fracture” because anterior flexion over a lap belt can cause compression of the anterior vertebral elements and distraction of posterior elements, resulting in injury to the spinal cord. Prevention of lap-belt injury can be accomplished by placing children aged 4–8 years and weighing less than 40 pounds in approved belt-positioning booster seats.

More than 50% of children with SCI have SCIWORA (described earlier in this chapter). The high incidence is thought to reflect the elasticity of the pediatric spine, which causes a stretch injury to the spinal cord and surrounding soft tissues without resulting in a bony fracture. Soft tissue damage can be detected on MRI scans but not on plain radiographs.

B. Classification

When performing a neurologic assessment of a child, it is important to take into consideration the child’s age and maturity level, which affect the ability to follow motor commands and respond to sensory testing. A creative and integrated play approach can help examiners obtain an accurate assessment of NLI.

C. Acute Management and Medical Complications

Similar to management in the adult population, airway, breathing, circulation, spinal stabilization, fluids, and pain control are mainstays in the acute setting following traumatic SCI. Spinal stabilization may be treated surgically with the use of fixation devices, or nonsurgically with an appropriate spinal orthosis. Initially, spinal shock with flaccid paralysis, sensory impairment, and bladder and bowel dysfunction is encountered, and this may last a few days to weeks. Supportive treatment and prevention of secondary pulmonary, cardiovascular, gastrointestinal, skin, and urinary complications are initiated in the intensive care unit.

In traumatic injuries, a concomitant brain injury should be suspected in a patient with an altered level of consciousness. Other common associated injuries
include fractures, intraabdominal and pelvic injuries, and pulmonary contusions or hemorrhage. The degree of trauma impact, as well as a younger age, correlates with the severity, presence of associated injuries, and overall mortality. Rehabilitation starts in the intensive care unit, and early mobilization is encouraged as soon as possible and when medically stable. Splints and positioning devices, along with an appropriate wearing schedule, can help prevent joint contractures and skin breakdown.

1. Respiratory support—A child with a high cervical injury and weak or absent diaphragmatic function requires ventilatory support, along with monitoring of oxygen saturation, blood gases, and end-tidal carbon dioxide. Even children who do not require a ventilator should be monitored for overnight oxygen desaturation and hypercarbia, given their restrictive respiratory dysfunction. Pulmonary toilette and postural drainage are important measures that help clear secretions. A cough-assist device may be needed for those with a thoracic-level injury and weak intercostal and abdominal muscles. In addition to respiratory muscle training in assisted cough techniques, pneumococcal and yearly influenza vaccines are recommended.

2. Nutritional requirements—Weight loss is usually seen in the acute stage of illness or injury. This may be worsened by the child’s refusal to eat or lack of appetite. The importance of adequate nutrition in the healing and rehabilitation process cannot be overemphasized. Initially, enteral feedings through a nasogastric or gastric tube may be required to provide adequate caloric intake. As the child begins to eat, care must be taken to avoid shifting to a pattern of overeating and excessive weight gain.

3. Venous thrombosis—Although venous thromboembolism is rare in the prepubertal child, it does occur. The risk is highest in the first few weeks after the injury. Prophylactic anticoagulation therapy is not routinely warranted except in cases associated with multiple risk factors, such as presence of a central venous line, hypercoagulable state, poor perfusion, and prior history of thrombosis. However, in the pubertal child, recommendations for prophylaxis and treatment follow those adult protocols. A child with a DVT usually presents with a swollen, warm extremity, with or without fever. Diagnosis can be confirmed with Doppler ultrasound. Upon confirmation, the affected limb should be immobilized and anticoagulation therapy begun immediately. Mobilization can be resumed once therapeutic levels or adequate heparinization have been achieved. For more detailed discussion, see Chapter 5.
4. **Autonomic dysfunction**—Autonomic dysreflexia (described earlier in this chapter and in Chapter 36) occurs in about 40% of pediatric patients with SCI, most commonly as a result of a distended bladder. If treatment of hypertension is required, a rapid-acting and short-duration medication such as nifedipine can be used; however, nitroglycerin paste is preferred. The latter is easier to administer in children (applied topically to the skin above the level of injury), and has the advantage of being easily stopped (ie, wiped away) once symptoms have resolved. Temperature regulation can also be impaired, placing children at risk for hyperthermia or hypothermia. Thus, it is important to educate both the child and caregiver about the need to wear clothing appropriate for the environmental temperature.

5. **Neurogenic bladder**—Clean intermittent catheterization (CIC) is taught to children 5–7 years of age and to those who are developmentally and functionally capable. Parents or caregivers are also taught the technique. Although, the child with tetraplegia may not be able to physically perform CIC, he or she should be taught how to direct others in this task. The goal of bladder management is to gain urinary continence, promote independence, minimize occurrence of infection, and protect the kidneys.

6. **Neurogenic bowel**—Similar to affected adults, children and adolescents with neurogenic bowel may not feel the urge to defecate or may be unable to perform this function. Management of neurogenic bowel in a child with SCI is similar to that of an adult. Bowel programs are generally initiated at 2–4 years of age. Timed toileting can be initiated by placing the child on a commode or toilet at the same time every day. The goal is to have adequate and regular bowel emptying, promote continence and independence, and prevent complications such as severe constipation, fecal impaction, and bowel obstruction.

7. **Hypercalcemia**—Hypercalcemia is primarily a complication in adolescent males. It usually develops during the first 12 weeks after injury as a result of rapid bone resorption associated with immobilization. Signs and symptoms include anorexia, vomiting, abdominal pain, lethargy, polydipsia, polyuria, and dehydration. Serum calcium levels are usually greater than 12 mg/dL when symptoms manifest. Management includes intravenous hydration with normal saline, early mobilization, limitation of calcium intake, and administration of intravenous pamidronate.

8. **Skin breakdown**—Pressure ulcers are common among pediatric SCI patients.
In both acute and chronic SCI, pressure ulcers cause a huge burden of care and may lead to further complications, such as sepsis and osteomyelitis. Prevention is key, through education of the child and family about proper positioning, pressure relief, avoiding moisture, use of pressure-relieving cushions or seating system, and at least daily inspection of the skin. Good hygiene and proper nutrition are crucial elements of skin care in children.

9. Orthopedic and musculoskeletal complications—Children who sustain an SCI before puberty are at an increased risk for developing scoliosis and hip subluxation or dislocation. Thoracolumbar spinal orthoses (TLSO; see Chapter 28) may be used to retard progression when curves are 20–40 degrees. They may also help with truncal support in the upright position. An appropriately fitting wheelchair and seating device is crucial to assist in prevention of pressure ulcers and pain associated with spinal deformity and pelvic obliquity. In patients with severe SCI, a molded seat and back may be needed to accommodate for the deformity. Spinal fusion is considered for rapidly progressive curves, curves greater than 40 degrees, and for problematic pain and seating issues.

Contractures often develop at the hips, knees, ankles, elbows, and wrists, and worsen after a growth spurt. Conservative management includes positioning, ROM, bracing, and spasticity management.

Pathologic fractures are a common complication given the risk for osteoporosis in patients with SCI. A high index of suspicion should be maintained when a child presents with a warm, swollen limb, with or without fever. Pain may not be reported, given the lack of sensory function.

The incidence of heterotopic ossification in pediatric SCI patients ranges from 3% to 18%, with the most common site being the area around the hip joint.

10. Pain—Early in the course of postspinal care, mechanical pain can be secondary to the inciting trauma or surgical intervention and is usually managed with analgesics, antiinflammatory agents, and narcotics. However, a more debilitating pain (dysesthiasia or neuropathic-type pain) may occur, characterized by burning, numbness, and tingling or hyperesthesia. It commonly follows a dermatomal distribution caudal to the zone of injury, but this is often difficult to accurately assess in a very young child. Physical modalities (eg, transcutaneous electrical stimulation, physical therapy, and hydrotherapy) can be used to help relieve pain. Pharmacologic measures include gabapentin (initiated at 5 mg/kg per dose, three times daily) and amitriptyline (started at 10–25 mg nightly for children younger than 10 years of age).
11. **Latex allergy**—Children with SCI are at risk for developing a latex allergy, most likely due to frequent exposure to latex-containing medical products early in life. Up to 18% of children display evidence of such an allergy, which is characterized by localized or generalized urticaria, wheezing, angioedema, or anaphylaxis. An allergy to latex should be suspected when a child develops unexplained intraoperative allergic reactions. Owing to the severity of the allergic reaction, children with SCI or with a history of myelomeningocele should be cared for in a latex-free environment as a prophylactic measure. If latex allergy is diagnosed, the patient should wear a medical alert tag and carry autoinjectable epinephrine in case of an anaphylactic reaction.

**D. Rehabilitation**

The sudden change in a child’s functional and physical capability after an acute event resulting in SCI can be overwhelming to both the patient and the family. The interdisciplinary approach used in an inpatient rehabilitation setting can help with this transition process. Goals include maintaining good health, preventing secondary complications, and promoting maximal and age-appropriate functional independence and must be appropriate to the patient’s age, level of injury, and neurologic recovery.

1. **Physical therapy**—Physical therapy focuses on bed mobility and transfer techniques, sitting, standing ability and tolerance, and eventual ambulation. A child or teen with a C6–7 injury can do level transfers, with or without a sliding board. A medical stroller is a more appropriate seating and mobility device for a child younger than 2 years of age. Most children can transition to a manual wheelchair between 18 and 26 months of age, and children as young as 24 months can safely operate a powered mobility device. For the tetraplegic patient with a high-level injury, a power wheelchair with a sip-and-puff, head array, or chin control may be needed for independent driving if hand controls are not amenable. Additional wheelchair specifications may include tilt, recline, ventilator tray, and standing features. “Standers” are commonly used to help facilitate upright posture and weight-bearing tolerance; however, caution should be used, given the risk for orthostasis in these patients. This risk can be minimized or prevented through the use of support hose, wrapping of the lower limbs, and abdominal binders to maintain blood pressure. Standing and walking may be achieved with or without an assistive device or orthosis. It is important to distinguish ambulatory status and recognize energy consumption in relation to daily activity. Paraplegic patients with low thoracic or high lumbar injuries may
be able to achieve limited community-level ambulation with an appropriate lower limb orthosis and assistive device. Those with injury at the L3 level and below can achieve independent community-level ambulation with an ankle–foot or supramalleolar orthosis. (For additional discussion of assistive devices, see Chapter 41.)

2. Occupational therapy—Through occupational therapy, the child or teen works to relearn ADLs, such as feeding, dressing, bathing, hygiene, and transfers; writing; computer skills; and leisure activities. At around age 5, a child can be independent in the majority of self-care activities with supervision. Children with high cervical injuries may not be expected to perform their own self-care but should be taught how to direct caregivers. Various types of adaptive equipment can be incorporated to facilitate independent function, such as a mobile arm support for the forearm that also aids shoulder and elbow movements in a patient with C4–5 tetraplegia. A dynamic hand orthosis makes use of the tenodesis effect to facilitate grasping in those with injury at the C6 level.

3. Discharge considerations—Architectural and environmental modifications and equipment may be needed in the home, school, or community; examples include installing grab bars, lift systems, and ramps. Transitioning back to school and into the community is a stressful event for both the patient and the family. Good communication with designated school liaisons prior to discharge regarding the patient’s medical status, medications, precautions, activity limitations, and necessary accommodations helps to facilitate this process. Referrals for recreation therapy, camps, adaptive sports, support groups, and resources in the community can be of help to the patient and family physically and, more importantly, psychosocially.


PROGNOSIS

Medical advances have given those who have sustained traumatic SCI a better quality of life and overall improved outcomes. Consequently, determining prognosis after traumatic SCI has become all the more imperative. The most important predictor of prognosis is an accurate physical examination as outlined by the ISNSCI (described earlier in this chapter). The initial NLI, initial motor strength, and whether the injury is neurologically complete or incomplete are the most important factors when determining recovery in the first year after traumatic SCI. Of these factors, completeness of the injury is the most predictive of long-term recovery and prognosis. Findings on MRI that correlate with poor functional recovery are spinal cord hemorrhage, long segments of spinal cord edema, and high cervical locations of SCI.

It is advisable to wait to prognosticate until the patient is out of spinal shock. Generally, an ASIA examination is performed at 72 hours postinjury and again at 1 month to predict short-term and long-term prognosis, respectively. Early return of deep tendon reflexes bodes well for better recovery. Poor prognostic factors include advanced age at time of injury, vertebral displacement of greater than 30%, and, most notably, completeness of injury. Most motor recovery has been found to occur within the first 6 months after injury. Recovery does continue to a lesser extent during the second year after injury, but at a much slower rate.

Patients with complete SCI can anticipate, on average, recovery of one root level below the NLI. However, functional lower extremity movement is often not attainable if the patient remains completely without motor and sensory innervation for more than 1 month postinjury. As well, it has been documented that conversion from complete to incomplete status more than 1 month after injury has little effect on the ultimate prognosis for recovery. On the other hand, more than 50% of tetraplegic patients with incomplete injury may become ambulatory.

Thorough patient examination and development of an expected prognosis not only assists in the construct of a comprehensive rehabilitation program but also helps to guide patient and family expectations. As acute care management of these patients continues to improve, it has become increasingly necessary for rehabilitation professionals to understand the course of SCI and to be able to offer reasonable expectations for the future.


Traumatic Brain Injury

Thomas K. Watanabe, MD
Michael H. Marino, MD

ESSENTIALS OF DIAGNOSIS

► Traumatic brain injury (TBI) may be mild, moderate, or severe.
► In mild TBI, duration of loss of consciousness (LOC) is <30 minutes, duration of post-traumatic amnesia (PTA) is <1 hour, and glasgow Coma scale (GCS) score is 13–15.
► In moderate TBI, duration of LOC is <24 hours, duration of PTA is between 1 and 7 days, and GCS score is 9–12.
► In severe TBI, duration of LOC is <24 hours, duration of PTA is <7 days, and GCS score is ≤ 8.
► Computed tomography (CT) scan of the head is the initial imaging modality of choice.

GENERAL CONSIDERATIONS

Traumatic brain injury (TBI) is a major cause of morbidity in the United States and the world. A wide range of deficits can be seen, including physical, cognitive, and behavioral problems, and difficulties encountered in one area may hinder the patient’s progress in other areas. TBI is characterized by a change in
neurologic functioning and can be classified as mild, moderate, or severe. Because some neuromedical problems seen after TBI are rather specific to the brain injury population, the rehabilitation physician must be familiar with diagnosing and treating these complications. TBI rehabilitation encompasses the spectrum of care from the intensive care unit (ICU) through acute inpatient rehabilitation, and outpatient care that extends to family, community, vocational, and avocational activities. Appropriate rehabilitation improves functional outcome, reduces complications, and enhances the quality of life for individuals with TBI and those in their social network.

Epidemiology & Demographics

The Centers for Disease Control and Prevention (CDC) estimates that there are 1.7 million TBIs annually in the United States. Of these, approximately 75% are considered to be mild TBI, or concussions. Despite this fact, brain injury is thought to be a contributing factor in nearly one third of all injury-related deaths. Of the 1.7 million patients with TBIs annually, 80% are treated and released from an emergency department; however, 275,000 are hospitalized. The CDC estimates that 52,000 people die each year from injuries related to head trauma. Currently there is no estimate for the number of brain injuries that occur in which people never present to an emergency department or seek medical care. The CDC estimates the total of direct and indirect costs related to TBI to be $60 billion annually.

Falls are the leading cause of all TBIs and are responsible for 35% of head injuries in the general population. Falls are an even greater cause of TBI in children and the elderly, accounting for 50% of TBIs in children younger than 14 years and for 61% of TBIs in adults aged 65-plus. Motor vehicle accidents and traffic-related accidents cause 17% of head injuries. Struck by or against events (eg, colliding with a moving or stationary object) cause another 16% of head injuries. Assaults account for 10% of head injuries. Brain injury is more common in males than females across all ages. Additionally, alcohol intoxication is involved in 12% of hospitalizations due to head injury and in nearly 50% of all head injuries. Blast injury is a leading cause of TBI in military personnel. Blast injuries involve force applied to the brain and body from explosion-generated pressure waves and also from flying debris.

American Association of Neurological Surgeons: Traumatic brain injury.
PATHOGENESIS

Changes in the brain due to trauma are the result of a complex interplay between the direct force and the resultant cascade of neurochemical and hemodynamic changes. Primary injuries are those caused by the direct force of the trauma, while secondary injuries are a result of the cascade of intracerebral changes. Primary injuries include focal pathology such as contusions, hematomas (subdural, epidural, subarachnoid, intraparenchymal), lacerations, and direct injury from penetrating or missile injuries. Primary injury can also be diffuse in nature as is the case in diffuse axonal injury.

Contusions result from bruising of the surface (cortex) of the brain. Areas of the cortex that are most susceptible to contusion injury are those that are in close proximity to irregular bony areas of the skull vault. These areas include the anterior temporal lobes and the inferior frontal lobes.

Hematomas are hemorrhages in and around the brain. They are defined by their location and by the type of injured blood vessels that result in the hemorrhage. Epidural hematomas are located between the dura mater and the skull. They result from injury to arteries and classically involve the middle meningeal artery but can also occur due to middle meningeal vein injury. Skull fractures are present in over 90% of cases. Epidural hematomas appear convex on neuroimaging.

Subdural hematomas are located in the subdural space between the dura and arachnoid. They result from injury to bridging veins. Risk factors for subdural hematoma include older age and alcoholism. These hematomas have a concave appearance on neuroimaging.
Subarachnoid hematomas are located in the space between the arachnoid and pia mater. They are also the result of injury to microvessels. Traumatic subarachnoid hematoma is frequently found near areas of contusion. Blood products in the subarachnoid space increase the risk for communicating hydrocephalus due to obstruction of arachnoid villi, and also increase the risk of vasospasm.

Intraparenchymal hematomas are located within the brain tissue itself and are also referred to as intracerebral hemorrhages. They are most commonly found in association with cortical contusions in the frontal and temporal lobes. Intraparenchymal hematomas are the result of severe cortical contusions, lacerations, or penetrating injury.

Primary injuries also can be diffuse in nature, as is the case with diffuse axonal injury. Diffuse axonal injury is caused by sheer and tensile strains on axons as a result of angular acceleration of the brain. The axons of the cerebral white matter, corpus callosum, and brainstem are particularly vulnerable to this type of injury. Diffuse axonal injury is a complex entity in which mechanical stretch of an axon leads to changes in axon permeability, resulting in disruption of axon transport and impaired cellular metabolism. This can cause immediate or delayed cell death.

Although much attention is, understandably, focused on the primary injury, it would be foolish to overlook the importance of secondary injury. Secondary injuries significantly contribute to morbidity and mortality. Cerebral edema has been described as a hallmark finding in severe TBI. Cerebral edema is a result of brain swelling from vasogenic edema and vascular dysregulation, which leads to increased cerebral blood volume. Cerebral edema plays a prominent role in the development of elevated intracranial pressure (ICP). Elevated ICP, or intracranial hypertension, compromises cerebral perfusion pressures, putting the patient at risk for ischemic injury. Additionally, intracranial hypertension can produce brain herniation syndromes. Focal injuries such as hematomas also contribute to secondary injury via mass effect and edema, resulting in compression of blood vessels, alterations of cerebral perfusion pressures, and brain herniation syndromes. Areas at greatest risk of secondary ischemic injury post-TBI include the hippocampus, basal ganglia, and cerebellum. Excitotoxicity is another form of secondary injury in which damaged neurons release large amounts of excitatory neurotransmitters, such as glutamate, which build up to toxic levels causing cell death. Apoptosis has also been identified as a form of secondary injury as damaged cells undergo programmed cell death.

Focal injuries may present clinically with a loss of function directly
corresponding to the region of the brain that is injured. However, it is important to remember that both focal and diffuse injuries can exist at the same time in the same patient. In addition, the presence of secondary injuries can alter the clinical picture. For these reasons, diffuse injury and secondary injury may mask focal deficits that would be anticipated from focal pathology.


CLINICAL FINDINGS

The most important aspect in diagnosing a brain injury is a clinical history of trauma. Brain injury results from a traumatic force and may occur after direct trauma to the head, or trauma to the body that is then transmitted to the head. Neurologic functioning is objectively measured using the Glasgow Coma Scale (GCS; Table 13–1). Duration of loss of consciousness (LOC) and duration of post-traumatic amnesia (PTA) are key features of this evaluation.

Table 13–1 Glasgow Coma Scale.ª
Symptoms & Signs

A rehabilitation professional performing an assessment of a patient with TBI will likely be confronted by a wide spectrum of clinical findings. Disorders of consciousness, weakness, impaired cognition, aphasia, apraxia, incoordination, visuospatial dysfunction, impaired balance, and spasticity are some of the most common clinical findings in brain-injured patients. Owing to the complex interplay among primary injury, secondary injury, focal injury, and diffuse injury, these deficits are less likely to exist in isolation or in discrete patterns, as might be the case in stroke syndromes. Instead, they are more likely to exist in complex combinations of deficits. Some of these clinical findings are outlined in greater detail below.

A. Disorders of Consciousness

Severe TBI can result in disorders of consciousness, the defining behavioral features of which are listed in Table 13–2. Disorders of consciousness are manifested by profound impact on alertness, cognition, and behavior. The most severe of these disorders is coma. In the comatose state there is an absence of sleep–wake cycles, the eyes remain closed, and the patient is unable to be aroused. The loss of arousal includes the absence of spontaneous arousal and stimulus-induced arousal. Coma is typically a self-limited condition that resolves within 2–4 weeks.
Some cases of coma progress into the other disorders of consciousness, vegetative state and minimally conscious state. The vegetative state typically represents a transitional state between coma and minimal consciousness or consciousness. The Multi-Society Task Force has stated that all of the following conditions must be present to diagnose vegetative state: (1) intermittent wakefulness manifested by sleep–wake cycles (ie, times during which the eyes are open); (2) no evidence of sustained reproducible, purposeful, or voluntary behavioral responses to visual, auditory, tactile, or noxious stimuli; and (3) no evidence of language comprehension or expression.

The terms *persistent vegetative state* and *permanent vegetative state* are frequently the source of great confusion and misuse by the general public and also within the medical community. The terms are often incorrectly applied or inappropriately used interchangeably. The Multi-Society Task Force determined that the term *persistent* should apply when the vegetative state is present for 4 or more weeks. The term *permanent vegetative state* is used to denote a time point after which the probability for recovery of consciousness is poor. The Task Force determined that the vegetative state should be considered permanent when it has been present for 3 months or more after hypoxic–ischemic, metabolic, and

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### Table 13–2 Behavioral features of disorders of consciousness.

<table>
<thead>
<tr>
<th>Behavior</th>
<th>Coma</th>
<th>Vegetative</th>
<th>Minimally Conscious</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eye opening</td>
<td>None</td>
<td>Spontaneous</td>
<td>Spontaneous</td>
</tr>
<tr>
<td>Spontaneous movement</td>
<td>None</td>
<td>Reflexive/Patterned</td>
<td>Automatic/Object manipulation</td>
</tr>
<tr>
<td>Response to pain</td>
<td>None/Posturing</td>
<td>Posturing/Withdrawal</td>
<td>Localization</td>
</tr>
<tr>
<td>Visual response</td>
<td>None</td>
<td>Startle</td>
<td>Object recognition/Pursuit</td>
</tr>
<tr>
<td>Affective response</td>
<td>None</td>
<td>Random</td>
<td>Contingent</td>
</tr>
<tr>
<td>Commands</td>
<td>None</td>
<td>None</td>
<td>Inconsistent</td>
</tr>
<tr>
<td>Verbalization</td>
<td>None</td>
<td>Random vocalization</td>
<td>Intelligible words</td>
</tr>
<tr>
<td>Communication</td>
<td>None</td>
<td>None</td>
<td>Unreliable</td>
</tr>
</tbody>
</table>

congenital brain injuries, and only after at least 12 months following a TBI.

The minimally conscious state was defined by the Aspen Neurobehavioral Working Group as “a condition of severely altered consciousness in that there is minimal but definite behavioral evidence of conscious awareness.” The diagnosis is based on the bedside examination revealing at least one of the following behaviors: (1) simple command following, (2) intelligible verbalization, (3) recognizable verbal or gestural “yes–no” responses, and (4) movements or emotional responses triggered by relevant environmental stimuli that cannot be attributed to reflexive activity.

The Working Group also identified two behaviors that signify the emergence from a minimally conscious state: (1) reliable demonstration of interactive communication, and (2) functional object use. It is important to note that for a patient to meet the criteria for emergence from a minimally conscious state, reliable communication or functional object use must be consistent, not merely reproducible.

Assessment of disorders of consciousness is best performed using a standardized neurobehavioral rating scale. A brief bedside neurologic examination has poor reliability for determining LOC. For example, 30–40% of patients who cannot speak and do not follow commands are incorrectly diagnosed as being vegetative. The use of a standardized neurobehavioral rating scale minimizes the risk of both diagnostic and prognostic error. Although several formal rating scales are available for use, the Disorders of Consciousness Task Force determined that the Coma Recovery Scale–Revised (CRS-R) is the most acceptable scale. The CRS-R is a widely used and accepted tool for diagnosing and monitoring progression of disorders of consciousness. Its six subscales each address a functional system: auditory, visual, motor, oromotor/verbal, communication, and arousal. Within each subscale, items are hierarchically arranged so that the lowest items reflect brainstem-mediated functions and the highest, cortical functions. A sample CRS-R scoring sheet is shown in Figure 13–1.
Coma Recovery Scale – Revised c2004
Record Sheet

This form should only be used in conjunction with the CRS-R Administration and Scoring Manual which defines guidelines for standardized application of the scale

<table>
<thead>
<tr>
<th>Patient:</th>
<th>Diagnosis:</th>
<th>Etiology:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date of onset:</td>
<td>Date of Examination:</td>
<td></td>
</tr>
</tbody>
</table>

| DATE | Admission | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 | 12 | 13 | 14 | 15 | 16 |
|------|-----------|---|---|---|---|---|---|---|---|---|----|----|----|----|----|----|----|

<table>
<thead>
<tr>
<th>WEEK</th>
<th></th>
</tr>
</thead>
</table>

**AUDITORY FUNCTIONS**
4 Consistent Movement to Command*
3 Reproducible Movement to Command*
2 Localization to Sound
1 Auditory Startle
0 None

**VISUAL FUNCTIONS**
5 Object Recognition*
4 Object Localization: Reaching*
3 Visual Pursuit*
2 Fixation*
1 Visual Startle
0 None

**MOTOR FUNCTIONS**
6 Functional Object Use**
5 Automatic Motor Response*
4 Object Manipulation*
3 Localization to Noxious Stimulation*
2 Flexion Withdrawal
1 Abnormal Posturing
0 None/Facetoid

**OMOTOR/VERBAL FUNCTIONS**
3 Intelligible Verbalization*
2 Vocalization/Oral Movement
1 Oral Reflexive Movement
0 None

**COMMUNICATION SCALE**
2 Functional: Accurate**
1 Non-functional: Intentional*
0 None

**AROUSAL SCALE**
3 Attention
2 Eye opening without stimulation
1 Eye opening with stimulation
0 no arousal response

**TOTAL SCORE**

Denotes emergence from MCS**
Denotes MCS*
**Figure 13–1** Coma Recovery scale–Revised (CRs-R). MCs, minimally conscious state. (Reproduced, with permission, from Hirschberg R, Giacino JT: The vegetative and minimally conscious states: Diagnosis, prognosis and treatment. Neurol Clin 2001;29:773–786.)


**B. Impaired Cognition**

A broad spectrum of cognitive impairments can be seen after TBI. Information processing and attention, general intellectual functioning, memory, spatial cognition, and executive functions are among the prime cognitive domains affected by TBI. The patient may indicate a lack of awareness of the deficits, even when these are significant. When evaluating a patient for cognitive dysfunction post-TBI, it is important to take into account any other conditions that may be affecting his or her abilities. For example, preexisting conditions such as learning disabilities or attention-deficit/hyperactivity disorder may affect cognitive abilities. Factors related to the brain injury itself, such as seizures, medication side effects, and pain, can also affect cognitive abilities. Mood disturbances such as depression have also been shown to negatively influence cognitive test scores.

Information processing involves both the speed and accuracy of responses to
external stimuli. The control of information processing is not well localized in the brain. Instead, it is controlled by broad networks, including multiple cortical and subcortical structures. Delayed information processing is manifested in TBI patients by prolonged response times. Diffuse injury disrupting cognitive networks results in the patient having to expend more mental effort and energy to perform a task that previously would have been easily accomplished. This increased energy expenditure results in both mental and physical fatigue.

Anosognosia refers to the loss of awareness of deficits. It is a condition in which the patient does not fully acknowledge that there is any neurologic dysfunction. Anosognosia has been identified as a barrier to rehabilitation, often due to the patient’s reluctance to participate in a formal rehabilitation process. It is unclear if anosognosia is a result of purely psychological denial, purely cognitive dysfunction from disruption of several cognitive domains, or is multifactorial in nature.

Overall intellectual functioning is impaired in patients with TBI. This can be measured by a variety of formal evaluations; however, the Wechsler Adult Intelligence Scale is widely used. This scale generates an intelligence quotient (IQ) score, but more importantly, allows for evaluation of a broad range of cognitive capacities ranging from digit span to language, attention, and vocabulary. Again, it is important to note that IQ scores will be heavily influenced by slow information processing, aphasia, and inattention.

Memory impairment is a hallmark feature of traumatic brain injury. Initially, impairments in memory manifest as post-traumatic amnesia (PTA). PTA is defined as the timeframe in which brain-injured patients are unable to encode or retain new information. It is manifested by confusion, agitation, perseveration, and disorientation. The duration of PTA is a powerful predictor of long-term functional outcome post-TBI (see Prognosis, below). As such, formal tools have been devised to evaluate the progression and emergence of PTA. Two such tools are the Galveston Orientation and Amnesia Test (GOAT) and the Orientation-Log (O-log). Once a patient attains a score above 75 on the GOAT or above 34 on the O-log on two consecutive evaluations, he or she is considered to have emerged from the period of PTA. After emergence from PTA, there is a great variability between individuals with respect to the severity of memory impairments. Again, it is prudent to consider the effects of pain, sleep disturbances, mood disturbances, and medication side effects on learning and memory. Although learning and memory are controlled by multiple neurologic structures and networks, the medial temporal lobe is integral to these functions. There is significant correlation between memory impairment and decreased
volumes in the hippocampi and medial temporal white matter. It remains unclear whether memory deficits are due to dysfunctions in encoding, consolidation, or retrieval.

Spatial cognition skills are also affected post-TBI. The term spatial cognition is broad and encompasses multiple functions, including visual scanning, exploring one’s environment, constructional praxis, route finding, and leaning of spatial patterns and complex scenes. Hemispatial neglect can also be seen post-TBI and is associated with injury to the right parietal lobe.

Executive functions can also be impaired in the brain-injured patient. Executive functions are the processes of the brain responsible for higher order complex activities such as planning, judgment, and decision making. These processes are reliant upon working memory, prospective memory, strategic planning, cognitive flexibility, abstract reasoning, and self-monitoring to function properly. The frontal lobe is thought to be responsible for executive function, in conjunction with broad cortical and subcortical circuits. The frontal lobe executive function networks have been divided into the dorsolateral, orbital, and medial prefrontal networks. The dorsolateral prefrontal network is thought to be responsible for abstract reasoning, planning, and working memory; the orbital prefrontal network, for social and emotional aspects of behavior, emotion-based learning, and social judgment. The medial prefrontal network is thought to be responsible for motivational and attentional processes, which include initiation, inhibition, and maintenance of behavior. The orbital and medial prefrontal areas of the brain are more susceptible to injury from trauma. Frontal lobe and temporal lobe injury can also result in a change in personality and social interactions. The spectrum of personality change is highly variable between patients. Some will become socially disinhibited, manifesting vulgar behavior, irresponsibility, and aggression. Others may become abulic and apathetic. The clinical interview and obtaining a history from family members may assist with recognition of these changes in personality.


C. Spasticity and the Upper Motor Neuron Syndrome

The upper motor neuron syndrome (UMNS) results from damage to the corticospinal tract. Its characteristic findings can be divided into positive or negative signs. Positive signs include increased muscle stretch reflexes, spasms, and co-contraction of agonist and antagonist muscle groups. This constellation of signs is often collectively referred to as “spasticity” and represents motor overactivity. The negative signs of UMNS include decreased force production and dexterity of movement. These signs are associated with loss of control of voluntary movement and represent motor underactivity. UMNS is a common finding in TBI. Approximately 25% of TBI patients will develop UMNS during inpatient rehabilitation. Given the diffuse nature of intracranial injury in TBI, the presentation of spasticity may be highly variable in this population. The presence of long bone fracture and peripheral nerve injuries can complicate the clinical picture of UMNS due to mobility restrictions and the flaccidity and areflexia of lower motor neuron injury. There exists a broad spectrum of treatment options for UMNS in brain-injured patients. These treatments include therapeutic modalities, splinting and casting, oral systemic medications, intrathecal therapy, and chemodenervation. For a more comprehensive review of UMNS and
Imaging Studies

A. Conventional Imaging Techniques

Computed tomography (CT) scan of the head is the initial imaging modality of choice in brain-injured patients. CT scan is able to quickly detect intracerebral hemorrhages, skull fractures, edema, and signs of increased ICP. CT is superior to magnetic resonance imaging (MRI) in detecting injury to the skull vault, whereas MRI has greater sensitivity for detecting intracranial abnormalities. In particular, MRI is superior to CT in the detection of diffuse axonal injury as well as injury to the brainstem, posterior fossa, and subtemporal and subfrontal regions. Findings of diffuse axonal injury may be absent or subtle on CT scan. Small petechial hemorrhages at the junction of gray matter and white matter in the corpus callosum may be seen on CT. MRI susceptibility-weighted images are useful for detecting areas of microhemorrhage associated with this injury. MRI is contraindicated in patients with pacemakers, metallic implants, ocular foreign bodies, and certain vascular clips.

B. Advanced Neuroimaging Techniques

Although not routinely available in clinical use, several advanced neuroimaging techniques show promise for assessment of TBI. Diffusor tensor imaging (DTI) is an MRI sequence used for evaluating the structural integrity of white matter tracts in the brain. DTI utilizes fractional anisotropy and apparent diffusion
coefficient, which are measures of water diffusion in the brain. This technique can provide a roadmap of white matter tracts and their relative integrity. Unfortunately, fiber tracking algorithms are not well developed and lack sensitivity, and currently the primary use of DTI is as a research tool.

Magnetic resonance spectroscopy (MRS) is used to detect levels of neurometabolites post-TBI. Metabolites measured include N-acetylaspartate, creatinine, choline, glutamate, lactate, and myoinositol. It is beyond the scope of this chapter to discuss the variations in these metabolites in TBI. However, MRS is used to detect TBI when conventional neuroimaging is normal, to predict outcome, to assess severity of injury, and to determine the stage of brain recovery. It is currently limited to research applications owing to complexities of data interpretation, which are influenced by severity of injury, time of measurement, and location examined within the brain.

Functional MRI measures activity within the brain while performing a task. It relies on the concept that cerebral blood flow changes depending on neuronal activity and need for oxygen. Changes in the blood oxygen level–dependent (BOLD) signal can indicate areas of the brain or networks that are preferentially activated during a specified task. Again, this technique is primarily limited to the research realm due to problems with standardization of measurement.

Single-photon emission tomography (SPECT) imaging is another technique that evaluates cerebral perfusion. SPECT is useful for demonstrating areas of hypoperfusion post-TBI but lacks the sensitivity for smaller lesions detectable by MRI. SPECT is readily available in both research and clinical use. Positron emission tomography (PET) imaging evaluates cerebral metabolism by measuring glucose utilization. PET is not widely available and is expensive.


The physiatrist who cares for patients with TBI must be aware of the common medical and neurologic complications that may arise in these patients. Some of these complications rarely occur outside the realm of TBI and may be unfamiliar to physicians who are consulted to address neuromedical problems. This section highlights the diagnosis and treatment of several problems that, if delayed or missed, may adversely affect recovery.

**Elevated Intracranial Pressure & Hydrocephalus**

Acutely, ICP is often elevated after trauma due to increases in volume within the closed space of the cranium. Sources of increased volume include bleeding, tissue edema, and increased cerebrospinal fluid (CSF). Increased ICP often manifests as a decline in neurologic level of function. A CT scan of the brain is the most common diagnostic test used to assess for this problem and can identify all of the above sources of increased volume. Interventions to address these complications may include surgical evacuation of hemorrhage, administration of mannitol or hypertonic saline to decrease edema, craniectomy to increase the cranial volume, and drainage of the excess CSF.

In the postacute setting, hydrocephalus is common, but the interpretation of the finding of ventricular dilation on head CT can be difficult. With more severe TBIs, hydrocephalus ex vacuo may occur; this is an increase in ventricular size that results from a decrease in brain tissue volume. Because cranial volume does not change, CSF volume increases to compensate for the loss of tissue volume. It may be difficult to determine in these cases whether the hydrocephalus is clinically relevant. Additionally, clinical findings that may be suggestive of significant hydrocephalus, such as urinary incontinence, worsening gait,
cognitive changes, may not be evident in a patient who has significant deficits related to TBI. Increasing ventricular size on serial head CT, cisternography, and a CSF tap test are all tools that are used to determine whether a shunt is indicated, but there is no gold standard. The most common type of shunt is the ventriculoperitoneal shunt. Some shunts have programmable valves whereby the opening pressure can be changed to allow adjustments in CSF drainage. A programmable valve can help prevent complications of over- or underdrainage. Overdrainage may lead to symptoms of orthostatic headache, nausea, dizziness, and mental status changes.


**Venous Thromboembolism**

Venous thromboembolism (VTE), which includes deep vein thrombosis (DVT) and pulmonary embolus, is a common and potentially fatal complication seen in patients after TBI. The incidence of DVT among TBI patients admitted to a rehabilitation unit has been reported as 20%, while the incidence of previous unidentified DVT was reported as 5.5%. Many risk factors for DVT are present in patients who sustain a TBI. These include long bone fractures, immobility, blood transfusions, spinal cord injury, and central venous access. Some patients may have inherited or acquired hypercoagulability due to conditions such as protein C or S deficiencies, or malignancy. The mainstay of prophylaxis, heparin, may be contraindicated after TBI as the risk of intracerebral hemorrhage is increased. Physicians must evaluate the relative risk of VTE versus bleeding when using heparin products. Nonpharmacologic DVT prophylaxis options include intermittent pneumatic compression garments and compression stockings. Vena cava filters decrease the incidence of pulmonary embolus but do not help prevent DVTs. The primary diagnostic tool for DVT is venous Doppler ultrasonography. There is no consensus regarding the use of periodic screening ultrasonography. The d-dimer test is sensitive but not very specific. Risk assessment profiles have been developed for hospitalized patients in an attempt to help determine relative risk of DVT. When DVTs occur, warfarin is the usual method of treatment, but again, the risk of intracranial bleeding must be considered, and vena cava filter is an option in high-risk patients to prevent pulmonary embolus. (DVT management is further discussed in Chapters 5 and
Seizures

The reported incidence of seizures after TBI varies depending on the study population and methods used to identify seizures. One study utilizing continuous electroencephalographic (EEG) monitoring detected seizures in 22% of patients admitted to an ICU with moderate to severe TBI. The risk of post-traumatic seizures and epilepsy (two or more seizures) is increased for patients who have sustained skull fractures, cortical contusions, and subdural hemorrhages. The risk also rises with increased age and severity of brain injury. Randomized studies have demonstrated the efficacy of phenytoin and valproic acid for seizure prophylaxis after TBI, but only within the first week postinjury. Because there is no evidence that medications help prevent seizures beyond this time period, the recommendation is that prophylactic use be discontinued after 7 days. All antiepileptic medications have side effects in the TBI population that may decrease cognition, adversely affect behavior, or cause medical problems, and there is no clinical agreement as to how long antiepileptic drugs should be continued in patients who experience one or more seizures. In addition to the typical signs and symptoms associated with seizures, patients may demonstrate seizure-associated alterations in functional performance, alertness, or behavior that might otherwise be ascribed to neurologic deficits due to TBI. In such patients, an EEG may also be considered.

Inadequate Nutrition or Hydration

Maintenance of adequate nutrition and hydration is important to maximize
recovery after TBI. However, there are several reasons why patients may not meet their nutritional needs. Many cognitive deficits associated with TBI can adversely affect oral intake; these include lack of alertness, inattention, and decreased initiation. Appetite may be altered as a direct result of the brain injury or due to concomitant cranial nerve injuries affecting taste (cranial nerves VII and IX) or smell (cranial nerve I). Medications may also lead to alterations in appetite, as can gastrointestinal problems such as gastroparesis or constipation. Dietary restrictions related to dysphagia (altered textures or thickened liquids) can also adversely affect oral intake.

Dysphagia is a common problem seen in TBI patients. A bedside swallowing evaluation may be performed by a speech therapist, but this test lacks sensitivity for detecting aspiration. Videofluoroscopy and fiberoptic endoscopic evaluations are useful tests that also evaluate the safety of oral intake. These assessments can help determine whether dietary modifications or strategies to improve swallowing (e.g., chin tuck or double swallow) will decrease the likelihood of aspiration. Cognitive deficits resulting from TBI may make the implementation of strategies to improve swallowing more difficult. Medications may affect swallowing directly, or through effects on cognition.

Frequent weight measurements, calorie counts, and laboratory measures (e.g., electrolytes and prealbumin) may aid in the assessment of whether patients are meeting their nutritional needs. Some patients may exhibit significant increase in basal metabolic rate after TBI, so one cannot base nutritional needs on data obtained from the general population. Indirect calorimetry may be useful if patients do not appear to be meeting their needs based on the preceding measures. Malabsorption and conditions that lead to excessive fluid loss may also lead to inadequate nutrition and hydration, even though intake appears to be adequate. Patients with severe dysphagia or other conditions that do not allow adequate oral intake may require enteral feeding and hydration. It is important that a suitable route for nutrition be established within the first week after injury.


Dysautonomia

Autonomic dysfunction is a well-known complication of severe TBI, with a
reported incidence between 8% and 33%. The term paroxysmal autonomic instability with dystonia (PAID) was proposed by Blackman when he developed the following criteria for diagnosis based on a literature review and clinical experience: severe brain injury, temperature of at least 38.5°C, pulse at least 130 beats per minute, tachypnea, agitation, diaphoresis, and dystonia, with at least one episode of these features daily for at least 3 days. Baguley retrospectively reviewed records of 35 patients with dysautonomia and compared them with 35 control patients matched for gender and Glasgow Coma Scale scores. Clinical features of the group with PAID included increased evidence of diffuse axonal and brainstem injuries on CT, as well as greater reports of preadmission hypoxia. The group with PAID had a longer rehabilitation length of stay, greater duration of PTA, and lower Glasgow Outcome Scale (GOS) and Functional Independence Measure (FIM) scores at discharge. (Both of these tools are discussed in more detail later, under Prognosis.)

Symptoms may also be consistent with other medical complications, so a systematic approach should be taken when evaluating patients who demonstrate signs of autonomic instability to rule out other causes for the findings. Other diagnoses to consider include infection, neuroleptic malignant syndrome and, in patients with concomitant spinal cord injury, autonomic dysreflexia. Several medications have been used to treat PAID, perhaps related to the multifactorial nature of the syndrome. Narcotics, β blockers, and gabapentin are among the more widely utilized medications, but there is no consensus regarding which medication might be most effective. Noxious stimuli may trigger these episodes, so a thorough evaluation for causes of discomfort should be performed routinely.


Upper Motor Neuron Syndrome

Complications related to the UMNS can lead to significant functional deficits. Problems arise related to muscle overactivity, weakness, and contracture.
Although these conditions are discussed elsewhere in this text (see Chapter 6), it is worth reviewing them in the context of TBI, because certain features of brain injury may affect clinical course and treatment decisions. Muscle overactivity and weakness are commonly seen after TBI, and both may lead to muscle stiffness and contractures with loss of range of motion in multiple joints as well as the spine. This may be very pronounced for patients who have PAID (see preceding discussion). Because PAID may be triggered by noxious stimuli, the clinician must use caution when proceeding with interventions such as casts, which may cause pain. Although spasticity and dystonia may be present in several limbs, systemic medications should be used cautiously since most of these drugs can cause sedation and worsen cognitive deficits related to TBI. Because of this, focal interventions (botulinum toxin injections, nerve blocks, and motor point blocks) may take a more prominent role in care of this population than in conditions where cognitive function is better preserved. Neuro-orthopedic interventions may also be appropriate to improve positioning or function, or both, by addressing contractures and restoring function of a joint via muscle transfers.


Heterotopic Ossification

Heterotopic ossification is discussed elsewhere in this text (see Chapter 5) but warrants mention here as some aspects of this orthopedic disorder are particular to TBI. Although the pathophysiology is unclear, risk factors include hypertonicity, decreased movement of the affected joint, severity of brain injury, and dysautonomia. The most common locations for heterotopic ossification after TBI are hips, followed by shoulders, elbows, and knees. The incidence has been reported to be as high as 73%, but clinically significant disease occurs less frequently. The utility of medications for prophylaxis or acute treatment of heterotopic ossification after TBI is uncertain. Surgical intervention may be
considered when specific functional goals, active or passive, are identified. This is not usually performed until 6–18 months after injury to allow bone maturation and to decrease the likelihood of recurrence.


**Neuroendocrine & Electrolyte Abnormalities**

Up to 30–40% of selected groups of TBI patients demonstrate at least one endocrine abnormality. Risk factors include basilar skull fracture, hypothalamic edema, prolonged unresponsiveness, hyponatremia, and hypotension. Identification and treatment of hormone deficiencies is warranted because recovery after brain injury may be slowed in the absence of treatment. Also, many of the clinical findings associated with these deficiencies mimic (and likely functionally worsen) deficits seen after TBI. For example, cognitive deficits, weakness, and fatigue are common problems associated with TBI but are also consistent with hypothyroidism, growth hormone deficiency, hypogonadism, and Addison’s disease (adrenocorticotropic hormone deficiency). Consensus guidelines exist that support the use of routine screening for some patients with TBI.

One of the most common endocrine abnormalities seen after TBI is the syndrome of inappropriate antidiuretic hormone secretion (SIADH), which leads to hyponatremia. Nausea, weakness, and mental status changes are problems seen with hyponatremia. The primary treatment is fluid restriction, and demeclocycline is an alternative if fluid restriction fails to resolve the problem. Other causes of hyponatremia in TBI patients include cerebral salt-wasting syndrome and psychogenic polydipsia. For any electrolyte abnormality, iatrogenic causes should also be considered in the differential diagnosis. While SIADH is caused by excessive antidiuretic hormone (ADH) secretion, diabetes
insipidus is caused by insufficient ADH secretion. This abnormality is usually associated with craniofacial trauma. Hypernatremia, polydipsia, and high urine output are the main clinical findings. Vasopressin is the primary treatment for central diabetes insipidus. Table 13–3 summarizes findings and treatment for these abnormalities.

Table 13–3 Common electrolyte abnormalities associated with TBI.

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Causes</th>
<th>Clinical Findings</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyponatremia</td>
<td>SIADH</td>
<td>For all etiologies—mental status changes, hyperreflexia, seizure, plus:</td>
<td>Fluid restriction, demeclocycline</td>
</tr>
<tr>
<td></td>
<td>Cerebral salt-wasting syndrome</td>
<td>Clinically euvoletic, high urine osmolality and urine sodium</td>
<td>Sodium and fluid replacement</td>
</tr>
<tr>
<td></td>
<td>Psychogenic polydipsia</td>
<td>Hypovolemia, high urine osmolality and urine sodium</td>
<td>Fluid restriction, clozapine</td>
</tr>
<tr>
<td>Hyponatremia</td>
<td>Diabetes insipidus (DI), diuretics, dehydration</td>
<td>Lethargy, weakness, mental status changes, thirst, polyuria (for central DI)</td>
<td>Fluid replacement, DDAVP (for central DI)</td>
</tr>
<tr>
<td>Hypokalemia</td>
<td>Diuretics, hypomagnesemia, excess catecholamines</td>
<td>Lethargy, weakness, hyporeflexia, arrhythmia</td>
<td>Correct underlying disorder, potassium replacement</td>
</tr>
<tr>
<td>Hyperkalemia</td>
<td>Hypoaldosteronism, hemolyzed blood specimen, renal dysfunction</td>
<td>Muscle weakness, cardiac conduction block, peaked T waves on ECG</td>
<td>Correct underlying disorder, Kayexalate, diuretics</td>
</tr>
</tbody>
</table>

SIADH, syndrome of inappropriate secretion of diuretic hormone; DDAVP, vasopressin; ECG, electrocardiogram.


Other Trauma-Associated Injuries

It may be difficult to identify peripheral orthopedic and nerve injuries in patients who sustain severe TBI in the setting of multiple trauma due to their compromised cognitive status or ability to communicate. Pain and weakness, the most common symptoms of these injuries, may be difficult to detect. Therefore, it is not surprising that these injuries may not be identified in these patients until after they are admitted to inpatient rehabilitation. The incidence of nerve injuries was reported to be 34% in a cohort of TBI patients in one study. Peripheral nerve injuries may be suspected when focal hypotonicity or focal diminished muscle stretch reflexes are noted, even in patients who are aphasic or unable to follow commands for formal manual muscle testing. Increased suspicion for these injuries may lead to earlier identification and treatment for these common complications. Shoulder trauma may lead to brachial plexus injuries. Lumbosacral plexopathies may result from pelvic trauma or retroperitoneal hemorrhage. Some fractures are also associated with nerve injuries, such as humeral fractures and radial nerve injury. Another cause of neuropathy in this population is critical illness polyneuropathy. This is a more symmetric weakness, and has significant implications for the time course of functional recovery. Electrodiagnostic testing in the primary tool for identifying neuropathies and myopathies.

The incidence of fractures that are first identified in rehabilitation in the TBI population has been reported as 11%, with a mean delay in diagnosis of 57 days. Most of these fractures were not of significant clinical consequence. It is often not until patients are more alert and able to report pain or demonstrate focal functional deficits that these fractures become clinically detectable. The most appropriate timing of fracture repair for patients with moderate to severe TBI is not well established. Early repair may decrease the incidence of some complications associated with fractures, but other studies suggest that early repair may lead to a higher incidence of medical complications that may adversely affect brain injury recovery.

Sleep Dysfunction

Sleep disturbances are very common after TBI, affecting up to 70% of patients in some studies. Recognition of these disorders is important because they may have a significant effect on patients’ daily physical and cognitive function as well as participation in rehabilitative activities. There are many reasons why patients may not get adequate sleep. Proper identification of the cause is important in determining the most appropriate treatment. Assessing the environment may be very helpful. This is especially important in a hospital setting, although altering the environment may be more difficult in that setting. Pain and stress may also have an adverse effect on sleep initiation and maintenance. Medications may interfere with sleep; alternatively, sedating medications given during the day may lead to daytime sleep, which will also disrupt nocturnal sleep. Sleep disorders that may be related to or predate the TBI, such as obstructive sleep apnea and restless legs syndrome, will also negatively affect the quality of sleep. Chronic sleep problems may lead to maladaptive behaviors that may worsen the situation. Counseling regarding basic sleep hygiene principles may help address these problems.

A number of medications are available to help restore regular sleep, including antidepressants (especially trazodone and tricyclic antidepressants), benzodiazepines, and nonbenzodiazepine sedatives such as zolpidem, melatonin, and diphenhydramine. Because of their sedating properties, these agents have cognitive side effects, so it is important to determine that the medication being utilized is having a beneficial effect on daytime performance. Some of these medications may have an idiosyncratic effect of worsening sleep, so efforts must be made to assess for this outcome. A sleep log is useful in both the inpatient and home setting to determine whether the problem is with sleep initiation or maintenance, and may also be used to quantify other barriers to uninterrupted sleep (eg, nocturia, pain, environmental effects).

Daytime fatigue is a very common complaint in both the inpatient and outpatient settings. It is common in mild TBI populations as well as more severely injured individuals. There are many etiologies to consider in addition to poor sleep at night. Damage to structures in the brain, such as the reticular activating system, will affect alertness. Many medications used to treat common medical problems have sedating side effects (eg, antihypertensives, anxiolytics), as do medications prescribed to address trauma-related complications such as pain. Specific endocrine abnormalities may lead to daytime fatigue, as can depression. If a cause cannot be identified and addressed, consideration should
be given to the use of medications to enhance alertness, such as dextroamphetamine, methylphenidate, and modafinil, but evidence supporting their use is limited.


Post-Traumatic Headaches

Headaches are very common after TBI, with an incidence reported to be between 30% and 90%, depending on the population studied, time since injury, and instruments used. Some reports suggest that the incidence of post-traumatic headache (PTH) is higher after milder TBI when compared with more severe injuries, but this is not a consistent finding. Occasionally, PTH is related to a serious intracranial complication such as infection, hydrocephalus, or hemorrhage, so these problems should be ruled out in patients with more severe headaches, acute onset of headaches, or those who demonstrate other signs and symptoms consistent with neurologic compromise.

Several types of headaches are commonly seen after TBI (Table 13–4). It is important to try to determine the type of headache since treatment will be based on this classification. Some patients may present with more than one type of headache. Headache logs are very useful tools to help determine the type of headache as well as efficacy of the headache interventions. The logs may also help patients to identify activities that provoke headaches, which may help identify TBI-associated complications that are leading to headaches (eg, visual deficits) or activity modification.

Table 13–4 Types of headaches commonly seen after TBI.
Chronic headaches are often more difficult to treat. If prior interventions have not been successful, reconsideration of headache typology may be warranted. Cognitive-behavioral interventions should be considered. Rebound headaches are a known complication of chronic pain medication used to treat headaches. Increasing doses of medications are required to control the pain, and headaches return soon after the medications are metabolized. Treatment of rebound headaches requires the slow tapering of pain medications, which will initially result in an increase in pain. Occasionally patients may need to be hospitalized in order to be safely withdrawn from the offending pain medication while adequately controlling pain by other means.

Visual Deficits

Several different types of visual difficulties may result from trauma to the head. Compounding the problem, it may be very difficult to assess visual function in cognitively impaired patients. Visual evoked responses can provide some information regarding neurologic injury to the visual system even in patients who cannot participate in a visual examination due to severe cognitive deficits. For patients who are aroused but cannot follow commands, some estimate of vision (and cognition) may be made by assessing tracking or quantifying the amount of time that gaze is focused on a visual field with salient images compared with, for example, a blank card. Visual inattention and visual field deficits will also need to be considered when assessing a patient’s visual abilities.

Injuries to the optic nerve may be a direct result of trauma to the orbital area. Ptosis may lead the clinician to suspect a cranial nerve III injury with resultant deficits in extraocular movements and accompanying visual disturbances. Evaluation of cranial nerves IV and VI may be performed in a patient who has the ability to track objects. A dysconjugate gaze and accommodation insufficiency may also be seen with milder TBIs, and as well as causing visual complaints may lead to headaches. Some of these injuries may improve with vision therapy, including the use of prisms, patching, and central occlusion.

TREATMENT

Spectrum of Care

The rehabilitation of persons with TBI may be considered to begin as soon as care is initiated. Efforts to minimize the extent of neurologic injury aid in the rehabilitative process. Prevention of medical complications that may hinder restoration of function is an important aspect of rehabilitation. Care also extends well beyond the period of hospitalization, as efforts are made to allow patients to be as independent as possible, including activities in the home, community, school, and workplace. Survivors of TBI and their families and friends often need lifelong services to maintain a high quality of life.

Acute Care
An important aspect of controlling secondary brain injury acutely after trauma is the control of ICP and the maintenance of cerebral blood flow. Causes of increased ICP and possible interventions were discussed earlier, under Complications. ICP increases logarithmically as the intracranial volume increases. Increased ICP may lead to herniation as well as inadequate cerebral blood flow, worsening the extent of the brain injury. Cerebral blood flow is monitored by calculating cerebral perfusion pressure (CPP), which is controlled by two variables, ICP and mean arterial blood pressure (MAP), such that CPP = MAP - ICP. Therefore, both ICP and systemic blood pressure must be managed to maintain an adequate CPP. After severe TBI, research suggests that the optimal CPP is between 60 mm Hg and 70 mm Hg.

Various therapies have been studied to try to minimize the extent of secondary brain injury. Because there are so many different mechanisms of secondary injury, it is not surprising that many different types of agents and modalities have been utilized, including N-methyl-D-aspartate receptor antagonists, calcium channel blockers, hypothermia, antiinflammatory agents, and various growth factors. One of the more promising agents is the hormone progesterone, which may work through several different pathways to decrease secondary brain injury after trauma.


Rehabilitation

Rehabilitation after TBI relies on a transdisciplinary approach. Patients have a wide variety of needs and deficits, so a coordinated team is required to address the many aspects of TBI rehabilitation. The team must be able to manage complex neuromedical problems, as mentioned previously; provide therapies that appropriately address physical cognitive and behavioral deficits; and also address the psychological and social needs of patients and their families. Therefore, the team often includes physical, occupational, and speech therapists; psychologists or neuropsychologists, or both; recreational therapists; social
workers; case managers; rehabilitation nurses; and physicians. The composition of the team may change as the patient progresses from inpatient rehabilitation to the outpatient and community setting. At that point, vocational or school reentry personnel may also become involved.

Several aspects of the neuromedical care of patients with TBI have been discussed earlier. When a patient is admitted to a rehabilitation hospital it is important to assess the medications he or she had been taking in the acute care setting. The patient’s medication regimen should also be assessed periodically for the duration of care (inpatient and outpatient), as the requirements for these medications may change. Many medications have the potential to slow recovery or decrease cognition, in addition to other side effects. A sound pharmacologic strategy is to minimize medications (ie, discontinue any that are no longer needed), substitute by choosing a medication from a class of drugs based on side effect profile, and possibly augment, by utilizing a medication that may promote recovery. Some commonly prescribed medications that may cause sedation include antiepileptic medications, centrally acting antihypertensive medications, pain medications, anxiolytics, and antipsychotic medications. Clearly, there is often a role for these classes of medications for many patients with TBI, but the potential for unwanted side effects should be considered. Some of these medications have also been shown to slow recovery after experimental brain injuries in different animal models.

A. Behavioral Problems after TBI

Among the most challenging problems in the rehabilitation of patients with TBI are behavioral problems, including agitation, aggression, and irritability. These problems may interfere with needed medical care, affect patient and staff safety, make a discharge to community more difficult, and be a major barrier to community reentry. These behaviors may be exacerbated by other neuromedical problems such as pain, confusion, medication effects or interactions, depression, and sleep dysfunction. Therefore, these and other potentially treatable causes should be considered and addressed as appropriate. It is also important to identify specific target behaviors that the team deems important. That is, some behavioral problems may be more tolerable than others. Efforts should be made to quantify the target behaviors in order to objectively determine whether chosen interventions are, in fact, effective. A more general assessment tool, such as the Agitated Behavior Scale may also be used.

While clinicians often rely on medications to address behavioral problems, there is no consensus regarding which medications to use, as all of these
Mediations may have unwanted side effects. A Cochrane review of studies that have evaluated the efficacy of medications to address agitation found the most support for the use of propranolol, although a survey of physicians who regularly treat patients with TBI showed carbamazepine to be the most commonly used medication. With the exception of extreme agitation that jeopardizes safety, the use of “as-needed” medications to treat agitation should be avoided. It is very difficult to assess efficacy of medications in such cases, because different staff members will have varying thresholds for deciding when to administer them. Also, agitation is often self-limited, and may be waning on its own by the time the medication is taking effect, leading to the incorrect assumption that the medication achieved the desired effect. As noted, many of the medications used to treat agitation have undesirable side effects, making it even more important to demonstrate benefit given the risks. The use of scheduled medications along with objective and systematic measurement of behavioral problems will increase the likelihood of determining whether the medication is having the desired effect (ie, fewer or less intense behavioral episodes per unit time). Sufficient time should be provided between changes in medication regimens to obtain enough data to determine efficacy, especially in cases where the behavioral problems are variable. Pharmacologic options may include anticonvulsants, antipsychotics, stimulants, β blockers, and serotonergic medications.

Behavioral approaches may also be undertaken, governed to some degree by the cognitive abilities of the patient. For instance, an elaborate reward system may not be useful for a patient who is extremely impulsive or one who cannot understand or remember how the reward system works. For some patients, the treatment team may consider evaluating the “ABCs” of behavior: antecedent, behavior, and consequence. A stimulus (antecedent) often provokes a certain behavior, and often the consequence of the behavior reinforces (reward) or extinguishes it. Assessing the “ABCs” may allow the treatment team to formulate a behavioral plan that can be consistently implemented and then reassessed to determine whether the plan needs to be modified.


B. Evaluation and Treatment of Cognitive Deficits
As previously described, great variability in cognitive deficits is seen after TBI, consistent with the variable and diffuse nature of the extent of damage to the brain. However, some areas of the brain are relatively susceptible to traumatic injury; therefore, common patterns of neurologic deficit occur.

**Arousal** is perhaps the most important aspect of cognition in that patients must be aroused to perform any functional task or participate in therapy. Without arousal, one cannot perceive or respond to any sensory input from the environment. The reticular activating system is a very important neurologic structure for maintaining arousal. Strategies that may benefit the patient’s state of arousal include promoting proper sleep, minimizing use of sedating medications, ensuring rest breaks, and alternating activities. Some medications, including stimulants (eg, dextroamphetamine, methylphenidate) and dopaminergic agents (eg, amantadine, bromocriptine) may also improve arousal.

**Attention** refers to being able to select one stimulus from a field of many stimuli. It can be considered a more focused arousal. Several types of attentional deficits are seen after TBI. Patients may demonstrate distractibility and can then only perform well in environments with minimal visual and auditory stimulation. Deficits in divided attention lead to difficulties in performing more than one specific task simultaneously. Hemispatial neglect is another form of disordered attention. Strategies may be implemented that address these specific deficits, such as maintaining a nondistracting environment, encouraging a patient to perform one task at a time, or using visual cues on the affected side to address hemispatial neglect. As patients improve, these compensatory strategies may be slowly withdrawn. The efficacy of medications to address deficits in attention is not well established. Dopaminergic and stimulant medications have been the most widely studied, and one evidence-based review reported that the agent with the best evidence in this regard is methylphenidate.

Disorders of **executive function** are quite common after TBI, related in part to the vulnerability of the frontal lobes to injury with head trauma. Executive function refers to the ability to control impulses in order to act appropriately, adapt to different situations, and demonstrate planning and goal-directed behavior. Deficits in these areas may be quite disabling during community reentry such as vocational and avocation activities. The patient may be unable to plan and follow through with even basic daily tasks and may also have great difficulty maintaining socially acceptable behavior. Strategies that may help with executive deficits include training that targets specific problems, such as an undesirable behavior, or developing a plan to organize and carry out a task. One of the challenges is that patients often have difficulty generalizing these
strategies to even slightly different situations, which is especially problematic in the community as opposed to a clinic or hospital setting.

Memory impairment is another very common complication of TBI. As mentioned previously, the length of PTA is one of the more sensitive ways to evaluate severity of TBI. Anterograde memory problems may persist, even as a patient emerges from PTA. Deficits in arousal attention will also make memory tasks more difficult. Several different strategies are utilized to address memory deficits. Skill acquisition may develop through procedural learning, where tasks can be learned automatically, without the need for conscious awareness. For some patients, compensatory strategies work very well; memory books or electronic devices with calendar features are examples. Patients must be evaluated as to the appropriateness of devices and trained as to how to use them. There is a paucity of methodologically rigorous studies supporting the use of medications to improve memory after TBI.

Language impairments after TBI can be quite varied. While they may mimic to some degree the well-characterized aphasias seen in stroke, they are more often a blend of several different impairments consistent with the more diffuse nature of TBI. Therefore, treatment strategies often need to address several different pathologic processes. For instance, a patient who has difficulty following verbal commands may have hearing deficits and poor attention in addition to a receptive language component. Reading may also be compromised by distractibility, visual neglect, or other visual deficits, in addition to problems with comprehension. Verbal expression can be less effective due to disorganized speech and social inappropriateness in addition to more characteristic expressive aphasia. Therefore, the social and behavioral aspects of impaired functional communication need to be addressed in addition to the identified discrete language deficits.


C. The Challenge of Community Reentry

Many of the strategies that are implemented in the clinic or hospital setting to improve function for patients with TBI are reproducible in the community. Other problems that are not faced in the clinical setting will arise in the community. For instance, other individuals may not be available to provide the verbal cues that the patient may need to initiate tasks or carry them out appropriately. The household may be a very distracting and overstimulating environment. Alternatively, patients may find themselves very socially isolated in the community, perhaps leading to depression. A patient who relies on consistency and a routinized pattern will likely find it difficult to maintain this structure in “the real world.” Access to alcohol or other drugs will be much easier at home compared with the hospital setting. School and vocational activities rely on many skills that will need to be addressed on an outpatient basis. Given the importance of activities and participation, there are clearly many more aspects to a meaningful recovery from TBI than basic (but important) tasks such as ambulation and activities of daily living.

Community reentry skills for hospitalized patients should begin before discharge. Mobility in the community requires a more varied skill set than does ambulation in the hospital. Car transfers, ambulation on uneven surfaces, and negotiating busy and distracting environments are all skills that are necessary for a safe return to the community. The social aspects of being in the community should also be assessed and, if available, caregivers should participate in the community reentry activities to provide them with insight and better prepare them for the challenges they will face.

Patients should develop an activity pattern that provides structure and also encourages them to remain active. Ideally this activity pattern will include both activities such as therapy that will help with continued skill acquisition, as well as social activities that may help prevent social isolation and lessen the risk of depression, which is very common in the TBI population. Incidence of major depression was found to be 33% in a group of patients followed for 1 year after TBI. A regular activity pattern may also help patients maintain a regular sleep cycle.

Return to school or work will be a goal for many patients after TBI. The likelihood of successful return to work is increased for patients with a higher
level of education and stronger work history, and decreased for individuals with more severe injuries and continued cognitive and behavioral deficits. Clinicians may need to work closely with schools or employers to provide appropriate accommodations to support successful return to academic or vocational activities. An individualized assessment of work tasks and the work environment should be done, along with an evaluation of a patient’s strengths and weaknesses, to develop strategies and therapy activities that take all of these variables into account.


D. Substance Abuse

Alcohol use is a risk factor for TBI. Between 35% and 50% of TBIs involve intoxicated individuals, and 50–66% of those hospitalized for TBI have a history of alcohol or other substance abuse. Intoxicated patients as well as those with a history of alcohol use have an increased risk of medical complications, longer acute hospital length of stay, and longer duration of PTA. It is important to obtain an accurate history regarding substance abuse because prior abuse may increase the risk of subsequent abuse, although it is less clear whether TBI increases the risk of substance abuse in patients who do not have a prior abuse history. The CAGE questionnaire is one tool that can be used to screen for prior alcohol abuse. Alcohol use is a risk factor for slower recovery from TBI, and animal models have helped explain some of the reasons for this finding. Ongoing alcohol use is also related to poorer long-term outcomes such as return to work.

Special Populations

A. Disorders of Consciousness

The challenges of assessing patients with disorders of consciousness were discussed earlier, under Clinical Findings. Assessment techniques and treatment goals must be modified and individualized to the patient’s abilities. Evaluations must include repeated assessments because inconsistency of performance is common in this population, leading to difficulty in determining whether responses are random or related to a command or stimulus. Perhaps the most important intervention is maintaining medical stability to allow the brain the best opportunity for continued recovery while preventing complications that may adversely affect function (described earlier).

There is limited evidence supporting specific rehabilitation interventions that may improve outcomes. A recent randomized, blinded, placebo-controlled trial that studied the safety and efficacy of amantadine in improving recovery for patients in a minimally conscious or vegetative state did demonstrate an improved rate of recovery for the patients who received amantadine. Other medications for which there is published evidence of efficacy for this population include amitriptyline and bromocriptine. Hyperbaric oxygen, deep brain stimulation, and sensory stimulation are other examples of interventions that have been attempted with varying amounts of success to improve recovery.

When considering an intervention such as a medication in an attempt to improve cognition, it is important to choose reliable, reproducible, objective measures that can identify the small changes in the rate of responsiveness that may occur. For example, arousal might be measured by recording how often eyes are open or closed at defined intervals during specific time periods in the day. Improvements in awareness may be noted by the changes in percentage of time spent visually attending to a salient object versus a blank card. Perhaps as important is the usefulness of systematic objective data collection in identifying a decline in function that may be missed in the absence of such data. This may lead to initiation of a neuromedical workup and earlier identification of a potentially treatable complication.


B. Mild TBI and Concussions

Using the criteria presented earlier, the majority of TBIs are categorized as mild. Symptoms usually resolve within a few weeks for most patients with mild TBI. Patients who sustain a sports-related concussion tend to have quicker recovery than those who sustain other forms of TBI. In one study, 90% of collegiate football players recovered within 7 days. In a civilian population, Ponsford identified several risk factors for a protracted recovery: history of prior TBI, history of neurologic or psychiatric problems, female gender, injury due to motor vehicle crash, and student status. A study of U.S. military service members (mean of 9.4 months postinjury) identified possible symptom exaggeration, poor effort, traumatic stress, and depression as most predictive of postconcussive symptom reporting.

The Diagnostic and Statistical Manual of Mental Disorders (DSM) criteria for postconcussional disorder specify a history of head trauma causing significant cerebral concussion, evidence of cognitive decline on neuropsychological testing, and three or more symptoms lasting greater than 3 months. The symptoms are fatigue, disordered sleep, headache, vertigo or dizziness, irritability, anxiety or depression, personality changes, and apathy. Because many of these symptoms can occur in the absence of TBI, it is sometimes difficult to determine whether they are related to the initial injury. This problem is more relevant to diagnosis of postconcussive disorder than to treatment as the interventions likely will not differ based on cause.

Acute evaluation and treatment for concussions and return to play guidelines for sports concussion have been discussed elsewhere (see Chapter 29). When patients are slow to recover from mild TBIs, other etiologies for their persisting symptoms should be considered. For headaches, the prior working diagnosis regarding headache typology may be incorrect. Undiagnosed visual problems may be causing the headaches. Cranial or cervical trauma may not have fully resolved. Regarding daytime fatigue and cognitive complaints, symptoms might be related to a sleep disorder, side effects of medications, or depression. Maladaptive behaviors regarding pain may have developed. Similarly patients may be consciously or subconsciously motivated to over-report symptoms or exaggerate deficits on examination. Psychosocial factors, expectations regarding the injury, and personality traits may all contribute to the variation in recovery rates, and if these issues are identified, interventions can be employed.
PROGNOSIS

Determining a long-term prognosis for a brain-injured patient is one of the most challenging tasks for the rehabilitation physician, as well as one of the most frustrating issues for patients and their families. There will always be a great deal of uncertainty regarding long-term functional outcome due to the variable rates at which patients recover and the complexities of the injury itself. Uncertainty also arises from the myriad ways in which outcome is measured. A multitude of methods are available for measuring outcome post-TBI. These methods include but are not limited to discharge disposition, return to independent living, return to prior level of work or school, return to prior level of function, and Functional Independence Measure (FIM) score change. The Community Integration Questionnaire (CIQ) and the Mayo Portland Adaptability Inventory are used to measure outcome involving function in the community setting. Some of the more commonly used standardized assessment tools include the Glasgow Outcome Scale (GOS; Table 13–5), the Coma Recovery Scale–Revised (CRS-R), and the Disability Rating Scale (DRS).

Table 13–5 Glasgow Outcome Scale.
The FIM is well known and is in widespread use across all areas of rehabilitation. It is an 18-item scale with domains measuring self-care, communication, sphincter control, mobility, locomotion, and social-cognitive function. It is reliable, validated, and best used to mark progress in acute inpatient rehabilitation. It has a ceiling effect for higher functional levels and is less sensitive to changes in patients postdischarge from acute inpatient rehabilitation.

The CIQ is a 15-item scale that measures home integrations, social integration, and productive activities. It is easy to administer, requires no formal training, and can be self-scored or scored by a proxy. Overall, reliability is mixed. The Mayo Portland Adaptability Inventory consists of three subscales (Ability Index, Adjustment Index, and Participation Index) designed to assess physical, cognitive, social, emotional, and behavioral difficulties posthospitalization.

The GOS is widely used and has both advantages and disadvantages. Its advantages are that it is widely used and contains broad terms that are easily understandable to the layperson. It addresses major functional outcomes that are of prime importance to patients and families. Its disadvantages include the fact that the categories are so broad that there exists great variability of patient function within each category. Additionally, the broadness of the categories makes it relatively insensitive to change. There is also some concern about reliability and subjective scoring by the rater, as there is no formal or standard

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<td>Dead</td>
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<td>2</td>
<td>Vegetative state (&quot;alive but unconscious&quot;)</td>
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<td>3</td>
<td>Severe disability (&quot;conscious but dependent&quot;)—unable to live alone for more than 24 hours: daily assistance of another person is needed for physical and/or cognitive impairments.</td>
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<td>4</td>
<td>Moderate disability (&quot;independent but disabled&quot;)—independent at home, able to utilize public transportation, able to work in supported environment.</td>
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<td>5</td>
<td>Good recovery (&quot;mild to no residual deficits&quot;)—capacity to resume normal occupational and social activities, possible minor residual physical or cognitive deficits.</td>
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protocol for rating. The Glasgow Outcome Scale Extended (GOS-E) addresses the limitation of the GOS. The GOS-E is divided into eight categories (Dead, Vegetative State, Lower Severe Disability, Upper Severe Disability, Lower Moderate Disability, Upper Moderate Disability, Lower Good Recovery, and Upper Good Recovery). The GOS-E is administered via a structural interview.

The DRS is another widely used standardized assessment scale for outcome measurement. Advantages of the DRS include its ability to be used on patients at very low levels of function from acute hospitalization all the way through to patients functioning in the community. The scale can be scored by trained healthcare professionals or family members without formal training. Scores range from 0 to 29, with 29 indicating extreme vegetative state and 0 indicating no disability. Unfortunately, the DRS is relatively insensitive for measuring change in patients with lower levels of disability.

The CTD-R was discussed earlier in reference to disorders of consciousness. It is very sensitive for patients at lower functional levels and in determining level of consciousness. However, it has a ceiling effect in that it is insensitive at detecting change in patients at higher functional levels. The CRS-R can be time consuming and requires formal training and administration by a rehabilitation professional.

Utilizing threshold values to convey prognosis is a clinically useful way to inform families and patients about outcome. As defined by Kothari, a threshold value is a “value of a predictor variable above or below which a certain outcome is especially likely or unlikely.” In his chapter on prognosis in *Brain Injury Medicine*, Kothari lays out a blueprint for using threshold values and approaching patients and family members for a discussion about prognosis. By using these threshold values, the clinician can provide practical information to patients and family that is evidence based. These threshold values are based on the GOS as the outcome measure.

1. Severe Disability on GOS is unlikely when:
   a. Time to follow commands is less than 2 weeks
   b. Duration of PTA is less than 2 months

2. Good Recovery on GOS is unlikely when:
   a. Time to follow commands is longer than 1 month
   b. Duration of PTA is greater than 3 months
   c. Age is greater than 65 years and TBI is severe

With regard to prognosis for disorders of consciousness, the preceding
guidelines using an evidence-based approach with practical threshold values still apply. In addition, the natural history of recovery from disorders of consciousness has been described. Approximately one third of patients who remain in a vegetative state for 3 months after a TBI regain consciousness by 1 year postinjury. However, if the brain injury is nontraumatic, only 5% of patients who were vegetative at 3 months regain consciousness at 1 year postinjury. If patients remain in a vegetative state for 6 months postinjury, the probability of regaining consciousness at 1 year drops to 15% for traumatic injuries and near zero for nontraumatic injuries. Nearly 50% of patients who were deemed minimally conscious on admission to inpatient rehabilitation attained a level of “no to moderate disability” on the DRS at 1 year postinjury. Only 3% of patients who were deemed vegetative at time of admission to inpatient rehabilitation reached a comparable level on DRS. There is also evidence to suggest that late recovery from disorders of consciousness is possible beyond 1 year postinjury. In one series of 50 vegetative patients, 24% emerged from a minimally conscious state between 19 and 25 months postinjury. In another series of 18 patients who were minimally conscious at the time of discharge from rehabilitation, nearly one third recovered to an independent level of cognitive or motor function between 2 and 5 years postinjury.


Malec J: The Mayo Portland Adaptability Inventory. The Center for Outcome Measurement in Brain Injury. 2005. Available at:


Although stroke was once thought of as an incurable and static disease, numerous advances have been made over the past decade in the prevention, diagnosis, management, and rehabilitation of this disorder. In particular, the treatment and rehabilitation paradigm has improved significantly as a result of advances in acute interventions, risk reduction, medical devices, therapeutic modalities and exercise, robotics, diagnostic imaging techniques, and our overall understanding of the disease process itself. This resulted in a 35% reduction in the population-based death rate due to stroke between 1998 and 2008. Even with this progress, stroke remains a leading cause of death and disability in the world. Multidisciplinary stroke rehabilitation remains the primary treatment for post-stroke disability and should begin as soon as possible to optimize functional recovery and avoid potential complications and setbacks.

**ACUTE DIAGNOSIS & MANAGEMENT OF STROKE**

Stroke is a sudden-onset neurologic dysfunction resulting from focal disruption to the cerebrovascular system that requires rapid diagnosis and intervention. Acute neurologic symptoms are a medical emergency that justify immediate transport to the emergency department of an acute-care hospital for evaluation and treatment. It is vital to differentiate hemorrhagic from nonhemorrhagic (thrombotic or thromboembolic) strokes as soon as possible after onset of symptoms. Noncontrast computed tomography (CT) scan (Figure 14–1) is highly
sensitive for acute bleeding and is commonly used for this purpose. Intravenous tissue plasminogen activator (tPA) should be considered for selected patients with acute thrombotic stroke within 3 hours of symptom onset, but other acute interventions can also be considered after that timeline.

▲ Figure 14–1 Computed tomography image showing evidence of an acute ischemic stroke in the left middle cerebral artery distribution.

A thorough neurologic examination, combined with immediate neuroimaging study is indicated to evaluate acute neurologic symptoms suggestive of stroke. Magnetic resonance imaging (MRI) (Figure 14–2) is more sensitive for detecting posterior fossa lesions and acute ischemia within 24 hours of the stroke, especially using diffusion-weighted studies (Figure 14–2C). Patients with cardioembolic strokes caused by atrial fibrillation or associated with a proven embolic source in the heart and great vessels should be considered for anticoagulant therapy. Acute strokes that occur simultaneously in two areas of
the brain subserved by different blood vessels are considered to be embolic until proven otherwise. Magnetic resonance angiography can help identify and characterize occlusion or stenosis of major cerebral vessels and the presence of cerebral aneurysms of moderate or large size. (Figure 14–3).

▲ Figure 14–2 Magnetic resonance imaging scan showing an acute right deep cerebral white matter infarction involving the right corona radiata and periventricular regions. **A:** T2 imaging. **B:** Fluid-attenuated inversion recovery imaging. **C:** Diffusion-weighted imaging.
**Figure 14–3** Magnetic resonance angiography imaging showing (A) normal circle of Willis and (B) circle of Willis in a stroke patient, demonstrating decreased flow in the left middle cerebral artery as well as other changes.


DEFINITIONS & GENERAL CONSIDERATIONS

Stroke, or cerebrovascular accident (CVA), is characterized by an acute onset of a neurologic deficit that persists for at least 24 hours and is the result of a focal lesion to the cerebrovascular system. A transient ischemic attack (TIA) is an acute-onset neurologic deficit that (apparently) resolves within 24 hours. However, approximately 17% of patients with TIAs suffer strokes within the next 3 months. In many patients with TIAs or other strokes with seemingly complete recovery, neuropsychological testing reveals long-lasting or permanent subclinical deficits; hence these events are associated with true damage, and all clinicians involved in post-stroke care should address risk factors aggressively to prevent future strokes.

Stroke is the fourth most common cause of death in the United States, behind heart disease, cancer, and chronic lower respiratory disease. Approximately 795,000 strokes occur per year in the United States, resulting in 135,000 deaths; approximately 610,000 of these strokes are initial incidents, and 185,000 are recurrent. Stroke is also the leading cause of long-term disability in the United States. Among elderly survivors 6 months post-stroke, approximately 50% have residual hemiparesis and 30% require assistance with activities of daily living or walking.

In 87% of patients, strokes are ischemic in origin, caused by occlusion of a blood vessel or other restriction of blood flow to a specific region of the brain. *Thrombotic infarcts* are the most common and are usually caused by occlusion of large cerebral arteries or their branches, most often as a result of atherosclerosis and progressive stenosis. *Embolic infarcts* are a subset of thrombotic infarcts and occur from distal passage of a thrombus or plaque from the heart, aortic arch, or proximal carotid arteries. *Lacunar infarcts* result from occlusion of small penetrating branches of cerebral arteries, often those that supply the basal ganglia, thalamus, internal capsule, and pons. They are often a consequence of atherosclerosis or degenerative changes in the arterial walls of these vessels secondary to longstanding hypertension or diabetes mellitus, or both. Up to half of single lacunar infarcts are clinically silent.

*Hemorrhagic strokes* (intracranial hemorrhages) account for 13% of all strokes. 3% of strokes are *subarachnoid hemorrhages* (SAHs), which usually result from rupture of intracranial arterial aneurysms. *Intracerebral hemorrhages* (ICHs), also called *intraparenchymal hemorrhages*, account for 10% of all strokes; they are often associated with hypertension and sometimes result from rupture of a vascular anomaly, such as an arteriovenous malformation. Intraventricular hemorrhage (IVH) can occur with any of these incidents.

PATHOGENESIS
Risk factors for stroke can be divided into nonmodifiable and modifiable factors. The modifiable risk factors encompass those associated with certain health habits and lifestyle-influenced diseases, reinforcing the central role of preventive efforts in reducing stroke morbidity and mortality. The major modifiable risk factors are high blood pressure, disorders of heart rhythm, smoking tobacco, dyslipidemias, physical inactivity, diabetes mellitus, sleep apnea, and chronic renal disease. Identification and management of these and other modifiable factors is crucial to the primary and secondary prevention of stroke and needs to be incorporated into the medical care of all stroke survivors undergoing rehabilitation.

Nonmodifiable Risk Factors

Nonmodifiable risk factors are age, sex, race, geographic location, and heredity. The risk of stroke doubles every decade after 55 years of age. Females have a higher lifelong but lower age-adjusted stroke incidence than men (except over 85 years of age). Blacks have almost twice the risk and Hispanics have approximately 1.5 times the risk of stroke compared with non-Hispanic whites. There is a substantially higher incidence in the southeastern United States. Family history is important, as documented ischemic stroke in a parent before age 65 years is associated with a threefold increased risk of stroke. The presence of preexisting arterial disease in any form is also a nonmodifiable risk factor; stroke and coronary artery disease share many risk factors contributing to atherosclerosis; therefore, experiencing one increases the risk of developing the other.


Modifiable Risk Factors

A. Ischemic Stroke: Thrombotic Strokes

1. Atherosclerosis—Hypertension is the most common modifiable risk factor
for both ischemic and hemorrhagic stroke. In ischemic stroke victims, it likely accelerates the development of atherosclerosis and formation of atheromatous plaques. The plaque itself can result in stenosis and subsequent ischemia or provide a source of emboli, causing occlusion distal to the site of the lesion. More than 77% of patients who experience a first CVA have a blood pressure greater than 140/90 mm Hg. Diastolic hypertension may increase the risk of stroke up to seven fold. Hence, lowering blood pressure below 140/90 mm Hg (135/85 mm Hg for patients with added risk factors, such as diabetes and renal disease) results in a significant reduction in risk over time. Subjects with pressures less than 120/80 mm Hg have approximately half the lifetime risk of their hypertensive counterparts. However, especially during early rehabilitation started within several days of a stroke, blood pressure should be lowered cautiously, paying attention to neurologic status and performance in therapy, as rapid reduction risks clinical worsening and even enlargement of the ischemic stroke. Vasomotor instability can occur immediately following a stroke; the accompanying postural hypotension adds to that risk and should be vigilantly avoided.

Other important risk factors to address during rehabilitation, including diabetes mellitus, hyperlipidemia, and cigarette smoking, have been implicated in accelerated atherosclerosis. Smokers have two- to fourfold increased risk of CVA, and cessation has been shown to reduce that risk over time. The presence of diabetes mellitus nearly triples the risk of stroke; however, no conclusive evidence links tight blood glucose control to a decreased risk of stroke. Hyperlipidemia increases the risk of both heart disease and stroke. Treatment with HMG-CoA reductase inhibitors (statins) can reduce this risk in individuals with coronary artery disease and diabetes mellitus by up to 25%. An elevated high-density lipoprotein level is also protective. Underlying contributing factors to diabetes and hyperlipidemia, such as obesity and inactivity, themselves risk factors, should also be addressed. A creatinine level greater than 1.5 mg/dL has been associated with an elevated risk, likely due to accelerated atherosclerosis.

Antiplatelet therapy is advocated for primary and secondary prevention of stroke in patients with known atherosclerotic disease or a history of noncardioembolic ischemic stroke or TIA. Low-dose aspirin (81–325 mg daily) or clopidogrel is usually prescribed for primary and secondary prevention. Inpatients who suffer strokes while taking aspirin alone, secondary prevention with clopidogrel or the combination of aspirin and extended-release dipyridamole is usually initiated. Combining aspirin and clopidogrel may reduce
the risk of further strokes but may also increase hemorrhage risk and is not routinely recommended unless the patient has other indications (ie, acute coronary syndrome or coronary stents). Cilostazol is used (off label) for secondary stroke prevention and has proven efficacy in patients of Asian origin, while ticagrelor is used to prevent thrombotic events after acute coronary syndromes.

Atherosclerosis often affects the carotid artery, especially near the carotid bulb. This and other areas may be subject to shear stresses from turbulent flow, resulting in a predilection to develop stenosis. A carotid bruit during physical examination may indicate carotid stenosis, although it does not necessarily correlate with severity. Studies have shown that **symptomatic carotid stenosis** (> 70%) has been associated with a substantial increased risk of ipsilateral and contralateral stroke and warrants referral for carotid endarterectomy. Although the risk of acute perioperative stroke or death in patients who undergo the procedure is 1.9% higher than in patients receiving medical therapy, the absolute risk reduction of endarterectomy over the next 5 years is 5.9% (55% relative risk reduction). Less-invasive endovascular angioplasty and stenting can also be considered, but this option remains controversial.


Willey JZ, Xu Q, Boden-Albala B, et al: Lipid profile components and risk of
2. Other vascular disorders—A variety of disorders can affect circulation to the brain. Autoimmune inflammatory vasculopathies produce changes in the vessel walls that may stimulate platelet adhesion and aggregation, resulting in thrombosis and distal embolism. It is vital that these vasculopathies be diagnosed and treated aggressively to prevent further strokes. With the exception of giant cell arteritis (temporal arteritis), these disorders usually affect small to medium-sized cerebral vessels.

Carotid and vertebrobasilar artery dissections can be post-traumatic or spontaneous and may lead to thrombus formation, arterial occlusion by narrowing of the vessel lumen, and embolization. Dissections should be treated emergently with therapeutic parenteral anticoagulation to prevent clot propagation and embolization distally. Migraines with aura (classic migraines) increase the risk of thrombotic stroke; sometimes the stroke can occur in the same vascular territory that produced transient neurologic symptoms during previous migraine attacks. Often, the role of migraine is questioned if there are other coexisting risk factors, especially smoking or estrogen use. Venous sinus thrombosis is often associated with otitis or sinusitis or a hypercoagulable state; anticoagulation is recommended.

Cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is the most common hereditary stroke disorder and is thought to be caused by a mutation of the Notch 3 gene on chromosome 19. It is characterized by a progressive degeneration of the cerebral and extracerebral vascular smooth muscle cells leading to migraine headaches, TIAs, and strokes; the latter usually occur between 40 and 50 years of age. Fibromuscular dysplasia is usually an autosomal-dominant disorder characterized by fibrous changes to arterial structures, which may predispose children and young adults to thrombus formation and embolic strokes. Moyamoya disease is sometimes inherited as an autosomal-recessive disorder but also occurs in patients with severe atherosclerosis. It is characterized by progressive stenosis of the distal carotid arteries along with the adjacent middle cerebral artery branches and the presence of a fine network of collateral vessels at the base of the brain. This leads to the development of multiple arteriovenous malformations that can steal blood from surrounding tissue leading to infarction, or rupture and bleeding.
3. Hematologic disorders—Sickle cell anemia, hypercoagulable states, thrombocytosis, polycythemia, and leukemias can all predispose a patient to cerebral ischemia. **Sickle cell anemia** typically affects the internal carotid artery or proximal portion of the middle cerebral or anterior cerebral artery. **Hypercoagulable conditions** include hereditary coagulopathies (factor V Leiden gene, protein C or S deficiency, and others) and acquired conditions, such as oral contraceptive use, cancer and the postpartum, post-traumatic or postsurgical state. **Oral contraceptives** in even low doses are associated with higher risk of stroke; additional risk factors such as smoking or migraine headaches, magnify this risk enormously.

4. Tobacco, drug, and alcohol abuse—Tobacco use is a major risk factor associated with ischemic stroke because of its role in accelerating atherosclerosis. Alcohol in moderation (less than 2 drinks per day) has been shown to cut the risk of stroke in half, whereas excess alcohol intake (greater than 5 drinks per day) can triple the risk of stroke. Insufflation of cocaine hydrochloride or smoking alkaloid (“crack”) cocaine can cause vasospasm resulting in an ischemic stroke. Intravenous abuse of heroin or other drugs predisposes individuals to infective endocarditis and consequent embolic strokes. Amphetamines, ephedrine, and phenylpropanolamine are stimulants that can raise blood pressure precipitously; these drugs are more commonly associated with ICH but may also cause vasospasm and cerebral ischemia.

5. Other risk factors—Sleep apnea, physical inactivity, and pregnancy are also associated with an increased incidence of stroke. **Sleep apnea** has been shown to increase the risk two- to fourfold, depending on severity. **Physical inactivity** is linked to multiple other risk factors for stroke but may itself be independently associated with stroke. More ischemic strokes occur in the winter months when people are less active, and moderate to vigorous intensity exercise is associated with a lower incidence of stroke. **Pregnancy** without the presence of preeclampsia is a risk factor for both ischemic and hemorrhagic strokes, especially in blacks and older women. Postpartum strokes are associated with cardiovascular disease, but not strokes that occur before delivery. Stroke during pregnancy usually necessitates termination of the pregnancy to enable aggressive treatment of the mother.

B. Embolic (Cardioembolic) Stroke: Cardiac Disorders

Patients with atrial fibrillation, especially those with valvular disease, have a markedly increased chance of suffering an embolic stroke each year. Most will require lifetime therapeutic anticoagulation, which often is started or continued during acute rehabilitation. Warfarin (INR goal of 2–3) has been shown to decrease the risk of stroke by 61% in patients with atrial fibrillation. Dabigatran, a direct thrombin inhibitor, and rivaroxaban and apixaban, direct factor Xa inhibitors, also reduce the risk of stroke from nonvalvular atrial fibrillation. When anticoagulation is contraindicated, antiplatelet agents are recommended even though they are inferior to warfarin for embolic prophylaxis. Dilated cardiomyopathy and myocardial infarction are associated with intracardiac mural thrombus formation leading to cerebral embolism. The risk of development of mural thrombus correlates with the size of the myocardial infarct; hence patients with large cardiac lesions may require anticoagulation. Structural heart diseases, particularly mitral valve stenosis and prolapse, have also been associated with increased risk but usually do not warrant prophylactic anticoagulation. Mechanical prosthetic heart valves, however, are highly prone to platelet adhesion and thrombus formation and therefore necessitate long-term anticoagulation therapy with warfarin (INR goal of 2.5–3.5). Left ventricular assistive device implantation is also associated with elevated stroke risk, which may be further increased by postoperative infections.

Theoretically, atrial septal defect and patent foramen ovale allow for blood flow between the right and left sides of the heart and may allow for passage of embolic material from the venous circulation in the extremities to the brain. Thus, a deep venous thrombosis in the leg may give rise to a “paradoxical” embolic CVA. Conditions that increase right heart pressures, such as multiple pulmonary emboli and severe obstructive sleep apnea, can increase the flow of blood through the septal opening. Intravascular repair of these defects is sometimes considered in patients with cryptogenic stroke who are found to have a patent foramen ovale; however, repair is not proven to reduce future stroke risk.
Both infective and marantic endocarditis can lead to formation of vegetations on the valve leaflets, which may also lead to thromboembolic events. Infective endocarditis is usually seen in intravenous drug users and patients with valvular disease or prosthetic valves and can be bacterial or fungal in etiology. Prolonged antibiotic treatment is critical. Marantic endocarditis is typically seen in cancer patients, most often those with adenocarcinomas of the lung or gastrointestinal tract. Anticoagulation should be strongly considered in these patients.


C. Hemorrhagic Stroke

1. Intracerebral hemorrhage—ICH is usually associated with hypertension. Common affected areas include the basal ganglia, thalamus, pons, cerebellum, and occipital lobes. In older patients, particularly when hemorrhage occurs in other areas, the most common cause is thought to be angiodysplasia from cerebral amyloid angiopathy. These individuals often have lobar hemorrhages at multiple sites.

Other causes of ICH are wide and varied. Heavy alcohol intake raises the risk of ICH in selected populations. Vascular malformations such as cerebral angiomas and aneurysms may also cause other symptoms, such as seizures and headaches, prior to any actual bleeding. The initial presentation of some ischemic strokes, especially embolic, may be accompanied by hemorrhagic transformation, which may lead to clinical worsening. Patients with coagulation deficits, such as hemophilia and von Willebrand’s disease, and those receiving anticoagulation therapy are at an increased risk for developing spontaneous ICH. Intravenous tissue plasminogen activator (tPA) administered acutely within 4.5
hours of stroke onset is associated with a 7% risk of symptomatic ICH. Vascular malformations and primary or metastatic brain tumors also predispose patients to ICH. In general, all ICH of uncertain etiology warrants follow-up contrast imaging to rule out tumors or other mass lesions obscured by acute blood on initial evaluation.


2. Subarachnoid hemorrhage—SAHs usually result from a ruptured saccular (berry) aneurysm or arteriovenous malformation, but can also be due to trauma. The classic presenting symptom is sudden onset of an unusually severe headache followed frequently by vomiting, neck stiffness, and collapse with loss of consciousness. Since bleeding occurs mainly in the subarachnoid space, prominent focal neurologic deficits are less common on neurologic examination. One exception is an aneurysm of the posterior communicating artery or middle cerebral artery, which may result in ipsilateral cranial nerve III palsy from local compression. Associated vasospasm can cause focal infarctions, which greatly complicate acute symptoms and eventual disability. Intracranial blood can be very irritating to surrounding neural tissues and is associated with cerebral edema. Symptoms vary considerably, depending on the amount of cerebral edema surrounding the hemorrhage, and the capacity of the brain to accommodate the edema.

Ischemic strokes due to thrombosis or emboli often produce focal neurologic deficits that correlate with the region of the brain supplied by the underlying vascular lesion. A thorough neurologic examination can often localize the lesion. However, physical examination alone cannot differentiate between hemorrhagic and thrombotic stroke; thus, imaging studies are needed acutely. The vascular territories and usual neurologic examination findings for major large vessel cerebral infarctions are shown in Table 14–1. Major brainstem stroke syndromes are listed in Table 14–2. Lacunar infarcts in the brain result from occlusion of small penetrating branches of the major cerebral arteries and are frequently clinically silent and incidental findings on brain imaging. Although neurologic deficits can vary widely, five distinctive lacunar syndromes that occur frequently have been identified (Table 14–3). ICH is more variable in presentation owing to variations in size and location, particularly the lobar hemorrhages. Pontine, cerebellar, and deep cerebral (putamen and thalamic) hemorrhages often causes omnolence or coma because of their location proximal to the reticular activating system. Hemorrhages in the basal ganglia cause hemiparesis and, if sizable or surrounded by edema, can cause hemianopia, inattention, or aphasia. Thalamic hemorrhages usually cause hemisensory deficits. Brainstem hemorrhages can cause hemiparesis or quadriplegia, severe dysphagia, dysarthria, and gaze deficits. Cerebellar hemorrhages can cause ataxia of the trunk or limbs and vertigo.

Table 14–1 Possible symptoms of strokes involving major cerebral blood vessels.
<table>
<thead>
<tr>
<th>Vascular Territory and Possible Areas Damaged</th>
<th>Possible Neurologic Deficits</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Anterior cerebral artery</strong>&lt;br&gt;(medial aspect of frontal or parietal lobes)</td>
<td>Contralateral hemiparesis of the leg&lt;br&gt;Contralateral hemisensory deficits in leg&lt;br&gt;Impaired bladder inhibition&lt;br&gt;Personality changes&lt;br&gt;Ideomotor apraxia&lt;br&gt;Abulia&lt;br&gt;Gegenhalten rigidity, alien arm or hand</td>
</tr>
<tr>
<td>Dominant hemisphere</td>
<td>Transcortical motor or mixed aphasia</td>
</tr>
<tr>
<td>Severe damage or bilateral lesions</td>
<td>Akinetic mutism&lt;br&gt;Paraplegia</td>
</tr>
<tr>
<td><strong>Superior division of MCA</strong>&lt;br&gt;(lateral aspect of frontal or frontoparietal areas)</td>
<td>Contralateral hemiparesis of the face, hand, and arm &gt; leg&lt;br&gt;Contralateral hemisensory deficits in the face, hand, and arm &gt; leg</td>
</tr>
<tr>
<td>Dominant hemisphere</td>
<td>Expressive (Broca’s) aphasia</td>
</tr>
<tr>
<td>Nondominant hemisphere</td>
<td>Hemineglect, constructional and dressing apraxia</td>
</tr>
<tr>
<td><strong>Inferior division of MCA</strong>&lt;br&gt;(lateral aspect of posterior parietal or temporoparietal areas)</td>
<td>Contralateral homonymous hemianopsia&lt;br&gt;Homonymous quadrantanopsia&lt;br&gt;Contralateral graphesthesia and stereognosis&lt;br&gt;Anosognosia&lt;br&gt;Dressing and constructional apraxia</td>
</tr>
<tr>
<td>Dominant hemisphere</td>
<td>Receptive (Wernicke’s) aphasia</td>
</tr>
<tr>
<td>Nondominant hemisphere</td>
<td>Left visual neglect</td>
</tr>
<tr>
<td>Bilateral lesions</td>
<td>Simultagnosia</td>
</tr>
<tr>
<td><strong>Posterior cerebral artery</strong>&lt;br&gt;(temporo-occipital or occipital areas)</td>
<td>Contralateral homonymous hemianopsia&lt;br&gt;Vertical gaze palsy&lt;br&gt;Oculomotor nerve palsy&lt;br&gt;Ataxia&lt;br&gt;Internuclear ophthalmoplegia&lt;br&gt;Anomic or transcortical receptive aphasia&lt;br&gt;Alexia without agraphia&lt;br&gt;Visual agnosia&lt;br&gt;Prosopagnosia</td>
</tr>
<tr>
<td>Bilateral lesions</td>
<td>Cortical blindness, memory impairment&lt;br&gt;Simultagnosia</td>
</tr>
<tr>
<td><strong>Basilar artery branches</strong>&lt;br&gt;(brainstem or cerebellum)</td>
<td>Unilateral or bilateral abducens nerve palsy&lt;br&gt;Impaired horizontal eye movements&lt;br&gt;Ipsilateral hemiparesis or quadriplegia&lt;br&gt;Ipsilateral hemisensory deficits&lt;br&gt;Balance disturbance&lt;br&gt;Contralateral ataxia&lt;br&gt;Dysarthria&lt;br&gt;Dysphagia</td>
</tr>
<tr>
<td><strong>Bilateral basilar artery branches</strong>&lt;br&gt;(Ventral portion of pons is infarcted and tegmentum spared)</td>
<td>Locked-in syndrome: Only eye opening, vertical eye movements, pain, and temperature sensation spared&lt;br&gt;Quadriplegia&lt;br&gt;Mutism</td>
</tr>
</tbody>
</table>

MCA, middle cerebral artery.
Table 14–2 Major brainstem stroke syndromes.

<table>
<thead>
<tr>
<th>Vascular Territory</th>
<th>Neurologic Deficits</th>
</tr>
</thead>
</table>
| Weber syndrome (occlusion of interpeduncular branches of posterior cerebral or posterior choroidal artery to base of medial midbrain) | Ipsilateral oculomotor palsy  
Contralateral hemiplegia                                                              |
| Benedikt syndrome (occlusion of interpeduncular branches of posterior cerebral or basilar artery to lateral midbrain)  | Ipsilateral oculomotor nerve palsy and mydriasis  
Contralateral sensory loss  
Contralateral ataxia, tremor, chorea, and athetosis secondary to damage to red nucleus |
| Millard–Gubler syndrome (occlusion of circumferential branches of basilar artery to medial pons)  | Ipsilateral abducens and facial nerve palsy  
Contralateral hemiplegia  
Contralateral sensory loss                                                             |
| Occlusion of superior cerebellar artery to lateral portion of rostral pons         | Impaired optokinetic nystagmus  
Contralateral loss of vibration, position, pain, temperature, and tactile sensation |
| Occlusion of anterior inferior cerebellar artery to lateral portion of caudal pons | Ipsilateral facial weakness  
Abducens nerve palsy  
Contralateral hemiplegia  
Contralateral sensory loss                                                                 |
| Lateral medullary syndrome or Wallenberg syndrome  
(most cases due to vertebral artery or posterior inferior cerebellar artery infraction) | Ipsilateral cerebellar ataxia  
Ipsilateral Horner syndrome (ptosis, anhydrosis, and miosis)  
Impaired pain and temperature sensation of ipsilateral face and contralateral body  
Vocal cord paralysis, dysphagia, dysarthria  
Vertigo, nausea, vomiting, and hiccups  
Nystagmus or diplopia, or both                                                          |
| Medial medullary syndrome                                                          | Ipsilateral hypoglossal nerve palsy  
Contralateral hemiplegia  
Contralateral sensory loss                                                                 |
In addition to a neurologic examination, physical examination should also include a comparison of left and right blood pressures and pulse strength, auscultation of the neck for carotid bruits, careful cardiac examination for arrhythmias or murmurs, and (especially in those with headaches) palpation of the temporal arteries. An ophthalmoscopic examination can also be useful, especially in patients with hemorrhagic strokes or diabetes mellitus. Visual confrontation examination of the diabetic stroke patient should be performed with each eye separately.


**DIFFERENTIAL DIAGNOSIS**
Primary focal involvement of the cerebral circulation is what differentiates a stroke or TIA from other causes of acute central nervous system dysfunction such as multiple sclerosis, brain tumors, seizures, toxic or metabolic disturbances, and anoxic or traumatic brain injuries.

**TREATMENT**

Numerous advances have recently been made in the prevention, treatment, and rehabilitation of stroke. This has resulted in a significant reduction in the population-based death rate over the past decade. Once thought of as an incurable and static disease, stroke is now viewed as a treatable acute condition. In some dramatic cases, intervention with fibrinolytic therapy or mechanical thrombectomy has led to complete resolution of symptoms in days with apparent full recovery. Early interdisciplinary rehabilitation remains the primary recommended treatment for post-stroke disability.

**Rehabilitation**

Rehabilitation is a holistic process involving multiple health care professionals that should begin almost immediately after stroke to maximize the survivor’s potential for functional recovery. Even patients intubated in the intensive care unit should receive range-of-motion exercises and frequent repositioning in bed to prevent future complications such as decubitus ulcers and contractures that can further impede progress.

**A. Mechanisms of Stroke Recovery**

Some of the initial recovery from stroke is presumed to result from resolution of the penumbra or cerebral edema. The penumbra is the margin of reversible ischemia surrounding the infarcted core of the CVA. Edema around the lesion can impair brain function, and resolution of this response may lead to significant recovery. Following ICH and SAH, resorption of blood and resolution of associated cerebral edema are likely responsible for early clinical improvement. The axons of partially spared pathways may reinnervate or sprout over several months or longer, contributing to later recovery.

It is now widely accepted that with stimulation the brain has a great potential for neuroplasticity-induced changes after stroke. A large body of evidence has shown that the brain can often change and continue to regain function months or years after the initial event. These changes are associated with alterations in
neurotransmitters and their synaptic receptors, and much research has been
directed toward determining whether pharmacologic modulation of these
neurotransmitters can facilitate changes. Medications that have been studied
include those that affect noradrenergic, dopaminergic, cholinergic, and
serotonergic receptors. Recently published studies suggest that serotonin
reuptake inhibitors (SSRIs), widely used for prophylaxis or treatment of post-
stroke depression, may facilitate motor recovery. The evidence supporting the
benefits of the other classes of medication has been weaker.

acute ischaemic stroke (FLAME): A randomised placebo-controlled trial.

Dombovy ML, Aggarwal U: Stroke rehabilitation. In Grabois M, Garrison SJ,
Hart KA, Lehmkuhl LD (Eds): Physical Medicine and Rehabilitation: The

**B. Goals of Acute and Postacute Rehabilitation**

The basic goal of intensive post-stroke rehabilitation is to maximize
independence, and the vast majority of stroke survivors will respond to intensive
interdisciplinary rehabilitation by making gains. However, some rehabilitation
hospitals and units restrict access to such intensive treatment for stroke patients
who lack realistic goals. For example, a patient who lives alone and suffers a
severe right brain stroke with residual perceptual and motor weakness and likely
permanent need for assistance may have the unrealistic goal of returning home
independently. Such a patient may be rejected for admission to some units,
especially if resources for placement (in custodial nursing facilities) are lacking.
It should be understood that such a patient would likely make significant gains in
intensive rehabilitation and ultimately require substantially less assistance after
discharge than if sent directly from an acute hospital to a nursing facility. Such a
reduction in the amount (and cost) of care may justify admission to intensive
rehabilitation (but the patient would have to agree, at some point, to change his
or her discharge goal).

Generally, acute stroke patients are evaluated in the acute care hospital by a
physical therapist or rehabilitation physician, who determines the level of
mobility impairment and the ability to tolerate therapy. Self-care deficits are
identified by nursing and, when needed, occupational therapy providers.
Occupational therapy participation is particularly important in the acute hospital when evaluating the patient for whom discharge home is a consideration. Assessment by a speech therapist is important acutely to determine whether dysphagia is severe enough to warrant nothing-by-mouth (NPO) status, a modified consistency diet, or enteral feeding.


C. Alternatives to Acute Intensive Inpatient Rehabilitation

The term subacute care has been applied to a broad range of medical and rehabilitative services and settings that provide care to patients following acute management in a hospital setting. Medicare has five levels of skilled nursing facility rehabilitation. The two most intensive of these are considered subacute level of care, providing 1.5–2.5 hours of therapy per day, 5 days per week. For Medicare to pay for skilled nursing facility rehabilitation, the patient must have had a 3-day inpatient hospitalization in the past 30 days (observation status is not included) and require skilled nursing or therapy services.

A long-term acute care hospital is a hospital that specializes in the medical treatment (and sometimes rehabilitation) of medically complex patients who require an extended stay in a hospital setting; often the medical focus involves weaning patients from ventilators. Some stroke patients referred for acute intensive rehabilitation who have potentially unstable medical problems should be considered for treatment in a long-term acute care hospital; once the complex medical problems stabilize and the patient demonstrates consistent tolerance of therapies, transfer to an acute intensive rehabilitation unit may be appropriate.

Studies have shown that for Medicare fee-for-service beneficiaries needing rehabilitation after stroke, comprehensive treatment in intensive rehabilitation hospitals leads to, on average, greater functional improvement and more frequent community discharge than patients receiving subacute rehabilitation services at skilled nursing facilities.

D. General Medical Management and Preventive Care

1. Deep vein thrombosis (DVT) and pulmonary embolism—DVT occurs in the hemiparetic lower extremity 60–75% of the time. In the vast majority of stroke patients, DVT is not painful; only 3–5% have clinical symptoms (limb swelling, fever, pain, or a combination). DVT risk factors include advanced age, severe limb paresis, immobility, dehydration, previous venous thrombosis, and the presence of neoplasm. Pulmonary embolism occurs in 1–2% of stroke patients. Half of patients with symptomatic pulmonary embolism die at presentation; thus, it is now a standard of medical practice that thromboembolic prophylaxis be initiated or otherwise addressed for all acute stroke patients and continued during intensive rehabilitation.

DVT is diagnosed by ultrasonography, impedance plethysmography, contrast venography, or D-dimer assays. Currently, diagnosis is usually made by duplex ultrasonography, which is more sensitive and specific in the thigh than in the calf. For pulmonary embolism, pulmonary angiography is most sensitive and specific, but radionuclide ventilation perfusion scan is also used. 90% of pulmonary emboli are associated with proximal lower limb DVT. Prophylaxis includes subcutaneous heparin given every 8 hours, low-molecular-weight heparin, or intermittent pneumatic compression if patients have contraindications to prophylactic anticoagulation. If pneumatic compression devices are used it is essential to monitor the skin. It is generally safe to combine antiplatelet therapy with prophylactic anticoagulation.

The goal of therapeutic treatment of acute DVT is to prevent extension and propagation of the clot. If anticoagulation is contraindicated, an inferior vena cava filter should be considered. These filters have an acceptable safety profile in stroke patients. If possible, therapeutic anticoagulation should be started later, to limit clot propagation. If the patient lacks other procoagulant risk factors and is likely to regain ambulatory ability, a retrievable interior vena caval filter should be inserted.

Screening all patients for DVT at the time of admission to the rehabilitation unit is controversial, largely because most clots diagnosed will be in the calf and may not warrant any therapeutic treatment. Screening is more strongly indicated if the patient has had no prophylaxis or a gap in prophylaxis prior to admission.
A calf clot should be followed with a repeat study within 1 week, to determine if the clot is propagating; if so, therapeutic anticoagulation is indicated.


2. Cardiac precautions—Acute exacerbations of cardiac-related disease can occur during postacute stroke rehabilitation. Common problems include angina, uncontrolled hypertension, hypotension, myocardial infarction, congestive heart failure, atrial fibrillation, and ventricular arrhythmia. Less common is neurocardiogenic injury, which can cause electrocardiographic changes and troponin elevation. Clinical signs include slower-than-anticipated progress, excessive fatigue, lethargy, and mental status changes. Cardiac precautions should be communicated to all therapies for all patients with known or suspected cardiac disease. For all patients, activity should be terminated and the patient evaluated urgently if new-onset cardiopulmonary symptoms occur. Otherwise, cardiac precautions may include pulse and blood pressure parameters, such as avoidance of blood pressure greater than 170/95 or pulse greater than 75% of the roughly calculated maximal heart rate (220 minus age). Such values do not represent target vital sign parameters; rather, therapists should allow the patient to rest if the parameters are exceeded. If parameters are consistently exceeded, the physician should reevaluate them or adjust medication.
For patients with more unstable or recent-onset cardiac disease (eg, myocardial infarction at the time of the acute stroke), more stringent cardiac precautions should be used: pulse and blood pressure should be taken at the beginning of each therapy session, and therapy should be halted if the heart rate decreases more than 10 beats per minute below baseline or increases more than 20 beats above baseline, if systolic blood pressure decreases more than 20 mm Hg below baseline or is less than 90 mm Hg, or if diastolic blood pressure increases more than 20 mm Hg.

Stroke and coronary artery disease are interrelated comorbidly. The risk of acute stroke is increased after myocardial infarction (highest in the 3 months following the event). One third of stroke patients have coronary artery disease, and 3% of stroke patients die from cardiac events within 3 months of the stroke. In addition, stroke may occasionally be the presenting sign of a myocardial infarction.

3. **Hypertension**—Hypertension is a major risk factor for recurrent strokes, and it behooves the rehabilitation physician to address this serious problem for all post-stroke patients. The general goal is to normalize blood pressure over time. During the first week after stroke onset, blood pressure should be gradually lowered to normal levels (< 140/90 mm Hg). For patients admitted to the rehabilitation unit within that first week, the rehabilitation physician may need to accept a modestly high blood pressure at first, then gradually increase antihypertensive medication. Patients with extensive or severe focal intracranial atherosclerosis and stenosis may require more prolonged maintenance of “permissive” hypertension. At the time of inpatient rehabilitation admission, it is important for the rehabilitation physician to review and follow the recommendations of the referring acute care physician.

Any time antihypertensive medication is adjusted, blood pressure should be checked more than once daily by nurses, and at least once daily during therapies. The patient should be observed carefully for signs of neurologic deterioration. Hypotensive episodes need to be avoided and generally warrant immediate reduction in medication dosage. Dehydration will compound the hypotensive effects of medication, and intravenous fluids should be urgently considered if this is suspected.

Normalizing blood pressure is important to reduce the long-term risk of another stroke. In patients who are otherwise stable and asymptomatic, transient episodes of hypertension, even at severe levels, raise the risk of stroke minimally, whereas sudden large reductions of blood pressure can precipitate
syncope or aggravate cerebral ischemia acutely. Short-acting antihypertensives, particularly sublingual calcium channel blockers, should therefore be avoided. Hydralazine or labetalol given in low doses every 6–8 hours may be helpful in slowly bringing down excessively high blood pressure.


4. Respiratory care, aspiration from dysphagia, and pneumonia—Aspiration commonly occurs after stroke as a result of impaired swallowing and failure of protective reflexes. Risk factors for aspiration pneumonia are decreased level of consciousness, presence of a tracheostomy, emesis, reflux, nasogastric tube feeding, and dysphagia of any kind. Dysphagia may be associated with an increase in the normal aspiration of oral and airway secretions. Aspiration of those secretions or regurgitated gastrointestinal contents can cause aspiration pneumonia or chemical pneumonitis.

Stroke patients with cough and fever should be examined closely for the development of congestion or clinical pneumonia and undergo a chest radiograph. Aspiration, which occurs in as many as 70% of patients, can be completely silent and is missed on bedside swallowing evaluations in 40–60% of patients. Predictors of aspiration include abnormal cough, dysphonia, dysarthria, and a wet voice quality after swallow. Predictors of aspiration studied with video fluoroscopic swallowing study (VFSS) include delayed initiation of swallow, decreased pharyngeal peristalsis, or both.

A patient who fails a bedside screening test on the rehabilitation unit should not be allowed to eat until a speech therapist has deemed it safe to proceed with a modified diet. Until then, the patient may require feedings through a nasogastric or percutaneous endoscopic gastrostomy (PEG) tube.

5. Tracheostomy—Severe neurologic deficits from stroke may initially be associated with cardiorespiratory collapse, necessitating intubation. Prolonged intubation, in turn, mandates placement of a tracheostomy (see Chapter 38). Patients who are admitted to an intensive rehabilitation unit after such events are generally debilitated from prolonged illness and lacking in respiratory muscle strength and stamina. Pulmonary congestion, often residual from pulmonary complications shortly after the stroke, must be treated aggressively (ie, using chest physiotherapy, nebulizer treatments, and suctioning). Complications that
may result from a tracheostomy include tracheoesophageal fistula, pneumothorax, tracheal or subglottic stenosis, tracheomalacia, or injury to the recurrent laryngeal nerve. It is important to rule out these problems before attempting to plug the tracheostomy tube. For this reason, an ear–nose–throat consultation is recommended before plugging.

Decannulation may also be difficult owing to subjective dyspnea related to respiratory muscle weakness. The general approach is to reduce tube size (at least to no. 6, and, for a small female, no. 4), and then plug the tracheostomy. Cuffed tubing must be replaced with uncuffed tubing if possible. A helpful intermediate step is to use a Passy-Muir valve, which permits inhalation through the tracheostomy tube, and exhalation through the mouth and past the vocal cords, enabling phonation (see Chapter 38). Oxygen saturation should be monitored regularly (every 2 hours is recommended) When the patient demonstrates tolerance of plugging for more than 24 hours (without unplugging), the tracheostomy tube can generally be removed.


6. Gastrointestinal precautions—Gastrointestinal bleeding occurs in 3–5% of acute stroke patients. Stroke patients should receive gastrointestinal prophylaxis if they are NPO, receiving tube feedings, or have a history of gastric bleeding, gastroesophageal reflux disease, or peptic ulcer disease. According to the American College of Gastroenterology, American College of Cardiology Foundation, and the American Heart Association Expert Consensus Panel, patients using antiplatelet medications or nonsteroidal antiinflammatory drugs should be considered for long-term gastrointestinal prophylaxis due to risk of bleeding from antiplatelet agents. Patients using oral corticosteroids, aspirin or dual antiplatelet therapy, or anticoagulation therapy should be prescribed a proton pump inhibitor, especially if aged 60 years or older.

7. **Nutrition**—For the patient receiving tube feedings, several options are available. Bolus feedings are the most physiologic and can be given at usual meal times, if tolerated without eructation or vomiting. If continuous feedings are needed, they should be given during evenings and overnight to avoid interference with daytime therapies. As patients tolerate increasing amounts of oral food, tube feedings should be gradually decreased. Calorie counts of oral food should be checked periodically to assure adequate enteral supplementation. Reduction of tube feedings usually encourages appetite and enhances intake of oral food.

Complications of dysphagia include dehydration and malnutrition. Hypoalbuminemia and other signs of malnourishment are common in patients admitted to rehabilitation units and correlate with a prolonged length of stay, slower rate of functional gain, higher risk of infection, and decubitus ulcers. Experimental studies have found that protein synthesis is suppressed in the ischemic penumbra. Small studies have shown that inadequate protein intake, low zinc intake, and low antioxidant capacity are associated with expansion of ischemia-induced brain damage. A PEG tube should be seriously considered if a patient is unable to tolerate adequate amounts of thickened liquid and a pureed diet by 5–7 days post-stroke (or, if NPO, by 24–48 hours). Hypoalbuminemia may respond to protein supplementation orally, or by PEG or nasogastric tube.

Patients taking thickened liquids are also at risk of dehydration and azotemia; if these complications occur, short courses of intravenous fluid supplementation may be appropriate. If supported nutritionally, most patients with severe dysphagia will recover to the point at which an oral diet can be tolerated. Recovery of swallowing function in most patients with brainstem strokes usually occurs in the first 3 weeks post-stroke.
8. Bowel and bladder management—Although the incidence of bowel incontinence in stroke patients is 31%, it typically resolves in the first 2 weeks; however, if brain damage is severe, bowel incontinence may persist. Incontinence may also be aggravated or caused by diarrhea or genitourinary infection, inability to transfer or manage clothing, or communication impairments. Sometimes a timed toileting schedule (ie, after each meal) helps. Hydration, fiber, stool softeners, and cathartics may be necessary for constipation. Suppositories and small enemas can often help to time defecation.

Urinary incontinence occurs in 50–70% of patients during the first month after stroke and in 15% after 6 months. Following most strokes, cortical inhibition of the bladder is reduced and patients experience increased urgency, which occurs at lower filling volumes than before the stroke. However, the pontine micturition center is usually preserved; therefore, reflex voiding typically has normal synchrony of the internal sphincter relaxation and detrusor contraction. Reasons for urinary incontinence include severe central nervous system damage, genitourinary infection, fecal impaction, difficulty with transfers, communication deficits, confusion, and misperception of bladder sensation.

All post-stroke patients are at risk of incomplete bladder emptying and should have measurement of postvoid residual (PVR) volume of urine. It is vital to differentiate incontinence caused by an overflowing areflexic bladder from an adequately emptying hypertonic or uninhibited bladder. The former produces a large PVR, while the latter results in a small PVR. Measurement of PVR can now be done with a bedside portable ultrasonic machine (Bladderscan). Patients with large PVRs (> 250 mL) require intermittent catheterization (or an indwelling catheter, if absolutely necessary). Diabetic patients may develop a sensory autonomic neuropathy leading to hypotonia and urinary retention. For patients with large PVRs, rectal examination helps to rule out fecal impaction or to identify a large obstructing prostate in males. α-Adrenergic blocking agents should be considered to relax the internal urinary sphincter and improve urinary outflow, and anticholinergic medications should be identified and discontinued where possible. If urinary retention persists, urologic consultation is recommended.

On the other hand, patients with urgency incontinence and small PVRs may be helped by timed voiding and judicious use of anticholinergics; if such patients
experience symptoms during only part of the day, short-acting anticholinergics to cover the time of bothersome symptoms, rather than time-release medications lasting 24 hours, should be used.

9. **Skin care and prevention of decubitus ulcers**—Post-stroke patients at greatest risk of pressure ulcers include those with mobility deficits, sensory impairments, compromised skin vascularity, urinary or fecal incontinence, frailty, low body mass, and malnutrition. Skin should be clean and dry and nutritional deficiencies addressed. Padded heel boots, heel-relieving ankle foot splints (Multi-Podus, manufactured by RCAI and others; PRAFO, by Anatomical Concepts and others), egg-crate mattresses, waterbeds, low-pressure hospital bed mattresses, and soft padding are helpful but do not replace the need to change the patient’s position to relieve pressure. This should take place every 30 minutes if the patient is sitting in a wheelchair, and at least every 2 hours when the patient is in bed. All post-stroke patients with mobility or sensory impairment should receive regular skin checks. Areas of particular concern include bony prominences. Decubitus ulcers that develop require pressure relief and, if necessary, surgical consultation.


10. **Contracture prevention**—Stroke patients often are flaccid immediately following their strokes, but over time experience gradually increasing tone in the hemiparetic side. Spastic tone usually affects the flexors of the upper extremity and extensors of the lower extremity more than their antagonists, leading to muscle imbalance and increasing the risk of developing contractures. Common sites of soft tissue contractures are listed in Table 14–4. The “muscles to stretch” are muscles that commonly have early return of tone and spasticity and therefore are at risk of tightening. The following joint ranges tend to be increasingly limited over time and require frequent evaluation by a therapist, nurse, or physician: shoulder abduction, flexion, and external rotation; finger, wrist, and elbow extension; and ankle dorsiflexion and eversion. Stroke survivors with more severe weakness are particularly at risk of developing contractures, which can limit movement, impede functional progress, and sometimes cause pain. Splints, serial casting, passive range-of-motion machines, robotic therapy, or
prolonged positioning can help limit contractures. It is vital that patients’
contracted limbs be moved through range of motion several times daily. On days
when there is no therapy, family members or nurses should implement these
stretching exercises. It should be noted that single or serial casting at the ankle,
knee, wrist, or elbow—may improve range of motion, but may not affect the level
of spasticity. Treatment of spasticity with medication or motor point blocks may
be necessary in order to perform effective stretching or splinting. It is the
clinician’s responsibility to monitor muscles at risk of shortening and to institute
more vigorous range of motion or other treatments if needed.

Table 14–4 Common sites of contracture in stroke patients, and muscles
requiring range of motion.a

<table>
<thead>
<tr>
<th>Joint</th>
<th>Direction to Stretch</th>
<th>Muscles to Stretch</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder</td>
<td>Flexion, abduction</td>
<td>Latissimus, pectoralis, teres major</td>
</tr>
<tr>
<td></td>
<td>External rotation</td>
<td>Subscapularis</td>
</tr>
<tr>
<td>Metacarpophalangeal and interphalangeal</td>
<td>Extension</td>
<td>All finger flexors, thumb flexors</td>
</tr>
<tr>
<td>Wrist</td>
<td>Extension</td>
<td>Flexor carpi radialis, flexor carpi ulnaris</td>
</tr>
<tr>
<td>Hip</td>
<td>Abduction</td>
<td>Adductors</td>
</tr>
<tr>
<td>Ankle</td>
<td>Dorsiflexion, Eversion</td>
<td>Gastrocnemius, soleus, tibialis posterior, tibialis anterior</td>
</tr>
</tbody>
</table>

aOther muscles may also need stretching.

In stroke patients, flexor spasticity involving the hips or knees may be a sign
of nociception in the lower extremities or pelvic area. The clinician should rule
out urinary tract infection, other rectal or pelvic pathology, ingrown toenail,
treatable arthritis, tissue damage of the pelvis or lower extremities, or fecal
impaction.
Reintroduction to the Community

A. Discharge Planning

Optimal transition of the moderately or severely impaired stroke survivor to the community is complex and often requires considerable preparation. All persons who will assist the stroke survivor after discharge should receive hands-on training by the therapists prior to discharge. Trained family members develop a more realistic outlook on the patient’s abilities and needs, and become more confident about their own capability to care for the patient. Regardless of severity, stroke patients who undergo comprehensive discharge planning and whose families undergo training are more likely to be discharged home.

It is also important for the rehabilitation physician to communicate with the patient’s primary care and other treating physicians. This sometimes requires direct telephone communication, particularly if there is an active medical problem that requires close followup. In most cases, providing the patient with copies of laboratory and radiology reports, a typed summary note of the medical and rehabilitation course since the stroke, and a full list of ongoing diagnoses, medical problems, and medications is sufficient; the patient and family should be given explicit instructions to arrange followup with the primary care physician within 1 week of discharge and to bring the medical papers to the physician’s office. A CD copy of the patient’s neurologic imaging studies is also helpful, particularly when patients are returning to areas distant from the rehabilitation facility. The rehabilitation physician should invite the patient and family to call with questions after discharge home.

B. Social Isolation and Driving

Driving can help maintain an individual’s independence, access to community resources, access to the place of employment, and overall quality of life. The loss of driving privileges may contribute to vocational disability, financial stress, loss of self-esteem, and social isolation. Evaluation of ability to drive after a stroke usually requires an on-road assessment, which may be expensive and is typically not covered by insurance. Psychological tests that assess multiple cognitive domains relevant to driving appear to have the best reproducibility in predicting fitness to drive in stroke patients, but no single battery of tests or off-road driving simulation perfectly predicts competence to drive on the road.

Motor, sensory, visual, or cognitive impairments can affect a person’s ability to drive after a stroke. Visuospatial and attention deficits, slowed motor
processing, homonymous hemianopia, and right cerebral hemisphere lesions most likely predict poor on-road driving ability. Strokes may also impede reaction time, judgment, and multitasking ability, all of which are essential for safe driving and the ability to respond emergently on the road. Stroke survivors with any residual deficits should, at discharge from inpatient care, be explicitly told not to drive until further recovery has taken place, and until they have been evaluated with an on-road assessment. A significant role for the physician is to determine if it is safe and appropriate for the patient to undergo the on-road assessment. The American Medical Association’s (AMA) Guide to Assessing and Counseling Older Drivers provides a useful brief battery of office tests that are applicable to stroke survivors, including the Trail Making B and Clock Drawing Tests.

Therapists can provide important objective functional status and observational data to the physician to help make that decision. Drivers of commercial vehicles will need additional evaluation by a commercial driving instructor before return to work. Patients who lack awareness of their deficits, exhibit poor judgment, have significant hemineglect, or who express the intention to drive after discharge despite counseling otherwise should be reported to their state motor vehicle department.

Persistent weakness of an arm or leg may require adaptive equipment for the vehicle, which mandates an evaluation by a certified driver rehabilitation specialist (CDRS). Equipment that can be helpful for stroke survivors includes spinner knobs for one-handed control of steering, right-to-left gas pedal linkage for right lower extremity weakness, and left-to right turn signal linkage for left arm weakness. Adaptive equipment not only requires installation, but also requires that the patient be trained and certified competent in its use.

It is important that the clinician become familiar with state or national regulations that specify medical requirements for driving, including visual fields, visual acuity, and required waiting time after seizure or other unpredictable change of consciousness. Visual deficits may require formal testing by an ophthalmologist, neuroophthalmologist, or optometrist to clear the patient for return to driving. Many states and countries also have legal reporting requirements and regulations and offer varying legal protections for physicians who report potentially impaired drivers. The National Highway Traffic Safety Administration and AMA maintain that information for the United States in the Guide to Assessing and Counseling Older Drivers.

If an individual cannot drive, he or she may be able to use public transportation independently or with assistance. Using public transportation
requires a skill set that warrants evaluation and training by a physical or occupational therapist as part of overall therapy for community integration (Table 14–5). If the patient has to walk to an access point to board a bus or train, it must be determined whether this can be done safely. Considerations include the width of the streets and the patient’s speed of ambulation, and the therapist may need to evaluate and train the patient outdoors. Many communities offer separate accessible vehicles to transport patients with a variety of disabilities, but a physician usually must certify patients’ need for these. Post-stroke patients with persistent mobility deficits often are limited socially by architectural barriers in buildings, such as steep steps required to exit a residence. Physicians should team with social workers to explore options and certify need for accessible housing or equipment such as stair glides to enable access to the community. If patients are truly unable to exit their homes, police and fire departments in their communities should be notified to be prepared to assist the patient in emergencies.

Table 14–5 Evaluation of issues related to stroke survivors’ use of public transportation.
<table>
<thead>
<tr>
<th>Physical</th>
<th>Cognitive</th>
<th>Communication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Is assistance required to exit the residence?</td>
<td>1. Can the patient find and recognize the point of access?</td>
<td>1. Can the patient communicate the destination to the driver or conductor?</td>
</tr>
<tr>
<td>2. Is assistance required to ambulate (or use wheelchair) to reach the point of access (bus or train stop)?</td>
<td>2. Once on the public conveyance, can the patient recognize the final destination?</td>
<td>2. Can the patient read maps or a schedule?</td>
</tr>
<tr>
<td>3. How wide are streets between the patient's home and access point?</td>
<td>3. Can the patient tolerate noise and crowding on a vehicle?</td>
<td>3. Can the patient communicate with other passengers to get to a seat or to the door to exit?</td>
</tr>
<tr>
<td>4. Is assistance needed to board the public conveyance?</td>
<td>4. Can the patient reliably and accurately pay for the trip?</td>
<td></td>
</tr>
<tr>
<td>5. Could the patient safely stand on the vehicle?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. What accommodations are available?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Kneeling bus, wheelchair lifts, dedicated transport vehicles, dedicated seats on vehicles)</td>
<td></td>
<td></td>
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</table>
C. Vocational Training

Approximately 40% of people return to work (RTW) after suffering a stroke. There is a direct correlation among RTW, age, and disability, with younger patients and less severe disability favored. Difficulty returning to work can have a significant impact on family relationships, level of intimacy, economic situation, and leisure activities of the stroke survivor. Negative prognostic factors for RTW include living alone, severe functional impairment, and speech disorders. Positive prognostic factors include professional support and early involvement of an occupational physician. There is a positive correlation between RTW and the time of resumption of driving. Education and income are independent predictors of RTW among stroke patients during the first post-stroke years.

The stroke patient who improves to the point where RTW in some capacity is possible should be referred for vocational services. Testing of skills related to a particular job can sometimes be performed by a physical therapist, occupational therapist, or speech therapist, and these evaluations may provide enough information for the physician to clear the patient for RTW. At other times, more formal vocational evaluation, counseling, and testing may be needed. For further discussion, see Chapter 34.

Stroke survivors often have residual physical and cognitive deficits that interfere with work and need to be addressed in the vocational evaluation. Physical and cognitive fatigue, errors of omission, and deficits in attention, concentration, divided attention, judgment, time management, organization and
planning, sequencing of tasks, and cognitive processing speed may limit vocational competence and performance. Patients with impairment of any of these areas likely will not be able to return to critical jobs where any error could lead to a catastrophe. RTW can be facilitated if there are colleagues of the patient, superiors, or support personnel who can check work and assure that any errors the patient makes will be uncovered before they cause damage. It may be impossible to develop adequate job samples for some professions. Formal neuropsychological testing shortly before planned RTW may be needed to characterize residual deficits that may interfere with work and to optimize compensatory strategies.

Day programs sometimes provide supervised vocational activity for severely impaired stroke survivors. In home-based programs disabled individuals perform a variety of jobs from the home; however, such individuals are usually not reintegrated into community employment. Transitional employment provides job placement, training, and support services, usually for 18 months or less, to help people transition to independent or supported employment. Supported employment is the most utilized successful strategy for placing severely disabled stroke survivors into integrated community employment. It may require ongoing support and assistance after placement, including counseling, transportation, and housing. Sheltered workshops that pay subminimum wages to people with diminished earning capacity may provide job experience but rarely lead to competitive employment. Stroke survivors deemed totally disabled for competitive employment may still be able to contribute to society through participation in volunteer activities.


COMPLICATIONS OF STROKE

Post-Stroke Disability

A. Upper Limb: Activities of Daily Living Dysfunction

Voluntary upper limb movement is a vital goal of rehabilitation. Factors predictive of upper limb functional recovery include the initial degree of motor impairment and the timing of onset of return of voluntary movement. Patients with return of voluntary finger extension and abduction of the shoulder in the hemiparetic side within 72 hours of the stroke have a 98% probability of recovering some dexterity within 6 months of stroke. This drops to 25% and 14% if no voluntary finger extension or shoulder abduction is made within 72 hours and 5 days, respectively. MRI and transcranial magnetic stimulation can assess the extent of disruption in the corticobasal motor pathways and may also be useful in prediction of upper limb motor recovery.

Many tools are available for facilitating motor and functional recovery during rehabilitation. Task- and context-specific training is well accepted in motor relearning; patients are trained at tasks that are relevant for their individual needs. However, the benefits of repetitive task training for arm or hand recovery of function remain uncertain. Teaching compensatory strategies that can be used to perform activities of daily living (ADLs) with the unaffected side is an important component of therapy, certainly until the affected arm reaches sufficient motor recovery to allow the patient to begin to participate in self-care.

Mental imagery, which involves imagining physical movement prior to attempting to move, has been shown to be beneficial in upper limb functional recovery. In mirror therapy, the patient views an image of the intact limb moving normally and imagines that it is the hemiparetic limb that is moving. This creates an illusion of perfect bilateral synchronization. It is important to keep the paretic hand behind the mirror, hidden from the view of the patient. Mirror therapy has several advantages: it is inexpensive, easily practiced by the patient, and easy for the patient or family to set up for home use. Mirror therapy may improve motor function, ADLs, and visual spatial neglect.

Post-stroke patients often obtain significant motor return of the upper extremity but do not use the limb functionally; this is termed learned nonuse. This can occur when patients compensate by relying entirely on the unaffected side early and ignoring the paretic side. Therapies that force patients to use the paretic side may prevent or reverse the development of such maladaptive learned behavior. Constraint-induced movement therapy (CIMT) involves training the
affected side while the unaffected arm is restrained, usually with a large oven-style mitt. Increasingly difficult tasks for the hemiparetic arm are gradually introduced (“shaping.”) The standard high-intensity version of this therapy is provided 5 hours per day, 5 days per week, for 2 weeks. Modified CIMT involves 1–3 hours per day, two to three times weekly, for up to several months. For both intensities, the patient is required to wear the restraint for 90% of waking hours. This form of therapy is only suitable for highly motivated individuals without severe cognitive deficits. The patient should also have a minimal level of voluntary control (10 degrees of active finger extension and 20 degrees of active wrist extension) in order to participate in CIMT. CIMT and modified CIMT for upper limb function show strong evidence of benefit for recovery of arm function in patients 3 months or more post-stroke; however, evidence of benefit for recovery of hand function is lacking. CIMT in its most intensive form should not be employed during acute rehabilitation before 2 months post-stroke.

Although many patients regain ambulatory function after a stroke, over 50% of patients with upper limb impairments fail to regain functional use of the hemiparetic arm, and less than 25% of stroke patients recover complete functional use of the hemiparetic arm. **Upper limb orthotics** are widely used to improve functional use of the impaired limb. Orthotics may be helpful to prevent contractures and reduce pain that occurs when the upper limb assumes uncomfortable postures at rest. So-called functional orthotics are useful to prevent pain and improve comfort during functional activities. However, studies have revealed no significant effects of splinting on upper limb neurologic status or spasticity. Dynamic extensor splints, often with outriggers and springs or elastic bands to substitute for paretic or plegic finger extensors, are useful as training devices (eg, Saeboflex) and with training can sometimes be used functionally. A patient with moderate return of finger flexors can use a dynamic extensor splint to perform repetitive grasp-and-release exercises. When combined with exercises for the proximal arm, dynamic extensor splints can be effective in improving the precision of preposition of the hand.

For motor rehabilitation, **functional electrical stimulation** (FES), also called **neuromuscular electrical stimulation** (NMES), stimulates the intact lower motor neuron by means of surface electrodes to contract paralyzed or paretic muscles. To produce functional results, multichannel FES devices recruit muscles in a programmed synergistic sequence, allowing a specific functional movement pattern. For the upper limb, devices can be programmed to stimulate multiple muscle groups to mimic hand grasp and release, a lateral pinch, or key
grip (eg, Bioness H200). Using these devices, even a patient with a plegic hand may be able to hold a glass or book or handle small objects such as a spoon or pen. FES devices that incorporate surface electromyography (used to detect voluntary muscle activity) can provide patients with neuromuscular education through visual or auditory biofeedback. The value of such devices is twofold. Training can be structured so that the patient strives to reach a threshold of volitional muscle contraction (sensed by surface electromyography), which triggers stimulation of the muscle by FES, causing it to contract more vigorously. Alternatively, the training can be structured so that the patient focuses on subthreshold control, which may improve muscular control during less vigorous effort. Electrostimulation, with or without the use of electromyographic biofeedback, has uncertain benefits in terms of functional motor recovery in the arm or hand but can provide strong focal stimulation of weak or paralyzed muscles to complement other passive and active treatments by the therapist.

The emergence of robotics in the field of medicine has created models for novel approaches in stroke rehabilitation. A robotic-assistive device can provide several forms of training, including passive, active assisted, active, and resisted movement. The goal of robotic devices is to assist or correct repetitious movements, with training occurring as assistance from the robot is decreased. Robot-assisted training for arm function can enhance and document motor recovery; however, the effects in hand motor recovery are uncertain. The devices are complex and expensive, and require considerable supervision by therapists.

Table 14–6 lists some of the choices of adaptive equipment available for assisting stroke survivors in ADLs. It is important to recognize appropriate patient and caregiver needs when choosing suitable adaptive equipment.

| Table 14–6 | Adaptive equipment to enhance upper extremity function. |


B. Lower Limb: Ambulatory and Mobility Dysfunction

Almost two thirds of stroke survivors have gait and mobility dysfunction. Although the majority of patients eventually gain independent ambulation by 6 months post-stroke, about 30% do not. Of patients in the community who achieve independent ambulation post-stroke, only 50% can complete a 6-minute walk test (6MWT), and of those, only 40% are able to ambulate their predicted normal walking distance. Acute treatment on a specialized stroke unit and intensive rehabilitation within 6 months of stroke can significantly enhance recovery of ambulation.

In the early recovery period following stroke that results in severe motor deficits, motor function consists of only early synergistic patterns. Ambulation is difficult owing to poor postural control, collapse of the paretic lower extremity during stance, and difficulty advancing the paretic leg in swing phase. Therefore, early physical therapy should include exercises to improve upright trunk control, and gait training should include balance, posture, and weight shifting. Ambulation can progress when spasticity becomes more prominent and voluntary movement takes place within synergistic patterns. Early ambulation usually requires an assistive device (ie, cane or hemi-walker), an ankle–foot orthosis (AFO), and physical assistance by a therapist.

Gait deviations in hemiplegia occur bilaterally, although there is seemingly unilateral motor disruption. The deviations include reduced stride and step lengths, asymmetric step length, increased stride time, reduced walking velocity, uneven cadence, foot drop, steppage, moderately wide base of support, and greater toe-out angles. There is also an increase in double support time. An AFO is useful in patients lacking sufficient strength and coordination to clear the foot in swing phase of the gait cycle. The use of an AFO increases walking velocities, increases step length on the involved side, reduces steppage, and can increase stance phase duration on the involved side. By limiting ankle motion an AFO can also often stabilize the knee during stance and weight bearing. However, if the patient has insufficient quadriceps strength to prevent knee collapse, a knee–ankle–foot orthosis (KAFO) or knee immobilizer may be needed to start gait training. AFOs can also improve metabolic energy expenditure during gait, helping to counteract the additional expenditure (50–67%) required by impaired
individuals compared with controlled normal individuals walking at the same speed. If there is no return of dorsiflexion, a solid ankle AFO is usually appropriate. As the patient becomes stronger (particularly by developing reflexive or volitional dorsiflexion or knee extension sufficient to prevent knee collapse), an articulated AFO with a joint that allows ankle movement may be more appropriate.

Therapy for gait deficits includes overground training (assisted by a therapist), partial body weight–supported treadmill training (eg, LiteGait), aerobic treadmill training, aquatic gait training, and repetitive task training with standing, weight shifting, and transfers. In most physical therapy gyms, therapies consist of assisted gait over a level surface and specific exercises to improve components of gait. Four weeks of mechanically assisted gait training with partial body weight support results in greater likelihood of independent ambulation in patients who are nonambulatory up to 3 months post-stroke, and the benefits are maintained with increased distance and speed of gait maintained at 6 months.

**Robot-aided therapies** can provide safe, intensive, and task-oriented rehabilitation. Robotic gait trainers include robot-driven foot plates and external exoskeletal orthoses. Detectors in these devices can sense patient effort and modify the assistance that is given to supplement patient musculature. With robot-aided therapy, even nonambulatory patients can participate intensively in all motor aspects of ambulation, which would be otherwise difficult with overground techniques alone. Partial body weight support with a harness can be combined with these devices to enhance training. However, several factors limit their use; the more complex devices are very expensive, require extensive therapist supervision, and are considered investigational at this time.

**FES**, described earlier in the context of upper limb rehabilitation, can aid in restoration of motor control by stimulating peripheral nerves through surface or implantable electrodes to obtain coordinated movement patterns. FES devices are used to correct drop foot in hemiplegic patients. These devices usually consist of a switch triggered during early swing phase (by the heel lifting off the ground or the angle of the tibia with the ground), a leg cuff with electrode, and an external control. With a heel switch, lifting of the heel during gait triggers electrical stimulation to the leg cuff, which activates the common peroneal nerve or the tibialis anterior muscle directly, leading to dorsiflexion and clearing the foot during the swing phase. Combining such FES use with other types of gait training results in greater improvement in gait dysfunction, faster rehabilitation time, and enhancement of endurance.
The choices of durable medical equipment and adaptive devices to enhance gait training (ie, canes and walkers) must be appropriate for individual patient needs. Additionally, wheelchairs may be needed for most or all mobility for some patients. A wheelchair for stroke patients should have a lowered seat height to allow the patient to use the unaffected lower limb to assist in propulsion of the wheelchair. Other possible adaptations include a one-arm drive mechanism, specialized cushions for comfort and positioning, backrests, trunk and head supports, and anti-tippers for those with movement disorders, cognitive deficits, or agitation. (Further discussion of assistive devices for stroke patients appears in Chapter 41.)

Poorer prognosis for achieving ambulatory status is seen with older age, low initial level of consciousness, low initial ADL performance, greater extent of cognitive dysfunction, paresis of limbs, previous hemiplegia, homonymous hemianopia, visual extinction, constructional apraxia, large size of stroke, visuospatial construction problems, urinary incontinence, and female gender. Nevertheless, even patients who will not achieve independence in ambulation benefit from gait training. Improvements in transfers can occur, and reduction in the amount of assistance needed to stand can have a significant impact on the care burden assumed by the family, accessibility to places within the home, and general quality of life.


States RA, Pappas E, Salem Y: Overground physical therapy gait training for
C. Dysphagia

Dysphagia occurs in more than half of patients post-stroke. As soon as possible after admission to a rehabilitation unit, patients should be assessed by a speech therapist, and the dietary consistency established in the acute care hospital should be verified as safe. A videofluoroscopic-modified barium swallow study or fiberoptic endoscopic evaluation of swallowing can help provide objective information about the risk of aspiration and safety of various consistencies of dietary solids and liquids.

The restoration of normal swallowing can be approached directly or indirectly. Direct methods include fluid and diet modification, repetitive practice of safe swallowing strategies, and optimizing the position and posture while eating (neck flexion or turning, or both). Indirect methods include oral musculature exercises and stimulation of the oral and pharyngeal structures. NMES (eg, VitalStim) and thermal tactile stimulation may be helpful when combined with direct methods. Repetitive transcranial magnetic stimulation is also associated with improvement in functional swallowing. Dysphagia usually improves within a few weeks following stroke; however, it can persist, requiring long-term intervention or alternative feeding strategies. In cases of severe dysphagia with recurrent pneumonia from aspiration of saliva and mucus, usually from brainstem lesions, patients may require a tracheostomy and periodic suctioning to manage the pulmonary secretions. In extreme cases (fortunately rare), patients may need to be considered for surgical procedures to seal off the oropharynx from the airway. Additional discussion of dysphagia appears in Chapter 38.


**D. Balance**

Balance deficits are a major determinant of a patient’s functional abilities and risk of falling post-stroke. Impaired balance is associated with low ambulatory activity, contributing to restricted social activity and physical deconditioning in post-stroke patients. The location of the stroke correlates with recovery of balance. In general, patients with left hemispheric strokes have a higher chance of independent standing compared with those having right hemispheric strokes. Patients whose stroke involves the parietoinsular vestibular complex or posterior fossa with severe damage to cerebellar and vestibular pathways have a poorer prognosis for recovery of balance. Visuospatial hemineglect and impaired vertical midline awareness, seen especially in those with nondominant parietal lobe damage, further add to postural instability and balance dysfunction.

Patients’ balance dysfunction can be measured with tools such as the Berg Balance Scale, measures of weight distribution, and postural sway during static sitting or standing. Computerized posturography can help determine objectively the neurologic subsystem deficits contributing to balance dysfunction, but its use has not been proven superior to standard evaluation and treatment for improving balance in stroke survivors.

A major goal of balance rehabilitation includes correcting asymmetry of weight bearing. It is common practice for therapists to use multiple sensory modalities while providing manual and verbal feedback. As an example, a patient may stand in front of a mirror, which gives visual feedback. The therapist encourages the patient verbally to stand straight and vertical, while also giving the patient manual feedback to maintain that posture. The therapist may also allow the patient to move or fall slightly out of the vertical position, then give additional tactile feedback by nudging the patient back to vertical. With such feedback, balance ability in most patients gradually improves over time. Targeted balance training during visual deprivation can also improve balance performance, most likely by means of somatosensory and vestibular inputs. Assistive devices such as canes and walkers are also important for improving balance and symmetry during gait training.


E. Aphasia

Aphasia is a disorder of language processing caused by dysfunction in a particular brain region. Between 14% and 38% of acute stroke patients experience aphasia, which is associated with disability after stroke and higher mortality. All forms of communication, both verbal and nonverbal, may be impaired. The speech center is located in the left hemisphere in 99% of right-handed and 60% of left-handed people, and overall, 93% of aphasic patients have right hemiparesis. Left hemiparesis with aphasia in a right-handed patient is rare; it suggests that the person was born left handed and taught to be right handed during childhood.

The two most frequently seen types of aphasia seen after stroke are Broca’s aphasia (expressive aphasia) and global aphasia, both of which usually involve the structures subserved by the middle cerebral artery. In contrast, Wernicke’s aphasia affects more posterior structures, often causing visual perceptual deficits from posterior parietal damage or behavioral deficits from temporal lobe damage. Particularly in Wernicke’s aphasia, during the first weeks of recovery, the patient may have no awareness of a communication problem, leading to confusion and frustration. As the patient improves and can self-monitor speech output, he or she may note expressive errors and become hesitant; leading the clinician to surmise that the deficit in a patient with Wernicke’s aphasia is in expression, not comprehension. The greatest amount of recovery occurs in the first 2–3 months after stroke onset, but significant, albeit slower recovery can take place thereafter. Treatment is divided into three phases. During the first phase, starting upon admission to the rehabilitation unit, the patient and family are educated about communication, and the baseline status of the patient’s deficits is established. During this time the speech therapist plays a vitally important role in reducing family and patient anxiety and hopelessness related to
the deficits. The second phase, lasting up to 6 months, involves intensive treatment and ongoing evaluation of emerging skills and compensatory strategies. Phase three involves group treatment, and the patient is encouraged to practice in the community. This phase also includes vocational and psychosocial counseling. In all phases of treatment, patients need to be pushed to speak and communicate, while families need to be encouraged to create opportunities for the patient to participate actively in conversation and decision making. Further details about aphasia and its treatment appear in Chapter 38.


F. Apraxia

Apraxia is the loss of ability to carry out volitional motor activities despite understanding the task, and in the absence of motor, sensory, or cognitive impairment. This is a motor-planning problem, often associated with lesions of the parietal lobe. Ideomotor apraxia is suspected when complex actions can be carried out independently, such as in response to a sensory stimulus, but not when the patient is commanded to perform the action. Patients with ideational apraxia have difficulty correctly making use of objects or tools to carry out a sequence of volitional actions to accomplish a task. Speech apraxia, which often accompanies expressive aphasia, refers to difficulties in forming expressive speech, despite the ability to move speech-related musculature.

Dressing and constructional apraxia are related to impairment of the nondominant parietal lobe, which is related to spatial disorientation and neglect more than motor planning deficit. The patient with dressing apraxia cannot orient clothing correctly, and, for example, puts a shirt on backward or upside down. In constructional apraxia, the patient makes errors copying diagrams or drawing pictures, often leaving off details on the side of the drawing opposite to the side of the hemiparesis. Patients with oculomotor apraxia have difficulty fixating their gaze volitionally; Balint’s syndrome includes this deficit along with optic ataxia (errors in reaching or pointing to an object using visual information) and simultagnosia (inability to use information across the entire
visual field and inability to see more than one object at a time in a picture). Bilateral lesions of the parietooccipital lobes cause this syndrome.

Treatment should focus on functional activities, which are structured and practiced. If correction does not occur through rehabilitation, modification of the environment should be implemented. For example, sharp objects such as knives should be removed or monitored, to prevent inadvertent harm to the patient.

Alien hand syndrome occurs rarely, often in association with frontal lobe damage that extends to the corpus callosum. The relatively intact motor cortex ipsilateral to the infarct is cut off from the intact contralateral prefrontal executive center that plans and volitionally executes movement. The result is that the affected limb moves autonomously, albeit in complex ways that are often misinterpreted by observers as intentional. (Contrast this syndrome with movement disorders, discussed in section E of Medical Complications, below, which are clearly purposeless and involuntary.) The hand often tends to grasp whatever it comes into contact with, which can lead to socially embarrassing situations. Other manifestations include automatic tic-like behavior, such as the hand picking up and clicking pens, or flicking light switches every time upon entering a room. The arm may levitate involuntarily, throwing the person off balance. Intermanual conflict, which refers to the hemiparetic hand attacking the opposite hand or preventing it from performing a task, is often seen. Fortunately, the syndrome almost always fades over time. Patients with this syndrome need reassurance that they are not insane. Simple measures include restraining the limb by putting the hand in a pocket inside the waist belt while walking, or under a bed cover at night. Incorporation of the limb into ADL and instrumental activities of daily living (IADL) training as much as possible is important.


G. Sensory Deficits

Somatosensory deficits are present in 80% or more of stroke survivors. These cause problems in exploration and manipulation of the environment, which decrease quality of life and personal safety. Sensory loss ranges from complete hemianesthesia of all sensation to dissociated impairment of somatosensory modalities. Touch, temperature, pressure, pain, and vibration are impaired in 53–64% of stroke survivors. Impaired stereognosis is the most common
somatosensory deficit after a stroke. Loss of proprioception in the upper extremity is more disabling than loss of pain and temperature sensation.

Damage to the ventroposterior nucleus of the thalamus generally causes profound contralateral sensory loss of all sensory modalities. Brainstem strokes that damage the spinothalamic tract and the trigeminal sensory nucleus cause loss of pain and temperature sensation, which can affect the ipsilateral face and contralateral lower body. Corticoparietal strokes often impair proprioception, stereognosis, and recognition of texture, which usually spares the trunk. Somatosensory processing is often impaired, noted by delayed recognition of stimuli (compared with the intact side). Somatosensory impairment tends to be more severe and long lasting in right hemispheric strokes compared with left.

Physician action is important for treatment of sensory disturbances. Dysesthesias may be painful and should be treated to maximize rehabilitative performance. The presence of pain in a limb with impaired sensation strongly suggests central pain, which may respond to gabapentin, pregabalin, or other more direct or adjunctive analgesics. The rehabilitation physician needs to monitor skin regularly in patients with decreased sensation. Sensory loss can predispose to skin breakdown, injury, and burn injuries from heating pads.

All stroke patients should receive an occupational therapy evaluation of sensation. Two test batteries frequently used to assess sensory deficits and recovery from stroke are the Nottingham Sensory Assessment and Rivermead Assessment of Somatosensory Performance. The therapist generally also tests for two-point discrimination, or uses Semmes Weinstein filaments for more objective verification of deficits in sensory thresholds. The therapist can then design interventions to help the patient compensate for the deficit to regain function. For example, a patient with severe loss of hand sensation (common in middle cerebral artery strokes) will tend to drop objects held in the affected hand; one strategy is to train the patient to look at the hand at all times when it is used.

Impaired sensory function correlates with the quality of upper limb movement, force control, fine manipulation, and sensory ataxia (dyscoordination of movement when the patient does not look at the limb). Very intensive active motor retraining may be associated with some sensory recovery, but often sensory function improves even without specific sensory training. The effect of formal active sensory retraining, which is helpful in sensory loss associated with peripheral nerve damage, is somewhat uncertain. This typically consists of progressive tactile object recognition (finding, handling, and recognizing small objects of decreasing size buried in a bowl of rice). Data are stronger for passive
sensory stimulation, which involves repetitive electrical stimulation of the hypesthetic body part at various frequencies and intensities, all above the threshold for perception; this is theorized to induce neuroplasticity passively.


**H. Visual Deficits: Hemianopsia**

Up to one third of stroke survivors have visual field loss. This may occur acutely from damage to any neurologic structure that affects vision or visual processing. Often, the deficit does not involve primary visual pathways and improves or resolves over time. Unfortunately, homonymous hemianopsia resulting from damage to the optic tracts, optic radiation, or occipital lobes usually does not recover. Efforts at restoring vision have, at best, led to minimal functional gains. Computerized Vision Restoration (Novavision) is a computerized training program consisting of random presentation of stimuli at the edge of the hemianopsia, to try to stimulate recovery of vision. It is associated with limited reported increase in useful field of vision, possibly by stimulating compensatory scanning.

The primary rehabilitative treatment for this disorder consists of training the patient in all therapies to scan visually to the affected side. Other treatments of hemianopsia involve repetitive sensorimotor stimulation to force attention to the affected side and prisms to bring objects in the hemianoptic visual field into the intact field. Fresnel paste-on prisms can be placed on the lateral aspect of an eyeglass lens ipsilateral to the hemianopia. When a client looks into the blind hemifield or the edge of the blind area, double vision is experienced. One image is the normal view seen by the eye looking through the plain lens. The eye that looks through the prism sees the image displaced by the prism by approximately 20 degrees. In homonymous hemianopsia expansion of visual field an average of 5 degrees is reported. Studies have shown that such prisms do not improve ADL function in stroke patients with hemianopsia. However, they are worth trying, as up to two thirds of patients report subjective improvements in ADLs using this technique. The prisms may make it possible for some patients to accomplish necessary tasks more safely, such as walking straight down a hallway without hitting a wall, or walking outside without bumping into people.

If premorbid low vision is also present, a variety of aides and techniques for
this disorder may also improve functional deficits associated with hemianopsia. Bright lighting, handheld magnifiers, enlarged print, and high-contrast tape or markers to indicate hazards and identify or locate items can be helpful. Maneuvers to improve reading in patients with field cuts or low vision include using a colored ribbon or ruler under a line of text or on the left side of text, placing the finger at the beginning of each line, and reinforcing scanning.

The intact visual system can compensate well for small optical ophthalmologic or eye alignment imperfections; after a stroke of almost any type, such compensation deteriorates, and imperfections become noticeable and may interfere with function. Patients with persistent diplopia may benefit from using an eye patch on either eye. This should be done as little as can be tolerated, because natural recovery will occur faster with greater exposure to the problem. Post-stroke patients with visual deficits, whether preexisting or stroke related, should undergo evaluation by an ophthalmologist or neuroophthalmologist. Patients with severe visual loss should be referred early in their rehabilitation to a low-vision therapist.


**I. Visuoperceptual Deficits: Hemi-inattention (Neglect)**

It should be noted that homonymous hemianopsia is not synonymous with hemi-inattention (neglect). The patient with pure visual problem may lack neglect and be able to scan and compensate well for the visual loss. However, others with or without intact visual fields may be severely impaired by visual or perceptual neglect. Neglect is associated with significant functional and visual difficulties, including bumping into objects on the hemiparetic side, diplopia, difficulties with ocular convergence, impaired saccadic movement, oversensitivity to light, nystagmus, and dry eyes. Patients with neglect should be evaluated by an ophthalmologist for intraophthalmic or other eye health problems that could complicate the neurologic deficits. Neuro-ophthalmology consultation can be
helpful for localizing neurologic lesions and for developing short- and long-range plans for intervention. Patients with any evidence of perceptual neglect should be prohibited from operating a motor vehicle.

Although no single technique has been shown in controlled studies to increase or speed recovery from neglect, the patient with visual or perceptual neglect should receive general rehabilitation (mobility and self-care training) from multiple disciplines. This should include visual, tactile, vestibular, and auditory stimuli to increase awareness of and sensitivity to sensory input on the affected side. In most patients with left-sided neglect, language is a strength, and language tasks can be employed for visual and auditory stimulation. The environment should be modified to stimulate sensory input using the affected side during part of the day when the patient is well rested and alert. If the patient is fatigued or the neglect is too severe, however, such stimulation may be counterproductive and should be limited. Computer games, in particular those requiring motor input (eg, Wii) can be helpful. Medications to improve alertness and arousal may also help improve participation.


**J. Memory and Executive Functioning**

Memory is the cognitive ability to encode, store, and retrieve information. It is subcategorized by retention time (short-and long-term memory) and type of material encoded (declarative and procedural). Patients found to have memory impairment causing difficulties in rehabilitation or adaptive functioning should undergo a comprehensive memory assessment, ideally by a neuropsychologist. Medications that may impede memory and cognition, such as benzodiazepines or other sedatives, should be minimized or avoided.

Nursing and therapy sessions should include strategies that make use of preserved cognitive abilities to reduce the patient’s disabilities, such as notebooks or log books, audiotapes, electronic organizers, and audio alarms. Usually, occupational and speech therapists are involved in training patients to use such devices. Approaches aimed at enhancing memory function include
modifying patients’ rooms, the nursing unit, and therapy environment to be similar to the patients’ usual living environment as much as possible to encourage generalization. Activities, therapies, and meals should take place at the same time each day. Nursing and therapy staff should coordinate their efforts to maintain consistency and limit introduction of unexpected stimuli. Patients may also manifest attention and concentration deficits, errors of action, and inability to perform tasks requiring divided attention. Rehabilitation should include development and training in activity patterns that emphasize reduction of environmental distractors, completion of one task at a time, and repeated training sessions in complex daily activities that will be necessary after discharge. Neuropsychological evaluation is essential for these patients in the rehabilitation setting, even if the deficits seem mild; it should be repeated prior to attempting return to work, even if recovery seems complete. Occupational therapy can sometimes create a work-related job sample activity to help test the patient and clarify deficits. For patients with persistent deficits, referral for vocational rehabilitation and dialog with employers about workplace accommodation may be appropriate.


Medical Complications

A. Spasticity

The primary treatment of spasticity in stroke patients is range-of-motion and stretching exercises, targeting muscles most at risk (see Table 14–4). Relaxation techniques, motor reeducation, biofeedback, avoidance of noxious stimuli (especially skin breakdown), and optimization of positioning can all be helpful adjuncts to stretching. Modalities, splints, and casts are also helpful. The direct application of vibratory stimuli has antispastic effects in the hemiplegic upper limbs of post-stroke patients. Splinting joints most at risk, such as the wrist and hand or the ankle, may reduce discomfort and prevent contractures, but if the spasticity is too great, the splints can cause skin irritation or breakdown, which in turn can increase the spasticity. Serial casting is valuable for stretching a joint with a contracture but to be most effective may need to be combined with focal
or systemic antispasticity medication. Oral antispasticity drugs must be used with caution, as stroke patients may be more susceptible to adverse cognitive effects of these medications than patients with intact brains. Spasticity is discussed in detail in Chapter 6.


B. Shoulder Pain

Shoulder pain affects as many as 75% of post-stroke patients. Patients with hemiplegic shoulder pain (HSP) may have associated prolonged hospitalization, decreased quality of life, poor functional recovery, disturbed sleep, and depression. HSP is associated with severity of the paresis, poor upper limb functional recovery at 12 weeks, and prolonged rehabilitation stay. Many causes of HSP have been postulated (Table 14–7). Clinically, patients with HSP often have localized tenderness over the biceps brachii and supraspinatus tendons. Pain may be present at rest but is generally greater during range-of-motion activities (greatest with external rotation and abduction) or with dependent arm position. Central pain from damage to sensory pathways may be present regardless of range of motion or position and may magnify pain from any of the causes listed in Table 14–7.

Table 14–7 Possible causes of hemiplegic shoulder pain.
Once shoulder pain develops, trials of analgesics (starting with acetaminophen), antiinflammatories, or antispasticity agents may be attempted. Physical and occupational therapists may utilize muscle energy techniques and shoulder joint mobilization techniques to reduce pain and improve range of motion; often, such treatment reduces pain substantially. Adjunctive use of transcutaneous electrical nerve stimulation and NMES can also be considered. NMES has been shown to improve pain-free passive shoulder range of motion. Transcutaneous NMES (T-NMES) and NMES with implanted electrodes have shown efficacy in relieving HSP, most likely from altering central pain modulation rather than from reduction of subluxation. If central pain is suspected, adjunctive medication such as gabapentin, pregabalin, or tricyclic agents should also be considered and may be more efficacious compared with narcotics. In patients who do not respond to these agents, steroid injection of the shoulder joint or sheaths of painful tendons may be useful. For patients with adhesive capsulitis, analgesia and injections can help in tolerating proper positioning, range of motion, and stretching, which are essential to long-term management.

Prevention of HSP should begin immediately post-stroke. The basic strategy for prevention includes attention to shoulder positioning and handling. The shoulder should be positioned as much as tolerated in external rotation and abduction. Foam supports and shoulder strapping may be helpful. A subluxed shoulder should be supported by a lap tray or arm trough while the patient is
sitting. Early post-stroke application of T-NMES has been proven to reduce HSP. Shoulder slings should be used judiciously. If reduction of subluxation decreases discomfort, then a sling should be considered and evaluated during upright activities. Slings may also be helpful during ambulation to protect the arm from trauma and injury. However, for every sling that is prescribed, the physician must advise the patient to take the arm out of the sling and perform range-of-motion exercises several times daily.


C. Complex Regional Pain Syndrome

Complex regional pain syndrome (CRPS) occurs in 12.5–28% of post-stroke patients. Other common terms for this condition are reflex sympathetic dystrophy and shoulder hand syndrome. CRP Sgenerally develops within 3 months of an acute stroke, and very rarely develops after 5 months. The syndrome involves severe pain, hyperesthesia, alldynia, vasomotor dysfunction, edema, and, if untreated, atrophy of soft tissues of the shoulder and hand, and patchy demineralization of the bones of the extremity (Sudeck’s atrophy). Often, swelling of the hands and fingers causes extension of the fingers, in contrast to the usual tendency of the fingers to assume a flexed position during recovery from stroke. Movement tends to aggravate pain, and patients often resist range of
motion and exercise, risking the development of severe contractures and atrophy, even if pain eventually improves. CRPS generally resolves within 1 year in 35% of post-stroke patients.

CRPS is thought to be a consequence of immobility, sensory dysfunction, or imbalance, and possibly extreme central sensitization. Tissue damage may also play a role. The diagnosis of CRPS is made clinically, largely by physical examination. Radiographs of the affected limb should be performed to rule out fracture or other bony pathology. Treatment approaches for CRPS should include the positioning and range-of-motion exercises used for HSP. Modalities that may evoke discomfort, such as vigorous therapeutic heat and cold, electrical stimulation, and aggressive range of motion, must be moderated in intensity to avoid overstimulation, aggravation of severe pain, and worsening of the condition. Control of edema is limited to limb elevation and gentle massage. Addition of adjunctive medications used for neuropathic pain, particularly gabapentin, may be helpful. Oral high-dose tapering steroids for 10–20 days are likely to reduce pain and swelling and improve toleration of physical therapy, beginning several days after initiating the medication. Oral steroids may be risky in patients with “brittle” diabetes. Regional anesthesia with stellate ganglion block can be considered for patients not receiving anticoagulants. The rehabilitation physician must assure that all pain-relieving therapies ordered are followed immediately by range-of-motion and, if at all possible, active exercises for the limb, including isometric strengthening and stress loading of the limb. This will counteract the effects of immobilization, improve motor recovery, and may possibly reduce the sensory imbalance contributing to the CRPS.


D. Hydrocephalus

Hydrocephalus is an excessive accumulation of cerebrospinal fluid within the ventricles or subarachnoid space, or both, and often complicates subarachnoid hemorrhage (SAH), intracerebral hemorrhage (ICH), and intraventricular hemorrhage (IVH). IVH, in particular, can lead to arachnoiditis from blood in the cerebrospinal fluid, causing obstructive hydrocephalus even several weeks after the acute event—sometimes after admission to the rehabilitation unit. Mechanical compression of cerebrospinal fluid flow channels from intracerebral hematoma or surrounding edema can also cause obstructive hydrocephalus. Communicating hydrocephalus is often seen after hemicraniectomy, which is usually performed because of excessively elevated intracranial pressure from a large bleed, infarct, or cerebral edema; hydrocephalus often persists after cranioplasty.

The early signs and symptoms of hydrocephalus reflect elevated intracranial pressure and include headaches, nausea, vomiting, decreased appetite, and changes in personality or behavior. At later stages there is compression of the midbrain structures, causing diplopia from third and sixth cranial nerve palsies, lethargy, drowsiness, coma, and “setting-sun” sign due to impairment of upward gaze. Vital signs may show bradycardia, systemic hypertension, and altered respiratory rate. Papilledema may be seen with funduscopic examination. CT or MRI should be performed diagnostically and to measure ventricular size in patients with suspected hydrocephalus. Following confirmation of new or enlarging hydrocephalus, an immediate neurosurgical evaluation is indicated to determine the need for more intensive medical monitoring, and whether ventricular shunting is indicated.


E. Movement Disorders

The prevalence of nonspastic post-stroke movement disorders ranges from 1.1% to 3.9%. Most symptoms of these disorders are induced by lesions of the basal ganglia, thalamus, or frontal subcortical pathways. Movement disorders seen in
patients immediately post-stroke, especially chorea and hemiballismus, often resolve spontaneously within 2 weeks; however, many persist. Hemichorea and nonspastic dystonia are the most frequent movement disorders post-stroke.

Nonspastic dystonia, which usually occurs several months post-stroke, is characterized by disturbed voluntary muscle contraction that results in twisting, turning, and posturing of the limbs, neck, or trunk. It may be focal, segmental, or generalized. Approximately one third of patients eventually have complete recovery, and nearly two thirds have partial recovery. Focal disorders such as cervical dystonia and blepharospasm may respond to chemodenervation of selected hyperactive muscles with botulinum toxin. Generalized dystonia may respond to benzodiazepines (clonazepam, diazepam), baclofen, anticholinergic agents, or tetrabenazine. Rehabilitation techniques such as biofeedback, massage, and repetitive exercise of affected muscle groups need to be individualized. Dystonia and spasticity can coincide during stroke recovery; in such cases, antispasticity medication may offer only partial relief.

Hemichorea involves involuntary, often brief, irregular flowing movements that interfere with volitional movement. Hemiballismus, which arises from damage to the subthalamic nucleus, causes severe flinging of extremities when the patient tries to move. Potentially helpful medications include dopamine-blocking agents (eg, antipsychotics and tetrabenazine). Clonazepam, valproate, and reserpine have also been helpful to decrease the involuntary movements. Stereotactic thalamotomy and deep brain (thalamic) stimulation have helped some patients with chronic dysfunction refractory to medication.

Post-stroke secondary Parkinson’s disease (see Chapter 19) often occurs months after a stroke. Depression and dementia are frequently seen in these patients. Medical treatment includes dopaminergic and anticholinergic medications, although patients with secondary Parkinson’s disease respond less predictably to these agents than those with primary disease. In evaluating patients, the clinician must consider the possibility that stroke can occur in a patient with primary Parkinson’s disease, leading to worsening of symptoms or unmasking of previously silent disease. Although a trial of antiparkinsonian medication may be worthwhile, it is also important, as in all post-stroke movement disorders, to weigh the adverse effects of medications against observed therapeutic efficacy and to wean medications in nonresponders. (For further details, see Chapter 19.)

Post-stroke tremors are usually delayed in onset and often respond poorly to drug treatment. This is particularly true for rubral tremors (midbrain damage) and palatal tremors. Intention tremors, which interfere with voluntary movement,
are often associated with cerebellar dysfunction and usually do not respond to medication or surgery. Mobility and ADL training with weighted adaptive equipment or wrist and ankle weights may be helpful. Preexisting essential tremor may interfere with rehabilitation and can be treated with propranolol, anticholinergics, and reduction in caffeine intake.

Myoclonus is seen in the palate and face of patients with brainstem strokes, and elsewhere in those with thalamus and basal ganglia damage. Clonazepam and valproate can be helpful. It should be noted that, as disturbing these symptoms may appear to the examiner, they may hardly be noticed by the patient, may interfere minimally with function, and may not require drug suppression.

F. Mood Disturbances
Depression and anxiety are common sequelae of stroke and occur in at least one third of post-stroke patients within 1 year. Although neuroanatomic correlates have been suggested, studies have shown that post-stroke depression shares characteristics with depression in older patients with other chronic diseases, suggesting that it represents a psychological reaction to loss, rather than a physical or neurochemical response post-stroke. However, the positive functional and emotional responses to antidepressants post-stroke, even in patients without clinical depression, suggest that cerebral damage may be associated with a change in CNS levels of serotonin or norepinephrine. Nondepressed post-stroke patients given SSRIs are significantly less likely to develop post-stroke depression than those not given the medications.

In the rehabilitation setting, severe depression may impede participation in therapy and limit functional progress; when this occurs, depression should be treated aggressively. Among patients affected severely by post-stroke depression, there is a higher frequency of death, including death by suicide. Yet despite this, many patients are not treated or are undertreated.

Management of depression includes psychotherapy and pharmacotherapy. Psychotherapy alone is generally not effective in treating severe post-stroke
depression but may elevate mood and provide insight to patients affected mildly. Antidepressant medications, including SSRIs and tricyclic agents, are effective in treating post-stroke depression and reducing depression rating scale scores. Stimulants such as low-dose methylphenidate may also elevate mood. Clinicians must consider potential side effects of pharmacotherapy prior to initiating treatment or prophylaxis.

Certain emotional disturbances occur almost universally post-stroke; these include hyper irritability, anxiety, and decreased control of anger. Other frequently occurring problems include emotional lability and pathologic laughing or crying. The latter condition, known as pseudobulbar affect, is estimated to affect 11–34% of patients with cerebrovascular disease, usually spontaneously, without emotive stimulus. In contrast, in emotional lability, the response is developed from an emotive stimulus but is out of proportion to the stimulus and outside of expected emotional control. Emotional lability affects approximately 20–25% of post-stroke patients within the first 6 months, and 10–15% at 12 months post-stroke. Pseudobulbar affect develops most frequently in patients with lesions of the basis pontis. Additionally, lesions of the internal capsule, cerebral peduncles, and cerebellum increase the frequency of both emotional lability and pseudobulbar affect.

Although crying is most often seen in patients with pseudobulbar affect, diagnosable depressive disorders or other depressive symptoms often are not seen. Family and friends may respond emotionally and with vigorous sympathy to the patient, and this reaction often causes more distress to the patient than his or her own crying. In the case of laughing, family may mistakenly think that the patient is mocking or laughing at them. Patients may worry that they are becoming insane. Invariably, patients affected by these disorders describe distress and embarrassment, and avoid social contact. If pseudobulbar affect is suspected, the clinician should ask patients who are crying if they are truly upset, and if there is something that is making them sad. If not, then educating the patient and family about the condition, and emotional lability, can be very helpful in reducing distress. Antidepressants are often effective in reducing the frequency of both conditions following stroke.


Hackett ML, Anderson CS, House A, Xia J: Interventions for treating
G. Sleep Disturbances

Sleep disorders are the most unrecognized and undertreated modifiable risk factor for developing a stroke. Stroke also can unmask or further exacerbate sleep disturbances such as obstructive sleep apnea, periodic limb movement, and restless legs syndrome. Sleep-disordered breathing, mostly in the form of obstructive sleep apnea, affects almost 50% of post-stroke patients. Immediately following acute stroke there is an increase in the incidence of central sleep apnea, which usually completely resolves within several months. Sleep–wake disorders, mostly in the form of insomnia, affect 20–40% of post-stroke patients. Excessive daytime sleepiness frequently follows thalamic or midbrain strokes. Sleep disorders are associated with poor rehabilitation outcome after stroke. Therefore, effective treatment of sleep disorders is important for primary and secondary stroke prevention and improvement in overall rehabilitation outcomes.

Post-stroke patients should be risk stratified for sleep-disordered breathing, and polysomnography should be utilized in those suspected of the disorder to confirm the diagnosis and initiate treatment. If possible, patients diagnosed with obstructive sleep apnea should be fitted with a comfortable mask and equipment prescribed prior to discharge home. In such patients, use of continuous positive airway pressure (CPAP) has been shown to improve functional recovery and depressive symptoms, and in those with moderate to severe apnea, to decrease future stroke recurrences. Patients with obstructive sleep apnea often report dramatically improved daytime fatigue with the use of CPAP.
Patients with insomnia, whose strokes involve the basal ganglia, corona radiata, or pyramidal tract, should also be screened for periodic limb movement and restless legs syndrome. Treatment of these disorders in such patients, usually with dopaminergic drugs such as ropinirole, can enhance stroke recovery. Treatment of insomnia, whatever the cause and manifestation, should also include behavioral and environmental interventions, such as relaxation induction techniques, maintaining the bedroom as a quiet place, limiting bedtime stimulation, and keeping a consistent bedtime. Limiting daytime napping may also be helpful.

Pharmacologic interventions at bedtime include judicious use of hypnotics or other sedating drugs such as trazodone. It should be noted that many hypnotic or sedating drugs will worsen obstructive sleep apnea. Pain that interferes with sleep needs to be treated; patients with central pain may benefit from gabapentin at bedtime. Those with nocturia may respond to limitation of evening fluid intake and short-acting anticholinergics at bedtime. If depression is present, scheduling sedating antidepressants at bedtime may induce or deepen sleep.


H. Daytime Fatigue

The incidence of post-stroke fatigue (PSF) is estimated at 30–68%. Unlike fatigue in normal individuals, PSF, which is a common and a long-term sequela of stroke, is unrelated to prior exertion and unrelieved after rest. Multifactorial causes are generally thought to contribute to PSF, with depression having the strongest correlation post-stroke. Other possibly influential factors include nutritional imbalance, systemic disorders (infection, congestive heart failure, hypothyroidism, anemia, etc), medication side effects, sleep disorders (see preceding discussion), and emotional disturbances. PSF has a negative impact on functional ability and independence, and it can lead to poor physical activity level in patients and increased caregiver burden. PSF is also associated with worse survival at 2–3 years after stroke.

Several possible interventions to treat fatigue include pharmacotherapy (antidepressants, stimulants, modafinil, and dopaminergics), psychotherapy, and physical therapy to increase endurance. Systemic disorders and nutritional
imbalance should also be identified and treated.


I. Falls and Injuries

Falls are frequent occurrences after stroke, with an estimated incidence of 73% within 6 months of discharge home. In a hospital setting the incidence of falls is 14–65%, and nearly one third of these patients suffer injuries, usually contusions or abrasions, but occasionally fractures. For those on anticoagulants, the risk of major hemorrhagic sequelae from a fall is greater (but generally not so great that it constitutes an absolute contraindication to anticoagulation during inpatient rehabilitation). As osteoporosis is a significant complication of stroke, the risk of fractures in post-stroke patients is quadruple that of nonstroke control populations. The most common site of fracture in stroke survivors is hip fracture on the hemiplegic side. Additionally, although falls may motivate some patients to adhere better to safety precautions, many patients are left in fear of further falls, leading to avoidance and decrease of physical activity. Even with aggressive fall prevention programs in an inpatient rehabilitation setting, falls still frequently occur. Generally, the highest risk of falls is seen in patients with impaired balance, visuospatial hemineglect, and poor performance in ADLs. Patients with leg weakness, sensory loss, foot problems, incontinence, and visual deficits are also considered to be at high risk of falling. As the majority of post-stroke survivors suffer from one or more of these problems, it is appropriate to apply fall prevention strategies (Table 14–8) to all patients.

Table 14–8 Strategies for preventing falls and injuries from falls in post-stroke patients.
Multifactorial interventions reduce the rate of falls but not the risk of falling. To prevent falls, the preceding risk factors need to be addressed in physical or occupational therapies. Cognitive, psychological, and emotional factors, particularly anxiety when standing or ambulating, should also be addressed. On the rehabilitation unit, judicious use of bed and wheelchair alarms and restraints needs to be considered. Some patients respond to education, while others need constant reminders. Confused patients who become agitated with alarms or restraints may respond positively to the use of an enclosure bed. Alternatively, a confused, agitated patient may need a one-on-one companion or behavioral technician to ensure safety. It should be noted that assistive devices such as canes, walkers, wheelchairs, and bedrails are sometimes ordered to promote safety; however, the effect of these devices should be evaluated carefully, as they may actually increase the likelihood of falling for some patients.

Pharmacotherapy with use of low-dose vitamin D supplementation has been shown to reduce falls in female patients in an institutional setting. If present, osteoporosis should be treated to reduce fracture risk.

Batchelor F, Hill K, Mackintosh S, Said C: What works in fall prevention after


**PROGNOSIS**

Although more than half of all stroke survivors achieve functional independence, approximately one third remain permanently disabled, requiring assistance with ambulation, and one fourth are dependent for ADLs. In addition, 20–25% of all stroke survivors are permanently institutionalized, usually in nursing facilities. Potential factors that may adversely influence functional outcomes and response to rehabilitation post-stroke are listed in Table 14–9.

**Table 14–9** Potential factors that may adversely affect prognosis and outcome following stroke.
The prognosis in elderly patients is often worse than in younger patients, but this often reflects a poor premorbid functional level and the presence of multiple other medical comorbidities, including dementia, diabetes mellitus, cardiac disease, prior strokes, electrocardiographic abnormalities, or alcohol abuse. Delay in initiation of acute medical care, especially fibrinolytic therapy, reduces the likelihood of a good functional recovery.

Patients with lacunar strokes have an excellent prognosis, as most achieve complete or nearly complete recovery. Prognosis following other ischemic strokes and ICH is less predictable; poor potential for recovery is associated with large lesions, bilateral lesions, low level of consciousness, and certain neurologic deficits. Anatomically, large brainstem hemorrhages have the worst prognosis due to likelihood of coma and death. Patients with large cerebellar, putamen, and thalamic hemorrhages are also at risk of coma, or of severe disability if they survive. Those with small hemorrhages, including brainstem lesions, often have an excellent chance of survival and recovery, as the area of actual ischemic damage can be very small; hence many of the deficits may improve as hemorrhage and surrounding edema resolve.

Neurologic deficits causing neglect and affecting cognition, communication, sensation, and vision have been associated with poor outcomes. Clearly impaired cognition, severe aphasia, abulia, and neglect impair learning of new skills. Visual deficits present a challenge to rehabilitation, especially when accompanied by sensorimotor and communication deficits. In addition, poor
sitting balance, incontinence persisting for more than 1–2 weeks, minimal motor recovery, or a flaccid limb at 4 weeks have also been associated with decreased functional recovery and decreased likelihood of discharge to the community.

The clinician and the care team should always take into account psychological and social factors that may be obstacles to functional recovery. Depression and poor coping skills have been correlated with poorer outcomes and should be addressed with counseling and pharmacologic interventions as early as possible. The lack of social supports should always be addressed, as it usually severely reduces a patient’s realistic options for discharge to community and complicates follow-up care.


Multiple sclerosis (MS) most often affects Caucasian females, with onset of symptoms in the second and third decades of life.

No definitive etiology has been determined.

There are four clinical patterns; the most common is relapsing-remitting MS.

Magnetic resonance imaging (MRI) of the brain and spinal cord is the most useful diagnostic tool for detecting MS.

Multiple sclerosis (MS) is an immune-mediated disease of the central nervous system that most commonly affects young to middle-aged adults. It is characterized by multiple white matter plaques of demyelination that pose major threats to function by producing impairments to cognition, vision, speech, swallowing, muscle function, and bowel and bladder function. Because of its chronic, progressive nature, disability may evolve throughout the course of a patient’s lifetime. The costs of disease, both direct and indirect, are immense.
Direct medical costs have been estimated to be in excess of $10 billion per year in the United States. Indirect costs include those related to employment (reduced, or unemployment), disability-related assistive devices and home modifications, and personal care. Physiatric and rehabilitative intervention is crucial to ensure functional and symptomatic improvement over the course of a patient’s life, to improve quality of life, and to decrease the burden of disease.

Most patients with MS are diagnosed while in their 20s and 30s, traditionally prime working and childbearing years. Although the incidence of disease is difficult to ascertain, it is estimated that 400,000 people in the United States are living with MS. Worldwide, prevalence probably exceeds 2.5 million people. Areas with the highest prevalence include northern Europe, southern Australia, and the middle portion of North America. Additionally, MS is one of the most common causes of nontraumatic disability in young adults. The disease is more common in women, with a female-to-male ratio of approximately 2–3:1. Caucasians also tend to be affected more than blacks and Asians.

Although the cause of MS remains unknown, several different factors appear to play a role. One theory focuses on environmental factors associated with geographic location. Living farther away from the equator has been found to convey a higher risk of developing MS, although the Inuit population is an exception. A second notable difference relates to migration patterns. Researchers have noted that the risk of MS decreases when individuals migrate from areas of high to low geographic risk, but the opposite is not true. Although the relationship between distance from the equator and MS is unclear, the significance of vitamin D–promoting sun exposure in relation to development of MS has gained prominence. There is a known increased risk of development of MS in those with low levels of vitamin D. Research is ongoing to determine the effect of vitamin D levels on symptoms in patients previously diagnosed with the disease. Giving additional credence to the environmental influence is the variability seen in disease manifestations and age of onset. This variability implies that while individuals may have a genetic predisposition to develop the disease, environmental factors may continue to mitigate susceptibility. It remains unclear whether a critical period for physiologically necessary sun exposure exists during the lifespan.

The most prominent theories of causation focus on a postinfectious etiology and an autoimmune process, with the latter influenced by genetic predisposition. Numerous research studies have investigated the possibility that an infectious agent initiates the disease process in MS. Viruses that have been studied include rabies, herpes simplex virus 6 (HSV-6), measles, coronavirus, canine distemper
virus, human T-lymphotropic virus 1 (HTLV-1), and Epstein-Barr virus (EBV). *Chlamydia pneumoniae*, a bacterium, has also been studied. Although none of these agents has been proven to cause MS, many experts believe, based on cerebrospinal fluid studies, that an infectious agent does play a role. The persistence of oligoclonal immunoglobulin G (IgG) in the cerebrospinal fluid of patients with MS is consistent with infectious or parainfectious disorders of the central nervous system; however, the specificity of IgG in cerebrospinal fluid remains unknown. Of the viral candidates, the relationship between EBV and MS has been established most strongly, as individuals who are seronegative for EBV have a very low risk of developing MS. Exposure to EBV may be most important during adolescence or early adulthood, as there are many more adults who are seropositive for EBV but do not have MS. Moreover, infectious mononucleosis has been determined to be a risk factor for MS. Again, it remains unclear at which point of exposure any of these factors could ignite the inflammatory cascade.

A genetic predisposition, in combination with environmental triggers, has also been proposed as a mechanism for MS. Multiple genetic linkage studies have confirmed a linkage with the major histocompatibility (MHC) region, as well as less well-defined linkages to other zones that code for interleukins. The presence of serotype HLA-DR2 in DR-positive families is associated with a greater chance of developing the disease. The risk of concordant MS is 30% with monozygotic twins, 5% with dizygotic twins, and between 2% and 4% for first-degree family members of people with MS.

Despite many theories and ongoing research, no clear single etiology has been elucidated, and the disease likely represents a combination of environmental, infectious, and genetic factors.


Fox RJ: Multiple sclerosis. Cleveland Clinic Center for Continuing Education. Available at: http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/neuro
PATHOGENESIS

MS was first defined as its own disease entity by French neurologist Jean-Martin Charcot in 1868. Historically, MS has also been known as disseminated sclerosis or encephalomyelitis disseminata, based on descriptions of affected brains and spinal cords. Charcot called the disease *sclerose en plaques*, based on his own clinical findings and those of others. Charcot also noticed a triad of symptoms similar among patients with the disease: diplopia, dysarthria, and ataxia.

The disease is characterized by multiple lesions occurring in the white matter of the brain and spinal cord, causing demyelination. Typically these lesions are found adjacent to the lateral ventricles, corpus callosum, periaqueductal region, optic nerves, optic chiasm, and optic tracts, as well as white matter tracts of the spinal cord. The process is believed to be autoimmune mediated, whereby an individual’s immune system attacks the nervous system in response to some unknown trigger. This process is believed to occur in genetically susceptible individuals. The plaques consist of inflammatory infiltrates composed of lymphocytes and macrophages, and there is evidence of astrocytic proliferation and gliosis. This inflammatory process is caused by T cells. T cells cross the blood–brain barrier through leaks, and begin to attack the myelin. An additional release of other inflammatory cytokines, such as interleukin-2, γ interferon, and tumor necrosis factor, also occurs.

T cells specifically attack oligodendrocytes, the cells responsible for production and maintenance of myelin. Myelin helps to insulate axons, and if damaged, results in loss of impulse strength between nodes of Ranvier. This slows or blocks nerve conduction, thus impeding normal nerve impulses and compromising related functions. As the disease worsens, transection of the axons may occur, correlated with irreversible disability. An additional insult to the axon is mediated not only by demyelination, but also by the proliferation of sodium channels localized within the membrane. In an attempt to reestablish normal conduction, there is an increased entry of sodium, slowing of nerve conduction, and potentially, even conduction block.

In between exacerbations, a repair process occurs, or remyelination. However, oligodendrocytes cannot completely rebuild the cell’s myelin sheath, and repeated attacks lead to successively fewer effective remyelinations. The myelin segments become thinner and shorter until a scarlike plaque is built up around the damaged axons. Return of function depends on the degree of
remyelination, although this can be altered by fatigue, heat, or additional comorbidities.

Researchers have recently described four different pathologic subtypes of MS. They are: cell-mediated destruction of myelin, cell-mediated myelin destruction with immunoglobulin and activated complement, primary oligodendrogliopathy with apoptosis, and finally neurodegenerative oligodendrogliopathy.


CLINICAL FINDINGS

 Symptoms and Signs

MS remains a clinical diagnosis. Patient presentations are varied and complex, occasionally making the diagnosis a challenge to the practitioner. The first presenting symptoms are commonly optic neuritis or sensory disturbance. Other symptoms that may prompt patients to seek evaluation are tremor, ataxia, cognitive problems, and dysarthria. Many patients report symptoms of weakness,
bowel and bladder dysfunction, fatigue, balance deficits, and pain, for which the differential diagnosis is broad (see later discussion).

The National Multiple Sclerosis Society classifies MS into four main clinical patterns, which are contrasted in Table 15–1. This classification system is helpful in clarifying prognostic and therapeutic decisions. Of the four patterns, the last two are far less common than the first two.

Table 15–1 Classification of multiple sclerosis.

<table>
<thead>
<tr>
<th>Disease Pattern</th>
<th>Percentage of Patients Affected</th>
<th>Characteristic Findings</th>
<th>Clinical Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Relapsing-remitting</td>
<td>55–65%</td>
<td>Sudden neurologic decline that resolves over 4–8 weeks</td>
<td>Remission after exacerbations; return to baseline or residual associated disability</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Most common symptoms are tingling/weakness in limbs, visual disturbances</td>
<td>Stable between exacerbations</td>
</tr>
<tr>
<td>Secondary-progressive</td>
<td>25%</td>
<td>Develops from relapsing-remitting form of disease</td>
<td>Similar to above, but deterioration with each exacerbation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Functional deficits accumulate between attacks</td>
<td>50% of relapsing remitting convert to secondary progressive within 10 years</td>
</tr>
<tr>
<td>Primary-progressive</td>
<td>10%</td>
<td>Older age at diagnosis (50+ years)</td>
<td>Progressive disease without remissions</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Men and women affected equally</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Initial symptoms are most often motor and continuous, without relapsing-remitting periods</td>
<td></td>
</tr>
<tr>
<td>Progressive-relapsing</td>
<td>5%</td>
<td>Aggressive onset and often rapid worsening of symptoms, necessitating increasing support</td>
<td>Swiftly progressive disease, with high mortality rate</td>
</tr>
</tbody>
</table>

A. Relapsing-Remitting MS

This is the most common pattern of MS, affecting approximately 55–65% of patients. Affected individuals have discrete attacks of new or worsened neurologic symptoms that emerge over a few days and may resolve over a 4–8-week period with or without corticosteroid treatment. The most common initial
complaint is tingling or weakness of a limb. Others may complain of visual disturbances or optic neuritis—transient unilateral visual impairment lasting days to weeks, which may be associated with retrobulbar pain. In general, patients with sensory symptoms, and patients whose symptoms fully remit after early exacerbations, demonstrate better long-term prognosis. All relapsing-remitting patients remit after an exacerbation, but they may have residual disability.

**B. Secondary-Progressive MS**

About 25% of patients are in the secondary progressive category. This pattern begins as relapsing-remitting MS; however, patients deteriorate between attacks and, overtime, functional deficits accumulate. Many patients who have a relapsing-remitting pattern of disease initially eventually develop the secondary-progressive type. The reason for this is not known. These patients also tend to have fewer attacks and show less gadolinium enhancement on magnetic resonance imaging (MRI) than during the relapsing-remitting phase (see later discussion).

**C. Primary-Progressive MS**

The primary-progressive pattern of disease accounts for approximately 10% of all MS patients. These individuals tend to be older when the disease manifests (~50 years), and both men and women are affected equally. This pattern is characterized by a progressive disease course without attacks. Unlike patients with relapsing-remitting MS, those with primary-progressive MS usually have an initial complaint of motor symptoms. Patients present with asymmetric paraparesis, and symptoms tend to progress more rapidly, validating observations that patients who begin with motor symptoms fare less well than those with sensory complaints. No disease-modifying therapies have been approved for the treatment of patients with primary-progressive MS.

**D. Progressive-Relapsing MS**

This pattern affects about 5% of patients with MS. This disease pattern is aggressive, frequently affecting the brainstem. Patients typically have a swift decline, characterized by an increasing number of relapses as well as worsening residual impairment, and may require mechanical ventilation. Mortality from the disease is common.
Laboratory & Imaging Studies

The diagnostic workup of a patient with MS may include laboratory tests, neuroimaging studies of the brain and the spinal cord, as well as neurophysiologic studies. To rule out the presence of another disease process, diagnostic and laboratory tests should be performed that screen for other neurologic, rheumatologic, infectious, metabolic, and cardiac causes. Additionally, lumbar puncture may be performed for cerebrospinal fluid analysis. Oligoclonal bands are present in the cerebrospinal fluid of many patients with MS. Their presence is not pathognomonic for MS, and pleocytosis is usually noted.

A. Magnetic Resonance Imaging

MRI has become the most useful diagnostic tool in the evaluation of patients with MS. Typically, MS lesions are ovoid, greater than 5mm in diameter, and located in the periventricular white matter. Such lesions may be called Dawson’s fingers, as they surround the deep veins of the brain, perpendicular to the long axis of the lateral ventricles. MRI may be repeated over time if there is uncertainty about the diagnosis. This may clarify whether lesions are disseminated in time. Gadolinium should be used to determine if any of the lesions represent acute disease (Figure 15–1). The contrast medium crosses the blood–brain barrier, and lesions that have developed between 4 and 12 weeks prior to imaging will enhance on T1 images. Imaging is useful for diagnosis as well as determining a patient’s response to disease-modifying therapy. With fluid-attenuated inversion recovery (FLAIR) sequencing, cerebrospinal fluid is suppressed and appears dark; this allows for improved imaging of the lesions compared with traditional T2 imaging (Figure 15–2 and 15–3). Over time, the T2 hyperintense lesions atrophy, producing “black holes” on T1 image sequences. These black holes signify areas of axonal damage and demyelination. The relevance and importance of spinal cord imaging in MS has increased over time.
Figure 15–1 Magnetic resonance imaging scan of a patient with MS. The T1 image at left was taken before infusion of gadolinium. The image at right is postinfusion. Note the enhancing lesion in the right frontal lobe. Such T1 “black holes” are associated with cognitive decline and atrophy.
Figure 15–2 Sagittal FLAIR image showing hyperintense lesions.
MRI has demonstrated utility in clinical trials as a marker of disease activity in patients. Although not all pathologic changes are represented on imaging, gadolinium enhancement and new T2 lesions provide objective measurements of disease activity. Practitioners vary in their use of MRI in the clinical setting; however, MRI is commonly used in combination with other diagnostic and physical examination findings in evaluating the efficacy of disease-modifying therapy as well as in determining prognosis.

Traditional MRI focuses on white matter changes. New technologies such as diffusion tensor imaging and magnetization transfer imaging are being implemented to further study gray matter changes and their subsequent clinical and functional effects. Although the use of imaging in clinical practice continues to increase, it is important to avoid relying solely on imaging when making diagnostic decisions.

**B. Evoked Potentials**

Visual evoked potentials, brainstem evoked potentials, and somatosensory evoked potentials can be used alone or in combination in the evaluation of
suspected MS. In patients with MS, latencies are usually prolonged. Visual evoked potentials can be used to assess the presence of subclinical optic neuritis.

**Diagnostic Criteria**

Over the years, the diagnosis of MS has evolved. In the 1960s, MS was a clinically made diagnosis; by the 1980s, criteria for diagnosis were expanded to include laboratory tests. In 2005, updated criteria were proposed. The most recent modification came in 2010 with the McDonald criteria, which rely on clinical presentation, MRI findings, and results of additional tests such as cerebrospinal fluid analysis and visual evoked potentials. The diagnosis of MS has been made when the following criteria were met: two or more occurrences of central nervous system symptoms, separated by time and space; a lesion that occurred in different locations in the central nervous system at different points; and dissemination in space or time, or both. The 2010 revised criteria allow for diagnosis of MS to be made based on clinical presentation and the use of MRI at a single point in time. A “clinical attack” of MS is typically defined as a neurologic change that persists for more than 24 hours. Infections should be screened for immediately. The “dissemination in space” criterion is satisfied by one or more T2 lesions in the periventricular, juxtacortical, infratentorial, or spinal cord. The “dissemination in time” criterion is defined as the presence of a gadolinium-enhancing or new T2 lesion on followup imaging, regardless of the timing of the prior MRI.


**Differential Diagnosis**

The differential diagnosis for MS includes many conditions with systemic manifestations (Table 15–2), and the diagnosis may be one of exclusion. A systematic workup of symptoms usually distinguishes those that mimic MS,
such as vascular, infectious, and neoplastic diseases, from true central demyelinating pathology.

**Table 15–2** Differential diagnosis of multiple sclerosis.

<table>
<thead>
<tr>
<th>Neuromyelitis optica</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute disseminated encephalomyelitis (ADEM)</td>
</tr>
<tr>
<td>Transverse myelitis</td>
</tr>
<tr>
<td>Neurosyphilis</td>
</tr>
<tr>
<td>Cerebral autosomal-dominant arteriopathy with subcortical</td>
</tr>
<tr>
<td>leukoencephalopathy (CADASIL)</td>
</tr>
<tr>
<td>Behçet's syndrome</td>
</tr>
<tr>
<td>Neurosarcoidosis</td>
</tr>
<tr>
<td>Vitamin B\textsubscript{12} deficiency</td>
</tr>
<tr>
<td>Folate deficiency</td>
</tr>
<tr>
<td>Vasculitic process</td>
</tr>
<tr>
<td>Mixed connective tissue disease</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td>Lyme disease</td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
</tr>
<tr>
<td>Sjögren's syndrome</td>
</tr>
<tr>
<td>Carcinoma</td>
</tr>
<tr>
<td>Wegener's granulomatosis</td>
</tr>
<tr>
<td>Hypercoagulable state</td>
</tr>
<tr>
<td>Migraine</td>
</tr>
<tr>
<td>Hypertension</td>
</tr>
<tr>
<td>Mitochondrial disorders</td>
</tr>
</tbody>
</table>

**TREATMENT**

Like the disease itself, the treatment of MS is varied and complex, encompassing a wide range of treatment measures. A comprehensive approach, individualized for each patient, is needed that best utilizes the existing therapeutic options. Despite the availability of many disease-modifying agents, the progressive nature of the disease requires physiatric involvement at every stage to optimize
strength and function, and prevent deterioration in the patient’s quality of life.

**Pharmacotherapy**

Because MS is an immune-mediated disease, pharmacotherapy, including use of various immunoactive drugs, is a cornerstone of treatment. Three main categories of drugs are available: (1) immunosuppressants, (2), disease-modifying drugs, and (3) drugs that block extracellular processes. Corticosteroids are usually the first-line agent when patients present with acute disease.

**A. Corticosteroids**

Methylprednisolone is used in short bursts for acute attacks or relapses. It has both antiinflammatory and antiedematous effects. Dosage is generally 500–1000 mg/day intravenously, for 3–5 days, which may be followed by an oral taper. Despite lack of consensus on regimen, dose, or duration, a Cochrane review from 2009 found no significant clinical or radiologic differences between patients receiving oral versus intravenous steroids. Corticosteroids reduce severity and duration of exacerbations, and recent data also demonstrate their long-term benefit in patients with MS. However, while they hasten recovery, steroids do not prevent further attacks or alter the progression of disease. Furthermore, use of these drugs should be weighed against the known long-term risks of treatment (ie, avascular necrosis, osteoporosis).

**B. Disease-Modifying Drugs**

1. **Interferons**—Several immunomodulators used in the treatment of MS are variants of interferon β. Interferons are proteins that play a role against viral, microbial, and neoplastic insults, and also help to regulate the immune system. When early treatments with interferon γ caused an increase in exacerbations, interferon β was employed to suppress interferon γ. Subsequently, interferon β was found to decrease T-cell activation and inhibit leakage in the blood–brain barrier, reducing the extent of demyelination (see Pathogenesis, earlier).

   Interferons are available in different formulations for use weekly, every other day, and three times a week. Patients treated with all three Food and Drug Administration (FDA)–approved interferons have demonstrated little to no progression of disease on followup MRI. Relapse rates also decreased among patients taking these drugs. Side effects are similar and include injection site
reactions, flulike symptoms, and development of neutralizing antibodies, which lessen efficacy. Table 15–3 summarizes information about these and other commonly prescribed disease-modifying drugs.

Table 15–3 Disease-modifying drugs used in the treatment of multiple sclerosis.

<table>
<thead>
<tr>
<th>Name</th>
<th>Route</th>
<th>Frequency</th>
<th>Monitoring</th>
<th>Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>IFN-β1a (Avonex)</td>
<td>IM</td>
<td>Weekly</td>
<td>CBC, LFTs, TSH</td>
<td>Myalgia, hepatotoxicity</td>
</tr>
<tr>
<td>IFN-β1a (Rebif)</td>
<td>SQ-autoinjector</td>
<td>3 times daily</td>
<td>CBC, LFTs, TSH</td>
<td>Myalgia, injection site reactions</td>
</tr>
<tr>
<td>IFN-β1b (Betaseron, Extavia)</td>
<td>SQ-autoinjector</td>
<td>Every other day</td>
<td>CBC, LFTs, TSH</td>
<td>Myalgia, injection site reactions</td>
</tr>
<tr>
<td>Glatiramer acetate (Copaxone)</td>
<td>SQ-autoinjector</td>
<td>Daily</td>
<td>None</td>
<td>Injection site reactions, lipoatrophy, systemic reaction (chest pain, shortness of breath)</td>
</tr>
<tr>
<td>Mitoxtantrone (Novantrone)</td>
<td>IV</td>
<td>~ Every 3 months</td>
<td>CBC, LFTs, UA, Echo</td>
<td>Male and female infertility, cardiotoxicity, leukemia, hair loss, amenorrhea</td>
</tr>
<tr>
<td>Natalizumab (Tysabri)</td>
<td>IV</td>
<td>Every 28 days</td>
<td>LFTs</td>
<td>Hypersensitivity, progressive multifocal leukoencephalopathy, death</td>
</tr>
<tr>
<td>Fingolimod (Gilenya)</td>
<td>Capsule</td>
<td>Daily</td>
<td>LFTs, CBC</td>
<td>Cardiac (bradycardia), vision (macular edema), infections, varicella exposure, teratogenic</td>
</tr>
</tbody>
</table>

IM, intramuscular; SQ, subcutaneous; IV, intravenous; CBC, complete blood count; LFTs, liver function tests; TSH, thyroid-stimulating hormone; UA, urinalysis; Echo, echocardiogram.

²Agents are listed by order of introduction.

2. Glatiramer acetate (Copaxone)—Glatiramer acetate is a first-line drug for patients with relapsing MS. The drug is a synthetic mix of polypeptides consisting of four amino acids (glutamic acid, lysine, alanine, and tyrosine). The proposed mechanism of action involves binding to class II major histocompatibility antigens, with inhibition of T cells, thus, blocking immunologic attack. Dosage is 20 mcg/day subcutaneously. Adverse effects include reaction site irritation and (rarely) arthritic complaints. Some patients experience a transient response termed immediate postinjection reaction, which
consists of chest tightness, flushing, and dyspnea, beginning soon after the injection and lasting no longer than 20 minutes. In a 15-year study, many patients who took glatiramer demonstrated few relapses and minimal disease progression.

**3. Natalizumab (Tysabri)**—Natalizumab contains humanized neutralizing IgG4-κ monoclonal antibodies that act against leukocyte α4 integrins. By blocking such integrins, natalizumab impedes the movement of mononuclear leukocytes into the central nervous system, resulting in less inflammation. The drug, indicated for relapsing-remitting MS, is administered intravenously every 28 days. As noted by Bomprezza and colleagues, this medication is “typically reserved for patients with clinically or radiographically extremely active disease either as initial therapy or when initial therapy has been ineffective or poorly tolerated.” Its side effect profile mandates that patients receiving natalizumab receive rigorous ongoing clinical surveillance. The most significant and severe of these side effects is the risk of developing progressive multifocal leukoencephalopathy (PML), a rare demyelinating disorder of the brain caused by the John Cunningham (JC) virus. The risk of JC virus infection increases after 24 months of natalizumab therapy. The JC antibody virus test has helped to stratify according to risk patients who are about to begin or are already receiving natalizumab therapy. Factors that increase the risk of developing PML include more than 24 infusions of natalizumab, prior immunosuppressant use, and positive JC antibody status.

**4. Mitoxantrone (Novantrone)**—Mitoxantrone is the only chemotherapeutic agent approved for use in patients with MS. It acts by reducing lymphocyte proliferation. It is considered a second-line treatment for patients with secondary-progressive or worsening relapsing-remitting MS. Mitoxantrone is given as an intravenous infusion over 30 minutes every 3 months at 12 mg/m² for a 2–3-year period, with a maximum cumulative dose of 140 mg/m². The drug is associated with significant side effects, including risk of leukemia development and reduced ejection fraction of the left ventricle. Duration of use is limited to approximately 2 years due to cardiotoxicity and monitoring with echocardiography is routine. Studies have indicated that mitoxantrone may decrease exacerbations and reduce the number of lesions seen on MRI. Moreover, these effects may persist for several years after discontinuation of the medication.

**5. Fingolimod (Gilenya)**—Fingolimod gained FDA approval in 2010 as a
first-line agent for patients with MS. It is also the first oral agent approved for the treatment of relapsing-remitting MS. The recommended dosage is 0.5 mg daily. The drug is an oral sphingosine-1 phosphate (S1P) receptor modulator that binds to lymphocytes, sequestering them and thereby downregulating their expression.

Two pivotal studies evaluated the efficacy of fingolimod against placebo (FREEDOMS) or interferon β1a (TRANSFORMS). These studies showed that fingolimod resulted in significant reduction in annualized relapse rates (ARR) compared with placebo or IFN-β1a. Additionally, fingolimod achieved reduction in new T2 or gadolinium-enhancing lesions and time to disease progression. Two deaths occurred in patients receiving a dosage of 1.25 mg daily, one due to disseminated primary varicella zoster infection and the other to herpes simplex encephalitis. For this reason, varicella antibody titers should be obtained before beginning drug therapy. If the antibody titer is low, varicella vaccination is advised. Animal models have shown fingolimod to have neuroprotective effects.

Side effects include development of bradycardia or atrioventricular node block. Thus, before a patient begins therapy with the drug, electrocardiography should be performed and a cardiology consultation scheduled, if necessary. Patients should be monitored for 6 hours following administration of the first dose as the risk of heart block is elevated during this period. A followup electrocardiogram at the end of the 6 hours is also advised. Other side effects include risk of macular edema, infections, mild changes in pulmonary function testing, and elevated transaminases. Patients should have ophthalmologic testing within 4 months of beginning the drug to screen for macular edema. Laboratory testing for hematologic and liver profile assessment should also be performed quarterly. The safety of this medication in pregnancy has not been established and caution is advised when administering the drug to patients of childbearing age.

C. Other Agents

1. Teriflunomide (Aubagio)—Teriflunomide is the active metabolite of leflunomide, a drug used in the treatment of rheumatoid arthritis and approved for treatment of relapsing MS. It causes reversible inhibition of the mitochondrial enzyme dihydroorotate dehydrogenase (DHODH), which plays a crucial role in pyrimidine synthesis for new DNA replication. Because the salvage pathway is spared, quiescent T cells are not affected. However, the drug reduces T- and B-cell replication, activation, and function when autoantigens are present. In clinical studies, teriflunomide, a once-daily oral agent, significantly
reduced the ARR and the number of new lesions seen on MRI, and slowed disability progression compared with placebo. Several other studies involving active comparator and a study using teriflunomide in clinically isolated syndrome, a first clinical event suggestive of MS, are underway. Side effects include elevated transaminase levels, hair thinning, nausea, and diarrhea. This drug has been given a pregnancy category X rating and should be used with caution in patients of childbearing age. Rapid elimination with cholestyramine is possible should a patient become pregnant while taking the drug.

2. Dimethyl fumarate (Tecfidera)—Dimethyl fumarate (DMF) is the third oral agent, after fingolimod and teriflunomide, approved by the FDA for the treatment of relapsing forms of MS. Fumaric acid esters (FAE) have previously been used in the treatment of patients with psoriasis. Although the mechanism of action of DMF is not fully understood, the drug is believed to help downregulate several of the inflammatory mechanisms responsible for apoptosis of activated T cells that will ultimately transmit across the blood–brain barrier. In two pivotal clinical trials involving DMF (DEFINE, a placebo-controlled study using oral fumarate at two different dosages, and CONFIRM, in which an active comparator of glatiramer acetate and two dosages of oral fumarate were compared to placebo), dimethyl fumarate significantly reduced the ARR and number of new enhancing lesions compared with placebo. In the DEFINE trial, improvement in disability was also demonstrated. Dosing is twice daily, and the most common side effects include flushing and gastrointestinal upset.


Rehabilitation & Exercise

Prescribing exercise and designing therapy protocols for MS patients presents a challenge because the disease is heterogeneous and unpredictable. The goal should be to maximize individual physical, emotional, social, and vocational independence and improve quality of life. Because MS is a progressive disease, physicians need to anticipate the future needs of the patient based on his or her clinical course. Rehabilitation of patients with MS, unlike that of many other disorders, is an ongoing process.

For most MS patients an outpatient course of therapy is beneficial. More complex cases with extensive needs will require a multidisciplinary approach offered by inpatient rehabilitation. Acute inpatient rehabilitation is utilized frequently after exacerbations that result in dramatic decreases in function, strength, or ability to complete activities of daily living (ADLs). Multiple studies show a statistically significant improvement of disability and impairment with therapy. A home exercise program is crucial to maximize carryover of effects. Therapy more than twice a week may be too fatiguing for patients and is not advised. Also, timing of therapy may be best in the morning due to fatigue and
temperature-related increase in symptoms. Therapy offered usually consists of ADLs, range of motion, strengthening, balance, coordination, and gait training. Improvements can be seen in mobility, balance, transfers, self-care, bladder control, fitness, and the use of adaptive devices.

There is no evidence that exercise is detrimental to MS patients, or that it influences the disease process negatively. Most MS patients have a normal cardiovascular response to exercise, but some patients with severe symptoms may have a blunted response to exercise. With advanced disease, exercise tolerance may be reduced secondary to decreased aerobic capacity, possibly related to deconditioning or respiratory muscle dysfunction.

Regular exercise has a positive effect on the fitness of MS patients, but they may require a longer training program to achieve cardiovascular benefits. Weak muscles can be strengthened in MS patients and this may translate into improved function. Exercise has been shown to have a beneficial effect on MS disability and quality of life, but not on impairment. Aerobic training improves maximum exercise capacity for ambulating MS patients, but there is minimal evidence that exercise benefits nonambulatory patients.

### A. Gait and Ambulation Problems

Most patients with MS have difficulty with ambulation. Gait difficulties may be the result of weakness, spasticity, fatigue, decreased proprioception, visual loss, or cerebellar dysfunction. Safety of ambulation is a great concern in many MS patients, and an assistive device is usually required within 20 years of the onset of symptoms. One third of ambulating patients with MS are frequent fallers. Poor balance and the use of an assistive device are the best predictors for falls. Strategies should be implemented to avoid falls in patients with balance deficits and weakness, as many are osteoporotic and fractures occurring as a result of falls are debilitating and can lead to significant decline in function.

The typical gait pattern in a patient with MS shows decreased stride length; increased cadence; slow speed; decreased rotation of the hips, knees, and ankles; increased trunk flexion; and decreased vertical lift. Wireless neuromuscular electrical stimulators (NMES) have been introduced to improve foot drop by means of peroneal nerve stimulation. Both the Bioness L300 and Walkaide expand on the traditional ankle–foot orthosis model in assisting with foot clearance that is challenged by weakness or spasticity.

In clinical trials, patients taking dalfampridine were shown to have increased walking speeds; it is the only drug for which functional improvements have been noted. Dalfampridine is believed to work by inhibiting potassium channels.
thereby increasing conduction of action potential. Dosage is 10 mg twice a day. A major side effect of the drug is its propensity to cause seizures.

**B. Spasticity**

Addressing spasticity with physical therapy or pharmacotherapy (oral, injectable, or intrathecal) is valuable in maintaining function. The use of oral medications is frequently limited by the accompanying side effects (ie, fatigue and cognitive clouding). Botulinum toxin therapy is an option for patients with focal areas of spasticity. Many patients with MS find the use of intrathecal baclofen to be a better alternative for the treatment of spasticity because fatigue is not a side effect. Patients must be selected carefully for intrathecal baclofen therapy, and screening with an intrathecal trial is recommended. Timing of the intervention is critical and it should be performed before significant lower limb weakness occurs. Additional discussion of spasticity management appears in Chapter 6.

**C. Fatigue**

The most common symptom of MS is lack of energy, or fatigue, which is frequently exacerbated by heat. This occurs more frequently in women than men. The etiology of the fatigue is unclear; however, it may be related to a decreased central motor drive or decreased muscle fiber aerobic capacity, possibly related to sympathetic dysfunction or deconditioning. Fatigue management includes energy conservation, maintaining a cool environment, improving sleep, and avoiding smoking, alcohol, and caffeine. It is important to differentiate between central and peripheral fatigue. Central fatigue is frequently associated with central nervous system impairment and is not alleviated with rest. Patients complain of difficulty with arousal, attention, and task completion. Peripheral fatigue is secondary to muscular fatigue and improves with rest.

MS patients may be vulnerable to effects of increased heat with exercise. Multiple studies have been conducted to examine the benefits for patients of cooling vests used during periods of exercise, with modest results. Drinking cool water or ice slurries is beneficial. Environmental cooling efforts such as fans or air conditioners should be used for MS patients with heat sensitivity.

Screening for metabolic disorders, anemia, and mood disturbance is paramount, as all are known to produce fatigue. Sleeping disorders are common in patients with MS. In addition to activity modification, pharmacologic treatment of fatigue is recommended. The use of stimulants (methylphenidate, dextroamphetamine/amphetamine) may help with focus and attention. The use of modafinil and armodafinil may improve symptoms of fatigue. Studies
investigating the use of modafinil in the treatment of MS-related fatigue have yielded mixed results, the majority of which support its use.

**D. Cognition**

Function may be limited by cognition, as 50% of MS patients have some cognitive deficits. Difficulty with memory, attention, information processing, fluency, executive function, and visuospatial function are frequently seen. Cognitive dysfunction correlates with the number and location of lesions on MRI.


Neurophysiologic studies can play a significant role in the assessment of peripheral nervous system disorders and suspected neuromuscular dysfunction. Neurophysiologic evaluation provides objective information about the diagnosis, localization, nature, and severity of the pathologic processes involved. Information obtained about the peripheral neuromuscular system can be used to plan and prescribe a therapeutic program. It also provides information on prognosis and the effectiveness of a treatment program. This chapter reviews basic principles of electrodiagnostic studies and the technical aspects of obtaining biologic signals from study subjects.

**NEUROMUSCULAR ANATOMY & PHYSIOLOGY**

The peripheral nervous system consists of the nerve root, the motor efferent from the anterior horn cell, and the afferent sensory root with dorsal root ganglion, usually located in the neural foramina—both of which combine to form a mixed spinal nerve. The mixed spinal nerve splits into the ventral and dorsal rami. The ventral rami from these nerves coalesce to form trunks, divisions, and cords, which ultimately become terminal nerve branches of the brachial plexus (to innervate the upper limbs) and the lumbosacral plexus (to innervate the lower limbs). The dorsal rami of the spinal nerves innervate the paraspinal muscles (Figure 16–1).
Nerve axons have electrical properties common to all excitable cells. The axons not only conduct the propagating electrical potential but also transport nutritional and trophic substances, the latter maintaining the metabolic integrity of the peripheral nerve. There is extracellular preponderance of sodium (Na) ions and intracellular preponderance of potassium (K) ions. At rest an excess positive charge outside the cell and negative charge inside the cell is maintained as a resting transmembrane potential by the cell wall. Voltage-gated sodium (Na\(^+\)) and potassium (K\(^+\)) channels are present in the cell wall. The density of the Na channels is high at the nodes of Ranvier in the myelinated axons. The influx of Na with efflux of K that occurs upon opening of these channels produces the action potential that is propagated along the axon. When a weak current is applied to a nerve, negative charges from the negative pole (cathode) make the inside of the cell relatively more positive, causing depolarization. After 10–30 mV of depolarization, the membrane potential reaches a critical threshold for
generation of an action potential, which is bidirectional.

Motor action potentials that are generated at the cortical level travel through the brainstem and spinal cord to the muscle. At the level of the muscle the action potentials traverse the neuromuscular junction. The neuromuscular junction is a simple relay between the nerve terminals of a motor neuron and the skeletal muscle fibers innervated by that nerve. The electrical potential that arrives at the nerve terminal is converted at the neuromuscular junction to a chemical signal. The neuromuscular junction contains the presynaptic nerve terminal, a specialized postsynaptic muscle membrane, and the endplate, which underlies the nerve terminal (Figure 16–2).
Chemical transmission across the synaptic cleft is made possible by acetylcholine (ACh), which is synthesized from acetyl coenzyme-A and choline by the enzyme choline acetyltransferase in the nerve terminal. ACh is present in
three compartments in the presynaptic terminal. The largest compartment is a reserve of ACh, or main store, with 300,000 quanta available. A smaller compartment, which serves as a “mobilization store,” is adjacent to the endplate zone with approximately 10,000 quanta of ACh available for movement to the nerve terminal. The smaller synaptic vesicles loaded with ACh aggregate near discrete areas of the presynaptic membrane in active zones, available for immediate release. In the resting state, a small quantity of ACh is released intermittently; this release is dependent on intracellular calcium (Ca) concentration that allows for binding of the ACh molecule to the postsynaptic membrane. Once released, the ACh diffuses across the synaptic cleft and binds to ACh receptors (AChR), which are concentrated on the postsynaptic folds.

Binding of ACh to its receptor causes the ion channel in the center of the AChR to open, resulting in a flux of Na, which depolarizes the muscle membrane at the junctional region. If only a single quantum of ACh is released, a nonpropagating depolarization is produced, resulting in a miniature endplate potential. The action of ACh is quickly terminated by the release of acetylcholinesterase, which breaks the molecule down into acetic acid and choline.

In contrast to intermittent random release of quanta of ACh at rest, there is coordinated release of up to 100 quanta of ACh into the primary synaptic cleft when an action potential is generated from the motor axon. As the action potential travels, depolarizing the presynaptic terminal, the voltage-gated Ca channels open, allowing Ca to diffuse across the presynaptic terminal. This triggers a coordinated movement of ACh vesicles toward the synaptic membrane, resulting in fusion and release of ACh to the synaptic cleft. This much larger release of ACh diffuses across the synaptic cleft, binding to a large number of ACh receptors and causing a larger nonpropagating depolarization of the postsynaptic membrane. This nonpropagating potential is called the endplate potential.

If this potential reaches the critical threshold for depolarization, an action potential is propagated. This action potential invades the tubule system of the muscle, resulting in release of Ca from sarcoplasmic reticulum and triggering muscle fiber contraction in all the muscle fibers innervated by the axon. Coordinated generation of a large number of motor unit action potentials is needed to contract the muscle. The larger a muscle is, the more motor unit action potentials are needed to activate it.

The sensory cell body is located at the dorsal root ganglion, which is at the level of the neural foramen and distal to the sensory nerve root. The sensory
response thus is obtainable with stimulation distal to the ganglion in any lesion proximal to the dorsal root ganglion (preganglionic lesion) despite the loss of sensation in the distribution of the nerve root because of the intact axon distal to the ganglion.


NERVE CONDUCTION INJURIES

Classification

Peripheral nerve injuries are most commonly classified according to Seddon’s scheme, which identifies three classes of injury. The terms corresponding to these classes—neurapraxia (temporary loss of nerve conduction without structural change of the axon), axonotmesis (disruption of the axons), and neurotmesis (complete disruption of the axons and surrounding connective tissue)—are generally used by electromyographers (Table 16–1). Sunderland’s classification of nerve injury, although more extensive, is not helpful to the electromyographer and may be more useful to the peripheral nerve surgeon.

Table 16–1 Classification of nerve injuries.
Amplitude is the most important measure in the nerve conduction study because it reflects the number of conducting nerve fibers. If some of these fibers are blocked (as occurs in neurapraxia), then the amplitude with stimulation proximal to the block will be lower in a manner proportionate to the number of fibers blocked. Latency reflects information about the fastest conducting fibers. Conduction velocity reflects the fastest time between two stimulating electrodes. This measure is not sensitive enough to assess focal axonal loss but measures pathologic changes involving the myelin. Conduction velocities can be normal in patients with focal axon loss neuropathies in which up to 75% of the motor nerve fibers have been lost, especially if the loss primarily affects the slower conducting axons. With proximal stimulation and greater distance from the recording electrode in a normal nerve, there is desynchronization and phase cancellation of the response, which decreases the amplitude of the motor response. Although some physiologic dispersion and loss of amplitude is normal, the loss of amplitude proximally of 25–30% or greater may be evidence of a conduction block in most of the nerves.

The mildest injury, neurapraxia, has the best prognosis for recovery. Neurotmesis, which results in absent motor amplitude throughout the course of the nerve, has the worst prognosis, given the disruption of axons and epineurium that occurs. This problem may require surgical repair by a skilled peripheral

<table>
<thead>
<tr>
<th></th>
<th>Neurapraxia</th>
<th>Axonotmesis</th>
<th>Mixed Injury (Neurapraxia and Axonotmesis)</th>
<th>Neurotmesis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Motor</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Proximal</td>
<td>Decreased amplitude</td>
<td>Decreased amplitude</td>
<td>Partial decreased amplitude No response</td>
<td></td>
</tr>
<tr>
<td>Distal</td>
<td>Normal amplitude</td>
<td>Decreased amplitude</td>
<td>Partial decreased amplitude</td>
<td>No response</td>
</tr>
<tr>
<td><strong>Sensory</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Proximal</td>
<td>Decreased amplitude</td>
<td>Decreased amplitude</td>
<td>Partial decreased amplitude</td>
<td>No response</td>
</tr>
<tr>
<td>Distal</td>
<td>Normal amplitude</td>
<td>Decreased amplitude</td>
<td>Partial decreased amplitude</td>
<td>No response</td>
</tr>
<tr>
<td>Demyelination</td>
<td>Present</td>
<td>Absent</td>
<td>Partially present</td>
<td>Absent</td>
</tr>
<tr>
<td>Wallerian degeneration</td>
<td>Absent</td>
<td>Present</td>
<td>Partially present</td>
<td>Present</td>
</tr>
<tr>
<td><strong>Perineurium intact</strong></td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>
nerve surgeon. Outcomes are often guarded, although the surgical approximation of nerve fibers is improving. The prognosis for recovery depends on many factors, including proximity of the lesion to the muscle(s) being innervated, integrity of the muscle itself, and age and health of the patient. In partial axon loss lesions, the previously mentioned factors should be taken into consideration for recovery along with severity of the lesion. In general, upper brachial plexopathy has a better prognosis than lower brachial plexopathy, because of the length of axonal regeneration needed in each instance.

After a nerve injury, changes in amplitude of the compound muscle action potential occur over a period of 6–8 days, and in the sensory nerve action potential over a period of 8–12 days; therefore, nerve conduction studies performed within 1–2 weeks after a traumatic injury can determine whether there is axon loss and, if present, the severity of it. Changes seen on needle electromyography may evolve over 3–4 weeks and are more helpful in localizing the axonal loss lesion (Table 16–2). Electrodiagnostic studies can establish where a lesion is located and whether it is generalized, multifocal, or focal. Additionally, they can indicate whether the underlying process involves demyelination or axon loss.

Table 16–2 Electrodiagnostic changes associated with pathologic processes.
<table>
<thead>
<tr>
<th>NCS Changes</th>
<th>Segmental Demyelination</th>
<th>Partial Axon Loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>CMAP amplitude</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal</td>
<td>Normal</td>
<td>Decreased</td>
</tr>
<tr>
<td>Proximal</td>
<td>May decrease (temporal dispersion)</td>
<td>Decreased</td>
</tr>
<tr>
<td>CMAP duration</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Proximal</td>
<td>Prolonged (temporal dispersion)</td>
<td>Normal</td>
</tr>
<tr>
<td>Distal latency</td>
<td>Prolonged</td>
<td>Normal range</td>
</tr>
<tr>
<td>Conduction velocity</td>
<td>Slow</td>
<td>Normal range</td>
</tr>
</tbody>
</table>

| EMG Changes                                     |                         |                   |
| Fibrillations and PSW                           | No                      | Yes               |
| MUAP changes                                    |                         |                   |
| Acute                                           | None                    | None              |
| Subacute                                        | None                    | Normal amplitude/wide duration polyphasic units |
| Chronic                                         | None                    | Large amplitude/wide duration polyphasic units |
| MUAP recruitment                                | Normal                  | Decreased         |

NCS, nerve conduction studies; CMAP, compound muscle action potential; EMG, electromyography; PSW, positive sharp wave; MUAP, motor unit action potential.
Electromyographic Studies

Electrodiagnostic studies are biased toward evaluating the large, myelinated (type A) peripheral nerve fibers, and not the smaller type C pain fibers. Two different types of studies are performed: (1) motor and sensory nerve conduction studies (NCS), and (2) needle electromyography (EMG). The EMG study analyzes motor units and spontaneous potentials seen in the muscles after axon loss nerve injury. NCS and needle EMG together are commonly referred to as “EMG.” These two studies are used in combination to assess and diagnose multiple neuromuscular problems.

The changes noted on NCS or needle EMG are dependent on the type of injury and the time from injury. These tests can help confirm a suspected peripheral nerve injury or, alternately, suggest other explanations for a patient’s pain, muscle atrophy, or weakness. One advantage of early electrodiagnostic testing is its ability to detect evidence of conduction block and continuity of nerve fibers. It can also help to rule out premorbid problems such as underlying peripheral neuropathy, which may influence a patient’s recovery. One disadvantage of early electrodiagnostic testing is its limited prognostic value, as testing may take place before neurophysiologic changes have had time to evolve.

In the hands of a skilled electromyographer, electrodiagnostic studies have a specificity greater than 90%. The results of these studies should always be interpreted in combination with the clinical history and physical examination. An electrodiagnostic study is not an extension of the physical examination; it only confirms a clinical impression. If there are electrodiagnostic findings in the absence of clinical findings, the test does not add to the diagnosis or treatment. When inconsistencies are noted, the clinician should consider whether affected nerves were compared with similar nerves in the contralateral limb, whether the patient has a different underlying medical condition, or whether the electrodiagnostic study was thorough and technically competent. Patients should ideally be evaluated by an electromyographer who can perform a complete electrodiagnostic consultation, interpreting electrodiagnostic results in the context of the patient’s history, physical examination, and other pertinent studies such as imaging.

NERVE CONDUCTION STUDIES

NCS commonly assess either sensory or motor nerves, although mixed nerves can also be assessed using the same methods. These impulses are recorded as sensory nerve action potentials (SNAPs), compound muscle action potentials (CMAPs), or compound nerve action potentials (CNAPs). Measurements of conduction velocity and distal latency are reflective of the speed of propagation of action potentials via large myelinated axons. As such, slowing of these measurements may suggest dysmyelination. The conduction velocity can also be slow in remyelination after dysmyelination, reinnervation after axon loss injury, cold temperature, wallerian degeneration of the fastest conducting fibers, and axon stenosis. Amplitude of SNAPs is reflective of the number of axons in a sensory nerve. CMAP amplitude is reflective of the combined function of the motor neurons, the neuromuscular junction, and the striated muscle.

In NCS, nerves are stimulated percutaneously using electrical impulses. A recording electrode (identified as G1) is placed over the nerve or muscle, a reference electrode (G2) is placed nearby to filter out extraneous electrical
activity, and a ground is placed between stimulus and recording electrode to reduce stimulus artifact.

Supramaximal stimulation is an important concept in NCS. A submaximal stimulus will trigger a response in some, but not all, axons in a nerve. A maximal response will activate the entire group of axons. Once this point is reached, further increase in the intensity of stimulation will not change the response. When conducting a study, the electromyographer must increase the stimulus until no change in amplitude is noted between a response at a lower intensity of stimulus and the next higher level. This helps assure that accurate data are being obtained and recorded. Further increase in the stimulus beyond this point decreases the latency owing to distal migration of the cathode.

▶ Sensory Nerve Action Potentials (Figure 16–3)
<table>
<thead>
<tr>
<th>Stim Site</th>
<th>Rec Site</th>
<th>Onset (ms)</th>
<th>Peak (ms)</th>
<th>NP Amp (µV)</th>
<th>PP Amp (µV)</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
<th>Temp (°C)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wrist</td>
<td>DigV</td>
<td>2.70</td>
<td>3.65</td>
<td>62.5</td>
<td>112.9</td>
<td>14</td>
<td>51.9</td>
<td>32.1</td>
</tr>
</tbody>
</table>
SNAPs are recorded using two electrodes and a common ground. The active (G1) electrode is placed superficial to the nerve. A second electrode, the reference electrode (G2), is placed 4–5 cm distally. An electrical stimulus is delivered, creating an electrical field that propagates down the length of the nerve. The initial portion of the electrical field is considered positive. As it approaches the G1 electrode, this is recorded as an initial positive or downward deflection in the action potential waveform. As the impulse continues to pass beneath the G1 electrode, a large negative deflection is produced in the waveform. As the impulse travels away from the G1 electrode, a final positive portion of the nerve action potential waveform is seen. The distance between the G1 and G2 electrodes is important and should be at least 4 cm. If the electrodes are too close together, the signal may be distorted, with the beginning of the signal approaching the G2 before the end of the signal passes over the G1; this will cause attenuation of the amplitude.

SNAPs are useful in NCS because they establish the integrity of peripheral sensory nerves, which are often the first affected in conditions such as diabetic polyneuropathy or brachial plexopathy, and aid in assessment of sensory involvement in a pathologic process. Additionally, they can help distinguish between preganglionic and postganglionic lesions. The cell bodies of sensory neurons form in the dorsal root ganglia, which lie within the intervertebral foramina, where spinal nerve roots exit the spinal canal. In a preganglionic lesion, the distal sensory nerve remains intact and exhibits normal SNAPs even when there is a clinical sensory deficit. Any lesion that affects the nerve distal to the lesion causes a decrease in SNAP amplitude along with the clinical sensory deficit. This characteristic can help distinguish anterior horn cell disease or radiculopathy from plexopathy, or peripheral neuropathy.

SNAPs can be studied using an antidromic or orthodromic technique. Antidromic studies measure impulse propagation in the opposite direction from the physiologic response. Orthodromic studies measure propagation in the same direction as the physiologic response. Normative data for sensory nerve studies are included at the end of the chapter (see Box 16–1).
<table>
<thead>
<tr>
<th>Nerve</th>
<th>Conduction Velocity (m/s)</th>
<th>Distal Sensory Latency (ms)</th>
<th>SNAP (μV)</th>
<th>Distance (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median (index finger)</td>
<td>&gt; 40.0</td>
<td>&lt; 3.5 Side-to-side comparison &lt; 0.4</td>
<td>&gt; 20</td>
<td>14</td>
</tr>
<tr>
<td>Ulnar (little finger)</td>
<td>&gt; 45.0</td>
<td>&lt; 3.1 Side-to-side comparison &lt; 0.4</td>
<td>&gt; 18</td>
<td>14</td>
</tr>
<tr>
<td>Radial (first web space)</td>
<td>&gt; 40.0</td>
<td>&lt; 2.5</td>
<td>&gt; 20</td>
<td>10</td>
</tr>
<tr>
<td>Median versus radial (thumb in extension)</td>
<td>&lt; 2.5 Latency difference &lt; 0.4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median versus ulnar (ring finger sensory)</td>
<td>Latency difference &lt; 0.4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median versus ulnar (palm orthodromic)</td>
<td>Latency difference &lt; 0.4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Combined sensory index (CSI)&lt;sup&gt;a&lt;/sup&gt;</td>
<td>&lt; 1.0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lateral antebrachial cutaneous</td>
<td>61.6 ± 4.2</td>
<td>2.3 ± 0.2</td>
<td>18.9 ± 9.9 (&gt; 5.0)</td>
<td>14</td>
</tr>
<tr>
<td>Medial antebrachial cutaneous</td>
<td>62.7 ± 4.9</td>
<td>2.2 ± 0.2</td>
<td>11.4 ± 5.2 (&gt; 3.0) Compare to normal side</td>
<td>14</td>
</tr>
<tr>
<td>Deep peroneal sensory (first web space)</td>
<td>42.0 ± 5.0</td>
<td>2.9 ± 0.4</td>
<td>3.4 ± 1.2 Compare to normal side</td>
<td>12</td>
</tr>
<tr>
<td>Superficial peroneal, medial dorsal cutaneous branch Age adjusted</td>
<td>&lt; 30 = 42 31–50 = 40 51–70 = 39 &gt; 71 = 36</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sural Age adjusted</td>
<td>&lt; 30 = 43 31–50 = 39 51–70 = 36 &gt; 71 = 35</td>
<td>&lt; 30 = 3.2 31–50 = 3.5 51–70 = 3.6 &gt; 71 = 3.9</td>
<td>&lt; 50 = 8 31–50 = 5 51–70 = 5 &gt; 71 = present</td>
<td>14</td>
</tr>
<tr>
<td>Saphenous</td>
<td>Range 37–66</td>
<td>Range 2.1–3.8</td>
<td>Range 1–15 Compare to normal side</td>
<td>14</td>
</tr>
<tr>
<td>Lateral femoral cutaneous</td>
<td>Range 42–65</td>
<td>2.6 ± 0.2</td>
<td>Range 5–25 Compare to normal side</td>
<td>14</td>
</tr>
<tr>
<td>Plantar (tibial) sensory Medial plantar</td>
<td>3.2 ± 0.3</td>
<td>3.1 ± 0.3</td>
<td>Compare to normal side &gt; 10 &gt; 8</td>
<td>14</td>
</tr>
</tbody>
</table>

SNAP, sensory nerve action potential.

<sup>a</sup>Combines latency difference between (1) radial/median thumb, (2) ulnar/median ring finger, and (3) ulnar/median palm orthodromic.
Primary causes of error in SNAP measurement are cool limb temperature, increased or decreased distance between G1 and G2 electrodes, submaximal stimulus, edema, and noise artifact. Because the recorded potentials are of low amplitude, anything affecting impedance, such as edema, thick callused skin, or poor contact between the skin and electrodes, can affect the waveform morphology. Crossed wires, presence of another electrical device nearby, dry skin, and too little contact gel can all distort results. Cold temperature of the limb increases the latency and slows the conduction velocity but increases the SNAP amplitude.

► Compound Muscle Action Potentials (Figure 16–4)
Top Wave 1 Distal onset latency, 1–2 Amplitude
Bottom Wave 1 Proximal onset latency

R Median–thenar

<table>
<thead>
<tr>
<th>Sites</th>
<th>Rec Site</th>
<th>Lat (ms)</th>
<th>Amp (mV)</th>
<th>Rel Amp (%)</th>
<th>Segments</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
<th>Temp (°C)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wrist</td>
<td>APB</td>
<td>3.10</td>
<td>10.6</td>
<td>100</td>
<td>Wrist-APB</td>
<td>8</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Elbow</td>
<td>APB</td>
<td>6.85</td>
<td>10.6</td>
<td>99.7</td>
<td>Elbow-Wrist</td>
<td>21.7</td>
<td>57.9</td>
<td>32.1</td>
</tr>
</tbody>
</table>
In recording CMAPs, electrode location is key, as amplitude of the CMAP decreases with distance of the G1 electrode from the endplate zone of the muscle. The G2 electrode is placed over an inactive area, such as the tendon–muscular junction. In contrast to sensory studies, the G2 location is not at a fixed distance. For motor studies, the impulse is conducted from stimulus to motor endplate, and exhibits an initial negative, or upward, deflection. This is followed by a sharp rise and then a positive deflection as the impulse passes the recording electrode. A larger stimulus is required to obtain CMAPs than SNAPs, and waveforms are larger and well defined. If an initial positive deflection occurs, the G1 recording electrode may not be positioned over the motor endplate. Moving the active electrode to eliminate any initial downward deflection ensures that the onset latency is accurate. Given the importance of this latency in determining whether the result is normal or abnormal, it is imperative that the conduction technique be optimal. In fact, to obtain the most accurate distal latency, it is recommended that the onset latency be measured at a gain of 200 μV, magnifying the result so that the true onset of the waveform can be detected. Normative data for motor studies are included at the end of the chapter (see Box 16–2).
### Box 16–2 Normal values for motor nerves.

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Distal Motor Latency (ms)</th>
<th>CMAP (mV)</th>
<th>Conduction Velocity (m/s)</th>
<th>Distance (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median (APB)</td>
<td>&lt; 4.2</td>
<td>&gt; 4.0</td>
<td>&gt; 49.0</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Side-to-side comparison &lt; 0.6</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar (ADM)</td>
<td>&lt; 3.5</td>
<td>&gt; 4.0</td>
<td>&gt; 49.0</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Side-to-side comparison &lt; 0.6</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median vs. ulnar (APB versus ADM motor)</td>
<td>Latency difference &lt; 1.0</td>
<td></td>
<td></td>
<td>8</td>
</tr>
<tr>
<td>Axillary</td>
<td>3.9 ± 0.5</td>
<td>Compare to normal side</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Musculocutaneous</td>
<td>4.5 ± 0.6</td>
<td>Compare to normal side</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Suprascapular Supraspinatus</td>
<td>2.7 ± 0.5</td>
<td>Compare to normal side</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infraspinatus</td>
<td>3.3 ± 0.5</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Radial (EIP)</td>
<td>2.4 ± 0.5</td>
<td>Side-to-side comparison &lt; 50% abnormal</td>
<td>61.6 ± 5.9</td>
<td>8</td>
</tr>
<tr>
<td>Deep peroneal (EDB)</td>
<td>&lt; 6.0</td>
<td>&gt; 2.0</td>
<td>&gt; 40.0</td>
<td>8</td>
</tr>
<tr>
<td>Tibial (AH)</td>
<td>&lt; 4.8</td>
<td>&gt; 2.0</td>
<td>&gt; 40.0</td>
<td>10</td>
</tr>
<tr>
<td>Common peroneal</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tibialis anterior</td>
<td>3.0 ± 0.6 (4.2)</td>
<td>3.9 ± 1.2</td>
<td>66.3 ± 12.9</td>
<td></td>
</tr>
<tr>
<td>Peroneus longus</td>
<td>3.0 ± 0.6 (4.6)</td>
<td>5.9 ± 2.4</td>
<td>55.3 ± 10.2</td>
<td></td>
</tr>
<tr>
<td>Femoral (VM)</td>
<td>6.0 ± 0.7 (7.4)</td>
<td>12.1 ± 5.1 (3.7)</td>
<td>Compare to normal side</td>
<td></td>
</tr>
<tr>
<td>F wave&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Minimum latency</td>
<td>Chronodispersion</td>
<td>Median 4 ms</td>
<td>Ulnar 4 ms</td>
</tr>
<tr>
<td></td>
<td>Median 22–30</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ulnar 22–31</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Peroneal 37–53</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tibial 40–59</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tibial H reflex (soleus)&lt;sup&gt;b&lt;/sup&gt;</td>
<td>25–34</td>
<td>Side-to-side comparison &lt; 1.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Facial</td>
<td>3.6 ± 0.35 (&lt; 4.1)</td>
<td>Compare to normal side</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spinal accessory</td>
<td>Upper trapezius 1.8–3.0</td>
<td>Compare to normal side</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mid trapezius 2.6–3.4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Low trapezius 4.0–5.2</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

CMAP, compound muscle action potential; APB, abductor pollicis brevis; ADM, abductor digiti minimi; EIP, extensor indicis proprius; EDB, extensor digitorum brevis; AH, abductor hallucis; VM, vastus medialis.

<sup>a</sup>Consider age, height and limb length.

<sup>b</sup>Consider age and limb length.
Motor NCS are useful for objective assessment of the anterior horn cell, motor nerve, neuromuscular junction, and muscle fiber. The amplitude of the response is decreased in anterior horn cell disease, axon loss lesion of the motor nerve, neuromuscular junction disorders such as Lambert–Eaton myasthenic syndrome, and in myopathy, where there is muscle fiber loss. These conduction studies are thus helpful in identifying the location of focal disease, evaluating the severity of peripheral disease, and defining the extent of neurologic injury.

The primary causes of error in CMAP measurement are cool temperature, submaximal stimulation (because of inappropriate stimulator location), low or high intensity of the stimulus delivered, inadequate pressure on the nerve, poor placement of the G1 electrode, and measurement error. Measurement error will affect conduction velocity and distal latency. Amplitude is affected if G1 is not placed over the motor endplate and with submaximal stimulation. Additionally, an initial positive deflection of the response may be seen if electrode placement is not over the muscle endplate. Cool limb temperature causes increased amplitude, increased distal latency, and decreased conduction velocity.


Late Responses

F wave and H reflex recordings are NCS techniques that can be useful in the context of other findings from the clinical and electrodiagnostic examination. They are most often used when the results of NCS are equivocal and are particularly valuable in the diagnosis of conditions that begin more proximally, such as acute inflammatory demyelinating polyradiculoneuropathy, in which F and H latencies are prolonged. They are also useful in the setting of acute paralysis or paresis with pronounced weakness, where their presence confirms intact function of the peripheral nerve. Occasionally these responses can be valuable in evaluating metabolic or vasculitic proximal plexus or nerve root pathology. H reflexes obtained from the tibial nerve are often used and may be helpful in the diagnosis of S1 radiculopathy, and to differentiate it from L5 root involvement.

A. Wave (Figure 16–5)
M wave
T1 M latency
T2 Minimum F latency

Mean latency: Average of 10 F latencies
Range or chronodispersion: Difference between minimum and maximum F latency
%F or persistence: Total number of F waves/Total number of stimulations
Left ulnar–Abductor digiti minimi

<table>
<thead>
<tr>
<th>Stats</th>
<th>F Latency (ms)</th>
<th>F Persistence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>F Min Lat</td>
<td>28.50</td>
<td></td>
</tr>
<tr>
<td>F Max Lat</td>
<td>31.25</td>
<td></td>
</tr>
<tr>
<td>F Mean Lat</td>
<td>29.62</td>
<td></td>
</tr>
<tr>
<td>F Range</td>
<td>2.75</td>
<td>100</td>
</tr>
</tbody>
</table>
The F wave recording measures a stimulus as it travels antidromically along the motor axon to the body of the anterior horn cell in the spinal cord. This action triggers a secondary discharge within the same anterior horn cell, due to summation of inhibitory and excitatory influences on the cell. The inhibitory influences are the result of Renshaw inhibition, refractory period of the axon hillock, central inhibition, and the collision of the distally propagating response and proximal spread of the stimulus from the distal stimulation. Excitatory influences are dendritic stimulation of the neighboring motor neurons, current sink produced at the dendrites, and reversal of the refractory period at the axon hillock. Only a small proportion (2%) of the available anterior horn cells are activated by the antidromic stimulation. The resultant response is approximately 5% of the size of the CMAP that is obtained with stimulation. The stimulus provided is a supramaximal stimulus of the nerve distally, such as in the motor NCS.

Termed F waves, because they were first measured in the foot, these responses have long latencies and represent nerve conduction from periphery to spinal cord. Their waveform, amplitude, and latency change from stimulus to stimulus. F waves are studied from distal muscles in the limbs. The more proximal the stimulation site, the more difficult it is to isolate the response due to superimposition of the obtained CMAP and the F wave. For this reason, F waves are most often obtained with stimulation of the median or ulnar nerves at the wrist and the tibial or peroneal nerves at the ankle, with recording in the hand and foot muscles.

A recording electrode (G1) is placed on the muscle belly, the reference electrode (G2) is placed distal to this location, and a supramaximal stimulus is delivered. F waves are much smaller than the initial motor response (often called the M wave) and are therefore recorded at a lower gain. A minimum of 10 responses is necessary for evaluation of latency. Minimum latency is measured to the earliest recorded F wave. These measurements can be obscured by muscle contraction and may also be absent in normal nerves.

Prolongation of the minimal latency and increased delay between minimum and maximal latencies (chronodispersion) are evidence of dysmyelination. Decreased persistence of the F waves and presence of repeater waves (waveforms having the same characteristics) may suggest axon loss or proximal neurapraxic block. Increased amplitude ratios of F and M waves may suggest an upper motor neuron lesion.


**B. Reflex (Figure 16–6)**
Figure 16–6 H reflex.

Discovered by Paul Hoffman in 1910, the H reflex is a late response that is used
to measure the electrical activity of muscles following stimulus of the Ia afferent fibers. In normal adults this reflex can be recorded reliably over a limited number of muscles, including the soleus (with stimulation of the tibial nerve), flexor carpi radialis (with stimulation of the median nerve), and quadriceps (with stimulation of the femoral nerve). H reflexes are also frequently found in the plantar muscles. The H reflex is activated by a long-duration and small-intensity electrical pulse that stimulates the Ia sensory fibers in the nerve being tested. The Ia fibers typically have the lowest electrical threshold for creating an action potential. This ascending potential synapses to the anterior horn cells and elicits a motor response similar to the monosynaptic reflex response of the tendon tap. The motor response that is recorded over the muscle innervated by the nerve being stimulated is the H reflex. The initial small stimulus of the Ia fibers elicits a small excitatory postsynaptic potential. As this submaximal response is slowly increased in intensity, a more defined, larger H reflex response occurs as more muscle fibers are recruited. Further increase in the stimulus extinguishes the H reflex response and produces the F wave response. This increased stimulus also begins to cause a true motor response (the M wave).

The H reflex is useful for assessing proximal nerve segments that are inaccessible through routine NCS. It can help establish the diagnosis of a clinically suspected radiculopathy, plexopathy, or neuropathy when routine NCS are normal. It can also be used in the evaluation of spasticity. The reflex is easily obtained in all nerves in children younger than 2 years of age. However, it can be difficult to obtain in adults in all the nerves for the following reasons: (1) not all nerves are easily accessible with surface electrodes, (2) the afferent stimulus may not be sufficient to activate a motor response, and (3) in some mixed muscle nerves, the Ia afferent threshold is so close to the activation of the motor fibers that the H response is effectively effaced. Therefore, positioning, stimulus, and recording variables must be closely monitored to ensure the accuracy of the response. The response is typically measured in terms of latency, maximal H amplitude, and H-to M-wave amplitude ratio.


Goldberg G, Sridhara CR: Clinical neurophysiology of the peripheral nervous
Blink Reflex (Figure 16–7)
### Table

<table>
<thead>
<tr>
<th>Sites</th>
<th>Muscle</th>
<th>R1 (ms)</th>
<th>R2 Ipsi (ms)</th>
<th>R2 Contra (ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stim L Rec Ipsi</td>
<td>L Orb Oculi</td>
<td>10.15</td>
<td>31.30</td>
<td></td>
</tr>
<tr>
<td>Stim L Rec Cont</td>
<td>R Orb Oculi</td>
<td></td>
<td></td>
<td>27.70</td>
</tr>
<tr>
<td>Stim R Rec Ipsi</td>
<td>R Orb Oculi</td>
<td>10.70</td>
<td>30.40</td>
<td></td>
</tr>
<tr>
<td>Stim R Rec Cont</td>
<td>L Orb Oculi</td>
<td></td>
<td></td>
<td>27.20</td>
</tr>
</tbody>
</table>
Evaluation of the blink reflex is one of the most commonly performed cranial nerve studies. The underlying mechanisms of this reflex were not fully understood until 1952 when Kugelberg used electrical stimulation to analyze the supraorbital nerve. The reflex involves an afferent arc of the trigeminal nerve and an efferent arc through the facial nerve. It can be used to assess lesions of the facial and trigeminal nerves, peripheral neuropathies, and also in the evaluation of patients with multiple sclerosis or other brainstem lesions.

The reflex is best recorded with the patient relaxed and lying quietly on the examination table. The active (G1) electrodes are placed over the midpoint of the orbicularis oculi muscles on the lower lids, and reference (G2) electrodes are placed over the outer canthus of the eyes. The ground electrode is placed over the chin. Stimulus is administered to one side at a time over the supraorbital notch, with the cathode over the nerve and the anode directed away from the midline. The response is recorded simultaneously over both eyes. An early (R1) response is obtained ipsilaterally, and subsequent (R2) responses are obtained both ipsilateral and contralateral to the stimulation. The stimulus intensity is adjusted to elicit a stable R1 response with repeated stimulations. The stimulus interval should be at least 5–10 seconds to avoid habituation of the reflex response. Technical errors can easily confound test results or eradicate the response. The most common errors are using either a sweep speed that is too fast or a gain that is too low. Background muscle activity can also obfuscate the response.

The R1 is an oligosynaptic reflex and is thought to have an ipsilateral course through the lower pons. It is generally a stable response in relationship to latency and amplitude. The R2 is a polysynaptic response with variable latencies and amplitudes likely to be mediated through the pons and the lateral medulla in a more complicated route. A third response (R3) may occur if the stimulus threshold is high enough to evoke activation of the smaller diameter, high-threshold afferent sensory fibers. The R1 and R2 latencies are measured with stimulation of the supraorbital nerves bilaterally, and at least five stimulations are used in obtaining the latencies (Table 16–3).

Table 16–3 Blink reflex: normal values.
In lesions of the trigeminal nerve, responses will be delayed or absent when stimulating the affected side and present on the unaffected side. In idiopathic trigeminal neuralgia or with atypical facial pain, responses are usually normal. Abnormality of the R1 response may indicate a space-occupying lesion affecting the sensory root, a lesion of the brainstem, sensory neuropathy of the trigeminal nerve, or neuropathy of the facial nerve on the ipsilateral side. In facial nerve lesions, the R2 response latency is delayed on the affected side regardless of the side receiving stimulus apart from the abnormality of the R1 latency on the ipsilateral side (Table 16–4). In peripheral cranial nerve VII paralysis, prognosis is good when blink reflex is normal or only R1 is delayed. Prognosis for recovery is poor in patients with an absent blink reflex. Abnormal R2 responses with normal R1 responses indicate a central lesion of the spinal tract and nucleus of the trigeminal nerve in the medulla.

Table 16–4 Blink reflex—interpretation of abnormalities.
**NEEDLE ELECTROMYOGRAFHY**

Needle EMG is performed by inserting a needle directly into the muscle and recording the insertional activity, spontaneous activity, and motor unit action potentials (MUAPs) from the muscles in the vicinity of the needle (Figure 16–8). Different potentials have different characteristic amplitudes, durations, and waveforms, which indicate whether the potential is a motor unit or a spontaneous potential (Table 16–5). The spontaneous potentials (fibrillations and positive sharp waves) are muscle fiber potentials that are recorded from muscles that have sustained denervation of their nerve axons secondary to wallerian degeneration or as a result of local muscle injury (see Figure 16–8). Normal muscle is electrically silent at rest except at the neuromuscular junction. Fibrillations and positive sharp waves may be graded on a 1–4 scale, depending on their prevalence with needle testing. This rating does not correlate with degree of axonal loss; only the amplitude of the CMAP is proportional to the degree of axonal loss or muscle fiber loss.
**Figure 16–8** Motor unit action potential.

**Table 16–5** Characteristics of EMG potentials.
Complex repetitive discharges (CRDs) and myotonic discharges are high-frequency discharges that are generated in muscle fibers as a result of abnormal recurrent muscle fiber group contraction. Myotonia sounds like a “dive bomber” on the audio track of the EMG machine and may be present in myotonic dystrophy, paramyotonia, and hyperkalemic periodic paralysis. Needle movement generally evokes these potentials. CRDs are caused by ephaptic discharge in a group of muscle fibers that start and stop suddenly and have a motorbike–like sound on the audio track. These potentials may be noted in patients with chronic neuropathies, chronic axon loss lesions, Duchenne muscular dystrophy, or spinal muscular atrophy.

Fasciculations and myokymic discharges are potentials that are generated in the motor unit potentials. Fasciculations are involuntary and irregularly firing single MUAPs that may be benign or indicative of anterior horn cell or peripheral nerve pathology. If accompanied by abnormalities of spontaneous activity or of MUAPs, they are considered pathologic; otherwise, they are considered benign. Myokymic discharges are grouped fasciculations and are seen in radiation plexopathy, myelopathy, chronic entrapment, radiculopathy, or acute idiopathic demyelinating radiculoneuropathy. Myokymic discharges have a semirhythmic sound like that of a marching soldier on the audio track.

The MUAP is the electrophysiologic correlate of the discharge from one anterior horn cell and coordinated summated electrical activity of all the muscle fibers innervated by that anterior horn cell. The shape and size of the MUAP

<table>
<thead>
<tr>
<th>Spontaneous Activity</th>
<th>Amplitude</th>
<th>Duration</th>
<th>Shape</th>
<th>Rate</th>
<th>Firing Pattern</th>
<th>Sound</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fibrillations</td>
<td>20–300 μV</td>
<td>&lt; 5 ms</td>
<td>Biphasic or triphasic</td>
<td>2–20/s</td>
<td>Regular</td>
<td>Raindrops</td>
</tr>
<tr>
<td>Positive sharp waves (PSWs)</td>
<td>20–300 μV</td>
<td>10–100 ms</td>
<td>Positive spike with long tail</td>
<td>5–10/s</td>
<td>Regular</td>
<td>Dull thud</td>
</tr>
<tr>
<td>Fasciculation</td>
<td>500–5000 μV</td>
<td>5–20 ms</td>
<td>Biphasic, triphasic, or polyphasic</td>
<td>1–5 s between discharges</td>
<td>Irregular</td>
<td>Popcorn</td>
</tr>
<tr>
<td>Myotonia</td>
<td>10–1000 μV</td>
<td>Variable</td>
<td>Resemble fibrillations and PSWs</td>
<td>15–150 Hz</td>
<td>High-frequency, wax and wane</td>
<td>Dive bomber</td>
</tr>
<tr>
<td>Complex repetitive discharges</td>
<td>100–1000 μV</td>
<td>Stable</td>
<td>Polyphasic stable</td>
<td>10–50 Hz</td>
<td>High-frequency, constant</td>
<td>Motorbike</td>
</tr>
<tr>
<td>Myokymia</td>
<td>500–1000 μV</td>
<td>Stable</td>
<td>Polyphasic</td>
<td>Up to 50 Hz</td>
<td>Semirhythmic</td>
<td>Marching soldiers</td>
</tr>
</tbody>
</table>
depend on the location of the needle electrode in relation to the activated muscle fibers. Small needle movement may cause significant change in the shape and size of the MUAP. The amplitude and duration of the MUAP is measured when the rise time of the MUAP is the shortest; the sound of the unit is a sharp “pop” on the audio track of the machine. There are several parameters of the MUAP, as illustrated in Figure 16–8.

Another EMG parameter is recruitment, or the extent to which more motor unit potentials can fire in response to increasing muscle contraction. Recruitment is full and spontaneous when axonal integrity is preserved but becomes more limited with axon loss. Decreased recruitment is often the first finding noted on needle EMG in axon disruption. This should be evaluated, along with the presence of fibrillations and positive sharp waves, before concluding that there is evidence of acute axonal loss as decreased recruitment may also be present in patients with partial neurapraxic block.

When patients can recruit only a few MUAPs, the central nervous system may compensate by increasing the firing rate of the MUAP. A trained electromyographer will be able to hear this decrease in recruitment on the audio track. Patients with decreased recruitment usually demonstrate corresponding clinical motor weakness on physical examination. Nerves have the capacity to regenerate (at a rate of 1 inch per month). Over time, the fibrillations and positive waves become smaller due to atrophy of the muscle fiber or disappear with reinnervation or fibrosis of the muscle fiber. The MUAPs may have multiple phases (polyphasic MUAPs), suggesting immature axonlets that are conducting at different rates, but as axons mature, the number of phases may decrease and amplitude of the MUAP may increase as a result of better summation of the individual muscle fiber action potentials. Once the reinnervation is completed, the firing rate may sound more normal due to better recruitment of the MUAPs.


Herbison G: EMG: Waveform Analysis. 33rd annual course in Electrodiagnostic Medicine and Clinical Neurophysiology, Jefferson
ELECTRODIAGNOSTIC INSTRUMENTATION

A specialized instrument, the electromyograph, is used to perform electrodiagnostic studies. The physiologic signals it records are tiny and must be amplified to be displayed and measured. Three electrodes are always attached to the patient and include an active, a reference, and a ground electrode. The electrical activity produced by the anatomic structures in the patient’s body is transmitted through volume conduction and is converted into an electrical signal. The signal is transmitted to the instrument by the electrodes. Increased impedance in the electrode–patient interface is related to poor contact and may cause significant “movement artifact” of the biologic signals. Use of conducting gel between the electrodes and the skin surface, lightly abrading the skin to remove the stratum corneum of the skin, use of short electrode leads, and use of shielded cables decrease the impedance and improve recording of the small biologic potentials. Several types of recording needle electrodes are available, as listed in Table 16–6.

Table 16–6 Types of needle electrodes.

<table>
<thead>
<tr>
<th>Type of Electrode</th>
<th>Monopolar</th>
<th>Concentric</th>
<th>Bipolar</th>
<th>Single Fiber</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recording surface</td>
<td>0.15–0.20 mm²</td>
<td>0.05–0.08 mm²</td>
<td>0.05–0.08 mm²</td>
<td>0.025 mm²</td>
</tr>
<tr>
<td>Reference electrode</td>
<td>Separate at a distance</td>
<td>Needle cannula</td>
<td>Second wire in cannula</td>
<td>Needle cannula</td>
</tr>
<tr>
<td>Ground electrode</td>
<td>Separate at a distance</td>
<td>Separate at a distance</td>
<td>Needle cannula</td>
<td>Separate at a distance</td>
</tr>
<tr>
<td>MUAP recording</td>
<td>Less selective</td>
<td>Good, isolate individual MUAP</td>
<td>Excellent, least artifact</td>
<td></td>
</tr>
<tr>
<td>Patient tolerance</td>
<td>Better</td>
<td>Lower</td>
<td>Lowest</td>
<td>Low</td>
</tr>
</tbody>
</table>

MUAP, motor unit action potential.
The preamplifier is a differential amplifier to which the cables are attached in the electromyographic system. It amplifies the difference between the two signals that are generated at the active and reference electrodes, rejecting any signal that is common to the two inputs. In other words, it amplifies the difference in electrical potential between the active and ground electrode, and that between the reference and the ground electrode. Any common interference potentials (eg, 60-cycle interference from an electrical line) is rejected by the differential amplifier. The electrode impedance of both active and reference electrodes should be of close match for the preamplifier to carry out the common mode rejection.

The EMG signal utilizes frequencies lower than 10,000 Hz, whereas most of the amplifier noise involves frequencies above this level. Thus, a filter cutoff frequency of 10,000 Hz (low-pass filter) allows the biological signal to be recorded without extraneous noise. Frequencies below 20 Hz in the EMG recording are usually a result of movement, and a filter cutoff of 20 Hz (high-pass filter) will minimize the noise. Filter settings are also used for nerve conduction velocity studies (Table 16–7). An ideal amplifier converts a low-voltage waveform to a higher voltage copy of the same waveform. To do this, the input impedance of the preamplifier has to be very high. The amplifier filters the signal as it acquires it in real time, with only minimal filtering or distortion of the waveform.

**Table 16–7** Filter settings for electromyography and nerve conduction studies.

<table>
<thead>
<tr>
<th>Signal</th>
<th>High-Pass Filter (Hz)</th>
<th>Low-Pass Filter (Hz)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensory nerve action potential (SNAP)</td>
<td>20</td>
<td>2000</td>
</tr>
<tr>
<td>Compound muscle action potential (CMAP)</td>
<td>10</td>
<td>10,000</td>
</tr>
<tr>
<td>Electromyography (EMG)</td>
<td>20</td>
<td>10,000</td>
</tr>
</tbody>
</table>
POLYNEUROPATHY

Diabetes mellitus is the most common cause of peripheral neuropathy worldwide. In clinical practice, the two conditions often seem to be synonymous: more than half of all patients with diabetes have neuropathy, and half the patients with neuropathy have diabetes. Disease-related changes in diabetic individuals with neuropathy cause a wide range of presentations, reflecting acute or chronic symptoms in a variety of anatomic locations, such as skin, nerve root, vasculature, and autonomic nervous system. The discussion that follows focuses on three characteristic presentations: distal symmetric polyneuropathy, diabetic amyotrophy, and diabetic autonomic neuropathy. Characteristic features of each are contrasted in Table 17–1. 

Focal mononeuropathy caused by diabetes produces symptoms similar to those of compressive or entrapment neuropathies, which are discussed at the end of this chapter.

Table 17–1 Key features of polyneuropathies
DISTAL SYMMETRIC POLYNEUROPATHY

ESSENTIALS OF DIAGNOSIS

- Most common form of peripheral neuropathy in diabetic patients as well as worldwide.
- Findings are chronic, distal, symmetric, and sensory predominant.
- Symptoms include tingling or burning pain, sensory loss, or numbness.
- Electrodiagnostic studies show axonal demyelinating neuropathy.

General Considerations

Distal symmetric polyneuropathy (DSP) is the most common form of peripheral neuropathy in diabetic patients, and also the most common form worldwide. DSP is a major risk factor in foot ulceration and eventual limb amputation. Neuropathy, sensory loss, and distal weakness are major risk factors for falls, and the confluence of these findings in patients with DSP increases the fall risk.
sevenfold. For these reasons, DSP is a significant cause of disability and reduced quality of life. Neuropathy may progress even in patients who achieve excellent glycemic control. Additional risk factors for neuropathy in diabetic patients, independent of glucose control, include obesity and dyslipidemia.

### Clinical Findings

#### A. Symptoms and Signs

Typical findings in DSP are chronic, distal, symmetric, sensory predominant, and very painful. DSP can cause a variety of positive and negative symptoms or it can be asymptomatic. Positive symptoms include prickling, tingling, or burning. Negative symptoms consist of sensory loss or numbness. Severe neuropathy can result in painless injury. Pain is length dependent and involves the feet, toes, calves, and hands. Pain is worse with walking but is most severe at night, often leading patients to sleep with their feet on top of the covers or make a “tent” with the sheet. Patients present with reduced distal sensation to pinprick, cold temperature, and vibration, often in a stocking-and-glove distribution. Deep tendon reflexes are reduced or absent, and proprioception or joint position sense is likewise reduced. Distal muscles in the hand and feet may atrophy. Gait can be ataxic, with a positive Romberg’s sign.

#### B. Laboratory Findings

Patients often have elevated hemoglobin A$_{1c}$ levels, reflecting difficulty in attaining good blood glucose control.

#### C. Electrophysiologic Findings

Similar to other axonal demyelinating neuropathies, the central feature of DSP is reduced sensory nerve action potential (SNAP) amplitudes and conduction velocities with mild motor conduction slowing in the distal extremity. Distal latencies may be prolonged, and lower extremities are affected before upper extremities. Diagnosis can be challenging as DSP preferentially involves small nerve fibers, which may result in a normal nerve conduction study. Needle electromyography (EMG) usually reveals spontaneous activity with active denervation of the distal muscles of the legs. With severe disease, fibrillations and positive sharp waves may be seen in the hand muscles.
Complications

Diabetic neuropathy is a major cause of morbidity and mortality. Pain is a frequent complaint. It is estimated that 20% of patients with DSP experience severe pain. DSP is also a leading risk factor for foot ulceration, infection, and eventual foot or limb amputation.

Treatment

First-line treatment involves diet, blood glucose control, and exercise counseling. Experts believe the only therapy that is effective at reducing risk and slowing progression of diabetic neuropathy is aggressive glycemic control. Currently there are no treatments that reverse DSP. Treatment of neuropathic pain is challenging and often requires multiple pharmacologic agents. Medications used include anticonvulsive agents such as gabapentin, serotonin norepinephrine reuptake inhibitors such as duloxetine, and tricyclic antidepressants such as amitriptyline. Counseling to teach patients about fall risk, as well as formal gait evaluation to assess patients on uneven or irregular surfaces, where they are most likely to fall, are beneficial measures.

Prognosis

As previously noted, aggressive glycemic control may be the only therapy effective at reducing risk and slowing progression of diabetic neuropathy; however, this does not eliminate or reverse progression of the disease.

DIABETIC AMYOTROPHY

ESSENTIALS OF DIAGNOSIS

- Rapid onset of pain and weakness in one leg, involving the proximal muscles first (vastus lateralis, adductors, buttock muscles), eventually progressing to distal leg muscles.
- Electrodiagnostic studies show axonal loss more than demyelination.
General Considerations

Patients with diabetic amyotrophy usually present with unilateral thigh pain. Also known as diabetic lumbosacral radiculoplexus neuropathy, diabetic amyotrophy should be considered in the differential diagnosis of radiating back pain, focal leg weakness, or lumbosacral radiculopathy in patients with diabetes mellitus. The condition typically affects older patients with noninsulin-dependent diabetes mellitus and, unlike DSP, is not related to glycemic control or duration of diabetes. Proper diagnosis is important to ensure appropriate treatment and prevent unnecessary lumbar procedures and surgeries.

Clinical Findings

A. Symptoms and Signs

Classically patients with diabetic amyotrophy present with acute onset of severe unilateral back, hip, or thigh pain, or some combination of these, with pain described as deep, aching, burning, or tingling. The condition evolves, producing progressive weakness and atrophy. Symptoms begin proximally and spread throughout the limb, with distal involvement usually arising later in the progression. Pain is exacerbated at night. If involved, the contralateral limb is affected months later. The lower extremities are preferentially affected, but some upper extremity involvement, characterized by pain and weakness, is also common. Diabetic amyotrophy can also manifest with autonomic symptoms, including changes in sexual, bowel, and bladder function and unexplained weight loss.

B. Laboratory Findings

Cerebrospinal fluid (CSF) examination reveals elevated protein without pleocytosis.

C. Electrophysiologic Findings

Electrodiagnostic studies demonstrate evidence of an axonal polyradiculoneuropathy with active denervation of the hip girdle, distal muscles, and paraspinal muscles.
D. Diagnostic Imaging
Radiographic, magnetic resonance imaging (MRI), and computed tomography scans of the lumbosacral and pelvic region should be obtained to rule out spinal cord lesions, spondylosis, nerve root compression, or a pelvic mass.

E. Nerve Biopsy
Nerve biopsies demonstrate microvasculitis.

Complications
Diabetic amyotrophy often causes significant pain and weakness that leads to impaired functioning, resulting in wheelchair dependence and severe disability.

Treatment
The clinical presentation, CSF findings, histopathology, and discordance with metabolic severity in diabetic amyotrophy all support an autoimmune etiology. Immunosuppressive agents or corticosteroid therapy may be beneficial in some patients, but conclusive studies on efficacy are lacking. Many patients require aggressive pain control with opiates.

Prognosis
The disease course is progressive over weeks to months but is usually self-limiting. While some improvement can be expected, patients are often left with permanent deficits.

DIABETIC AUTONOMIC NEUROPATHY

ESSENTIALS OF DIAGNOSIS

★ Diabetic patients with poor glucose control, prolonged duration of disease, or peripheral neuropathy are at risk.
Manifestations include orthostasis, gastroparesis, and erectile dysfunction.

Cardiac dysautonomia is suggested by orthostatic hypotension, presyncope, or exercise intolerance.

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### General Considerations

Diabetic autonomic neuropathy (DAN) is a common but underdiagnosed manifestation of diabetes mellitus that has a wide array of clinical presentations and can affect most systems in the body. The presence of DAN is associated with an increased risk of cardiac death and can result in other complications such as orthostatic hypotension, erectile dys-function, gastroparesis, and hypoglycemia. Every internal organ as well as the skin has autonomic innervation and thus a potential for neuropathy causing dysautonomia. Patients with poor glucose control, prolonged diabetes, or peripheral neuropathy are at risk for DAN.

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### Clinical Findings

#### A. Symptoms and Signs

Most patients are asymptomatic or present with vague symptoms. Cardiovascular symptoms and signs include orthostasis, arrhythmia, silent ischemia, and exercise intolerance. Gastrointestinal manifestations include nausea, early satiety, and constipation or diarrhea. Genitourinary symptoms and signs are erectile dysfunction, reduced vaginal lubrication, and neurogenic bladder. Cutaneous signs manifest as anhidrosis, dry skin, hair loss, and heat intolerance. Central symptoms and signs include asymptomatic hypoglycemia and a reduced hypoxia-induced ventilatory drive. A validated self-report clinical questionnaire for autonomic symptoms is recommended as it can improve diagnostic sensitivity for diabetic dysautonomia.

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#### B. Electrophysiologic Findings

Autonomic neuropathy can be evaluated in the EMG laboratory with studies such as heart rate variation of the R-R interval, with breathing and sympathetic skin response. Because autonomic function varies with age, matched control values must be used.
C. Other Tests

Additional tests that confirm certain aspects of diabetic dysautonomia include gastric emptying studies; colonoscopy; blood pressure variability to grip, standing, and tilt; nocturnal penile plethysmography; measurement of postvoid residuals; and thermoregulatory sweat testing.

Treatment

Although there is no definitive treatment of DAN, strategies should be implemented that provide symptomatic relief of the associated manifestations and assist patients in achieving good blood glucose control. Blood pressure fluctuations can be controlled using midodrine or fludrocortisone.

Prognosis

Detection of diabetic dysautonomia is critical because cardiac dysautonomia increases all-cause mortality risk two- to fivefold. It can also lead to further comorbidities such as accelerated neuropathy, malignant arrhythmias, and labile blood pressures, increasing the patient’s risk for a life-threatening cardiovascular or neurovascular event.

Infectious neuropathies are a significant cause of neuropathy worldwide. The mechanism of injury can involve the infectious agent directly, result from the body’s immune reaction to the infection, or occur secondary to drug toxicity from agents used to treat the infection.
LEPROSY

Leprosy is one of the most common treatable causes of peripheral neuropathy in the world. Clinically the disease usually manifests in the skin and the nerves. Pure neural leprosy (PNL) occurs in 4–10% of patients as a purely neuritic form of the disease. The lack of skin lesions can make it difficult to diagnose. Mononeuritis is often the most common presentation in patients with PNL, and the ulnar nerve is most commonly affected. Cranial nerve involvement is seen in 18% of PNL patients, with the facial and trigeminal nerves most often affected. The primary risk factor for development of neuropathy is the presence of a skin lesion overlying a nerve trunk. There is no significant correlation between the severity of the clinical signs and the histopathologic findings.

Leprosy-related neuropathy can be confirmed by EMG or nerve biopsy results. Leprosy causes a predominantly axonal loss neuropathy, which is most severe in the lower limbs. Distal symmetric and sensorimotor polyneuropathy is seen more often than mononeuropathy. Needle EMG reveals denervation in the small muscles of the hands and feet. In addition, the sympathetic reaction of the skin is almost always reduced.

Multidrug therapy using rifampicin, clofazimine, and dapsone is the usual treatment approach, but prevention of leprosy is the primary objective.

HUMAN IMMUNODEFICIENCY VIRUS

Peripheral neuropathy is the primary neurologic complaint of people infected with human immunodeficiency virus (HIV), occurring in 35% of patients with AIDS. Clinical manifestations include acute or chronic inflammatory neuropathy, polyradiculopathy, and distal symmetric neuropathy (DSN), which is the most common form of neuropathy. Signs and symptoms include dysesthesias, paresthesias, numbness, and decreased sensation to pain, temperature, and vibration, which occurs in the feet initially and then moves to the hands. Ankle reflexes are almost always absent. Several independent factors are associated with the development of DSN, including severity of HIV infection (nadir CD4+ count and plasma HIV RNA load), diabetes mellitus, alcohol abuse, race, and perhaps most commonly, antiretroviral medications such as stavudine, didanosine, and zalcitabine.

EMG reveals distal and symmetric degeneration of sensory and motor axons. Patients with HIV-related neuropathy can also present with inflammatory demyelinating polyneuropathy, multiple mononeuropathies, autonomic neuropathy, or polyradiculopathy. Nerve biopsies in HIV patients with DSN
show axonal degeneration due to proinflammatory cytokines.

Both treatment and prevention should include highly active antiretroviral therapy (HAART). Over the past two decades, the incidence of DSN among HIV-infected patients has decreased as the percentage of patients receiving HAART increased. But while early initiation of HAART therapy may decrease the risk of developing DSN as a result of the viral infection, some patients may develop peripheral neuropathy in response to the HAART medications themselves. The medications used to manage neuropathic pain in diabetic polyneuropathy (ie, gabapentin, amitriptyline), discussed earlier, can also be effective in treating HIV patients with neuropathic pain.

**LYME DISEASE**

Infection with *Borrelia burgdorferi*, which causes Lyme disease, can affect the peripheral nervous system and present clinically as a subacute cranial neuropathy. It can also present as a painful asymmetric radiculopathy with pleocytosis in CSF and intrathecal antibodies against *B burgdorferi*. Chronic manifestations of Lyme disease can occur years after the tick bite that transmitted the organism. Patients may present with skin manifestations and associated peripheral neuropathy characterized as a moderate, symmetric sensory polyneuropathy, occasionally with an exaggerated pain response. Diagnostic studies reveal axonal polyneuropathy on EMG and nerve biopsy. Treatment consists of an often prolonged course of antibiotics (doxycycline, amoxicillin).

**HEPATITIS C**

Several types of neuropathy are associated with hepatitis C virus (HCV) infection. These include polyneuropathy, mononeuropathy or mononeuropathy multiplex, and cranial neuropathy. It has been estimated that 10% of individuals infected with HCV will have EMG findings of axonal sensory or sensorimotor polyneuropathy. No correlation has been shown between viral load and the presence of polyneuropathy, nor is there an association with duration of infection or sex of the patient. However, the incidence of polyneuropathy increases significantly with age.

Clinically HCV polyneuropathy presents as a predominantly sensory or sensorimotor neuropathy. Diagnostic studies reveal axonal polyneuropathy and are usually associated with cryoglobulin-positive patients. Nerve biopsy reveals
fascicular axon loss, which suggests that the pathophysiology is ischemic.

Treatment is not completely effective, but some patients have improved with antiviral medication or corticosteroid treatment, or both.


**TOXIN- & MEDICATION-INDUCED NEUROPATHY**

Numerous toxins and medications have been implicated in the development of neuropathy. Although objective proof of causation may be lacking, it is important to try to identify causative agents owing to the potential reversibility of toxin-induced neuropathy. Chemotherapeutic agents, nucleoside analogs, heavy metals, and toxins have a clear association with neuropathy, although in some instances this is only a rare temporal association. It can be challenging to identify a neuropathy caused by chronic drug exposure. The pathophysiologic features will most likely be axonal; however, some agents cause demyelination and conduction block, which mimics immune-mediated neuropathy. *Table 17–2* outlines information about some of the more common medication- and toxin-induced neuropathies.

*Table 17–2* Key features of toxin- and medication-induced neuropathies.
<table>
<thead>
<tr>
<th>Agent</th>
<th>Symptoms and Signs</th>
<th>Electrodiagnostic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sensory (Sensory &gt; Motor) Axonal</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alcohol</td>
<td>Distal symmetric sensory loss; hands are affected late</td>
<td><strong>NCS:</strong> decreased SNAP and CMAP, reflecting a distal symmetric axonal sensorimotor neuropathy</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>Mild sensory stocking-and-glove distribution; toxic levels produce numbness, tingling, gait dysfunction with mild weakness</td>
<td><strong>NCS:</strong> mildly reduced SNAPs and motor conduction velocity <strong>NEE:</strong> normal</td>
</tr>
<tr>
<td>Nitrofurantoin</td>
<td>Pain with paresthesias and gait dysfunction; can progress to quadriplegia</td>
<td><strong>NCS:</strong> absent (severely decreased) sensory and motor amplitudes, normal conduction velocity <strong>NEE:</strong> neuropathic changes</td>
</tr>
<tr>
<td>Cisplatin</td>
<td>Affects dorsal root ganglion and large myelinated sensory nerves; numbness, painful paresthesias, decreased deep tendon reflexes, decreased proprioception</td>
<td><strong>NCS:</strong> reduced or absent SNAP, mildly prolonged latency <strong>NEE:</strong> normal</td>
</tr>
<tr>
<td>Lithium</td>
<td>Distal sensory loss with weakness</td>
<td><strong>NCS:</strong> absent SNAP and CMAP in lower extremity, significantly decreased SNAP and CMAP in upper extremity with normal conduction velocity</td>
</tr>
<tr>
<td>Thallium</td>
<td>Painful dysesthesias with preserved reflexes (characteristic of small fiber pathology)</td>
<td><strong>EMG:</strong> normal (secondary to small fiber loss), decreased SNAP (late in course)</td>
</tr>
<tr>
<td><strong>Motor (Motor &gt; Sensory) Axonal</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lead</td>
<td>Weakness in upper extremity preferentially affecting radial nerve, associated with atrophy of hand and foot intrinsic; all sensory spared</td>
<td><strong>NCS:</strong> mildly decreased amplitude and conduction velocity <strong>NEE:</strong> neuropathic changes with fibrillations and positive sharp waves</td>
</tr>
<tr>
<td>Organophosphates</td>
<td>Weakness (length dependent) distal followed by proximal; upper motor neuron signs (spasticity) secondary to cortical spinal tract involvement</td>
<td><strong>EMG:</strong> sensorimotor axonal neuropathy with decreased conduction velocity</td>
</tr>
<tr>
<td><strong>Motor (Motor &gt; Sensory) with Conduction Slowing</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Amiodarone</td>
<td>Symmetric distal sensorimotor neuropathy or rapidly progressive motor predominant neuropathy</td>
<td><strong>EMG:</strong> predominantly slowed conduction velocity or predominant SNAP reduction <strong>Sural nerve biopsy:</strong> axonal loss or demyelination</td>
</tr>
<tr>
<td>Arsenic</td>
<td>High dose: flaccid quadriplegia, facial weakness, respiratory failure (mimics AIDP) Chronic exposure: dermatologic symptoms first, then length-dependent painful neuropathy; few motor symptoms</td>
<td><strong>EMG:</strong> decreased amplitudes (motor &gt; sensory), prolonged F waves, partial conduction block <strong>Laboratory:</strong> urine arsenic &gt; 25 mcg/24 h; elevated levels in hair and nails in chronic cases</td>
</tr>
</tbody>
</table>

NCS, nerve conduction study; SNAP, sensory nerve action potential; CMAP, compound muscle action potential; NEE, needle electrical examination; EMG, electromyography; AIDP, acute inflammatory demyelinating polyneuropathy.

CHARCOT-MARIE-TOOTH DISEASE

ESSENTIALS OF DIAGNOSIS

- Most common inherited disorder of the peripheral nervous system
- Disease process involves a length-dependent axonal degeneration.
- Patients present clinically with a combination of lower motor neuron deficits and sensory signs.
- Characterized by progressive muscle weakness and atrophy, sensory loss, foot deformities, and steppage gait.

General Considerations

Charcot-Marie-Tooth disease (CMT), also known as hereditary motor and sensory neuropathy (HMSN), is the most common form of inherited peripheral neuropathy, with an estimated prevalence of 1 in 2500. The two main types of CMT, identified as CMT1 and CMT2, can be distinguished through electrophysiologic testing. Approximately 80% of patients have CMT1. Inheritance patterns can be autosomal dominant, autosomal recessive, or X-linked. In 90% of cases the inheritance pattern is autosomal dominant.

Pathogenesis

More than 40 causative genes expressed in Schwann cells and neurons have been associated with CMT. Simply stated, the pathophysiologic process can be attributed to a demyelinating process (CMT1), axonal loss (CMT2), or a
Clinical Findings

A. Symptoms and Signs
Patients often present in the first or second decade of life with slowly progressive distal and symmetric muscle weakness and atrophy that affects intrinsic foot and peroneal muscles. A positive family history is common. Early signs include tripping on uneven surfaces, frequent ankle sprains, difficulty heel walking due to weak ankle dorsiflexion, and tight heel cords. These symptoms often cause a steppage gait. Common phenotypical features include foot deformities such as hammer toes and pes cavus. Sensory symptoms include loss of vibration and joint position sense, and decreased pain and temperature sensation in a stocking-and-glove distribution. Muscle stretch reflexes disappear early in the ankle, and later in the patella and upper limbs. Later in the course of the disease the muscles of the hands can be affected.

B. Electrophysiologic Findings
Nerve conduction velocity is symmetrically slowed, at less than 38 m/sec (normal is > 45 m/sec) in the demyelinating type of CMT (CMT1). Patients with CMT2 have normal nerve conduction velocities but reduced amplitudes of compound muscle action potentials (CMAPs) and SNAPs secondary to axonal loss. Patients with intermediate forms of CMT show signs of both axonal loss and demyelination and decreased nerve conduction velocity in the range of 25–45 m/sec.

C. Nerve Biopsy
Sural nerve biopsies, if performed, show segmental demyelination and onion bulb formation in CMT1, and axonal loss, few or no onion bulbs, and no evidence of demyelination in CMT2.

D. Special Tests
Genetic testing can be useful to determine the inheritance pattern for family planning decisions and to obtain information about the cause and prognosis of the disease.
Treatment

There is currently no medication or intervention that reverses CMT. Physical therapy and occupational therapy should be utilized to maintain range of motion and function. Orthotic devices and assistive equipment can improve safety and function if needed. Occasionally surgical intervention for hands and feet is necessary to maintain function. Patients should be cautioned to avoid certain neurotoxic drugs, especially vincristine, as well as excessive alcohol intake. Ongoing clinical trials are investigating potential treatment options, including a therapeutic progesterone antagonist, ascorbic acid, and gene therapy.

Prognosis

CMT does not affect mortality rates. Patients usually remain ambulatory throughout their lives, but often require ankle-foot orthotics.


GUILLAIN-BARRÉ SYNDROME

Guillain-Barré syndrome (GBS) is described in detail in Chapter 19. This autoimmune polyradiculoneuropathy is characterized by progressive, symmetric weakness of the limbs. Reflexes are typically absent or greatly diminished and sensory deficits may or may not be present.

CSF evaluation reveals elevated protein levels (> 0.55 g/L) with a normal white blood cell count (< 5/mm³; < 5–10 × 10⁶ cells/L), referred to as albuminocytologic disassociation. However, CSF protein level is normal in 10% of patients. CSF protein may not rise until 1–2 weeks after the onset of weakness. MRI is sensitive but not specific in GBS diagnosis. Nerve root enhancement with gadolinium strongly suggests GBS; however, this is
commonly found in inflammatory conditions.

Electrodiagnostic testing differentiates the more common demyelinating form of GBS (acute inflammatory polyradiculoneuropathy [AIDP]) from axonal forms (acute motor axonal neuropathy [AMAN], or acute motor sensory axonal neuropathy [AMSAN]). A delay in F-wave latencies may be the earliest abnormality noted, as the nerve roots are typically affected first. Nerve conduction studies demonstrate a demyelinating pattern with greater motor involvement than sensory. These studies reveal slowing, prolongation of distal latencies, and prolongation of F-waves. Stimulation of distal and proximal sites will display temporal dispersion and a partial conduction block, findings consistent with an acute demyelinating process. These changes should be present in at least two nerves in regions not typical for the compressive mononeuropathies. Needle electrode examination demonstrates decreased motor unit recruitment. Axonal variants of GBS reveal absent or markedly reduced CMAPs and abnormal insertional activity on needle examination.


CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

ESSENTIALS OF DIAGNOSIS

- Patients classically present with sensory and motor symptoms in the distal and proximal segments of all four limbs with areflexia.
- Diagnosis is based on clinical presentation, CSF examination, and electrodiagnostic studies.
- Typical workup shows elevated protein counts in CSF and heterogeneous slowing of nerve conduction.
General Considerations

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a demyelinating neuropathy of suspected inflammatory or autoimmune origin, also considered by some to be the chronic counterpart of GBS. CIDP is considered a predominantly motor neuropathy, but in one study up to 30% of patients has a sensory variant. CIDP can be associated with hepatitis C, inflammatory bowel disease, lymphoma, HIV/AIDS, organ transplantation, and connective tissue disorder.

Patients with acquired demyelinating neuropathies (CIDP, AIDP), in contrast to hereditary neuropathies, often present with late onset, rapid progression, focal onset or advancement, preceding infection or immunization, or both, and do not have a family history of similar findings.

Clinical Findings

A. Symptoms and Signs

CIDP has a clinically heterogeneous presentation. It is a grossly symmetric motor and sensory diffuse polyneuropathy with generalized areflexia that develops over 8 weeks (in contrast to GBS, which develops over 4 weeks). CIDP affects the distal and proximal segments of all four limbs, and it can be a monophasic, relapsing, or progressive disorder.

B. Laboratory Findings

CSF analysis usually shows a cytoalbuminologic dissociation with elevated protein and minimal or absent pleocytosis. An elevated white blood cell count in CSF is more indicative of infection (eg, HIV, Lyme disease), inflammation (eg, sarcoidosis), or malignancy (eg, lymphoma). The presence of monoclonal gammopathy is suggestive of hematologic disease.

C. Electrophysiologic Findings

Electrodiagnostic findings of demyelination include prolonged distal and peak motor and sensory latencies, prolonged F-wave latencies, temporal dispersion, and conduction block. The presence of nonuniform slowing of nerve conduction, temporal dispersion, and conduction block, in contrast to uniform findings, is more common in acquired (CIDP, AIDP) versus inherited demyelinating neuropathies.
D. Nerve Biopsy

Nerve biopsies may be useful in select patients in whom the diagnosis of inherited versus acquired demyelinating neuropathy is unclear, but because CIDP often affects proximal motor nerves, a routine sural nerve biopsy may fail to show diagnostic pathologic changes.

E. Imaging Studies

MRI with contrast can reveal enhancement at the root level secondary to the breakdown of the blood–nerve barrier. Root hypertrophy can also be seen at the cervical and lumbar level, which may account for clinical symptoms consistent with cervical or lumbar stenosis.

Treatment

Standard treatment includes corticosteroids (commonly oral prednisone), intravenous immunoglobulin (IVIG), and possibly plasmapheresis. IVIG has been shown to be slightly more effective than steroids. Evidence shows up to 20% of patients do not need treatment, but this requires good clinical evaluation and regular monitoring of disease progression. Long-term steroid use should be accompanied by vitamin D supplementation, weight-bearing exercise, and possibly bisphosphonates. Targeted immunotherapies, including immunosuppressives and cytotoxic drugs, are required for refractory cases or when steroids and IVIG therapy are contraindicated. Rare cases of acute-onset chronic inflammatory demyelinating polyradiculoneuropathy (A-CIDP) require continuous immunotherapy. Physical therapy with emphasis on balance and proprioception, assistive devices, including ankle-foot orthoses, and home safety evaluation can be very beneficial.

Prognosis

Current treatment with corticosteroids, IVIG, and plasma-pheresis is effective in 80% of patients. Long-term maintenance treatment is often required, as many patients will relapse during dose tapering. Despite extensive treatment, 15–20% of patients are considered nonresponders and will develop a progressive decline in function.


FOCAL NEUROPATHIES

ESSENTIALS OF DIAGNOSIS

► Focal neuropathies usually represent a focal area of demyelination.
► Nerve conduction studies reveal a focal conduction block with segmental slowing and amplitude drop.
► Prognosis is usually favorable but more severe axonotmesis can be seen.

BRACHIAL PLEXOPATHY

► General Considerations

The brachial plexus runs through the axilla and consists of the anterior rami of roots C5 through T1. The brachial plexus is frequently injured as the result of trauma—from gunshot wounds, knife wounds, a fall on an outstretched arm, or fractures. Brachial plexopathy is also associated with traction during birth, shoulder dislocation, positioning during surgery, radiation, primary nerve tumors, and metastatic disease. Occasionally, injury to the plexus is idiopathic.
Clinical Findings

A. Symptoms and Signs

Specific types of brachial plexus injury can be differentiated based on the history and physical examination findings (Table 17–3). In the upper trunk, Erb-Duchenne palsy affects the shoulder and upper arm muscles while sparing the hand. A “stinger,” seen in sports injuries, is a transient stretch of the brachial plexus that usually resolves spontaneously after a short duration. A Pancoast tumor of the apex of the lung affects the lower trunk (C8–T1 roots) of the plexus, as does a Klumpke’s palsy. The posterior cord of the plexus can be injured with a shoulder dislocation, causing weakness of axillary and radial innervated muscles.

Table 17–3 Key features of brachial plexopathies.

<table>
<thead>
<tr>
<th>Plexopathy</th>
<th>Etiology</th>
<th>Symptoms and Signs</th>
<th>Electrodiagnostic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic outlet syndrome</td>
<td>Cervical rib or scalene pathology</td>
<td>Pain, weakness, paresthesias of hand</td>
<td>Decreased ulnar SNAP and median CMAP amplitude</td>
</tr>
<tr>
<td>Erb’s palsy</td>
<td>Plexus traction</td>
<td>Shoulder weakness</td>
<td>Slowing through the plexus, denervation of shoulder muscles</td>
</tr>
<tr>
<td>Lower trunk injury (Klumpke, Pancoast)</td>
<td>Tumor compression (Pancoast), plexus traction (Klumpke)</td>
<td>Hand weakness</td>
<td>Slowing through the plexus, denervation of hand muscles</td>
</tr>
<tr>
<td>Parsonage Turner syndrome</td>
<td>Possible immune cause</td>
<td>Shoulder girdle weakness</td>
<td>Slowing through the plexus</td>
</tr>
</tbody>
</table>

EMG, electromyography; NCS, nerve conduction study; SNAP, sensory nerve action potential; CMAP, compound muscle action potential.

Patients with idiopathic brachial neuropathy (IBN), or Parsonage-Turner syndrome, present with C5–C6 dermatomal deficits and weak shoulder girdle musculature. This syndrome occurs sporadically after the third decade of life and is usually unilateral. It is associated with pregnancy, vaccination, trauma, and infection. An immune pathology has been suggested, but the exact etiology remains unknown. Presenting symptoms include pain in the C5–C6 dermatome and weakness in the C5–C6 myotome, primarily the shoulder girdle musculature.

Thoracic outlet syndrome is a brachial plexopathy caused by a cervical rib or
scalene compression. The compression may cause vascular or neurologic compromise, or occasionally both. Neurologic symptoms include pain and paresthesias of the hand and medial forearm with weakness of the hand intrinsics.

**B. Electrophysiologic Findings**

Electrodiagnostic evaluation of a brachial plexopathy usually reveals reduced sensory and motor amplitude proximal to the lesion with stimulation at Erb’s point (supraclavicular), in conjunction with nerve conduction velocity slowing across the lesion. Needle EMG may show denervation of the affected distribution of muscles, with long-duration polyphasic motor unit potentials and reduced recruitment. Median and ulnar F-wave latency may be prolonged.

EMG findings in IBN resemble those for other brachial plexus injuries, with slowing through the plexus and denervation of the muscles involved, although mild cases may be purely demyelinating. In thoracic outlet syndrome, EMG with nerve conduction studies typically reveals a reduced ulnar SNAP amplitude and reduced median CMAP amplitude, prolonged ulnar F wave, and denervation of the hand intrinsic muscles.

**Treatment**

There is no definitive treatment for brachial plexus lesions, and an individualized plan must be developed. This may include therapy for range of motion and strengthening, orthoses, and surgical intervention with nerve or muscle transfer, or both. Patients with thoracic outlet syndrome may require cervical rib resection. Those with IBN usually improve gradually over a period of months and have a favorable prognosis.


**AXILLARY NERVE MONONEUROPATHY**

**General Considerations**

The axillary nerve is the terminal branch arising from the posterior cord of the brachial plexus, which receives input from C5 and C6. It primarily innervates the
deltoid and then branches to innervate the teres minor and become the lateral sensory cutaneous nerve. The two most common points of entrapment are at the glenoid rim and the quadrilateral space, where the posterior circumflex humeral artery may also be occluded, causing quadrilateral space syndrome. Nerve entrapment can be a result of compression from muscular contraction through the axilla in sports such as rowing or the improper use of crutches. Axillary neuropathy is prevalent in 45% of humeral dislocations.

**Clinical Findings**

Patients may complain of numbness or isolated sensory deficits over the deltoid belly in the upper arm. Shoulder flattening sometimes occurs as a result of deltoid atrophy and limited abduction after the first 30 degrees in the range of motion.

EMG can be utilized to detect abnormalities of the deltoid and teres minor. Nerve stimulation of Erb’s point with pickup over the bulk of the deltoid muscle may reveal a reduced amplitude or slowed conduction velocity. Needle EMG may show spontaneous activity and reduced recruitment in the deltoid or teres minor. MRI can assess the quadrilateral space, and a subclavian arteriogram can be used to confirm artery occlusion.

**Treatment**

Treatment depends on the etiology of the insult, but patients with stretching injuries tend to have a worse prognosis. Initially, conservative therapy, including gentle stretching through internal rotation, should be tried. If no clinical or EMG progress is seen within 2–3 months postinjury, a decision to try surgical repair should be made as the literature shows optimal success when this is accomplished between 3 and 6 months postinjury.


**SUPRASCAPULAR NERVE MONONEUROPATHY**
General Considerations

The suprascapular nerve branches off of the upper trunk of the brachial plexus and receives input from the C5 and C6 nerve roots. It supplies motor innervation to the supraspinatus and infraspinatus muscles, as well as sensory innervation to the deep muscle and capsule of the glenohumeral joint. Most commonly this nerve becomes entrapped by the transverse scapular ligament in the suprascapular notch. However, in 50% of individuals the infraspinatus branch travels within a tunnel roofed by the spinoglenoid ligament and can become entrapped there. Suprascapular entrapment may occur from repetitive overhead activities (volleyball, tennis, pitching) or forcible compression (gymnastics, football) and is seen in 28% of full-thickness rotator cuff tears.

Clinical Findings

Patients primarily complain of shoulder pain, which is throbbing in nature and may run along the superior border of the scapula with extension into the shoulder. Pain usually does not radiate and may increase with motion. Injury at the supraspinatus notch can lead to atrophy of the supraspinatus and infraspinatus muscles after a few months, resulting in weakness of abduction and external rotation.

Physical examination of the shoulder joint can elicit pain with abduction and external rotation. EMG may show prolonged latency of the supraspinatus and infraspinatus, as well as selective denervation of these areas with sparing of other muscles supplied by C5 and C6. Relief from an anesthetic injection into the suprascapular notch can confirm the diagnosis more definitively.

Treatment

For patients whose injuries have occurred secondary to athletics, avoiding repetitive trauma or movement can help relieve symptoms. Early exploration is essential in complicated fractures. If surgery is necessary, the entrapped nerve is released by cutting the transverse scapular ligament, which is compressing the suprascapular nerve.

MUSCULOCUTANEOUS NERVE ENTRAPMENT
General Considerations

The musculocutaneous nerve branches off of the lateral cord of the brachial plexus and receives input from C5, C6, and C7. It pierces and supplies the coracobrachialis, biceps, and brachialis muscles, and then descends along the lateral aspect of the forearm, where it becomes the lateral cutaneous nerve. Entrapment of this nerve is rare but may result from compression by the coracobrachialis secondary to strenuous exercise or trauma (eg, proximal humeral fracture or shoulder dislocation).

Clinical Findings

The biceps is most often affected by damage to this nerve. Patients often have weakness with elbow flexion as well as an absent stretch reflex of the biceps. Pain is usually located in the proximal elbow and forearm; hypesthesia may continue down the radial aspect of the forearm. These symptoms tend to be exacerbated by elbow extension.

The diagnosis of musculocutaneous nerve entrapment is made primarily on the basis of physical examination and EMG findings. Decreased amplitude and increased latency of the musculocutaneous nerve can confirm diagnosis. Needle EMG findings may be present in the biceps brachii, brachialis, and coracobrachialis muscles.

Treatment

Methods of treatment for this entrapment have not been fully explored owing to the paucity of cases studied. If a trial of conservative therapy does not produce improvement, patients may be referred for surgery.

LONG THORACIC NERVE MONONEUROPATHY

General Considerations

Entrapment of the long thoracic nerve is extremely rare. The nerve originates from separate branches of C5, C6, and C7 before descending on the lateral aspect of the rib cage and innervating the serratus anterior muscle. Impairment of nerve function may result from trauma, traction during sports activities such
as volleyball and archery, or intraoperatively during radical mastectomy.

Clinical Findings

Patients report pain in the shoulder that is exacerbated by tilting the head away from the affected side. Patients may also note an inability to flex at the shoulder while keeping the arm straight. Scapular winging with medial displacement of the superior angle of the scapula is also present in these cases and may be more noticeable when the patient is sitting straight in a high-backed chair.

Diagnosis is primarily based on physical examination findings that demonstrate the patient’s inability to raise the arm straight up, as well as scapular winging. Confirmation can be obtained by EMG showing isolated spontaneous activity, motor unit changes, or reduced recruitment of the serratus anterior. Erb’s point stimulation with pickup over the serratus may reveal delays.

Treatment

There is a high rate of spontaneous recovery in these types of injuries, so conservative therapy is the mainstay of treatment. Surgery is indicated for patients whose symptoms persist over 1–2 years and those who show lack of improvement as assessed by EMG.


MEDIAN NERVE MONONEUROPATHY
1. Carpal Tunnel Syndrome

General Considerations

Carpal tunnel syndrome (CTS) is the most common entrapment neuropathy, carrying a 10% lifetime risk in the general population. The median nerve as well as nine flexor tendons course through this anatomic tunnel at the junction of the wrist. Bony carpal structures surround the tunnel on the dorsal and lateral sides, while the thick transverse ligament stretches across the volar surface. Approximately 2–2.5 cm distal to the entrance, the tunnel narrows, and it is at this point that many patients develop compression of the median nerve causing slowed median nerve conduction and associated symptoms.

The most common CTS patient is a middle-aged woman, reflecting increased prevalence of CTS among women of 5.8% compared with 0.6% in men. Other risk factors for development of CTS include increasing age (50–60 years); obesity; pregnancy; chronic use of a single-point cane, crutches, or wheelchair; and occupational hazard. Certain disease processes can also predispose individuals to CTS, including acromegaly, rheumatoid arthritis, hypothyroidism, lupus erythematosus, amyloidosis, hyperparathyroidism, Lyme disease, and conditions that cause prolonged wrist or finger hyperflexion. Lastly, trauma to the forearm or wrist can cause symptoms of CTS, especially in Colles’ fracture, hamate fracture, or a crushing injury.

Clinical Findings

A. Symptoms and Signs

The most prominent symptoms of CTS are numbness or tingling of the hand in the median nerve distribution, with the index and middle digits being most commonly affected. Often, symptoms spare the skin of the thenar eminence as this part of the hand is innervated by the palmar cutaneous branch of the median nerve, which branches off 3 cm proximal to the carpal tunnel. However, patients may present atypically with symptoms involving the entire hand or only areas within the ulnar nerve distribution and still be found to have CTS. In up to 36.5% of patients, pain and numbness can travel proximal to the wrist, up to the elbow, or even to the shoulder.

Symptoms are exacerbated by movement or positions that require wrist flexion. Patients report dropping objects and worsening symptoms while driving, holding the phone, typing, and often while sleeping. Shaking the hands relieves
the symptoms for a short period of time in patients with mild disease.

**B. Electrophysiologic Findings**

There is no gold standard test for diagnosing CTS; however, combined findings from physical examination and EMG have proven to be the most effective in narrowing the diagnosis. Key findings of muscle atrophy and decreased strength on physical examination may point toward median nerve pathology. A careful sensory examination can help to distinguish single versus multiple nerve involvement. Thenar atrophy has been found to increase the predictive value of CTS, but this finding is rare. The Phalen and Tinel tests are commonly used to elicit symptoms but are not diagnostic without EMG correlation.

Several EMG findings help to differentiate CTS from other pathology. Prolonged SNAP or CMAP distal latency with wrist stimulation and segmental slowing through a midpalm–wrist portion of the median nerve are typically found. A median–radial SNAP comparison that reveals a median delay is also relevant. For a definitive diagnosis, these findings must be correlated with findings from the physical examination. The Combined Sensory Index (CSI) is the sum of three parameters tested and has been shown to have high sensitivity and specificity for CTS (*Table 17–4*).

---

**Table 17–4** Combined Sensory Index (CSI).

<table>
<thead>
<tr>
<th>CSI (≤ 0.9 ms) = Median–ulnar antidromic SNAP latency difference at 14 cm +</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median–radial thumb antidromic SNAP latency difference at 10 cm +</td>
</tr>
<tr>
<td>Median–ulnar midpalmar orthodromic latency difference at 8 cm</td>
</tr>
</tbody>
</table>

SNAP, sensory nerve action potential.

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**Treatment**

Conservative and surgical options are available for patients with CTS. Conservative options include oral corticosteroids or steroid injections, wrist
splinting, and activity modification. It is recommended that patients try conservative therapy before surgery; however, after 2–7 weeks of no improvement, patients should try an alternative conservative option or be taken to surgery. Carpal tunnel release involves splitting the flexor retinaculum to relieve pressure put on the median nerve. Studies have shown that surgery has better long-term outcomes than conservative therapy, but a conservative trial is undergone in an attempt to avoid risks that surgery presents.

2. Anterior Interosseus Syndrome

General Considerations

The anterior interosseus nerve branches from the median nerve about 5–8 cm below the elbow. It innervates the flexor pollicis longus, flexor digitorum profundus (I and II), and pronator quadrates and is not responsible for any dermatomal innervation. Entrapment of this nerve is rare, and most cases are due to viral neuritis. The deep position of the nerve protects it from most trauma; however, damage has resulted from puncture wounds in the forearm, including complications of venipuncture. Other reported causes include poorly constructed casting, sleeping on the arm, and excessive exercise.

Clinical Findings

A. Symptoms and Signs

Patients with anterior interosseus syndrome demonstrate weakness while flexing the distal phalanx of the thumb as well as the second and third digits. Often, patients report pain in the proximal forearm that may last hours or days and is followed by hand weakness, most commonly in the thumb and index finger. Pain is exacerbated by exercise and relieved with rest. They may also report deterioration of their handwriting. The pronator quadrates is affected in some cases, although objective pronator weakness is rare because the pronator teres is unaffected. History and physical examination data are important in this diagnosis. The pinch sign is used to look for weakness of the flexor pollicis longus and flexor digitorum profundus (Figure 17–1). Weakness of the pronator quadrates can be elicited by flexing the elbow to remove influence of the pronator teres and then asking the patient to pronate the forearm.
Figure 17–1 Inability to make the “OK” sign is a characteristic finding in anterior interosseus syndrome. Damage to the anterior interosseus nerve is indicated by the pinch sign—the patient’s inability to flex the thumb and DIP of the index finger.

B. Electrophysiologic Findings
EMG is used to confirm diagnosis. Spontaneous activity may be seen in the flexor digitorum profundus, pronator quadrates, or flexor pollicis longus.

C. Diagnostic Imaging
MRI is sometimes used as adjunctive imaging and is most helpful in the acute phase because of the increased extracellular water uptake.

Treatment
Conservative therapy may include arm rest, immobilization, corticosteroid injections, or use of nonsteroidal antiinflammatory drugs (NSAIDs), and is the preferred approach. Symptoms resolve with such treatment in most patients within 6 months to 1 year of initial presentation, although complete recovery is more common in the flexor digitorum profundus than the flexor pollicis longus. Release surgery may be necessary in the event of trauma or known compression
of the anterior interosseus nerve. Some patients also undergo surgery if their symptoms have not improved after 6 months of conservative therapy; however, this time frame varies and is primarily dependent on patient and surgeon choice.

3. Pronator Syndrome

General Considerations

The median nerve can become entrapped at the elbow in four different locations: by a thickened lacertus fibrosis, the arch of the flexor digitorum superficialis, the pronator teres, or the ligament of Struthers (discussed below). In 80% of individuals the median nerve dives between the superficial and deep heads of the pronator teres, and this is the most common site of entrapment. Risk factors for entrapment include activities involving repetitive, forceful pronation and supination (eg, rowing, pitching, racquet sports, and grip-intensive sports or employment). Nerve damage can also result from trauma at the elbow, muscle hypertrophy, or a persistent median artery.

Clinical Findings

The parameters for diagnosis of pronator syndrome are ill defined and depend primarily on the history and physical examination. The most common physical finding is tenderness to palpation over the pronator teres. Muscle weakness and sensory findings vary depending on the extent of nerve involvement. Detection of numbness over the thenar eminence on sensory examination is an important finding that helps to avoid misdiagnosis of CTS. To confirm the diagnosis, some physicians perform steroid injections into the pronator teres to observe for symptom relief.

A. Symptoms and Signs

The presentation of pronator syndrome is much like that of CTS, with symptoms often occurring in the dominant hand. Patients report weakness in the thumb, numbness and tingling throughout the hand, and pain that increases with activity. Pronation is intact in most patients, although they may complain of pain directly over the pronator teres. Of importance, numbness is usually not felt in the thenar eminence and symptoms are absent at night.
B. Electrophysiologic Findings

EMG detects abnormalities in only a small percentage of affected patients and is mainly used to rule out other neuropathies, including CTS.

Treatment

As with anterior interosseus syndrome, the mainstay of treatment for patients with pronator syndrome is nonoperative. Treatment includes activity modification, rest, immobilization, NSAIDs, oral corticosteroids, and steroid injections directly into the pronator teres. If after 3 months of such treatment the patient has not improved, release surgery is pursued to prevent intraneural fibrosis, which can lead to irreversible nerve damage. Surgical treatment can be challenging because the site of nerve compression is not always apparent; however, up to 90% of patients who undergo release procedures experience relief of symptoms after decompression.

4. Median Nerve Entrapment at the Ligament of Struthers

In 1848, John Struthers described a ligament that connected the medial epicondyle to a rarely seen supracondylar process. The process is quite rare and the presence of the ligament is even more unique. In a small number of patients with these anomalies, the median nerve and brachial artery run underneath the ligament and can become entrapped. Symptoms include elbow pain, weakness in pronation, and tenderness over the ligament. Treatment is primarily surgical and involves the removal of the bony process and release of the ligament.


ULNAR NERVE MONONEUROPATHY

1. Cubital Tunnel Syndrome

General Considerations

Cubital tunnel syndrome is the second most common neuropathy of the upper extremity. The cubital tunnel, located just distal to the medial epicondyle, is composed of the flexor carpi ulnaris aponeurosis, medial collateral ligament, joint capsule, and olecranon. The tunnel is narrowest during elbow flexion due to compression by the aponeurosis, but the medial collateral ligament also contributes to the increased pressure by bulging medially.

Clinical Findings

A. Symptoms and Signs

Patients most commonly present with numbness and tingling in the ulnar distribution, although they may have hand weakness or problems with grip. Pain in the ulnar side of the forearm may occur and is often worse at night. Symptoms are exacerbated by resting the elbow on a hard surface or repetitive use of the elbow and forearm. Finger adduction is weakened, and patients may report that the fifth digit “gets caught” when putting their hands in their pocket (Wartenberg’s sign). Other difficulties include opening bottles and buttoning buttons, and patients may state that they “feel clumsy.”

B. Diagnostic Studies

Provocative tests include the Tinel test, along the course of the ulnar nerve, and the elbow flexion test. In the flexion test, the patient is asked to hold the arm in complete elbow flexion and wrist extension for 3 minutes. Onset of increasing pain, numbness, or tingling indicates a positive result. Electrodiagnostic studies
show weakness in the flexor digitorum profundus and sparing of the flexor carpi ulnaris, because the nerve fibers travel to the latter branch before the aponeurosis. The exact site of compression can be identified by stimulating the nerve at multiple sites along the cubital tunnel. A nonlinear decline in velocity or amplitude confirms the lesion location. Recently, investigators have looked at effectiveness of ultrasound in diagnosing cubital tunnel syndrome. One study found that the ulnar nerve diameter was significantly smaller in patients with a confirmed lesion than in patients with normal elbows. As in other compressive syndromes, clinical and electrophysiologic findings together can confirm the diagnosis.

**Treatment**

There is no gold standard for treatment of this syndrome; rather, the duration and intensity of symptoms guide management. Between 65% and 89% of patients with mild to moderate symptoms show improvement with conservative therapy (activity modification, splinting, physical therapy). Many surgical options exist if conservative therapy fails or there is significant motor weakness, In situ decompression and subcutaneous anterior transposition are the two most successful options available; both decrease the tension placed on the ulnar nerve itself. Other options include intramuscular transposition, submuscular transposition, medial epicondylectomy, and endoscopic decompression. Factors influencing procedure choice include surgeon’s experience, location of the lesion, and anatomic variation.

2. **Ulnar Nerve Entrapment at Guyon’s Canal**

**General Considerations**

Entrapment of the ulnar nerve at the wrist is rare compared with entrapment at the elbow; however, when it does occur, Guyon’s canal is the most common location. The canal is made up of tendons, ligaments, and bones and is approximately 4 cm long. Common causes of symptoms include ganglions, fractures, anomalous muscle bellies, and repetitive trauma. Cyclists, metal cutters, and mechanics are at risk, because repetitive wrist dorsiflexion causes trauma to the ulnar artery which travels through the canal with the nerve.
Clinical Findings

A. Symptoms and Signs
Depending on the location of the lesion, patients can present with purely motor, purely sensory, or mixed symptoms. Patients may report numbness or tingling in their ring and small fingers, impairments in two-point discrimination, and grip weakness. Loss of the interossei muscles with preservation of hypothenar function and sensation occurs if the lesion is at or distal to the hook of hamate. Physical examination may reveal Wartenberg’s sign (due to unopposed abductor digiti quinti) or palmaris brevis sign. Tenderness over the hook of hamate indicates possibility of fracture, and a timed Allen’s test may reveal infarction as the cause of symptoms.

B. Diagnostic Studies
Depending on the history and physical examination findings, imaging (radiographs or MRI) may be necessary to confirm the etiology (fracture, tumor, or ganglion). Nerve conduction findings on electrodiagnostic study can differ depending on lesion location. Confirmatory findings include a reduced or absent SNAP with slowing through the wrist and a decreased CMAP from the abductor digiti quinti and the first dorsal interosseous.

Treatment
In cases of repetitive trauma with no evidence of fracture or mass, a trial of conservative therapy, including NSAIDs, splinting, and discontinuation of provocative activities, is first attempted. Surgery is indicated if conservative treatment is unsuccessful or there is evidence of a mass. The nerve must be exposed from the forearm to the hand distal to nerve bifurcation to ensure that all the compressive forces are removed. For fractures at the hook of hamate, the bone must be excised before decompression and neurolysis.


Murata K, Shih JT, Tsai TIM: Causes of ulnar tunnel syndrome: A
RADIAL NERVE MONONEUROPATHY

1. Lesions of the Superficial Radial Nerve

General Considerations

Lesions of the superficial radial nerve are extremely rare. They are also referred to as cheiralgia paresthetica or Wartenberg’s syndrome, as he described them in detail in 1932. The superficial radial nerve branches from the radial nerve at the lateral forearm, becoming superficial approximately 9 cm proximal to the radial styloid. Compression at the wrist by watches or handcuffs, as well as iatrogenic injury during repair of de Quervain’s tenosynovitis, are common causes.

Clinical Findings

A. Symptoms and Signs

Patients present with primarily sensory symptoms that include pain or dysesthesias radiating to the thumb and index finger. Patients may experience symptoms in all of the radial nerve distribution or only a few digits. This variation is due to innervation overlap with the lateral antebrachial cutaneous nerve.

B. Diagnostic Studies

Clinical examination findings include thickening of the radial nerve at the site of injury and a positive Tinel’s sign. EMG studies may reveal decreased amplitude of the SNAP at the dorsum of the hand and diminished conduction velocity when compared with the contralateral radial SNAP or the ipsilateral lateral antebrachial cutaneous nerve. Some clinicians perform lidocaine nerve blocks to distinguish between superficial radial nerve and lateral cutaneous nerve involvement, but this is not standard practice.
Treatment

Spontaneous resolution of symptoms is common; therefore, conservative management is the mainstay of treatment if symptoms have persisted for less than 6 months and the offending agent can be removed. Nerve exploration and possible neurolysis are indicated in the following instances: the lesion is iatrogenic or post-traumatic, de Quervain’s disease is also present, or symptoms have persisted for longer than 6 months. Currently there are no good data comparing results for differing conservative regimens versus surgery at time of diagnosis.

2. Radial Nerve Entrapment at the Elbow

General Considerations

Radial nerve lesions are the least common of the nontraumatic entrapment syndromes. The radial nerve is easily entrapped around the elbow and proximal forearm, causing two different syndromes: posterior interosseus nerve syndrome (PINS) and radial tunnel syndrome (RTS). PINS is caused by entrapment of the posterior interosseus branch of the radial nerve by the arcade of Frohse, which is a fascial plane that connects the two heads of the supinator. RTS occurs when the posterior interosseus nerve becomes entrapped within a tunnel in the proximal forearm past the proximal joint of the supinator. Causes of radial nerve entrapment at the elbow and proximal forearm most commonly include lipomas, trauma, and activities involving repetitive pronation, and up to 60% are found to be spontaneous.

Clinical Findings

A. Symptoms and Signs

Patients may describe onset of pain in the lateral forearm, which may have lasted only 1–3 days. In PINS, patients tend to have a pure motor syndrome affecting the wrist and finger extensors, although the paralysis may be limited to only a few fingers. RTS presents as a primarily sensory syndrome with pain in the lateral epicondyle that radiates down to the back of the hand. No weakness is found in supination because in both syndromes the entrapment occurs after the radial nerve has innervated the supinator. However, motor and sensory
symptoms can vary in both syndromes, making it even more difficult to make a clear distinction.

**B. Diagnostic Studies**

The diagnosis of entrapment of the radial nerve at the elbow is mainly clinical. Provocative testing for radial nerve entrapment includes pain elicited with resisted middle finger extension (middle finger test) and worsening symptoms when the forearm is passively pronated with a flexed wrist. EMG may be used to identify slowed conduction across the site of entrapment, especially when the arm is supinated against resistance, or to see denervation of all muscles innervated by the radial nerve. Computed tomography can identify lipomas or ganglions that may be causing symptoms, and patient relief after anesthetic injection to the radial tunnel can confirm RTS.

**Treatment**

Conservative treatment for 6–8 weeks is recommended for most patients, especially if there is no evidence of a mass causing the compression. Treatment includes rest, use of NSAIDs, avoidance of activities that exacerbate symptoms, and splinting to maintain supination and wrist extension. If the symptoms do not improve or worsen to incomplete or complete palsy, surgical decompression should be considered. Decompression is successful in 60–70% of cases, enabling individuals to return to light activities in 10 days and back to work within 6 weeks.

**3. Radial Nerve Entrapment at the Shoulder**

**General Considerations**

Most radial nerve lesions at the shoulder are secondary to trauma. The close proximity of the radial nerve to the humerus and its decreased mobility as it courses through the lateral intermuscular septum make it susceptible to being lacerated or entrapped by fracture segments. Consequently, it is the most common nerve lesion complicating long bone fractures. Up to 10–15% of humeral fractures are associated with radial nerve palsy, especially if classified as a Holstein Lewis fracture.
Clinical Findings

A. Symptoms and Signs

Patients often present with acute symptoms after trauma, sleeping on an extended shoulder overnight (so-called Saturday night palsy), or after several weeks of using crutches. Wrist extension is almost always affected, resulting in a complete wrist drop with weakened extension of the metacarpophalangeal joints as well. Numbness and tingling may be reported in the radial distribution (dorsum of the hand and first two digits). Patients may experience some weakness in elbow flexion and supination. Because these functions are carried out by muscles with different innervation, marked deficits are not always reported.

B. Diagnostic Studies

The context of injury is helpful to guide physical examination and further testing. EMG can result in slowed conduction through the site of injury with stimulation proximally over Erb’s point. Axonotmesis can result in diminished amplitude or complete lack of motor conduction and sensory action potentials, whereas focal demyelination reveals focal slowing through the site of injury with spared conduction distally. Needle examination may reveal the extent of damage to the nerve based on ability to recruit motor units and presence of spontaneous activity in the muscles sampled. Ultrasound has been investigated as a tool to help guide treatment decisions in humeral fracture, but it is not yet widely used in practice.

Treatment

Conservative management is widely accepted as the first step, with up to 88% of patients spontaneously recovering unless the lesion is secondary to an open fracture. Conservative measures include NSAIDs, rest, avoiding provocative activities, and splinting, sometimes with a spring-loaded extensor brace for the fingers if paralysis is longstanding. Early versus late exploration of closed fractures remains controversial, but open fractures require exploration immediately or at delayed closure in order to preserve as much nerve function as possible.


LATERAL FEMORAL CUTANEOUS MONONEUROPATHY

**General Considerations**

Neuropathy of the lateral femoral nerve, also known as meralgia paresthetica, is caused by entrapment of the lateral femoral cutaneous nerve (LFCN) as it travels under the inguinal ligament or as it pierces the fascia lata. The nerve is formed by contributions of the L2–L3 nerve roots. It travels laterally under the psoas muscles and across the iliacus muscle. The nerve exits the pelvis under the inguinal ligament just medial to the anterior superior iliac spine and then pierces the fascia lata. Possible causes of compression of the LFCN are listed in Table 17–5.

Table 17–5 Causes of lateral femoral nerve compression.
Clinical Findings

A. Symptoms and Signs
Clinical manifestations include burning pain, numbness, and tingling located at the lateral aspect of the thigh. The symptoms are aggravated by local pressure over the anterior superior iliac spine and relieved by hip flexion.

B. Diagnostic Studies
Nerve conduction studies reveal a prolonged latency for the LFCN with stimulation proximal to the compression. The amplitude is likely to be reduced with slowed nerve conduction. Needle EMG is normal. MRI findings include alteration in size and signal of the entrapped nerve.

Treatment
Pressure on the LFCN can relieved by avoiding heavy belts or tight clothing that may be irritating or compressing the nerve in the region of the inguinal ligament. Injection of lidocaine 1% is sometimes effective in reducing symptoms. Refractory cases may require surgical decompression.
The peroneal nerve arises primarily from the L5 nerve root, with additional input from S1 and S2 and, minimally, from L4. It travels as part of the sciatic nerve through the thigh and separates at the upper level of the popliteal space as the common peroneal nerve. In the distal thigh the peroneal nerve gives off one motor branch to the short head of the bicep femoris muscle. It continues distal to the head of the fibula and wraps around the neck. It is exposed to the bony prominence of the fibula. At the fibula neck the nerve passes through a tunnel, and then divides into its final two branches.

Peroneal nerve entrapment is the most common nerve entrapment in the lower limbs, with entrapment at the fibula head being the most common variant. Possible causes are listed in Table 17–6.

### Table 17–6 Causes of peroneal nerve entrapment.

<table>
<thead>
<tr>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic heel sitting, such as in yoga</td>
</tr>
<tr>
<td>Kneeling</td>
</tr>
<tr>
<td>Crossed legs while sitting</td>
</tr>
<tr>
<td>Wearing of knee pads as part of work</td>
</tr>
<tr>
<td>Surgery</td>
</tr>
<tr>
<td>Ganglion cyst</td>
</tr>
</tbody>
</table>

#### Clinical Findings

Clinical manifestations include pain, palpable mass, and foot drop. Motor function is affected to a greater degree than sensory. Imaging such as radiography and MRI can help in diagnosis but the most definitive study is EMG. Proximal stimulation may reveal reduced amplitude and nerve conduction slowing of the CMAP, and the SNAP amplitude may be reduced distally. Needle EMG may reveal findings such as denervation, reduced recruitment, and changes to motor unit potentials in the deep peroneal and superficial peroneal distribution.
Treatment

Treatment recommendations include behavior modification, massage, physiotherapy, and in some refractory cases, surgery to decompress the nerve.

DEEP PERONEAL NERVE ENTRAPMENT

General Considerations

Deep peroneal nerve compression at the ankle is also known as anterior tarsal tunnel syndrome. The points of possible compression are deep to the superior and inferior extensor retinacula or at the level of the talonavicular joint as it travels deep to the extensor hallucis longus tendon. Distally the deep peroneal nerve may also be entrapped at the level of the first and second tarsometatarsal joints as it travels in a tight tunnel beneath the extensor hallucis brevis muscle. Possible causes of entrapment are listed in Table 17–7.

Table 17–7 Causes of deep peroneal nerve entrapment.

<table>
<thead>
<tr>
<th>Causes of deep peroneal nerve entrapment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stretching of the nerve secondary to ankle instability</td>
</tr>
<tr>
<td>Direct trauma to the dorsum of the foot</td>
</tr>
<tr>
<td>Hypertrophic extensor hallucis brevis muscle</td>
</tr>
<tr>
<td>Os intermetatarsum in the proximal first metatarsal space</td>
</tr>
<tr>
<td>Dorsal degenerative spurs at the talonavicular joint</td>
</tr>
<tr>
<td>Tight-fitting shoes</td>
</tr>
</tbody>
</table>

Clinical Findings

A. Symptoms and Signs

Clinical manifestations include dysesthesias along the dorsomedial aspect of the foot and weakness of the extensor digitorum brevis muscle. On examination it is important to inspect the foot for masses or other space-occupying lesions. The
extensor digitorum brevis may be atrophied, and sensory examination may reveal numbness and paresthesias in the first web space. Percussion of the deep peroneal nerve and a reproduction of symptoms (Tinel’s sign) may help in diagnosis.

**B. Diagnostic Studies**

MRI findings may include atrophy and edema of the anterior compartment muscles. EMG study reveals prolonged latency and nerve conduction slowing with stimulation proximal to the lesion. Needle EMG may reveal denervation and motor unit potential changes in the extensor digitorum brevis.

**Treatment**

In treating patients, conservative measures such as orthoses and accommodative shoes should be tried first. If these measures are unsuccessful, other options such as corticosteroid injections or surgical release may be required.

**SUPERFICIAL PERONEAL NERVE ENTRAPMENT**

**General Considerations**

Superficial peroneal neuropathy is caused by entrapment of the superficial peroneal nerve as it exits through the deep fascia of the lateral leg. The superficial peroneal nerve descends the leg within a fascial plane between the peroneus longus and extensor digitorum longus muscles. The nerve exits through the deep fascia of the lateral leg compartment. Possible causes of entrapment are listed in **Table 17–8**.

<table>
<thead>
<tr>
<th>Table 17–8 Causes of superficial peroneal nerve entrapment.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overstretching during ankle injuries involving inversion and plantar flexion</td>
</tr>
<tr>
<td>Thickening of the deep fascia of the lateral leg</td>
</tr>
<tr>
<td>Lateral compartment muscle hernia or fascial defect</td>
</tr>
</tbody>
</table>
Clinical Findings

A. Symptoms and Signs
Clinical manifestations include tingling and paresthesias along the lateral aspect of the lower leg and dorsum of the foot with sparing of the first web space. Pain is typically exacerbated by activity. On physical examination point tenderness may be elicited 10–12 cm above the lateral malleolus where the nerve exits the deep fascia.

B. Diagnostic Studies
Electrodiagnostic evaluation reveals prolonged latency and nerve conduction slowing with stimulation proximal to the entrapment. Needle EMG is normal, as the peroneus longus and brevis are innervated proximally.

Treatment
Treatment options should include lateral wedges in the shoes and physical therapy for ankle rehabilitation. If these measures are ineffective, other options such as corticosteroid injection or surgical release can be tried.

Tarsal tunnel syndrome is caused by compression of the posterior tibial nerve or its associated branches within the tarsal tunnel, a fibrous–osseous space that extends from the posteromedial aspect of the ankle to the plantar aspect of the foot. The tunnel is divided into two compartments: proximal, at the level of the tibiotalar joint, and distal, at the level of the subtalar joint. The tarsal tunnel contains the posterior tibial nerve and its branches. The posterior tibial nerve provides motor and sensory function to the plantar muscles of the foot and toes. Possible causes of entrapment are listed in Table 17–9.

Table 17–9 Causes of posterior tibial nerve entrapment.
Clinical manifestations include paresthesias along the plantar aspect of the foot and toes. Tinel’s sign over the tarsal tunnel and muscle weakness of the plantar muscles of the foot can be seen. MRI findings include increased size and signal of the tibial nerve and its branches, denervation edema of the plantar muscles of the foot, and enhancement of the tarsal tunnel on postgadolinium images.

SURAL NEUROPATHY

Sural neuropathy is caused by entrapment of the sural nerve at the level of the fifth metatarsal base, where the sural nerve bifurcates into lateral and medial terminal branches. The sural nerve is a pure sensory nerve arising in the midleg region. The nerve is formed by a branch of the tibial nerve (medial sural nerve) and a branch of the common peroneal nerve (lateral sural cutaneous nerve). The nerve courses downward between the two heads of the gastrocnemius and pierces the deep fascia from within at the upper calf. More distally it courses along the posterolateral aspects of the leg, behind the lateral malleolus and along the lateral aspect of the foot. It supplies sensation to the lateral aspect of ankle and foot up to the base of the fifth toe. Causes of entrapment are listed in Table 17–10.

**Table 17–10** Causes of sural nerve entrapment.
Clinical manifestation includes paresthesia and or pain along the lateral ankle and foot, which is exacerbated by inversion and plantar flexion of the foot, and chronic calf pain exacerbated by physical activity. Treatment options include an accommodative shoe that relieves extrinsic pressure on the nerve or treatment of the underlying pathology. With intractable cases surgical release can be attempted.

<table>
<thead>
<tr>
<th>Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute trauma, including fractures of the fifth metatarsal base, talus, calcaneus, or cuboid</td>
</tr>
<tr>
<td>Traction injury with secondary fibrosis of the nerve</td>
</tr>
<tr>
<td>Tendinosis of the Achilles or peroneal tendons</td>
</tr>
<tr>
<td>Space-occupying lesions such as ganglia</td>
</tr>
<tr>
<td>Gastrocnemius injury</td>
</tr>
</tbody>
</table>


Myopathies

David Brown, DO
Krystle Williams, MD
Sara Cuccurullo, MD

The term *myopathy* refers to a muscle fiber disorder that can have a variety of etiologies. Myopathies present as pure motor syndromes without any disturbance of sensory or autonomic function. Deep tendon reflexes are usually preserved. In most myopathies, symptoms tend to be unilateral and affect proximal muscles preferentially, although there are exceptions. Because many myopathies cause progressive impairment of patients’ daily functioning, supportive therapy is often necessary to address the physical and psychological effects of these disorders. The final section of this chapter outlines strategies that are often used in rehabilitation of patients with myopathies.

**EVALUATION OF MYOPATHIC DISORDERS**

By far the most frequent symptom of patients presenting with myopathic disease is weakness. Clinically, it is important to differentiate weakness from easy fatigability. A hallmark of myopathy is the inability to generate a forceful contraction. It is important to observe patients performing activities such as walking, climbing stairs, and arising from a sitting, kneeling, squatting, or reclining position or using the arms overhead. Difficulty in performing these tasks signifies weakness rather than fatigue. Patients with complaints of fatigue often describe a subjective loss of energy. In myopathic disorders, objective muscle weakness and loss of function usually accompany fatigue. Pathologic fatigue not accompanied by muscle cramps upon exercise testing or repetitive electrophysiologic testing usually suggests a disorder of the neuromuscular junction (such as myasthenia gravis or Lambert-Eaton myasthenic syndrome),
rather than a myopathy. If the patient develops fatigue along with frank swelling and cramps with exercise, then certain metabolic myopathies may be suspected.

Myalgias (muscle pain) and muscle aches may be a presenting complaint in patients being evaluated for myopathy. Most myopathies and muscle diseases are not associated with severe myalgias or muscles that are very tender to palpation. Severe myalgias and tenderness often accompany fasciitis, infectious myositis, and some metabolic myopathies.

Myopathies can be classified as hereditary or acquired. Information about the progression of the disease process is very important in helping to classify the specific etiology of myopathy. In patients who have deteriorating strength, it is important to make note of whether the rate of progression is over days, weeks, months, or years. A detailed family history and pedigree chart is very useful in clarifying suspected hereditary myopathies. Table 18–1 contrasts the key features of various hereditary and acquired myopathies.

Table 18–1 Etiology of myopathies.
Generally, myopathic electromyogram (EMG) findings will reveal low-amplitude, short-duration motor units. There can, however, be exceptions to this general rule. Early recruitment is another feature of myopathies. Nerve conduction velocities will be normal with normal sensory responses. Compound muscle action potentials (CMAPs) can be small in amplitude. Muscle biopsy may be necessary to confirm a diagnosis of myopathy and ascertain the specific type. Additionally, genetic testing has become an important tool in diagnosis, as advances in the field of genetics have yielded a more detailed understanding of the pathophysiology of myopathies.
HEREDITARY MYOPATHIES

The hereditary myopathies can be grouped into three categories: muscular dystrophies, including Duchenne and Becker variants; congenital myopathies; and metabolic myopathies, comprising the glycogen storage diseases and channelopathies.

MUSCULAR DYSTROPHIES

Muscular dystrophies are a progressive, heterogeneous group of neuromuscular disorders. They are often hereditary. These disorders are characterized by histologic abnormalities that include extensive muscle necrosis and fibrosis, with fat and connective tissue infiltration. Table 18–2 compares the key features of the major classifications of muscular dystrophies.

Table 18–2 Common dystrophic myopathies.
<table>
<thead>
<tr>
<th></th>
<th>Duchenne Muscular Dystrophy (DMD)</th>
<th>Becker Muscular Dystrophy (BMD)</th>
<th>Limb-Girdle Muscular Dystrophy (LGMD)</th>
<th>Facioscapulohumeral Muscular Dystrophy (FSH)</th>
<th>Myotonic Dystrophy</th>
<th>Emery-Dreifuss Muscular Dystrophy*</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Etiology</strong></td>
<td>X-linked recessive (Xp21), spontaneous, most common form of muscular dystrophy, affecting 1 in 3500 male infants</td>
<td>X-linked recessive</td>
<td>Two forms: autosomal dominant (LGMD1), autosomal recessive (LGMD2)</td>
<td>Two forms: FSH1 and FSH2 (classical form)</td>
<td>Two forms: DM1 and DM2, autosomal dominant</td>
<td>Two forms: EMD1 and EMD2, X-linked recessive</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>Early childhood (3–5 y)</td>
<td>Adulthood</td>
<td>LGMD1: later onset, in adulthood, LGMD2: childhood or adolescence</td>
<td>FSH1: infantile form, &lt; 2 y, FSH2: early adulthood (2nd–3rd decade)</td>
<td>DM1: infancy through adulthood, DM2: 20–60 y</td>
<td>Usually in teenage years, but can vary from neonatal period to adulthood</td>
</tr>
<tr>
<td><strong>Symptoms and signs</strong></td>
<td>Proximal muscle weakness (pelvic girdle), Abnormal muscle stretch reflexes, Increased lumbar lordosis, Ambulation difficulties: toe walking (&lt; 5 y), clumsy running (&lt; 7 y), Gower's sign: difficulty rising from floor due to hip and knee extensor weakness, Calf pseudohypertrophy with fat and fibrous tissue, Contractures: iliotibial band (first), Achilles tendon, Scoliosis, causing cardiomyopathy and restrictive lung disease, Possible mental retardation, Wheelchair by age 12 y, Extraocular muscles are spared</td>
<td>Proximal weakness, Calf pseudohypertrophy, Cardiomyopathy, Less mental retardation than DMD</td>
<td>Pelvic and shoulder girdle weakness, Distal muscles, facial muscles, and extraocular muscles are spared, Atrophy of affected muscle groups, Contractures of elbow or heel cords, depending on subtype, May develop cardiomyopathies, Normal intellect</td>
<td>Proximal muscle weakness, Facial droop, Weak eye closing, Weak forehead wrinkling, Arm atrophy with deltoid and forearm sparing (Popeye arm), Cataracts (dry sclera), Retinopathy, Lip protrusion, Transverse smile, Frontal balding, Testicular atrophy, Extracocular muscles are spared, Inability to whistle</td>
<td>Weakness: distal &gt; proximal myotonia with sustained grip, “Hatchet face” (wasting of the temporals and masseter), Frontal balding, Poor vision, Ptosis, Impotence, Hypertrichosis, Mental retardation, Cardiac abnormalities, Endocrine abnormalities, Congenital myotonic dystrophy—“shark mouth” appearance, facial diplegia, possible clubfoot</td>
<td>Early involvement of biceps, triceps, tibialis anterior and peroneal muscles, Later involvement of shoulder and pelvic girdle muscles, Contractures in elbows and ankles, Rigid spine, with neck extension contractures, By 2nd decade, a dilitated cardiomyopathy with conduction defects can occur, Cardiac arrhythmias may lead to death</td>
</tr>
<tr>
<td>Laboratory findings</td>
<td>Muscle biopsy: no dystrophin; internal nuclei variation in fiber size; Blood: increased CK and aldolase; ECG: abnormal</td>
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<tr>
<td>Electrodiagnostic findings</td>
<td>NCS— SNAP: normal; CMAP: (+/-) decreased amplitude; EMG—AA, ER, small MUAP at first</td>
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<tr>
<td>Electrodiagnostic findings</td>
<td>NCS— SNAP: normal; CMAP: (+/-) decreased amplitude; EMG—AA, ER, small MUAP at first</td>
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<tr>
<td>Electrodiagnostic findings</td>
<td>NCS— SNAP: normal; CMAP: decreased amplitude in the involved muscle; EMG—AA, ER, small MUAP</td>
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<tr>
<td>Electrodiagnostic findings</td>
<td>NCS: SNAP: normal; CMAP: decreased amplitude in affected muscles; EMG: AA, ER, small MUAP</td>
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<tr>
<td>Treatment</td>
<td>Rehabilitation, scoliosis surgery before vital capacity drops below 35% (usually due to curvature of &gt; 30 degrees)</td>
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<tr>
<td>Treatment</td>
<td>Rehabilitation, bracing, tendon lengthening, possible scoliosis surgery</td>
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<tr>
<td>Treatment</td>
<td>Physical and occupational therapy to manage weakness, contractures</td>
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<tr>
<td>Treatment</td>
<td>Rehabilitation, bracing, ADL management</td>
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<tr>
<td>Treatment</td>
<td>Monitoring and treatment of cardiac arrhythmias, physical therapy for joint mobility and contracture prevention</td>
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<tr>
<td>Prognosis</td>
<td>Severely progressive (death by 20s)</td>
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<tr>
<td>Prognosis</td>
<td>Slowly progressive</td>
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<tr>
<td>Prognosis</td>
<td>Slowly progressive</td>
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<tr>
<td>Prognosis</td>
<td>Slowly progressive</td>
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<tr>
<td>Prognosis</td>
<td>Slowly progressive</td>
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</tbody>
</table>

CK, creatine phosphokinase; ECG, electrocardiogram; NCS, nerve conduction studies; SNAP, sensory nerve action potential; CMAP, compound muscle action potential; EMG, electromyogram; AA, abnormal activity; ER, early recruitment; MUAP, motor unit action potential; ADL, activities of daily living.

*Also known as humerocoperoneal muscular dystrophy.

*Key muscle to test in FSH is tibialis anterior.

The dystrophinopathies encompass a spectrum of hereditary muscle diseases in which insufficient dystrophin is produced in the muscle cells. Dystrophin is a large, rodlike cytoskeletal protein on the inner surface of muscle fibers. It is part of the dystrophin–glycoprotein complex, which bridges the inner cytoskeleton and the extracellular matrix in muscle. Mutation of the gene leads to the muscle disorders, Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD).

Serum creatine kinase (CK) concentrations are 10–20 times the upper limit of normal in patients with both DMD and BMD and peak at around age 3 years. Serum CK is also mildly increased in 70% percent of DMD and 50% of BMD female carriers.

Electrodiagnostic testing in dystrophinopathies is of limited value, especially when there is a family history of the disorder. Diagnosis requires genetic testing for identifiable mutations in the dystrophin gene and, if this is unsuccessful, a muscle biopsy. Evaluation and treatment of pediatric patients with Duchenne and Becker muscular dystrophies is described in detail in Chapter 20, and readers are referred to that chapter for additional information. An overview of each condition is presented here, along with other disorders in this group, to facilitate comparison of key features of the various dystrophinopathies.

**DUCHENNE MUSCULAR DYSTROPHY (see also chapter 20)**

**ESSENTIALS OF DIAGNOSIS**

- X-linked recessive disorder, more common in males.
- Weakness and hypotonia are often present at birth.
- Child may attain developmental milestones until age 4 or 5 years, then experience difficulty running and jumping.
- Weakness is proximal and manifests with positive Gower’s sign and lumbar lordosis.
- Muscle biopsy specimens show damage to muscle fibers.
Western blot test shows reduced or absent dystrophin (0–3%).

**General Considerations**

DMD is an X-linked disorder caused by an abnormality in the Xp21 gene locus. It is the most common of the muscular dystrophies, affecting up to 1 in 3500 male infants at birth, and only rarely affecting female infants.

**Clinical Findings**

**A. Symptoms and Signs**

Affected infants may manifest weakness and hypotonia or have no obvious abnormality at birth. Symptoms and signs become more apparent as the child develops, which usually prompts diagnosis by age 4 or 5 years. Toe walking is common; this is a compensatory adaptation to knee extensor weakness. A lordotic posture of the lumbar spine is adopted to compensate for hip extensor weakness. Calf pseudohypertrophy and calf pain are other common findings. Gower’s sign, which involves the patient using hands and arms to walk up the body in order to stand up, is another symptom. Patients have varying degrees of cognitive impairment.

The earliest signs of weakness are seen in the neck flexors during the preschool years. The child is often wheelchair bound by the age of 12. Pulmonary function declines gradually. Tachyarrhythmias and cardiomyopathies may develop. Smooth muscle is often involved, with patients developing gastroparesis.

**B. Diagnostic Studies**

Very high CK levels and reduced or absent dystrophin are characteristic findings. Muscle biopsy findings reveal scattered necrotic and regenerating muscle fibers. The Western blot test reveals 0–3% of the normal amount of dystrophin present in muscle tissue.

**Treatment**
Treatment with prednisone helps maintain strength and prolongs ambulation by 2 years. The optimal dose of prednisone is 0.75 mg/kg per day, and benefits may continue for up to 3 years. Supportive therapy is necessary to address the physical and psychological effects of the disease: contracture management requires splinting and bracing; ambulatory decline requires assistive devices; spinal weakness requires appropriate assistance such as proper seating. Half of all DMD patients develop scoliosis between 12 and 15 years of age, which correlates with the adolescent growth spurt. For curvature that has progressed past 20–40 degrees, spinal arthrodesis has been shown to be the only effective treatment. Spinal orthoses are generally not used for prevention of scoliosis in those with DMD. Unfortunately, most patients with DMD die in their late teens or early 20s from ventilatory or cardiac failure.

BECKER MUSCULAR DYSTROPHY (see also Chapter 20)

ESSENTIALS OF DIAGNOSIS

- X-linked recessive disorder.
- Less common than DMD, with later onset (usually after 12 years of age) and slower progression.
- Like DMD, Becker muscular dystrophy affects only boys with rare exceptions.
- Western blot test shows 20–80% of the normal levels of dystrophin.

General Considerations

BMD is an X-linked disorder with an incidence of 5 per 100,000 people. In 10% of cases, the genetic defect is the result of a spontaneous mutation. The same gene locus is affected as in DMD (ie, Xp21), and expression is allelic.

Clinical Findings

A. Symptoms and Signs
BMD is a less severe form of muscular dystrophy than DMD, with a slower rate of progression. Patients typically present after age 12 years, when increasing weakness and disability prompt evaluation. Great variability in phenotypic expression exists, and a wide spectrum of clinical phenotypes can be seen. In contrast to DMD, many patients remain ambulatory past the age of 15 years. Nonetheless, 50% of patients lose the ability to ambulate independently by the fourth decade. Cardiac abnormalities are similar to those described earlier for DMD.

**B. Diagnostic Studies**

Characteristic findings are seen on muscle biopsy evaluation. Muscle is replaced with fat and connective tissue. Reduced levels of dystrophin are noted on the Western blot test (20–80% of normal).

**Treatment**

The approach to treatment is similar to that for DMD, discussed earlier. Many patients survive beyond 30 years of age.

**LIMB-GIRDLE MUSCULAR DYSTROPHY**

**ESSENTIALS OF DIAGNOSIS**

- Equal occurrence in males and females.
- Autosomal-dominant (LGMD1) or autosomal-recessive (LGMD2) mode of inheritance.
- Predominantly affects the pelvic or shoulder girdle musculature, or both.

**General Considerations**

Limb-girdle muscular dystrophy (LGMD) is a hereditary dystrophy that affects males and females equally. Inheritance may be either autosomal dominant or
autosomal recessive, designated LGMD1 or LGMD2, respectively. Genotypic subtypes of each are given alphabetical subclasses, as LGMD1A, LGMD1B, etc. In patients with LGMD1 subtypes, disease onset is usually later, in adulthood. Patients with LGMD2 subtypes usually have onset during childhood or adolescence. Many of the LGMD2 subtypes have been linked to gene defects causing abnormalities of the sarcolemmal-associated proteins.

Clinical Findings

A. Symptoms and Signs
Pelvic and shoulder girdle weakness are common presenting symptoms among patients with all forms of LGMD. Distal muscles are spared as well as facial and extraocular muscles. The rate of progression is slower in LGMD than in DMD. The age of onset can vary from childhood through adulthood, depending on the type. Cardiomyopathies are associated with many types. Pseudohypertrophy of the calf muscles may occur. Atrophy of affected muscle groups along with early contractures of the elbow and heel cords may develop, depending on the subtype. Low back pain may be a prominent symptom in affected patients. Intellect is usually normal.

B. Diagnostic Studies
CK levels are often very high but can vary according to the subtype. Muscle biopsy evaluation can help evaluate different sarcolemmal-associated proteins, including sarcoglycans, dystroglycans, calpain-3, dysferlin, fukutin-related protein, telethonin, and titin, which can help determine subtypes.

Treatment
Treatment consists of supportive care, which may include physical therapy and occupational therapy to support activities of daily living (ADLs) and ambulation. Splinting and stretching are required for contracture management. The course of disease is slowly progressive and may lead to significant disability, depending on the mode of inheritance and subtype.

Fascioscapulohumeral Muscular Dystrophy
ESSENTIALS OF DIAGNOSIS

- Autosomal-dominant inheritance linked to chromosome 4q35 locus.
- Caused by a DNA fragment deletion of D4Z4 at the telomere region.
- Second most common inherited muscular dystrophy in adults.
- Patients usually become symptomatic before age 20.
- Predominantly affects the facial and shoulder girdle muscles.

-General Considerations

Fascioscapulohumeral muscular dystrophy (FSHD) is the third most common form of muscular dystrophy, after DMD and BMD, and the second most common form in the adult population. The disease has two forms: a rapidly progressive infantile form, which manifests within the first two years of life, and the classic form, with onset usually in the second or third decade.

-Clinical Findings

A. Symptoms and Signs

Infantile-onset FSHD is associated with severe weakness that leads to diagnosis within the first 2 years of life. For patients with the classic form of the disease, the onset of weakness is highly variable, ranging from 3 to 44 years, although presentation is generally before age 20. Weakness of the facial muscles may be asymmetric. Shoulder girdle weakness causes scapular winging. Humeral weakness and wasting can occur with sparing of the forearm muscles. In the leg, the tibialis anterior muscle is often affected and may cause a drop foot.

Some patients with FSHD have only mild weakness over the course of their lifetime. Other patients appear to experience a late exacerbation of muscle weakness. After having mild weakness for years, they suddenly develop a marked increase in weakness in the typical distribution over several years, leading to significant disability.
B. Diagnostic Studies

CK levels range from normal to mildly elevated. Muscle biopsy findings may show necrosis. Genetic testing may reveal abnormalities at the D4Z4 region.

Treatment

Treatment of patients consists of supportive care, which may include physical therapy and occupational therapy to support ADLs and ambulation. Splinting and stretching are required for contracture management. The use of prednisone has not been found to be helpful. Ankle-foot orthotics may be helpful in cases of drop foot. Affected individuals with classic FSHD usually have a normal lifespan.

MYOTONIC DYSTROPHY

ESSENTIALS OF DIAGNOSIS

- Autosomal-dominant inheritance.
- The most common inherited neuromuscular disorder of adults.
- Limb weakness starts distally then progresses to the proximal muscles.
- Atrophy and weakness of the facial muscles leads to a “hatchet face” appearance.
- Molecular genetic testing reveals an unstable CTG trinucleotide repeat in the MPK gene.

General Considerations

Myotonic dystrophy is the most common inherited neuromuscular disorder of adults. Myotonia refers to a state of delayed relaxation or sustained contraction of skeletal muscle. There are two common types, designated DM1 and DM2. DM1 can present at any age, including infancy. Most patients with DM2 become
symptomatic between the ages of 20 and 60 years, although the onset can occur in childhood. Age of onset is inversely correlated with the number of repeat links, and exhibits genetic anticipation.

Clinical Findings

A. Symptoms and Signs

DM1, the congenital form of the disease, is associated with severe weakness. However, weakness may not be evident in the adult-onset form. Other clinical signs include delayed relaxation of the fingers after grip, and characteristic facial features, seen in adults with longstanding DM1. These patients have a long thin face with temporal and masseter wasting, sometimes referred to as a “hatchet face. “Adult males often have frontal balding.

DM1 is a systemic disorder affecting the gastrointestinal tract, ventilatory muscles, cardiac muscles, the eyes, and the endocrine system. Cardiac abnormalities are common, and 90% of patients have conduction defects. Neurobehavioral abnormalities are also common.

B. Diagnostic Studies

Molecular genetic testing reveals unstable CTG trinucleotide repeats within the region of the myosin–protein kinase (MPK) gene at 19q13.3. CK levels can be normal or mildly elevated. On muscle biopsy evaluation less necrosis is seen than in the other dystrophies. EMG reveals waxing and waning discharges.

Treatment

Symptomatic, painful myotonia can be treated with agents such as mexiletine or membrane stabilizers such as carbamazepine or phenytoin sodium. These agents have been shown to reduce the symptoms, although with little functional gain. Risk of sudden death increases with male sex, duration of disease, and age.

EMERY-DREIFUSS MUSCULAR DYSTROPHY
ESSENTIALS OF DIAGNOSIS

- Two variants, one of which is X-linked recessive.
- Affects males and females equally.
- Patients usually become symptomatic in the teenage years, but age of presentation can vary.
- Weakness occurs in the biceps brachii, triceps, anterior tibialis, and peroneal muscles.
- Elbow flexion contractures are a hallmark of the disease.

General Considerations

Emery-Dreifuss muscular dystrophy (EMD), also known as humeroperoneal muscular dystrophy, refers to a group of muscular dystrophies characterized by weakness of shoulder and pelvic girdle muscles, contractures, and cardiac conduction abnormalities. There are two main types, EMD1 and EMD2.

Clinical Findings

A. Symptoms and Signs

Age of presentation can vary from the neonatal period to the third decade; however, patients usually become symptomatic by the teenage years. There is early involvement of the humeroperoneal muscles (eg, biceps, triceps, tibialis anterior, and peroneal muscles). Severe contractures of the elbow (flexion) are a hallmark of the disease. Ankle (equinus) contractures, along with spinalrigidity and neck extension, are also characteristic.

Weakness is slowly progressive, and eventually the shoulder and pelvic girdle muscles become involved. A dilated cardiomyopathy often develops in affected individuals by age 20, along with various conduction defects.

B. Diagnostic Studies
Serum CK levels range from normal to mildly elevated. Muscle biopsy findings reveal muscle fiber atrophy in more than 95% of patients. Electrocardiography may show sinus bradycardia or conduction blocks. EMG shows myopathic motor unit action potentials (MUAPs).

**Treatment**

Physical therapy may help maintain joint mobility and prevent contractures. Cardiac arrhythmias in early adult life can lead to death. Careful monitoring is essential as EMD patients with arrhythmias may require pacemaker placement.

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**CONGENITAL MYOPATHIES**

Congenital myopathies describe a group of myopathies present at birth that are distinct from congenital muscular dystrophies. The incidence of these disorders is low, and microscopic evaluation of muscle biopsy specimens is generally required to identify the underlying defect and confirm the diagnosis. Included in this group are central core, nemaline rod, fiber-type disproportion, multicore–minicore, and centronuclear myopathies. **Table 18–3** summarizes the features of the major classifications of the congenital myopathies.

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**Table 18–3** Common congenital myopathies.
<table>
<thead>
<tr>
<th>Etiology</th>
<th>Central Core Myopathy</th>
<th>Nemaline Rod Myopathy</th>
<th>Fiber-Type Disproportion</th>
<th>Multicore–Minicore Myopathy</th>
<th>Centronuclear Myopathy</th>
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<tbody>
<tr>
<td>Onset</td>
<td>Infancy</td>
<td>Infancy</td>
<td>Infancy</td>
<td>Infancy and early childhood</td>
<td>Infancy</td>
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<td>Symptoms and signs</td>
<td>Floppy infant, hypotonia</td>
<td>Floppy infant, hypotonia</td>
<td>Floppy infant, hypotonia</td>
<td>Proximal muscle weakness</td>
<td>Floppy infant, hypotonia</td>
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<tr>
<td></td>
<td>Proximal weakness</td>
<td>Diffuse weakness</td>
<td>Hip contractures</td>
<td>Delayed milestones</td>
<td>Ptosis</td>
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<td>Congenital hip dislocation</td>
<td>Facial involvement</td>
<td>Hip dislocation</td>
<td>Ambulation achieved</td>
<td>Extraocular muscle</td>
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<tr>
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<td>Delayed milestones</td>
<td>Narrowed, long face</td>
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<td>later in life</td>
<td>involvement</td>
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<td></td>
<td>Associated with malignant</td>
<td>High, arched palate</td>
<td></td>
<td>Neck and trunk muscles</td>
<td>Facial diplegia</td>
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<td></td>
<td>hyperthermia</td>
<td>Foot drop</td>
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<td>contracted, causing</td>
<td>Respiratory</td>
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<td></td>
<td>Facial muscles spared</td>
<td>Extraocular muscles spared</td>
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<td>rigid spine or scoliosis</td>
<td>insufficiency</td>
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<tr>
<td>Laboratory findings</td>
<td>Muscle biopsy: central cores in</td>
<td>Muscle biopsy: rod-shaped bodies</td>
<td>Muscle biopsy: multiple</td>
<td>Muscle biopsy: central</td>
<td>Muscle biopsy: central</td>
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<td></td>
<td>type I fibers; absent mitochondria</td>
<td>in sarcolemna</td>
<td>small minicores within</td>
<td>location of fiber nuclei,</td>
<td>location of fiber</td>
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<td></td>
<td>Gene mutation and ryanodine receptor</td>
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<td>muscle fibers</td>
<td>nuclei, forming chains</td>
<td>nuclei, forming chains</td>
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<tr>
<td>Electrodiagnostic findings</td>
<td>EMG: ER, small MUAP</td>
<td>EMG: AA, ER, normal to small MUAP</td>
<td>EMG: ER, small MUAP</td>
<td>EMG: AA, ER, normal to small MUAP</td>
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<tr>
<td>Treatment</td>
<td>Bracing for leg weakness and</td>
<td>Rehabilitation for ambulation and</td>
<td>Rehabilitation, bracing</td>
<td>Rehabilitation, antiseizure</td>
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<td>management</td>
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<tr>
<td>Prognosis</td>
<td>Nonprogressive</td>
<td>Dependent on severity of disease;</td>
<td>Variable; sometimes</td>
<td>Variable (see text</td>
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<td></td>
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<td>neonatal form is often fatal before</td>
<td>nonprogressive</td>
<td>discussion)</td>
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<td>age 1 y</td>
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CK, creatine kinase; EMG, electromyogram; ER, early recruitment; AA, abnormal activity; MUAP, motor unit action potential.
CENTRAL CORE MYOPATHY

ESSENTIALS OF DIAGNOSIS

► Autosomal-dominant inheritance.
► Symptoms may be present at birth or manifest in early childhood.
► Mild to severe weakness affecting the proximal muscles of the legs more than the arms.
► Facial muscles are spared.
► Biopsy specimens show changes to the central core of type 1 muscle fibers.

Central core myopathy is caused by mutations in the ryanodine receptor gene (*RYR1*) on chromosome 19q13.1. Patients may be symptomatic at birth or present in early childhood with mild to moderate proximal leg weakness. The degree of weakness is variable. Motor milestones are delayed. Some patients may never ambulate. Facial weakness, if present, is mild. Contractures are uncommon. Mild respiratory muscle weakness as well as skeletal deformities may be seen in some patients.

Muscle biopsy evaluation shows structural alterations in the center of type 1 muscle fibers. These changes in the core are noticeable on NADH-tetrazolium reductase (NADH-TR) staining. CK levels are normal to mildly elevated. EMG may display fibrillations.

Treatment consists of supportive care, including physical therapy as needed to address ambulatory issues, and bracing and stretching as needed to control contractures. Patients are at risk for developing malignant hyperthermia with general anesthesia. The clinical course is nonprogressive.

NEMALINE ROD MYOPATHY
ESSENTIALS OF DIAGNOSIS

- Autosomal-dominant and autosomal-recessive forms.
- An infantile form causes severe generalized weakness that is often rapidly fatal.
- A more common mild, progressive form can manifest from birth to early childhood.
- Results in proximal and distal muscle weakness.
- Electron microscopy shows rod bodies in the sarcolemma and perinuclear regions.

Nemaline rod myopathy takes its name from the rod bodies that are characteristically seen on electron microscopy, giving the appearance of threads in the subsarcolemma and perinuclear regions. The disease manifests in two forms that vary in severity. It is the most common of the congenital myopathies.

In the infantile form, severe weakness and hypotonia are present at birth. The more common form is mild and slowly progressive. Proximal and distal muscle weakness with low muscle bulk is present. Motor milestones are often delayed. Patients also exhibit a wide-based, waddling gait. Some affected children have minor anomalies including a long thin face, high arched palate, and pectus excavatum.

Serum CK levels are normal to slightly elevated. In the severe form, EMG reveals fibrillations and small MUAPs. Muscle biopsy findings show nemaline rods in the sarcolemma.

Treatment consists of supportive care, including physical and occupational therapy as required for ambulation and ADL deficits. Prognosis depends on the severity of the disease. In the severe neonatal form, death due to respiratory failure often occurs during the first year of life.

FIBER-TYPE DISPROPORTION MYOPATHY
ESSENTIALS OF DIAGNOSIS

- Least common of the congenital myopathies.
- May be autosomal dominant or autosomal recessive.
- Characterized by weakness at birth with a weak cry and suck.
- One third of patients have central nervous system abnormalities.
- Biopsy specimens show small type 1 muscle fibers compared with type 2.
- Many cases are nonprogressive, and symptoms may improve with age.

In fiber-type disproportion, the least common of the congenital myopathies, generalized weakness and hypotonia are present at birth, along with a weak cry and suck. Dysmorphic features that may be present include a rigid spine, kyphoscoliosis, arthrogryposis, hip dislocations, and facial dysmorphism.

Electron microscopy does not show consistent structural changes as seen in other congenital myopathies. Serum CK levels range from normal to mildly elevated. EMG shows early recruitment, myopathic MUAPs, and some fibrillations. Muscle biopsy evaluation shows small type 1 muscle fibers compared with type 2.

Treatment consists of supportive care with physical therapy. Prognosis is variable. The disease is sometimes nonprogressive, and symptoms in this group of patients may improve with age.

MULTICORE–MINICORE MYOPATHY

ESSENTIALS OF DIAGNOSIS

- Predominantly autosomal recessive.
- Symptoms are usually observed in infancy and early childhood.
- Proximal muscle weakness is slowly progressive.
- Microscopy shows multiple minicores within muscle fibers of biopsy
Autosomal-recessive mutations in the genes SEPN1 and RYR1 account for half of all reported cases of multicore–minicore myopathy. Patients with this form of myopathy manifest weakness in infancy or early childhood that involves primarily proximal muscles. Milestones are often delayed, with ambulation achieved later in life. Neck extensors and trunk muscles are frequently contracted, leading to spinal rigidity. Scoliosis occurs in two-thirds of patients, and respiratory muscle involvement is disproportionate to the degree of scoliosis. Cardiomyopathy may also develop.

Diagnosis is based on clinical findings and the presence of multiple minicores within muscle fibers on muscle biopsy evaluation. Serum CK levels are normal. EMG shows early recruitment of small MUAPs. Treatment consists of supportive care, including physical therapy and occupational therapy as needed to address strength and functional deficits. Prognosis is variable.

**CENTRONUCLEAR MYOPATHY**

**ESSENTIALS OF DIAGNOSIS**

- Heterogeneous group of disorders; most common form is X-linked.
- Presents in the neonatal period with severe hypotonia and generalized weakness.
- Evaluation of biopsy specimens shows myonuclei in muscle fibers.

Centronuclear myopathy, also referred to as myotubular myopathy, is caused by a mutation in MTM1, a myotubularin gene involved in muscle cell growth and differentiation. Mothers of affected infants often have a history of polyhydramnios during pregnancy.

Infants have severe hypotonia and generalized weakness in the neonatal period. Developmental milestones are delayed. Ptosis and ophthalmoparesis develop later, after the newborn period. Respiratory complications are common,
and affected infants usually require ventilator support and feeding tubes. Serum CK is normal or slightly elevated. EMG shows myopathic findings, which include early recruitment of short-duration, small-amplitude MUAPs. Diagnosis is confirmed by muscle biopsy findings that show myonuclei in the center of muscle fibers.

Treatment consists of supportive care as needed, including physical and occupational therapy to address ambulation and ADL deficits, and pulmonary rehabilitation to increase ventilatory volume. X-linked myotubular myopathy is usually fatal in infancy, although prognosis is dependent on the type of the genetic mutation. Long-term survivors may require advanced medical support.


**METABOLIC MYOPATHIES**

**GLYCOGEN STORAGE DISEASES**

The process of glycogen synthesis and breakdown is essential to the maintenance of an adequate glucose concentration in muscles, which in turn is necessary for the generation of ATP. Disorders of glycogen metabolism can result in the abnormal storage of glycogen, the mechanism that underlies the glycogen storage diseases. **Table 18–4** summarizes information about three of these diseases (types II, V, and VII) that can cause myopathy.

**Table 18–4** Glycogen storage diseases (GSDs) that can cause myopathy.
<table>
<thead>
<tr>
<th>Etiology</th>
<th>Lysosomal Acid Maltase Deficiency (Pompe's Disease)</th>
<th>Myophosphorylase Deficiency (McArdle's Disease)</th>
<th>Phosphofructokinase Deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Autosomal recessive</td>
<td>Autosomal recessive</td>
<td>Autosomal recessive</td>
</tr>
<tr>
<td></td>
<td>Glycogen storage disease type II</td>
<td>Glycogen storage disease type V</td>
<td>Glycogen storage disease type VII</td>
</tr>
<tr>
<td></td>
<td>Deficiency of maltase leads to accumulation of glycogen in lysosomes and cytoplasm</td>
<td>Caused by mutations in the gene encoding muscle phosphorylase on chromosome 11</td>
<td>Deficiency of phosphofructokinase leads to buildup of glycogen</td>
</tr>
<tr>
<td></td>
<td>Incidence: 1 in 40,000</td>
<td>Incidence: 1 in 100,000</td>
<td>Incidence: unknown (least common of the GSDs)</td>
</tr>
<tr>
<td>Onset</td>
<td>Infancy to adulthood</td>
<td>Usually &lt;15 y</td>
<td>Childhood</td>
</tr>
<tr>
<td>Symptoms and signs</td>
<td>Hypotonia in limb-girdle distribution</td>
<td>Exercise intolerance</td>
<td>Exercise intolerance</td>
</tr>
<tr>
<td></td>
<td>Tongue enlargement</td>
<td>Easy fatigability</td>
<td>Muscle pain</td>
</tr>
<tr>
<td></td>
<td>Cardiomyopathy</td>
<td>Muscle stiffness</td>
<td>Contractures</td>
</tr>
<tr>
<td></td>
<td>Hepatomegaly</td>
<td>Cramping</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Respiratory insufficiency</td>
<td>Second-wind phenomenon: brief rest improves symptoms</td>
<td></td>
</tr>
<tr>
<td>Laboratory findings</td>
<td>Blood: increased CK levels during attacks</td>
<td>Urine: myoglobinuria</td>
<td>Blood: elevated CK level</td>
</tr>
<tr>
<td></td>
<td>Muscle biopsy: vacuoles in type I and type II fibers</td>
<td>Muscle biopsy: excess glycogen, absent phosphorylase</td>
<td></td>
</tr>
<tr>
<td>Electrodiagnostic findings</td>
<td>NCS— • SNAP: normal</td>
<td>NCS— • SNAP: normal</td>
<td>NCS—normal</td>
</tr>
<tr>
<td></td>
<td>• CMAP: normal</td>
<td>• CMAP: normal</td>
<td>EMG—myopathic features with abnormal spontaneous activity</td>
</tr>
<tr>
<td></td>
<td>EMG—abundant myopathic discharges</td>
<td>EMG—electrical silence during attacks (contracture)</td>
<td></td>
</tr>
<tr>
<td>Complications</td>
<td>Cardiac insufficiency</td>
<td>Strenuous exercise can precipitate myolysis (possibly causing renal failure and death)</td>
<td>Exertional rhabdomyolysis</td>
</tr>
<tr>
<td></td>
<td>Respiratory failure</td>
<td></td>
<td>Myoglobinuria</td>
</tr>
<tr>
<td>Treatment</td>
<td>IV recombinant α-glucosidase enzyme</td>
<td>Avoidance of intense isometrics</td>
<td>Avoidance of strenuous exercise</td>
</tr>
<tr>
<td></td>
<td>Supportive care</td>
<td>Supportive care</td>
<td>Supportive care</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Death within first year of life without treatment</td>
<td>Favorable</td>
<td>Favorable</td>
</tr>
</tbody>
</table>

CK, creatine kinase; NCS, nerve conduction studies; SNAP, sensory nerve action potential; CMAP, compound muscle action potential; EMG, electromyogram.
Defects in the muscle ion channels (ie, calcium, chloride, potassium, and sodium) can cause myotonia and episodes of sudden weakness or paralysis. Clinical manifestations of these disorders, part of a group of diseases known as channelopathies, are presented in Table 18–5. Short exercise tests, electrodiagnostic studies, and a careful clinical examination help confirm the diagnosis in patients with these disorders.

**Table 18–5** Channelopathies that can cause myopathy.
<table>
<thead>
<tr>
<th>Biology</th>
<th>Hypokalemic Periodic Paralysis</th>
<th>Hypokalemic Periodic Paralysis</th>
<th>Myotonia Congenita (Thomson's Disease)</th>
<th>Paramyotonia Congenita</th>
<th>Generalized Myotonia (Becker's Disease)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Autosomal dominant</td>
<td>Autosomal dominant</td>
<td>Autosomal dominant</td>
<td>Autosomal dominant</td>
<td>Autosomal recessive</td>
<td>Autonomic recessive</td>
</tr>
<tr>
<td>Multiple secondary causes</td>
<td>Type 1: Ca²⁺ channel defect</td>
<td>Affects Cl⁻ channels</td>
<td>Type 2: Na⁺ channel defect</td>
<td>Affects Ca²⁺ channels</td>
<td>Affects Cl⁻ channels</td>
</tr>
<tr>
<td>Affects Ca²⁺ channels</td>
<td>Caused by a mutation in the gene coding for diphosphorylase-sensitive calcium channels in skeletal muscle</td>
<td>Caused by a mutation in the gene coding for diphosphorylase-sensitive calcium channels in skeletal muscle</td>
<td>Caused by a defect in the gene CLCN1, which causes a defect in skeletal muscle chloride ion channels.</td>
<td>Caused by a mutation in the sodium channel SCN4A, leading to prolonged depolarization</td>
<td>The other major form of myotonia congenita, but presents later in life, with more severe myotonia</td>
</tr>
<tr>
<td>Caused by a defect in the gene regulating production of the protein SCN4A in sodium channels of skeletal muscle</td>
<td>Weakness starts in the legs and spreads proximally</td>
<td>Severe spasms exacerbated by the cold and improved with warmth and exercise</td>
<td>Muscle hypertrophy</td>
<td>Exacerbated by exercise period, pregnancy, emotional stress, and anesthetics</td>
<td>Exacerbated by cold and exercise</td>
</tr>
<tr>
<td>Onset</td>
<td>2nd decade of life</td>
<td>Early in 2nd decade of life</td>
<td>Birth–adulthood</td>
<td>Birth–childhood</td>
<td>Childhood</td>
</tr>
<tr>
<td>Symptoms and signs</td>
<td>Proximal muscle weakness</td>
<td>Weakness starts in the legs and spreads proximally</td>
<td>Severe spasms exacerbated by the cold and improved with warmth and exercise</td>
<td>Stiffness</td>
<td>Myotonia</td>
</tr>
<tr>
<td>Paresthesias of the lips and lower limbs</td>
<td>Attacks last 12–24 hours</td>
<td>Muscle hypertrophy</td>
<td>Weakness</td>
<td>Weakness</td>
<td></td>
</tr>
<tr>
<td>Myotonia</td>
<td>Myotonia seen in the eyelids</td>
<td>Exacerbated by rest after exercise, stress, and a high-carbohydrate diet</td>
<td>Fatigue</td>
<td>Fatigue</td>
<td></td>
</tr>
<tr>
<td>Attacks last 10–60 min</td>
<td>Exacerbated by cold exposure</td>
<td>Myotonia</td>
<td>Myotonia</td>
<td>Myotonia</td>
<td></td>
</tr>
<tr>
<td>May be aborted with exercise</td>
<td>Exacerbated by cold exposure</td>
<td>Dysphagia</td>
<td>Exacerbated by cold and exercise</td>
<td>Exacerbated by cold and exercise</td>
<td></td>
</tr>
<tr>
<td>Laboratory findings</td>
<td>Blood: high K⁺ during attack</td>
<td>Blood: low K⁺</td>
<td>Blood: normal CK levels</td>
<td>Muscle biopsy: fiber size variation</td>
<td>Blood: mildly elevated serum CK levels</td>
</tr>
<tr>
<td>Electrodagnostic findings</td>
<td>NCS—</td>
<td>NCS—</td>
<td>NCS—</td>
<td>NCS—</td>
<td>NCS—</td>
</tr>
<tr>
<td>CMAP: normal</td>
<td>CMAP: normal</td>
<td>CMAP: normal</td>
<td>CMAP: normal</td>
<td>CMAP: normal</td>
<td>CMAP: normal</td>
</tr>
<tr>
<td>EMG—increased insertional activity</td>
<td>EMG—normal between attacks</td>
<td>EMG—myotonic discharges at rest and volitional activity</td>
<td>EMG—myotonic discharges at rest and volitional activity</td>
<td>EMG—myotonic discharges at rest and volitional activity</td>
<td>EMG—myotonic discharges at rest and volitional activity</td>
</tr>
<tr>
<td>Complications</td>
<td>Myotonic reaction with general anesthesia</td>
<td>Progressive proximal myopathy</td>
<td>Chronic joint problems</td>
<td>Extreme sensitivity to cold</td>
<td>Same as myotonia congenita</td>
</tr>
<tr>
<td>Treatment</td>
<td>Diet: high carbohydrate</td>
<td>Diet: K⁺ supplement</td>
<td>Medications: procainamide, phenytoin, quinine</td>
<td>Avoid myotonia—triggering events, warm extremities</td>
<td>Same as myotonia congenita</td>
</tr>
</tbody>
</table>

CK, creatine kinase; EMG, electromyogram; NCS, nerve conduction studies; SNAP, sensory nerve action potential; CMAP, compound muscle action potential.
ACQUIRED MYOPATHIES

Acquired myopathies encompass a wide range of inflammatory, toxic, and systemic disorders that can produce myopathic symptoms (see Table 18–1). Included in this broad group are autoimmune, infectious, drug-induced, and endocrine-related myopathies.

INFLAMMATORY MYOPATHIES

Distinct types of inflammatory myopathies can be associated with connective tissue disorders, neoplasms, or infection, or occur in isolation. The incidence of inflammatory myopathy is 1 in 100,000 per year. Dermatomyositis and polymyositis are autoimmune disorders that can be associated with other mixed connective tissue disorders; the pathogenesis of inclusion body myopathy is unknown. These three disorders are discussed further in Chapter 22, from the perspective of pediatric rehabilitation. Inflammatory myopathy can also result from viral, bacterial, fungal, or parasitic infections.

DERMATOMYOSITIS

ESSENTIALS OF DIAGNOSIS

► Onset from childhood through adulthood.
► Affects female more frequently than males.
► Proximal greater than distal weakness with pain develops over weeks to months.
Heliotrope rash is classic; rash can occur on sun-exposed skin, knees, and chest.
May have associated cardiac, pulmonary, gastrointestinal, and joint maladies.

General Considerations

In dermatomyositis, an autoimmune process causes a vasculopathy that may be complement mediated and associated with malignancy. Disease onset may occur at any age from infancy through adulthood.

Clinical Findings

A. Symptoms and Signs

Weakness occurs over weeks to months. The proximal muscles of the arms and legs are affected first, and then distal involvement follows. Difficulty arising from a chair and raising the arms above the head are typical early functional deficits.

A rash over the eyelids (heliotrope) is classic. A rash can occur on sun-exposed skin, over the knees, knuckles (Gottron papules), back, chest, and hips. The rash may occur before or after the onset of weakness. Systemic vasculopathies may develop. Cardiac arrhythmias, conduction defects, and reduced ejection fraction can occur. A restrictive pulmonary condition occurs in approximately 20% of cases. Patients have a risk of aspiration pneumonia.

Speech and chewing may be affected, and dysphagia can occur in 30% of cases. Gastrointestinal involvement includes dysphagia, aspiration, and gastric paresis due to smooth muscle compromise. Arthralgias and contractures occur in some cases.

B. Diagnostic Studies

Serum tests reveal an elevated CK level, which is usually 50 times greater than normal. Alanine aminotransferase (ALT), aspartate aminotransferase (AST), myoglobin, aldolase, and lactate dehydrogenase levels may be elevated. Antinuclear antibodies (ANA) may be positive. Myositis-specific-antibodies are elevated in a small number of cases. Mi-2 and Jo-1 are examples.
Fibrillations and positive sharp waves are common EMG findings, along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy findings show perifascicular atrophy, inflammatory infiltrates, and perivascular infiltrates.

**Complications**

Patients with dermatomyositis have an increased risk of malignancy, interstitial lung disease, and cardiac disease. Gastroparesis and dysphagia leading to aspiration pneumonia may also occur.

**Treatment**

Immunosuppressive therapy is the first-line treatment. Patients are started on intravenous methylprednisolone, 1 g daily for 3 days, and then transitioned to oral maintenance doses for weeks to months afterwards. CK levels and clinical strength testing should be monitored. Medications that may be used as the second line of treatment if corticosteroids are not efficacious or contraindicated include methotrexate, cyclophosphamide, chlorambucil, cyclosporine, azathioprine, and intravenous immunoglobulin (IVIG). The majority of patients require ongoing treatment.

### POLYMYOSITIS

**ESSENTIALS OF DIAGNOSIS**

- Affects patients older than age 20.
- Affects female more frequently than males.
- Symmetric weakness of the proximal arms and legs.
- Myalgia usually occurs but may not be present initially.
General Considerations

Polymyositis is an autoimmune disorder that results in a cell-mediated assault on muscle fibers.

Clinical Findings

A. Symptoms and Signs

Patients are generally older than age 20 and have progressive proximal weakness over weeks to months. Distal weakness of the arms and legs follow. There can be some facial weakness. Patients can have myalgias initially, or later in the disease course, after weakness develops. As with dermatomyositis, patients have difficulty arising from a chair or raising their arms overhead.

Cardiac arrhythmias and conduction defects, reduced ejection fraction, and congestive heart failure can occur. Approximately 20% of patients have restrictive pulmonary disease. Approximately 45% of patients develop polyarthritis.

B. Diagnostic Studies

Serum tests reveal a highly elevated CK level, often 50 times greater than normal level. ANA may be positive. Aldolase, aminotransferases, myoglobin, ALT, and AST may be elevated.

On EMG, fibrillations and positive sharp waves are common findings, along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy findings show endomysial muscle fibers invaded by T cells.

Complications

The risk of malignancy is lower than for dermatomyositis. However, patients are at risk for developing interstitial lung disease and dysphagia.

Treatment

Treatment is the same as for dermatomyositis, and relies on immunosuppressive therapy as the first-line treatment.
ESSENTIALS OF DIAGNOSIS

- Slowly progressive weakness affects both proximal and distal muscles
- Onset generally after age 50 years, predominantly in males
- Quadriceps, finger, and wrist flexors, and ankle dorsiflexors often become weak initially
- Muscle biopsy specimens may show inclusion bodies of β-amyloid

General Considerations

Inclusion body myositis (IBM) is a rare inflammatory myositis that produces symptoms similar to those of polymyositis. The onset of disease is idiopathic, and diagnosis is made clinically.

Clinical Findings

A. Symptoms and Signs

IBM commonly affects males over age 50 years, involving both proximal and distal muscles. In the majority of patients, the quadriceps, finger and wrist flexors, and ankle dorsiflexors are affected first. The clinical course is marked by slowly progressive muscle weakness. Close to 50% of patients may develop dysphagia.

Weakness can be asymmetric, and some patients have sensory peripheral neuropathies. IBM is not associated with cardiac or pulmonary disease.

B. Diagnostic Studies

Serum tests reveal an elevated CK level that is usually 10 times greater than normal. On EMG, a mild axonal sensory neuropathy is found in some patients. Fibrillations and positive sharp waves are common findings, along with small, short-duration, polyphasic MUAPs, and early recruitment.
Muscle biopsy results are variable, including necrosis, endomysial inflammation, and inclusion bodies of β-amyloid.

**Treatment**

Supportive therapies, including physical therapy and occupational therapy may be needed to address ambulation and ADL deficits; immunosuppressive treatment is not helpful. IBM may not respond as well to treatment as polymyositis.

**SARCOID MYOPATHY**

**ESSENTIALS OF DIAGNOSIS**

- Weakness can be proximal or distal.
- Some patients may have focal myalgias.
- Pulmonary involvement is common.

**General Considerations**

Sarcoid myopathy is a multiorgan disease of noncaseating granulomas that often affects the lungs and can involve muscles as well. The disease is more prevalent in blacks than in whites, and affects women more often than men.

**Clinical Findings**

**A. Symptoms and Signs**

Patients may have deep local muscle tenderness and arthralgias, generalized weakness, and atrophy of muscles. The myositis is usually not severe and may even be asymptomatic. Symptoms of restrictive pulmonary disease are common.
B. Diagnostic Studies

Serum tests show normal to mild CK elevation. EMG findings are normal in some patients. More often, however, EMG shows fibrillations and positive sharp waves along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy evaluation shows noncaseating granulomas.

Treatment

The treatment is directed at moderating the systemic expression of sarcoidosis. Corticosteroids can be used to treat the myositis, if necessary.

INFECTION-INDUCED MYOPATHIES

1. Viral Myopathies

The most common causes of viral myopathy are human immunodeficiency virus (HIV), influenza, and leukemia viruses. Herpes simplex, cytomegalovirus, adenovirus, Epstein-Barr, and respiratory syncytial virus are less common causes. All viral-induced myopathies present clinically with myalgias and weakness after the onset of the viral illness. HIV and influenza-induced myopathies are discussed here as examples of this category.

Human Immunodeficiency Virus

Patients infected with HIV may develop myopathy as a result of the medications required to keep viral replication in check. The severity of myopathy is not directly associated with the degree of immunosuppression. Antiretroviral medications such as zidovudine (AZT) can cause muscle weakness and elevated muscle enzymes. Patients may present with an inflammatory myopathy and may have a concurrent HIV neuropathy with sensory loss or paresthesias. Weakness is progressive, symmetric, proximal, and painful. Symptoms resemble those of polymyositis. Serum tests show an elevated CK level. EMG shows fibrillations and positive sharp waves along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy results show endomysial inflammation. High-dose corticosteroid therapy is effective in alleviating symptoms; however, patients may subsequently develop steroid toxicity.
Influenza Virus

Myopathy sometimes occurs as a complication in patients who are infected with influenza A, B, or C. Severe pain and swelling in the calf muscles of children may arise as the upper respiratory infection symptoms begin to dissipate. In adults, a proximal or general muscle weakness can result. Symptoms tend to be more severe in adults than in children. Myoglobinuria can occur, and in rare cases, acute myoglobinuric renal failure can develop.

Serum tests show an elevated CK level. EMG shows fibrillations and positive sharp waves along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy results show necrotic muscle fibers. The disease is generally self-limiting, and treatment consists of supportive therapy such as pain management and physical therapies, as needed.

2. Bacterial Myopathies

Bacterial infections that commonly cause myopathies include *Staphylococcus aureus*, *Escherichia coli*, streptococci, *Legionella*, and *Yersinia*. *Borrelia burgdorferi* in rare cases can cause a true myositis associated with Lyme disease. In bacterial myopathy, the disease occurs as the result of hematogenous or contiguous spread of organisms in adjacent tissue.

Patients present with a history of febrile illness, and with muscle pain and tenderness, especially in the quadriceps, deltoids, and gluteal muscles. Diagnostic findings are consistent with bacterial infections. Serum tests reveal an elevated or normal CK, and an elevated erythrocyte sedimentation rate. Blood cultures may initially show no growth of organisms.

Treatment consists of appropriate antibiotics prescribed as soon as practical, because patients can become septic if not treated early in the illness. Mortality rates can range from 1% to 10% in complicated cases.

3. Parasitic Infection Myopathies

Parasitic infections, including protozoans, cestodes (tapeworms), and nematodes (unsegmented roundworms), can cause myopathies. In this category are toxoplasmosis, cysticercosis, and trichinosis. Patients with all of these parasitic infections present with myalgias and varying degrees of weakness. Those with toxoplasmosis and trichinosis also present with fever.
Toxoplasmosis

Toxoplasmosis results from infection with *Toxoplasma gondii*, a protozoan. Myositis can occur in patients with toxoplasmosis, producing initial symptoms of fever, weakness, and myalgias. Eating undercooked food can transmit oocysts; common symptoms are fever with lymphadenopathy. Immunocompromised patients are at greatest risk of developing systemic disease such as pneumonia and meningoencephalitis.

Serum tests show an elevated CK level. EMG shows fibrillations and positive sharp waves, along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy findings show cysts of *T. gondii*. Treatment is directed at the causative organism and consists of pyrimethamine and sulfadiazine.

Cysticercosis

Cysticercosis is caused by *Taenia solium*, the pork tapeworm. The disease is transmitted by eating contaminated, undercooked meat. Complications include encephalopathy, seizures, and neurologic deficits.

Infected patients present with muscles that are painful and mildly weak. Pseudohypertrophy may develop. Serum tests show an elevated CK level. EMG shows fibrillations and positive sharp waves, along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy shows larvae, fibrotic changes, eosinophils, plasma cells, macrophages, and lymphocytes.

Treatment includes niclosamide and paromomycin, along with corticosteroids, to kill the adult tapeworm. Praziquantel reduces the size and number of granuloma cysts.

Trichinosis

Trichinosis is a disease caused by *Trichinella spiralis*, a nematode. This is the most common parasite of skeletal muscle. Symptoms develop 2–12 days after eating undercooked meat infected with larvae. Generalized weakness and muscle pain, fever, diarrhea, and abdominal pain are common symptoms. Myocarditis and meningoencephalitis may develop if the heart muscle and central nervous system are invaded.

Serum tests show an elevated CK levels. EMG shows fibrillations and
positive sharp waves along with small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy findings show larvae, cyst granulomas, and eosinophils. Treatment consists of niclosamide and paromomycin, along with corticosteroids.


TOXIC MYOPATHIES

Many substances can cause muscle necrosis either directly or indirectly. Direct effects can be focal or general. Indirectly, substances can cause an immune response or an electrolyte imbalance.

ALCOHOL-RELATED MYOPATHIES

ESSENTIALS OF DIAGNOSIS

- Weakness may be acutely related to alcohol poisoning.
- Weakness may be progressive in the chronic disease state.
- May manifest as cardiomyopathy.

General Considerations
Alcohol abuse, especially among chronic or binge users, produces a range of physiologic effects that often includes myopathy. Severity of myopathy correlates with the amount of alcohol ingested.

**Clinical Findings**

**A. Symptoms and Signs**

Binge drinking can cause an acute necrotizing myopathy. Affected patients have severe muscle pain, swelling, and cramping. Acute renal failure may result from myoglobinuria.

Acute hypokalemia associated with alcohol abuse can result in generalized weakness. Chronic alcoholism results in a proximal muscle weakness or a limb girdle weakness. Concomitant alcoholic neuropathy is common. Rhabdomyolysis is followed by acute renal failure.

**B. Diagnostic Studies**

Serum tests for acute alcohol toxicity show an elevated CK level and low potassium level. In patients with acute hypokalemic myopathy, potassium level is less than 2 mEq/L. In patients with chronic alcoholism, the CK level may be normal or slightly elevated. EMG findings may reveal positive waves and fibrillations, small MUAPs, and early recruitment. Muscle biopsy results show areas of necrosis.

**Treatment**

Treatment involves supportive therapy during alcohol withdrawal, and should always include referral to an appropriate substance abuse program.

**COCAININE-INDUCED MYOPATHY**

**ESSENTIALS OF DIAGNOSIS**

- Onset of myalgias is usually 1 hour after cocaine use by any route.
Myalgia is usually accompanied by other symptoms (eg, delirium, seizure) but may be the presenting symptom.

Muscle injury can occur after initial or repeated use of cocaine. Moderate to severe myalgias develop after cocaine use. Severe rhabdomyolysis, acute renal failure, and compartment syndrome can develop depending on the degree of muscle injury. Serum tests show a CK elevation (variable), and electrolyte abnormalities (ie, hyperkalemia, hypocalcemia). Treatment consists of supportive care and referral to an appropriate substance abuse program.

MEDICATION-INDUCED MYOPATHIES

STATINS

ESSENTIALS OF DIAGNOSIS

- Myalgias and proximal weakness.
- Concurrent use of a statin and other medications.

Statins used alone have a slightly less than 1% incidence of causing a severe myopathy. The risk rises for toxic myopathy when these drugs are used in combination with the following medications: fibric acids, niacin, erythromycin, cyclosporine, and ezetimibe. Hepatic or renal dysfunction also increases the risk. Symptoms of muscle injury can occur anytime during treatment.

The exact mechanism by which cholesterol-lowering drugs cause myopathy is not clear. Serum tests show an elevated CK level. EMG of weak muscles reveals positive waves and fibrillations, small, short-duration, polyphasic MUAPs, and early recruitment. Muscle biopsy specimens show scattered necrosis. Discontinuance of the cholesterol-lowering drug is required. If symptoms do not resolve, a course of corticosteroids may be needed. After discontinuing the statin, weakness may persist or increase for 6 months.

GLUCOCORTICOIDS (STEROID MYOPATHY)
GLUCOCORTICOIDS (STEROID MYOPATHY)

ESSENTIALS OF DIAGNOSIS

► Proximal muscle weakness, affecting the legs more than the arms.
► Muscle weakness is not associated with pain or tenderness.
► Coincides with the use of glucocorticoids.
► May occur within weeks of corticosteroid use initiation; however, often occurs after chronic use.

Patients who undergo lengthy treatment with glucocorticoids may develop a form of muscle weakness termed steroid myopathy. Muscle tenderness and pain do not occur. A cushingoid appearance is present in chronic corticosteroid users as a side effect of therapy. Doses of greater than 30 mg/day have higher risk of producing side effects. Acute severe weakness can occur in patients receiving high-dose intravenous corticosteroids. Respiratory muscles may be involved in patients with systemic malignancy.

Serum tests show normal CK and low potassium levels, resulting from high levels of glucocorticoids. EMG findings are normal, because type 2 fibers are primarily affected. Early recruitment may be seen. Muscle biopsy evaluation shows atrophy of type 2 fibers.

Tapering the dose of corticosteroids alleviates symptoms. Muscle strength eventually improves after the drug is discontinued.

MYOPATHIES ASSOCIATED WITH SYSTEMIC DISEASE

CRITICAL ILLNESS MYOPATHY

ESSENTIALS OF DIAGNOSIS

- Generalized weakness involving the extremities, truncal muscles, and muscles of respiration.
- Occurs in conjunction with severe illness.
- Often manifested by the patient’s inability to be weaned from a ventilator.

General Considerations

Critical illness myopathy, also referred to as ICU myopathy, is a form of generalized weakness involving the extremities, truncal muscles, and muscles of respiration that frequently occurs in conjunction with severe illness. Intervenous corticosteroid use is the strongest risk factor for developing critical illness myopathy.

Clinical Findings

A. Symptoms and Signs

Patients with critical illness myopathy often cannot be weaned from a ventilator, indicating the severity of the myopathy. The extremity and truncal muscles are also weak. Sepsis or multiorgan compromise is a common antecedent event, with weakness following. Corticosteroids or neuromuscular blocking agents may have been used.

A critical illness neuropathy may also be present. Distinguishing between the two, and determining the contribution of each to the weakness, can be difficult, especially if voluntary motor recruitment is compromised. Deep vein thrombosis may develop, along with other complications related to being immobilized for
long periods of time.

**B. Diagnostic Studies**

Serum tests show moderately elevated CK levels. Motor conductions studies show low amplitudes with normal latencies and motor velocities. Sensory amplitudes should be normal. Insertional activity is abnormal, with fibrillations and positive sharp waves; small, short-duration, polyphasic MUAPs; and early recruitment. Muscle biopsy findings include variable scattered necroses, and type 2 or type 1 fiber atrophy.

**Treatment**

Treatment of the underlying maladies is crucial, along with withdrawal or tapering of neuromuscular blocking agents and corticosteroids. Physical and occupational therapy may be prescribed. Strength usually returns over the course of weeks to months.

### ENDOCRINE MYOPATHIES

#### THYROTOXIC MYOPATHY

**ESSENTIALS OF DIAGNOSIS**

- Proximal muscle weakness and atrophy.
- Myalgias may or may not occur.
- Occurs in patients with common features of hyperthyroidism.

**General Considerations**

Thyrotoxic myopathy is a complication of hyperthyroidism that is more likely to develop in patients as they age.
Clinical Findings

A. Symptoms and Signs
Overt symptoms of hyperthyroidism include nervousness, palpitations, psychosis, diarrhea, and weight loss. Weakness may be present but is usually not a primary complaint of patients. Myalgias occur with dysphagia and respiratory decline. Thyrotoxic hypokalemic periodic paralysis can occur as well. Bulbar and respiratory muscles may be affected, although this is very rare.

B. Diagnostic Studies
Serum tests show normal CK level and altered thyroid hormone levels. The thyroid-stimulating hormone (TSH) level is low; triiodothyronine (T₃) is elevated; thyroxine (T₄) may be elevated; and potassium levels are low in patients with thyrotoxic periodic paralysis. EMG results are usually normal. Muscle biopsy findings are unremarkable, showing nonspecific atrophy.

Treatment
Muscle weakness usually improves with treatment of the hyperthyroidism. Anticholinesterase inhibitors will not produce improvement in patients with thyrotoxic myopathy. Full recovery usually occurs within a few months.

HYPOTHYROID MYOPATHY

ESSENTIALS OF DIAGNOSIS

- Proximal muscle weakness.
- Myalgias and cramps.
- Occurs in patients with common features of hypothyroidism.

General Considerations
Hypothyroid disease is more common in women than men, producing a range of symptoms that often includes myopathy.

► **Clinical Findings**

**A. Symptoms and Signs**
Patients develop proximal muscle weakness with cramping and myalgias. Hair loss and thickened skin are general features of the disease. Severe muscle weakness and rhabdomyolysis may occur.

**B. Diagnostic Studies**
Serum tests show CK levels that are 10–100 times normal, with low T₃ and T₄ and elevated TSH levels. EMG findings may be normal or may reveal low-amplitude polyphasic MUAPs. Muscle biopsy findings may be normal or reveal nonspecific changes.

► **Treatment**
Thyroid hormone replacement therapy should improve weakness. Recovery occurs over several weeks to months, depending on the severity of muscle damage.

**HYPERPARATHYROID MYOPATHY**

► **ESSENTIALS OF DIAGNOSIS**

► Proximal muscle weakness and atrophy, worse in lower extremities.
► Myalgias may or may not occur.
► Occurs in patients with common features of hyperparathyroidism.
General Considerations

Hyperparathyroid myopathy develops in fewer than 10% of patients with primary hyperparathyroidism.

Clinical Findings

A. Symptoms and Signs

Elevated parathyroid hormone levels in patients with primary hyperparathyroidism eventually cause osteomalacia. This leads to microfractures with a high incidence of muscle involvement. Bone pain is common as microfractures develop. Fibrous tissue replaces mineralized bone, leading to osteitis fibrosis.

Weakness is proximal and affects the legs more often than the arms. Neck extensors can become weak, causing a dropped head syndrome. Elevated parathyroid hormone is associated with the triad of osteitis fibrosis, kidney stones, and duodenal ulcers. Myopathy is not common in patients with hypoparathyroidism. Patients may have a coexisting neuropathy. Dysphagia and respiratory involvement can occur.

B. Diagnostic Studies

Serum tests show elevated parathyroid hormone, elevated calcium, low phosphate, and normal CK levels. EMG findings include small, short-duration, polyphasic MUAPs; early recruitment; and normal insertion of activity. Muscle biopsy results show muscle fiber atrophy.

Treatment

For primary hyperparathyroid disease, the treatment is surgical resection of the gland or adenoma. Chronic renal disease causes a secondary hyperparathyroidism: for this, vitamin D replacement is helpful, and renal transplantation is the ultimate solution. Full recovery within weeks of treatment is expected.

CUSHING’S SYNDROME
ESSENTIALS OF DIAGNOSIS

- Painless proximal muscle weakness.
- Cushingoid appearance.
- Results from excess glucocorticoids.

General Considerations

Cushing’s syndrome results from exposure to very high levels of cortisol, most often in response to glucocorticosteroid medications or tumors of the pituitary or adrenal glands. Tumors of the pituitary gland produce adrenocorticotropic hormone (ACTH), triggering the adrenal glands to produce large amounts of cortisol (Cushing’s disease). Tumors in the adrenal gland itself, high levels of corticotrophic-releasing hormone (CRH), or ectopic ACTH syndromes can also produce similar symptoms. Steroid myopathy, discussed earlier in the chapter, can occur in patients who receive long-term corticosteroid therapy. It is the most common endocrine-related myopathy.

Clinical Findings

A. Symptoms and Signs

Patients with Cushing syndrome have a characteristic appearance that includes truncal obesity, hirsutism, and acne. Other features of the disorder are glucose intolerance and hypertension. Respiratory muscle involvement may occur, although it is rare.

B. Diagnostic Studies

Serum tests show normal CK level. EMG shows normal insertional activity and MUAPs, but early recruitment may be seen. Muscle biopsy evaluation shows atrophy of type 2b fibers.
Treatment

If an adrenal tumor is the cause, it must be excised. For iatrogenic causes of Cushing’s syndrome, reduction of corticosteroid dosing is required. In either case, exercise should be encouraged to prevent further atrophy. Recovery of strength usually occurs over a few months.

ACROMEGALY

Acromegaly is a rare disorder that occurs when excess growth hormone is produced after puberty, often as a result of a tumor of the anterior pituitary gland. Severity of muscle weakness correlates with duration of acromegaly as opposed to the levels of growth hormone.

Proximal muscle weakness without atrophy can develop slowly. Patients have the typical bony overgrowth appearance of acromegaly. Symptoms may be due to a pituitary adenoma. Bony overgrowth can lead to nerve root and spinal cord compression. Serum test show a normal CK level. EMG shows small, short-duration MUAPs. Muscle biopsy findings show muscle fiber atrophy.

Surgical resection of a pituitary adenoma, if present, with resultant lowering of growth hormone level should improve the symptoms of myopathy and usually results in improved muscle strength.

DIABETIC MUSCLE INFARCTION

ESSENTIALS OF DIAGNOSIS

- Pain with swelling and tenderness at the site of infarction.
- Quadriceps, hamstrings, gastrocnemius, or thigh adductors are common sites.

General Considerations

Diabetic muscle infarction is a complication of diabetes mellitus that may occur in patients with poorly controlled disease.
Clinical Findings

A. Symptoms and Signs

Diabetic muscle infarction is a focal process. There is acute pain, swelling, and tenderness over the affected area. Patients usually have the other features of severe diabetes, including nephropathy, neuropathy, and renal failure. Muscle biopsy may cause hemorrhage. Symptoms may recur in the contralateral leg.

B. Diagnostic Studies

Serum tests show a normal CK level. EMG shows increased insertional activity, positive waves and fibrillations, and small MUAPs at the site. Magnetic resonance imaging, computed tomography, and ultrasound scans all show abnormal findings at the site. Muscle biopsy evaluation shows large areas of necrosis.

Treatment

Treatment consists of immobilization of the affected site and pain control. Muscle pain and swelling usually resolve after several weeks.

REHABILITATION IN MYOPATHIC DISORDERS

Comprehensive management of the various clinical problems associated with myopathies is necessary to maximize function. For patients with both rapidly and slowly progressive myopathies, a submaximal strengthening program is recommended. Therapeutic modalities should be performed regularly to prevent the development of contractures, although they may be inevitable in some myopathic conditions. These include regularly prescribed periods of standing and walking if the patient is capable of being upright, passive stretching of the muscles and joints with a daily home exercise program, and night splinting if necessary.

Surgical release of contractures and appropriate bracing should be immediately implemented after the loss of ambulation in patients with myopathies such as DMD. This approach can increase additional walking time by up to 2–3 years. The management of spinal deformity with orthoses is ineffective in DMD patients. Spinal arthrodesis is the only effective treatment for scoliosis in DMD patients. Improved pulmonary toilet can be achieved in
patients with respiratory difficulties through assisted cough, incentive spirometry, percussion, and postural drainage. The swallowing mechanisms of patients can be monitored using a fluoroscopic video dynamic swallowing evaluation. Poor nutritional status, labored feeding, and symptoms of dysphagia are indications for initiation of supplemental enteral feedings via nasogastric tube or gastrostomy.


Normal volitional control of movement depends on the balanced relationship of the cortical, subcortical, cerebellar, spinal, and peripheral nervous systems. Neurologic dysfunction can result from compromise at any point in this balance resulting in motor or sensory impairment, or both. This chapter reviews essential features of common diseases encountered in neurorehabilitation, including amyotrophic lateral sclerosis, Guillain-Barré syndrome, polio and post-polio syndrome, and Parkinson’s disease, with a focus on diagnosis, treatment, and rehabilitation for each disorder.

AMYOTROPHIC LATERAL SCLEROSIS

ESSENTIALS OF DIAGNOSIS

► Most widely known motor neuron disease.
► Insidious onset and rapidly progressive course.
► Most common presentation is painless asymmetric limb weakness and atrophy.
► Diagnosis is primarily clinical, based on the presence of upper and lower motor neuron signs.
**General Considerations**

In its classic form, amyotrophic lateral sclerosis (ALS) is a rapidly progressive neurodegenerative disease affecting both upper and lower motor neurons that results in destruction of the anterior horn cells, motor cranial nerve nuclei, and corticospinal–bulbar tracts. Patients most commonly present with painless asymmetric limb weakness, but other symptoms associated with upper and lower motor neuron degeneration include spasticity, pathologic reflexes, and muscular atrophy. The worldwide prevalence is 5–7 in 100,000, and men are more commonly affected than women at a ratio of 1.5:1.0. The disease onset is typically between 40 and 60 years of age (mean: 58 years).

A group of atypical motor neuron diseases has been studied that usually but do not always evolve into ALS. These disorders—progressive muscular atrophy (PMA), progressive lateral sclerosis (PLS), and progressive bulbar palsy (PBP)—are distinct but interrelated disorders, perhaps representing variants within a spectrum of ALS disease.

**A. Progressive Muscular Atrophy**

PMA is a sporadic disease affecting only the anterior horn cells without involvement of the upper motor neurons (UMNs). Hence, those affected exhibit only lower motor neuron (LMN) signs. As such, the most common presenting symptom of PMA is distal limb weakness with muscle atrophy. The natural history of PMA and ALS demonstrate such a high degree of similarity as to often be indistinguishable, and in one study of PMA subjects, 35% of patients developed UMN signs, mirroring the classic ALS phenotype.

**B. Progressive Lateral Sclerosis**

PLS is also a rare sporadic motor neuron disorder. The initial manifestations of the disease are dominated by UMN features. The chief complaint is progressive asymmetric spasticity, most commonly in the legs followed by the arms or bulbar muscles. The disease progression appears to be slower than in ALS, occurring over years to decades. The life expectancy of patients with PLS is also better than that of ALS patients, ranging between 7 and 14 years. Ultimately, approximately 45% of PLS patients go on to develop LMN symptoms and progress to ALS.

**C. Progressive Bulbar Palsy**
In rare instances motor neuron disease is limited to the bulbar muscles. Patients with this condition most often present with dysarthria or dysphagia, or both.

**Pathogenesis**

Although the cause of ALS is unknown, it may share common biologic mechanisms with disorders such as Parkinson’s, Alzheimer’s, and other neurodegenerative diseases. One widely held hypothesis postulates an interaction between genetic susceptibility and environmental exposure in sporadic ALS. Investigations are currently underway to find susceptibility genes or materials within the environment that predispose certain individuals to sporadic ALS. Evidence supports several different causative mechanisms leading to the demise of motor neurons: nucleic acid changes, oxidative damage, mitochondrial dysfunction, growth factor expression, protein aggregation, inflammatory cascades, derangement of cytoskeletal elements and axonal transport, excitotoxicity, and apoptosis. How these mechanisms work to achieve motor neuron death is unknown, but a variety of factors, either in sequence or in combination, most likely predispose certain individuals to develop the ALS phenotype.

**Clinical Findings**

**A. Symptoms and Signs**

The initial clinical symptoms of ALS can vary greatly. Clinical diagnosis relies on the demonstration of UMN and LMN signs, although either may be absent early in the course of the disease. Signs of LMN involvement include muscle weakness, atrophy, diminished or absent deep tendon reflexes, and fasciculations. Identification of UMN signs can be more subjective as they can be transient, developing and then disappearing as the more clinically dominant LMN features manifest. In addition, signs indicative of UMN pathology—sustained clonus, hyperactive deep tendon reflexes, spasticity, and a Babinski sign—may not manifest clinically in some patients with ALS. Additional UMN features to be aware of are forced yawning, retained reflexes in an atrophied and weakened limb, pseudobulbar affect, and pathologic reflexes (eg, Hoffman sign’s and a heightened jaw, gag, or snout reflex). Pseudobulbar affect is an interesting feature in what is primarily known as a pure motor disorder, and refers to uncontrollable episodes of laughing, crying, or other emotional displays.
The most common presentation of ALS is a patient with a combination of UMN and LMN features. Typically patients present to the physician with persistent, unexplained weakness and atrophy. These signs are frequently painless and asymmetric at onset. Neurologic examination may reveal hyperreflexia, increased muscle tone, and clonus. Some patients may not have sought early medical evaluation or may have done so, but their physicians did not recognize the significance of the symptoms. In these instances their disease may evolve from limb-dominant ALS to bulbar-dominant ALS. Such patients present with dysarthria, dysphagia, and aspiration. Head drop is often seen and results from weakness in the neck extensors, causing a chin-on-chest deformity that requires cervical bracing.

Other signs and symptoms associated with ALS are loss of manual dexterity, cachexia, fatigue, and diffuse musculoskeletal complaints. Fasciculations, particularly in the setting of weakness, should raise a strong suspicion of motor neuron disease. Wasting and fasciculations on examination of the tongue indicate LMN involvement of the cranial nerves. Muscle cramping is frequently elicited during manual muscle testing but can occur anywhere in the body, including the trunk. Ventilatory difficulties and paradoxical abdominal movements usually indicate diaphragmatic weakness and necessitate pulmonary function testing.

The presence of cognitive and behavioral deficits in ALS was recognized in the 19th century; however, for many years this was overlooked as an area of clinical investigation and research. These deficits can exist in both sporadic and familial forms of the disease and are frequently the initial symptoms. Cognitive impairment is most prominent in the domain of executive function and language. This impairment may appear in a clinical setting as disorganization, mental inflexibility, word finding difficulty, and fluent semantic dementia. Behavioral difficulties are displayed in social and personal interactions. Patients may be unable to receive and interpret nonverbal cuing, and they may become disinhibited or withdrawn.

**B. Laboratory Findings**

General laboratory workup includes a chemistry panel, complete blood count (CBC), and creatine kinase concentration. Based on the patient’s history and presentation, additional laboratory evaluation may include antibody testing to assess immune-mediated causes and serology testing to rule out infectious etiologies. If there is a family history of motor neuron disease, DNA mutational analysis is warranted. A muscle biopsy can be done if myopathy is suspected, but findings are not diagnostic for ALS.
C. Diagnostic Studies

1. Electrodiagnostic studies—Electrodiagnostic testing improves diagnostic accuracy and should be performed on every patient suspected of having a motor neuron disease. Testing offers insight into the rate of progression and helps to exclude other diagnoses that may mimic ALS. Nerve conduction studies (NCS) with electromyography (EMG) can confirm a pattern of active or chronic denervation and demonstrate fasciculation potentials in multiple muscles innervated by multiple segments in multiple regions. Although these findings are not diagnostic of ALS, they have been shown to be a consistent pattern. Most often NCS indicates a normal sensory response, normal or low motor amplitudes, and up to a 25% decrease in conduction velocities. Needle EMG shows a pattern of decreased recruitment, normal to large motor unit action potentials, and abnormal spontaneous activity such as positive sharp waves, complex repetitive discharges, and fibrillation potentials. Single-fiber EMG and repetitive stimulation can also be included in this study.

2. Other tests—Magnetic resonance imaging of the brain and spinal cord can exclude other diagnoses such as cord compression, syrinx, tethered cord, stroke, or tumor. Pulmonary function testing is invaluable in assessing the patient’s current level of respiratory impairment. For a patient who presents with bulbar muscle weakness, referral to speech therapy for a video-assisted swallow evaluation is warranted to determine the risk of choking or aspiration.

Differential Diagnosis

For a patient with a combination of UMN and LMN signs, the differential diagnostic considerations are limited. ALS may mimic hereditary spastic paraparesis if the patient exhibits spastic paraparesis. Myasthenia gravis must also be considered in a patient with a bulbar presentation of ALS. Other disorders deserving of consideration include inflammatory myopathies, X-linked spinobulbar muscular atrophy, head and neck cancer, and syringobulbia.

Complications

As ALS progresses, patients experience complications consistent with continued neurodegeneration of the motor system. Respiratory problems are the most common serious complication of ALS. Dysfunction in the upper airway and
expiratory muscles leads to impaired swallow and inadequate cough. This decreased ability to clear secretions and protect the airway leads to an increased risk of aspiration with swallowing. Weakness in the inspiratory muscles, including the diaphragm and external intercostal muscles, results in retention of carbon dioxide. Nocturnal hypoventilation is a common problem in patients with ALS, a consequence of reduced neural output to the respiratory muscles coupled with decreased ventilatory drive that occurs during sleep. Ultimately this restrictive lung disease leads to respiratory failure, necessitating cough-assistive devices, mechanical ventilation, and tracheostomy.

The use of noninvasive positive-pressure ventilation (NIPPV) has been shown to be beneficial by improving duration and quality of life. NIPPV can serve as an effective temporizing measure before the progression of weakness and onset of bulbar symptoms make more invasive ventilation such as tracheostomy or laryngeal diversion necessary for continued survival. Because of the multiple adverse effects on the respiratory system, a pulmonary specialist is an integral part of ALS management. Frequent monitoring of pulmonary function provides valuable information on disease progression and prognosis. In addition, objective measurements of pulmonary function may help patients and families address crucial decisions concerning more invasive interventions or end-of-life issues.

Bulbar muscle weakness as a result of motor neuron involvement in the brainstem causes dysfunction in the lips, tongue, pharyngeal and laryngeal muscles. Swallowing and cough can be impaired, making clearance of oral secretions and ingestion of adequate nutrition difficult. In addition, deficits in speech production can make attempts at communication frustrating. Early referral to speech therapy for a comprehensive evaluation of speech and swallowing function can address these issues by providing patients with compensatory strategies to maintain intelligible speech and adequate oral nutrition. Recent development of sophisticated computer-based augmentative communication devices has greatly enhanced the ability to communicate when phonation is no longer possible.

Dysphagia can be addressed initially by instruction in compensatory swallowing techniques such as chin tuck, head turn, and double swallow. This instruction is essential in helping to maintain oral nutrition and decrease the risk of aspiration. ALS patients often have difficulty with certain dry foods and thin liquids. Modifying the diet by thickening liquids, moistening solids, and changing food texture can help. The addition of high-calorie liquid supplements helps to maintain caloric requirements and stave off weight loss and cachexia.
Because malnutrition can precipitate increased fatigue, muscle breakdown, and risk of death, a referral to a registered dietician can be invaluable in maintaining appropriate nutrition through diet and supplementation. When nutrition can no longer be maintained orally, the option of enteric feeding must be broached with the patient and family. Placement of a feeding tube must be strongly considered with weight loss greater than 10%, increase in the time required for oral feedings, and the occurrence of aspiration pneumonia. Feeding tubes have been shown to stabilize weight, decrease fatigue, and prolong survival.

Spasticity is treated if it impairs function, prevents bracing, or causes pain.

Treatment

Currently, no treatment options are able to reverse or arrest the progression of ALS. Consequently, the management of ALS revolves around slowing the inexorable progression of symptoms, minimizing complications, maintaining independent patient function, and ensuring meaningful quality of life. Optimal management of patients with ALS and their families is an arduous task and cannot be accomplished singly. The interdisciplinary approach provides management of patient care issues by a team composed of physicians, therapists, social workers, and mental health professionals. Physiatrists offer the training and expertise to direct the rehabilitation team as well as to oversee the goals of treatment. A comprehensive program of rehabilitation is essential in maximizing functional capacity, maintaining function, optimizing mobility within the home and community, and preventing physical deformity.

Riluzole remains the only Food and Drug Administration (FDA)-approved pharmacologic treatment available that has been proven to affect the natural history of ALS and slow its progression. A Cochrane Database review in 2007 concluded that riluzole at a dose of 100 mg daily prolonged median survival by 2–3 months. Other studies suggest an even greater benefit ranging from 4 to 20 months. Unfortunately, this drug comes at considerable expense and may not be affordable for all ALS patients. The most common side effects are asthenia, nausea, and reversible hepatotoxicity, requiring monitoring of liver function. In recent years there has been an increase in ALS research as well as clinical trials for promising medications that have the potential to slow progression of the disease. The search for potential therapeutic targets based on motor neuron degeneration may lead to novel treatments for ALS.

As in all other motor neuron diseases, the essential condition in ALS is musculoskeletal weakness, which ultimately causes the majority of
complications. Exercise training is not absolutely contraindicated, and a program of aerobic, strengthening, and endurance exercise prescribed with a commonsense approach can yield physical benefits such as improved cardiovascular endurance and increased muscle efficiency. Patients may also experience improvements in pain, appetite, sleep, and psychological well-being. Ideal therapeutic modalities include aquatherapy, stationary bicycling, and other low-impact activities. However, patients should be advised not to exercise to exhaustion and should be educated on the symptoms of overwork weakness, such as increased muscle cramping, fasciculations, and prolonged shortness of breath.

It is important to discuss end-of-life issues with ALS patients and their families. Physicians should outline available treatment options and choices, and a social worker should be involved, when possible, to help the patient designate a durable power of attorney as well as to assist in drafting a living will.

## Prognosis

Sadly, ALS remains a rapidly fatal disease, with a median survival of 3–5 years. However, longer survival is not unseen. About 15% of ALS patients survive 5 years after initial diagnosis, and 5% survive past 10 years. Younger age at symptom onset, male gender, and limb-dominant symptoms all offer improved chances at long-term survival. Poor prognostic factors include older age at symptom onset, bulbar and pulmonary dysfunction early in the clinical course, a short period from symptom onset to diagnosis, and the presence of frontotemporal dementia.

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Mitchell JD, Borasio GD: Amyotrophic lateral sclerosis. Lancet
GUILLAIN-BARRÉ SYNDROME

ESSENTIALS OF DIAGNOSIS

► Most common cause of acute neuromuscular paralysis.
► Recognized as a syndrome of disorders with various subtypes.
► Characterized by progressive onset of symmetric limb weakness.
► Cranial nerve involvement occurs in 45–75% of patients, most often after the limbs are affected, and severe pain in 50%.
► Weakness of oropharyngeal and respiratory muscles affects 40% of presenting patients.
► Can be confirmed with electrodiagnostic testing, which also differentiates demyelinating from axonal forms.

General Considerations

With the eradication of poliomyelitis in the Western world, Guillain-Barré syndrome (GBS) has become the most common cause of acute neuromuscular paralysis, with an annual incidence of 1–2 per 100,000. The male-to-female ratio is 1.5:1. Most patients are older males, although GBS has been reported in all age groups. In the United States the disease appears to have a bimodal distribution, with the first peak occurring in young adulthood (15–35 years of
GBS is characterized by a progressive, symmetric weakness of the limbs. Reflexes are typically absent or greatly diminished, and sensory deficits may be present. Today it is recognized that GBS is not a single disorder but a syndrome of several subtypes of acute immune-mediated polyneuropathies. The term *acute inflammatory demyelinating polyradiculoneuropathy (AIDP)* is often used synonymously with GBS, and it accurately describes the histopathologic features seen in this disease. AIDP is the most common subtype, accounting for 95% of GBS cases in Europe and North America. Although rare, accounting for only 5–10% of all US cases, axonal subtypes also occur, as an acute motor axonal neuropathy (AMAN) or an acute motor sensory axonal neuropathy (AMSAN). The prevalence of axonal subtypes is higher in South America and Asia, comprising 30% of GBS cases in those regions. Miller-Fisher syndrome (MFS) is a subtype of GBS that manifests in a unique triad of ataxia, areflexia, and ophthalmoplegia.

### Clinical Findings

GBS is considered to be a postinfectious, immune-mediated disease that attacks peripheral nerves, leading to progressive, symmetric onset of limb weakness. Up to two thirds of patients report an infectious illness in the weeks before symptom onset. Respiratory infections are most often reported, followed by gastrointestinal infections. The most commonly identified infectious agent is *Campylobacter jejuni*, but other causative agents include cytomegalovirus, Epstein-Barr virus, *Mycoplasma pneumonia*, and *Haemophilus influenzae*. There is also a well-recognized association between GBS and acute human immunodeficiency virus (HIV) infection.

### A. Symptoms and Signs

The typical patient with GBS, which in most cases is the AIDP subtype, presents 2–4 weeks after a relatively benign upper respiratory or gastrointestinal illness with complaints of bilateral weakness in the lower extremities and dysesthesias in the fingers. The classic clinical picture of weakness is one that is ascending and symmetric in nature with involvement of the lower extremities occurring before the upper extremities. Proximal muscles may be involved earlier in the course than distal muscles. Signs of axial involvement may also be seen, with weakness in the trunk and bulbar muscles. The patient may complain of difficulty standing or walking secondary to ophthalmoparesis or proprioception.
deficits. Shortness of breath may be present due to respiratory muscle weakness.

The onset of weakness is typically acute, with progression over days to weeks. Maximal weakness usually occurs by 2 weeks. By 4 weeks, 98% of all cases have reached maximal weakness. The severity of weakness over the course of GBS can be variable, from mild paresis to complete tetraplegia.

Involvement of cranial nerves is observed in 45–75% of patients with GBS. Most frequently, cranial nerves III–VI and IX–XII are affected. Common symptoms with cranial nerve involvement include ophthalmoplegia, dysphagia, dysarthria, diplopia, and facial droop. In most cases, facial symptoms appear after the limbs and trunk are affected. The Miller-Fisher variant is unique in that cranial nerve deficits appear first, with minimal weakness in the limbs.

Loss of sensation is variable and is usually mild. Sensory symptoms often precede muscle weakness, manifested as paresthesias or numbness beginning in the toes and fingertips and migrating proximally. Distal loss of proprioception, light touch, and vibration sense may also be observed.

Pain can be a significant complaint, and on initial presentation approximately 50% of patients rate their pain as severe. The most common locations of pain are in the shoulder girdle, buttocks, thighs, and back. Characterized as deep, aching, and throbbing in nature, pain initially may be nociceptive from acute nerve inflammation, but it can evolve during the course of the illness into neuropathic pain as sensory nerve degeneration followed by regeneration occurs. Neuropathic pain persists indefinitely in 5–10% of patients.

Involvement of the autonomic nervous system has also been observed in patients with GBS. Autonomic dysfunction in the sympathetic and parasympathetic systems can manifest with hypertension, cardiac arrhythmias, tachycardia, bradycardia, facial flushing, orthostatic hypotension, and diaphoresis. Urinary retention and constipation can also occur. The likelihood of dysautonomia is higher in patients with severe weakness and respiratory failure.

Upon presentation, 40% of patients complain of weakness in oropharyngeal or respiratory muscles. Typical symptoms include shortness of breath, dyspnea on exertion, difficulty swallowing, and slurred speech. These patients must be monitored for respiratory failure with sequential measurement of forced vital capacity every 2–4 hours initially because up to one third will require ventilatory support at some point during the course of the disease.

B. Laboratory and Diagnostic Studies

The diagnosis of GBS is generally made based on clinical features. Laboratory studies such as CBC and chemistry panel are less useful but are utilized to
exclude other potential diagnoses.

1. **Cerebrospinal fluid studies**—Lumbar puncture for cerebrospinal fluid (CSF) studies is recommended and routinely performed in patients presenting with rapidly progressive weakness. The characteristic finding on CSF analysis is albuminocytologic disassociation, which is an elevated CSF protein level greater than 0.55 g/L with normal white blood cell count (<5–10 × 10⁶ cells/L). The elevated CSF protein is believed to reflect widespread inflammation within the nerve roots. However, a normal CSF protein level does not rule out GBS, as the level in 10% of patients may be normal. Additionally, CSF protein may not rise until 1–2 weeks after the onset of weakness.

2. **Imaging studies**—MRI is sensitive but not specific in GBS diagnosis. The use of gadolinium with resulting nerve root enhancement is strongly suggestive of GBS; however, this nonspecific feature is commonly seen in inflammatory conditions where the blood–nerve barrier has been disrupted.

3. **Electrodiagnostic studies**—Electrodiagnostic testing is the most useful confirmatory test for GBS, able to differentiate between the more common demyelinating form (AIDP) and axonal forms (AMAN, AMSAN). Details of these findings are presented in Chapter 17.

#### Complications

Severe, early symptoms of GBS significantly increase the risk of complications during hospitalization. The care of acutely affected patients involves close monitoring of respiratory and cardiovascular status. Depending on the patient’s initial presentation and course, management of these issues may best be accomplished in an intensive care unit (ICU). Severe muscle weakness may affect the diaphragm and intercostal muscles to such an extent that intubation and ventilatory support may be indicated. Autonomic dysfunction can lead to blood pressure fluctuations and cardiac arrhythmias, complicating the use of vasoactive and sedative medications. In addition, bladder areflexia and adynamic ileus can occur with derangement of the parasympathetic system.

Because of the high percentage of cranial nerve involvement in patients with GBS, symptoms of dysarthria and dysphagia may precede aspiration and lead to pneumonia. The prolonged immobilization that can occur with weak or paralyzed muscles is a significant risk factor for development of deep vein
thrombosis and pressure ulcers.

### Treatment

The management of GBS consists of supportive care, disease-modifying treatment, and rehabilitation. Patients should be admitted to a hospital for close monitoring until the course of the disease has reached a plateau or undergone reversal. Continued progression may result in a neuromuscular emergency with complications related to respiratory insufficiency and cardiac arrhythmia. Primarily because of respiratory failure, about one third of GBS patients require ICU admission. Chemical prophylaxis to prevent deep vein thrombosis along with frequent repositioning for pressure relief will minimize problems related to immobility. Continued care is also needed to minimize complications from neurogenic bowel and bladder, and pain.

Immunomodulatory therapy is beneficial in patients with GBS, hastening recovery. Intravenous immunoglobulin (IVIG) and plasma exchange have proven equally effective. IVIG is usually the preferred treatment because of greater availability and convenience. Administration is for 5 consecutive days at 0.4 g/kg per day. Plasma exchange administered five times over the course of 2 weeks is beneficial when started within 4 weeks of symptom onset. The exact mechanism of action is unclear but likely involves removal of autoantibodies, complement, immune complexes, or other humoral factors involved in the pathogenesis of GBS.

Given the marked degree of weakness, potential complications, protracted recovery, and possible long-term functional deficits, rehabilitation of patients with GBS should begin in the acute phase. Patients with persistent functional impairments may require transfer to an inpatient rehabilitation unit. Initial therapeutics should begin with range of motion, positioning, and splinting followed by early mobilization to minimize the effects of prolonged bed rest. As strength improves the patient can be advanced to ambulation and self-care activities utilizing adaptive equipment and assistive devices. After discharge, outpatient therapy and a home program are essential in continued functional gains and a return to independence.

### Prognosis

Although rare, death may occur from acute complications such as respiratory
distress syndrome, cardiac arrest, sepsis, pneumonia, and venous thromboembolic disease. Most often, mortality is due to complications of prolonged intubation and paralysis or from severe autonomic instability. GBS-related deaths usually occur in ventilator-dependent patients, and mortality is increased in elderly patients, particularly those with underlying pulmonary disease and the need for mechanical ventilation. Although most patients (up to 85%) with GBS make a full and functional recovery, 2–12% will die from complications related to GBS, and a significant percentage of survivors will have persistent symptoms.

After symptoms reach a plateau, which usually occurs within 4 weeks from onset, the recovery phase of GBS begins. This period typically lasts from 6 to 12 months, but for some patients it may take up to 3 years. Although the exact prevalence is uncertain, up to 50,000 persons in the United States may have long-term functional deficits from GBS. Estimates indicate 15–20% of patients will have moderate residual deficits, and 1–10% will be left severely disabled. Long-term sequelae of GBS include persistent fatigue, weakness, imbalance, and residual sensory changes. Approximately 7–15% of patients have permanent neurologic sequelae, including bilateral foot drop, intrinsic hand muscle wasting, sensory ataxia, and dysesthesia. Patients may also exhibit long-term differences in pain intensity, fatigability, and functional impairment compared with healthy controls. In a small percentage (~10%) of patients, an acute relapse occurs after initial improvement or stabilization after treatment.


ESSENTIALS OF DIAGNOSIS

Polio:

- Results from infection with an RNA virus that invades the motor cells of the anterior horn cells (LMN) and spinal cord.
- Common symptoms include fever, malaise, headache, and myalgias.
- Patients may also report gastrointestinal and respiratory symptoms.
- Paralysis is asymmetric, with involvement of lower extremities greater than that of upper extremities.

Post-polio syndrome:

- Confirmed history of paralytic polio followed by partial or fairly complete neurologic and functional recovery and a period of stability for at least 15 years.
- Onset of two or more of the following symptoms since achieving stability: unaccustomed fatigue, new weakness in muscles previously affected or in unaffected muscles, joint pain, functional loss, cold intolerance, or new atrophy.
- No other medical explanation for the new health problems.

General Considerations

Poliomyelitis is a disease caused by an RNA virus that invades the motor nerve cells of the anterior horn cells (LMNs), and spinal cord, producing weakness in affected bulbar or spinal myotomes, or both. Before the advent of the Salk trivalent inactivated polio vaccine in 1956, and the Sabin trivalent oral live vaccine in 1962, polio was the most common viral infection of the central nervous system, affecting more than 600,000 children per year worldwide.

Polio has been eradicated in the United States since 1979; however, there may remain as many as 1.5 million polio survivors in this country, and millions more worldwide, where a small percentage of initial cases continue to be reported. Post-polio syndrome (PPS) is a neurologic disorder that occurs in polio
survivors 20 or more years after recovery from the initial infection. New-onset weakness is the hallmark symptom. PPS has been reported to affect anywhere from 20% to 60% of polio survivors.

**Pathogenesis**

Poliomyelitis infection can be classified into several distinct phases (acute, recovery, and post-polio syndrome) based on the onset of infection and course of the disease. During the *acute phase* of the disease, often lasting several days, 95% of the anterior horn cells are affected. Because these cells control the skeletal muscles of the trunk and limbs, diffuse and severe paralysis results. The *recovery phase* is a period of relative stability, when anterior horn cell survival, axonal sprouting, and muscle hypertrophy occurs and allows for improved strength. The recovery phase can be spread out over a period of months, during which therapeutics used to treat the complications experienced in the acute phase are instituted, such as rehabilitation, and treatment of orthopedic and respiratory complications. After 20–40 years of stability, between 20% and 60% of previously stable individuals experience late-onset worsening, or new-onset involvement, often with symptoms of fatigue, muscular weakness, pain, and atrophy, resulting in further functional impairment and progressive debility. Although this condition may be in part due to medical complications, or the process of normal aging, a *post-polio syndrome* has been described (PPS). The length of time since acute poliovirus infection, the presence of permanent residual impairment after recovery from the acute illness, and both overuse and disuse of neurons all increase the risk of developing PPS. While PPS is a diagnosis of exclusion and the cause remains uncertain, various explanations have been proposed. These focus on viral reactivation and replication, enterovirus infection, ongoing infection at the spinal level, effects of aging and muscle overuse or disuse, chronic denervation exceeding reinnervation, dysfunction of surviving motor neurons, and distal degeneration of axons in enlarged motor units.

**Clinical Findings**

**A. Symptoms and Signs**

1. **Polio**—Poliovirus infection primarily affects the musculoskeletal and neurologic systems. Initial symptoms include fever, malaise, headache,
gastrointestinal, and respiratory symptoms. Worsening disease leads to myalgias and paralysis, affecting asymmetrically the lower extremities more commonly than the upper extremities. Maximal recovery can take years, and improvement may be complete or only partial.

In a patient with clinical evidence of an earlier polio infection, the physical examination should support findings of a LMN process, with absent or diminished reflexes, decreased muscle tone, asymmetric atrophy and weakness or paralysis of the limbs or trunk musculature, and spared sensation. Neurologic signs of fasciculation and peripheral neuropathies may also be seen in polio survivors.

A. LOWER EXTREMITY FINDINGS—Although the poliomyelitis virus can affect muscles in any extremity, resulting in partial or complete paralysis and impairment, the quadriceps and hip abductors are most often involved. Tibialis anterior muscle involvement is also commonly found, and usually results in a complete muscle paralysis.

Knee hyperextension and genu recurvatum is seen with early complete quadriceps paralysis. To ensure stance stability, the residual hip extensors and calf musculature are recruited to control the limb, shift body forces anterior to the knee, and prevent knee flexion. Knee pain and joint instability often become significant as the disease progress, and there is increased posterior torque and anterior compression applied to the joint and ligamentous structures.

Individuals with early complete quadriceps paralysis demonstrate characteristic compensatory features. Stance stability while loading the limb is achieved by thigh retraction and increased foot plantar flexion. Mid and terminal stance stability is gained by decreasing ankle plantar flexion. When using crutches, double limb support with bilateral knee and hip hyperextension, and complete foot contact, is required to advance the crutches. Ipsilateral heel rise and knee flexion begins after crutches are advanced, and body weight shifted to the contralateral limb. Initial swing phase incorporates a quick and excessive hip flexion. Terminal swing knee extension remains incomplete due to lack of functioning quadriceps, and a rapid thigh retraction and ankle plantar flexion are used to prepare for weight acceptance.

The late development of quadriceps weakness results in various substitutive motions that may be observed in the gait cycle. Functionally, the same mechanics of relative knee hyperextension are used to substitute for quadriceps weakness and obtain knee stability. Initial contact is made with a small degree of heel strike, and a loading response of minimal knee flexion. To maintain knee
extension stability, ankle dorsiflexion remains limited at mid and terminal stance. There is limited knee flexion in preswing, reduced hip flexion with initial swing, and delayed knee extension at midswing, with ankle dorsiflexion to ensure foot clearance.

**B. Upper extremity findings**—Although muscles of the lower extremities are often involved more frequently and extensively, the upper extremity can also be affected. Loss of shoulder muscle strength can result in difficulties with activities of daily living, and loss of independence, as hand placement in space is dependent on preserved shoulder range of motion.

Wrist involvement can also occur, as affected individuals place a large amount of pressure on their wrists when transferring, or walking with ambulatory devices. Over time, this can lead to wrist pain, carpal tunnel syndrome, or bony subluxation and arthritic deformities. Paralysis of the intrinsic muscles of the hand, not uncommon in PPS, also results in loss of function. Various orthopedic complications can occur, and can be addressed with the use of orthotic devices, and orthopedic interventions.

**2. Post-polio syndrome**—The hallmark of PPS remains weakness and atrophy, which often translates to worsening or loss of function. Polio survivors are thought to lose strength at a greater rate than that associated with normal aging. The weakness may occur in muscles previously affected by the disease, or in those thought to have been unaffected. Many symptoms seem to be related to overuse of weak muscles, with delayed recovery after heavy use, and pain from chronic stresses on joints, ligaments, and tendons in affected extremities. Other PPS symptoms may include muscle atrophy, swallowing or breathing problems, cold intolerance, fatigue, and joint and muscle pain. The muscle imbalance that results from quadriceps paralysis in PPS patients is demonstrated in two gait patterns, early complete quadriceps paralysis, and late quadriceps weakness (described earlier). Shoulder weakness is noted in 95% of patients with PPS and has a close correlation with lower extremity involvement.

Restrictive lung disease, associated with chronic alveolar hypoventilation is also commonly seen, especially in polio survivors with initial respiratory muscle involvement. Shortness of breath, dyspnea with exertion, and chronic respiratory infections may be common complaints. In those individuals who may have initially presented with bulbar involvement, new onset or worsening dysphagia may manifest with vague complaints of difficulty swallowing, coughing, or choking. In limbs affected by paralysis, polio survivors often report cold intolerance; however, a general inability of these individuals to thermoregulate
has been reported, causing them to limit their exposure to cold. These features results from several factors, including alterations in the sympathetic nervous system and paralysis of muscles in the extremities that are needed to help maintain dynamic blood flow.

Fatigue, worsened with activity and increased use, is the most common complaint of polio survivors, occurring in up to 87% of individuals. Fatigue can also affect cognition in terms of memory and concentration. The pain associated with PPS is most often due to muscle imbalance, poor posture and body mechanics, repetitive stress, and a worsening of underlying osteoarthritis that may already affect the joint. It is usually more pronounced at the end of the day, and typically is described as muscular aching, burning, or cramping.

Polio survivors require a comprehensive and detailed musculoskeletal evaluation, including history and physical examination, which should be repeated annually. Initially, simple manual muscle testing is useful in measuring strength, and various asymmetries that may exist. It is important to note patient use of compensatory movements or strategies on muscle testing as the individuals attempt to maintain stability of the joint or muscle involved.

**B. Diagnostic Studies**

Although a history of polio or PPS cannot be proven by electrodiagnostic studies alone, certain EMG findings are suggestive, and are helpful in ruling out neuromuscular or other disease processes. EMG and NCS changes consistent with prior anterior horn cell disease include increased amplitude and duration of motor unit potentials, increased percentage of polyphasic potentials and, decreased maximum motor unit recruitment in weak muscles. Fibrillations and sharp waves may also be noted.

Spinal imaging such as MRI, computed tomography (CT), or myelography may be helpful to exclude spinal conditions such as spondylosis, spinal stenosis, myelopathy or radiculopathy, as well as neoplasm. Plain radiographs can help in the diagnosis of osteoarthritis. Laboratory studies may include routine evaluation of blood chemistry, thyroid function, and muscle enzymes.

If bulbar symptoms exist and swallowing is affected, a dysphagia evaluation, including a modified barium swallow evaluation or functional evaluation of swallowing (FES), may be performed. Symptoms of fatigue and sleep dysfunction may be explored through pulmonary function and nighttime sleep study evaluations.
Differential Diagnosis

Many medical and neurologic conditions have symptoms that overlap with those of PPS, and these should be considered in the differential diagnosis (Table 19–1).

Table 19–1 Differential diagnosis of polio.
<table>
<thead>
<tr>
<th>Symptoms and Signs in Polio</th>
<th>Disorders that Can Produce Similar Symptoms</th>
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<tr>
<td>Weakness</td>
<td>Adult spinal muscular atrophy</td>
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<td>Amyotrophic lateral sclerosis (ALS)</td>
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<td></td>
<td>Deconditioning</td>
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<td>Multiple sclerosis</td>
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<td>Myasthenia gravis</td>
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<td>Myelopathy</td>
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<td>Myopathy</td>
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<td>Peripheral neuropathy</td>
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<td>Radiculopathy</td>
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<td>Pain</td>
<td>Bursitis</td>
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<td>Fibromyalgia</td>
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<td>Myofascial pain syndrome</td>
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<td>Osteoarthritis</td>
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<td>Peripheral neuropathy</td>
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<td></td>
<td>Radiculopathy</td>
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<td></td>
<td>Tendinitis</td>
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<td>Respiratory problems</td>
<td>Cardiac disease</td>
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<td>Chronic obstructive pulmonary disease</td>
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<tr>
<td>Cold intolerance</td>
<td>Peripheral vascular disease (PVD)</td>
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<tr>
<td>ALS–upper motor neuron (UMN) signs</td>
<td>Multiple sclerosis</td>
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<tr>
<td></td>
<td>Stroke</td>
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<td></td>
<td>Tumor</td>
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<tr>
<td></td>
<td>Exclusion of medical, orthopedic, rheumatologic, or neurologic diseases</td>
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<tr>
<td></td>
<td>Other—depression, anemia, thyroid, respiratory disease, PVD, chronic infection</td>
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<tr>
<td>Fatigue</td>
<td>Anemia</td>
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<td></td>
<td>Cancer</td>
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<td>Cardiac dysfunction</td>
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<td>Chronic infection</td>
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<td>Chronic systemic infection</td>
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<td>Deconditioning</td>
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<td></td>
<td>Depression</td>
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<td></td>
<td>Fibromyalgia</td>
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<td></td>
<td>Medication side effect</td>
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<td></td>
<td>Respiratory dysfunction</td>
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<td></td>
<td>Sleep disorder</td>
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<td>Thyroid dysfunction</td>
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<td>Swallowing problems</td>
<td>Gastroesophageal reflux</td>
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<td>Mass lesion</td>
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<td>Stricture</td>
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Complications

Polio complications may include permanent nerve damage, mild febrile illness, headache, sore throat, nausea, vomiting, painful muscle spasms, oropharyngeal dysphagia, nasal regurgitation, and pulmonary aspiration.

PPS is characterized by progressive weakness and gait difficulties that often lead to loss of balance and falling. Many of these falls result in injury and fracture, which further reduces functional ability. Osteopenia and osteoporosis are common in polio survivors due to weakness, deconditioning, paralysis, and atrophy of affected extremities. This puts these individuals at increased risk of fracture and pain conditions. Other common orthopedic complications found in PPS include fixed flexion deformities, hyperextension or lateral instability of the hip or knee joint, progressive instability of joints, osteoporosis, fractures, osteoarthritis, scoliosis, and cervical spondylosis.

Neurologic complications in PPS include poor concentration due to mental fatigue, bulbar weakness, progressive wasting and weakness in already affected extremities, muscle weakness causing respiratory insufficiency, and sleep apnea in those with bulbar involvement. Supplemental oxygen may be used in individuals with loss of endurance due to progressive respiratory compromise. Aspiration pneumonia and malnutrition may be evident should disease-associated dysphagia become severe.

The severe physical stresses of post-polio disability contribute to the development of progressive orthopedic, respiratory, neurologic, and general medical abnormalities. Many of these complications are potentially treatable, and most individuals can be helped to understand and manage increasing disability. Functional deterioration does not necessarily progress once it has occurred, and the associated fatigue and reduced mobility may often progress slowly or stabilize.


Treatment

Medical treatment targets the presenting symptoms of weakness, fatigue, pain, respiratory complaints, cold intolerance, and dysphagia. Procedures such as trigger point injections, joint injections, acupuncture, and various anesthetic procedures may be used to treat pain. Surgery may be recommended for certain orthopedic conditions that may occur in the polio survivor. The accompanying bed rest that often results after surgery may result in significant recuperative difficulties, and should be avoided by starting postoperative rehabilitation and ambulation early. Fatigue should be addressed by treating any contributing concomitant medical conditions.

A. Medical Treatment

1. Pharmacotherapy—There is no specific pharmacologic treatment for polio survivors. Corticosteroids and immunoglobulin have been suggested to target the immunologic and inflammatory process of the disease with mixed or little benefit on pain, fatigue, or strength. Pyridostigmine, which may improve the transmission of nerve–muscle fiber at the neuromuscular junction or motor plate, has been tried with mixed results to improve strength. Lamotrigine and bromocriptine, may be beneficial but require further investigation. Coenzyme Q10 has been tested because of potential positive effects on mitochondria and protective antioxidant properties with little benefit noted in strength or fatigue.

2. Physical therapy and exercise—Several studies report the beneficial effects of exercise in the treatment of individuals with PPS. It is essential to strike a balance so that exercise regimens help alleviate symptoms without causing increased weakness and fatigue in damaged muscles. Submaximal aerobic training and low-intensity muscular strengthening may have positive effects on
muscular strength and cardiorespiratory reserves. Aquatherapy has demonstrated a positive impact on pain reduction, improved mobility, and enhanced muscular functioning.

A muscular strengthening program can be implemented for PPS patients that allows individuals to regain muscle strength without adverse side effects. Strengthening is usually applied to those muscle groups that are capable of at least antigravity movement on manual muscle testing. Isolation is not advised for deficient muscles because of their propensity for excessive fatigue. A moderate and progressive intensity exercise protocol should be used so as not to go beyond the muscular fatigue threshold. The response to exercise should be reassessed regularly and adjusted if there is any complaint of resultant pain or worsening of fatigue. Periods of rest must be incorporated into the exercise program.

Lack of endurance is commonly reported by PPS patients. In polio survivors, energy consumption for ambulation and activities of daily living is increased. Studies have shown significant improvements in the maximum volume of oxygen utilization (VO$_{2\text{max}}$) and respiratory volume, as well as a decrease in maximal heart rate (HR$_{\text{max}}$) with aerobic conditioning programs. The positive impact of aerobic training programs such as bicycle riding and fast walking, which incorporate large muscle groups, shows that capacities to adapt to exercise training are proportionally similar in PPS patients and healthy subjects.

Aquatherapy has often been a preferred modality for individuals with PPS. It allows muscular strengthening to be implemented in a controlled resistance environment and also provides active assistive benefits for deficient muscles as a result of the intrinsic buoyancy of water. The warmth of the water and its antigravity effect may also contribute to pain reduction.

Most therapeutic studies in PPS describe an 8- to 16-week training program, with exercise sessions two to three times weekly for durations of 20–40 minutes. Training programs should always be of moderate intensity, with an exercise threshold of 70% of HR$_{\text{max}}$. Improvements have been shown in strength, as well as cardiac and pulmonary parameters (HR$_{\text{max}}$, VO$_{2\text{max}}$, and blood pressure); however, positive effects have only been demonstrated for the short term. Individuals with impaired respiratory function should be educated to recognize the signs of developing chest infections and have a prophylactic supply of antibiotics on hand, receive regular flu and pneumococcal immunizations, avoid smoking, and maintain ideal body weight.

The fatigue in PPS has been described by individuals as a feeling of having to force oneself to perform simple daily activities that were managed effortlessly
before. Part of any therapy program must include instruction in energy conservation. Healthy, effective, and efficient lifestyle changes may include maintaining a stable weight, sitting and rest breaks when needed, daily planning to minimize repeated transfers or repetitive activities, environmental changes in the home for safety and energy consumption, and using assistive walking devices or orthoses.

3. Bracing and orthotics—Orthotics are used by up to 50% of PPS individuals to support and assist the movement of weakened lower extremities while walking, standing, and transferring, while also providing increased control and improved energy efficiency. When considering whether to prescribe an orthotic device, the provider must consider the patient’s prior orthotic use, pain, fall history, new muscle weakness, and fatigue levels. Studies have shown that patients with PPS have 28% slower gait velocity, 9% greater energy consumption, and 40% greater energy cost.

The lack of normal supporting musculature in those with PPS often leads to advanced arthritic degenerative joint changes and ligamentous laxity, with associated pain. In addition to overuse, pain most often results from angular deformities that stress the supporting structure of the involved joint. The goal of orthotic use is not to restore proper limb alignment, but to prevent extremes in joint motion and reduce pain to an acceptable level. The extremity that may be failing often is the intact side, as a result of the chronic compensatory stresses placed on the unaffected side. The most effective treatment may be to restore the more severely involved leg to a more functional state with the use of orthotics. Heavy, bulky or inappropriate orthotics, however, may adversely affect mobility and can increase risk of falling.


B. Surgical Treatment

Orthopedic surgery interventions may be needed in the treatment of polio survivors to relieve pain, correct deformities, transfer muscles to counter muscle imbalance, provide stability to unstable joints, and allow for more effective use of orthoses. Most surgeries are performed on the upper and lower extremities.

The shoulder joint, which is important for hand placement in space, is dependent on muscle strength for active mobility. With overuse and improper loading, rotator cuff injuries are common. These injuries often require local arthroscopic debridement or surgical repair. The use of ambulatory aides, such as canes or crutches, may lead to wrist and hand involvement. Chronic pressure loading often results in carpal tunnel syndrome requiring surgical release, as well as advanced arthritis. Joint arthrodesis may be needed to relieve pain and enhance stability. For loss of function or paralysis that occurs in the muscles of the hand, procedures such as tendon transfers and capsulodesis may be performed.

For enhanced functional ambulation and energy efficiency, it is important to maintain full hip and knee range of motion in PPS patients. In select patients, total hip arthroplasty has been used to achieve this goal. Hip adduction deformity, iliobibial band contracture, leg length discrepancy, and valgus foot deformity can predispose to valgum alignment at the knee. Painful genu recurvatum deformity, knee flexion contractures, and progressive arthritic worsening of the knee joint are commonly seen. Surgical tendon release or lengthening as well as potential total knee arthroplasty are possible treatment options. Equinus contracture at the ankle, cavus foot deformity and forefoot equinus, and various other foot muscle imbalances may be corrected with Achilles tendon lengthening, plantar fascia releases, as well as hindfoot triple arthrodesis for stability.


Prognosis

Individuals with abortive polio infections recover completely. Many other cases
of polio result in only temporary paralysis. Nerve impulses often return to the formerly paralyzed muscle within 1 month and recovery is usually complete in 6–8 months. Paralysis remaining 1 year after the initial viral infection is likely to be permanent.

In cases of spinal polio, if the affected nerve cells are completely destroyed, paralysis is permanent. The cells that are not destroyed, but lose function temporarily, may recover within a period of several weeks. Half the patients with spinal polio recover fully; one quarter recovers with mild disability, and the remaining quarter have severe disability. Spinal polio is rarely fatal.

Consequences of polio with respiratory involvement include suffocation or pneumonia from aspiration of secretions. It is estimated that 5–10% of patients with paralytic polio die due to paralysis of the muscles of respiration. Bulbar polio can be fatal if respiratory support is not provided, but outcome is improved significantly with respiratory support.

Between 20% and 60% of individuals who survive paralytic polio in childhood develop additional symptoms decades after recovering from the acute infection. PPS is not an infectious process and is usually not life-threatening; however, about 25% of PPS patients may have respiratory complications. The prevention of the late deterioration of polio survivors should be attempted by a comprehensive interdisciplinary approach, including medical, rehabilitation, and surgical treatments, as well as the use of orthotics. Because of the potential for functional decline, PPS individuals should be closely monitored and appropriate interventions instituted at appropriate times to maximize patient function and independence.


PARKINSON’S DISEASE
Unilateral onset of slowly progressive motor symptoms, with persistent asymmetry primarily affecting the side of onset.

Most common motor symptoms are bradykinesia and rigidity (all patients); resting tremor (4–6 Hz) occurs in 60–70% of patients.

Postural instability and gait disorders worsen with disease progression.

Nonmotor symptoms include autonomic dysfunction (especially constipation); sensory symptoms (especially olfactory); sleep disturbance; and, with advanced disease, neuropsychiatric symptoms (depression, dementia).

Clinical course of 10 years or more (sometimes decades).

General Considerations

Parkinson’s disease (PD) is the most common neurodegenerative movement disorder, affecting 1% of the population older than 60 years of age and up to 4% of people aged 80 years or older. The mean age of onset is approximately 60 years; however, in up to 10% of patients the disease appears before the age of 40 years. It is an inexorably progressive disease that usually leads to severe motor disability, often accompanied by a constellation of nonmotor complications. Although symptomatic treatment is highly effective in the early and moderate stages of the disease, severe complications of long-term treatment are common and disabling, and to date there is no curative treatment or effective means to delay the progression of the disease.

Pathogenesis

PD is caused by degeneration of dopaminergic neurons in the substantia nigra, with consequent loss of dopaminergic innervations in the striatum and dysfunction of the cortical–subcortical extrapyramidal motor network. The disease is characterized pathologically by loss of pigmented neurons in the substantia nigra and microscopically by the presence of Lewy bodies,
characteristic intracellular inclusions that are mostly composed of abnormally structured and misfolded α-synuclein, a protein of unknown function expressed mostly in the brain.

Although the etiology of PD is unknown, genetic factors may play an important role in the pathogenesis of the disease. Most patients do not have any known genetic abnormality, but mutations of the SNCA, LRRK2, DJ1, PINK1, and GAD genes and other less common mutations have been associated with increased risk of developing PD, and an estimated 5–10% of cases, especially early-onset cases, are due to genetic mutations. The mechanisms leading to premature neuronal death are unknown, but mitochondrial dysfunction, oxidative stress, abnormal α-synuclein structure and folding, and impaired protein clearance have been implicated in the pathogenesis of the disease.

Exposure to pesticides and heavy metals are among the environmental factors that have been associated with PD, while tobacco smoking appears to have a protective effect.


Clinical Findings

The disease is slowly progressive, manifesting initially with only minimal motor impairment, usually limited to one side of the body, and progressing over years, often decades, to cause severe and global disability. In addition to motor symptoms, virtually all patients with PD experience a number of nonmotor symptoms that also become more severe as the disease progresses. These nonmotor manifestations affect quality of life and at times become the source of additional severe disability. Finally, in the moderate and advanced stages of the disease, many patients experience characteristic motor and nonmotor symptoms that are due to chronic dopaminergic treatment.
A. Symptoms and Signs

1. Motor symptoms—The initial motor symptoms of PD are always unilateral, affecting either side irrespective of the individual’s handedness. Any of the cardinal motor features of PD can present at onset of the disease, and while a significant number of affected individuals will never develop tremor in the course of their disease, every individual with PD will always have bradykinesias and rigidity.

   Tremor is present in 60–70% of patients and is often the first manifestation of the disease. The lack of tremor does not exclude a diagnosis of PD. PD tremor has a low frequency of 3–5 Hz, but in younger individuals the frequency can be higher. It affects the hands and arms, but not infrequently the legs, chin, lips, and tongue. The tremor is most prominent at rest but is also often present while an individual is sustaining certain postures, especially of the arms and hands.

   Bradykinesia refers to the slowing of movements and is often accompanied by akinesia that manifests with paucity of movements and decreased spontaneous motor activity. The slowing is most evident when the patient is asked to perform repetitive tests such as finger tapping or foot tapping, and often increases with repetitions.

   Rigidity refers to hypertonicity that affects both flexors and extensors in the limbs, and the axial muscles in the trunk. During passive limb mobilization the rigidity manifests with a typical catch-and-release quality that is referred to as cogwheeling.

   Gait disorder and abnormal posture can occur in mild stages of the disease and worsen with disease progression. Asymmetric decrease of arm swing and stride asymmetry are present in early stages. Later, the typical shuffling gait becomes apparent, with short steps, initial toe–foot contact, decreased associated hip and trunk movements, and stooped posture. Arm and hand tremor often increase while the individual walks. Turning becomes difficult, requiring multiple steps referred to as en bloc turning. Festination is an unusual gait phenomenon occurring in more advanced stages of disease; it is characterized by involuntary acceleration and inability to stop that often culminates in a fall. Posture is consistently affected by PD, with forward and lateral leaning, usually toward the side more severely affected. An extreme accentuation of postural abnormality that involves severe bending forward (camptocormia) can lead to permanent spinal and trunk deformity. Striatal hand and striatal foot are typical postures of the hands and feet that are also usually more prominent on the more severely affected side.

   Balance impairment and postural instability typically occur later in the
disease, due to abnormal posture with off-center gravity, impairment of postural mechanisms, and abnormal gait dynamics. This leads to falls and, eventually, loss of independent mobility. Falling forward (propulsion) and backward (retropulsion) may occur in response to external forces. Spontaneous loss of balance indicates severe balance disturbance.

Freezing of gait and start hesitation are frequently seen in patients with PD. Freezing of gait refers to sudden gait arrest and “freezing” in place, with inability to move the legs and reinitiate walking that lasts for several seconds. Patients describe a feeling of being glued to the ground, or a magnet that holds them from moving, typically when walking though doorways, elevator doors, or in narrow, crowded spaces that cannot be managed using visual or other sensory tricks (described later, under Exercise Therapy). Start hesitation is the inability to initiate gait after standing that affects many patients and may be physiologically related to freezing.

Other typical motor manifestations of PD include dysarthria and hypomimia. Dysarthria usually manifests with lower voice volume (hypophonia) and monotonous and poorly articulated and differentiated syllables and phonemes. The speech is usually slowed, but occasionally PD dysarthria may manifest with tachyphemia, a paradoxical speech acceleration. Patients with hypomimia, a loss of facial expression and spontaneous facial mobility, have a staring look and decreased spontaneous blinking. The typical “masked facies” appearance leads to a diminished ability to express emotions through facial expression.

2. Nonmotor symptoms—Common nonmotor manifestations of PD include autonomic dysfunction, sleep disorders, neuropsychiatric symptoms and sensory symptoms. As the disease progresses, nonmotor manifestations can become severe and in some cases the major source of disability.

Autonomic dysfunction presents with varying degrees of severity in all PD patients and often worsens with antiparkinsonian pharmacologic treatment. Constipation is nearly universal and can precede the onset of motor symptoms. Orthostatic hypotension is often mild and asymptomatic early in the disease course but can become severe in advanced stages, leading to syncopal episodes. Sexual dysfunction is common, often manifesting early in the disease. Urinary frequency and urgency are very common and can lead to incontinence. Excessive sweating and abnormal thermoregulation are also frequent symptoms.

PD patients often report sensory symptoms, such as body aches, diffuse or localized pain, tingling, and numbness, in the absence of objective sensory deficits. Olfactory dysfunction is common, often precedes the onset of motor
symptoms, and is a potential predictor for PD in otherwise asymptomatic individuals. Blurred vision, intermittent or permanent diplopia, decreased contrast sensitivity and light–darkness adaptation, and other visual disturbances can interfere with daily activities such as reading or driving.

Sleep disturbances such as insomnia with sleep fragmentation and early awakening, vivid dreams, nightmares, and other rapid eye movement (REM) sleep disorders, are experienced by most PD patients. REM sleep disorders often precede the onset of motor symptoms. It has been postulated that REM abnormalities may be used as a presymptomatic marker of PD. REM sleep behavior disorders are characterized by vivid dreams that are partially acted out with talking, screaming, swearing, moving, often with punching or kicking, often with involuntary physical harm to the bedmate. Daytime somnolence, also ubiquitous in PD, may be related to the abnormal sleep pattern or may be an independent nonmotor manifestation of the disease.

Neuropsychiatric disorders are common and can be severe and disabling. Cognitive impairment and dementia such as apathy, executive dysfunction, attention disorders, and other cognitive problems are frequent in PD patients. Dementia may also develop in advanced stages, especially in older patients. The dementia risk varies considerably, with estimates that suggest the majority of older individuals with more advanced disease will develop dementia.

Depression is very common in PD patients and can adversely affect quality of life. The clinical profile of depression in PD is defined by apathy, loss of motivation, and anhedonia. Often, however, sadness, guilt, or melancholia are predominant. Of note, PD patients, while meeting clinical criteria for depression, often do not perceive themselves as depressed. In addition, PD depression is often resistant to pharmacologic treatment. Anxiety is ubiquitous in PD at all stages of the disease, may accompany depression or be an isolated disorder, and may interfere with sleep.


B. Diagnostic Studies

The diagnosis of PD rests on clinical evaluation, nuclear medicine testing, and exclusion of other causes of parkinsonism. Specific clinical criteria are used for a diagnosis of PD and to differentiate the disease from Parkinson-like disorders (or parkinsonism). Both the UK Parkinson’s Disease Society Brain Bank (Table 19–2) and the National Institute of Neurological Disorders and Stroke (NINDS) (Table 19–3) have established precise diagnostic criteria for PD that are considered the standard for an accurate diagnosis.

Table 19–2 UK Parkinson’s Disease Society Brain Bank’s clinical criteria for the diagnosis of probable Parkinson’s disease.
Table 19–3 National Institute of Neurological Disorders and Stroke (NINDS) diagnostic criteria for Parkinson’s disease (PD).

<table>
<thead>
<tr>
<th>Step</th>
<th>Criteria</th>
</tr>
</thead>
</table>
| Step 1 | Bradykinesia  
At least 1 of the following criteria:  
- 4–6 Hz resting tremor  
- Postural instability not caused by primary visual, vestibular, cerebellar, or proprioceptive dysfunction  
- Rigidity |
| Step 2 | Exclude other causes of parkinsonism |
| Step 3 | At least 3 of the following supportive (prospective) criteria:  
- Clinical course of ≥ 10 y  
- Excellent response (70–100%) to levodopa  
- Levodopa response for ≥ 5 y  
- Persistent asymmetry primarily affecting side of onset  
- Progressive disorder  
- Rest tremor  
- Severe levodopa-induced chorea (dyskinesia)  
- Unilateral onset |

| **Group A Features**  
|----------------------|-----------------------------|
| **(Characteristic of PD)** | **Group B Features**  
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Resting tremor</td>
<td>Features unusual early in clinical course</td>
</tr>
<tr>
<td>Bradykinesia</td>
<td>Prominent postural instability in first 3 y after symptom onset</td>
</tr>
<tr>
<td>Rigidity</td>
<td>Freezing phenomenon in first 3 y</td>
</tr>
<tr>
<td>Asymmetric onset</td>
<td>Hallucinations unrelated to medications in first 3 y</td>
</tr>
<tr>
<td></td>
<td>Dementia preceding motor symptoms or in first year</td>
</tr>
<tr>
<td></td>
<td>Supranuclear gaze palsy (other than restriction of upward gaze) or slowing of vertical saccades</td>
</tr>
<tr>
<td></td>
<td>Severe, symptomatic dysautonomia unrelated to medications</td>
</tr>
<tr>
<td></td>
<td>Documentation of condition known to produce parkinsonism and plausibly connected to patient’s symptoms (eg, suitably located focal brain lesions or neuroleptic use within past 6 mo)</td>
</tr>
</tbody>
</table>

**Criteria for definite PD**
- All criteria for probable Parkinson’s are met, and
- Histopathologic confirmation of diagnosis is obtained at autopsy

**Criteria for probable PD**
- At least 3 of the 4 features in group A are present, and
- None of the features in group B is present (note: symptom duration ≥3 y is necessary to meet this requirement), and
- Substantial and sustained response to levodopa or a dopamine agonist has been documented

**Criteria for possible PD**
- At least 2 of the 4 features in group A are present; at least 1 of these is tremor or bradykinesia, and
- Either none of the features in group B is present or symptoms have been present ≥3 y and none of the features in group B is present, and
- Either substantial and sustained response to levodopa or a dopamine agonist has been documented or the patient has not had an adequate trial of levodopa or a dopamine agonist

Magnetic resonance imaging and other imaging studies are required to rule out secondary forms of parkinsonism resulting from vascular or structural lesions affecting the basal ganglia. In 2011 the FDA-approved radio labeled ioflupane (\(^{123}\)I) for single-photon emission CT studies (DaT Scan) to aid in the diagnosis of PD and differentiate PD from other diseases. Ioflupane binds to presynaptic dopamine transport proteins in the striatum. Decreased uptake indicates loss of dopaminergic innervations and can confirm the diagnosis. The scan is costly, performed only in specialized centers, and lacks full diagnostic accuracy, especially in early stages of the disease.


**Differential Diagnosis**

A number of primary and acquired neurologic disorders have symptoms that overlap with PD and should be considered in the differential diagnosis. Many Parkinson-like diseases are collectively referred to as parkinsonism. Among these diseases, progressive supranuclear palsy (PSP), multiple system atrophy (MSA), corticobasal degeneration (CBD), and other less common neurodegenerative diseases have distinct clinical features and, unlike PD, generally do not respond to levodopa.

Parkinson-like features may be prominent in dementing disorders such as frontotemporal dementia. Essential tremor or other tremor syndromes, especially when asymmetric and associated with other neurologic features, may be difficult to differentiate from early PD.

Parkinsonism can also result from vascular disease affecting the basal ganglia; hydrocephalus; exposure to toxins or medications such as neuroleptics, antiemetics, or calcium channel blockers; or structural lesions in the brain affecting the basal ganglia.

In patients with young or early-onset PD, Wilson’s disease and other metabolic disorders should be considered.
Complications

In addition to increased motor disability, autonomic dysfunction, and neuropsychiatric disorders, several other complications can develop in the late stages of PD. In more advanced stages of the disease speech problems with dysarthria, hypophonia, tachyphemia, or other forms of speech disorder become prominent and may compromise the ability to communicate verbally. Dysphagia also becomes clinically significant in the late stages of disease and can lead to aspiration pneumonia. Visual problems with diplopia, loss of depth perception, and accommodation problems are also common in advanced PD. With increased motor disability the risks of falls and fractures also increases. An underlying vitamin D deficiency and lower bone mineral density may contribute to the risk of fractures.


Treatment

Although there is no curative treatment for PD, nor a proven intervention to delay its progression, symptomatic treatment with medications and rehabilitation intervention can be very effective in mitigating the symptoms and allowing years of a relative functional activity and physical independence. In more advanced stages, surgical implantation of brain electrodes (deep brain stimulation) may help restore function in individuals who have developed severe and disabling
A. Pharmacologic Treatment

Numerous drugs are available to treat PD. Different drugs are used at different stages of disease and in different age groups. As the disease progress most patients need to use a combination of multiple drugs and often take medications several times per day. Guidelines for the optimal treatment of patients with PD have been developed by the American Academy of Neurology.

1. Levodopa—The most effective drug for PD is levodopa, which is always used in combination with carbidopa, a peripheral dopa decarboxylase inhibitor, to minimize nausea and other peripheral dopaminergic side effects. It is usually well tolerated and is very effective in controlling many manifestations of the disease, maintaining its efficacy for years. With long-term treatment, most patients develop dyskinesias, wearing-off, and other motor and nonmotor complications that can be disabling and gravely diminish quality of life (see later discussion).

2. Dopamine agonists—These drugs are often used in early stages of PD but are poorly tolerated by older individuals. Several dopamine agonists are available in the United States (pramipexole, ropinirole, rotigotine) and some offer the advantage of being administered once daily. Though not as effective as levodopa, their use is not associated with dyskinesias or significant motor fluctuations. Patients may, however, have significant gastrointestinal, autonomic, and psychiatric side effects.

3. Monoamine oxidase inhibitors—The selective monoamine oxidase type B inhibitors, selegiline and rasagiline, have mild symptomatic benefit and can be used in conjunction with levodopa and other drugs to ameliorate the motor fluctuations of advanced disease. They are well tolerated, but can (especially selegiline) interact with other drugs and foods and cause hypertensive crisis and
other serious medical problems. Rasagiline is a safer drug and there is some evidence that it may help delay the progression of the disease in newly diagnosed patients.

4. **Catechol-O-methyl transferase inhibitors**—Central and peripheral inhibitors of the enzyme catechol-O-methyl transferase (COMT) decrease the enzymatic metabolism of levodopa and dopamine and prolong the duration of its effect, and are therefore used in combination with levodopa in patients who have developed wearing-off and on–off fluctuations (see later discussion). Entacapone is a peripheral COMT inhibitor that is generally well tolerated and often is used in combination formulations with levodopa and carbidopa. Tolcapone is a highly effective central and peripheral COMT inhibitor, but its use is limited by serious and potentially fatal liver disease requiring periodic liver panel monitoring during the initial months of treatment.

5. **Amantadine and anticholinergics**—Amantadine, another well-tolerated drug, has mild antiparkinsonian effects and may be helpful in patients with tremor. It can be used as initial treatment or in combinations with other drugs.

Anticholinergic drugs such as trihexyphenidyl or benztropine have limited efficacy but may be effective in patients with tremor resistant to other drugs. They can cause constipation, urinary retention, confusion, and other side effects and should not be used in older patients.

6. **Other drugs**—**Neuropsychiatric drugs** are commonly used in the treatment of PD. Among these are clonazepam and other benzodiazepines for anxiety and for REM sleep disorder; hypnotic agents, including zolpidem and zaleplon, for insomnia; alertness-promoting agents, such as modafinil and armodafinil, for fatigue and daytime somnolence; cholinesterase inhibitors, such as rivastigmine and donepezil, and glutamate antagonists, such as memantine, for dementia and cognitive problems. *Botulinum toxin* is often used for patients with concomitant dystonia or injected salivary glands to diminish sialorrhea. *Pressor agents* such as fludrocortisone and midodrine are used in patients with more severe orthostatic hypotension.


7. Complications of dopaminergic treatment—

A. **Motor complications**—Most PD patients treated with levodopa for prolonged periods will experience motor response fluctuations, dyskinesias, and other long-term complications. *Dyskinesias* are involuntary hyperkinetic movements, usually choreic or choreoathetotic and, less frequently, dystonia, that manifest as a mild motor overflow during active movements or occur independently from any movements. In a minority of patients dyskinesias can be severe and disabling, interfering with any motor activity and at times causing loss of balance and falls. *Motor fluctuations* are a peculiar pharmacologic effect of long-term levodopa treatment that leads to shorter duration of benefit from each individual dose. As a consequence patients often experience return of symptoms a few hours after they ingested the last dose (“off” status), and, after taking the next dose, return to an increased level of function and well-being (“on” status). This phenomena is called “wearing off.” Over time these fluctuations may become unpredictable with random transitions from “on” to “off” status (so-called on–off fluctuations).

B. **Nonmotor complications**—*Compulsive and obsessive behavior* such as compulsive gambling, shopping, eating, hypersexuality, and other compulsive behavior can complicate treatment with dopamine agonists.

C. **Illusions, hallucinations, and psychosis**—Drug-induced hallucination can
be caused by several dopaminergic drugs and are more common in patients with cognitive dysfunction. Benign illusions and hallucinations are usually visual and transient, and the patient retains insight into the nonreality of the imagined object (often animals or insects). More structured hallucinations with imaginary individuals and multisensory hallucinations indicate a state of toxicity and, if untreated, can lead to a psychotic outbreak.


B. Surgical Treatment
Deep brain stimulation is an established surgical technique that can help improve the symptoms of patients with severe dyskinesias and motor fluctuations for whom pharmacologic intervention has become ineffective. Electrodes are stereotactically implanted bilaterally in the subthalamic nucleus or the internal segment of the globus pallidus and attached to a thoracic-implanted microcurrent generator. Specific stimulation parameters can be set and adjusted using an external device. In addition to the surgical risks, deep brain stimulation has been associated with several cognitive, behavioral, and psychiatric complications, and often patients experience worsening of speech and gait after surgery. These complications are more frequent in older patients and in patients with more advanced PD.


C. Exercise Therapy
The main areas of rehabilitation and exercise methods for individuals with PD
are development of compensatory strategies, motor skill learning, management of secondary sequelae, and education to optimize physical activity and decrease falls.

1. Enhancing compensatory strategies and motor skill learning—The characteristic posturing and gait in PD is a bradykinetic, short-stepped, shuffling, forward-flexed gait with asymmetric arm swing, which usually translates into difficulties with movement sequences such as walking, turning, writing, transfers, and falls. Cognitive strategies can be used to teach individuals to move more easily and maintain postural stability. Compensatory strategies attempt to bypass the defective basal ganglia, and learning strategies encourage improvements through practice. Motor training can often assist patients to move and walk more freely, and with improved balance. Hypokinesia, or reduced movement amplitude and speed, can often be helped with external cueing strategies, such as music, using a rhythmic beat like that provided by a metronome, verbal cueing, or visual cues such as marking lines on the floor. This external auditory, visual, or verbal cueing often results in increased stride length and rate of ambulation. Individuals with PD are taught to try to bypass the defective basal ganglia, and instead use the frontal cortex to regulate size and speed of movements by consciously thinking about the desired movements.

In PD, the ability to move normally is not lost; rather the activation of the motor activity is affected. Other common therapy strategies used include the PD–BIG techniques of thinking big, such as visualizing walking with large, long steps, and mentally rehearsing the movement pattern before performing the activity. Segmentation teaches patients to break down complex motor sequences into pieces and focus on the performance of each individual segment of the activity.

The optimal mix of interventions varies according to the stage of disease, progression, the individual’s physical ability, his or her capacity for learning, and age. In earlier stages, the capacity to learn new motor skills is preserved. In those with moderate to severe disease, teaching of compensatory strategies (eg, task repetition, drilling a given desired movement or action sequence, segmentation, and using external cues and reminders) is encouraged. When patients are more severely affected, have cognitive impairments limiting skill acquisition, or are very old with multiple comorbidities, instruction in compensatory strategies instead of new motor skill learning is recommended.

2. Managing secondary sequelae—Along with teaching compensatory strategies and new motor skill learning, physical impairments that may exist
such as weakness, loss of range of motion, and diminished aerobic capacity are targeted in hopes of improving balance, gait, and functional ability. Individuals with PD demonstrate altered posture, including forward trunk flexion, decreased axial flexibility, decreased muscle strength, and lack of spinal extension range of motion. These structural limitations contribute to loss of postural control, gait impairment, loss of balance, and functional decline. The management of musculoskeletal limitations by using exercises that targets axial flexibility, balance, gait, and overall strength is a vital part of any program.

3. Promoting physical activity and preventing falls—Minimizing the possibility of falls should be a goal in any therapy program for individuals with PD. It has been suggested, that between 50% and 70% of people with PD experience falls over a 12-month period, a figure that is higher than the 30% fall rate reported for community-dwelling older persons. Many falls occur when the individual is attempting multitasking activities or long complex movement sequences, such as turning while walking, walking and carrying objects, or even walking and talking. The episodic inability to initiate or continue stepping (freezing) places an individual with PD at increased risk for falls. Along with the interventions discussed, patients and caregivers should be educated about risk factors for falls as well as how to prevent slips and falls. Assistive walking devices, such as a wheeled walker or light canes that incorporate visual cueing in gait mechanics, may help PD patients to improve gait and decrease fall risk.

Because PD is a chronic, progressive disease, sustained exercise may be necessary to maintain the benefits obtained through a rehabilitation program. The potential neuroprotective effect of vigorous exercise, forced use, and repetitive exercise has also been proposed. Animal studies lend support to this idea. For instance, motorized treadmill running twice daily for 10 days enhanced motor performance and brain neurochemistry in two different rat models of PD. Other studies have suggested a reduced relative risk for developing the disease in those who reported moderate to vigorous exercise participation. At a minimum, incorporation of exercise assists individuals to maintain functional ability. It is suggested that vigorous exercise begin as soon as possible after diagnosis is made, and that it continue throughout the course of the disease, with modifications according to disease progression and exercise ability.

The incorporation of vigorous exercise early in the disease process has been suggested as a potential means to delay symptom onset, retard disease progression, prevent secondary sequelae of normal aging, and improve aerobic capacity. Although recommendations as to specific exercise types, dosages, frequency, and intensities are lacking, an exercise program should incorporate an
individual’s needs, functional limitations, balance impairment, fall history, lifestyle, and personal interests in hopes of ensuring the best possible compliance.

The PD literature has suggested daily to three times weekly exercise for 6- to 12-week periods to improve spinal flexibility, and at least three times weekly exercise for 4 months to improve cardiovascular fitness. For muscular strengthening resistance training, 2–3 days weekly for a minimum of 6 weeks is suggested. Recently, various integrative treatment modalities have been proposed as having a role in PD while promoting health and well-being. Among these modalities, which have shown varying degrees of positive results, are tai chi, Pilates, yoga, body-weight-supported treadmill training, and whole body vibration.

Ahlskog E: Does vigorous exercise have no protective effect in Parkinson’s disease? Neurology 2011;77:288–294.


Prognosis

PD invariably progresses with time. The Hoehn and Yahr scale (Table 19–4) is commonly used to describe the symptoms and estimate the time frame through which the five stages of the disease progress. Untreated, individuals may lose the ability to ambulate, and hence independence in activities of daily living, and remain bedridden. Pharmacotherapy has improved the prognosis with respect to motor symptoms; however, the undesired side effects of levodopa, including dyskinesia, may occur in up to 50% of individuals after 5 years of drug use.
Table 19–4 Hoehn and Yahr Scale for evaluation of Parkinson’s disease.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No signs of disease</td>
</tr>
<tr>
<td>1</td>
<td>Unilateral disease</td>
</tr>
<tr>
<td>1.5</td>
<td>Unilateral plus axial involvement</td>
</tr>
<tr>
<td>2</td>
<td>Bilateral disease</td>
</tr>
<tr>
<td>2.5</td>
<td>Mild bilateral disease with recovery on pull test</td>
</tr>
<tr>
<td>3</td>
<td>Mild to moderate bilateral disease; some postural instability; physically independent</td>
</tr>
<tr>
<td>4</td>
<td>Severe disability; still able to walk or stand unassisted</td>
</tr>
<tr>
<td>5</td>
<td>Wheelchair bound or bedridden unless aided.</td>
</tr>
</tbody>
</table>

Age remains the best predictor of disease progression. The rate of motor decline is often greater in those with less involvement at the time of diagnosis. Disability is initially related to motor symptoms; however, as the disease advances, motor symptoms that do not respond adequately to medication, such as swallowing and speech difficulties, gait and balance problems, and cognitive deficits, increase disability status. In the advanced stages of the disease, patients have worsening autonomic disturbances, sleep problems, mood alterations, and cognitive decline.

Life expectancy in people with PD is reduced. Risk factors for increased mortality include cognitive decline, dementia, old age at onset, a more advanced course, and the presence of swallowing problems. Death from aspiration pneumonia is twice as common in those with PD as in the healthy population.


Pediatric rehabilitation encompasses a wide variety of conditions, from neurologic disorders to musculoskeletal abnormalities that affect growth and development. Developmental delay, specifically in gross and fine motor activity, is a common concern of parents and caregivers; thus an understanding of normal growth and developmental patterns is essential (Table 20–1). Causes of developmental dysfunction in motor and cognitive skills range from congenital and genetic abnormalities to acquired illnesses and injuries of the neurologic and musculoskeletal systems. A thorough history and examination is key to diagnosis and management. Goals for habilitation or rehabilitation will depend on the child’s age and development.

Table 20–1 Pediatric developmental milestones.
|----------|-----------------------------------------------------------------------------|----------------------------------------------------------------------------|--------------------------------------------------------|------------------------------------------------------|----------------------------------------------------------|
| Newborn  | Arms and legs flexed  
Poor head control                                    | Hands fist ed  
Involuntary grasp reflex                                           | Can fixate on a face at 8–15 inches  
Visual acuity 20/400                                    | Startles or widens eyes to sound  
Variation in crying                                       | Fixates on a face in preference to other objects         |
| 2 months | Head lag on pull to sit  
Lifts head in prone  
Head erect when held upright                                            | Grasp reflex disappears  
Hands open and relaxed  
Hands to midline  
Holds objects put in hand                                      | Can track horizontally and vertically                           | Coos and laughs  
Vocalizes with vowel sounds                                  | Social smile  
Responds                                                  |
| 3 months | No head lag on pull to sit  
Can lift chest when prone                                               | Reaches and swipes a toy                                              | Can track a ring in a circular motion  
Stares at own hand                                            | Coos and laughs  
Vocalizes with vowel sounds                                  | Interested in image in mirror  
Smiles; playful  
Laughs at active stimuli                                      |
| 4 months | Rolls over back to side                                                   | Voluntary grasp                                                        | Localizes bull’s eye in near and far position          | Squeals                                               | (+) Gaze monitoring                                         |
| 6 months | Can sit with support  
Rolls over front to back                                      | Raking motion  
Transfers object from hand to hand                               | Can look for a dropp ed spoon  
Pulls a cord to obtain a disc                              | Babble s with consonant sounds  
Turns to orient to name                                       | Basic emotions emerge:  
happiness, interest,  
surprise, fear, anger,  
sadness, and disgust                                         |
| 9 months | Sits without support  
Can sit from supine  
Pulls to stand  
Crawl  
Cruises along furniture                                            | Radial digital grasp  
Immature pincer grasp  
Uses forefinger to poke or roll an object  
| Can look for a hidden object  
Turns cup right side up                                           | Says “mama,” “dada”  
nonspecifically polysyllabic babbling  
(+). Joint attention                                          | (+) Stranger anxiety  
Engages in back-and-forth play and peek-a-boo  
Attachment to preferred caregiver                             |
| 11 months| Stands alone  
Walks with hands held                                               | Puts small objects in a cup                                           | Makes object association                                 | Says “mama,” “dada”  
specifically  
Gives toy with gesture                                        | Shows or offers a toy to adult                               |
| 12 months| Stands alone  
Can take a few steps                                               | Mature pincer grasp  
Turns pages in a book                                          | Demonstrates object permanence  
Attends to a picture                                        | Says at least 1 word clearly  
Can identify objects                                           | Points to an object to obtain it  
Attachment forms  
Symbolic play                                                   |
| 15 months| Begins to walk alone  
Gait is wide-based                                                    | Spontaneous scribbles  
Builds a tower of 2 blocks                                        | Looks for a toy that was displaced  
Can put a circular shape in a puzzle                       | Says 2 words in addition to  
“mama” and “dada”  
Gives a toy on request without gesture  
Combines jargon and gesture                                  | Greets people with “hi”  
Recognizes image of self in mirror                            |
| 18 months| Can climb into an adult chair  
Begins to run  
Walks up stairs with help                                           | Builds a tower of 3 blocks  
Imitates a vertical line                                            | Deferred imitation  
Can put 4 shapes in a formboard                             | Uses > 5 words; follows simple instructions  
Can identify 4 body parts                                     | Points to share an experience  
Uses the word “no”                                               |
## GENERAL CONSIDERATIONS IN PEDIATRIC EVALUATION

In the evaluation of pediatric patients, the importance of the chief complaint and
reason for the visit cannot be overemphasized. Traditionally part of the basic history, this component is sometimes omitted or inferred by time-pressed providers. Of similar importance are the parents’, caregiver’s, or patient’s goals and expectations, which will shape the direction of later care and management.

In addition to obtaining past medical and surgical histories, the examiner should explore the patient’s birth history. Information about prenatal care, maternal illness or conditions, medications, mode of delivery, age of gestation at delivery, and any perinatal and postnatal complications (including treatment course in the neonatal intensive care unit [NICU]) can provide important clues that may help in the differential diagnosis. Queries should be made regarding developmental milestones and functional abilities, including the age at which milestones were achieved, delays, and any skills that were lost or have regressed (see Table 20–1). Relevant information from the family, personal, and social history, including concerns relating to school performance and social interactions, medications and allergies, and immunizations, should be noted.

The physical and neurologic examination of a child, especially an infant or toddler, can be challenging and may be limited by the patient’s inability to cooperate or communicate with the examiner. The examination can be facilitated by developing a rapport with the parent or caregiver and child before touching the child, and, particularly with infants and young children, by incorporating play and use of toys, music, and songs in the assessment. The examination starts by simply observing the child, noting any dysmorphic features or gross abnormalities such as head or facial asymmetry, neck and limb posturing, or limited movements and chest deformities (pectus carinatum or excavatum). Patterns of breathing and the presence of a tracheostomy or a feeding tube should be noted.

In the extremities, the examiner may notice limb-length discrepancy, hemihypertrophy, or uneven skinfolds, especially at the thighs, which may be indicative of hip dysplasia. Active and passive range of motions are tested and any limitation of the joint or contractures, whether fixed or flexible, are noted. Common foot alignment abnormalities with bony deformities include talipes equinovarus (clubfoot), pes planus or pes planovalgus (flat feet), and pes cavus. Skin tags, hypopigmentation or hyperpigmentations, whorl patterns, birth marks, or hirsutism should be noted as these may be associated with a genetic syndrome. Table 20–2 lists distinctive clinical findings associated with common genetic syndromes. In examining the back and spine, the examiner should observe for presence of a sacral dimple or sinus, hair tuft, or abnormal spinal curvatures.
Table 20–2 Clinical characteristics of common genetic abnormalities and syndromes.
<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Genetics/Inheritance Pattern</th>
<th>Clinical Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Down syndrome</td>
<td>Trisomy 21</td>
<td>Hypotonia, flat facies, slanted palpebral fissures, small ears, mental deficiency, endocardial cushion defect (40%), short neck, hyperflexible joints, high risk for C1–2 subluxation</td>
</tr>
<tr>
<td>Edward syndrome</td>
<td>Trisomy 18</td>
<td>Clenched hand with overlapping fingers, short sternum, low-arched dermal ridge, patterning on fingertips, feeble cry, ventricular or atrial septal defect, hypotonia and hypoplasia of skeletal muscles</td>
</tr>
<tr>
<td>Patau syndrome</td>
<td>Trisomy 13</td>
<td>Holoprosencephaly, microcephaly, severe mental retardation, polydactyly, narrow convex fingernails, prominent (rocker-bottom) heel, cleft lip or palate, cardiac abnormality</td>
</tr>
<tr>
<td>Klippel-Feil syndrome</td>
<td>Mutation in GDF6 and GDF3; autosomal dominant</td>
<td>Fusion of any cervical vertebra 2–7, restricted neck range of motion, short neck, low hair line, spina bifida, scoliosis</td>
</tr>
<tr>
<td>Cornelia De Lange syndrome</td>
<td>Autosomal dominant; mutation in Nipped-B homolog</td>
<td>Synophrys (unibrow); hirsutism; thin, downturning upper lip; micromelia or limb deficiency; low-pitched cry; mental retardation</td>
</tr>
<tr>
<td>Turner syndrome</td>
<td>45 XO</td>
<td>Short stature, lymphedema, webbed neck from cystic hygroma in infancy, coarctation of the aorta, bicuspid aortic valve, horseshoe kidney, attention deficit hyperactivity disorder, amenorrhea</td>
</tr>
<tr>
<td>Noonan syndrome</td>
<td>Autosomal dominant</td>
<td>Webbed neck, pectus excavatum, cryptorchidism, pulmonic stenosis, cardiac defects, short stature, scoliosis</td>
</tr>
<tr>
<td>Prader-Willi syndrome</td>
<td>Deletion of paternal chromosome 15</td>
<td>Hypotonia, obesity, small hands and feet, scoliosis, excessive appetite, mental retardation</td>
</tr>
<tr>
<td>Angelman (“happy puppet”) syndrome</td>
<td>Deletion of maternal chromosome 15</td>
<td>“Puppet-like” gait, ataxia, jerky arm movements, paroxysms of laughter, developmental delay, speech impairment</td>
</tr>
<tr>
<td>VATER association</td>
<td>Unknown</td>
<td>Vertebral anomalies, anal atresia, tracheo-esophageal fistula, radial dysplasia, renal anomaly</td>
</tr>
<tr>
<td>Ataxia–telangiectasia syndrome</td>
<td>Autosomal recessive, ATM gene mutation, chromosome 11</td>
<td>Progressive ataxia, telangiectasia, dysarthria, lymphopenia, immune deficit</td>
</tr>
<tr>
<td>Rett syndrome</td>
<td>MeCP2 mutation</td>
<td>Regression of milestones, hand-wringing or handwashing, dystonia, breath-holding, unsteady gait, severe constipation</td>
</tr>
<tr>
<td>Friedreich's ataxia</td>
<td>Autosomal recessive, abnormal frataxin protein (trinucleotide repeat)</td>
<td>Progressive ataxic gait, dysarthria, muscle weakness, decreased proprioception and vibration sense, cardiomyopathy</td>
</tr>
</tbody>
</table>
As part of the neurologic assessment, the child’s level of alertness, attentiveness, and social interaction, including speech/language and communication, are observed with reference to age-appropriate expectations. Cranial nerve function is assessed for deficits and abnormalities. The motor assessment includes evaluation of muscle tone, strength, and the presence of abnormal or involuntary movements. If formal manual muscle testing cannot be accurately performed (eg, in younger patients), a description of movements or ability to perform tasks may suffice. If the patient is able to walk, any gait deviations, abnormal or compensatory mechanisms involving limb posture, trunk alignment, step and stride lengths, base of support, and balance should be noted. Sensory and cerebellar functions are also assessed. Reflex testing includes muscle stretch responses; the presence of primitive or infantile reflexes, and their abnormal persistence, should be noted (Table 20–3).

Table 20–3  Primitive reflexes.

<table>
<thead>
<tr>
<th>Reflex</th>
<th>Appearance</th>
<th>Disappearance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moro</td>
<td>Birth</td>
<td>4 months</td>
</tr>
<tr>
<td>Hand grasp</td>
<td>Birth</td>
<td>3 months</td>
</tr>
<tr>
<td>Atonic neck reflex</td>
<td>2 weeks</td>
<td>6 months</td>
</tr>
<tr>
<td>Head righting</td>
<td>46 months</td>
<td>Persists voluntarily</td>
</tr>
<tr>
<td>Protective equilibrium</td>
<td>4–6 months</td>
<td>Persists voluntarily</td>
</tr>
<tr>
<td>Parachute</td>
<td>8–9 months</td>
<td>Persists voluntarily</td>
</tr>
</tbody>
</table>

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CEREBRAL PALSY
ESSENTIALS OF DIAGNOSIS

- A group of disorders affecting movement and posture, and causing activity limitation.
- Attributed to nonprogressive injury and disturbances in the developing fetal or infant brain.
- Often accompanied by disturbances of sensation, perception, cognition, communication, and behavior; epilepsy; and secondary musculoskeletal problems.

General Considerations

Cerebral palsy is the most common cause of motor disability affecting children. In the United States, the Centers for Disease Control and Prevention estimates that 3.3 per 1000 children have cerebral palsy. This is similar to European data of 1.5–3 per 1000 live births. The incidence has increased over time—a fact that may be attributed to technical and medical advances, which have increased the survival of low-birth-weight and premature infants. Although there is no racial or ethnic predilection, the incidence of cerebral palsy has been correlated with low socioeconomic status.

Pathogenesis

Cerebral palsy is caused by injury or developmental disturbances to the immature brain. While the actual etiology may not always be well understood, certain risk factors may contribute to its occurrence. These include prematurity, infection, inflammation, trauma, and coagulation disorders leading to intrauterine or perinatal strokes.

The most common risk factor for cerebral palsy is prematurity. Infants born at or before 28 weeks of gestation, with low or very low birth weights (1000–1499 g) are at particularly high risk given the potential for multiple medical complications, including intraventricular hemorrhage. Maternal risk factors include infections such as chorioamnionitis, endocrine disorders (especially
thyroid disease), fever during labor, multiple births, and placental insufficiency or abnormality. Intrauterine infections and postnatal sepsis, meningitis and encephalitis, asphyxia, hypoxic ischemic encephalopathy, and toxins are also important causes. Kernicterus (bilirubin deposition at the basal ganglia due to severe jaundice) is rarely encountered in the United States but may be seen in developing nations. Traumatic brain injury, including abusive head trauma ("shaken baby" syndrome), is also a significant cause. Whether structural brain anomalies such as lissencephaly or holoprosencephaly should be classified under cerebral palsy is debated, as both are congenital disorders rather than caused by injury. However, clinical manifestations and management for both conditions are similar to cerebral palsy.

Classification

The classification of cerebral palsy has evolved since the condition first became a focus of medical research by Dr John Little in the 19th century. Historically, cerebral palsy has been grouped into four main motor syndrome categories: spastic, dyskinetic, ataxic–hypotonic, and mixed type. Spasticity, as defined by the Task Force on Childhood Motor Disorders, is hypertonia manifested as resistance to a rapid stretch or change in joint angle. Dystonia is a movement disorder that causes twisting and repetitive, abnormal postures, or both. Although the spastic pattern occurs most commonly, a mix of spasticity and dystonia is also seen. Children whose movements are predominantly dystonic tend to have lesions in the deep brain structures, including the basal ganglia and thalamus. The ataxic–hypotonic form of cerebral palsy is the least common.

Owing to the clinical complexity and diversity of cerebral palsy, there has been a push for development of classification systems that would better reflect the underlying pathology and would improve standardization of treatment. In 1997, Palisano and colleagues established a more reliable quantitative classification of gross motor function that places more focus on what the child is capable of doing with regard to sitting, walking, and wheeled mobility. The Gross Motor Function Classification Scale (GMFCS) is now widely used to help guide intervention and management (Figure 20–1). In 2004, the International Workshop on the Definition and Classification of Cerebral Palsy recommended a more comprehensive approach to the specification of motor abnormalities, including the nature and type of the movement disorder, functional motor abilities, accompanying impairments, anatomic and neuroimaging findings, and causation and timing of injury.
GMFCS Level I
Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.

GMFCS Level II
Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.

GMFCS Level III
Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when travelling long distances and may self-propel for shorter distances.

GMFCS Level IV
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

GMFCS Level IV
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.
Clinical Findings

A. Symptoms and Signs

Numerous illnesses cause motor deficits that can mimic cerebral palsy. Diagnosis is usually made between the ages of 2 and 5 years as notable persistent developmental delay, weakness, abnormal muscle tightness, or the child’s inability to walk prompts further investigation. A detailed history and physical examination are essential in narrowing the diagnosis. Along with the past medical history, the examiner should carefully review the birth history (including NICU course), family history, achievement of developmental milestones, functional impairments, and early hand preference (in children younger than age 2). Parental or caregiver concerns about weakness, abnormal muscle tightness, poor head or neck control, and unusual means of mobility (eg, bunny hopping and combat or commando crawling) should be noted.

A thorough neurologic examination should begin with observation of the child’s movement and posture. Muscle tone is assessed by gently moving the joints through their appropriate range of motion and evaluating the degree of resistance. The Modified Ashworth and Tardieu scales are useful tools in evaluating spasticity. Range of motion of each joint is evaluated for muscle tightness or contractures. Along with hypertonia, children with cerebral palsy may present with loss of selective motor control, muscle weakness, and impaired balance.

1. Altered muscle tone, motor control, and movement—Children with hypotonic cerebral palsy usually have difficulty maintaining a sitting position owing to weakness of the neck and trunk and a frog-leg posturing of the legs, whereas those with spastic quadriplegia have hips that cross at the midline or extensor posturing of the legs. The arms may be in a flexor pattern posture with shoulders adducted and internally rotated, elbows flexed, forearm pronated, and hands fisted. Hemiplegic children show early hand preference and decreased ability for bilateral motor tasks. Walking onset may be delayed but almost all children eventually become ambulatory, developing an abnormal gait pattern similar to that of a patient with stroke, and manifesting a spastic flexor pattern.
synergy of the upper limb and extensor pattern in the lower limb.

Children with diplegic cerebral palsy tend to have better trunk control in sitting than those with quadriplegia, but have impaired dynamic balance and may need support or an assistive device to stand and walk. They tend to ambulate with a scissoring and tiptoe (equinus) pattern because of hip adductor and ankle plantarflexor spasticity. A crouched gait pattern is seen in older children as a result of lower extremity weakness and progressive contractures.

In dystonic or dyskinetic cerebral palsy, muscle tone may be variable or fluctuating and there is sustained muscle contraction, with twisting or repetitive movements usually triggered or pronounced with voluntary effort. Children with pure dystonic cerebral palsy rarely have joint contractures. Some patients may have other types of abnormal movements, such as chorea and athetosis.

2. Altered or persistent reflexes—Muscle stretch reflexes are usually hyperreflexive, and clonus may be elicited, especially at the ankles and knees. Persistence of primitive infantile reflexes (eg, Moro, asymmetric tonic neck reflex, and palmar and plantar grasp) past 6 months of age should raise red flags for cortical developmental abnormality. The emergence of parachute, equilibrium, or righting reflexes may be delayed or impaired, and sometimes these postural or protective reactions fail to develop at all.

3. Secondary musculoskeletal findings—Although the brain disorder or injury is one that is static, secondary musculoskeletal problems are likely to evolve and progress as the child grows. Equinus deformity resulting from spasticity and contracture of the gastrocnemius–soleus complex is a common finding. Equinovarus deformity has the additional varus or inversion component and is caused by tightness of the posterior tibialis muscle. Other foot deformities include equinoplanovalgus and equinocavovarus.

At the knee, flexion contractures occur due to spasticity and tightness of the hamstring muscles. Such contractures may result in a crouch gait pattern among ambulatory patients and, in severe cases, lead to posterior pelvic tilt and decreased lumbar lordosis in the seated position. Patella alta (high-riding patella) can occur due to increased muscle pull of the rectus femoris muscle and may result in a stiff-knee gait, characterized by difficulty in flexing the knee during swing phase. Genu valgum is often associated with significant femoral anteversion and hip internal rotation.

Special attention should be given to evaluating the patient for hip displacement, which is caused by spasticity of the hip adductors and underdevelopment of the acetabulum. Hip subluxation is defined as a migration
percentage of greater than 30% on radiographic examination; this may eventually lead to frank dislocation (Figure 20–2). Limited hip abduction and external rotation, along with a positive Galeazzi’s sign (ie, the weaker leg in hemiplegic patients is shorter) and pain on passive range of motion at the hips should raise suspicion of hip pathology and prompt an imaging study. Up to one third of children with cerebral palsy develop hip displacement, with the severity of displacement directly related to the GMFCS level (most severe in levels IV and V). Windswept deformity occurs when one hip is adducted and internally rotated, while the opposite side is abducted and externally rotated. Hip surveillance is recommended as early as age 2, followed by serial hip radiographs, tone management, and orthopedic intervention as necessary, with the goal of preventing hip dislocation, which can lead to pain, decreased mobility, and difficulty in seating and positioning. Other common rotational limb abnormalities include femoral anteversion and excessive internal tibial torsion.

![Figure 20–2](image)

▲ Figure 20–2 Right hip superolateral dislocation with acetabular dysplasia, and mild left hip subluxation. Bilateral coxa valga deformity can be seen.

Neuromuscular scoliosis is a common problem in children with cerebral
palsy, with a reported incidence of up to 67%. Most cases occur in patients classified as GMFCS levels IV and V. Severe spinal curvatures, including kyphosis, can impede proper positioning and seating, cause skin irritation or breakdown, and compromise respiratory function.

Osteopenia is also common, placing children with cerebral palsy at increased risk for long bone fractures with minimal trauma. No clear guidelines are available with regard to treatment. Most clinicians recommend vitamin D and calcium supplementation while encouraging weight-bearing activities as much as possible for bone health. Bisphosphonate treatments are reserved for patients who have experienced multiple fractures or those with vertebral compression fractures.

4. Other associated findings—

A. **Seizure disorders**—Although seizure occurrence varies among the different cerebral palsy groups, children with hemiplegia or those with severe motor impairments (ie, quadriplegia) often have epilepsy and related disorders.

B. **Sensory problems**—Sensory assessment is difficult, particularly among infants and children with cognitive impairment. Those with hemiplegic cerebral palsy may have concomitant hemisensory loss or impairment to light touch and pain. Visual disturbances and ophthalmologic abnormalities are frequently noted in children with cerebral palsy. Strabismus is the most common finding. Other abnormalities include cortical visual impairment, amblyopia, convergence disorder, refraction errors, and astigmatism. Hearing loss or impairment is relatively rare but may be noted among patients with a concomitant genetic syndrome or abnormality.

C. **Oromotor dysfunction**—Dysarthria, dysphagia, and sialorrhea may lead to communication difficulties, decreased caloric intake, and aspiration pneumonia. A swallow function test is usually warranted to help guide feeding and prevent aspiration (see Chapter 38), and referral to a feeding therapist is recommended for assistance with strategies and oromotor strengthening.

D. **Speech and language problems**—Speech and language delay is also common and tends to be more severe among children with quadriplegic cerebral palsy. Verbal skills may be significantly impaired in a child with normal cognition; thus, other forms of communication (eg, sign language or use of an augmentative communication device) should be explored.
E. COGNITIVE AND BEHAVIORAL ISSUES—Cognition level varies among children with cerebral palsy, but mental retardation is more commonly associated with the spastic quadriplegic pattern. Patients with hemiplegic and diplegic motor dysfunction may have normal intelligence or some degree of learning disability requiring special education services with an individualized education plan (IEP) or resource classes. It is important to note, however, that severity of motor and expressive language impairments does not equate with mental retardation. Cognition level can be normal or even above average, especially in those with lesions involving extrapyramidal structures (eg, ataxic, dystonic, or athetoid), and an accurate assessment of cognitive skills is needed to ensure appropriate educational and vocational support. Psychological, behavioral, and emotional problems include attention deficit/hyperactivity disorder (ADHD), impulsivity, aggression, passivity, and low self-esteem. Some children may have autistic or pervasive disorder characteristics.

B. Imaging Studies

Neuroimaging is important in identifying lesions or abnormalities that support the diagnosis of cerebral palsy, while elucidating the pathophysiology behind the clinical presentation. Magnetic resonance imaging (MRI) is the imaging study of choice as it may help in clarifying the onset or time of injury. Periventricular leukomalacia is a classic finding that commonly occurs as a result of intraventricular hemorrhage in premature infants (Figure 20–3). Among term infants, neuroimaging findings are often normal and no specific lesions may be identified. Infants with intrauterine infarction tend to show focal ischemic brain changes within a single or multiple vascular distributions. Unilateral middle cerebral artery infarction (Figure 20–4) is seen in approximately one third of children with hemiplegic cerebral palsy. A coagulation panel should be obtained to check for a possible hematologic disorder contributing to the occurrence of stroke in a child with cerebral palsy.
▲ Figure 20–3 Magnetic resonance imaging scan of the brain showing periventricular leukomalacia (arrows).
Figure 20–4 Encephalomalacia consistent with remote left middle cerebral artery infarction.

Differential Diagnosis

In making the diagnosis of cerebral palsy it is important to establish, through the history and physical examination, that there has been no regression or loss of previously acquired skills, and that findings do not suggest a progressive or degenerative central nervous disorder. Functional impairments may worsen over time in children with cerebral palsy as a result of secondary musculoskeletal complications from abnormal muscle tone and weakness, but no new neurologic findings should be noted on subsequent examinations. Migrational or developmental anomalies such as holoprosencephaly and lissencephaly (smooth brain) may be detected and often are associated with dysmorphic features or other congenital anomalies. In these cases, genetic testing may be warranted. Important differential diagnoses for patients evincing the spastic diplegic pattern include neuromuscular disorders and myopathy (especially for children with hypotonic-type findings), and Rett syndrome. Hereditary spastic paraparesis and spinal cord lesions such as tumor or tethered cord may also mimic cerebral palsy, and imaging of the spine may be warranted to rule out these conditions.
especially when symptoms seem progressive or the history does not reveal risk factors for cerebral palsy. The workup to identify metabolic causes includes, but is not limited to, a basic metabolic panel; plasma amino acid analysis; arterial acid–base levels; ammonia, lactate, pyruvate, and bilirubin levels; and urinalysis (organic acid analysis). Lumbar puncture to obtain cerebrospinal fluid for evaluation of neurotransmitter disorders or glucose transporter deficiency may be requested, as these conditions are highly associated with seizures and movement disorders.

Treatment

The most favorable habilitation or rehabilitation results are achieved when goals are realistic. Striving to achieve outcomes that are beyond the patient’s reach can ultimately leave both child and family discouraged. Frequent review of the patient’s progress is key, while simultaneously inviting feedback from primary caregivers throughout the rehabilitation process. The rehabilitation goals for a child with cerebral palsy are unique to each patient and ideally should combine the goals of both the caregivers and the rehabilitation team.

A. Physical and Occupational Therapy

There is significant statistical evidence to verify improved functional mobility through muscle strengthening in patients with cerebral palsy whose muscle weakness impaired their functional mobility but whose muscles were still able to generate enough volitional movement to be trained. The National Guideline Clearinghouse has published a best evidence statement on strengthening for individuals with cerebral palsy, aged 4–20 years, who demonstrate muscle weakness. It recommends strength training in small numbers of repetitions until fatigued, with rest periods between exercises. Strength training should be task-specific and tailored to the upper or lower extremities, or both, as indicated. Several techniques can be used to enhance developmental stimulation and promote functional improvements.

Aquatic therapy can be helpful in achieving goals related to increased endurance by forming more efficient movement patterns and assisting in respiration and phonation. Warm water relaxes taut muscles, leading to a decrease in tone and, hence, to the development of more efficient movement patterns. In addition, hydrostatic pressure activates sensory receptors and increases external lung pressure, thereby facilitating breathing and speaking.
Constraint-induced movement therapy (CIMT) and bimanual training may be used to improve upper limb and fine motor function in patients with hemiplegic cerebral palsy. CIMT may be used to help facilitate use and function of the plegic and neglected extremity. Studies comparing CIMT with bimanual training have been inconclusive in determining whether one is more advantageous than the other; improvements in limb use and function, as well as quality of life, are reported with either technique, as well as both in combination. Other therapeutic modalities include neuromuscular electrical stimulation, biofeedback, and therapeutic horseback riding or hippotherapy, and partial body weight–supported treadmill training.

B. Speech and Swallow Therapy

Oral aversion is often the initial problem in feeding, especially among ill infants who have been receiving long-term parenteral or enteral tube feedings. Oromotor stimulation and desensitization techniques can be applied. Children with oromotor weakness and dysphagia should be evaluated by a feeding therapist. A speech and language pathologist can also address issues relating to swallowing, as well as speech and language deficits, including verbal and nonverbal communication. (Chapter 38 covers this evaluation in detail.) With advances in technology, several adaptive computer devices are now being utilized as adjuncts for communication.

C. Nutritional Supplementation

For patients with moderate to severe dysphagia, placement of a nasogastric or gastrostomy tube may be indicated to ensure safe and appropriate nutrition. Gastroesophageal reflux (GER) is highly prevalent, especially among premature infants, and should be properly managed through a combination of positioning and medications to decrease the gastric pH and prevent vomiting. A Nissen fundoplication may be performed upon placement of a gastrostomy tube in children with severe GER.

D. Management of Oromotor Problems

Children with cerebral palsy often have difficulty managing oral secretions due to oromotor weakness and poor swallowing. This may lead to anterior drooling (sialorrhea) or posterior drooling and aspiration. Management options include anticholinergic medications such as glycopyrrolate, scopolamine patch, and atropine drops. Botulinum toxin can also be injected into the salivary glands, and
more definitive surgical intervention can be performed (eg, salivary duct ligation or excision).

E. Casting, Orthotics, and Assistive Devices

1. Serial casting—For a flexible joint contracture, a cast (soft, reinforced soft, or bivalve plaster or fiberglass) is applied for 1–4 weeks and changed every 7–10 days. This cycle continues while the joint range of motion, gait, and other physical findings are reassessed until the ankle or knee range of motion goals are achieved. The goals of this extensive therapy may include increasing the passive range of motion of ankle dorsiflexion or knee extension, or both, in order to improve brace fit, increase range of motion during daily stretching routines, advance overall function, or delay more invasive procedures.

2. Orthoses—Various orthotic devices are commonly recommended for children with cerebral palsy. Goals for application include providing stability, preventing contractures, improving range of motion, improving positioning, and promoting better alignment of the foot–ankle in standing and walking. The most common lower limb orthosis is the ankle–foot orthosis (AFO). This can be solid or articulated depending on the goal and the patient’s level of function. Generally for children with spastic equinus gait but with some ankle dorsiflexion movement, an articulated AFO with a plantar flexion stop can help improve the abnormal gait pattern. For children with a crouched gait, a ground reaction ankle–foot orthosis may help to decrease hyperdorsiflexion at the ankle, providing increased knee extension movement.

   Upper limb orthoses may range from a custom molded resting hand splint to a neoprene sleeve at the wrist or hand to help prevent excessive wrist flexion and support the wrist in extension to facilitate grasp. Dynamic splints provide low-load continuous stretch to a joint and may be used to improve range of motion or to facilitate certain functional tasks.

   Spinal orthoses vary from soft to rigid. The orthosis can provide compression and sensory feedback to facilitate trunk support in children with weak truncal musculature. For those with neuromuscular scoliosis, a properly fitting, supportive, custom seating device is favored over rigid spinal bracing, as the latter has not been proven to be effective in correcting the spinal curvature. Compliance is an issue as the brace requires at least 23 hours per day of wear.)

3. Equipment and assistive or adaptive devices—Options include custom seating devices, such as a medical stroller or wheelchair (manual or power),
ramps, standers, gait trainers and walkers, and crutches (Figure 20–5). Lift systems are offered to families of children who are fully dependent in transfers when the child’s increasing weight makes such transfers difficult. Bath equipment (eg, shower chair, tub bench, and commode) also makes it safer and easier for children to perform or caregivers assist with activities of daily living. Home modifications may be needed to accommodate adaptive equipment, including installation of grab bars that will allow some independence in activities of daily living for household ambulators.

▲ Figure 20–5 Gait trainer and stander.

**F. Spasticity Management**

Spasticity management requires a team approach, which includes the patient (if able), caregiver, and rehabilitation team. Goals may include alleviation of pain, increase in function and mobility, improvement in range and flexibility, and assistance in alleviating caregiver burden. Providers should remember that underlying the abnormal muscle tone is weakness. Several treatment options can be used, singly or in combination, depending on the location, distribution, and severity of spasticity, and how it affects the patient’s function and care.

1. **Pharmacotherapy**—

   A. **Oral medications**—Oral medications are indicated for patients requiring generalized spasticity management. There is evidence supporting short-term
spasticity treatment with diazepam in children with cerebral palsy, and some evidence supporting tizanidine. However, evidence is insufficient to support or refute improved *motor function* using these medications, or dantrolene or baclofen (oral or intrathecal). Table 20–4 lists common oral medications used for spasticity and dystonia management.

Table 20–4 Oral medications used in the treatment of spasticity and dystonia.
<table>
<thead>
<tr>
<th>Medication</th>
<th>Class</th>
<th>Dosage</th>
<th>Formulation⁴</th>
<th>Side-Effects</th>
<th>Precautions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>GABA₂ agonist</td>
<td>Initial: 10–20 mg/day divided 3 times daily; slowly titrate up Children 1–4 y: 2.5–5 mg 2–3 times daily Children 5–12 y: 2.5–10 mg 3 times daily Maximum: 80–120 mg/day</td>
<td>Tablet: 10 mg, 20 mg Oral suspension: 10 mg/mL (compounded)</td>
<td>Drowsiness, CNS depression, fatigue, weakness, constipation, decreased seizure threshold</td>
<td>Do not stop abruptly Hallucination, seizure, pruritus with abrupt withdrawal (especially with intrathecal delivery)</td>
</tr>
<tr>
<td>Diazepam</td>
<td>GABA₂ agonist</td>
<td>0.12–0.8 mg/kg/day in divided doses every 6–8 h</td>
<td>Solution: 1 mg/mL Tablet: 2 mg, 5 mg</td>
<td>Drowsiness, CNS and respiratory depression, hypotension, fatigue, weakness Paradoxical reactions: hyperactive or aggressive behavior, hallucinations</td>
<td>Monitor respiratory status Abrupt withdrawal may cause increased seizure activity</td>
</tr>
<tr>
<td>Tizanidine</td>
<td>a₂-Adrenergic agonist</td>
<td>Initial: 0.5–1 mg 3 times daily; slowly titrate up Maximum: 6 mg/day</td>
<td>Tablet: 2 mg, 4 mg</td>
<td>Drowsiness, fatigue, loss of appetite, nausea or vomiting (or both), nervousness, hallucinations, hypotension</td>
<td>Monitor respiratory status and blood pressure</td>
</tr>
<tr>
<td>Dantrolene sodium</td>
<td>Inhibits calcium release at sarcoplasmic reticulum of skeletal muscles</td>
<td>Initial: 0.5 mg/kg/dose 2 times daily; slowly increase Maximum: 3 mg/kg 4 times daily or 100 mg 4 times daily</td>
<td>Tablet: 25 mg, 50 mg, 100 mg</td>
<td>Lightheadedness, vertigo, weakness, malaise, diarrhea Potential hepatotoxicity</td>
<td>Monitor LFTs closely</td>
</tr>
<tr>
<td>Trihexyphenidyl⁵</td>
<td>Anticholinergic</td>
<td>Initial: 0.05 mg/kg 2 times daily; increase gradually in increments of 0.05 mg/kg/day weekly up to 0.25 mg/kg 3 times daily Or Start with 1–2 mg/day; slowly titrate up to 15 mg/day</td>
<td>Tablet: 2 mg, 5 mg Syrup: 2 mg/5 mL</td>
<td>Drowsiness, dizziness, constipation, urinary retention, flushing, nausea, nervousness, blurred vision, dry mouth</td>
<td>Gradually increase dosage; monitor for anticholinergic side effects</td>
</tr>
<tr>
<td>Clonazepam⁶</td>
<td>Benzodiazepine (GABA agonist)</td>
<td>Initial: 0.01–0.03 mg/kg/day divided 2 or 3 times daily Maintenance: 0.1–0.2 mg/kg/day divided 2 or 3 times daily</td>
<td>Tablet: 0.5 mg, 1 mg, 2 mg Wafer: orally disintegrating tablets 0.125 mg, 0.25 mg, 0.5 mg, 1 mg, 2 mg</td>
<td>Drowsiness, dizziness, ataxia, fatigue, CNS depression</td>
<td>Do not stop abruptly Monitor LFTs and CBC if long-term use</td>
</tr>
<tr>
<td>Carbidoa/Levodopa⁷</td>
<td>Dopamine agonist/precursor</td>
<td>25/100 formulation: may start with ¼ tablet 2 times daily and gradually titrate up to 1 tablet 3 or 4 times daily</td>
<td>Tablet: 10/100 mg, 25/100 mg, 25/250 mg Can be compounded for elixir</td>
<td>Gastrointestinal upset, nausea, vomiting (significant limiting factor), sedation, dyskinesia</td>
<td>May give additional carbidoa, or give with meals to Minimize gastrointestinal symptoms</td>
</tr>
</tbody>
</table>

GABA: γ-aminobutyric acid; CNS: central nervous system; LFTs: liver function tests; CBC: complete blood count.

⁴How supplied in the United States.
⁵Primarily used for dystonia.
B. **Focal treatment: Botulinum toxin**—In children, the recommended dose of botulinum toxin A used for chemodenervation is 16–20 units/kg, which may be repeated every 12 weeks, if needed. Dosing for different muscles to be injected depends on muscle size, severity of spasticity, and goals.

Alcohol blocks such as phenol, 3–6%, are also used for motor point or perineural injections. These are especially helpful when administered for obturator motor branch block to decrease hip adductor spasticity, or, in the musculocutaneous nerve, for upper limb spasticity. Effects are immediate and last longer than botulinum toxin. Side effects include pain over the injection site, dysesthesia, and possible tissue edema and scarring.

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2. **Surgical measures**—

A. **Orthopedic interventions**—General indications for orthopedic surgery include fixed contractures and rotational abnormalities affecting function, joint dislocation with pain, and shoe wear and hygiene problems. Interventions include muscle–tendon lengthening, tendon transfers, osteotomies, and arthrodesis procedures. Muscle–tendon lengthening procedures may include Z-type tendon lengthening or muscle lengthening. Z-type tendon lengthening increases joint range of motion, whereas muscle lengthening results in the muscle recessing closer to the muscle tendon junction, which disrupts Golgi receptors and thus decreases spasticity. The side effects of Z-type tendon lengthening may include tendon overlengthening or a shortened muscle.

   Tendon transfers utilize spastic muscles in order to fix a deformity by reattaching a spastic muscle to a weaker antagonist muscle group. A spastic rectus femoris may be transferred to the hamstrings to correct a stiff knee gait during swing phase. Or, a spastic anterior or posterior tibialis may be transferred to the lateral foot to help correct a varus foot deformity. Because tendon surgeries may need to be repeated, Graham and colleagues recommend waiting until a patient reaches 7–9 years of age before surgically intervening; this is a sufficient time for dystonia to completely appear, and also allows for a comprehensive gait analysis to be performed. A single-event multilevel surgery (SEMLS) may be performed that combines bone surgeries with tendon surgeries (which would otherwise likely need to be repeated if done prior).

   An osteotomy involves cutting bone to improve position and alignment. For the child who has a dysplastic hip with subluxation and coxa valga, a varus derotation osteotomy to the proximal aspect of an anteverted femur can improve hip coverage, and superior placement of muscle and tendons will result in more
power and improved energy conservation. An arthrodesis, or joint fusion, may be performed to correct an alignment deformity, stabilize a joint, or prevent recurrence of contracture.

b. Neurosurgery—Neurosurgical intervention may be performed to improve either focal or multifocal spasticity. Intervention may involve the brain, spinal cord, or peripheral nervous system and focuses mainly on decreasing abnormal muscle tone.

Intrathecal baclofen pump therapy may be considered in the treatment of patients with severe spasticity that is refractory to pharmacologic treatment or those in whom side effects are too great. It has been approved by the U.S. Food and Drug Administration for the treatment of spasticity related to various neurologic disorders, including cerebral palsy. The pump device is available in different sizes and contains a drug supply that may last up to 6 months, depending on the rate of dosing. It is usually inserted subfascially or subcutaneously into the abdomen. The following risks of baclofen pump complications have been reported: pump failure, 0.2%; catheter failure (fractures, kinks, dislodgement), 10%; and infection, less than 10%. Should intrathecal baclofen administration stop suddenly, the patient may suffer acute baclofen withdrawal. This should be treated with swift oral dosing of baclofen and intravenous benzodiazepines while the patient is medically monitored, until the problem is rectified. Symptoms of acute baclofen withdrawal can range from increased muscle stiffness, spasticity, and itching to life-threatening or fatal seizures and rhabdomyolysis from muscle breakdown. Baclofen overdose may likewise occur and can result from computer malfunction or programming errors. Caregivers must therefore have a good understanding of this treatment option and be able to monitor and report any significant change in tone that may warrant investigation and immediate medical intervention. Keeping appointments for dose adjustments and refills is crucial. Pump battery life is usually about 7 years.

Selective dorsal rhizotomy involves decreasing sensory input into the spinal cord by severing a portion of the dorsal or sensory roots intradurally at levels L2–S1. Intraoperative electromyographic monitoring can help identify sensory nerve roots that serve overactive lower motor neurons. The ideal candidate for this procedure is a child between 3 and 6 years of age, having spastic diplegic cerebral palsy, with good selective motor control and normal or near-normal cognition. The presence of significant dystonia and other movement disorders is a contraindication for this procedure. Postoperatively, an intensive rehabilitation program is imperative for strength and gait retraining.
Deep brain stimulation for dystonic cerebral palsy involves the stereotactic implantation of electrodes into the globus pallidus (internus) of the basal ganglia, either unilaterally or bilaterally, along with an implantable, programmable pulse generator. Postoperative programming often requires months to identify the optimal stimulation parameters.

**G. Measurement of Outcomes**

Given the complexity of cerebral palsy as a disorder, and the multimodal treatment that may be needed, goal setting should be clear prior to applying any intervention. The International Classification Model of Functioning, Disability and Health (ICF) put forward by the World Health Organization (WHO) recommends a framework for outcome measurements that focuses on four components: body structure, activity, participation, and environmental factors. For body structure and function, video gait analysis, the Gillette Gait Index, range of motion, and strength assessments can be used. The Gross Motor Function Measure (GMFM) and Functional Mobility Scale can be used to assess activity. The Child Health Questionnaire and quality of life measures touch on environmental and personal factors, addressing the final two components in the WHO framework.

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Cincinnati Children’s Hospital Medical Center: *Evidence-Based Care Guideline for Serial Casting of the Lower Extremity*. Cincinnati Children’s Hospital Medical Center, 2009:1–12.


SPINA BIFIDA

ESSENTIALS OF DIAGNOSIS

► Congenital disorder resulting from incomplete closure of the neural tube.
► Neural tissue may show varying degrees of involvement and resultant neurologic deficits.
► Myelomeningocele is commonly associated with neurosurgical, bladder, bowel, orthopedic, and musculoskeletal complications and cognitive deficits.

General Considerations

*Spina bifida* is a general term referring to a group of developmental defects involving incomplete closure of the neural tube. Neural tube defects (NTDs) stem from aberrations of gastrulation during the first month of gestation, when cells on the dorsal side of the fetus do not fuse anteriorly, leaving an opening within the spinal cord or the brain. As a result, incomplete development of the
spinal cord, brain, and meninges may arise. Spina bifida (Latin for “split spine”) can occur at all levels of the vertebral column but is most common in the lumbar and sacral areas.

Normally, closure of the neural tube occurs by the 26th day of gestation. Various types of NTDs may occur at different time points within the first 1–2 months of pregnancy. Spina bifida cystica, anencephaly, or craniorachischisis typically occur within 3–4 weeks. Spina bifida cystica is the most common and most severe class of NTDs that is compatible with life. Anencephaly is a form of neural tube defect that occurs at the cephalic end, resulting in absence of a major portion of the brain and skull. Craniorachischisis occurs in 10–20% of cases and generally leads to death around the time of birth because the entire brain and spinal cord remain open. Both anencephaly and craniorachischisis are incompatible with life.

A. Classification

There is wide phenotypic variability within the diagnosis of spina bifida, including spina bifida occulta, spina bifida cystica with meningocele, and spina bifida cystica with myelomeningocele (MMC). Another form, termed lipomyelomeningocele, may occur when an underlying MMC is covered with a lipoma (Figure 20–6).
Figure 20–6 Lipomeningocele at L5 and upper sacral level. The lipoma (arrow) tethers the spinal cord and extends through.

Spina bifida occulta is the mildest form and results in a small separation or gap in one or more vertebrae. It is present in up to 40% of Americans, is most common in the lumbar or sacral spine, and may produce only external findings (eg, an abnormal tuft of hair, collection of fat, or a small dimple or birthmark) without neurologic deficits, as neural tissues are not involved. It is often an incidental finding on plain radiographs when investigating low back pain later in life as these patients may be prone to spondylolisthesis or a pars interarticularis defect.

Spina bifida cystica with meningocele is a rare form in which the meninges push out through an opening in the vertebrae. Spinal cord development is often normal, and membranes can be removed with little or no damage to nerve pathways.

Spina bifida cystica with MMC is the most common and most severe form, accounting for 80–90% of all types of spinal dysraphism. In this condition, the spinal canal remains open along several vertebrae in the lumbosacral or thoracic spine. As a result, the spinal cord and meninges protrude at birth, leaving the sac exposed on the infant’s back, with or without a skin covering, increasing the likelihood of infections. In cases of MMC, the level of the lesion often affects function. Neurologic impairment includes paralysis below the level of the lesion (most often the lower limbs), neurogenic bowel and bladder, and spasticity. MMC is also highly associated with hydrocephalus, seizures, and orthopedic and other medical complications. The higher the level of spinal cord involvement seen, the greater is the degree of impairment and risk of associated brain malformations, with correspondingly worse cognitive and motor outcomes.

Diastematomyelia is the longitudinal splitting of the lower spinal cord in association with mesodermal elements such as bones or fibrous bands; this can occur alone or with MMC.

B. Epidemiology

Spina bifida is the most common birth defect affecting the central nervous system, and the most complex birth defect compatible with survival. The incidence of spina bifida is 1–2 per 1000 live births worldwide. It is high among Caucasians of European descent and Hispanics, and lower among Asians and those of Pacific ancestry. Spina bifida is also more common in females. Prevalence is declining in North America and western Europe because of dietary
fortification and advanced prenatal diagnosis, leading to more elective terminations.

C. Etiology and Risk Factors

NTD incidence has been linked with inadequate folic acid in pregnant women. Consequently, it is recommended that women intending to conceive begin low-dose folic acid supplementation, at approximately 0.4 mg/day, at least 3 months before conception and continue through the 12th week of pregnancy. Women who have had a child with an NTD, or who are taking anticonvulsants, required increased supplementation, at around 4–5 mg/day. However, even if every woman of childbearing age took dietary supplements, fortification would prevent only 50–70% of NTDs, a clue that the condition is multifactorial and other risk factors also play a role.

The level of the lesion, ethnicity, and socioeconomic status account for genetic heterogeneity. In addition, a maternal history of diabetes mellitus or high blood glucose levels early in pregnancy, obesity, low socioeconomic class, conception in mid spring, heat exposure, and maternal intake of anticonvulsant drugs (eg, valproic acid or carbamazepine) have been identified as risk factors. There is thought to be a genetic component to NTDs as the recurrence rate for having a second child with spina bifida ranges from 2.4% to 5%, or 1 in 25, and this rate doubles if two previous children have been affected. Similarly, when one parent has spina bifida, there is about a 4% chance of passing the disorder on to the child.

Clinical Findings

The approach to assessment of a newborn or child with spina bifida is similar to that of a patient with spinal cord dysfunction. Neonates and infants are usually not amenable to traditional manual muscle testing; thus, motor examination focuses on observation of posture, spontaneous and reflexive movements, muscle tone, and deep tendon reflexes (Table 20–5). Sensory and rectal examinations are also performed. Assessment also includes joint integrity and range of motion for contractures and deformities, which may be secondary to intra-uterine positioning.
### Table 20–5  Motor findings corresponding to level of lesion in spina bifida.

<table>
<thead>
<tr>
<th>Level of Involvement</th>
<th>Examination Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic</td>
<td>Flaccid with no spontaneous movement, “frog-legged” posture</td>
</tr>
<tr>
<td>High lumbar</td>
<td>Unopposed hip flexion and adduction, usually absent knee extension</td>
</tr>
<tr>
<td>Mid lumbar</td>
<td>Weak quadriceps or knee extension</td>
</tr>
</tbody>
</table>
| Low lumbar           | L4: unopposed ankle dorsiflexion and knee extension, weak gluteals  
                        | L5: greater knee flexion, relatively weak gluteals |
| Sacral               | Foot intrinsic weakness  
                        | Some active plantar flexion |

### A. Symptoms and Signs

Common NTDs are summarized in the section on Classification, earlier. Spinal shock may be present; the motor level may be asymmetric and may not correlate with the vertebral anomaly or lesion site. Common congenital musculoskeletal deformities include hip dysplasia, hip or knee flexion contractures, knee extension contractures, talipes equinovarus (clubfoot), and calcaneovalgus (see later discussion of common lower extremity deformities). The infant’s alertness, visual attentiveness, suck and swallow reflex, and capabilities should be noted, as hydrocephalus causing increased intracranial pressure and brain shift is a common complication even in the newborn period. Respirations may also be abnormal in neonates with brainstem compression. Nutritional intake, suck, and swallow may be impaired, necessitating referral for feeding therapy. Bladder and bowel function should be assessed. Decreased urine output and frequency may indicate a flaccid bladder, and catheterization may be needed to adequately empty the bladder.

### B. Laboratory and Imaging Studies

Diagnosis and treatment of spina bifida often begins before birth, usually between 16 and 18 weeks of gestation. Blood tests may reveal increases in
maternal serum α-fetoprotein (AFP), an enzyme produced by the fetus. Increased AFP may also be present in cases of multiple gestations, anencephaly, fetal death, or omphalocele, making the diagnosis unclear with maternal blood tests alone. A more specific way of measuring AFP is by amniocentesis (amniotic fluid sampling), because it is often sampled with acetylcholine esterase isoenzyme, an enzyme that is increased only in conjunction with spinal bifida. However, amniocentesis can be associated with higher risk of preterm labor or infection; thus, the benefits and risks of the procedure must be considered. High-resolution ultrasonography is scheduled between 16 and 24 weeks of gestation to help confirm the diagnosis.

### Treatment

Management starts prenatally and includes family education and counseling. Cesarean delivery is employed to reduce trauma to the exposed neural sac, ideally in a tertiary center with access to a specialized medical team that is familiar with the diagnosis. Surgical closure is recommended within 24–48 hours after birth to try to preserve existing neurologic function.

Intrauterine surgery for spina bifida may be performed between 19 and 25 weeks in an effort to preserve neuromuscular integrity and minimize iatrogenic damage. This procedure aims to improve neurologic outcome but may be associated with fetal and maternal risks, such as preterm birth, intraoperative complications, or uterine scar defects. The safety and efficacy of this procedure is still being investigated.

Rehabilitation focuses on maximizing functional capabilities, as well as preventing secondary complications such as skin breakdown and musculoskeletal deformities that may later impede function.

### A. Neurosurgical Complications

Chari II malformation occurs in over 90% of people with MMC and involves caudal displacement or herniation of the medulla, lower pons, fourth ventricle, or cerebellar vermis into the cervical canal. Complications stemming from this type of malformation include hydrocephalus, central ventilator dysfunction, stridor, upper airway obstruction, central sleep apnea, or aspiration. Of these, central ventilator dysfunction is the most frequent cause of death within this population.

Defects in myelination, hypoplasia, or aplasia are also frequent and likely congenital in cases of MMC. Dysgenesis of the corpus callosum or forebrain
may affect cognitive functions, behavior, and adaptation, including planning, organizing, initiating, and working memory, and cerebellar dysgenesis can lead to decreased coordination. Language and reading comprehension and math skills may also be poor, making early intervention in schooling important.

Ventriculomegaly is common in people with MMC (70–90%) because of obstruction caused by a Chiari II malformation or aqueductal stenosis. Most rapid progressions occur within the neonatal period; however, surveillance is maintained throughout infancy. Serial head ultrasound or computed tomography (CT) scans, head circumference measurement, and assessment of the anterior fontanelles are performed, and clinical signs indicative of increased intracranial pressure are noted (eg, decreased activity and alertness, poor feeding, irritability, vomiting). Treatment is with ventriculoperitoneal shunt placement. Shunt diversion dates back to the 1970s and has been credited with increasing the survival rate of people with MMC, but there have been concerns about its long-term effects, as shunting may lead to further malfunction and infection. Studies have also shown an increased incidence of cognitive deficits in hydrocephalic individuals with implanted shunts, as well as higher rates of ADHD (31% versus 17%). As a result, many centers only implant shunts when there is significant ventricular dilation and frequently monitor ventricular dilation over time with serial neuroimaging. Increased frequency of shunt malfunctions and infections is also associated with lower cognitive levels.

Tethered cord is a neurologic complication involving abnormal attachment of the spinal cord at its distal end. It often results from the spinal cord lesion itself or from scarring after surgery. Traction and stress from cord tethering can manifest as worsening of extremity spasticity or weakness, an ascending sensory deficit, or paresthesia, change in bowel or bladder habits, back or leg pain, or rapid progression of scoliosis. MRI or myelogram may help to confirm suspicion; however, tethered cord is mainly a clinical diagnosis as opposed to a radiographic abnormality. Treatment consists of surgical detethering of the cord. There is a 10–15% chance of recurrence postoperatively, making close followup crucial.

Syringomyelia is a tubular cavitation resulting from increased cerebrospinal fluid pressure caused by obstruction at the fourth ventricle or the foramen magnum, or both. It is most common at the cervical level and can lead to progressive scoliosis, paralysis (especially of the upper limbs), and impaired pain and temperature sensation. Treatment involves insertion of a syringopleural shunt.
B. Neurogenic Bladder

Spinal dysraphism is the most common cause of neurogenic bladder in newborns and is a significant cause of morbidity and mortality. Over time, patients develop increased bladder pressure, hydronephrosis, and increased risk of infection, leading to renal damage. Surveillance studies including yearly ultrasound of the kidney are recommended to look for hydronephrosis or renal parenchymal damage, which can be caused by vesicoureteral reflux, detrusor sphincter dyssynergia, structural abnormalities of the kidneys, and poor bladder compliance. Urodynamic studies may show different patterns of bladder abnormality indicative of an upper motor neuron lesion, such as uninhibited bladder contractions, decreased bladder compliance, and detrusor sphincter dyssynergia. Bladder areflexia is usually seen among patients with sacral involvement.

The main goal of treatment is to prevent renal damage and achieve social continence. The mainstay of urologic management is clean intermittent catheterization (CIC). For patients with a spastic or an overactive bladder, anticholinergic medications such as oxybutynin may help reduce voiding accidents in between catheterizations. The pediatric dose is 0.2 mg/kg per dose, two to four times daily, for children younger than 5 years of age, and 5 mg, two to three times daily, for those aged 5 years and older. For patients with a flaccid bladder, timed voiding can be tried; however, CIC may be needed when postvoid residuals remain high.

Surgical treatment is indicated for patients with persistent high intravesical pressure or hydronephrosis despite CIC. In the Mitrofanoff procedure with bladder augmentation, a tubularized segment of small bowel (usually the appendix) is used to create a conduit that can be catheterized through a stoma in the abdominal wall. Compliance with good bladder care is key to prevention of renal complications; thus, education and training of both caregivers and children is started as early as possible.

C. Neurogenic Bowel

Constipation is a major problem in patients with NTDs. Bowel incontinence occurs as a result of absent sensation of rectal fullness, lack of sphincteric control, or dyssynergia of intestinal peristalsis. The goal is to achieve regularity and continence through scheduled elimination, and avoid constipation. For patients with intact sphincteric tone, rectal stimulation with suppositories may be adequate. Those with a lower motor neuron lesion who lack sphincteric tone and have decreased rectal motility can benefit from a combination of high-fiber diet,
laxatives, stool softeners, cathartics, and enemas. The antegrade continence enema is a surgical procedure that is indicated for chronic constipation refractory to medical management. A continent diversion is made proximal to the anus, connecting the appendix or a small piece of the intestine to the abdominal wall, where a stoma is created. A calculated volume of enema (normal saline solution or tap water) is flowed through the stoma to flush the colon; stools can be eliminated in about 30–60 minutes. With this procedure and the Mitrofanoff procedure, described earlier, children with spina bifida can achieve greater independence in their bowel and bladder programs.

**D. Functional and Musculoskeletal Complications**

The level of the lesion in the spinal cord (ie, thoracic, lumbar, or sacral) affects mobility and function. Although walking is generally desired, it may not be the most efficient form of mobility. Ongoing discussions should be maintained with the parents or caregiver and patient regarding the child’s functional ability in relation to realistic goals for mobility and activities of daily living, taking into consideration the child’s age and developmental ability. Goals should be reviewed periodically as these may change as the child grows older.

Physical and occupational therapy are mainstays of treatment that help improve functional mobility and the ability to perform activities of daily living. Strengthening and stretching exercises, different types of orthotics, and assistive devices can be used to help aid ambulation, protect joints, and prevent further deformities and contractures. Hip, knee, and ankle pain are common problems that arise from muscle imbalance and joint malalignment in standing and walking. A gait analysis is essential to fully assess gait abnormalities and will help guide orthotic prescription and orthopedic management.

Children with spina bifida are at increased risk for development of joint contractures and bony deformities. Table 20–6 lists orthopedic and musculoskeletal abnormalities commonly associated with lesions at various neurologic levels. Children with lesions at the thoracic or high lumbar level are paraplegic and will not be ambulators. Upright standing can be facilitated with a framed orthosis, such as a parapodium or a regular stander. A swivel walker or reciprocating gait orthosis with crutches or a walker can promote therapeutic or household ambulation. Independent manual wheelchair propulsion can be achieved in children as young as 24 months. Motorized wheelchairs are useful for longer distance mobility or for individuals who are unable to self-propel their manual wheelchairs.
Table 20–6 Treatment options for common orthopedic and musculoskeletal impairments in children with neural tube defects.\textsuperscript{a}

<table>
<thead>
<tr>
<th>Level of Lesion</th>
<th>Mobility</th>
<th>Orthotic and Assistive Device Options</th>
<th>Common Orthopedic Problems</th>
<th>Treatment and Goals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic, high lumbar</td>
<td>Household or therapeutic ambulation Mainly wheelchair mobility</td>
<td>RGO, HKAFO, KAFO, plus walker or crutches</td>
<td>Hip subluxation or dislocation</td>
<td>Surgical correction to keep hip in place (if functionally needed), for pain relief, and to promote better sitting tolerance. Appropriate seating system; TLSO.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Kyphosis or scoliosis</td>
<td>Surgical correction for rapidly progressive scoliosis (for proper seating), to prevent pulmonary compromise and further deformity.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Hip and knee flexion contracture</td>
<td>Soft tissue release to promote proper seating or brace wear, or promote ambulation.</td>
</tr>
<tr>
<td>Mid lumbar</td>
<td>Limited community ambulation</td>
<td>KAFO, AFO, GRAFO, plus walker or crutches</td>
<td>Equinus, equinovarus deformity</td>
<td>Soft tissue release, osteotomy.</td>
</tr>
<tr>
<td>Low lumbar</td>
<td>Independent Community ambulation</td>
<td>AFO</td>
<td>Calcaneal deformity (due to excessive dorsiflexion)</td>
<td>Soft tissue release, osteotomy.</td>
</tr>
<tr>
<td>Sacral</td>
<td>Independent community ambulation</td>
<td>None</td>
<td>Cavus foot</td>
<td></td>
</tr>
</tbody>
</table>


\textsuperscript{a} Impairments are not exclusive to lesion level.

**E. Other Medical Issues**

Insensate skin is a pervasive problem that can lead to formation of pressure sores, chronic wounds from repetitive trauma, and infections. Strategies for preventing skin breakdown include daily inspection of skin; avoiding tight
clothing and ill-fitting orthotics or footwear; maintaining bladder and bowel continence; proper positioning, transfer technique, and pressure relief; and appropriate padding on equipment.

Over 70% of people with spina bifida develop an allergy to latex that can be mild, such as a skin rash, or severe and life threatening. Risk factors for latex allergy include a history of multiple surgeries (ie, multiple exposures to latex), and more than half of individuals with MMC and latex allergy have a history of more than one surgery. Thus, latex-free medical supplies and equipment should be utilized when providing care for patients with spina bifida.

Obesity is prevalent among patients, especially those who are nonambulatory, and may become a cause for concern during growth spurts and puberty. Excess weight has a negative impact on mobility as children age, and also puts them at risk for cardiovascular disease. Dietary modifications and incorporation of some form of physical activity are highly encouraged. Osteopenia leads to an increased risk of fractures in this population. There are no clear guidelines as to treatment and prophylaxis; common practices include calcium and vitamin D supplementation.

Patients with spina bifida may have cognitive deficits, with mean intelligence quotient (IQ) levels in the low average range. Factors that correlate with better intellectual functioning include a lower lesion level, absence of hydrocephalus, and decreased number of shunt revisions. Verbal IQ is usually better than performance or visual–perception. Children are sometimes referred as having a “cocktail personality”; that is, a child may appear to be chatty and argumentative, with an average IQ, but may be failing in school due to impairment in higher cognitive functions, such as conceptual reasoning, problem solving, and mental flexibility. Neuropsychological testing is important to properly identify the need for school accommodations, life skills classes, counseling, and cognitive remediation.


COMMON PEDIATRIC NEUROMUSCULAR DISEASES

1. Duchenne & Becker Muscular Dystrophies

ESSENTIALS OF DIAGNOSIS

- Muscle weakness (proximal greater than distal limbs), Gower’s maneuver, calf pseudohypertrophy, and neck flexor weakness.
- Markedly elevated serum creatinine kinase values, peaking by age 2 in Duchenne muscular dystrophy (DMD), which are usually 10–20 times the upper limit of normal or higher; increased aldolase levels may also be present.
- Myopathic changes (usually small polyphasic potentials) noted on electromyography.
- Muscle biopsy results revealing total absence of dystrophin in DMD or reduced dystrophin in Becker muscular dystrophy (BMD).
- Disease-causing mutation of the dystrophin gene noted on molecular genetic testing.
General Considerations

The muscular dystrophies are an inherited group of muscle disorders characterized by progressive muscular weakness. Both Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) are caused by mutations in the dystrophin gene, and as such are classified as dystrophinopathies. (For additional information about the dystrophinopathies, see Chapter 18.) DMD is the most common, and most severe, muscular dystrophy of childhood, and usually progresses quickly. BMD is clinically more heterogeneous, with a later onset and a slower, milder course. Patients with an intermediate phenotype may be classified as “outliers,” having either mild DMD or severe BMD.

DMD and BMD have an X-linked recessive inheritance, affecting only males and producing asymptomatic carriers in 50% of female offspring. DMD occurs in about 1 in 3500 male births; BMD is much less common, affecting 1 in 30,000 to 5 in 100,000 male births. Patients with clinical features suggestive of DMD or BMD, but without a clear X-linked pattern of inheritance, may have defects in other genes, including those encoding the dystrophin-associated glycoproteins.

Both DMD and BMD are caused by mutations in the dystrophin gene located on the X chromosome at Xp21.2. The majority of mutations are deletions, although point mutations and partial gene duplications have also been reported in a small proportion of affected individuals. The dystrophin protein molecule is a mechanical plasma membrane stabilizer located on the sarcolemma of skeletal and cardiac muscle cells that prevents protease degradation of the muscle fiber. Phenotypic differences among the dystrophinopathies are related to whether the reading frame for dystrophin is disrupted (as in DMD) or preserved (as in BMD).

Clinical Findings

The primary symptom in both DMD and BMD is generalized muscular weakness, as muscle fiber degeneration is the primary pathologic process. Patients with dystrophinopathies are initially ambulatory, but tend to have frequent falls as well as difficulty with running, hopping, jumping, climbing stairs, and getting up from a lying position. Clinical symptoms are similar to those seen in patients with myopathy and include fatigue and weakness affecting the proximal before distal limb muscles, and the lower before the upper extremities. Table 20–7 contrasts these and other relevant features used to
distinguish among the dystrophinopathies. Because of the genetic inheritance pattern, only boys manifest the classic symptoms of the disease. However, cases have been reported in which carrier girls manifested clinical findings to a milder degree.

Table 20–7 Key features in Duchenne, intermediate, and Becker muscular dystrophies.

<table>
<thead>
<tr>
<th>Type</th>
<th>Clinical Presentation</th>
<th>Laboratory Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duchenne muscular dystrophy (DMD)</td>
<td>Onset: age 2–5 years</td>
<td>Severe dystrophic changes</td>
</tr>
<tr>
<td></td>
<td>Pseudohypertrophy</td>
<td>Absence of dystrophin by immunochemistry</td>
</tr>
<tr>
<td></td>
<td>Diminished IQ</td>
<td>Dystrophin quantity: 0–5% of normal by</td>
</tr>
<tr>
<td></td>
<td>Cardiac involvement</td>
<td>Western blot</td>
</tr>
<tr>
<td></td>
<td>Wheelchair confinement: age 10–12 y</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Death: age 15–30 y</td>
<td></td>
</tr>
<tr>
<td>Intermediate</td>
<td>Intermediate severity</td>
<td>Dystrophin: 5–20% of normal</td>
</tr>
<tr>
<td></td>
<td>Wheelchair confinement: age 12–16 y</td>
<td></td>
</tr>
<tr>
<td>Becker muscular dystrophy (BMD)</td>
<td>Onset: variable</td>
<td>Less marked changes, normal appearance or</td>
</tr>
<tr>
<td></td>
<td>More benign course</td>
<td>reduced intensity on immunochemistry</td>
</tr>
<tr>
<td></td>
<td>Earlier, more severe cardiac involvement</td>
<td>Dystrophin: &gt; 20%</td>
</tr>
<tr>
<td></td>
<td>Wheelchair confinement: after age 16 y</td>
<td></td>
</tr>
</tbody>
</table>

A. Duchenne Muscular Dystrophy

Boys with DMD usually present with noticeable weakness and gait abnormality at age 2–3 years, although there is usually a history of some gross motor delay within in the first 2 years of life. Mean age of walking is about 18 months. Affected children have short stature, and by age 3–6 years exhibit the typical
ambulation pattern of exaggerated lumbar lordosis, broad-based gait, and toewalking. Walking ability progressively declines, leading to eventual wheelchair dependence by age 12. Physical examination reveals pseudohypertrophy of the calf (caused by replacement of muscle fibers with fatty tissue), exaggerated lumbar lordosis, a waddling gait, shortening of the Achilles tendons, and hyporeflexia or areflexia. Neck flexor weakness is a unique finding seen among DMD patients.

When arising from the floor, an affected child may roll onto the hands and knees in a prone position, extend the knees to raise the buttocks upward, and then “walk” the hands upward along the legs, pushing the torso upright with the arms, an action referred to as Gower’s maneuver. Clinicians should keep in mind that neither Gower’s maneuver nor lower extremity muscle pseudohypertrophy are pathognomonic of DMD, as these may also be present in other neuromuscular diseases. Musculoskeletal complications such as joint contractures and neuromuscular scoliosis are common especially as disease progresses.

Because the dystrophin isoform is expressed at some synapses in the brain, mild cognitive impairment of varying degrees is common, and younger children commonly have relatively decreased verbal skills. An occasional child may have average or above-average intelligence.

Patients with DMD usually die in their late teens or 20s from respiratory complications or cardiomyopathy. However, survival and quality of life in DMD have improved due to advances in respiratory care, including use of assisted ventilation, and better cardiac monitoring and management.

B. Becker Muscular Dystrophy

Children with BMD often present later than those with DMD, and may have near-normal lifespans. Mental retardation and contractures are not as common or severe, and there is relative preservation of neck flexor muscle strength. Muscle involvement is less severe and scoliosis is less common than in DMD. However, cardiac involvement is often more evident in children with BMD.

The onset of clinical symptoms ranges from 5 to 25 years of age, and patients with BMD typically remain ambulatory beyond age 16 and into adult life. There are two general patterns of BMD: the first is more severe, leading to wheelchair dependence by 25 years of age; the second is characterized by a milder, typically slow course and eventual wheelchair dependence in the fifth decade. Patients with BMD usually survive beyond the age of 30 years and have a mean age of death in the mid-40s, most commonly from heart failure or other
cardiac complications of dilated cardiomyopathy. Phenotypic “outliers” who have a mild DMD or severe BMD presentation are generally confined to wheelchairs between the ages of 12 and 16 years. Mean age and cause of death are the same as for BMD.

**Differential Diagnosis**

The diagnosis of DMD or BMD may be excluded in practically all cases if the dystrophin is normal in size and amount. Other muscular dystrophies to consider in the differential diagnosis include Emery-Dreifuss muscular dystrophy, limb-girdle muscular dystrophies, congenital muscular dystrophies, fascioscapulohumeral muscular dystrophy, myotonic dystrophy, and distal dystrophies. These are discussed elsewhere in this book (see Chapter 18 and Table 18–2). In addition to neuromuscular disorders, metabolic and multisystem diseases, such as disorders of glycogen metabolism, primary carnitine deficiencies, and mitochondrial myopathies, among many others, should be included in the differential diagnosis.

**Complications**

In addition to muscle weakness, cardiac, pulmonary, and orthopedic complications are frequently associated with DMD and BMD. The anticipation and early detection of organ involvement is important for optimal management. Pneumococcal and annual influenza vaccines are recommended for all patients 6 months of age and older, including (and especially) those with dystrophinopathy.

**A. Cardiac**

Dystrophin is present in myocardium and Purkinje fibers; hence, patients with DMD and BMD are predisposed to primary dilated cardiomyopathy. Electrocardiographic abnormalities are seen in nearly all DMD patients older than 13 years of age. The disease process in the heart is underway long before musculoskeletal symptoms appear; therefore, cardiac care should begin after the diagnosis of DMD and BMD. The incidence of symptomatic cardiomyopathy in DMD increases gradually in the teenage years; however, the majority of boys with DMD are relatively asymptomatic until late in the disease course. Early manifestations of heart failure in DMD often go unrecognized secondary to lack of classic signs and symptoms, physical inactivity, and musculoskeletal
limitations. Weight loss, cough, nausea and vomiting, orthopnea, and increased fatigue with a decreased ability to tolerate routine daily activities should be investigated. In contrast, patients with BMD usually manifest clinical signs of heart failure earlier in the course of the cardiomyopathy. In BMD, the development of cardiomyopathy carries a poor prognosis.

As the disease progresses, fibrosis can spread to the lateral free wall of the left ventricle. Echocardiography remains the standard noninvasive diagnostic modality for cardiomyopathy but is often limited in patients with DMD and BMD by scoliosis and poor echocardiographic acoustic windows. Heart failure and arrhythmias may develop in the late stages of the disease. Angiotensin-converting enzyme inhibitors, diuretics, and β blockers may be used to treat left ventricular dysfunction and overt heart failure but do not slow progression of heart disease. Individuals undergoing treatment with glucocorticoids warrant increased cardiac surveillance with specific monitoring for weight gain and hypertension. Complete cardiac evaluation of female carriers should begin after the teenage years and continue every 5 years thereafter. Death due to heart failure occurs in up to 40–50% of patients. In DMD, the rate of sudden death due to arrhythmias is about 5%.

B. Pulmonary

Pulmonary complications are leading causes of mortality in childhood neuromuscular diseases. Progressive muscle weakness leads to restrictive lung disease, hypoventilation, hypercarbia, and respiratory failure. Patients with decreased forced vital capacity (FVC) and peak cough flow can be supported using noninvasive ventilation to aid the inspiratory muscles and assisted-cough techniques to aid the expiratory muscles. FVC volumes show a linear decline between 10 and 20 years of age. Pulmonary referral and baseline pulmonary function tests should begin at age 10 years or prior to wheelchair confinement, and pulmonary evaluations should be monitored every 6 months after any one of the following occurs: wheelchair confinement, vital capacity less than 80% of predicted, or age 12 years. Respiratory management progresses in a stepwise approach based on ventilation parameters such as vital capacity, peak cough flow, oxygen saturation, and end-tidal CO$_2$. Deep lung inflation using a mechanical insufflation–exsufflation device is useful when vital capacity is decreased (< 40% of predicted).

Manual and mechanically assisted cough techniques are useful when respiratory infection is present and baseline peak cough flow (or vital capacity) is decreased. Nocturnal ventilation becomes necessary when patients exhibit
hypoventilation (patients with a vital capacity < 30% of predicted are at especially high risk). Optimally, the use of lung volume recruitment and assisted-cough techniques should always precede initiation of noninvasive ventilation. Daytime ventilation via continuous noninvasive assisted ventilation is indicated for extension of nocturnal ventilation into waking hours, abnormal deglutition due to dyspnea, inability to speak a full sentence without breathlessness, or symptoms of hypoventilation with a baseline pulse oximetry reading of less than 95% or blood or end-tidal CO$_2$ greater than 45 mm Hg while awake.

The final step is tracheostomy, as clinically indicated, for long-term use of noninvasive ventilation in patients who are unable to use noninvasive ventilation successfully, who have had repeated failed extubations during critical illness, or who need frequent direct tracheal suctioning via tracheostomy. Death from pulmonary failure occurs in the second or third decade of life in approximately 90% of affected individuals. The remaining 10% of deaths result from myocardial disease and its sequelae. Over the past 20 years, respiratory care for this latter group of patients has improved as a result of the development of supportive equipment and techniques. Consequently, dilated cardiomyopathy is increasing as the leading cause of death.

C. Musculoskeletal
Patients with DMD have risk factors for poor bone health due to decreased mobility, muscle weakness, and glucocorticoid therapy, which makes them prone to osteopenia, osteoporosis, kyphoscoliosis, and pathologic fractures. Fractures most commonly involve the arms and legs from falls, but may also involve the vertebrae. Long bone fracture occurs in about 20–40% of patients not receiving steroid treatment, most frequently between ages 8 and 11 years. The decline in proximal femur Z-score begins while patients are ambulatory and progresses rapidly when wheelchair-bound (Z-score, −1.7 to −3.9). Healing is unimpaired, but remobilization is essential.

Bone health may be assessed with serum tests (calcium, phosphate, alkaline phosphatase, and 25-hydroxyvitamin D) yearly or biannually. Additional measures include magnesium and parathyroid hormone levels; urine levels of calcium, sodium, and creatinine; and bone mineral density evaluation using dual-energy X-ray absorptiometry (DEXA) scanning (with a baseline reading obtained at age 3 or the start of glucocorticoid therapy). These tests should be repeated annually for patients at risk, especially those with a history of fractures, long-term glucocorticoid therapy, or a DEXA Z-score of less than −2. If kyphoscoliosis or back pain is noted, a spinal radiograph should be obtained to
assess for vertebral compression fracture. Bone age should be assessed (left wrist) using radiography in patients with growth failure (ie, height below the fifth percentile or faltering linear growth), whether on or off glucocorticoid therapy.

Vitamin D should be supplemented at serum concentrations below 20 ng/mL and considered in all children if levels cannot be maintained. Calcium intake and possible supplementation should be carried out in consultation with a dietician. Intravenous bisphosphonates are indicated for vertebral fracture. Oral bisphosphonates are controversial as a treatment or prophylactic measure. If possible, patients should perform standing and weight-bearing exercise, ideally for at least 30 minutes each day.

D. Nutrition and Weight Issues

Patients with DMD can be at risk of malnutrition or weight abnormalities, including being underweight or overweight for age. Weight should be monitored and controlled to avoid obesity. Obesity is most common between ages 9 and 13 years, and becomes problematic as activity decreases. Significant weight loss may occur at ages 17–21. Gastrointestinal dysfunction may be present as dystrophin is also expressed in smooth muscle. Symptoms may include dysphagia, heartburn, abdominal pain, gastroparesis, constipation, and pseudo-obstruction. Clinicians should maintain a low threshold for ordering nutritional and dietetic assessment at key time points in the treatment course (eg, time of diagnosis, initiation of glucocorticoids) and in any of the following scenarios: a patient who is underweight (< 10th percentile for age) or at risk of becoming overweight (> 95th percentile), unintentional weight loss or gain, planned major surgery, chronic constipation, or dysphagia.

E. Scoliosis

A progressive neuromuscular scoliosis develops in most children with DMD, although the prevalence of scoliosis varies from 33% to 100%. Among patients with DMD, 15% acquire scoliosis early, before wheelchair use, while the majority acquire scoliosis between 12 and 15 years of age. Ten to 15% of patients present with mild or no curve. Untreated, scoliosis (Cobb angles) can progress from 11 to 42 degrees per year; such progression is associated with hypoventilation and respiratory complications. Predicted FVC declines an average of 5% per year in nonsurgically treated patients. For every 10 degrees of thoracic scoliosis, vital capacity decreases about 4%.
**Treatment**

There is no cure for DMD and BMD. Glucocorticoids are the mainstay of treatment in DMD and should be offered to patients as an option. Steroids stabilize muscle strength and function in the short term and prolong ambulation ability for about 6 months to 2 years, although the overall long-term course is unchanged.

**A. Pharmacotherapy**

Prednisolone is administered at a dosage of 0.75 mg/kg per day, given daily or pulsed, for boys over the age of 5 years with DMD. Common side effects are weight gain, cushingoid facial appearance, short stature, decrease in linear growth, excessive hair growth, behavior changes, and osteoporosis, leading to increased risk of vertebral compression fractures and long bone fractures. Deflazacort is an oxazolone derivative of prednisone that has shown positive results when administered at a dosage of 0.9 mg/kg per day. Side effects are similar to prednisone, with a higher incidence of cataracts and short stature, but a lower incidence of weight gain.

The precise mechanism by which glucocorticoids act in muscular dystrophy is unknown; the drugs do not affect the dystrophin level but are believed to inhibit muscle proteolysis and help stabilize muscle fiber membranes. Although the dystrophinopathies are not primarily autoimmune diseases, available evidence suggests that humoral and cellular immune responses may contribute to the pathologic disease process. The long-term functional benefit of steroids in muscular dystrophy remains unclear, as does the choice of glucocorticoid, optimal dosage regimens, optimal age of administration, and timing of initiation and discontinuation of treatment. Often, emergence of adverse effects triggers discontinuation of treatment.

On the horizon are several investigational treatments for DMD, which include gene and cell therapy.

**B. Rehabilitation Management**

Therapeutic interventions in DMD and BMD are specifically aimed at maintaining function (particularly ambulation), slowing mechanical pulmonary decline, preventing contractures, and providing psychological support. Timed motor performance is used to predict time to wheelchair dependence by assessing speed of ambulation over a distance of 30 feet (eg, timed ambulation < 9 seconds is predictive of > 2 years to wheelchair dependence; > 12 seconds...
predicts < 1 year to wheelchair dependence). Decreased strength of hip extensors and ankle dorsiflexors is also predictive of cessation of ambulation. At this stage of disease progression, mild to moderate level of physical activity, appropriate orthotics prescription, clinically appropriate surgical interventions, proper seating, and wheelchair components become key considerations.

1. **Physical therapy and exercise**—Patients should receive physical therapy to encourage mobility and to prevent or reduce the risk of contractures. The mainstays of physical therapy are passive stretching exercises to prevent contractures of the iliotibial band, Achilles tendons, and hip flexors. All patients with DMD who are ambulatory or in the early nonambulatory stage should participate in regular submaximal (ie, gentle) exercise to avoid disuse muscle atrophy and other complications of inactivity. The role of exercise is somewhat controversial as a balance needs to be struck between preventing disuse atrophy and avoiding overwork weakness. No head-to-head studies currently exist, but experts recommend a combination of swimming-pool and recreation-based exercises for maintaining aerobic conditioning and pulmonary function. This should be carried out earlier in the course of DMD when greater numbers of relatively normal fibers are present. Exercise may be continued in the nonambulatory phase if medically safe. Low-resistance strength training is recommended at submaximal intensity, depending on the patient’s strength and tolerance. Eccentric (muscle lengthening) exercises should be avoided.

2. **Orthotics**—The functional stages of DMD are independent ambulation, assisted ambulation, and wheelchair mobility. Among muscles of the lower limbs, hip extensors are the first to demonstrate critical weakness. Weak hip extension leads to anterior pelvic tilt with compensatory lumbar lordosis (to move the center of gravity posterior to hip) and resultant shortening of the tensor fascia lata. Weak knee extensors lead to active equinus (tiptoe) posturing of the feet and ankles to maintain passive knee stability and keep the weight line anterior to the knees. Hip abduction develops to widen the base of support, but these muscles are also weakened, causing further shortening of the tensor fascia lata and iliotibial band; hence, patients develop a lateral pelvic tilt and Trendelenburg pattern of gait, furthering instability. During this stage, periods of prolonged immobilization should be avoided as it may cause disuse atrophy and hasten deterioration of the ability to walk. Contractures develop most commonly in the iliotibial band, Achilles tendon, hip, and knee, and later at the elbow and wrist. Contractures progress rapidly when ambulation is lost, although weakness
rather than contractures is a greater contributor to cause of loss of ambulation.

Lightweight plastic AFOs should be worn during sleep to provide a passive stretch, preventing the progression of equinovarus contractures. AFOs should not be worn during standing and walking as tiptoe gait is a compensatory mechanism for proximal weakness. Later in the course, standing and short-distance walking may be maintained by using lightweight long-leg braces or knee–ankle–foot orthoses. Surgery may be performed to release contractures of the hip flexors, iliotibial bands, and Achilles tendons. Immediate postoperative rehabilitation is key in gait-restorative management.

C. Surgical Management

1. For improved gait—Surgical interventions can prolong gait for about 1 year and should be carefully timed. When undergoing procedures requiring anesthesia or sedation, patients with neuromuscular diseases are at higher risk of significant reactions to inhaled anesthetics and certain muscle relaxants (eg, succinylcholine) that may include rhabdomyolysis, hyperkalemia, upper airway obstruction, hypoventilation, atelectasis, respiratory failure, difficulty weaning from mechanical ventilation, cardiac arrhythmias, heart failure, malignant hyperthermia, or sudden cardiac arrest.

2. For scoliosis—The development of scoliosis relates to age and strength more than to wheelchair dependence. A thoracic lumbar spine orthosis is generally not effective in prevention. Surveillance spinal radiographs should be obtained at least yearly. Surgical correction should be considered when FVC exceeds 40% or when the curve is greater than 30 degrees, as rapid curve progression with pulmonary compromise follows. Although scoliosis surgery may improve patient comfort and better seating, particularly for those confined to a wheelchair, studies have not shown that it decreases rate of pulmonary function loss, nor does it increase respiratory function or lifespan.


2. Spinal Muscular Atrophy

ESSENTIALS OF DIAGNOSIS

- Degeneration of α motor neurons in the spinal cord and motor nuclei in the brainstem results in progressive proximal muscle weakness and paralysis of varying severity.
- Autosomal-recessive disorder associated with a defect in the survival motor neuron (SMN) gene.
- Characteristic findings include a floppy infant, with generalized hypotonia, weakness, and motor delay.
- Electromyographic abnormalities show denervation potentials.
General Considerations

Spinal muscular atrophy (SMA) is the second most common fatal autosomal-recessive disorder after cystic fibrosis, with an estimated incidence of 1 in 6000–10,000 live births, and a carrier frequency of about 1 in 50. The disorder is classified into four types based on the age of onset and clinical course. Type 1 is also frequently referred to as infantile X-linked SMA. There is no cure for SMA, and the pathogenesis of the disorder is not completely understood.

The primary defect is in the survival motor neuron (SMN) gene, localized to the 5q13.2 region, although non-SMN variants are also possible. The homozygous biallelic deletion of SMN1 exon 7 is the most common mutation (found in 94% of classic presentations of SMA), but many compound heterozygous individuals have been identified in whom deletions and different point mutations have been detected. The region spanning these genes has a complex organization, including duplications, repetitive sequences, truncated genes, and pseudogenes, which makes molecular analysis of this condition difficult.

The SMN protein appears to play a role in mRNA synthesis in motor neurons and also may inhibit apoptosis. The level of SMN protein correlates with severity of disease.

Clinical Findings

A. Symptoms and Signs

Clinical features are highly suggestive of SMA, particularly in the severe variant that results in a floppy baby or weak child. Weakness is usually symmetric and more proximal than distal; generally it is greater in the legs than in the arms. The severity of weakness correlates with age of onset and with delayed motor milestones according to the clinical classification noted below. Sensation is preserved and deep tendon reflexes are more or less involved depending on age at onset and duration of disease. Attentiveness and intellect are always good.

1. Type 1 disease—SMA type 1, also known as infantile spinal muscular atrophy or Werdnig-Hoffman disease, is the most common and severe type, accounting for about 50% of patients diagnosed with SMA. In this form of the
disease, symptoms progress rapidly; most infants die before 1 year of age from respiratory failure. Classically infants with SMA type 1 have onset of clinical signs before 6 months of age, never acquire the ability to sit unsupported, and generally do not survive beyond age 2.

Clinically, all children with SMA type 1 have a combination of severe hypotonia; weakness (symmetric and more proximal than distal, with legs affected more than arms); atypical respiratory pattern (paradoxical breathing resulting from a spared diaphragm combined with weakened intercostals muscles); impaired head control, with sparing of the facial muscles (enabling alert expression, furrowed brow, and normal eye movements); and weakness of the bulbar muscles (resulting in weak cry, poor suck and swallow reflexes, pooling of secretions, aspiration, tongue fasciculations, and difficulty feeding over time). Deep tendon reflexes are absent or diminished but sensation is preserved. Aspiration pneumonia is an important cause of morbidity and mortality.

2. Types 2 through 4—SMA type 2 (intermediate form) and type 3 (Kugelberg-Welander disease) have a later onset and a less severe course than SMA type 1. Infants with type 2 disease typically are brought for evaluation between 3 and 15 months of age, whereas those with type 3, the least severe form, typically present with signs of weakness at or after 1 year of age and progress to a chronic course. In a study of children and adolescents with SMA types 2 and 3, muscle strength was reduced to a variable extent. Although the muscle weakness affected motor function, actions such as walking, transfer from lying or sitting to the standing position, and stair-climbing were possible in some children. The functional outcome depends primarily on the severity of muscle weakness at presentation rather than the age of onset, although earlier onset tends to correlate with greater weakness. Patients with adult-onset SMA (type 4) usually present in the second or third decade of life and with findings that are otherwise similar to type 3.

B. Laboratory and Imaging Studies

Testing for homozygous deletion of the SMN1 gene is performed; absence of SMN1 exon 7 confirms the diagnosis of SMA. Because point mutations can occur, sequencing of the gene should be pursued if the diagnosis is typical of SMA and only a single deletion is identified. In patients with suspected SMA who have a normal SMN1 gene by molecular genetic testing, the diagnosis of SMA is made clinically by electromyography and nerve conduction studies, and confirmed by muscle biopsy.
Electromyography shows abnormal spontaneous activity with fibrillations and positive sharp waves. The mean duration and amplitude of motor unit action potentials are increased, and many are polyphasic. Nerve conduction velocities are normal or slightly decreased, and sensory nerve action potentials are normal. Serum creatinine kinase concentration typically is normal or slightly elevated, although in rare cases it can be moderately elevated.

Muscle biopsy shows large groups of circular atrophic type 1 and 2 muscle fibers interspersed among fascicles of hypertrophied type 1 fibers. The enlarged fibers have been reinnervated by the sprouting of surviving nerves and are three to four times larger than normal. Histologic diagnosis may be more difficult to make in newborn infants because only widespread atrophy of muscle fibers may be seen. A later repeat biopsy is needed to demonstrate the mixture of hypertrophied and atrophic fibers seen after reinnervation occurs.

**Differential Diagnosis**

The differential diagnosis of a floppy infant includes but is not limited to arthrogryposis multiplex congenital, congenital myasthenic syndromes, congenital myopathies, hypoxic ischemic encephalopathy, metabolic disorders, Prader-Willi syndrome, myelopathy, and Zellweger syndrome.

**Treatment**

**A. Type 1 and 2 Disease**

Treatment for SMA is supportive and directed at providing nutrition and respiratory assistance as needed, and treating or preventing complications of weakness. Physical therapy may be helpful. Spinal bracing can be used to delay the development of progressive scoliosis caused by muscle weakness, as well as provide truncal support. However, spinal bracing applied to patients with SMA type 1 or 2 while in the sitting position significantly reduces expiratory tidal volume; thus, this option should be used cautiously. Proper seating systems are essential to help with upright posture.

Respiratory muscle weakness often results in difficulty clearing lower respiratory secretions and in hypoventilation during sleep. Important interventions include methods for mobilization, clearance of airway secretions, and respiratory support. The latter can be achieved through the use of manual or mechanical chest physiotherapy with postural drainage, and manual cough
assistance or use of mechanical insufflation–exsufflation devices. Noninvasive nasal ventilation is an alternative to tracheostomy and conventional ventilator support in some children with respiratory failure. Decisions about initiating ventilator support should be individualized, taking into account both medical status and family values.

The limited data relating to survival of patients born between 1980 and 2006 suggest that prolonged survival was related to aggressiveness of care. Ventilation for more than 16 hours a day, mechanical insufflation–exsufflation, and gastrostomy tube feeding were significantly and independently associated with prolonged survival independent of the year of birth in children with SMA type 1.

Formal genetic counseling should be provided to all couples with a family history of or a child with SMA syndrome. Parents should be advised of the limitations of current molecular testing: subjects who test negative for the heterozygous deletion may have two SMN1 copies on one chromosome 5 or be carriers of rare subtle mutations, and the occurrence of extremely rare de-novo mutations cannot be ruled out.

**B. Future Directions**

Gene therapy approaches have been evaluated for SMA, using viral vectors to replace SMN1 and therefore augment spinal cord SMN expression in mice. Several other mechanisms have been targeted in drug trials, such as neuroprotective drugs (eg, riluzole) to rescue motor neurons, creatine to improve energy metabolism, and albuterol for its anabolic properties and the molecular effect on SMN type 2 gene expression. Stem cell approaches offer promise as a cellular replacement strategy in the treatment of SMA and are currently receiving considerable attention.


3. Brachial Plexus Birth Palsy

**ESSENTIALS OF DIAGNOSIS**

- Upper limb weakness due to injury to one or more cervical and thoracic nerve roots (C5–T1).
- Injury may occur before, during, or after the birth process.
- Large infants and prolonged or difficult labor are important risk factors.

**General Considerations**

In the United States, the incidence of neonatal or brachial plexus birth palsy (BPBP) is approximately 1.5 in 1000 live births, according to recent epidemiologic reports. Perinatal risk factors include large-for-gestational-age infants (macrosomia), shoulder dystocia, breech delivery, prolonged labor, gestational diabetes, multiparous pregnancies, instrumented (vacuum or forceps-assisted) and difficult deliveries, and previous deliveries resulting in neonatal brachial plexus injury. In up to half of the cases, no risk factors are identified. The extent of brachial plexus injury, as well as prognosis, differs greatly among affected children. Earlier reports showed that up to 92% of patients had mild injury and spontaneously recovered in the first 2 months of life. However, in some reports, only 66% had full recovery and up to 15% had considerable permanent impairment.

**Classification**

BPBP is classified by severity of the pathology and by anatomic location. Nerve injury severity by pathoanatomy can be categorized using the Sedon and Sunderland classification of neuropraxia, axonotmesis, neurotmesis, or avulsion. The Narakas classification defines anatomic involvement and potential for recovery (Table 20–8).
Table 20–8 Narakas classification of brachial plexus birth palsy.

<table>
<thead>
<tr>
<th>Group</th>
<th>Disorder</th>
<th>Nerve Involvement</th>
<th>Clinical Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Upper plexus (classic Erb’s) palsy</td>
<td>C5–6 involvement</td>
<td>Absent shoulder abduction, external rotation, and biceps function 46% of cases. Most favorable prognosis, with full recovery by 4–6 months.</td>
</tr>
<tr>
<td>2</td>
<td>Extended upper plexus palsy</td>
<td>C5–7 involvement</td>
<td>Absent shoulder abduction, external rotation, and biceps function, and wrist extension. 30% of cases. Worse prognosis than group 1.</td>
</tr>
<tr>
<td>3</td>
<td>Total plexopathy</td>
<td>C5–8 involvement</td>
<td>Flail extremity, flexed wrist, closed fist. 20% of cases.</td>
</tr>
<tr>
<td>4</td>
<td>Total plexopathy with Horner’s syndrome</td>
<td>C5–T1 involvement</td>
<td>Flail extremity with Horner’s syndrome. Worse prognosis, associated with probable avulsion injury.</td>
</tr>
</tbody>
</table>

It is also important to determine, for prognostication purposes, whether the injury is preganglionic or postganglionic. In preganglionic avulsion injuries,
recovery of motor function does not occur spontaneously. The loss of phrenic, long thoracic, dorsal scapular, suprascapular, and thoracodorsal nerves and of the sympathetic chain in Horner’s syndrome are suggestive of a preganglionic injury and carry the worst prognosis for motor recovery.

Clinical Findings

A. Symptoms and Signs
Part of a comprehensive examination is to look for other injuries that may have occurred during birth. All extremities should be examined for evidence of fractures (including the clavicles). The infant or young child should be assessed for the presence of Horner’s syndrome, ipsilateral phrenic nerve palsy, or facial nerve palsy. The most common of these is Erb’s palsy. Affected children present with the arm extended, shoulder internally rotated, wrist flexed, and finger extended, resulting in a “waiter’s tip” posture. Passive range of motion and active muscle strength should be assessed. In infants or younger children, this requires the use of observation as they reach for objects, and assessment of reflexes. Palpation of the humeral head and shoulder may reveal glenohumeral head subluxation or dislocation. Muscle tightness or contractures of the pectoralis major, latissimus dorsi, and teres major, as well as scapular winging, may be present. Shoulder function can be evaluated using the modified Mallet classification. Other assessment tools include Toronto Score Test and Hospital for Sick Children Active Movement Scores.

B. Imaging Studies
Plain radiographs of the cervical spine and affected shoulder and clavicle are obtained to rule out fracture, subluxation, or dislocation. MRI is the imaging modality of choice as it allows visualization of the plexus itself, as well as neuromas. However, a computed tomography myelogram is better for identifying pseudomeningoceles associated with nerve root avulsion from the spinal cord.

C. Special Tests
Nerve conduction studies and electromyography play a limited role as they are not consistently predictive of specific nerve root damage and pose technical challenges in small infants and children. Baseline studies are obtained for use in following the natural history or postsurgical course. A nerve conduction study
revealing presence of sensory action potentials in peripheral nerves innervating a paralyzed muscle group is diagnostic of a root avulsion as the sensory root ganglion is located outside the spinal cord.

**Treatment**

Therapy should start immediately in infancy. Goals include maintaining passive range of motion, preventing contractures, muscle strengthening, and facilitating functional development. Serial casting and splinting may be used to prevent contractures at the hand, wrist, or elbow. Children with total or lower plexus injuries usually require primary surgical intervention. Those with upper lesions are usually managed with conservative treatment and watchful waiting initially. If active antigravity strength in the biceps or shoulder abductors is noted within the first 3 months of life, prognosis for recovery is excellent; these children need only continued conservative management. If no improvements are seen in 3–6 months, surgical exploration and microsurgical nerve reconstruction are advocated. Surgical techniques include neurolysis, neuroma resection, nerve grafting, and, more recently, nerve transfers.

One third of infants and children with BPBP have some residual dysfunction and thus must be followed closely. Shoulder deformities (eg, glenohumeral joint dysplasia and joint instability) occur as a result of muscle strength imbalance and longstanding internal rotation contracture owing to weakness of the infraspinatus and teres minor muscles. Secondary reconstructive procedures, including contracture release, muscle transfers, and humeral osteotomy to reposition the upper limb, are useful to improve and maximize function but do not restore normal function.


4. Congenital Muscular Torticollis

**ESSENTIALS OF DIAGNOSIS**

- Head tilt and limited neck range of motion due to contracture or shortening of the sternocleidomastoid muscle.
- May be associated with other musculoskeletal abnormalities, such as hip dysplasia.

**General Considerations**

Congenital muscular torticollis is the third most common congenital deformity presenting in the first week of life, after hip dysplasia and talipes equinovarus. Incidence is about 1 in 250 live births. There is a slight male predominance and the right side is more often affected than the left. Proposed causes include intrauterine crowding or vascular phenomena, fibrosis from peripartum bleeding, compartment syndrome, and primary sternocleidomastoid myopathy. There is a history of difficult birth in 30–60% of cases.

Torticollis is associated with concomitant developmental hip dysplasia, especially in neonates with a history of intrauterine overcrowding. Associated craniofacial deformities include positional or deformational plagiocephaly, which produces flattening of the occiput on the contralateral side, recessed eyebrow and zygoma, deviation of the chin point and nasal tip, inferior orbital dystopia on the affected side, and an inferiorly and posteriorly positioned ipsilateral ear. In some cases compensatory scoliosis may be noted. There is also associated developmental delay and decreased use (neglect) of the ipsilateral hemibody in an otherwise normal child.

**Clinical Findings**

**A. Symptoms and Signs**

Clinical characteristics are a lateral head tilt and chin rotation toward the
opposite side of the tilt, most commonly due to contracture or shortening of the sternocleidomastoid muscle with or without a palpable tumor (fibromatosis colli). Movement of the head and neck is limited and occasionally, on examination, the upper trapezius is also tight. A thorough physical and neurologic examination should be performed to assess the need for further investigation. Nonmuscular causes of torticollis include Sandifer’s syndrome (from gastroesophageal reflux), ocular deficiency, hearing deficit, and central nervous system abnormalities (eg, tumors, Arnold-Chiari malformation).

B. Imaging Studies

In most cases, infants with congenital muscular torticollis improve in response to a regimen of stretching exercises. If the infant does not show improvement despite aggressive physical therapy, a cervical spine radiograph should be obtained to check for congenital cervical vertebral anomalies. Ultrasound is the imaging modality of choice. In the normal patient, the sternocleidomastoid presents as a hypoechoic mass with echogenic lines. In the early stage, there is thickening of the muscle, which appears as weak or uneven echoes. In the later stage of fibrosis, diffuse hyperechoic signals are noted inside the muscle layer without significant blood flow signals. MRI is also used to assess the amount of fibrosis and aberrant dense connective tissue within the sternocleidomastoid and is the preferred imaging technique if evaluation for brain or cervical spine tumors is also desired.

Treatment

More than half the cases resolve spontaneously during the first year of life with minimal residual deficits. Early physical therapy should be initiated if there is lack of cervical range of motion as a result of fibrosis. The program includes manual stretching, proper positioning and handling, as well as strengthening of the contralateral neck muscles. Botulinum toxin injections have been shown to be safe and effective in those who fail to progress with conservative treatment. For resistant cases, surgical release or lengthening of the affected muscle is indicated. More recently, endoscopic correction has emerged as a treatment option that reportedly results in less visible scars and better cosmetic outcome.


### CONGENITAL LIMB DEFICIENCY

#### ESSENTIALS OF DIAGNOSIS

- Complete or partial loss of one or more limbs due to birth defect or failure of limb bud formation.
- May be associated with angular deformities, unstable joints, and inadequate surrounding or proximal musculature.
- Left terminal transradial deficiency is the most common upper limb deficiency.
- Fibular longitudinal is the most common congenital lower limb deficiency.

#### General Considerations

Congenital limb deficiencies occur when part or all of a limb bud fails to form
(at day 26) or differentiate (through 8 weeks of gestation) during the first trimester. The incidence is 4–8 per 10,000 live births, with a 3:1 ratio of upper to lower limbs. Most cases of congenital upper extremity deficiency have no hereditary implications and tend to be sporadic events. However, associated anomalies or syndromes are relatively common. Other risk factors include maternal infections, uterine abnormalities, chorionic villous sampling, and teratogen exposure (eg, alcohol, tobacco), and antiseizure medications (eg, phenytoin, valproic acid). Angular deformities, malrotation, unstable proximal joints, inadequate proximal musculature, or a flail, nonfunctional distal segment may be present, requiring conversion surgeries to establish a mechanically aligned limb and improve prosthetic fit.

Clinical Findings & Classification

The Frantz classification describes deficiencies as either terminal, representing the complete loss of the distal extremity, or intercalary, denoting the absence of intermediate parts with preserved proximal and distal parts of the limb. These main classes are then divided into horizontal and longitudinal deficits. Retained in this system is earlier nomenclature such as amelia (absence of a limb), phocomelia (flipper-like appendage attached to the trunk), and aphasis (absence of a finger or toe). The International Society for Prosthetics and Orthotics (ISPO) Classification is, however, preferred. It classifies limb deficiencies as either transverse or longitudinal. Transverse deficiencies have no distal remaining portions, whereas longitudinal deficiencies have distal portions. Longitudinal deficiencies name the bones that are affected; any bone not named is present and of normal form.

Treatment

Important considerations in prosthetic prescription in children are growth, comfort, durability, weight, simplicity, and cosmesis. Generally, children younger than 10 years of age require a new lower extremity socket every 1–2 years, with periodic lengthening and repairs. A team approach is key, with close surveillance for growth. Bony overgrowth is a common cause of poor socket fit and pain. This can be managed with socket revision. However, surgical revision may be needed if a bone becomes so prominent that it pierces through the skin.

Children with congenital limb deficiency have no sense of loss. Their
resilience allows them to compensate and still be highly functional. A prosthesis is mainly an aid and not a replacement; thus, rejection of its use does not equate to failure. A prosthesis for the lower limb is generally more accepted than one for the upper limb given the necessity for mobility and locomotion. Independent donning and doffing of prostheses can be achieved by children as young as 6 years of age.

**A. Upper Limb Deficiency**

The most common congenital limb deficiency is a left terminal transradial deficiency. Prosthetic fitting should follow the attainment of normal developmental milestones, with the first fitting for a unilateral deficiency occurring when the child achieves sitting balance at around 6–9 months. The goal is to facilitate bimanual activities, propping, symmetric crawling pattern, and an early prosthetic use pattern. The initial transradial prosthesis typically uses a self-suspending design with a supracondylar socket, and a hand, which is preferred by parents. By age 4–5 years, the child can operate all types of prosthetic components and controls.

For the transhumeral deficiency, the initial prosthesis may be suspended either by a harness or by silicone suction suspension. Active terminal devices should be prescribed shortly after the child begins to walk. Body-powered hooks can be used successfully at 2–3 years of age, once the child is strong enough and has the cognitive ability to operate them. By 4–5 years, a body-powered elbow may be used. In general, the higher the limb absence, the less the child accepts the prosthesis.

**B. Lower Limb Deficiency**

Fibular longitudinal deficiency is the most common congenital lower limb deficiency. Bilateral involvement occurs in 25% of cases; unilateral deficiency creates a problem because of the length discrepancy. If leg-length inequality is severe, Syme’s amputation may be performed along with fitting of a Syme’s prosthesis.

Partial proximal femoral focal deficiency (PFFD), also known as longitudinal deficiency of the femur, occurs in 1 in 50,000 births; 10–15% are bilateral. It consists of improper development of the proximal femur, with resultant stunting or shortening of the entire femur. Severe forms of PFFD usually require fusion of the shortened femur to the tibia and removal of the foot with a Syme’s amputation, leaving a residual limb that will accept an appropriate above-the-knee-prosthesis.
The lower limb–deficient child is fitted with a prosthesis when ready to pull up to standing position at 9–10 months. A jointless above-the-knee prosthesis is typically preferred for the toddler. The normal child does not establish heel-toe gait until around 2 years, and a prosthetic heel–toe pattern can be attained by 5 years of age when the child can sustain a single-leg stance. An articulated knee joint is added when the child is about 3–4 years of age.


BRAIN INJURY

1. Traumatic Brain Injury

General Considerations

Traumatic brain injury (TBI) is an important health problem in the United States and is often referred to as a “silent epidemic” given its long-term effects on cognition, sensation, language, and emotion, which may not be readily apparent at the time of injury. In children, TBI has a bimodal distribution, with the highest incidence initially among children from birth to 4 years and later among older adolescents aged 15–19 years. Almost half a million emergency department visits for TBI involve children from birth to 14 years of age. Falls are the leading cause of TBI and are highest among children from birth to age 4 years. Motor vehicle–traffic injuries are the second leading cause, but result in the most TBI-related deaths overall. Males are involved about 1.4 times more often than females regardless of age group. Discussion here focuses on moderate to severe TBI; for information about concussion injuries, see Chapter 29.

Abusive head trauma (AHT) is the term adopted by the American Academy of Pediatrics to describe the constellation of cerebral, spinal, and cranial injuries that result from an inflicted head injury to infants and young children. Also known as “shaken baby syndrome,” the injury occurs as the result of shaking (acceleration–deceleration force) combined with blunt impact trauma. It is the leading cause of substantial traumatic brain injury in infants (resulting in 20–
38% mortality) and is associated with significant neurologic and developmental impairment in up to 75% of survivors. In the United States, AHT occurs in 4000–5000 infants per year. A United Kingdom–based study reported an occurrence of 20–36 per 100,000 children per year.

Pathogenesis

Primary injuries may be caused by impact, deceleration, or rotational forces, or a combination of all three. A coup injury occurs under the site of impact, and a contrecoup injury on the side opposite the area impacted. Coup–contrecoup injury is associated with cerebral contusion as the brain hits the inner surfaces of the skull. Injuries related to acceleration–deceleration and rotational forces are associated with a shearing-type injury that causes disruption of membranes and axons (diffuse axonal injury). Occurrence of secondary hypoxic ischemic injury, cerebral edema, and subsequent metabolic cascades contribute to poor outcomes. Thus, early management to prevent these complications from ensuing is critical.

The classification of brain injury severity is based on the Glasgow Coma Scale score, duration of unconsciousness, and post-traumatic amnesia. A score of 8 or less indicates that the child is in a coma with severe injury; scores of 9–11 indicate moderate injury, and 13–15, mild injury. The longer the elapsed time until a patient reaches consciousness and the ability to respond to environment in any adaptive and meaningful way, the more severe is the injury. The Children’s Orientation and Amnesia Test (COAT) can be used to assess post-traumatic amnesia.

Clinical Findings

Among children who have experienced abusive injuries, subdural hemorrhage is the most common intracranial finding, and skull fractures are present in 15–27% of patients. Other important associated findings are apnea, retinal hemorrhages, rib and long bone fractures in different stages of healing, lacerations, bruising, visceral organ injuries, and burns. A concomitant spinal cord injury may occur but is infrequently documented and may be overlooked in nonfatal cases.

Clinical findings in patients with accidental causes of TBI reflect the mechanism of injury, as outlined earlier.
Complications & Treatment

A. Post-Traumatic Seizures and Epilepsy

The incidence of post-traumatic seizures is generally higher in children than adults and is associated with a lower Glasgow Coma Scale score and younger age. Seizures occur most often during the first 24 hours (immediate) to 7 days postinjury (early phase). Antiepileptic drugs (AEDs) such as phenytoin are often administered. There is no clear guideline regarding prophylactic AED use to prevent late seizures. Post-traumatic epilepsy refers to a seizure occurring after the first week of injury and is usually influenced by the severity of the head injury. Choice of AED therapy should take into consideration the clinical seizure pattern, side effect profile, and effect on cognitive functioning.

B. Central Autonomic Dysfunction

Central autonomic dysfunction is a syndrome of simultaneous sympathetic and muscle overactivity that is usually seen in the early postacute phase after a brain injury but may persist up to 6 months postinjury. Autonomic changes include hyperthermia, hypertension, tachycardia, tachypnea, profuse sweating (diaphoresis), agitation, and papillary dilation. Motor changes manifest as spasticity, dystonia, decerebrate or decorticate posturing, and rigidity.

Diagnosis is by exclusion of other possible causes (eg, infection, hypovolemia, or electrolyte imbalance) but requires a high index of suspicion. Management should be instituted as soon as possible to prevent secondary brain injury stemming from decreased cerebral tissue oxygenation, cerebral edema, and hemorrhagic stroke. Nonpharmacologic strategies include cooling blankets, ice packs, lowering room temperature, providing a quiet and calm environment, comfortable positioning, adequate hydration and nutrition, and pain relief. Medications for hypertension include β blockers such as propranolol and labetalol, and α-adrenergic agonists such as clonidine (given orally, intravenously, or by patch). Bromocriptine, a dopamine agonist, is helpful in patients with central fevers. Opiates, such as morphine, fentanyl, or oxycodone, are given as necessary for pain. For tone management, agents include γ-aminobutyric acid agonists such as diazepam, clonazepam, and baclofen; carbidopa/levodopa; dantrolene sodium; and, in some cases, intrathecal baclofen pump therapy.

C. Neuroendocrine Dysfunction
Hypernatremia with increased urine output from diabetes insipidus occurs due to a deficiency of antidiuretic hormone secondary to posterior pituitary dysfunction. Treatment consists of desmopressin acetate administration. Another condition that may occur is the syndrome of inappropriate antidiuretic hormone secretion (SIADH), in which hyponatremia resulting from water retention manifests as decreased urine output and decreased serum osmolality. Management involves restricting fluid while cautiously reestablishing normal serum sodium levels. Anterior pituitary function may be impaired. There are no specific guidelines for screening; however, this possibility should be investigated when problems such as growth failure or arrest, delayed puberty, amenorrhea, or increased fatigue are noted.

D. Respiratory Dysfunction

Part of the early management in TBI is securing a safe airway. Endotracheal intubation may be transitioned into tracheostomy tube placement for ventilatory and pulmonary support, secretion clearance, and long-term airway management. Eventual decannulation whenever possible is the goal. This is usually achieved by progressively downsizing the tracheotomy tube. Capping trials are carried out to ensure that oxygen saturations are maintained with comfortable breathing, and that secretions are cleared with a good cough.

E. Heterotopic Ossification

Heterotopic ossification occurs in about 14–23% of pediatric patients with TBI, most often among children aged 11 years and older. Those with multiple limb fracture are at higher risk. Common sites are around the hips and knees, and may manifest as limb swelling, pain, and decreased range of motion. Management include aggressive mobilization (once deep vein thrombosis is ruled out) and splinting to prevent contractures. Nonsteroidal antiinflammatory drugs such as indomethacin and ibuprofen are used to halt progression of the abnormal bone deposition. In severe cases or once the heterotopic bone has matured, surgical intervention may be considered if the site causes pain or limits range of motion and function. (For additional discussion, see Chapter 5.)

F. Nutritional Deficiencies

Acute brain injury is associated with increased energy expenditure and a hypermetabolic state. Adequate nutritional and hydration support needs to be maintained and closely followed. Initially, a nasogastric or nasojejunal tube is
inserted to start enteral feedings. For longer term support, a gastric tube is usually recommended with the goal of providing bolus feedings that mimic the typical meal-feeding schedule. In infants and younger children, an upper gastrointestinal series is obtained to screen for gastroesophageal reflux. If reflux is present, a Nissen fundoplication may be performed in conjunction with the gastrostomy placement to avoid reflux and aspiration. Once the child’s cognitive status improves, introduction of oral feedings may be initiated.

**G. Bladder and Bowel Management**

Whenever possible, children are transitioned postinjury to bladder and bowel elimination through removal of urinary catheters and resumption of controlled voiding. Once the Foley catheter is removed, bladder incontinence is most often associated with a neurogenic or uninhibited bladder. Children with TBI are usually able to empty the bladder, but bladder volumes are reduced. When the child’s cognitive status improves, timed voids can be instituted. For the child with a spastic bladder, anticholinergic medication such as oxybutynin may help achieve bladder continence.

Constipation is a common problem. Management includes adding dietary fiber, ensuring adequate fluid intake, and reviewing medications for those that may slow gastrointestinal motility. Stool softeners, laxatives, and suppositories are often needed to help establish a regular bowel program, along with a routine or set time of the day to eliminate stools.

**H. Associated Deficits and Impairments**

1. **Motor impairment and tone abnormalities**—Motor deficits depend on the extent and severity of injury. Focal damage or injury, such as penetrating trauma or cerebral contusion, will most likely result in hemiparesis. Balance, motor control, and coordination are often affected. Motor apraxia, tremors, and ataxia can also impede functional progress.

Spasticity management includes proper positioning and splinting, range of motion, and stretching. Oral medications similar to those used in the treatment of cerebral palsy may be administered (see Table 20–4). Because most such medications are potentially sedating, both the choice of agent and dosing are important. Dantrolene sodium is the least sedating medication but has side effects that include hepatotoxicity; therefore, liver function tests need to be monitored at least every 6 months. Focal treatment with botulinum toxin or phenol injections is common. For patients with severe, generalized spasticity, intrathecal baclofen pump therapy is an effective alternative to sedating
medication. (Additional discussion of spasticity appears in Chapter 6.)

2. Sensory impairments—Anosmia due to olfactory dysfunction is a common sequela of TBI. It can be partial or complete. Various types of visual impairments can occur, including reduced visual acuity; visual field defect; diplopia from injury to cranial nerves III, IV, and VI; and central visual dysfunction or even cortical blindness. Deficits depend on the site or impact of injury as well as associated visual pathways.

   Conductive hearing loss from middle ear dysfunction is more common than sensorineural hearing loss. Central auditory processing due to cortical tract damage may occur, making speech discrimination and learning more difficult. Vestibular dysfunction may also occur, manifesting with complaints of vertigo or balance problems.

3. Cognitive deficits—As the child emerges from a coma, arousal may remain impaired. No systematic studies involving pediatric patients have been carried out regarding the use of pharmacologic agents to improve arousal. However, amantadine has been used clinically as a neurostimulant with variable results. It is important to review the child’s medications periodically and eliminate or minimize potentially sedating medications as much as possible.

   Attention problems are common after TBI, and often there is a premorbid history of ADHD. Management includes behavioral interventions and medications.

   The ability to recall information is basic to learning. Often, after TBI, both immediate and delayed recall are impaired. These problems diminish over time, but less improvement is seen in patients with severe injuries. Cognitive therapy includes increased repetition, use of different organizational strategies, techniques to make learning more efficient, and use of compensatory techniques, such as a “memory book.”

   Executive function is often affected, especially in patients with frontal lobe injuries. Children may have difficulty with problem solving, organization, planning, decision making, self-regulation, and monitoring. This area is usually addressed by the speech/language pathologist in conjunction with cognitive therapy. Up to 50% of brain-injured children are at risk for behavioral problems and disorders, including anxiety, aggression, depression, and sleep problems. Children may be easily frustrated by the challenges of relearning previously acquired skills. Helpful techniques include maintaining a structured environment and daily routine, and providing positive reinforcement of desired behaviors. Sleep disorders, including insomnia and abnormal sleep–wake cycles, are
another common problem. Good sleep hygiene and a consistent schedule are encouraged. If needed, sleep-assistive medications can be prescribed, including melatonin, trazodone, and low-dose benzodiazepines.

I. Rehabilitation and Community Reintegration

The goal of rehabilitation after TBI is to help the child achieve the highest possible level of age-appropriate functional independence physically, cognitively, socially, and emotionally. Additional goals include reducing disability, preventing secondary complications and impairments, and educating the child and parent or caregiver about relevant therapeutic strategies. After this life-changing event, an interdisciplinary team approach is ideal to help with the transition back to home, school, and community. Environmental and architectural modifications may be needed to accommodate for physical limitations. Depending on the child’s cognitive level at discharge from the acute inpatient rehabilitation unit, continued intervention and services may be needed. Some children may require continued care at a post–acute rehabilitation facility; others may benefit from physical, occupation, or speech–language therapy provided in the outpatient or home setting.

Lifestyle and schedule changes may need to be made within the family. Return to school is facilitated by close communication regarding the child’s medical status, level of function, and accommodation needs. An IEP is best managed by a team that is knowledgeable and has a good understanding of the dynamic and changing needs of a child with a brain injury. The social worker can help in navigating the bureaucracy of the school system, and can provide information and referrals to community resources, such as family and peer support groups, adaptive sports programs, and leisure activities in the local community.

Christian C, Block R: Abusive head trauma in infants and children. Pediatrics
2. Nontraumatic Brain Injury

Acquired nontraumatic brain injuries are a significant cause of neurologic devastation in children. Causes include infections (eg, bacterial meningitis, viral encephalitis, and brain abscesses), metabolic problems (eg, anoxic or hypoxic ischemic encephalitis, seen among near-drowning victims and patients resuscitated after cardiopulmonary arrest), severe hypoglycemia, and central pontine myelinosus. Inflammatory and autoimmune causes include central nervous system lupus and anti-NMDA encephalopathy. Vascular insults, whether ischemic or embolic strokes, can occur among patients with sickle cell disease or as a complication after heart surgery. Hemorrhagic stroke may result from vascular malformations.

Brain tumors are another important cause of acquired brain injury in children. The annual incidence of brain tumors in patients younger than 19 years is 2.9–4.8 per 100,000 population. Central nervous system metastases from solid tumors are relatively uncommon in children, compared with adults. The peak age for malignant brain tumors is 3–4 years. Location is predominantly cerebellar (infratentorial) in young children and cerebral (supratentorial) in infants and adolescents. The most frequent tumors include astrocytomas, glioblastomas, and ependymomas. Craniopharyngiomas originate from remnants of embryonic tissue in the Rathke pouch, which later forms the anterior pituitary gland. These tumors vary from small, well-circumscribed, solid nodules to huge, multilocular cysts invading the sella turcica. Disruption of the hypothalamic–pituitary axis can lead to the full spectrum of endocrinopathies, along with visual disturbances (because of its location close to the optic chiasm).
Clinical Findings

A. Symptoms and Signs

Early symptoms of central nervous system tumors frequently are nonspecific but usually relate to the tumor location and the rate of growth. Supratentorial tumors often cause headache, limb weakness, sensory loss, seizures, deteriorating school performance, or personality change. Infratentorial tumors typically cause headache, vomiting, diplopia, and imbalance. Symptoms in infants and toddlers can include irritability, anorexia, head tilt, persistent vomiting, developmental delay or regression, macrocephaly (in infants with open sutures), or forced-downward deviation of the eyes (“sunset sign”). Children with brain tumors most often present with symptoms and signs of increased intracranial pressure due to obstruction of normal cerebrospinal fluid pathways. Concern should be raised by findings of morning headaches that are worsened with the Valsalva maneuver or are followed by vomiting. Fatigue, personality change, worsening school performance, persistent vomiting, neurologic findings (eg, ataxia, head tilt, weakness, vision changes, diplopia, papilledema, unusual somnolence, altered mental status, hemiparesis, seizures, cranial nerve palsies), endocrine disturbance (eg, growth deceleration, diabetes insipidus, precocious puberty), and stigmata of neurofibromatosis should all prompt immediate evaluation for the presence of a central nervous system tumor. Specific neurologic and endocrinologic symptoms may occur later in the illness and may suggest localization of the tumor.

Occasionally, children present with a classic syndrome that should raise suspicion of a tumor involving a specific location. The median duration of symptoms before diagnosis was 2–4 months in approximately 85% of children with malignant brain tumors. Diagnostic delay is less important than tumor aggressiveness in determining survival outcome.

B. Imaging Studies

Brain MRI remains the diagnostic imaging of choice. At times, MRI of the spine may also be warranted to check for metastasis.

Treatment

Surgery is usually the primary step in the treatment of brain tumors, with the goal of removing as much tumor as possible. If a tumor is located in a more
sensitive area, a biopsy may be done to determine the tumor cell type, which may further direct the course of management. The utility of chemotherapy or radiation therapy is determined by the tumor cell type, location, and spread. A ventriculoperitoneal shunt may be required to treat hydrocephalus, and steroids are helpful to decrease brain edema.

3. Posterior Fossa Syndrome

The posterior fossa syndrome may occur in children after resection of cerebellar tumors. The most common symptom is mutism, but variable presentations of oropharyngeal dyspraxia, poor oral intake, akinesia, impaired eye opening, transient cortical blindness, urinary retention, emotional lability, as well as neuropsychiatric and cognitive symptoms also occur. The first anecdotal reports date from the early 1970s and 1980s, when the term cerebellar mutism was coined, later expanded into cerebellar mutism and subsequent dysarthria (cerebellar MSD) syndrome. In 1994, Van Dongen and colleagues described the core features of this syndrome as (1) mutism after resection of a cerebellar mass lesion; (2) delayed onset of mutism after a brief interval of 1–2 days of relatively normal speech postsurgery; (3) transient mutism that lasts from 1 day to 6 months followed by a severe dysarthria, which recovers completely in 1–3 months; and (4) frequent association with neurobehavioral abnormalities. As other symptoms came to light, cerebellar MSD was replaced with the broader term posterior fossa syndrome. The pathophysiology and anatomic basis of this syndrome are poorly understood, and postoperative MRI has failed to reveal either a definite anatomic substrate or mechanism of injury. Recent single photon emission tomography findings suggest that these impairments may be secondary to supratentorial metabolic hypofunction following cerebellar surgery.


Garvin JH, Feldstein NA, Ghatan S: Congenital and childhood tumors. In
Interventional Pain Management

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Chronic pain is one of the most common medical problems encountered by physicians. It has been described as an unpleasant sensation that persists for at least 4 months, and often continues for an indefinite period of time. In the United States alone, more than 90 million people suffer from a variety of chronic pain syndromes. Chronic pain often disrupts family life, reduces function, and is a financial strain. Chronic pain is more common and causes more disability than cancer and heart disease combined. The drain on the U.S. economy produced by chronic pain is enormous, amounting to more than 100 billion dollars a year in medical expenses, lost work productivity, and insurance costs.

Use of interventional pain management procedures in the treatment of chronic benign and malignant pain can offer significant pain relief, improving function and reducing medical costs for patients, and provides a better quality of life and death for those with terminal disease. This chapter reviews indications, contraindications, complications, efficacy, and techniques for the most common interventional procedures used in the treatment of chronic pain.

SYMPATHETIC BLOCKS

General Considerations

The sympathetic nervous system works in conjunction with the parasympathetic nervous system to provide homeostasis for many bodily functions. Sympathetic
nerves provide innervation to a variety of structures, including the skin, blood vessels, and internal organs. The sympathetic nervous system can lead to ongoing chronic neuropathic pain involving the extremities, face, abdomen, and pelvis. Sympathetic nerve blocks are often used to treat a spectrum of neuropathic disorders from sympathetically maintained pain to complex regional pain syndromes. The pain can be mild and self-limited, as in a sunburn, or severe and chronic, as in trigeminal neuralgia, complex regional pain syndrome, and diabetic peripheral neuropathy. Sympathetic nerve blocks are used diagnostically to identify a neuropathic disorder (eg, complex regional pain syndrome) and therapeutically in the treatment of neuropathic conditions that have not responded to more conservative care.

1. Cervical Sympathetic Injection

- **Indications**

Cervical sympathetic injections are performed to treat neuropathic facial pain, such as trigeminal neuralgia, headaches (cluster, migraine), and atypical facial pain. These nerve blocks can also be used in the treatment of facial pain related to different types of cancer involving the head and neck. They are also used for neuropathic upper extremity pain, as seen with sympathetically maintained pain, radiculopathy, and complex regional pain syndromes. An increased temperature recorded in the involved extremity reflects a successful injection resulting from blockade of the sympathetic nerves. In regard to facial pain, the observation of a Horner’s syndrome postinjection, consisting of ptosis, enophthalmos, miosis, and anhidrosis, confirms an effective blockade.

- **Technique**

The cervical sympathetic ganglion, also known as the stellate ganglion, is located anterolateral to the C7 vertebral body and consists of a union between the inferior cervical and first thoracic sympathetic ganglia. The injection procedure is performed with the patient in the supine position. The anterolateral portion of the neck on the involved side is prepared and draped in a sterile fashion. The anterolateral aspect of the C7 vertebral body is located using fluoroscopic visualization. This is achieved by rotating the C-arm to the ipsilateral side and positioning the image intensifier such that a transforaminal
view is obtained with sharp endplates.

The skin overlying the C7 uncinate process is anesthetized with a local anesthetic. A 25- or 22-gauge, 3½-inch spinal needle is then inserted through the skin and advanced under fluoroscopic visualization until the needle tip makes contact at the base of the uncinate process. Next, the needle stylet is removed and tubing is attached to the needle hub. The other end of the tubing is attached to a syringe containing nonionic radiopaque contrast. The syringe is then aspirated to ascertain if the needle tip has entered any vascular structure.

The contrast is injected after negative aspiration under continuous fluoroscopy to further evaluate any violation of vascular structures. The contrast pattern of the properly placed needle tip should outline the surrounding muscular and osseous structures without any vascular uptake (Figure 21–1). The anteroposterior and lateral fluoroscopic views are used to confirm needle placement and contrast pattern. Lastly, the contrast syringe is exchanged for the syringe containing the injectate, which typically consists of a long-acting local anesthetic, such as bupivacaine.

▲ Figure 21–1 Cervical sympathetic injection. Posteroanterior view of spinal needle and contrast injection to confirm needle tip location for blocking the stellate (inferior cervical or cervicothoracic) ganglion. Tip, spinal needle tip; C5, C5 vertebra; C6, C6 vertebra; C7, C7 vertebra.
Side Effects & Complications

Side effects and complications associated with cervical sympathetic blocks can include infection, hematoma with subsequent airway compromise, injury to exiting nerve roots, vascular damage, and trauma to the spinal cord.

Outcome Studies

An evidence-based systematic review of sympathetic injections published by Day in 2008 shows a paucity of literature regarding the utility and outcome of all types of sympathetic injections. Only one prospective, double-blind, placebo-controlled study had been reported; the remainder of the included literature consisted of case reports, case series, and one retrospective study. Day concluded that the evidence was very low quality.

2. Thoracic Sympathetic Injection

Indications

Thoracic sympathetic injections are typically performed when neuropathic upper extremity pain does not respond to cervical sympathetic injections. In approximately 20% of people the thoracic sympathetic fibers that normally contribute to the stellate ganglion bypass it and merge with the brachial plexus. In those situations, blocking the T2 and T3 sympathetic ganglia, which are located anterolateral to their respective T2 and T3 vertebral bodies, provides relief of the neuropathic upper extremity pain.

There are two techniques for blocking the T2 and T3 sympathetic ganglion. The anterior (paratracheal) approach involves placing the spinal needle tip at the ventral surface of the C7 transverse process. Subsequent injection of local anesthetic then tracks in a superior and inferior direction blocking the T2 and T3 sympathetic ganglia. The risks associated with this technique include pneumothorax due to the proximity of the lung apex, and injury to the vertebral artery, which is located anterior to the C7 process. The posterior approach, which is described below, has a significantly greater success rate and is much safer than the anterior approach.
Technique

The patient is positioned prone when performing a T2 sympathetic ganglion injection. The neck and upper thoracic area are prepared and draped in a sterile manner on the symptomatic side. The C-arm is rotated obliquely approximately 10 degrees toward the ipsilateral side.

The skin is anesthetized with a local anesthetic and a 22-or 25-gauge, 3½-inch spinal needle is passed percutaneously and advanced under fluoroscopic guidance until the needle tip makes contact with the posterior lateral margin of the T2 vertebral body. The needle tip is then advanced carefully underneath the rib and further advanced in the lateral plane until the tip is located anterior to the costovertebral joint (Figure 21–2).

Figure 21–2 Thoracic sympathetic injection. Lateral view of spinal needles for thoracic T2 and T3 sympathetic ganglion injections with their tips located anterior to the costovertebral joint. Tip, spinal needle tip; T2, T2 vertebra; T3, T3 vertebra.

Side Effects & Complications

Side effects and complications associated with thoracic sympathetic injections
include nerve injury, pneumothorax, and intravascular injection.

**Outcome Studies**

The literature is lacking in regard to thoracic sympathetic injections.

3. Celiac Sympathetic Injection

**Indications**

Celiac plexus and splanchnic sympathetic blocks have been used predominately in the treatment of cancer-related abdominal pain but can also be used in patients with chronic nonmalignant abdominal pain after the failure of conservative treatment. The celiac plexus is located at the L1 level anterior to the aorta (Figure 21–3). The preganglionic fibers from T5 to T12 form the greater (T5–9), lesser (T10–11), and least splanchnic (T12) nerves, which travel along the lateral and anterolateral aspects of the T9–12 vertebral bodies. All three splanchnic nerves traverse the T12 vertebral body before passing through the diaphragm to synapse at the celiac plexus (see Figure 21–3).
Figure 21–3 Celiac plexus, splanchnic nerves (greater, lesser, and least), and surrounding structures. A, aorta; CP, celiac plexus; D, diaphragm; 9,10,11,12, thoracic sympathetic ganglion.

Multiple techniques for performing celiac plexus blocks have been described (Figure 21–4), including bilateral or unilateral paravertebral retrocrural injection (splanchnic sympathetic blocks), bilateral or unilateral posterior paravertebral antecrural injection, posterior transaortic injection, transintervertebral disc injection, anterior abdominal injection, and lateral abdominal injection. Most of these techniques can be performed using either fluoroscopic or computed tomographic (CT) guidance; the exceptions are the anterior and lateral abdominal approaches, which typically use CT guidance or transcutaneous ultrasound, in the case of the anterior abdominal approach.

Figure 21–4 Celiac plexus sympathetic injection techniques. (1) Unilateral posterior paravertebral retrocrural splanchnic injection. (2) Unilateral posterior paravertebral antecrural plexus injection. (3) Posterior transaortic plexus

Another unique approach to performing a celiac plexus injection involves the use of an upper gastrointestinal endoscope with an ultrasound curvilinear transducer (echoendoscope). The endoscope is positioned at the distal end of the esophagus and the aorta is identified longitudinally by ultrasound. The celiac artery is identified at the point where it branches off from the aorta as the endoscope is advanced distally toward the proximal stomach. A 22-gauge aspiration needle is inserted through and attached to the biopsy port and subsequently visualized by the ultrasound transducer anterior to the celiac artery prior to injection.

## Technique

It is beyond the scope of this chapter to describe every celiac plexus injection technique. The unilateral posterior paravertebral retrocrural technique (splanchnic sympathetic block) is presented as it is arguably the safest approach.

The patient is positioned prone and prepared and draped in a sterile manner. The C-arm is rotated obliquely to a position 15 degrees ipsilateral to the side of needle entrance at the L1 vertebral level. The skin entry site is anesthetized with a local anesthetic, and a 22-gauge, 6- or 8-inch needle (depending on the size of the patient) is passed through the skin and advanced until contact is made with the L1 vertebral body. The needle is then advanced under lateral visualization until the needle tip is positioned just anterior to the ventral aspect of the vertebral body.

The needle tip position is confirmed using anteroposterior and lateral fluoroscopic imaging. The stylet is removed and tubing is attached to the needle hub as well as the syringe containing the radiopaque contrast medium. The contrast is then injected under continuous fluoroscopy to assess contrast spread consistent with positioning of the needle tip in the retrocrural space and to ensure no vascular uptake. After confirmation of appropriate needle tip position the contrast syringe is replaced by a syringe containing 10 mL of a long-acting anesthetic, such as bupivacaine. In total, approximately 20–30 mL of a long-acting anesthetic is injected to block the greater, lower, and least splanchnic nerves.
Side Effects & Complications

Myriad side effects and complications can occur when performing a celiac plexus or splanchnic sympathetic nerve block, depending on the technique used. Side effects resulting from the sympathetic blockade include hypotension and diarrhea, which lead to vascular dilation and increased bowel motility, respectively. Therefore, patients should be well hydrated with intravenous fluids prior to a celiac plexus block, and an existing bowel obstruction is a contraindication for this procedure. Another contraindication is anticoagulation therapy.

The complications associated with a celiac plexus block include pneumothorax, nerve injury, bleeding, bowel injury, and infection. Other complications from a celiac plexus block using the anterior and lateral techniques include injury to the liver, stomach, small and large bowels, and pancreas; hemorrhage; infection; and fistula formation.

Outcome Studies

The literature regarding celiac plexus blocks is substantial when compared with that for other sympathetic injections. The evidence is strong with moderate quality of evidence for the use of celiac plexus blocks and neurolysis in the treatment of abdominal pain related to cancer.

4. Lumbar Sympathetic Injection

Indications

Lumbar sympathetic injections are typically performed in the treatment of acute and chronic neuropathic pain of the lower extremities resulting from disorders such as sympathetically maintained pain, complex regional pain syndrome, phantom limb pain, diabetic peripheral neuropathy, and herpes zoster, as well as for vascular insufficiency disorders. The lumbar sympathetic ganglia are located in front of the psoas muscle at the anterolateral inferior aspect of the L2 vertebra and at the anterolateral superior aspect of the L3 and L4 vertebrae.

Before a lumbar sympathetic injection is performed the patient should be hydrated with intravenous fluids to prevent hypotension as a result of vascular dilation. If there is bilateral involvement, the procedure is typically performed on
one side at a time to prevent hypotension by limiting the injection to one lower extremity.

### Technique

The patient is positioned prone on the procedure table and the area of the lumbar spine on the affected side is prepared and draped in sterile manner. The C-arm is rotated obliquely toward the affected side until the L3 transverse process is superimposed with the anterolateral aspect of the L3 vertebral body. The C-arm is then tilted until the superior endplate of L3 is squared off. Additionally, the C-arm may be tilted caudally or cranially if the transverse process is overlying the superior endplate of the L3 vertebral body to provide access to the superior endplate.

The skin overlying the needle insertion site is anesthetized with a local anesthetic. A 22-gauge, 6- or 8-inch spinal needle (depending on the patient’s body habitus) is inserted and advanced until contact is made with the upper outer corner of the anterolateral aspect of the L3 vertebral body. Once the spinal needle makes contact at the L3 vertebral body, the C-arm is positioned in a lateral view in order to advance the spinal needle under direct fluoroscopic visualization until the needle tip reaches the anterior portion of the L3 vertebral body. The use of a slight bend of the spinal needle tip helps the interventionist to more easily pass the spinal needle along the L3 vertebral body to reach the anterior portion of the L3 vertebral body in a lateral view.

The C-arm is repositioned in the anteroposterior position to confirm that the needle tip is located in the anterolateral position of the L3 vertebral body. Again, the C-arm is repositioned laterally and contrast is injected under continuous fluoroscopy to confirm contrast flow along the location of the L2, L3, and L4 sympathetic ganglia, as well as to ensure that there is no vascular uptake. The C-arm is then returned to the anteroposterior position to confirm contrast flow to the L2, L3, and L4 sympathetic ganglia (Figure 21–5). Finally, 20-mL of a long-acting anesthetic, such as bupivacaine, is injected to complete the lumbar sympathetic block.
Figure 21–5 Lumbar sympathetic injection. Lateral view of spinal needle placement and contrast injection. Tip, spinal needle tip; L2, L2 vertebra; L3, L3 vertebra; L4, L4 vertebra.

Side Effects & Complications

Side effects and complications of lumbar sympathetic blockade include bleeding, infection, nerve injury, neuritis, visceral injury, hypertension, epidural or spinal block, paralysis, and anesthetic toxicity from intravascular injection.

Outcome Studies

The literature regarding lumbar sympathetic injections in the treatment of neuropathic pain is quite limited. Although there are many case reports, case studies, and technique papers in the literature, only two randomized studies have been published. One of these compared lumbar sympathectomy radiofrequency denervation to phenol neurolysis; the other was a randomized, controlled clinical trial that assessed chemical lumbar sympathectomy in the treatment of ischemic rest pain. Despite the low number of clinical studies, the results are strong with moderate quality for percutaneous lumbar sympathectomy in the treatment of neuropathic pain and ischemic pain using radiofrequency denervation or
chemical neurolysis.

5. Superior Hypogastric Sympathetic Injection

**Indications**

Sympathetic blockade of the superior hypogastric plexus is indicated in the treatment of lower abdominal and pelvic pain caused by a number of disorders, including pain of the genitalia, rectum, bladder, uterus, vagina, or prostate; endometriosis; and cancer or other chronic pain disorders involving the descending colon, sigmoid colon, and rectum. The superior hypogastric plexus is located anteriorly from the lower L5 to upper S1 vertebral bodies and below the bifurcation of the iliac vessels. Approaches for performing this block using a posterior single needle, posterior bilateral needle, anterior needle, and transintervertebral disc technique have been described (Figure 21–6). All techniques can be performed using fluoroscopic or CT guidance. The anterior approach has also been described using ultrasound guidance. The bilateral needle technique, also known as the classic technique, is described in this section.

\[\text{Figure 21–6} \text{ Superior hypogastric plexus sympathetic injection using the}\]
transintervertebral disc technique; lateral view. Tip, spinal needle tip; L4, L4 vertebra; L5, L5 vertebra; S1, S1 vertebra.

### Technique

The posterior bilateral needle approach is performed with the patient prone. The lumbosacral area is prepared and draped in a sterile manner. The C-arm is tilted caudally until there is a sharp anteroposterior view of the L5–S1 intervertebral disc. The C-arm is then rotated obliquely 45 degrees to one side. The skin overlying the insertion site is draped in a sterile manner. A 22-gauge, 6- or 8-inch spinal needle (depending on the patient’s body habitus) is inserted and advanced under fluoroscopic or CT guidance to the anterolateral aspect of the inferior L5 vertebral body. Next, the same steps are repeated on the opposite side to place the second needle tip at the anterolateral aspect of the contralateral inferior L5 vertebral body. Contrast is then injected under fluoroscopic guidance through both needles to confirm proper needle placement based on contrast flow, and to verify the absence of vascular uptake. Finally, 10 mL of a long-acting anesthetic, such as bupivacaine, is injected through each of the needles to block the superior hypogastric plexus.

### Side Effects & Complications

The side effects and complications associated with sympathetic blockade of the superior hypogastric plexus depend on the technique employed to perform this procedure. With all techniques there is a risk of infection and bleeding, as well as bladder, rectal, and erectile dysfunction. In addition, the posterior single, posterior bilateral, and anterior needle techniques are associated with a risk of injury to vascular, nerve, and visceral structures. The transintervertebral disc approach appears to have little risk of the types of complications associated with these other techniques.

### Outcome Studies

The literature contains few high-quality studies investigating superior hypogastric plexus sympathetic blocks and outcome. One prospective randomized trial, which compared the classic posterior approach with the transintervertebral disc technique, reported strong results and moderate-quality
evidence. Both techniques produced the same degree of pain relief and morphine consumption trends throughout the followup period. However, there were some differences between the groups randomized to the classic posterior approach and the transverse vertebral disc technique. Two patients in the classic group did not receive any pain relief. Complications were reported in the classic group but not in the transdisc group. There was also a significant decrease in the procedure time for the transdisc group compared with the classic group.

6. Impar Ganglion Sympathetic Injection

**Indications**

Impar ganglion sympathetic block is used in the treatment of perineal pain and coccydynia. The impar ganglion, also known as the ganglion of Walther, is located in the retroperitoneal space ventral to the sacrococcygeal joint. It is the last ganglion of the sympathetic chain found in the spine.

The original technique described for blocking the impar ganglion involved insertion of a spinal needle bent at an angle 5–7 cm proximal to the needle tip. The needle was first inserted inferior and anterior to the tip of the coccyx and then passed superiorly through the anococcygeal ligament to the level of the sacrococcygeal joint. This approach is technically demanding and has an inherent risk of injury to the rectum and surrounding vessels. The transsacrococcygeal technique presented below is easier to perform and safer than the original procedure, and provides equivalent blockade of the ganglion.

**Technique**

The patient is placed in the prone position and the sacrococcygeal area and buttocks are prepared and draped in a sterile manner. The C-arm is positioned with an anteroposterior view over the sacrococcygeal joint. The C-arm is then tilted in a caudal direction until the sacrococcygeal joint is squared off. The skin overlying the joint space is anesthetized with a local anesthetic, and a 22-gauge, 3½-inch spinal needle is passed through the skin until contact is made with the joint. The C-arm is then repositioned laterally after which the spinal needle is passed through the sacrococcygeal joint until the needle tip is anterior to the joint space. Contrast is injected outlining the retroperitoneal space between the ventral surface of the sacrum and coccyx posteriorly, and the dorsal aspect of the
rectum anteriorly. Finally, 5 mL of a long-acting local anesthetic, such as bupivacaine, is injected to block the impar ganglion (Figure 21–7).

▲ Figure 21–7 Impar ganglion sympathetic injection. Lateral view of spinal needle placement and subsequent contrast injection.

▶ Side Effects & Complications

The impar ganglion block procedure described above is technically straightforward and rarely associated with any side effects or complications. As with all such procedures, bleeding and infection can occur, and puncture of the rectum is a rare occurrence.

▶ Outcome Studies

Again, the literature contains few published reports of impar ganglion block procedures. Most of these consist of case reports. One prospective case series described 16 consecutive patients with chronic perineal pain who underwent impar ganglion blocks using the transsacroccocygeal method. The patients were followed over a period of 2 months, and there was a statistically significant reduction in Visual Analogue Scale (VAS) scores. The results of this study were
considered strong based on low-quality evidence associated with its observational study design.

## General Considerations

Epidural fibrosis with or without adhesive arachnoiditis is a possible complication of spinal surgery. The fibrosis can be caused by manipulation of the supporting structures of the spine, bleeding into the epidural space following surgery, or leakage of disc material. Epidural fibrosis is related to inflammatory reactions that result in the entrapment of nerves within dense scar tissue. Arachnoiditis is most frequently seen in patients who have undergone multiple surgical procedures of the spine. Presumably, inflammation and compression of nerve roots by epidural scar or fibrosis (adhesions) are the mechanisms underlying persistent pain following back surgery, a ruptured or herniated disc, or a vertebral body fracture.

Percutaneous epidural lysis of adhesions (also referred to as epidural neuroplasty or epidural adhesiolysis) has been developed as a conservative procedure to reduce or eliminate adhesions or fibrosis. A semirigid catheter with a flexible tip is placed into the epidural space to mechanically loosen or remove adhesions from the nerve roots. Hypertonic saline can be injected through the catheter at the area of fibrosis to mechanically disrupt adhesions and potentially reduce perineural edema. Hyaluronidase can also be injected to assist with breakdown of scar tissue and allow for better infiltration of a local anesthetic and corticosteroid mixture dispensed through the catheter at the site of nerve root involvement.

## Indications

The indications for lysis of epidural adhesions include failed back surgery syndrome, chronic intractable back pain, and chronic radicular leg pain from a disc herniation. Local infection and sepsis are absolute contraindications to this procedure because of the potential for hematogenous spread via Batson’s plexus. Coagulopathy is another absolute contraindication because of potential compression of the spinal cord or thecal sac from a hematoma.

## Technique
The patient is positioned prone and the sacral hiatus is identified by palpation and fluoroscopy. A 16-gauge, 3½-inch styletted needle suitable for catheter placement is inserted and advanced into the sacral hiatus. An anteroposterior fluoroscopic view is obtained to ensure that the needle tip is midline and positioned slightly toward the side of pain just below the S3 foramen. The location of the needle in the epidural space is verified by the injection of contrast under biplanar fluoroscopy, producing an epidurogram that also identifies the area(s) of adhesions.

A styletted epidural catheter is used to perform the lysis of adhesions. To help steer the catheter in the epidural space, a small bend is made at the distal end of the stylet before it is reinserted into the catheter. The introducer needle stylet is withdrawn and the epidural catheter is carefully inserted through the needle to the level of the S3 sacral foramina. The catheter is steered gently by alternatively rotating the bent stylet from its proximal end and advancing or retracting the catheter to lyse the epidural adhesions under live fluoroscopy (Figure 21–8A). After mechanical lysis of the adhesions with the catheter, an additional 5–10 mL of contrast medium is slowly injected through the catheter to confirm the degree of adhesiolysis (Figure 21–8B). Hypertonic saline or hyaluronidase, or both, can be injected at this time through the epidural catheter to assist with the removal of scar tissue. A local anesthetic mixed with a corticosteroid is then injected after the lysis of adhesions through the catheter at the location of nerve root involvement to provide further therapeutic relief. The catheter is carefully removed after finishing the procedure so as not the shear any part of the catheter as it is withdrawn through the needle.
Figure 21-8 Epidural lysis of adhesions. **A:** Contrast injection revealing right-sided adhesions above the S1 nerve root represented by the filling defect. **B:** Contrast injection post–epidural lysis demonstrating elimination of adhesions with contrast flowing along the L4 and L5 nerve roots. L4, L4 vertebra; L5, L5 vertebra; S1, S1 vertebra.
## Side Effects & Complications

Possible side effects and complications of this procedure include increasing pain in the injection site or worsening of symptoms, transient increase in back and leg pain, catheter fracture, and ecchymosis or hematoma formation over the sacral hiatus. More severe complications of epidural lysis of adhesions include local infections, sepsis, bleeding and hematoma formation causing compression of the spinal cord and paralysis, transient nerve compression with temporary paresis, unintended subdural or subarachnoid injection of hypertonic saline, persistent sensory deficit in the lumbar and sacral dermatomes, persistent bowel or bladder dysfunction (or both), and sexual dysfunction. Lysis of adhesions in the cervical or thoracic spine must be exercised with caution due to the significant risk for spinal cord trauma.

## Outcome Studies

Epidural lysis of adhesions can reduce pain in 25% or more of patients who have lumbar radiculopathy plus low back pain refractory to conventional therapies for up to 1 year. Racz and colleagues reported that 65% of patients had therapeutic pain relief for 1–3 months but only 13% of patients had same pain relief for 3–6 months. Manchikanti and associates showed that there were no significant differences in pain relief among patients who underwent 1-day, 2-day, and 3-day procedures. In a prospective randomized, controlled study, Manchikanti and associates demonstrated long-term efficacy of pain relief from this procedure, with 97% of patients experiencing significant pain relief at 3 and 6 months, and 47% at 12 months. These patients also showed significant improvement in mental health and functional status, and a reduction in the use of narcotics. There appears to be no significant difference in treatment efficacy between normal saline and hypertonic saline or between hyaluronidase and hypertonic saline, although the use of hypertonic saline might reduce the number of patients who require additional treatments.

Manchikanti L, Pampati V, Fellows B, et al: Role of one day epidural
RADIOFREQUENCY NEUROLYSIS TECHNIQUES

General Considerations

The basic equipment needed to produce a radiofrequency (RF) tissue lesion from high-frequency waves includes a voltage generator, alternating current, and active and reference electrodes. The patient’s tissues serve as a resistor within the circuit and provide impedance. The active electrode is an insulated needle with an exposed tip, while the reference electrode is a large surface adhesive pad. This configuration leads to the greatest current concentration and heat being next to the tip, with diffusion of the current and heat at the large reference electrode. The current causes vibration of the electrons in the tissues near the RF probe, resulting in an increase in temperature. The greater the voltage and the tissue impedance, the higher the temperature that develops within the tissues.

The advantages of RF neurolysis include controlled lesion size, accurate temperature monitoring, limited need for anesthesia, precise probe placement, low incidence of morbidity or mortality, and rapid post-procedure recovery. The lesion size is dependent on the probe diameter, length of the uninsulated tip, temperature, time, and tissue vascularity. The lesion size is greater with a larger probe diameter, longer uninsulated tip, higher temperature, lower tissue vascularity, and longer lesioning time.

Pulsed RF neurolysis uses 10–30 ms bursts of high-frequency alternating current. Lesions created by this method are low temperature (cold RF) and nondestructive lesions. In producing an RF lesion the tissue that surrounds the tip of the electrode is exposed to an electromagnetic field.

Although the mechanism by which pulsed RF treatment works is not known, there are several theories. One theory is that the electromagnetic field may have a clinical neuromodulation effect rendering the nerve less likely to transmit painful impulses. Alternately, it may work in a manner similar to transcutaneous electrical nerve stimulation, activating both spinal and supraspinal mechanisms, which can reduce pain perception.
1. Radiofrequency Neurolysis of the Facet Joint

**Indications**

Patients with functionally limited spinal facet joint pain that is resistant to at least 3 months of conservative treatment are candidates for RF neurolysis or ablation (RFA). This condition cannot be definitively diagnosed by history, physical examination, or imaging studies. The current method of diagnosis is through facet joint injections or medial branch (facet joint) nerve blocks. The nerves that supply the facet joints from the cervical to the lumbar spine are the third occipital nerve, the medial branches of the dorsal rami, and the L5 dorsal ramus. Cervical and lumbar medial branch blocks have been shown to be target specific if anesthetic solutions are injected carefully at specific osseous target points, and contrast is necessary to ensure that inadvertent venous uptake does not occur. A dual-injection paradigm of facet joint or medial branch nerve injections is recommended for a more accurate diagnosis of facet joint pain because of the high false-positive rates associated with single lumbar and cervical facet joint or medial branch nerve blocks.

**Technique**

The patient is positioned prone with the head turned to the opposite side for cervical facet joint RFA in the same manner as for cervical medial branch nerve injections. The C-arm is positioned to visualize the segmental articular pillar and its waist. A hypodermic insulated RF probe (needle) is directed just medial to the waist of the articular pillar until bone is contacted. The needle is then slowly redirected just off the articular pillar laterally (Figure 21–9) and repositioned with lateral imaging to the anterior third of the pillar along the centroid plane. Needle placement is confirmed with anteroposterior imaging and electrical stimulation that ensures the probe is not close to other nearby neurologic structures. The medial branch nerve is anesthetized before RF lesioning at 80–90°C for at least 1 minute per lesion.
Figure 21–9 Radiofrequency (RF) neurolysis of the cervical facet joint, with the RF probe in position for cervical medial branch neurolysis; lateral view. The centroid is the geometric center of a two-dimensional object or shape that is defined as the intersection of the diagonals (solid lines). The cervical pillar in the lateral view projects as a parallelogram shape, and the cervical medial branch nerve (dashed line) travels in a plane that is parallel to the superior and inferior articular process that passes through the centroid (cervical pillar “waist”). The dashed oval area represents the approximate (not to scale) location of the RF lesion, which encompasses the medial branch nerve due to the position of the RF needle and the active tip along the centroid plane for maximal neurolysis. C5, C5 vertebra; C6, C6 vertebra; C7, C7 vertebra.

The patient is positioned prone for RF lesioning of the thoracic facet joints in the same manner as for medial branch nerve injections. The RF needle is inserted over the midline so that it can be advanced superiorly and laterally to lie parallel to the medial branch nerve as is crosses the target transverse process. The needle is advanced through the skin toward the lateral aspect of the transverse process and walked off laterally with the tip just over the superolateral edge. The T5 through T8 medial branch nerves are superior to the superolateral transverse process, necessitating a more superior position of the RF needle. Thoracic-level RFA lesions are created after electrical stimulation at 80–90°C for at least 1 minute.
The patient is positioned prone for RF lesioning of the lumbar facet joints. The RF probes are placed parallel to the nerves, unlike the perpendicular approach used for medial branch nerve blocks. This allows for optimal denervation of the medial branch nerves. The probe is placed inferior and lateral to the targeted medial branch and advanced under fluoroscopy until contact occurs at the junction of the superior articular process and the transverse process. An oblique “Scottie dog” view (Figure 21–10) is then obtained, which should show the needle residing parallel to the target nerve in the osseous groove. The needle is advanced to the proximal junction of the superior articular process and the transverse process for the L1–4 medial branch nerves, and the proximal junction of the S1 superior articular process and the sacral ala for the L5 dorsal ramus. A lateral view is then obtained to ensure that the needle is placed no further anterior than the posterior aspect of the foramen. Finally, the C-arm is repositioned in an anteroposterior projection to verify that the needle did not stray laterally while being advanced under oblique imaging. Electrical stimulation is performed as a safety precaution, the area is anesthetized, and an RFA lesion is created at 80–90°C for at least 1 minute.

Figure 21–10 Radiofrequency (RF) neurolysis of the lumbar facet joint, with the RF probe in position for lumbar medial branch neurolysis; oblique view. Tip, RF needle tip; Hub, RF needle hub; P, pedicle; TP, transverse process; SAP, superior articular process; L4, L4 vertebra; L5, L5 vertebra; S1, S1 vertebra.
Side Effects & Complications

Patients may feel increased soreness and local pain, especially in the first 3–5 days, but these symptoms usually disappear within 2 weeks. Other postoperative symptoms include itching, burning, and hypersensitivity, which usually subside in 4–6 weeks; gabapentin or tricyclic antidepressants can be very helpful in alleviating these symptoms. Improper needle placement can lead to permanent limb weakness, permanent sensory deficit, or persistent neuritis. In the cervical spine, the proximity to the vertebral artery, combined with the vascular nature of this anatomic region, makes intravascular injection or vascular trauma a distinct possibility. The injection of small amounts of local anesthetic into the vertebral arteries can result in seizures. In the thoracic spine, pneumothorax is a potential risk, given the proximity of the pleural space. No long-term complications or serious adverse effects have been described with facet RFA procedures when motor stimulation was performed before lesioning to prevent inadvertent ventral ramus or nerve root injury.

Needle electromyography (EMG) of the multifidi muscles should be performed if RFA of the facet fails to provide pain relief after several weeks. An EMG examination should show denervation potentials within the multifidi following this procedure, indicating that there was destruction of the medial branch nerves. If no denervation potentials are seen on EMG and the patient is still symptomatic, the RFA should be repeated.

Outcome Studies

Lord and colleagues reported in 1996 the only prospective, double-blind, controlled trial of RFA treatment for chronic cervical facet joint pain. Twenty-four subjects were randomized to an RFA or a sham RFA treatment group. RF treatments were conducted at 80°C degrees for 90 seconds in the RFA group and at 37°C in the control sham group. The median elapsed time until pain returned to 50% of the pretreatment level was 263 days in the treatment group versus 8 days in the sham treatment group. Dreyfuss and associates reported the first prospective study to treat only patients with lumbar facet joint pain proven with dual diagnostic medial branch blocks. A 90% denervation rate was confirmed using multifidi EMG 6 weeks after the procedure. At 1-year followup, nearly 90% of subjects reported at least 60% pain relief, and 60% of subjects had at least 90% pain relief. Overall, one systematic review, two randomized trials, four prospective studies, and three retrospective evaluations of RF medial branch
neurotomy have provided the best evidence to date of short-term relief and moderate evidence of long-term relief of chronic cervical and lumbar facet joint pain. There have been no reports of long-term adverse side effects secondary to facet joint RF neurolysis, including any risk for creating a Charcot joint.

2. Radiofrequency Neurolysis of the Sacroiliac Joint

Indications

Candidates for sacroiliac joint (SIJ) RFA are patients who have been diagnosed with chronic SIJ pain resistant to at least 3 months of conservative treatment, and who have experienced significant but transient relief after intraarticular SIJ corticosteroid injections.

Technique

One common technique for SIJ RFA is the bipolar technique, in which two RF probes are used to create denervation lesions. Under fluoroscopy, the first RF probe is inserted at the inferior joint margin. The second RF probe is placed more cephalad in the joint at a distance of less than 1cm. RF lesions are created at 80 °C. Another RF probe is then placed more cephalad in the sacroiliac joint at a distance of less than 1 cm from the second probe, and another lesion is created. Multiple subsequent lesions are then created in a repetitive alternating “leapfrog” manner, going as high in the joint as possible. An alternative approach is to place a single RF probe and advance it cephalad along the posterior capsule, creating overlapping lesions (Figure 21–11). Another technique is to create a lesion at the origin of the multiple nerve branches that are believed to innervate the sacroiliac joint.
Figure 21–11 Radiofrequency (RF) neurolysis of the sacroiliac joint (SIJ), using a single RF needle overlapping technique. Note contrast in the SIJ confirming probe placement within the posterior limb prior to RF neurolysis. Tip, RF needle tip; Hub, RF needle hub.

## Side Effects & Complications

The major side effect of SIJ lesioning is post-procedure pain. Care must be taken to avoid placing the RF needle too far lateral and traumatizing the sciatic nerve. There is a theoretical risk of dysesthesias if RF lesioning of the L5 dorsal ramus and lateral branches of the S1–3 dorsal rami is performed, as they provide sensory innervation to the skin of the buttock.

## Outcome Studies

Formal peer-reviewed outcome studies for SIJ RFA are lacking. An uncontrolled study by Ferrante and associates used a “leapfrog” technique along the posterior SIJ line. They reported that about 36% of patients experienced a 50% decrease in pain, evaluated by means of the VAS, for at least 6 months. Ferrante and associates also noted that a significantly higher proportion of nonresponders had pain with lateral flexion to the affected side, implying that the presence of facet
disease might have prevented these patients from experiencing at least 50% relief of their total back pain.

3. Radiofrequency Neurolysis of Dorsal Root Ganglia

Indications & Contraindications

Selection criteria for RF neurolysis of dorsal root ganglia (DRG) include radicular pain lasting more than 6 months with no response to conservative treatment, no indication for surgical intervention, and a positive but short-lived response to a selective nerve root block or transforaminal epidural injection. Contraindications include infection, coagulopathy, platelet dysfunction, neck or back pain alone without any limb pain, deafferentation pain in the involved limb, and, for procedures in the cervical and thoracic regions, severe cardiopulmonary disease.

DRG RF neurolysis can be performed using the traditional or pulsed methods. Pulsed RF is being used more frequently in the treatment of DRG-related pain than any other application, as the resulting temperature is below the threshold that causes irreversible nerve injury. The use of pulsed RF significantly reduces the risk of developing post-procedure neuritis.

Technique

The technique for probe placement in performing DRG RF neurolysis is the same whether using heat or cold (pulsed) RF. The probe is placed in the dorsal quadrant of the cervical (Figure 21–12), thoracic, or lumbar foramen. The RF probe is placed anteriorly for RFA of the second DRG (Figure 21–13). Sensory and motor stimulation is performed as a safety precaution and to improve the success rate of the procedure. The voltage at which the patient first perceives the stimulation in the appropriate dermatome is the sensory threshold. This threshold is usually around 0.4–0.7 V when the tip of the needle is next to the DRG using a frequency of 50 Hz. The frequency is changed to 2 Hz for motor stimulation, and the voltage intensity must increase to at least twice the sensory threshold before motor activity in the myotomal distribution is typically seen. This finding reflects the disassociation of stimulation that occurs at a point over the DRG where the sensory and motor nerves are still separate before crossing over into the ventral and dorsal rami, and indicates the proper probe placement site for
DRG RFA when using conventional RF. The probe is typically placed next to the DRG for pulsed RF, obtaining a sensory threshold at 0.1–0.2 V. Lesions for traditional RF are created at 80–90°C for 1–2 minutes and from 2–4 minutes at 42°C for pulsed RF ablation.

▲ Figure 21–12 Radiofrequency (RF) neurolysis of the C6 dorsal root ganglion; oblique (foraminal) view. Tip, RF needle tip; Hub, RF needle hub; P, pedicle; SAP, superior articular process; C5, C5 vertebra; C6, C6 vertebra; C7, C7 vertebra.
Figure 21-13 Radiofrequency (RF) neurolysis of the C2 dorsal root ganglion; anteroposterior view. Tip, RF needle tip; Hub, RF needle hub; C1, C1 vertebra; C2, C2 vertebra.

Side Effects & Complications

Possible risks include nerve injury, vascular trauma, and injection and entry into the subarachnoid space through the intervertebral foramen.

Outcome Studies

A limited case study report showed remarkable effectiveness of pulsed RF in patients with neuropathic pain syndromes who were poorly controlled with other oral and invasive treatments. Sluijter and colleagues demonstrated that 56% of patients with radicular pain had a global perceived effect of more than 75% pain relief. In this study, 8 of 15 patients reported successful treatment at 6 months followup; 3 of the 7 patients in the unsuccessful group reported that pain had improved on the treated side, but they felt pain on the contralateral side afterward. A more recent pilot study using pulsed RF for chronic cervical pain showed that 72% of patients experienced at least 50% pain relief 8 weeks
afterward, and 33% of patients continued to have good pain relief more than 1 year after treatment. Forouzanfar and associates compared subjects receiving conventional RF to a sham group in a prospective double-blind, randomized study of cervical DRG RF and found a significant reduction in pain in the treated compared with the sham group.

4. Cervical Sympathectomy Using Radiofrequency Neurolysis

Indications

Cervical sympathectomy RF lesioning is effective in the treatment of sympathetically mediated pain as well as pain secondary to vascular insufficiency in the face, neck, and upper limbs. This procedure is indicated when the duration of pain relief with sympathetic blocks using local anesthetics is not long lasting.

Technique

The patient is positioned supine with the head rotated to the asymptomatic opposite side. A foraminal view is obtained at the C6–7 level. The RF needle is advanced through the skin toward the superimposed uncinate process. The needle is then slightly withdrawn after bony contact is made to bring the needle tip out of the periosteum. Approximately 3–5 mL of contrast is injected after careful aspiration to assure that the RF needle tip is not in a blood vessel (Figure 21–14). A trial stimulation of both the sensory and motor nerves is performed prior to RF neurolysis to assess for any stimulation of nearby neural structures. A small volume of local anesthetic (0.5 mL) should be injected before lesioning. The RF is applied for 1 minute at 80°C. The cannula is then redirected for additional lesions at the C6 level. Further lesions can be created at the same session or at another time at the C7 or T1 levels, or both.
Because of the proximity to the cervical spinal canal, accidental RF lesioning of the neuraxial structures at this level can result in significant neurologic dysfunction, including quadriparesis. Unintentional lesioning of the phrenic nerve can result in diaphragmatic paralysis and respiratory insufficiency. Inadvertent lesioning of the recurrent laryngeal nerve can result in prolonged or permanent hoarseness. A permanent Horner’s syndrome can occur if the superior cervical sympathetic ganglion is damaged during the procedure. Pneumothorax is a distinct possibility, especially on the right side and with lesioning at the T1 level. The incidence of all of these complications can be decreased with careful use of trial stimulation and fluoroscopic guidance. The anatomic region in this area is highly vascular, increasing the risk for local and systemic anesthetic toxicity as well hematoma formation.
A recent retrospective review study demonstrated that 40% of patients who underwent RF lesioning of the stellate ganglion after responding to a diagnostic injection had 50% or more pain relief at a mean followup of 52 weeks.

5. Lumbar Sympathectomy Using Radiofrequency Neurolysis

### Indications

RF lesioning of the lumbar sympathetic chain is indicated for patients who have experienced only short-term pain relief following multiple lumbar sympathetic blocks with local anesthetic. Pain syndromes amenable to treatment include sympathetically mediated pain of the kidneys, ureters, genitalia, and lower limbs, such as phantom limb pain, complex regional pain syndrome, and a variety of peripheral neuropathies. Lumbar sympathetic ganglion RF lesioning can also be considered in patients suffering from pain secondary to vascular insufficiency of the lower limb.

### Technique

The approach to the lumbar sympathetic chain involves an oblique fluoroscopic technique similar to that used for sympathetic injections. The C-arm is positioned obliquely until the L3 transverse process converges with the vertebral body. The RF probe is advanced toward the upper outer quadrant of the L3 vertebral body until contact is made with the periosteum. The RF probe is then advanced to the anterior edge of the vertebral body under lateral fluoroscopy. Contrast is injected to confirm probe position and assess for unintentional intravascular placement (Figure 21–15). Once the needle tip is in the correct position, sensory stimulation at 50 Hz and motor stimulation at 2 Hz are performed to assess for nearby neural structures. During motor stimulation, there should be no movement in the lower limb with intensities up to 3 V. Approximately 1 mL of preservative-free anesthetic is injected before lesioning. An RF lesion is made between the anterior psoas fascia and the anterolateral vertebral body following proper stimulation.
RF lesioning at multiple levels is a more effective approach for lumbar sympathectomy. One RF lesion with a 10-mm exposed tip is adequate to produce a 10-mm lesion at the L2 vertebral level along the anterolateral aspect of the vertebral body. The RF probes at the L3 and L4 levels should be positioned initially at a point just posterior to the anterior aspect of the vertebral body for stimulation and lesioning. The RF probes are then moved approximately 5 mm anterior and a second stimulation followed by lesioning is performed at each of these levels. With this technique the cannulas are moved further away from the segmental nerves before creating the second lesion. This method creates a 15-mm “strip” lesion at the L3 and L4 levels and a 10-mm lesion at the L2 level. If the sympathetic disorder involves the foot, another 15-mm lesion may be necessary at the L5 level, as well.

**Side Effects & Complications**

Damage to the abdominal viscera, puncture of a ureter, or renal trauma during lumbar sympathetic RF lesioning are distinct possibilities. The incidence of
these complications is decreased if care is taken to place the needle just beyond the anterolateral margin of the vertebral body. RF lesioning in proximity to the genitofemoral nerve at the L2 vertebral level can result in persistent genitofemoral neuritis that can be difficult to treat. Probe placement that is too medial may result in trauma to the intervertebral disc, spinal cord, and exiting nerve roots.

### Outcome Studies

Lesioning at multiple lumbar sympathetic levels produced significant lower limb pain relief for 75% of patients over a period of at least 8 weeks in one prospective case series study.

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Zundert J: Percutaneous pulsed radiofrequency treatment of the cervical dorsal root ganglion in the treatment of chronic cervical pain syndrome: A
VERTEBROPLASTY & KYPHOPLASTY

General Considerations

Traditional treatment of painful compression fractures of spine has been almost exclusively nonoperative, including bed rest, nonsteroidal antiinflammatory drugs, oral or parenteral analgesics, muscle relaxants, and physical therapy with external back bracing. Surgical treatment of symptomatic vertebral compression fractures has consisted, in the past, of reduction and internal fixation using an open anterior or posterior approach. In the treatment of symptomatic vertebral compression fractures, surgical procedures have traditionally been reserved for actual or impending neurologic compromise. Although most patients have favorable outcomes with nonoperative treatment, some fail to respond and suffer from prolonged pain and immobility that can persist for life.

Vertebroplasty is a minimally invasive procedure used in the treatment of pain and instability caused by vertebral body compression fractures. The procedure involves percutaneous structural reinforcement of the compressed vertebral body using polymethylmethacrylate acrylic cement. The cement hardens upon delivery into the vertebral body, providing support and stabilization of the vertebral fracture or compression by eliminating micromovement of the fracture fragments. The main goal of vertebroplasty is to provide pain relief from the compression fracture by either stabilizing the fracture or destroying pain fibers from its exothermic reaction. In experienced hands vertebroplasty is a safe and effective procedure for rapid pain relief of acute vertebral compression fractures resulting from osteoporosis, hemangiomas, and metastatic tumor.

Kyphoplasty was developed to restore vertebral height and spinal alignment that is not possible with vertebroplasty. Kyphoplasty involves placing a catheter with a balloon tip through a large-gauge needle into the vertebral body. The balloon is then inflated, which partially restores vertebral height and creates a cavity for cement injection. The balloon is deflated and removed, after which the cement is injected into the vertebral body. In addition to restoring vertebral height and spinal alignment, kyphoplasty allows for the injection of cement under low pressure potentially reducing the risk for cement extrusion. However, kyphoplasty is performed under general anesthesia in an operating room and
requires considerably more time to complete than vertebroplasty. The primary goal of both procedures is pain relief, which they both equally provide to patients with compression fractures.

### Indications

Vertebroplasty and kyphoplasty are indicated for patients with osteoporotic compression fractures occurring from 2 weeks to 1 year prior, causing moderate to severe back pain that is unresponsive to conservative therapy. Both techniques have been successful in the treatment of spinal compression fracture secondary to metastatic tumor (Figure 21–16) or benign spinal tumors such as hemangiomas, although the success rate is lower compared with osteoporotic compression fractures. Absolute contraindications for either technique include infections, such as discitis, osteomyelitis, or sepsis. Relative contraindications include significant compromise of the spinal canal by retropulsed bone fragments or tumor, fracture older than 2 years, greater than 75% collapse of the vertebral body, disruption of the posterior vertebral body wall, fractures above T5, patients who cannot lie prone, and traumatic compression fractures.

![Figure 21–16](image.png)

> **Figure 21–16** Percutaneous vertebroplasty of a metastatic tumor. Radiofrequency (RF) ablation with destruction of a metastatic tumor within the
L2 vertebral body (see Figure 21–17). The L2 posterior vertebral body wall was destroyed by the tumor (see Figure 21–17). Therefore, contrast was injected into the thecal sac at the beginning of the procedure to assess for any retropulsion of radiolucent tumor that could lead to spinal cord compression during the cement injection. Tip, RF probe tip; L2, L2 vertebra.

CT (Figure 21–17) or magnetic resonance imaging (MRI) is recommended prior to vertebroplasty to assess the type of fracture as well as involvement of the epidural and foraminal space. Both procedures are generally contraindicated for compression fractures associated with a burst fracture of the endplate or a fracture involving the posterior wall of the vertebra.

▲ Figure 21–17 Computed tomography scan of the L2 vertebra. Soft tissue view demonstrating almost complete replacement of the vertebral body by metastatic tumor (gray material within the vertebral body). The posterior wall (dashed line) has been completely destroyed by tumor between the “X” marks. VB, vertebral body; P, pedicle; TP, transverse process; SP, spinous process.

A physical examination is performed to localize the level of the pain and rule out other causes of back pain, such as radiculopathy, disc degeneration, disc herniation, and facet disease. Ideally, the back pain should be at the level of the fracture or within one vertebral body inferior or superior to the fracture that can
be confirmed with an evaluation under fluoroscopy.

**Technique**

For vertebroplasty, the patient is positioned prone under conscious sedation or monitored anesthesia care. The C-arm is rotated obliquely in an ipsilateral direction so that the lateral margin of the pedicle lines up with the lateral border of the vertebral body. The C-arm is then tilted cranially or caudally until the pedicle is visualized between the superior and inferior endplates of the compressed vertebra. A large-gauge needle is inserted at the center of the pedicle under fluoroscopy and then advanced through the pedicle. The needle is kept parallel to the pedicle, and a lateral fluoroscopic view is obtained while the needle is advanced into the anterior third of the vertebral body using a twisting motion or gentle tapping of the needle with a sterile hammer. The needle stylet is removed, and approximately 3 mL of the cement mixed with contrast is injected into the vertebral body (Figure 21–18). The injection is stopped when the cement spreads into the posterior third of the vertebral body. The stylet is placed back into the needle and the needle is removed from the vertebra. Vertebroplasty can be performed using either a single or bilateral pedicular method, depending on the degree of vertebral filling with the cement and the skill of the physician.
Vertebroplasty. Lateral view of cement injection through a large-gauge Jamshidi needle into the osteoporotic compression fracture of the L5 vertebral body. Tip, Jamshidi needle tip; P, pedicle; F, foramen; L4, L4 vertebra; L5, L5 vertebra; S1, S1 vertebra.

Figure 21–19 Kyphoplasty (vertebral augmentation) using a balloon catheter.
The balloon is inflated with contrast, creating a cavity within the L4 vertebral body and restoring vertebral body height. Tip, balloon catheter tip; L3, L3 vertebra; L5, L5 vertebra; S1, S1 vertebra.

**Side Effects & Complications**

The complication rate associated with percutaneous vertebroplasty is 7–10% for treatment of vertebral compression fracture caused by malignant neoplasms, and 1–3% for treatment of osteoporotic vertebral compression fractures. The two major complications associated with vertebroplasty are pulmonary compromise and neurologic sequelae. A few cases involving serious complications have been reported, such as pulmonary, fat, and bone marrow embolism. The pulmonary complication rate is higher in vertebroplasty than kyphoplasty, which appears to be related to the high filling pressure for the bone cement, which can cause extrusion of bone cement into the vertebral venous system. Vertebroplasty requires a high-pressure injection using a low-viscosity cement, which can lead to cement leakage in a certain percentage of procedures because of the vertebral trabecular structure. However, vertebroplasty in general is a safe procedure when treating patients with intractable pain from osteoporotic compression fractures, with fewer than 1% having significant complications. Other complications include fracture of additional vertebral components or the rib, transient fever, increased pain, internal hemorrhage, nerve root irritation, and cord infarction.

The complications associated with kyphoplasty include cement leakage, infection, hematoma, adjacent and distant level fractures, pulmonary embolism, epidural abscess, spondylitis, discitis, bleeding, dural tears, fracture of the transverse processes, fractured ribs, and death. The majority of these complications are rare (eg, pulmonary embolism, 0.17%; spinal cord compression, 0.16%; radiculopathy 0.17%, overall mortality, 0.32%; and perioperative mortality, 0.01%). The most common complications associated with kyphoplasty are new fractures and cement leakage. The incidence of new fractures is 17% within the first year after kyphoplasty. The incidence of cement leakage is 8–14%, with only 0.001–0.04% being symptomatic.

**Outcome Studies**

Vertebroplasty can relieve or completely alleviate severe back pain in patients with compression fractures. A study by Jensen and colleagues evaluating
Percutaneous vertebroplasty in 231 patients showed a 90% success rate in the treatment of osteoporotic vertebral fractures, and an 80% success rate in painful or unstable neoplastic lesions and vertebral hemangiomas. Cortet and associates conducted an open prospective study, which showed that 90% of patients (29 patients with 47 fractures) with age-related or steroid-induced osteoporosis experienced pain relief and improved mobility at 24 hours postvertebroplasty. There were no reports of worsening of pain after the procedure. Another study reported good pain relief in 73% of patients with at least a 50% reduction in analgesic dose. This study also reported moderate pain relief in 29% of patients (37 patients, 52 vertebrae) with painful malignant vertebral lesions.

Two multicenter randomized, placebo-controlled studies published in 2009 compared vertebroplasty with a sham procedure in the treatment of painful osteoporotic vertebral compression fractures. In both sham groups, anesthetic was applied to the periosteum and cement was mixed to simulate its odor. In one study, pressure was provided to the back to simulate the procedure; in the other study, the vertebral body was lightly tapped with a blunt trocar resting on the lamina. Results of both studies showed no changes between the vertebroplasty and sham procedure groups in regard to reduction in pain, function, or quality of life. Both studies concluded that vertebroplasty showed no benefit in the treatment of painful osteoporotic vertebral compression fractures compared with the sham procedure.

There were multiple rebuttals from the medical community—in the form of published critiques, editorials, and position statements—to these two reports debunking the credibility and efficacy of vertebroplasty, which had prompted some commercial health insurance carriers to deny reimbursement for the procedure. The backlash from the medical community pointed out that both papers were seriously flawed in terms of study power, sample size, patient selection criteria, selection bias, placebo authenticity, crossover rate, and cement injection volume. This forceful response restored the status of vertebroplasty as an effective treatment for painful vertebral compression fractures with all insurance carriers. In addition, 1 year later the results of the VERTOS II open-label randomized trial was published, demonstrating that patients with acute osteoporotic vertebral compression fractures causing persistent pain received immediate and sustained pain relief with vertebroplasty. This study also showed that percutaneous vertebroplasty was effective, safe, cost-effective, and provided significantly greater pain relief compared with conservative treatment.

Kyphoplasty has been shown to be as effective as vertebroplasty in alleviating back pain resulting by from thoracic and lumbar vertebral
compression fractures based on low- to moderate-quality evidence. Complications are rare and often inconsequential for both kyphoplasty and vertebroplasty. Vertebroplasty and kyphoplasty are equally effective in the treatment of painful vertebral osteoporotic and malignant compression fractures. Both procedures result in reduction of pain, provide spinal stability, improve quality of life, and are safer and more cost-effective than open surgery, according to recent publications that include a systematic review and meta-analyses.


ENDOSCOPIC LUMBAR DISCECTOMY

General Considerations
Surgery of the spine has continued to advance over the past five decades with the development of less-invasive approaches to treat spinal disorders. The overall goal of such surgery is to treat the pathologic component while minimizing the degree of destruction to normal anatomy. This is and will continue to be the goal for all specialties of medicine, including spine surgery.

Great strides have been made by the spine surgery and interventional pain communities in developing minimally invasive techniques for performing discectomies and fusions at all levels of the spine. The skill set combination required for such procedures carries over to others, such as radiofrequency, vertebroplasty, kyphoplasty, spinal cord stimulation, peripheral nerve stimulation, and spinal discectomy. Another advance involves the use of endoscopic (arthroscopic) discectomy, either open or closed, in the treatment of radiculopathy resulting from lumbar disc pathology or spinal stenosis. This minimally invasive surgical procedure provides significant pain relief, preserves anatomy, reduces cost, improves function, and allows early return to work.

### Indications

Endoscopic lumbar discectomy is indicated for patients with persistent radiculopathy who have not responded to conservative treatment and have a structural pathologic problem of the lumbar spine that creates mechanical compromise to the involved nerve root(s). All patients should have demonstrated nonresponse to physical therapy and spinal injection procedures (eg, epidural or selective nerve root injections, or both) and have undergone diagnostic testing to confirm findings consistent with the radiculopathy, including MRI or CT imaging and electrodiagnostic studies. Imaging studies should demonstrate a pathologic finding—whether it be annular bulging, disc protrusion, or stenosis (central, lateral recess, foraminal)—that is consistent with the patient’s history and physical examination. Anticoagulation therapy may be a contraindication, depending on the necessity for, as well as the potential complications associated with, discontinuing its use.

### Technique

Endoscopic lumbar discectomy can be performed as an open or closed procedure using an interlaminar or a transforaminal approach. The following description is of the closed transforaminal technique. There are essentially three approaches,
described as the inside-out, outside-in, and far lateral approaches. While the techniques for all three are similar, subtle but significant differences for the outside-in and far lateral approaches make them more technically demanding to perform than the inside-out technique.

All three techniques utilize a similar posterolateral approach to the lumbar spine, which is analogous to performing lumbar discography. The inside-out technique uses a posterolateral approach to the spine for the removal of disc material from inside the disc space by placing instruments into the disc, creating a cavity that allows for removal of the herniated disc material. The outside-in technique uses a posterolateral approach to the spine to remove the disc protrusion from outside the disc within the central canal (intracanal) by using instruments in the anterior epidural space to remove the herniated disc material. The far lateral technique is similar to the outside-in technique but uses a more lateral approach to enter the spinal canal to remove the herniated disc material. The learning curve for performing endoscopic lumbar discectomy with any of these techniques is slow, long, and flat.

The outside-in technique is described here. Prior to starting the anesthesia, electrodes are placed on the patient for intraoperative monitoring. The procedure can be performed using either monitored anesthesia care or general anesthesia. The technique described here is based on the use of general anesthesia. After intubation and the induction of general anesthesia, the patient is positioned prone on the radiolucent operating table. Rolls or radiolucent frames are used to position the lumbar spine in flexion, which “opens” the foramina for greater access to the spinal canal and reduces epidural bleeding. The lumbosacral area is prepared and draped in a sterile fashion after the patient is properly positioned on the table.

A lumbar myelogram is performed before starting the surgery, which allows for fluoroscopic visualization of the thecal sac, passing nerve roots, and exiting nerve roots. The C-arm is positioned obliquely ipsilateral to the symptomatic side, with the degree of obliquity dependent on the location of the pathologic process. For example, in a patient with a central disc protrusion, the C-arm is positioned to a greater degree of obliqueness (∼70 degrees). This provides a flatter approach to the spinal canal, allowing the working cannula or sheath to be positioned within the anterior epidural space posterior to the central disc protrusion. On the other hand, in a patient with a posterior lateral disc protrusion, the C-arm is positioned to a lesser degree of obliqueness (∼45 degrees), which positions the working cannula posterior to the disc protrusion. Review of the diagnostic images (MRI or CT) obtained before surgery is imperative in
determining the best approach to the spinal canal based on the disc pathology. Review of the preoperative images will also determine how far laterally the surgeon can safely approach the spinal canal without violating the retroperitoneal space or peritoneal cavity.

A 22-gauge, 6-inch spinal needle is inserted through an incision made after the C-arm has been positioned. The spinal needle is advanced into the targeted disc space anterior to the superior articular process. A nonionic radiopaque contrast solution utilizing a 9:1 contrast-to-indigo carmine ratio is injected into the disc. The indigo carmine preferentially stains the nucleus pulposus, which helps identify nuclear material endoscopically. Next, a guidewire is passed through the needle into the disc space. To access the anterior epidural space, sequential dilators are used, alone or in conjunction with sharp cylindrical reamers (eg, when performing a partial facetectomy or removing vertebral osteophytes to decompress symptomatic stenosis). Finally, the working cannula is advanced over the last dilator and positioned posterior to or alongside the disc protrusion.

The endoscope is then passed through the working cannula to visualize the disc herniation and surrounding structures. The endoscope has a light source, instrument channel, and two irrigation ports. Under direct endoscopic visualization with irrigation, a variety of instruments (eg, cautery devices, forceps, rongeurs, chisels, rasps, curettes, and shavers) are passed through the instrument cannula to remove soft tissue and bone as well as control any bleeding. Ultimately, disc material and bone are removed to decompress the affected nerve roots in order to alleviate the radicular symptoms caused by disc or bone pathology (Figure 21–20).
**Figure 21–20** Endoscopic lumbar discectomy showing surgical results. **A:** Status post–endoscopic lumbar discectomy leading to decompression of the thecal sac (+). The extent of disc decompression involved the removal of disc material in the anterior epidural space (AES) and the posterior intervertebral disc (IVD), including the annulus. This exposed the thecal sac (+), anterior epidural
space (AES; notice the epidural fat (•)), posterior longitudinal ligament (X), L4 vertebral inferior endplate (L4), and L5 vertebral inferior endplate (L5). **B:** Endoscopic removal of disc material from a patient with a large disc extrusion. **C:** Endoscopic removal of facet and vertebral bone from a patient with foraminal and lateral recess stenosis.

### Side Effects & Complications

The complications associated with endoscopic lumbar discectomy include bleeding, infection, nerve root injury, dural tears, dysesthesias, hypesthesias, pseudocysts of the intervertebral disc or facet joint, discitis, recurrent disc herniation, retroperitoneal hematoma, and, rarely, seizures (caused by increased cervical epidural pressure from high flow or prolonged irrigation). Dysesthesias and hypesthesias, although infrequent, are the most common side effects associated with endoscopic lumbar discectomy and are self-limited in duration to several weeks. If persistent beyond several weeks, they typically respond to a selective nerve root injection or a lumbar sympathetic block.

The complication rates for endoscopic lumbar discectomy are consistent with those for microdiscectomy and open discectomy procedures. Serious neurologic complications are rare, as are nerve root injuries and dural tears. Little, if any, epidural fibrosis or scarring is seen with endoscopic lumbar discectomy, when compared with the incidence of epidural scarring in microdiscectomy and open discectomy.

### Outcome Studies

The literature contains several prospective and retrospective papers, as well as one systematic review evaluating the efficacy of endoscopic lumbar discectomy. Several papers compare the outcomes of endoscopic lumbar discectomy with those for microdiscectomy and open discectomy.

A systematic review published in 2010 assessed the effectiveness of transforaminal endoscopic discectomy and also compared it to microdiscectomy for the treatment of symptomatic lumbar disc herniations. One randomized controlled trial, 7 nonrandomized controlled trials, and 31 observational studies were identified based on inclusion criteria. The results from all three endoscopic techniques as a group were as follows: median improvement in leg pain reported using the VAS, 88%; median MacNab Global Perceived Effect (GPE) score,
85%; return to work, 90%; recurrence rate, 1.7%; complications, 2.8%; and reoperations, 7%. When the inside-out and far lateral (intracanal) techniques were compared with the outside-in technique, median improvement in leg pain reported using the VAS was 88% versus 83%, and median MacNab GPE was 86% versus 85%, respectively. The authors also evaluated the effectiveness of endoscopic lumbar discectomy based on the type of disc herniation, demonstrating a median MacNab GPE of 86% for lateral disc herniations, 90% for central disc herniations, and 83% for all disc herniations. Lastly, endoscopic lumbar discectomy was compared with open microdiscectomy. The authors found that there were no statistically significant differences between the two groups in terms of pain reduction, overall improvement, reoperation rate, or complication rate. Comparison of the transforaminal endoscopic group with the open microdiscectomy group showed median leg pain reduction reported using VAS of 89% versus 87%, median MacNab GPE of 84% versus 78%, reoperation rate of 6.8 versus 4.7%, and complication rate of 1.5% versus 1%, respectively. The results of this systematic review are consistent with other published reports that have assessed the efficacy, complications, quality of life, impact on early return to work, feasibility of outpatient procedure, and cost-effectiveness of transforaminal endoscopic lumbar discectomy in the treatment of disc herniations as well as stenosis leading to lumbar radiculopathy.

Transforaminal endoscopic lumbar discectomy offers many advantages over an open microdiscectomy procedure. Because it targets the pathologic process while leaving normal anatomy intact, the endoscopic procedure is less destructive than open microsurgery. With endoscopic discectomy, there is no injury to or stripping of the multifidus muscles, which is important in providing stability to the lumbar vertebral segment, nor is there epidural fibrosis or scarring, as can occur with a microdiscectomy. Additionally, the endoscopic approach is often preferable for a reoperation of a prior open procedure as the posterolateral approach to the spinal canal allows the surgeon to avoid the scar tissue from the original surgery. The endoscopic procedure is also more cost effective and does not require overnight hospitalization. Finally, the endoscopic technique provides the option of using monitored anesthesia care for patients who, because of comorbidities, are unable to undergo general anesthesia.

SPINAL CORD STIMULATION

General Considerations

Spinal cord stimulation (SCS) has been used to control intractable leg pain for more than 30 years. The SCS system stimulates the dorsal column of the spinal cord by means of tiny electrical impulses from small electrical wires placed on the spinal cord. This pain modality offers an option to patients with chronic back pain and can be particularly helpful in patients with chronic leg pain that is unresponsive to other medical treatment. SCS offers the medical community an effective treatment for pain that also reduces the costs associated with the treatment of chronic intractable pain in patients.

SCS involves the use of one or two lead wires, a number of electrodes, and a pulse generator or battery. The lead wire carries the electrical stimulation from the pulse generator or battery to the posterior column of the spinal cord. The mechanism of SCS is still unknown. Some investigators believe that stimulation of the dorsal columns closes the “gates” to pain transmission, while others believe that pain relief from SCS results from direct inhibition of pain pathways in the spinothalamic tracts rather than selective large fiber stimulation. Other possible mechanisms of SCS might involve supraspinal pain inhibition,
activation of central inhibitory mechanisms influencing sympathetic efferent neurons, and activation of putative neurotransmitters or neuromodulators. SCS does not totally eliminate the source of pain, but it interferes with the pain signal, providing relief that varies for each patient.

There are two different types of SCS systems: one that is totally implantable, and another with an internal SCS lead and an external radiofrequency (RF) transmitter. An external RF system might be best for the patient who requires a higher voltage or a multilead therapy for pain relief. The power source can be worn externally on a patient’s belt to transmit RF energy to the pulse generator that is implanted subcutaneously. The internal battery system is designed for a lower voltage or a one- to two-lead therapy, but the battery needs to be surgically replaced every 2–5 years, depending on usage. The SCS leads can be placed into the epidural space either percutaneously or with a small laminotomy. The long-term results from percutaneous epidural SCS implantation are comparable to those obtained from a laminotomy.

### Indications

Patients selected for the SCS procedure usually have had a disability due to intractable leg pain with or without back pain for more than 12 months. They have also tried all conservative forms of treatment, including medications, physical therapy, manipulations, injections, and other adjuvant treatments without any significant relief of the pain. Patients who have not responded to surgical treatment for their back and leg pain are also candidates for this modality; however, this procedure is best for those with limb rather than axial pain. The typical candidate should be free of drug dependence, psychologically suitable and stable, and be a highly motivated individual. The specific conditions considered for SCS treatment include radiculopathy, failed neck or back syndrome, epidural fibrosis or arachnoiditis (resulting in radiculopathy), postherpetic neuralgia, peripheral neuropathy, intercostal neuralgia, complex regional pain syndrome types 1 and 2, phantom limb pain, intractable angina, ischemic limb pain, interstitial cystitis, and headaches.

Absolute contraindications for SCS include local or systemic infection, and untreated coagulopathy (because of the potential for spinal cord compression from a hematoma). Relative contraindications for SCS are unmotivated or psychologically unstable patients, or patients who have exhibited drug-seeking behavior. Patients should try other types of conservative therapy before proceeding to SCS.
Technique

The patient is positioned prone on the procedure table with a bolster under the bottom of the rib cage. A large-gauge epidural needle is advanced under anteroposterior fluoroscopy into the T12–L1 interlaminar space using a loss-of-resistance technique. The needle enters at an angle of approximately 30 degrees from horizontal. The syringe is removed after entering the epidural space, and an SCS lead is passed through the needle into the space. The lead is advanced under fluoroscopic guidance, and is steered by alternately rotating the needle or lead as well as advancing or retracting the lead, being careful not to shear off the lead into the epidural space. The SCS lead tip should be placed in the space at the top of T9 (for lumbar coverage; Figure 21–21) or the top of C3 (for cervical coverage). The lead should be placed either in the midline for a bilateral stimulation pattern or to the symptomatic side for a unilateral stimulation pattern.
Figure 21–21 Lead placement for spinal cord stimulation (SCS). A: Anteroposterior view of an SCS percutaneous lead positioned in the posterior epidural space at the T9 vertebral level, right of midline, for right lower extremity stimulation. B. Lateral view. Tip, SCS lead cephalad tip; T9, T9 vertebra.

The lead is positioned for the best pain coverage. The patient is then sent
home for a trial stimulation of no less than 2 days and typically 5–7 days. The patient is kept on a prophylactic oral antibiotic during this time period to prevent infection. During the trial the patient keeps track of his or her pain relief, sleeping pattern, stimulation tolerance, pain coverage, use of medications, and functional activities. When the patient returns to the office these parameters are discussed to determine whether or not the trial was successful. If the trial was satisfactory, then the patient is scheduled for a permanent implantation.

The permanent placement procedure is very similar to that used for temporary placement. Once the lead is in place, it is permanently attached to the supraspinous ligament or fascia. The battery or radiofrequency receiver is then implanted into the fatty layer of the lower abdomen or one of the buttocks through a small incision. The patient is instructed on how to keep the wound dry and clean for the next 7–10 days to prevent infection. Prophylactic oral antibiotics are prescribed to prevent an infection. Bending or twisting of the trunk or neck (depending on lead placement) is limited after the procedure, as these movements can dislodge the lead.

#### Side Effects & Complications

Complications can occur but are generally minor. The most common complications include scar tissue formation and pain at the pocket or midline incision site. Less common complications include lead migration, lead fracture, and infection. Rare complications include an epidural hematoma, cord compression, paraplegia, and a pulmonary or fat embolism.

#### Outcome Studies

Generally in SCS, the more distally the radicular pain is located, the better are the results. Radicular pain in a unilateral limb responds best; however, with improved design, dual systems, and multiple electrode leads, it is possible to treat axial pain along with limb pain. Neuropathic pain also tends to respond well to SCS. There does not appear to be any difference in pain relief or complications between cervical and lumbar SCS.

LeDouxs demonstrated that 74% of patients with failed back syndrome were receiving 50% or better pain relief with SCS at 2 years. North and colleagues similarly found, in a prospective randomized study, that patients who had failed to obtain relief from a previous laminectomy did significantly better with SCS.
compared with a repeat lumbar laminectomy.

A study by Kemler and associates involving a randomized clinical trial of patients with complex regional pain syndrome demonstrated that the SCS group had significant pain reduction and improved quality of life compared with the non-SCS group. In a recent prospective case study by Barolat and co-workers, patients with chronic low back and leg pain generally did well with SCS for both back and leg pain. At 1 year in this study, 88% of the patients reported fair to excellent relief of leg pain, and 68.8% reported the same for low back pain relief.

SCS is an option for the improvement of pain and quality of life in a carefully selected subset of patients with chronic intractable pain. Because there are no reported long-term side effects, it is generally preferable as a first step when other less-invasive treatments have failed to produce acceptable control of pain. However, because of difficulty in conducting randomized, controlled trials, few high-quality studies have tested the efficacy and cost-effectiveness of SCS in chronic pain patients. Despite the positive findings, there is a need for randomized, controlled, long-term studies on the efficacy of SCS involving larger patient populations.


PERIPHERAL NERVE STIMULATION

General Considerations

Peripheral nerve stimulation (PNS) is a neuromodulation technique in which electrodes are implanted in close proximity to peripheral nerves and electrical current is applied to the nerves to ameliorate chronic pain. The enthusiasm for PNS in the management of chronic, intractable, noncancer pain has grown recently, and it has been shown to be beneficial in the management of headaches and complex regional pain syndromes. Peripheral nerve field stimulation (PNFS) has demonstrated efficacy in the treatment of well-localized, small areas of pain involving the abdomen, inguinal region, pelvis, face, occipital area, and low back. Peripheral nerve stimulation cross-talk (PNSCT) has demonstrated very promising results in the management of large areas of pain.

Indications

PNS is indicated for the treatment of chronic and severe neuropathic pain localized in a peripheral nerve distribution or body part that affects the patient’s ability to function and has not responded to less-invasive measures, such as local anesthetic or sympathetic block, and is not accessible by spinal cord or spinal nerve root stimulation. Neuropathic pain is broadly defined as pain initiated or caused by a primary lesion or dysfunction in the nervous system. This broad definition includes among other syndromes conditions such as headaches, facial pain, and coccydynia.

PNS has been used in the treatment of migraine, occipital, cervicogenic, and cluster headaches. Other indications include the treatment of neuropathic facial pain, such as supraorbital neuralgia and trigeminal neuralgia, and neuropathic limb pain, such as groin pain or complex regional pain syndrome. PNFS is a relatively new application that provides effective pain relief for axial pain such as low back pain, which has been difficult to treat using SCS.

Technique
PNS or PNFS has the same advantage as SCS and intrathecal drug delivery systems (ie, intrathecal pumps) in that patients undergo a temporary trial. The goal of this trial is to determine if the patient is a candidate for a permanent PNS or PNFS implant. The number of PNS leads and target level for stimulation are determined based on the patient’s pain problems.

The temporary trial involves the insertion of PNS stimulator leads into the subcutaneous tissue at the painful site overlying the nerves. The exposed portions of the leads are connected to an external pulse generator. The trial period generally lasts 5–7 days. The trial is considered effective if there is at least a 50% overall reduction in the baseline pain. Other aspects that are considered during the trial include patient tolerance to the stimulation, coverage of the painful area, sleep quality, functional improvement, and reduction in the use of breakthrough pain medications.

A permanent PNS or PNFS implant is indicated if a patient’s pain relief during the trial is greater than 50% compared with his or her baseline pain intensity, when this factor is combined with assessment of other parameters during the trial, taking into consideration the feasibility of making such an evaluation in a short time period. The permanent PNS or PNFS surgical implantation involves insertion and anchoring of the PNS leads into the subcutaneous space and implantation of an internal pulse generator (Figure 21–22).
Figure 21–22 Lead implantation for peripheral nerve stimulation (PNS). **A:** PNS leads implanted along the supraorbital and occipital nerves for treatment of migraine headaches. **B:** PNS leads implanted along the bilateral occipital nerves and upper cervical spine for treatment of occipital headaches and upper neck pain. **C:** PNS leads implanted along the lumbosacral spine for the treatment of low back pain that failed to respond to multiple lumbar fusions.

### Side Effects & Complications

The complication rate for PNS and PNFS is generally low, but both minor and major complications have been reported in the literature. The spinal canal is avoided with the use of PNS, thus eliminating the risk of bleeding and infection into the epidural space. Bleeding and infection, although infrequent, are associated with surgical implantation. Other possible complications include hardware complications such as skin erosion of components, lead migration, component breakage or disconnection, and superficial foreign body reaction to components. Persistent hardware pain occasionally can occur and be significant enough to result in explantation of the device. Sepsis is a rare but major complication. The complications of perineural fibrosis, described in the past with the use of plate or wraparound electrodes, are essentially unheard of with the current use of modern wire-type electrodes. Most complications require system revision or replacement, and lasting side effects are unlikely.

### Outcome Studies

Although there are no randomized, controlled studies demonstrating the efficacy of PNS, PNFS, or PNSCT, numerous published studies suggest that a significant proportion of patients with certain intractable pain syndromes derive benefit from PNS. A brief summary of their findings follows.

Two observational cohort studies assessed occipital nerve stimulation in the treatment of transformation migraine headaches in 15 and 20 patients, respectively, and found that the patients had less frequent, less severe, and less disabling headaches for at least 18 months of followup. Three observational cohort studies investigated the use of occipital nerve stimulation in the treatment of occipital neuralgia in 6, 8, and 14 patients, respectively, with a mean followup from 3 to 25 months. The data from these studies were retrospectively analyzed and demonstrated VAS scores that decreased from 50% to 90%, and in one study
several patients were able to return to work after the PNS implant. Two observational cohort studies, each involving 8 patients, evaluated occipital nerve stimulation in the treatment of cluster headaches. Patients were followed for a mean of 15 and 20 months, respectively, and showed a 40–90% reduction in attack frequency.

Similarly, two observational cohort studies looked at supraorbital nerve and a combination of supraorbital and infraorbital nerve stimulation in the treatment of neuropathic facial pain. The 16 and 11 patients were followed for 30 weeks and 27 months, respectively, and both studies reported a 50% reduction in facial pain and a reduction in the use of pain medications.

Three observational cohort studies assessed the use of peripheral nerve stimulation in the treatment of neuropathic limb pain with 32, 45, and 3 patients, and a mean followup of 2–4 years, 3 months, and 3–12 months, respectively. Hassenbusch and colleagues evaluated 32 patients with complex regional pain syndrome, among whom 30 had a successful stimulation trial and underwent a permanent system implant. Long-term good or fair symptom relief was experienced in nearly two thirds of patients. The VAS was reduced by 50%, vasomotor tone improved significantly, and activity levels increased, as well.

Mobbs and associates retrospectively studied 45 neuropathic pain patients with complex regional pain syndrome related to peripheral nerve trauma. Pain relief was obtained in 60% of patients and a significant improvement in activity levels in 47% of patients. Stinson and co-workers performed PNS in three patients with intractable postoperative inguinal pain and reported a 75–100% decrease in pain.

Several observational cohort studies have demonstrated favorable results when PNS was not being applied to stimulate single identifiable peripheral nerves but rather to generate electrical impulses from the diffuse subcutaneous network of afferent nerves. This latter technique (PNFS) been used in the treatment of neck, low back, chest, and abdominal pain. Paicius and colleagues performed PNFS for chronic lower back pain in six patients, five of whom previously underwent failed back surgery. The stimulation trial was successful in all patients. The followup duration was not clear in the study but the VAS for pain was reduced by 50%. Krutsch and associates reported the use of PNFS for chronic lower back pain in a patient with failed back surgery. The reported pain reduction was greater than 90% at the 12-month followup. Bernstein and co-workers used both spinal cord stimulation and PNFS in the treatment of low back and leg pain. Twenty patients underwent surgery. The combination of the two techniques provided greater benefit than either one alone in most patients. The followup duration and objective VAS were not clearly reported in this study.
Paicius and colleagues also described the use of PNFS for chronic abdominal pain in three patients. Followup was 6–12 months. The VAS for pain was reduced up to 90% on followup, with a reduction in pain medication and an improvement in function. Finally, Falco and associates described a new method for the clinical application of PNS. Their study demonstrated successful pain relief across large painful areas via PNSCT. The creation of an electrical circuit with interlead communication—and, therefore, stimulation from one lead to another despite large distances between the leads—was demonstrated for the first time in this study. Eighteen patients underwent implantation with the application of PNSCT. The numeric rating score was reduced from an average intensity level of 9.1 to 1.2, and patients achieved a significant decrease in pain medication usage, as well as significant functional improvement. Cadaveric experimentation confirmed the presence of the electrical circuit with PNS leads placed at a distance far apart from one another and verified that interlead stimulation (ie, cross-talk) does occur in subcutaneous fat over a great distance.


INTRATHecal PUMP DELIVERY SYSTEMS

General Considerations

The spinal infusion systems include intrathecal and epidural infusion devices. Spinal infusion pumps may be either programmable or nonprogrammable. The programmable pump is a computerized, battery-operated device that can dispense medication at different rates throughout the day as well as deliver a bolus. The nonprogrammable pumps are available in different sizes and different (but fixed) rates. A spinal infusion system consists of a pump and catheter (Figure 21–23), both of which are surgically placed under the skin. The catheter is inserted into the intrathecal space and connected to the pump, which is implanted into the subcutaneous tissue of the abdomen. The pump releases medication through the catheter into the intrathecal space.

▶ Figure 21–23 Intrathecal pump system. A. Medtronic SynchroMed EL
programmable pump. B. Medtronic intrathecal catheter.

Intrathecal infusion bypasses the blood–brain barrier, allowing for direct access to the brain and spinal cord neuroreceptors. Less medication is required for the desired result, with less chance of side effects from the medications. Morphine, baclofen, and recently ziconotide (an N-type calcium channel blocker) are the only medications currently approved by the Food and Drug Administration for infusion into human spinal fluid. However, several other medications have been used “off label” in the pump for pain control.

**Indications**

Intrathecal infusion of preservative-free morphine is designed for patients who suffer from chronic, malignant or nonmalignant intractable pain. These are patients who have not responded to the full range of pain treatment measures, including physical modalities, manipulations, injections, surgery, and other adjuvant treatments. Intrathecal infusion of baclofen is designed for patients who have severe spasticity of cerebral or spinal cord origin and have explored all forms of treatment without significant improvement. The appropriate candidate for an intrathecal pump has undergone a successful intrathecal trial with either an opioid for pain or baclofen for spasticity. The patient’s body size must be sufficient for implantation of the pump, there should be no issues regarding drug dependence, and the patient must be psychologically suitable and stable. Contraindications for an intrathecal pump include a local or systemic infection as well as coagulopathy.

**Technique**

The patient is positioned in a lateral decubitus position to allow for simultaneous access to the spine and abdomen. This avoids the need to prepare the patient twice in the operating room. A relative anteroposterior view of the lumbar spine is obtained, and a large epidural needle is advanced through the skin at a cephalad angle into the intrathecal space at the L3–4 level.

The stylet is removed, allowing free flow of cerebral spinal fluid. A catheter is passed through the needle into the intrathecal space and is advanced cephalad approximately to the T10 level. An incision is made along the epidural needle with dissection to the supraspinous ligament or to the fascia. The needle is removed, and an anchor is placed over the catheter and sutured to the ligament
or fascia. Next, a subcutaneous pocket large enough for the pump is created in the lower abdominal region. A pump catheter is passed from the abdominal pocket to the midline spine incision through a tunnel in either one or two passes. The pump catheter is attached to the intrathecal catheter as well as to the pump. An extra amount of the pump catheter is placed with the pump in the subcutaneous pocket and extra intrathecal catheter is placed at the spine incision site to accommodate for movement by the patient. The spine incision is then sutured closed with layered interrupted sutures follow by skin closure. The pump is sutured into the abdominal pocket typically with silk sutures, and then closed with layered interrupted sutures followed by skin closure.

**Side Effects & Complications**

Postsurgical complications can occur, including infection, bleeding, pain, discomfort around the implant site, hematoma, and seroma formation in the pocket. Other potential complications include symptoms of drug overdosage and underdosage resulting from component failure such as catheter occlusion, catheter fracture or dislodgement, leakage, catheter migration, arachnoiditis, and toxic spinal cord lesion. Patients can experience the symptoms of urinary retention, nausea, vomiting, itching, weakness, facial flushing, constipation, joint pain, muscle twitching, sedation, somnolence, respiratory depression, lack of effectiveness, and disorientation.

Intrathecal pump therapies are a safe option for most patients with intractable pain. Most complications are related to the implant procedure, and the incidence of long-term complications is very low. A few cases of spinal cord compression have been reported from an inflammatory mass at the intrathecal catheter tip. To prevent this serious complication, imaging is important in evaluating patients who develop uncontrollable pain and new neurologic findings after intrathecal catheter implantation. Patients who require high-dose intraspinal opioid therapy and those who receive drugs or mixtures that are not approved for intrathecal use should be monitored closely for signs of a catheter granulomatous mass or malfunction. Prompt diagnosis and treatment can preserve neurologic function.

**Outcome Studies**

Corrado and colleagues reported data on a prospective study that compared patients with intrathecal morphine pumps to those receiving oral medications.
They collected data on 40 patients with intractable chronic low back pain who were equally divided into two groups. Those who received the pump fared remarkably better, with significantly lower pain and lower disability index scores. Kumar and associates reported results of a prospective study of 16 patients who received intrathecal morphine for chronic nonmalignant pain, with a 29-month mean followup. Twelve patients (75%) had successful outcomes in terms of improvement in pain and quality of life, whereas 4 patients were considered to have failed treatment. Smith and co-workers demonstrated in a prospective randomized trial that cancer patients with recalcitrant pain had a statistically significant greater clinical response to intrathecal therapy than to medical management. The intrathecal pump patients had a more substantial reduction in VAS score and drug toxicity and also lived longer than those who received medical management.

The intrathecal pump is a safe and effective procedure if careful attention is paid to technique. Most patients have a good response to the pump, particularly after a successful trial. The major advantage of an implantable infusion pump is that a very small amount of medication is infused directly into the spine, reducing the risk of systemic side effects observed with equivalent amounts of oral medications. An intrathecal pump might be the only recourse for patients who have severe pain and cannot tolerate either the side effects or the interactions of oral pain medications.


Rehabilitation of Rheumatologic Disorders

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Deanna Janora, MD
Sajid Surve, DO

Rheumatologic diseases are commonly encountered in rehabilitation practice, in both inpatient and outpatient (ambulatory) settings. These diseases are diffuse and multifactorial. Most are poorly understood. In some diseases, such as rheumatoid arthritis and fibromyalgia, patients present with vague initial symptoms of fatigue and morning stiffness that can progress to a chronic debilitating process. In others, such as giant cell arteritis, immediate diagnosis and treatment is required to prevent disastrous consequences. Most rheumatologic diseases have limited treatment options, some of which (eg, glucocorticosteroids) have significant side effects.

The role of the physiatrist is to recognize common rheumatologic diseases, treat pain conditions that are common in patients with rheumatologic diseases, and assist in diagnosis and referral for proper treatment. A priority is to maintain function in patients by means of proper bracing and assistive devices, physical activities, and exercise programs. Because many rheumatologic diseases are chronic and debilitating, a team approach is preferred. The physiatrist should lead a team of professionals—including rheumatologists; psychiatrists; physical, occupational, and speech therapists; counselors; and social workers—whose combined efforts aid in diagnosing the disease, controlling pain, and improving function.

RHEUMATOID ARTHRITIS
Symmetric polyarticular inflammatory arthritis.

Diagnostic criteria include morning stiffness of joints, lasting at least 1 hour; involvement of three or more joints; and, in the hand joint, involvement of at least one metacarpophalangeal (MCP) or proximal interphalangeal (PIP) joint.

Additional features include the presence of rheumatoid nodules, positive serum rheumatoid factor (seen in 85% of patients), and bony erosions on radiographs.

General Considerations

Rheumatoid arthritis (RA) is the most common inflammatory arthritis, affecting more than 1.5 million adults in the United States. It has a slow, insidious onset and is a chronic, progressive, systemic rheumatic disease. RA affects more women than men, and more whites than other races, with the peak incidence in the third to sixth decades of life. It is thought to be caused by a combination of genetic, environmental, and immune-mediated factors. The most consistent genetic association is with class II major histocompatibility genes, especially those containing a specific five amino acid sequence in the hypervariable region of HLA-DR4. Other genetic polymorphisms also are found. RA is associated with several bacterial and viral infections; these include *Mycoplasma*, *Mycobacterium*, enteric bacteria, parvovirus, retroviruses, and Epstein-Barr virus.

Clinical Findings

Clinical criteria for diagnosis of RA are based on the classification criteria published in 1987 by the American Rheumatism Association, now the American College of Rheumatology (ACR; Table 22–1). The criteria were updated in 2010
to focus on features seen in earlier stages of the disease that are consistent with more erosive disease (Table 22–2). In using the updated criteria, each feature that is present is scored as 1 point, and a total score equal to or greater than 6 is indicative of RA.

Table 22–1 American College of Rheumatology (ACR) 1987 criteria for classification of rheumatoid arthritis.

<table>
<thead>
<tr>
<th>Clinical Criteria</th>
<th>Diagnostic Criteria (Higher Specificity)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>At least 6 months of:</strong></td>
<td>Presence of rheumatoid nodules</td>
</tr>
<tr>
<td>At least 1 hour of morning stiffness of affected joints</td>
<td>Positive serum rheumatoid factor</td>
</tr>
<tr>
<td>Simultaneous involvement of at least 3 joints with observable soft tissue swelling of fluid</td>
<td>Radiographic changes, including bony erosions at affected joints</td>
</tr>
<tr>
<td>Involvement of at least 1 joint in wrist/hand (MCP, PIP)</td>
<td></td>
</tr>
<tr>
<td>Simultaneous bilateral involvement of same joint</td>
<td></td>
</tr>
</tbody>
</table>

MCP, metacarpophalangeal; PIP, proximal interphalangeal.

Table 22–2 Revised ACR diagnostic criteria for rheumatoid arthritis—2010 update.
A. Symptoms and Signs

Onset is usually polyarticular and generally symmetric in the joints involved. The metacarpophalangeal (MCP), metatarsophalangeal (MTP), wrist, and proximal interphalangeal (PIP) joints are most frequently affected. Other joints that may be involved are the knee, hip, ankle, shoulder, and cervical spine. In addition to patients’ subjective reports of joint pain, inflammatory arthritides such as RA produce signs that include more than 1 hour of morning stiffness in affected joints, and tenderness and warmth to joint palpation. Joint effusions are also common.

A wide spectrum of severity is seen, reflecting variation in joint destruction and extraarticular organ involvement. Systemic manifestations can include rheumatoid nodules, cardiovascular or pulmonary disease, vasculitis, serositis, and eye disease. Felty’s syndrome is an uncommon but severe subset of RA characterized by neutropenia and splenomegaly.

B. Laboratory Findings

<table>
<thead>
<tr>
<th>Points</th>
<th>Swollen or Tender Joints</th>
<th>Laboratory Studies</th>
<th>Acute Phase Reactants</th>
<th>Duration of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td></td>
<td>Negative RF + CCP IgG (ACPA)</td>
<td>Normal CRP and ESR</td>
<td>Less than 6 weeks</td>
</tr>
<tr>
<td>1</td>
<td>1 large joint (shoulder, elbow, hip, knee, ankle)</td>
<td>Low positive RF or CCP IgG (ACPA)</td>
<td>Abnormal CRP or ESR</td>
<td>More than 6 weeks</td>
</tr>
<tr>
<td>2</td>
<td>2–10 large joints</td>
<td>High positive RF or CCP IgG (ACPA)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>1–3 small joints (not including DIP, first MTP, or first CMC, as these are commonly involved in OA)</td>
<td>Highly positive RF or CCP IgG (ACPA)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>4–10 small joints</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>&gt; 10 joints</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

RF, rheumatoid factor; CCP, cyclic citrullinated peptide; IgG, immunoglobulin G; ACPA, α-citrullinated protein antibody; ESR, erythrocyte sedimentation rate; DIP, distal interphalangeal; MTP, metatarsophalangeal; CMC, carpometacarpal; OA, osteoarthritis.

Despite the disease name, a positive serum rheumatoid factor (RF) test does not always indicate that a patient has RA. RF, an autoantibody that binds to the Fc portion of immunoglobulin G (IgG), is also present in other rheumatic diseases, including scleroderma, systemic lupus erythematosus, and Sjögren’s syndrome. Absence of serum RF is also seen in a small percentage of patients with RA.

Serum testing for cyclic citrullinated peptide (CCP) IgG or α-citrullinated protein antibody (ACPA) is more specific than, and equally as sensitive as, the serum RF test. CCP and ACPA are often positive years before a clinical diagnosis is made. They are also used as a marker for erosive disease.

Other pertinent laboratory findings seen with inflammatory arthritic processes include elevated peripheral white blood cell count with a “left shift” (elevated leukocytes in the differential) and an elevated erythrocyte sedimentation rate (ESR). Anemia of chronic disease and increase in platelets are also seen in patients with RA.

Synovial fluid analysis reveals moderate inflammatory (or group II) changes as well as decreased levels of complement C4 and C2, normal glucose, no evidence of crystals, and negative cultures and Gram stain (Table 22–3).

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Characteristic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Color</td>
<td>Yellow or straw</td>
</tr>
<tr>
<td>Opacity</td>
<td>Transparent to slightly cloudy</td>
</tr>
<tr>
<td>Viscosity</td>
<td>Variably decreased</td>
</tr>
<tr>
<td>Mucin clot test</td>
<td>Fair to poor</td>
</tr>
<tr>
<td>White blood cell (WBC) count</td>
<td>3000–50,000/mm³</td>
</tr>
<tr>
<td>WBC differential</td>
<td>&gt;70% polymorphonuclear leukocytes</td>
</tr>
</tbody>
</table>

**Table 22–3** Synovial fluid analysis in rheumatoid arthritis.

**C. Imaging Studies**

Radiographs are used for diagnosis and monitoring of disease course in RA. The hallmarks of RA are juxtaarticular soft tissue swelling, osteopenia and
osteoporosis, marginal bony erosions and cysts, diffuse joint space narrowing, and, at later stages, joint ankylosis. Subluxations occur, including swan-neck and boutonnière deformities in the fingers and ulnar deviation at the wrist (Figures 22–1 and 22–2). Later changes in RA are irreversible.

▲ Figure 22–1 Characteristic ulnar deviation in rheumatoid arthritis.

▲ Figure 22–2 Boutonnière deformity.

The chronic inflammatory processes affect the transverse and alar ligaments on the upper cervical vertebrae (C1–C2) and in combination with erosion of the odontoid process can lead to potentially significant instability at this joint.
D. Biopsy Findings

Findings on tissue biopsy include synovial lining hyperplasia, lymphocytic infiltration, and neoangiogenesis. There is destruction of periarticular bone and cartilage at joint margins. The synovial membrane extends, forming a pannus. The activation of multiple inflammatory mediators, including cytokines, tumor necrosis factor-alpha (TNF-α), and interleukin-1β, allows potential opportunities for targeted treatment. This same immune-mediated process also produces damage to tissues in other organs.

Complications

A. Joint Deformities

Hand and wrist deformities are seen in patients as the disease progresses. Early research showed that weakening of the extensor carpi ulnaris muscle by the erosive tenosynovitis leads to radial deviation of the wrist with compensatory ulnar deviation of the fingers (Figure 22–1), forming a zig-zag pattern. To prevent this deformity and maintain the functional use of the hand, a dynamic MCP extension assist splint can be used.

With ongoing inflammation at the wrist, the ulnar collateral ligament is destroyed. This allows the ulnar head to move up as a dorsal prominence. With this “piano key” styloid deformity, the ulna is easily depressed by the examiner’s fingers. Prefabricated wrist working splints are highly effective in reducing wrist pain after 4 weeks of splint wearing in RA patients with wrist arthritis.

Common deformities associated with RA in the hand include the swan-neck and boutonnière deformities (Figure 22–2). In swan-neck deformity shortening of the intrinsic muscles exerts tension on the dorsal tendon sheath, leading to hyperextension of the PIP joint. The distal interphalangeal (DIP) and MCP joints are in flexion.

Chronic inflammation of the PIP joint may cause the extensor hood to stretch or avulse. The PIP joint moves into excessive flexion, producing a boutonnière deformity. The DIP joint remains in hyperextension based on the pull of the extensor tendons. For patients with RA and a mobile deformity, silver ring splints and prefabricated thermoplastic splints are equally effective and acceptable. (These splints are further described in Chapter 28; see also Figures 28–1 and 28–2.)

Resting wrist and hand splints should not be used as a routine treatment of
patients with early RA as there is no significant difference made for grip strength, joint deformity, hand function, and pain.

The atlantoaxial joint is prone to subluxation in RA. Symptoms of cervical subluxation are pain radiating up into the occiput, painless sensory loss in the hands, paresthesias in the shoulders and arms with movement of the head, or slowly progressive spastic quadriplegia. Lateral cervical spine radiographs views should be examined for more than 3 mm of separation between the odontoid peg and the axial arch. Magnetic resonance imaging (MRI) is also helpful for evaluation of these joints.

Neurologic symptoms are not necessarily related to the degree of subluxation seen on radiographs and may be more correlated to variations in the diameter of the spinal canal. Findings of neurologic loss should be treated as urgent and require further evaluation by a surgeon.

B. Extraarticular Involvement

Although improvements in therapy have decreased the incidence of severe extraarticular complications associated with RA, it is important to be aware of them. One of the most common extraarticular findings is rheumatoid nodules. Rheumatoid nodules are usually noted as subcutaneous masses attached to the periosteum. They occur in 15–20% of patients with RA, most often on extensor surfaces or pressure points, such as the olecranon process and the proximal ulna, as well as on tendons. However, rheumatoid nodules have also been found in heart, lung, sclera, and the central nervous system of patients with RA. Activation of complement in the terminal arterioles causes increases in local histiocytes and fibroblasts as well as an influx of macrophages. These focal areas of granulation tissue grow and expand, leaving behind central necrosis due to destruction of the connective tissue matrix to become the palpable rheumatoid nodules.

Treatment

Management of RA encompasses a range of strategies centered on halting disease progression, maximizing functional ability, and supporting the patient’s overall health. Pharmacotherapy, therapeutic exercise, and use of adaptive devices are mainstays of treatment. Heating and cooling modalities may relieve pain associated with disease flares and inflammation. The relationship between dietary or lifestyle factors and exacerbations of disease in rheumatoid arthritis
Table 22–4  Best evidence for the use of nonbiologic disease-modifying antirheumatic drugs (DMARDs).
Table 22–5 Best evidence for the use of biologic disease-modifying antirheumatic drugs (DMARDs).

<table>
<thead>
<tr>
<th>Medication</th>
<th>Duration of Disease</th>
<th>Disease Activity</th>
<th>Indicators of Poor Prognosis Present</th>
</tr>
</thead>
<tbody>
<tr>
<td>Methotrexate</td>
<td>&lt; 6 mo</td>
<td>Moderate–high</td>
<td>Yes</td>
</tr>
<tr>
<td>Leflunomide</td>
<td>&gt; 24 mo</td>
<td>Moderate</td>
<td>Yes</td>
</tr>
<tr>
<td>Hydroxychloroquine</td>
<td>&lt; 24 mo</td>
<td>Low</td>
<td>No</td>
</tr>
<tr>
<td>Minocycline</td>
<td>&lt; 6 mo</td>
<td>Low</td>
<td>No</td>
</tr>
<tr>
<td>Sulfasalazine</td>
<td>&gt; 24 mo</td>
<td>Moderate</td>
<td>No</td>
</tr>
</tbody>
</table>

*aSee text for explanation.


B. Therapeutic Exercise

Nonpharmacologic treatments, including physical and occupational therapy, have a complementary role to drug therapy in managing inflammatory arthritis. Therapeutic exercise can maintain or improve joint range of motion, aerobic capacity, and muscle strength. Clinicians face three major challenges concerning the use of these treatments: limited research evidence, lack of knowledge of
providers of available treatments, and variability in delivery of multidisciplinary health care around the world.

Despite strong evidence suggesting that exercise is effective in improving disease-related characteristics and functional ability in RA patients, only 26% of patients with RA received a referral from rheumatologists for rehabilitation. However, the clinical and laboratory safety profiles for patients with RA participating in therapeutic exercise programs are good. No deleterious effects were found in any of the published studies.

Aerobic capacity training combined with muscle strength training is recommended for patients with RA. Both water-based and land-based aerobic capacity training show a positive effect on aerobic capacity and muscle strength. Caution must be used in prescribing high-intensity weight-bearing exercises for patients who have preexisting significant radiologic damage of large joints, as some patients may develop additional damage. During an acute flare, joint protection, including orthoses, and relative rest of affected joints are indicated. Isometric muscle exercises can be used until the flare has resolved.

Tai chi has been found to have statistically significant benefits on lower extremity range of motion, in particular ankle range of motion, for people with RA and has not been found to exacerbate symptoms of RA.

C. Orthoses and Adaptive Devices

Although there is evidence that foot orthoses reduce pain and improve functional ability, there is no consensus as to the recommended type. Foot orthoses range from simple cushioned insoles to custom-made rigid cast devices. There is evidence that extra-depth shoes and molded insoles decreases pain on weight-bearing activities such as standing, walking, and stair-climbing.

Patients with RA may use adaptive devices to help with activities of daily living or mobility. There is currently very limited evidence for the effect of assistive technology for adults with RA.

D. Occupational Therapy

Comprehensive occupational therapy is effective in improving function in people with moderate to severe arthritis. Specific interventions, including joint protection and hand exercises, have been found to be effective in patients with RA. High-quality evidence was found for beneficial effects of joint protection and patient education. There is also strong evidence for the efficacy of appropriate splinting to decrease joint pain.
Work disability is common in RA and accounts for a large fraction of the costs associated with the disease. Patients with RA are at risk of work disability from the very start of their symptoms. Twenty to 30% of patients with RA become permanently work disabled during the first 2–3 years of the disease. Risk factors for early work disability include a physically demanding job, older age, and lower educational level, as well as the level of functional disability in daily activities. People with RA who are work disabled have more joint involvement, radiographic damage, and laboratory abnormalities than people who are working. Work disability results from complex interactions of a medical disease, demographic variables, social conditions, and government policies. Although there is general agreement that vocational assessment and intervention should occur early in the course of RA, evidence for vocational rehabilitation is sadly lacking.


OSTEOARTHRITIS

ESSENTIALS OF DIAGNOSIS

- Most common joint disorder in the United States.
- “Wear-and-tear” degenerative disease that preferentially affects weight-bearing joints.
- Patients report dull, achy pain with stiffness; morning stiffness lasts less than 30 minutes.
- Radiographic imaging is the gold standard for diagnosis.

General Considerations

Osteoarthritis is the most common joint disorder in the United States, affecting approximately 27 million people. Although the precise cause of the disorder varies from case to case, osteoarthritis is generally considered to be a “wear-and-tear” type of degenerative disease. Regardless of etiology, the underlying mechanism of degeneration is thought to be a biomechanical or biochemical breakdown of the artrodial cartilage in synovial joints. Joints that are weight
bearing are preferentially affected; namely, the knees, hips, cervical and lumbar spine, as well as joints of the hands and feet, which are prone to overuse. Table 22–6 provides a more complete list of risk factors for developing osteoarthritis.

Table 22–6 Risk factors for osteoarthritis.

<table>
<thead>
<tr>
<th>Risk Factor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Obesity</td>
</tr>
<tr>
<td>Advanced age</td>
</tr>
<tr>
<td>Genetics</td>
</tr>
<tr>
<td>Previous trauma</td>
</tr>
<tr>
<td>Infection</td>
</tr>
<tr>
<td>Repetitive strain</td>
</tr>
<tr>
<td>Crystalline disease (ie, gout)</td>
</tr>
<tr>
<td>Metabolic disorders (ie, diabetes)</td>
</tr>
<tr>
<td>Compromised circulation (ie, avascular necrosis or sickle cell anemia)</td>
</tr>
<tr>
<td>Ligamentous laxity</td>
</tr>
<tr>
<td>Muscle weakness</td>
</tr>
</tbody>
</table>

**Clinical Findings**

In most patients, the major morbidity and presenting complaint is pain, usually described as dull and achy. This may be associated with stiffness, particularly morning stiffness lasting less than 30 minutes. In more advanced disease states, osteoarthritis is also associated with joint crepitus and loss of both active and passive range of motion.

Diagnosis may be made on the basis of clinical findings, although radiographic imaging is the gold standard for confirmation. Certain radiographic features are considered tantamount to diagnosis. These include bony hypertrophy, joint-space narrowing, subchondral cysts, bony sclerosis, and bone spur formation. Nonetheless, one of the main caveats of clinical presentation, as shown by recent studies, is that severity of disease in imaging is discordant to the amount of pain reported by patients. Therefore, appearance on imaging should not be used as a guide for symptomatic management.

In most patients osteoarthritis occurs as a primary disorder resulting from
degeneration over time. However, in certain cases, antecedent trauma or underlying disease may precipitate a secondary osteoarthritis. The main condition to consider and rule out in the differential diagnosis is RA (Table 22–7).

Table 22–7 Differential diagnoses for osteoarthritis.

<table>
<thead>
<tr>
<th>Differential diagnoses for osteoarthritis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td>Seronegative spondyloarthropathies</td>
</tr>
<tr>
<td>Bursitis or tendonitis</td>
</tr>
<tr>
<td>Lyme disease</td>
</tr>
<tr>
<td>Gout</td>
</tr>
<tr>
<td>Pseudogout</td>
</tr>
<tr>
<td>Crystal deposition disease</td>
</tr>
<tr>
<td>Septic arthritis</td>
</tr>
<tr>
<td>Neuropathic arthropathy (ie, Charcot joint)</td>
</tr>
</tbody>
</table>

Treatment

Treatment of osteoarthritis is largely empiric and is targeted at symptom management. Over-the-counter pain medications and lifestyle modification are typically sufficient for the majority of patients. Weight loss, if patients are overweight, is encouraged to reduce stress on painful joints. For osteoarthritis that requires further intervention, the ACR’s 2012 guidelines encourage practitioners to utilize both pharmacologic and nonpharmacologic modalities. Specific recommendations are tailored to the joint involved, based on current evidence (Tables 22–8 through 22–10). Orthotic use in patients with compression fractures of the spine due to osteoarthritis is further described in Chapter 28.

Table 22–8 Treatment of hand osteoarthritis (OA).
<table>
<thead>
<tr>
<th>Pharmacologic Therapy</th>
<th>Nonpharmacologic Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Conditionally recommended:</strong></td>
<td><strong>Conditionally recommended:</strong></td>
</tr>
<tr>
<td>Topical capsaicin</td>
<td>Assessment of ADLs (either by physician or by occupational</td>
</tr>
<tr>
<td>Topical NSAIDs, including trolamine</td>
<td>therapist)</td>
</tr>
<tr>
<td>salicylate&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Adaptive equipment as needed (ie, built-up eating utensils,</td>
</tr>
<tr>
<td>Oral NSAIDs, including COX-2 inhibitors</td>
<td>easy-open containers, etc)</td>
</tr>
<tr>
<td>Tramadol</td>
<td>Instruction in joint protection</td>
</tr>
<tr>
<td><strong>Conditionally not recommended:</strong></td>
<td>Instruction in self-use of thermal modalities</td>
</tr>
<tr>
<td>Opioid analgesics</td>
<td>Thumb spica splinting in basal joint OA</td>
</tr>
<tr>
<td>Intraarticular therapies</td>
<td></td>
</tr>
</tbody>
</table>

NSAIDs, nonsteroidal antiinflammatory drugs; COX, cyclooxygenase; ADLs, activities of daily living.
<sup>a</sup>For patients aged ≥ 75 years, topical NSAIDs are recommended over oral NSAIDs.


**Table 22–9** Treatment of knee osteoarthritis (OA).
<table>
<thead>
<tr>
<th><strong>Pharmacologic Therapy</strong></th>
<th><strong>Nonpharmacologic Therapy</strong></th>
</tr>
</thead>
</table>
| **Conditionally recommended:**  
Acetaminophen  
Oral NSAIDs, including COX-2 inhibitors  
Topical NSAIDs  
Tramadol  
Intraarticular corticosteroids  
**Conditionally not recommended:**  
Chondroitin sulfate  
Glucosamine  
Topical capsaicin  
**No recommendations (positive or negative):**  
Intraarticular hyaluronic acid  
Duloxetine  
Opioid analgesics | **Strongly recommended:**  
Aerobic or resistance-based land exercise  
Aquatic therapy  
Weight loss (if overweight)  
**Conditionally recommended:**  
Self-management programs  
Manual therapy in combination with supervised exercise  
Psychosocial interventions  
Medially directed patellar taping  
Medially wedged insoles for lateral compartment OA  
Laterally wedged subtalar strapped insoles for medial compartment OA  
Instruction in self-use of thermal modalities  
Use of walking aids, if needed  
Tai chi  
Acupuncture\(^a\)  
Transcutaneous electrical nerve stimulation\(^a\)  
**No recommendations (positive or negative):**  
Balance exercises, either alone or in combination with strengthening exercise  
Lateral wedged insoles  
Manual therapy alone  
Knee bracing  
Laterally directed patellar taping |

NSAIDs, nonsteroidal antiinflammatory drugs; COX, cyclooxygenase.  
\(^a\) Conditionally recommended only for patients with chronic moderate to severe pain for whom total knee replacement is indicated, but the patient is either unwilling or unable to receive surgical treatment.  
**Table 22–10** Treatment of hip osteoarthritis.

<table>
<thead>
<tr>
<th>Pharmacologic Therapy</th>
<th>Nonpharmacologic Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Conditionally recommended:</strong></td>
<td><strong>Strongly recommended:</strong></td>
</tr>
<tr>
<td>Acetaminophen</td>
<td>Aerobic or resistance-based land exercise</td>
</tr>
<tr>
<td>Oral NSAIDs, including COX-2 inhibitors</td>
<td>Aquatic therapy</td>
</tr>
<tr>
<td>Tramadol</td>
<td>Weight loss (if overweight)</td>
</tr>
<tr>
<td>Intraarticular corticosteroids</td>
<td><strong>Conditionally recommended:</strong></td>
</tr>
<tr>
<td><strong>Conditionally not recommended:</strong></td>
<td>Self-management programs</td>
</tr>
<tr>
<td>Chondroitin sulfate</td>
<td>Manual therapy in combination with supervised exercise</td>
</tr>
<tr>
<td>Glucosamine</td>
<td>Psychosocial interventions</td>
</tr>
<tr>
<td><strong>No recommendations (positive or negative):</strong></td>
<td>Instruction in self-use of thermal modalities</td>
</tr>
<tr>
<td>Topical NSAIDs</td>
<td>Use of walking aids, if needed</td>
</tr>
<tr>
<td>Intraarticular hyaluronic acid</td>
<td><strong>No recommendations (positive or negative):</strong></td>
</tr>
<tr>
<td>Duloxetine</td>
<td>Balance exercises, either alone or in combination with strengthening exercise</td>
</tr>
<tr>
<td>Opioid analgesics</td>
<td>Tai chi</td>
</tr>
<tr>
<td></td>
<td>Manual therapy alone</td>
</tr>
</tbody>
</table>

NSAIDs, nonsteroidal antiinflammatory drugs.


SERONEGATIVE SPONDYLOARTHRPATHY

Seronegative spondyloarthritis is a term that represents a group of disorders with similar etiology and clinical presentation. The most common disorders included in this category are ankylosing spondylitis, reactive arthritis, psoriatic arthritis, spondyloarthropathy associated with inflammatory bowel disease, and undifferentiated spondyloarthropathy. As a group, these conditions are closely linked to the presence of human leukocyte antigen (HLA)-B27, although the exact role this antigen plays in disease processes remains unclear. Clinically, they are associated primarily with spinal and sacroiliac enthesisopathy, as well as secondary extraarticular involvement of other structures such as the eye, skin, or intestinal tract. Recommendations for diagnosis and treatment of these conditions vary somewhat and are discussed separately for each clinical entity.

1. Ankylosing Spondylitis
General Considerations

Ankylosing spondylitis is the most common type of seronegative spondyloarthropathy, affecting approximately 1 in 1000 people. The male-to-female ratio is 3:1, and peak onset is between the ages of 20 and 40 years. The exact etiology is unknown, although genetics likely plays a role, and family history is a risk factor for developing the disease. The relationship between HLA-B27 and ankylosing spondylitis is strong, and 92% of white patients test positive for this antigen.

Clinical Findings

The hallmark of ankylosing spondylitis is sacroiliac joint inflammation, which characteristically ascends over time to involve progressively higher levels of the spine. The main presenting symptom is usually low back pain and stiffness, as well as fatigue. Extraarticular symptoms such as uveitis may also be present. Another common finding is enthesopathy, which may present as pain in the plantar fascia or Achilles tendon. Symptoms overall are improved with exercise or movement and exacerbated by inactivity. Symptoms are also generally worse in the morning.

The main diagnostic scheme for ankylosing spondylitis is the modified New York criteria, which combines clinical presentation with radiographic data (Table 22–11). Under this scheme, the diagnosis is definitive if the radiographic criterion and at least one clinical criterion are both met. On physical examination, the Schober test is classically used as a measure for lumbar spine
Although unnecessary to establish a diagnosis, the serum HLA-B27 level can be checked if the clinical evidence is borderline. Serum ESR and CRP levels may be helpful in evaluating disease progression and response to treatment. As the disease progresses and ascends in the spine, calcification and syndesmophytes of the thoracic and lumbar regions characteristically develop, leading eventually to bony fusion and the classic “bamboo spine” appearance on radiographic imaging.

**Treatment**

In the management of ankylosing spondylitis, the main treatment guidelines are the joint Assessments in Ankylosing Spondylitis International Society (ASAS) and European League Against Rheumatism (EULAR) guidelines. These have been modified slightly for adaptation within the United States regarding the use of anti-TNF medications. The main treatment considerations are that management should be individualized and multidisciplinary, involving both pharmacologic and nonpharmacologic therapies. Any extraarticular symptoms should be referred to an appropriate specialist for management.

**A. Pharmacotherapy**

For pain management, NSAIDs are the first-line choice, including selective cyclooxygenase (COX)-2 blockers. If patients are not responsive to NSAIDs, then the second-line choice is opiate analgesics. Corticosteroid injections into
locally affected joints are acceptable if warranted, although facet joint injections are of questionable value. The use of DMARDs such as sulfasalazine or methotrexate, which have shown great efficacy in the treatment in RA, is discouraged for patients with purely axial arthritis but may be considered for those with peripheral arthritis.

To be considered for anti-TNF therapy, patients must have a definitive diagnosis based on the modified New York criteria. They must have had active disease for at least 4 weeks, as evidenced by objective measures (ie, the Bath Ankylosing Spondylitis Activity Index [BASDAI] or Physician Global Assessment [PGA]), and must have shown nonresponse to at least two different NSAIDs for at least 3 months. Patients with peripheral arthritis must also have shown either a lack of response or intolerability to at least one DMARD, preferably sulfasalazine. In the choice of anti-TNF therapy, the ASAS/EULAR guidelines do not prefer one agent over another.

**B. Exercise and Rehabilitation**

From a nonpharmacologic standpoint, the main modality in the management of ankylosing spondylitis is exercise, either through a home exercise program or through land- or aquatic-based physical therapy. The goal of exercise should be improved flexibility and postural control. A pulmonary rehabilitation program may also be helpful in maximizing chest wall expansion. Patients should be encouraged to sleep on a firm mattress with a thin pillow to prevent worsening kyphosis. Bracing with a hyperextension brace (ie, Jewett brace) may be necessary to prevent worsening kyphosis in severe cases. For patients with severe deformity or debility, surgical correction via osteotomy may be required. Vertebral fractures are very common in end-stage disease and may also require surgical fixation. In patients with hip or shoulder joint involvement, joint replacement surgery may be considered in accordance with standard orthopedic guidelines.


2. Reactive Arthritis

General Considerations

Reactive arthritis (formerly known as Reiter’s syndrome) is a form of seronegative spondyloarthropathy that is classically preceded by an infectious process, usually a genitourinary or gastrointestinal infection. The disease is less common than ankylosing spondylitis, affecting approximately 35 in 100,000 people. In most cases, reactive arthritis is self-limiting, resolving spontaneously within 1 year. Approximately 15% of cases become chronic, lifelong conditions. The male-to-female ratio is equal when brought about by gastrointestinal causes, and 9:1 when precipitated by venereal disease. As with ankylosing spondylitis, the peak incidence is between the ages of 20 and 40 years.

Clinical Findings

In the typical clinical presentation, symptoms begin acutely between 2 and 6 weeks after the initial infectious process. Symptoms include acute conjunctivitis, urethritis, and asymmetric peripheral arthritis predominantly affecting joints in the lower extremities. Concordant low back pain is present in approximately half of patients, and other extraarticular findings may include prostatitis in men, cervicitis in women, fatigue, malaise, fever, and enthesopathy. The diagnosis can be made clinically based on the history and presence of preceding infection.

Serum ESR and CRP levels are characteristically high for reactive arthritis and track well with the disease process. Serum HLA-B27 levels are positive in approximately 60% of cases but are not required to make the diagnosis. Radiographic studies are usually negative in acute disease but may show an erosive arthritis of affected joints in chronic disease. Arthrocentesis of affected joints may be necessary to rule out septic arthritis and crystalline disease, and will show elevated synovial white blood cell counts with negative cultures.

Treatment

As with ankylosing spondylitis, ideal management involves both pharmacologic and nonpharmacologic treatment options. However, no clinical guidelines are presently available for reactive arthritis.
The use of antibiotics to treat the initial infectious cause is recommended. Some studies have evaluated the use of antibiotics in the treatment of the reactive arthritis itself, but results have been mixed. Patients should be referred to specialists to address their extraarticular symptoms. Pharmacologic management mirrors the treatment for ankylosing spondylitis, with NSAIDs being the first-line choice. Oral corticosteroids are generally considered helpful in refractory cases or severe disease. Intraarticular corticosteroid injections in affected joints are also beneficial. For patients who do not respond to NSAIDs, use of DMARDs is not well established in terms of efficacy but may be considered. Anti-TNF therapy similarly may be considered for patients who have not responded to other therapies, but their efficacy is not as well established when compared with results in patients with ankylosing spondylitis.

Nonpharmacologic treatment should be targeted toward symptom management. In particular, heat and cold modalities and range-of-motion exercises are preferred to preserve mobility. In chronic disease, bracing may be necessary to prevent contracture and reduce pain. One of the potential complications of reactive arthritis is muscle wasting around affected joints; therefore, strengthening exercises may be necessary to build up affected areas. In general, surgical intervention in reactive arthritis is rare.


3. Psoriatic Arthritis

General Considerations
Psoriatic arthritis is an inflammatory arthritis involving the peripheral joints that occurs in combination with psoriasis of the skin. The condition is rare within the general population, but in patients with psoriasis the occurrence is anywhere from 5% to 30%. The male-to-female ratio is equal, and peak incidence is between 35 and 55 years of age. A juvenile form also exists, having a peak incidence between the ages of 9 and 11 years. In most patients (60–80%), psoriasis precedes the arthritis symptoms, but for approximately 15%, the arthritis symptoms manifest first.

Clinical Findings

The Classification Criteria for Psoriatic Arthritis (CASPAR), which are extremely sensitive, are commonly used in evaluating patients for the disorder (Table 22–12). A positive diagnosis can be made on the basis of established peripheral inflammatory arthritis coupled with any three features from the list of criteria.

<table>
<thead>
<tr>
<th>Current psoriasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of psoriasis (without active disease)</td>
</tr>
<tr>
<td>Family history of psoriasis (without current or previous disease)</td>
</tr>
<tr>
<td>Dactylitis</td>
</tr>
<tr>
<td>Juxtaarticular new bone growth</td>
</tr>
<tr>
<td>Rheumatoid factor negative</td>
</tr>
<tr>
<td>Nail dystrophy</td>
</tr>
</tbody>
</table>

Table 22–12 CASPAR criteria for psoriatic arthritis.


Patients present with erythematous, scaly plaques on the skin consistent with psoriasis. Joint deformity and swelling from inflammatory arthritis tends to occur most commonly in the small joints of the wrists, hands, ankles, and feet, although they can occur anywhere. In the fingers and toes, erosion leads preferentially to flexion deformity. Extraarticular manifestations, such as uveitis
or conjunctivitis, can occur in up to 30% of psoriatic arthritis patients, which is a lower frequency than in ankylosing spondylitis.

Radiographs of affected joints show erosive changes, such as the classic “pencil-in-cup” appearance of the digits. Unlike patients with RA, those with psoriatic arthritis may also demonstrate new bone growth in the form of either ankylosis within the joint or heterotopic bone formation outside the joint.

While not necessary to establish diagnosis, serum ESR and CRP levels tend to be high and track the disease state well. Arthrocentesis is similarly not required for diagnosis but will show elevated synovial white blood cell counts with a majority of polymorphonuclear leukocytes.

**Treatment**

Management of psoriatic arthritis requires treatment of both the skin and the peripheral joints simultaneously. The EULAR 2012 guidelines on management of psoriatic arthritis recommend a combination of pharmacologic and nonpharmacologic therapies. As with other spondyloarthropathies, extraarticular symptoms should be managed by an appropriate specialist. For pharmacologic pain management, NSAIDs remain the first-line choice, with opioid analgesics as second line. Oral corticosteroids are not recommended as they may cause rebound worsening of the psoriasis. Skin plaques should be addressed with topical medications such as retinoic acid, or psoralen plus ultraviolet light therapy. The use of DMARDs such as methotrexate, sulfasalazine, and cyclosporine is recommended with proper patient selection and monitoring. The use of anti-TNF agents is recommended only for refractory patients with moderate to severe disease, and efficacy is not as well established as for ankylosing spondylitis.

Nonpharmacologic therapies should focus on symptomatic management. In particular, splinting and bracing of the hands may reduce pain and prevent contracture. Heating and cooling modalities are beneficial for disease flares and to treat inflammation. Both active and passive range of motion and isometric exercise are critical for preserving function, but precautions should be taken in eroded joints to prevent dislocation or fracture. Patient education in energy conservation may also be beneficial. For cases of severe contracture, surgical intervention may be necessary for arthroplasty, arthrodesis, or tendon lengthening. Synovectomy is recommended for chronic severe synovitis involving a single joint. Joint replacement surgery is also recommended if warranted through standard orthopedic guidelines.
4. Spondyloarthritis Associated with Inflammatory Bowel Disease

General Considerations

Spondyloarthritis associated with inflammatory bowel disease (sometimes referred to as enteropathic arthropathy) is a complication of ulcerative colitis and Crohn’s disease, affecting between 5% and 20% of these patients. In the general population, the incidence is rare. The male-to-female ratio is equal, and the disease affects whites more than other races. Peak incidence is between the ages of 15 and 35 years.

Clinical Findings

Patients present clinically in one of two ways, with symptoms that are either axial predominant or peripheral predominant. Those with axial-predominant disease have low back pain and stiffness that is improved with activity and worsened by prolonged positions. Arthritis symptoms are typically not tied to bowel symptoms. Peripheral-predominant disease is typically nonerosive and nondeforming, unlike reactive or psoriatic arthritis. This entity itself can be divided into two types, pauciarticular and polyarticular. The pauciarticular type tends to involve large joints, is asymmetric, and produces acute, self-limiting attacks. In this type, there is also a close correlation between bowel and joint symptoms. The polyarticular type, by contrast, tends to involve the small joints of the hands and feet, is more symmetric, and is chronic in nature. In this type, the bowel and joint symptoms are usually independent of one another. Regardless of type, spondyloarthritis associated with inflammatory bowel disease also includes extraarticular manifestations, namely, enthesopathy, uveitis,
fever, secondary amyloidosis, and skin involvement.

The diagnosis of spondyloarthropathy associated with inflammatory bowel disease is made clinically based on history and physical examination findings. With the exception of radiographs, which may be helpful in evaluating the sacroiliac joints in axial-type disease, imaging is seldom helpful. Laboratory studies show elevated serum ESR and CRP levels, which help to monitor the disease process but are not necessary for diagnosis. From 30% to 70% of affected patients are positive for HLA-B27; therefore, this marker is not a reliable tool for diagnosis. Arthrocentesis may be necessary to rule out septic arthritis or crystalline disease; synovial fluid analysis shows elevated white blood cell counts with negative cultures.

**Treatment**

No guidelines currently exist for the management of spondyloarthropathy associated with inflammatory bowel disease. A general recommendation for management is to treat the underlying bowel disease aggressively as this appears to have a positive impact on joint disease. Referral to a gastroenterologist is required to accomplish this goal. Although NSAIDs are generally acceptable for pharmacologic treatment of joint symptoms, in this case they may exacerbate bowel symptoms, and use of selective COX-2 inhibitors may be preferential. Oral or intraarticular corticosteroids are both recommended for management of acute inflammation. Among DMARDs, sulfasalazine is beneficial for peripheral-predominant but not axial-predominant disease. Methotrexate is used as an adjunct medication for inflammatory bowel disease to address bowel symptoms, but its efficacy in joint symptoms is unclear. The use of anti-TNF agents is indicated for severe or refractory cases, although its efficacy is not clearly established.

Nonpharmacologic therapies should be targeted toward maintaining mobility, flexibility, and postural control in axial-predominant disease, and symptomatic management in peripheral-predominant disease. Heating and cooling modalities may be helpful to this end. Because the disease process is nondeforming and nonerosive, the use of bracing or more aggressive measures is seldom required. For severe cases, total colectomy or surgical removal of affected bowel leads to remission of joint disease symptoms in peripheral-predominant disease associated with ulcerative colitis but not Crohn’s disease. Bowel surgery does not seem to have an effect on axial-predominant disease.
5. Undifferentiated Spondyloarthropathy

General Considerations

Undifferentiated spondyloarthropathy refers to disease processes that do not meet the diagnostic criteria for any of the preceding conditions, despite similar clinical presentation. Exact incidence and epidemiology is difficult to ascertain due to the nebulous nature of the condition.

Clinical Findings

The criteria that have been adopted to make this diagnosis are the Modified Amor Diagnostic Criteria. Using the inclusion criteria in Table 22–13, six points are required to make a diagnosis of undifferentiated spondyloarthropathy. If a patient has any of the following exclusion criteria, however, other causes must be examined: diagnosis of a specific spondyloarthropathy, radiographic evidence of grade 2 or higher sacroiliitis, precipitating genitourinary or gastrointestinal infection, psoriasis, keratoderma blennorrhagicum, inflammatory bowel disease, positive rheumatoid factor, and positive antinuclear antibody (ANA) titers.

**Table 22–13** Modified Amor diagnostic criteria for undifferentiated spondyloarthropathy.
No specific guidelines exist for the treatment of undifferentiated spondyloarthropathy, and management recommendations are the same as for ankylosing spondylitis, discussed earlier.

The mixed connective tissue disorders are a group of autoimmune diseases that manifest various clinical features characteristic of several different rheumatologic diseases. The most common of these disorders are lupus erythematosus, scleroderma, and polymyositis.

1. Systemic Lupus Erythematosus

**ESSENTIALS OF DIAGNOSIS**

- Chronic, progressive disease that affects every organ in the body.
- Joint pain is the most common symptom.
- Most patients are women who are diagnosed in early adulthood; however, lupus is also the most common connective tissue disease seen in children.

**General Considerations**

Systemic lupus erythematosus (SLE) is an autoimmune disease that produces a constellation of symptoms having the potential to affect every organ in the body. Joint arthralgias are common, as well as cutaneous, musculoskeletal, renal, neurologic, cardiovascular, pleural, pulmonary, and neuro-psychiatric
manifestations. SLE is nine times more common in women than in men and is also the most common major connective tissue disease seen in children. The prevalence is approximately 130 cases per 100,000 people in the United States, with African Americans, Hispanics, and Asians more frequently affected than non-Hispanic whites. Patients are most often diagnosed in early adulthood, and the progressive impairment of multiple body systems has significant implications for all areas of a patient’s life.

Clinical Findings

The ACR has published 11 criteria that outline the clinical manifestations of the disease (Table 22–14). Four or more of the 11 criteria must be met for a positive diagnosis.

Table 22–14 American College of Rheumatology criteria for classification of systemic lupus erythematosus (SLE).
<table>
<thead>
<tr>
<th>Criteria</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malar rash</td>
<td>Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds</td>
</tr>
<tr>
<td>Discoid rash</td>
<td>Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions</td>
</tr>
<tr>
<td>Photosensitivity</td>
<td>Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation</td>
</tr>
<tr>
<td>Oral ulcers</td>
<td>Oral or nasopharyngeal ulceration, usually painless, observed by physician</td>
</tr>
<tr>
<td>Nonspecific arthritis</td>
<td>Involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion</td>
</tr>
</tbody>
</table>
| Pleuritis or pericarditis     | 1. Pleuritis—convincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion  
Or  
2. Pericarditis documented by electrocardiogram or rub or evidence of pericardial effusion |
| Renal disorder               | 1. Persistent proteinuria > 0.5 g/day or > than 3+ if quantitation not performed  
Or  
2. Cellular casts—may be red cell, hemoglobin, granular, tubular, or mixed |
| Neurologic disorder           | 1. Seizures in the absence of offending drugs or known metabolic derangements; eg, uremia, ketoacidosis, or electrolyte imbalance  
Or  
2. Psychosis in the absence of offending drugs or known metabolic derangements, eg, uremia, ketoacidosis, or electrolyte imbalance |
| Hematologic disorder          | 1. Hemolytic anemia with reticulocytosis  
Or  
2. Leukopenia < 4000/mm³ on ≥ 2 occasions  
Or  
3. Lymphopenia < 1500/mm³ on ≥ 2 occasions  
Or  
4. Thrombocytopenia < 100,000/mm³ in the absence of offending drugs |
| Immunologic disorder          | 1. Anti-DNA: antibody to native DNA in abnormal titer  
Or  
2. Anti-Sm: presence of antibody to Sm nuclear antigen  
Or  
3. Positive finding of antiphospholipid antibodies on:  
a. an abnormal serum level of IgG or IgM anticardiolipin antibodies  
b. a positive test result for lupus anticoagulant using a standard method, or  
c. a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test |
| Positive antinuclear antibody (ANA) test | An abnormal titer of ANA by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs |

Note: The classification is based on 11 criteria. For the purpose of identifying patients in clinical studies, a person is defined as having SLE if any 4 or more of the 11 criteria are present, serially or simultaneously, during any interval of observation. Data from Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus [letter]. Arthritis Rheum 1997;40:1725.
A. Symptoms and Signs

1. Musculoskeletal and cutaneous—Musculocutaneous manifestations are the most common findings. Cutaneous lupus erythematosus (CLE) manifests in a variety of clinical forms, with three recognized subtypes: acute (ACLE), subacute (SCLE), and chronic (CCLE). ACLE may be localized, most often producing a malar (“butterfly”) rash over the face, or generalized. Patients with SCLE are highly photosensitive, with symptoms that are distributed predominantly on the upper back, shoulders, neck, and anterior chest. Classic discoid lupus erythematosus is the most common form of CCLE; patients present with indurated scaly plaques on the scalp, face, and ears, and with characteristic scarring and pigmentary change.

Musculoskeletal manifestations include painful joints, with or without erythema and synovitis. Almost all patients with SLE present with painful joints out of proportion to the amount of joint synovitis. Pain is usually symmetric and involves small joints of the hand, wrists, and knees but can involve any peripheral joint in the body. When effusions exist, they are small. In some patients, there is a progression to rheumatoid-like nonerosive hand deformities (Jaccoud’s syndrome). The major disabling type of joint disease in lupus is articular osteonecrosis, often induced by high-dose corticosteroids. Rare musculoskeletal manifestations of lupus include spontaneous tendon rupture, crystalline arthropathies, subcutaneous calcifications, and inflammatory myopathy. Fatigue is the most functional problem encountered.

2. Renal—Approximately 10% of patients with SLE develop end-stage renal disease (ESRD). In most patients, systemic disease activity diminishes as ESRD approaches. Consequently, the survival of SLE patients on dialysis (both hemodialysis and peritoneal) appears to be comparable to that of non-SLE patients.

3. Cardiovascular—Cardiac manifestations are common in patients with SLE. Pericarditis is the most frequent manifestation, followed by valvular and coronary arterial disease. Accelerated atherosclerosis is an important cause of mortality and morbidity in this disease. This pattern was confirmed in autopsy and epidemiologic studies, which identified hypercholesterolemia (and particularly its persistence in the first 3 years of disease), hypertension, and lupus itself as important risk factors for the development of accelerated atherosclerosis in these patients fueled by corticosteroid therapy.
4. **Pulmonary and respiratory**—Involvement of the respiratory system is common in patients with SLE. Pleuropulmonary manifestations are present in nearly 50% of patients during the disease course and may be the presenting symptoms in 4–5% of patients with SLE. Complications directly associated with the disease include pleuritis with or without pleural effusion, alveolitis, interstitial lung disease, lupus pneumonitis, pulmonary hemorrhage, pulmonary arterial hypertension, and pulmonary thromboembolic disease.

5. **Neurologic and psychiatric**—Neuropsychiatric syndromes in SLE include headache, seizures, cerebrovascular disease, psychosis, cranial neuropathy, and movement disorder. A higher prevalence of mood disorders and cognitive dysfunction has been demonstrated in studies involving systematic assessment of cognitive and psychiatric function. Psychiatric and neuropsychiatric evaluations should therefore be considered and cognitive remediation strategies implemented, if appropriate.

**B. Laboratory Findings**

Laboratory studies demonstrate a positive ANA titer. The presence of antibodies to double-stranded DNA (anti-dsDNA) is diagnostic, particularly in patients with renal disease, and can be used to predict flares. Although other antibodies may be present, they are generally not used to monitor disease progress. Thus, identification of new biomarkers to improve diagnostic potential is of great interest; among the most promising candidates are B-lymphocyte stimulator (BLyS), antiribosomal P protein, and anti-C1q antibodies.

Pericardial fluid levels tend to be small, and electrocardiographic, echocardiographic, and pathologic findings show abnormalities. Echo-Holter electrocardiography and isotopic myocardial perfusion scan techniques are useful in the clinical evaluation of patients with cardiac symptoms. At least 50% of patients with SLE have renal disease marked by the presence of proteinuria. Renal biopsy results are usually abnormal and are a good indication of prognosis.

**Treatment**

Corticosteroids have long been the cornerstone of therapy in SLE, and the addition of cyclophosphamide has contributed to preservation of renal function in patients with severe proliferative glomerulonephritis. More recently, in an effort to minimize drug toxicity and achieve equal effectiveness, other
immunosuppressive agents, including mycophenolate mofetil, have been introduced.

Rehabilitation measures should be geared toward addressing deconditioning and improving energy conservation techniques. Reducing inflammation is one of the major treatment strategies in these diseases. Exercise training has emerged as a potential therapeutic tool in counteracting systemic inflammation, thereby leading to better clinical outcomes. Pain should be addressed and managed during the rehabilitation process so as not to impede progress. Bracing to prevent or correct joint deformities should be implemented when necessary, and patients should be taught joint protection strategies.

During rehabilitation, cardiac abnormalities should be closely monitored with attention to cardiac precautions. Despite having a decreased cardiac capacity, SLE patients showed improvement in exercise tolerance, aerobic capacity, quality of life, and depression after a supervised cardiovascular training program. The fatigue resulting from both cardiac and renal disease can be a limiting factor in the rehabilitation of the patient. Antiembolic stockings (TEDS) may be helpful in managing peripheral edema in these patients.


2. Systemic Sclerosis

Systemic sclerosis, also known as scleroderma, is an autoimmune multisystemic disease marked by fibrosis of the skin and microvascular damage that can affect internal organs. Its incidence in the United States is about 19 cases per million per year. Systemic sclerosis is a disease that attacks predominately women of childbearing years but has been demonstrated in men also. It is believed that age, gender, and genetic background play a role in the disease. Survival rate is negatively impacted by older age of onset, male sex, scleroderma renal crisis, pulmonary fibrosis, pulmonary arterial hypertension, cancer, and the presence of antitopoisomerase and anti-U1 antibodies.

Rehabilitation strategies for systemic sclerosis should include preventing skin contractures as the disease progresses. The hands are of particular concern, because the disease is commonly accompanied by vascular hypoperfusion and neuropathy. Splinting may help early on in the disease but can become painful as the disease progresses. Range-of-motion exercises should be avoided during acute synovitis. Judicious use of modalities may help to alleviate the pain associated with the disease. However, careful skin precautions should be used to prevent burning or freezing the skin because of insensate extremities.
Polymyositis and dermatomyositis are characterized by proximal muscle weakness.

Inclusion body myositis is characterized by distal weakness.

---

**General Consideration**

Inflammatory myopathies can be classified into three major groups: polymyositis, dermatomyositis, and inclusion body myositis. These disorders were reviewed in Chapter 18, where they were contrasted with other disorders that cause myopathy. In both polymyositis and dermatomyositis, patients present with gradual proximal symmetric weakness over many months; those with dermatomyositis also have skin manifestations. Inclusion body myositis is rare, occurring primarily in men after 50 years of age. It has a slow, insidious onset that manifests as symmetric distal involvement.

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**Clinical Findings**

**A. Symptoms and Signs**

In both polymyositis and dermatomyositis, patients have normal sensation and preserved deep tendon reflexes until late in the disease. Pharyngeal and neck flexor muscles are often involved, resulting in dysphagia and head drop. Ocular muscles are most always spared. Weak intercostal muscles may cause respiratory problems late in the disease.

1. **Polymyositis**—Approximately one third of all patients with inflammatory myopathies present with primary idiopathic polymyositis. The disease course is characterized by slow onset, proximal muscle weakness, and muscle tenderness. Cardiac conduction disturbance occurs in 30% of patients, and 10% of patients develop interstitial lung disease, such as alveolitis and fibrosis. Systemic features include fever, malaise, weight loss, arthralgias, and Raynaud’s phenomenon.

2. **Dermatomyositis**—Primary idiopathic dermatomyositis produces the findings seen in polymyositis, along with skin lesions. Most common is a heliotropic, reddish, violaceous eruption on the upper eyelids. However, localized erythema over exposed areas of the chest, neck, chin, and malar region
may occur. Gottron’s sign, a violaceous erythematous eruption over the knuckles, is also characteristic. Underlying malignancy of the breast, lung, ovary, and stomach, and myeloproliferative disorders may be present in the adult form, usually in patients older than 60 years.

3. **Inclusion body myositis**—Patients with inclusion body myositis demonstrate thinning of the forearms and atrophy of the finger flexors and ankle dorsiflexors. Late in the disease, dysphagia and respiratory failure are common.

4. **Pediatric onset**—From 8% to 20% of all cases of myositis involve children. Pediatric patients with the disorder almost always present with a rash. Pediatric disease is associated with vasculitis of the skin, kidney, gastrointestinal tract, muscle, and brain. Subcutaneous calcifications are frequently present in children with dermatomyositis. Polymyositis or dermatomyositis may be associated with other connective tissue disorders, such as RA, SLE, systemic sclerosis, and mixed connective tissue disease.

**B. Laboratory Findings**

A chemistry panel shows elevated creatine kinase levels, usually greater than 10 times normal. Aspartate aminotransferase (AST), alanine aminotransferase (ALT), lactate dehydrogenase (LDH), and aldolase levels are also elevated. ANA testing is positive in 80% of patients.

**C. Diagnostic Studies**

Needle electromyography shows irritability of myofibrils on needle insertion, as well as fibrillations, positive sharp waves, complex repetitive discharges, and small, brief-duration polyphasic motor unit potentials with early recruitment. Muscle biopsy findings for patients with polymyositis reveal inflammatory infiltrates within the fascicle, with invasion of the individual muscle fiber.

**Treatment**

Exercise can be an important part of treatment in patients with idiopathic inflammatory myopathies, although bed rest may be required during severe inflammation when passive range-of-motion progressing to active range-of-motion exercises suffice. Improvements in overall function, ability to perform activities of daily living, and health-related quality of life have been reported
during remissions in adult patients with polymyositis, dermatomyositis, and, recently, inclusion body myositis, following different exercise regimens, with no signs of increased muscle inflammation. Exercise has also proved beneficial in children with inflammatory myopathies during remission periods of their disease.

It is possible that resistance training may reduce clinical disease activity and reduce expression of genes regulating inflammation and fibrosis in chronic polymyositis and dermatomyositis. Creatine kinase levels can be used to monitor patients’ muscle function when exercising, with the goal of preventing rhabdomyolysis. Swallowing and speech evaluations are important in later stages of the disease.

In the case of inclusion body myositis, forearm bracing and ankle foot orthoses may improve physical function. Unfortunately, patients with inclusion body myositis generally have a limited response to conventional treatments such as corticosteroids.


VASCULITIDES

The vasculitides are a set of related disorders characterized by blood vessel inflammation and destruction caused by inflammatory mediators that lead to end-organ injury. Although more common in adults, these disorders can also occur in childhood. The pathophysiology of these diseases is not well understood and is difficult to classify. In general, vasculitides can be classified into small, medium, or large vessel vasculitis.

Early diagnosis is critical in the rehabilitation of patients with vasculitides, especially giant cell arteritis. Glucocorticosteroids, which have widespread antiinflammatory effects, are the initial treatment for most of these disorders. Consequently, patients may experience significant side effects, including hyperglycemia, immune compromise, avascular necrosis, myopathy, hypertension, and cataract formation. Pain should be treated aggressively.
ESR and CRP values can be used to monitor the efficacy of treatments. Bone scans can be helpful in determining avascular necrosis, and dual X-ray absorptiometry (DEXA) scans can be used to monitor osteopenia. The patient’s symptoms offer the most reliable method of monitoring both disease progression and the patients’ ability to participate in exercise. Special precautions should be made to monitor for peripheral neuropathies, especially when using physical modalities. In patients with lung involvement, pulmonary function studies can help direct the rehabilitation program.

1. Polyarteritis Nodosa

Polyarteritis nodosa (PAN) is necrotizing inflammation of medium-sized or small arteries that spares the arterioles, venules, and capillaries. Early in the disease, patients may report nonspecific complaints of arthralgias, fatigue, and malaise. PAN can attack multiple organs, including the skin, peripheral nerves, gastrointestinal tract, and kidneys but usually spares the lungs. It is not associated with glomerulonephritis.

Vasculitic neuropathy is a common early sign of PAN. Clinically patients have a subacute, progressive, generalized but asymmetric, painful, sensorimotor polyneuropathy. However, mononeuritis multiplex affecting any nerve can be expected.

The diagnosis of PAN usually requires a tissue or nerve biopsy if neuropathy is evident. Therapeutic treatment includes corticosteroids followed by immunosuppressive drugs for patients with severe symptoms.

2. Giant Cell Arteritis

General Considerations

Giant cell arteritis (GCA) is a vasculitis affecting medium-and large-sized vessels. It has a predilection for the aorta and its branches. The term giant cell arteritis is often used interchangeably with temporal arteritis and cranial arteritis, but this is inaccurate. Although GCA usually involves the superficial temporal artery and other extracranial branches of the carotid, involvement of the aorta and its large branches is also common. Additional vascular manifestations include stroke, aortic aneurysm or dissection, and even aortic rupture. Cardiac manifestations include coronary artery disease, aortic valve insufficiency, or left ventricular dysfunction, which may occur independently from the valvular disease or hypertension.

GCA is the most common primary vasculitis in adults, affecting those older than 50 years of age. Incidence is about 18 per 100,000 per year but increases with age and reaches its peak in the eighth decade of life. The prevalence of GCA is highest in northern latitudes and in individuals of northern European descent. The disease affects significantly more whites than blacks and is two to six times more common in women than in men.

Clinical Findings

A. Symptoms and Signs

Headache is the most common presenting symptom. Nonspecific symptoms, such as malaise, wasting, myalgias, stiffness, and jaw claudication are common early in the disease course. Aortic aneurysm, dissection, and large vessel stenoses are complications of GCA that occur in approximately one third of patients. The worst complication of GCA is permanent visual loss. There is a strong correlation between polymyalgia rheumatica (described next) and symptomatic GCA. However, up to 20% of patients present without systemic symptoms, seeking evaluation of visual problems. If these patients elude early detection, the result is usually permanent visual loss. Early signs of blurred vision or transient visual loss, amaurosis fugax, and jaw claudication can be predictive, allowing for early diagnosis and treatment.
B. Laboratory and Diagnostic Studies

Laboratory findings, including elevated ESR and CRP, and thrombocytosis, may be helpful for diagnosis. However, the gold standard for diagnosis of GCA is a temporal artery biopsy that shows inflammation of the artery.

Complications

Aortic aneurysm, aortic dissection, and large vessel stenoses are complications that occur in approximately one third of patients. Other vascular complications include stroke and aortic rupture. Cardiac manifestations include coronary artery disease, aortic valve insufficiency, and left ventricular dysfunction, which may occur independently from the valvular disease or hypertension. The most common preventable complication of GCA is permanent visual loss; diagnosis is critical, and onset of symptoms is considered a medical emergency. Laboratory findings such as elevated ESR, CRP, and thrombocytosis may be helpful; however, the gold standard for diagnosis of GCA is temporal artery biopsy.

Treatment

Glucocorticosteroids are the mainstay of treatment for GCA and should be initiated immediately and aggressively to prevent or limit visual loss and ischemic stroke. Treatment should begin immediately, before performing the temporal artery biopsy. Unfortunately, treatment may prevent but not reverse the visual loss. However, when treatment is initiated within 24 hours of visual symptoms, 58% of patients have visual improvement, compared with only 6% of patients who improve after a delay in treatment. Once the symptoms are stabilized, a taper of the glucocorticosteroid dosage is recommended to minimize or forestall potential drug-related side effects, described earlier.

Melson MR, Weyand CM, Neuman NJ, Biouss V: The diagnosis of giant cell...


**POLYMYALGIA RHEUMATICA**

Polymyalgia rheumatica is the most common inflammatory rheumatic disease of elderly non-Hispanic whites. The cause is unknown, but a combination of genetic and environmental factors is believed to be involved. GCA is seen in about 30% of patients with polymyalgia rheumatica. Similar to that disease, polymyalgia rheumatica affects people over 50, and prevalence increases with advancing age. The average age of onset is 70 years, and 75% of patients are female. Most patients are of northern European ancestry, but any ethnic group may develop the disease.

The most common presenting feature is proximal muscle pain involving the bilateral shoulder and hip girdle, bilateral upper arm, and posterior neck musculature. Pain may be described as a constant, unremitting achy pain and patients may have difficulty arising from a chair or climbing stairs. Serologic testing demonstrates elevated ESR and CRP.

The only proven treatment is glucocorticosteroids. As noted earlier, side effects of such treatment can be significant; therefore, patients should be monitored closely to evaluate both response to treatment and drug-induced side effects. Within 3–4 weeks patients should report at least a 70% global improvement, and the ESR and CRP values should normalize.

**SJÖGREN’S SYNDROME**
Sjögren’s syndrome is a common autoimmune disease involving the connective tissue that has broad organ-specific and systemic manifestations. The incidence is much higher in women than in men and is higher in the elderly population, although individuals at any age may be affected. The average age of onset is the mid-50s. Individuals with primary Sjögren’s syndrome have an approximately 20 times increased risk of developing malignancies, specifically non-Hodgkin’s lymphoma, compared with the general population. There is also an association with other rheumatologic and autoimmune diseases, including fibromyalgia, autoimmune thyroid disease, multiple sclerosis, and spondyloarthropathy.

The most common presenting symptoms are xerophthalmia (dry eyes) and xerostomia (dry mouth), keratoconjunctivitis sicca, and parotid gland enlargement. Organs such as the lung and kidney (interstitial nephritis), as well as neurologic (central and peripheral) and gastrointestinal function, may also be affected. Parotid gland swelling may be palpable or visible on examination. Focal mononuclear cell infiltration of exocrine tissues and the presence of autoantibodies (anti-SSA/Ro, anti-SSB/La, and RF) are diagnostic.

The eyes may be tested by performing a Schirmer’s test. The test result is positive when there is less than 5 mm of wetting on a rectangular strip of filter paper. Ocular staining using rose Bengal or lissamine green can be used in scoring the results.

The rehabilitation of patients with Sjögren’s syndrome is primarily symptomatic. In primary Sjögren’s syndrome, pilocarpine and cevimeline have demonstrated efficacy for sicca features and topical cyclosporine for moderate or severe dry eye. Patients may require a swallowing and speech evaluation. Underlying autoimmune diseases as well as malignancy should be investigated.


**FIBROMYALGIA**
General Considerations

Fibromyalgia is a poorly understood disease that manifests with symptoms of widespread pain, muscle aches, and fatigue. It is the second most common disorder seen by rheumatologists, after RA. Fibromyalgia affects about 2% of the population, or an estimated 5 million adults in the United States. The disease is more common in women than men; in the US population, the prevalence of fibromyalgia in women is about 3.4%.

Many theories have been proposed to explain the pathogenesis of fibromyalgia; current thinking points to a combination of genetic, environmental, and psychosocial factors that precipitate onset of the disease. Convincing evidence, although small, suggests that pain results from increased sensitization of the central nervous system that usually begins with a physical or mental trauma. Fibromyalgia patients have an increased level of substance P in cerebral spinal fluid, as well as functional magnetic resonance imaging (fMRI) evidence of augmented pain processing. Thus, fibromyalgia may have more similarities to a chronic neuropathic pain syndrome than a typical rheumatologic disorder.

Clinical Findings

Characteristic clinical features of fibromyalgia include widespread body aches, pains, tenderness, fatigue, cognitive problems, sleep disturbance, body stiffness, depressive and anxiety symptoms, and impaired social and occupational functioning. Nonrestorative sleep is a common complaint, and a phasic α sleep activity pattern seen on electroencephalographic testing correlates with the clinical manifestations of fibromyalgia. The onset and course of the disease can be very variable; however, there is usually a significant delay in diagnosis and treatment.

Patients usually present with the complaint of “total body pain.” In 1990, the ACR defined the following criteria for diagnosis: a history of widespread pain lasting more than 3 months; pain existing in all four quadrants of the body (upper and lower, left and right); axial skeleton pain; and pain elicited to 4 kg of pressure specific to 11 of 18 predetermined tender points. By 2010, it was believed that tender point testing was laborious and underutilized in primary care centers and that more emphasis should be placed on patient symptoms. Consequently, the ACR proposed preliminary diagnostic criteria for fibromyalgia that abandoned the tender point count and placed increased
emphasis on patient symptoms in the form of a questionnaire. A later modification of the ACR 2010 criteria for use in surveys employed a self-report questionnaire (Fibromyalgia Survey Questionnaire [FSQ]) to assess patient symptoms. This has resulted in a quicker, symptom-focused diagnostic evaluation for fibromyalgia.

Diagnosis of fibromyalgia requires the exclusion of other rheumatologic and neurologic diseases. Patients with fibromyalgia commonly have many comorbid medical conditions, including painful neuropathies, circulatory disorders, depression, diabetes, and sleep disorders. A typical rheumatologic workup should be considered prior to confirming the diagnosis of fibromyalgia. Diseases to exclude are RA, cervical and lumbar radiculopathy, lupus, Lyme disease, HIV, hepatitis, hypothyroidism, and sleep apnea.

### Treatment

Because the disease course for most patients is extremely variable, ranging from mild to moderate daily pain to extreme pain resulting in functional incapacity, the rehabilitation of patients with fibromyalgia is complex and requires a multisystem approach tailored to the individual patient. The treatment goals should be to decrease pain, improve function, and restore sleep. It is critical that the patient’s family be involved in the treatment plan. The likelihood of a functional recovery is optimized for patients who receive early diagnosis and are motivated to participate in a multidisciplinary program geared to treatment of pain, depression, and rehabilitation.

**A. Pharmacotherapy**

Many analgesics and other classes of drugs have been used for symptom management, with varying degrees of success. These include opiates, NSAIDs, antidepressants, antispasmodics, and anticonvulsant agents. Pregabalin, duloxetine, and milnacipran have demonstrated efficacy in pain reduction and are indicated for use in fibromyalgia. These agents can be used in conjunction with other pharmacologic agents and treatments. Although opiate medications are widely prescribed, their use in the treatment of fibromyalgia remains controversial. Tramadol, a combination drug with weak μ-agonist activity, has demonstrated some efficacy in pain relief in fibromyalgia patients.

**B. Nonpharmacologic Measures**
Aerobic exercise is still the gold standard in the treatment of fibromyalgia. It is important to develop exercise programs that are tailored to the patient’s symptoms. A review of various aerobic exercise programs for fibromyalgia patients demonstrated that the greatest effect and lowest attrition occurred in programs of lower intensity compared with those of higher intensity. Aquatic therapy has been widely used for fibromyalgia, as well as most rheumatologic diseases. In one study, aquatic exercise not only decreased the pain and symptoms of fibromyalgia but also improved cognitive function. Balneotherapy, water immersion or spa therapy, is widely used in fibromyalgia and has also demonstrated efficacy in pain relief.

Other nonpharmacologic treatments that have shown some success in alleviating symptoms in fibromyalgia include tai chi, acupuncture, and cognitive and operant behavioral therapy. Osteopathic physicians have had some success with osteopathic manipulative treatment (OMT) used in conjunction with other forms of standard medical care. Finally, a newer and promising modality is repetitive transcranial magnetic stimulation (rTMS) of the primary motor cortex, which was initially studied in patients with depression. Although data are scarce, there is some evidence that patients with fibromyalgia may have improvement in both pain and depression.

Hauser W, Wolfe F: Diagnosis and diagnostic tests for fibromyalgia


Cardiac Rehabilitation

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Jamie Schmeer, DO

EPIDEMIOLOGY OF HEART DISEASE

Cardiovascular disease continues to be the leading cause of mortality in the United States for men and women, accounting for one third of all deaths. Nearly 16.3 million Americans have a history of coronary artery disease and 7.9 million have suffered a myocardial infarction. Direct and indirect costs of this condition approach $190.3 billion annually. That is $95.5 billion in direct costs (physicians and other professionals, hospital services, medication, and home health care) and $94.8 billion in indirect costs (lost productivity and mortality). Medical costs for coronary heart disease are projected to increase by 200% over the next 20 years, augmenting the need for widespread availability of cost-effective secondary prevention.

As cardiovascular disease is a substantial cause of disability, physiatrists must be well-versed in all aspects of cardiac rehabilitation. Numerous analyses of evidence-based medical studies have shown cardiac rehabilitation programs to be a safe and effective therapeutic intervention. These secondary prevention programs enhance quality of life and functional status, improve processes of care, and reduce recurrent myocardial infarction, hospitalization, and long-term mortality. Increasingly, cardiac rehabilitation patients have complex medical profiles; thus the need for physiatry involvement is growing. As medical specialists, physiatrists can contribute their unique knowledge and understanding of comprehensive functional assessment to this large patient population, which is in need of such expertise.

OVERVIEW OF CARDIAC REHABILITATION
Cardiac rehabilitation consists of comprehensive long-term services involving medical evaluation, prescribed exercise, cardiac risk factor modification, health education, counseling, and behavioral interventions. Its short-term goals are to control cardiac symptoms, enhance functional capacity, limit unfavorable psychological and physiologic effects of cardiac illness, and boost psychosocial and vocational status. The long-term goals are to alter the natural history of coronary artery disease, stabilize or reverse the progression of atherosclerosis, and lessen the risk of sudden death and reinfarction.

Indications for cardiac rehabilitation are listed in Table 23–1. Cardiac rehabilitation is considered reasonable and necessary in patients with any of the listed indications within the past 12 months. Additionally, patients with congestive heart failure, sustained ventricular tachycardia or fibrillation, and those who are survivors of sudden cardiac death may be candidates for rehabilitation on a case-by-case basis. Overall, more than 18 million Americans meet the requirements for cardiac rehabilitation.

Table 23–1 Indications for cardiac rehabilitation

<table>
<thead>
<tr>
<th>Acute myocardial infarction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stable angina pectoris</td>
</tr>
<tr>
<td>Percutaneous transluminal coronary angioplasty (coronary stenting)</td>
</tr>
<tr>
<td>Coronary artery bypass graft (CABG) surgery</td>
</tr>
<tr>
<td>Heart valve repair or replacement</td>
</tr>
<tr>
<td>Heart transplantation or heart–lung transplantation</td>
</tr>
</tbody>
</table>

**Possible indications:**

- Congestive heart failure
- Sustained ventricular tachycardia or fibrillation
- Survivor of sudden cardiac death

Cardiac rehabilitation is divided into three phases:

- **Phase 1** is the acute inpatient period, ranging from 1 to 14 days subsequent to the adverse cardiac event. The emphasis of this phase is to prevent the deleterious effects of bed rest. Therapy focuses on range of motion and early mobilization, progressing to activities of 5–7 metabolic equivalents (METs).
Upon discharge, the patient enters a transitional period, continuing activities at 5–7 METs, maintaining early mobilization, and gradually increasing endurance and community reintegration. Classically, this time period lasts 6 weeks after acute myocardial infarction to allow for scar formation and ends with a graded exercise stress test. Contemporary approaches allow for exercise training to be initiated sooner than 6 weeks, depending on the medical condition of the patient.

- **Phase 2** is the outpatient, medically supervised, comprehensive secondary prevention program, including exercise training with electrocardiographic monitoring, based on the parameters determined by the graded exercise stress test. It is typically initiated 1–6 weeks after hospital discharge. Individuals usually advance to activities of 7–8 METs.

- **Phase 3** is the independent lifelong maintenance period, consisting of regular exercise and lifestyle modifications.


BENEFITS OF CARDIAC REHABILITATION
Benefits of cardiac rehabilitation include an increase of tolerated METs and maximal oxygen consumption, a reduction of cardiac symptoms (chest pain, dyspnea on exertion, shortness of breath, and fatigue), a decrease in blood pressure and heart rate for any given level of physical activity, an improvement in blood lipid levels, an enhancement of psychosocial well-being, smoking cessation, and mortality reduction.

A meta-analysis of 48 randomized trials (8940 patients) found that exercise was associated with lower cardiac and all-cause mortality, as well as downward trends for revascularization procedures and nonfatal myocardial infarction. A separate meta-analysis of 63 randomized trials (21,295 patients) found that cardiac rehabilitation reduced recurrent myocardial infarction by 17% at 12 months and reduced mortality by 47% at 2 years.

One study of more than 600,000 Medicare patients hospitalized for acute coronary syndrome, percutaneous coronary intervention, or coronary artery bypass graft surgery found 12.2% of the cohort participated in cardiac rehabilitation. At 1 year, there was a 2.2% mortality rate for cardiac rehabilitation participants versus 5.3% for nonparticipants. The benefit persisted at 5 years, when the mortality rate for participants was 16.3% versus 24.6% for nonparticipants. This considerable risk reduction equals or exceeds the benefit of most drug and procedural interventions. Patients who attended 25 or more sessions had a 20% lower 5-year mortality rate than those who attended fewer than 25 sessions.

Another study of 30,000 patients who participated in at least one cardiac rehabilitation session found a dose-response relationship of cardiac rehabilitation upon followup at 4 years. Patients who attended all 36 sessions were 47% less likely to die and 31% less likely to have a myocardial infarction than those who attended only one session, 22% less likely to die and 23% less likely to have a myocardial infarction than those who attended 12 sessions, and 14% less likely to die and 12% less likely to have a myocardial infarction than those who participated in 24 sessions.


PATIENT EVALUATION

Before the patient begins phase 2 cardiac rehabilitation, several studies are recommended to assess risk, modify the treatment plan, and set exercise parameters for cardiovascular conditioning.

Electrocardiography & Echocardiography

An electrocardiogram (ECG) and echocardiogram are performed as screening tools for cardiac abnormalities such as left ventricular hypertrophy or dysfunction, previous myocardial infarction, conduction abnormalities, and valvular dysfunctions.

Exercise Testing

A. Submaximal Exercise Test

A submaximal exercise test is defined as a peak heart rate of 120 beats per minute, or 70% of the maximum predicted heart rate, and is typically performed 4–6 days after acute injury to provide additional data used in prognosis, activity prescription, and evaluation of medical therapy.

B. Graded Exercise Test

A graded exercise test, also referred to as an exercise tolerance test, should be sign- or symptom-limited and not terminated at a predefined target heart rate or work rate. It is performed at 2–3 weeks postinjury to determine risk stratification (assess prevalence of ischemia or arrhythmias, heart rate, and blood pressure
response to exercise), develop an exercise prescription, and quantify functional capacity of the patient. The graded exercise test most commonly utilizes the treadmill. Other modes that are used for patients with arthritis, obesity, or other comorbidities that may preclude an adequate treadmill test include arm ergometry and bicycle ergometry.

Table 23–2 summarizes circumstances in which the graded exercise test might be concluded before the patient reaches a maximal effort. If the test is submaximal, it will yield a lower peak heart rate and a consequential lower training target heart rate. The resultant prescribed exercise intensity will not provide the best possible benefits. In such circumstances, a second graded exercise test at 3–6 weeks postinjury may be performed to ascertain appropriate cardiac rehabilitation parameters.

Table 23–2 Graded exercise test end points.
C. Metabolic Equivalent Level

The peak MET level of patients entering cardiac rehabilitation is 4–6 METs. The
Bruce protocol, which is the most frequently used treadmill protocol, is often not the optimal choice as it begins at 4.6 METs and then increases by 2.5–3.0 METs with each 3-minute stage. Cardiac rehabilitation patients should exercise for about 10 minutes to assess functional capacity, heart rate, and blood pressure; however, they are often only able to complete one to two stages (< 6 minutes) of the Bruce protocol. The Modified Bruce protocol starts at 2.3 METs with 1.7 mph at 0% grade then 1.7 mph at 5% grade, but it proceeds with the same work rate profile as the standard Bruce protocol by stage 3 (1.7 mph and 10% grade, or 5 METs). The Naughton–Balke protocol starts at 2 mph at zero grade, speed is held, and the elevation is increased 3.5% every 3 minutes. This is often used for patients with poor exercise tolerance.

Patients with baseline ECG abnormalities due to left bundle branch block, preexcitation syndrome, left ventricular hypertrophy, digoxin therapy (or related medications), greater than 1 mm of resting ST-segment depression, or electronically passed ventricular rhythm may yield a false-positive graded exercise test. In such individuals and in those who are unable to tolerate a standard graded exercise test, pharmacologic stress imaging is needed.

### Radionuclide Perfusion Imaging

Radionuclide perfusion imaging can be completed after administration of intravenous thallium-201 or 99m-technetium sestamibi. Images obtained immediately after exercise are compared with images taken at rest to confirm reversible ischemia and scar. If a patient is unable to exercise adequately, an intravenous pharmacologic challenge using agents such as dipyridamole, adenosine, or dobutamine is performed that mimics the cardiac response to exercise, followed by imaging. Echocardiography can be used in conjunction with exercise or dobutamine to assess left ventricular function as well as global and regional wall motion abnormalities associated with transient ischemia occurring during cardiac stress and absent during rest. Stress echocardiography is more sensitive than exercise ECG for the diagnosis of myocardial ischemia.

The graded exercise test carries predictive value for both cardiac and all-cause mortality. Each 1-MET increase in exercise capacity is associated with a 12% improvement in survival. In addition to mortality, the graded exercise test can be used to stratify the patient’s risk for cardiac events during exercise (Table 23–3).
Table 23–3 Stratification of risk for cardiac events during exercise participation.

<table>
<thead>
<tr>
<th>Low Risk (all must be present)</th>
<th>Moderate Risk (≥ 1 present)</th>
<th>High Risk (≥ 1 present)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>EST/Recovery Findings</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No angina or symptoms</td>
<td>Angina or symptoms</td>
<td>Angina or symptoms</td>
</tr>
<tr>
<td>No ventricular arrhythmias</td>
<td>≥ 7 METs</td>
<td>&lt; 5 METs</td>
</tr>
<tr>
<td>Normal hemodynamics</td>
<td>Mild to moderate silent</td>
<td>Ventricular arrhythmias</td>
</tr>
<tr>
<td>Functional capacity ≥ 7 METs</td>
<td>ischemia (ST-segment</td>
<td>High silent ischemia</td>
</tr>
<tr>
<td></td>
<td>depression ≥ 2 mm)</td>
<td>(ST-segment depression ≥ 2 mm)</td>
</tr>
<tr>
<td></td>
<td>Abnormal hemodynamics</td>
<td>Abnormal hemodynamics</td>
</tr>
<tr>
<td><strong>Nonexercise Test Findings</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Resting EF ≥ 50%</td>
<td>Resting EF = 40–45%</td>
<td>Resting EF &lt; 40%</td>
</tr>
<tr>
<td>Uncomplicated MI or revascularization</td>
<td></td>
<td>History of cardiac arrest or sudden death</td>
</tr>
<tr>
<td>No ventricular arrhythmias</td>
<td></td>
<td>Complex dysrhythmias at rest</td>
</tr>
<tr>
<td>No CHF</td>
<td></td>
<td>Complicated MI or revascularization</td>
</tr>
<tr>
<td>No ischemia postevent or postprocedure</td>
<td></td>
<td>CHF</td>
</tr>
<tr>
<td>No clinical depression</td>
<td></td>
<td>Ischemia postevent or postprocedure</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Clinical depression</td>
</tr>
</tbody>
</table>

EST, symptom-limited exercise test; METs, metabolic equivalents; EF, ejection fraction; CHF, congestive heart failure; MI, myocardial infarction.


CORE COMPONENTS OF CARDIAC REHABILITATION

The American Heart Association (AHA) and the American Association of Cardiovascular and Pulmonary Rehabilitation have outlined the core components for cardiac rehabilitation programs (Table 23–4). Each component provides an opportunity by which to improve the overall health and functioning of the cardiac patient.

Table 23–4 Core components of a cardiac rehabilitation program.

<table>
<thead>
<tr>
<th>Patient assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipid management</td>
</tr>
<tr>
<td>Hypertension management</td>
</tr>
<tr>
<td>Smoking cessation</td>
</tr>
<tr>
<td>Diabetes management</td>
</tr>
<tr>
<td>Nutritional counseling</td>
</tr>
<tr>
<td>Weight management</td>
</tr>
<tr>
<td>Psychosocial counseling</td>
</tr>
<tr>
<td>Physical activity counseling</td>
</tr>
<tr>
<td>Exercise training</td>
</tr>
</tbody>
</table>

Patient Assessment

Patient assessment begins with a comprehensive history and physical examination, medication review, resting 12-lead ECG, risk profile, patient’s perceived health-related quality of life using a standardized questionnaire, and
graded exercise test. Modifiable and nonmodifiable cardiovascular risk factors are reviewed. The modifiable risk factors include physical inactivity, hypertension, smoking, dyslipidemia, overweight or obesity, diabetes, metabolic syndrome, and the emerging factors of elevated lipoprotein-a, elevated homocysteine, prothrombotic states, and elevated C-reactive protein. The nonmodifiable risk factors include increased age, male gender, prior cardiovascular disease, family history, and lower socioeconomic status.

**Lipid Management**

Dyslipidemia is a major risk factor for coronary heart disease. The American Association of Clinical Endocrinologists’ Guidelines and National Cholesterol Education Program Adult Treatment Panel (NCEP-ATP) III have set the following goals for individuals with coronary heart disease:

- Fasting total cholesterol: less than 200 mg/dL
- Low-density lipoprotein cholesterol (LDL-C): less than 100 mg/dL (< 70 mg/dL for high-risk patients)
- High-density lipoprotein cholesterol (HDL-C): 60 mg/dL or greater
- Triglycerides: 150 mg/dL

Therapeutic lifestyle changes should be initiated if the LDL-C level is above goal. In addition to increased regular exercise and weight reduction, patients need to reduce intake of saturated fat and cholesterol. Dietary methods to reduce LDL-C include increased intake of soluble fiber and plant stanols or sterols. These and other dietary recommendations are described in detail under Nutritional Counseling, below.

Therapeutic lifestyle changes often do not allow high-risk patients to achieve LDL-C targets. Statins (HMG-CoA reductase inhibitors), which reduce cholesterol production in the liver, are first-line pharmacotherapy. In one randomized controlled trial of more than 20,000 high-risk patients, those who received simvastatin demonstrated a 24% reduction in major cardiovascular events and a 17% reduction in cardiovascular death in the 5-year followup period. These benefits persisted for an additional 5 years. Although statins are generally well tolerated, myalgias are reported by 5–15% of patients. In rare cases, statins can cause myopathy and rhabdomyolysis, and the risk increases with higher doses. Some patients see an asymptomatic increase in liver enzymes,
particularly within the first 6 months of treatment. This often reverses on stopping the statin or with dose reduction. Prescribing statins with combination therapy using niacin, fibrates, bile acid resins, or ezetimibe is often necessary to enable patients to reach target LDL-C levels. (For additional discussion of hyperlipidemia, see Chapter 42.)


Hypertension Management

A review of medication compliance, use of nonprescription drugs that affect blood pressure, and assessment for ortho-static hypotension are included in the initial history. Patients with blood pressure greater than 130/80 mm Hg are advised to make lifestyle modifications, including regular aerobic physical activity of at least 30 minutes’ duration per day on most days, weight reduction (goal body mass index of 18.5–24.9 kg/m²), salt intake reduction (65 mmol/day or 1.5 g/day of sodium; 3.8 g/day of salt), and potassium intake limitation (120 mmol/day or 4.7 g/day). Adjustments in medications should be made to attain the goal blood pressure of less than 140/90 mm Hg in the general population, less than 130/80 mm Hg for high-risk patients with coronary artery disease or comorbidities such as diabetes and renal insufficiency, and less than 120/80 mm
Hg for patients with left ventricular dysfunction. (For additional discussion of hypertension, see Chapter 42.)


► Smoking Cessation

A smoking history includes current smoking status, past smoking history, secondhand smoke exposure, and smokeless tobacco use. Individuals who smoke pipes and cigars are at increased risk, but the amount of published scientific data and level of risk are much less than for cigarette smokers. Counseling, education, and pharmacologic assistance with nicotine replacement and bupropion should be considered with the goal of termination of tobacco use. The World Health Organization indicates that within 1 year of smoking cessation, coronary heart disease risk decreases by 50% and within 15 years, the relative risk of dying from coronary heart disease approaches that of a lifetime nonsmoker. Current smokeless tobacco use increases cardiovascular disease incidence by 27%.


► Diabetes Management

Diabetic patients should be questioned about medication and dietary compliance and monitoring of blood glucose levels. The aim is to enhance glycemic control. Physical activity reduces glucose intolerance and insulin resistance. The target for hemoglobin A1c concentration is less than 7%. Patients should receive individualized medical nutrition therapy to achieve treatment goals.
Nutritional Counseling

The initial step in dietary assessment is to record baseline daily caloric intake; dietary content of nutrients, saturated fat, trans-fat, cholesterol, and sodium; and balance with energy output. Daily energy expenditure should include moderate physical activity (200 kcal/day). The most effective dietary strategies for prevention of coronary heart disease are to increase fruits, vegetables, and whole grain intake; substitute nonhydrogenated unsaturated fats for saturated and trans-fats; and increase intake of omega-3 fatty acids.

Omega-3 fatty acids are polyunsaturated fatty acids that favorably influence factors related to cardiovascular disease, such as inflammation, platelet function and thrombosis, arrhythmias, telomere shortening, hypertriglyceridemia, adhesion molecule expression in plaque, nitric oxide–induced endothelial relaxation, and elevated blood pressure. There are no known serious adverse effects. Omega-3 fatty acids are found in fish (eg, salmon, albacore tuna, sardines), walnuts, seeds, dark green leafy vegetables (eg, Brussels sprouts, kale, spinach, salad greens), soybeans, and vegetable oils (canola, soybean, flaxseed). Reductions in LDL-C and triglyceride levels and increases in HDL-C level are seen with greater intake of omega-3 fatty acids. For secondary prevention, the recommendation is at least one 200–400 g serving of fatty fish or a fish oil supplement containing 900 mg of omega-3 fatty acids several times per week.

Soluble fiber found in oatmeal, kidney beans, apples, pears, barley, and prunes reduces cholesterol absorption. Five to ten grams or more of soluble fiber a day decreases total cholesterol and LDL-C, increases HDL-C, but has no effect on triglycerides. Phytosterols (plant sterols and stanols structurally similar to cholesterol and found primarily in vegetable oils and in lower amounts in cereals, fruits, and vegetables) lower LDL-C as a competitive agonist with cholesterol for gastrointestinal absorption.

In the Therapeutic Lifestyle Changes Diet, the AHA dietary recommendations for people with cardiovascular disease or those who are at high risk, 25–35% of total calories are permitted to be from fat (<7% saturated fat, up to 10% polyunsaturated fat, up to 20% monounsaturated fat, and no trans-fats). Protein makes up 15% of total calories, and complex carbohydrates (whole grains, fruits, and vegetables) make up 50–60%. Dietary cholesterol should be less than 200 mg daily. To reduce LDL-C, addition of 10–25 g of soluble fiber or 2 g of plant-derived sterols or stanols, or both, is suggested. Patients should consume oily fish containing omega-3 fatty acids at least twice weekly. Other recommendations include reducing intake of sodium (< 2.3 g/day or
approximately 1 teaspoon of salt) and limiting beverages and foods with added sugars.

Encouraging alcohol in the diet of a cardiac rehabilitation participant is not recommended. The potential benefits do not outweigh the addictive nature and adverse consequences of overconsuming alcohol. For those individuals who already drink alcohol as part of their customary diet, the AHA recommends limiting intake to not more than one alcoholic drink per day for women and two for men. There is evidence that links light to moderate wine consumption with a reduction in morbidity and mortality of cardiovascular disease; however, it is not seen with all alcoholic beverages. It has been postulated that the favorable effects of wine are linked to other factors in the beverage (ie, resveratrol), not the alcohol itself. Alcohol may adversely interact with many cardiac medications, and patients should use commonsense and report any concerning symptoms to their health provider.

There is a rising interest in plant-based diets, in which a larger proportion of daily food intake is made up of vegetables, grains, legumes, nuts, seeds, and fruits, rather than meats, poultry, fish, dairy products, and eggs. A plant-based diet does not require one to be vegetarian or vegan and accepts all foods in moderation with an emphasis on plants. In a study of more than 20,000 subjects with median follow-up time of 10.5 years, high consumption of fruits and vegetables (> 475 g/day or 6 servings) was inversely associated with coronary heart disease incidence compared with a low intake (< 241 g/day or 3 servings).

A popular plant-based diet is the Mediterranean diet. With this approach, red meat is limited to a few times monthly while fish or poultry is allowed twice weekly. Vegetables and fruits are emphasized. There is a decreased use of salt, butter, and added sugar, and use of olive oil is encouraged. The Mediterranean diet is associated with a significant reduction in adverse cardiac events as well as cardiovascular and all-cause mortality.


Movva R, Figueredo VM: Alcohol and the heart: To abstain or not to abstain? Int J Cardiol 2012; doi:10.1016/j.ijcard.2012.01.030
Weight Management

Obesity is an independent risk factor for cardiovascular disease and adversely influences other risk factors. Each cardiac rehabilitation participant should have baseline weight, height, waist circumference, and body mass index (BMI) documented. The goal BMI is 18.5–24.9 kg/m². It is important to recognize the patient’s muscle mass, because BMI overestimates body fat in muscular individuals and underestimates body fat in those with less muscle mass. The target waist circumference is less than 40 inches for men and less than 35 inches for women. If BMI or waist circumference is above goal, the patient should reduce his or her weight by 10% at a rate of 1–2 pounds per week over 6 months, which will allow for better adherence to lifestyle changes, help to preserve lean body mass, and produce less water weight loss. Patients should aim for an energy deficit of 500–1000 kcal/day, which is achieved by a combination of increased caloric expenditure and decreased caloric intake.

Psychosocial Management

Psychosocial factors have been linked to the pathogenesis and prognosis of heart disease. Routine screening to identify depression, anxiety, and substance abuse in all cardiac rehabilitation patients is recommended. Psychological interventions that reduce distress in cardiac patients also reduce morbidity and mortality. After a myocardial infarction, 20–45% of patients experience depression, and such patients have a fivefold greater risk of dropping out of cardiac rehabilitation. Anxiety is associated with increased risk of adverse events.
after myocardial infarction and may also interfere with cardiac rehabilitation participation.

Psychosocial interventions on the part of the cardiac rehabilitation staff enhance adherence to the program. These include education about the health and mood benefits of exercise, emphasizing the social support from other patients, making exercise as enjoyable as possible, developing realistic goals, engaging patients’ family members, and encouraging patients to seek psychological and pharmacologic treatment for depression if needed. Depressed cardiac patients have a more than fourfold increased risk of mortality than their nondepressed counterparts (22% versus 5%). In one study, prevalence of depressive symptoms decreased 64% following cardiac rehabilitation, from 17% to 6%. Patients with depression who complete cardiac rehabilitation demonstrate an improved fitness level that correlates favorably with a reduction in depressive symptoms and a 73% lower mortality rate (8% versus 30%).


Physical Activity Counseling

Exercise capacity, as well as occupational and recreational activity levels, should be established at baseline. The target is to perform 30–60 minutes of moderate physical activity most, if not all days of the week, to improve survival. In cardiovascular disease patients who exercise regularly, there is a 33% reduction in all-cause mortality. Patients should be educated in various ways to boost daily activity, including walking, doing housework, climbing stairs, working in the garden, parking far away from one’s destination, riding a bike, and planning family outings that include physical activity. Sitting less than 8 hours a day and meeting the physical activity recommendation of the World Health Organization independently protected against all-cause mortality. Patients should be advised to
reduce screen time, including watching television or working on a computer. Sitting for screen time is related to increased cardiovascular events and all-cause mortality regardless of physical activity. Screen time of more than 4 hours a day increased risk of cardiovascular disease by 125% and all-cause mortality by 48% as compared with individuals who had less than 2 hours of daily screen time.


Exercise Training

Once the symptom-limited stress test is completed, an exercise prescription may be developed. The prescription should include the type, intensity, duration, and frequency of exercise.

A. Exercise Type

Isotonic, rhythmic, aerobic exercise using large muscle groups, such as walking, jogging, swimming, and cycling, is preferred for the cardiac rehabilitation patient. After participating in endurance training for several weeks, the addition of strength training is recommended. The caloric effect and influence on risk-factor modification is less with strength training than with endurance exercise; however, the resultant increase in muscle mass correlates with increased strength, basal metabolic rate, and functional mobility. Patients who have congestive heart failure, uncontrolled arrhythmias, systolic blood pressure greater than 160 mm Hg, diastolic blood pressure greater than 100 mm Hg, severe valvular disease, or unstable angina should avoid strength training as these patients may decompensate. High-intensity isometric exercises should be avoided in cardiac rehabilitation patients due to the resultant increase in
afterload.

**B. Intensity**

There are several methods to determine the intensity of exercise. The AHA method for healthy adults is to calculate the maximum heart rate \( \text{HR}_{\text{max}} = 220 - \text{Age} \) and then exercise at a target heart rate of 70–85% of \( \text{HR}_{\text{max}} \). For cardiac rehabilitation patients, the goal is to exercise at 70–85% of maximal attainable heart rate on their symptom-limited graded exercise test. Depending on their level of conditioning, they may only be able to initiate the program at 50% of peak HR. The Karvonen method calculates the target heart rate as 0.4–0.6 \( \text{HR}_{\text{max}} - \text{Resting HR} \) + Resting HR. The oxygen consumption method uses 60–80% of maximal oxygen consumption. The Borg Rating of Perceived Exertion is a linear scale used to indicate the degree of perceived physical exertion while exercising. It correlates linearly with heart rate, ventilation, oxygen consumption, and lactate levels. Cardiac rehabilitation patients should exercise at a Borg rating from 11 to 15.

Patients taking β-blockers must reduce their submaximal and peak exercising heart rates by 20–40 beats per minute. Published data are available to determine target heart rate in such patients, and they can rely on the Borg scale. In patients with fixed-rate pacemakers, target heart rate cannot be used to guide exercise intensity, and patients must rely on the Borg scale. For dual-chamber and rate-responsive devices, the calculated target heart rate can guide exercise intensity.

**C. Duration and Frequency**

To achieve cardiovascular fitness one must exercise for 20–30 minutes at the target heart rate or conditioning level to start, and then progress to 30–60 minutes four to six times per week. Poorly conditioned patients with baseline exercise capacity of less than 3 METs should begin with short training sessions (3–10 minutes each) two to three times daily at a target intensity of 40–50% of maximum heart rate, with gradual progression as tolerated. The standard outpatient cardiac rehabilitation program frequency is three times weekly for 12 weeks.

An exercise session starts with the warmup designed to increase joint readiness, open existing collaterals, and prevent sudden changes in peripheral vascular resistance before the maximum contraction of the skeletal muscles occurs. Next is the training phase, during which exercises are completed at the
prescribed intensity and duration. Sessions end with a cool-down, consisting of a gradual reduction in exercise intensity allowing for redistribution of blood from the extremities. This redistribution reduces stiffness and soreness of muscles and joints and helps prevent sudden decline in venous return, reducing the risk of postexercise hypotension or syncope.


ACTIVITY PRECAUTIONS IN THE CARDIAC REHABILITATION PATIENT

➤ Post–Myocardial Infarction

After an acute myocardial infarction, exercise training may begin within 3–4 weeks with a gradual increase in activity over 6–8 weeks. Most individuals return to work by 8 weeks postinfarction. The extent of the myocardial infarction, development of complications, patient’s age, comorbid medical conditions, and physical demands of the individual’s job may all lengthen the period of disability postinfarction.

➤ Post–Catheterization or Angioplasty

There are few restrictions after a cardiac catheterization or percutaneous coronary intervention (PCI). Factors influencing the recovery after these procedures include the underlying reason for the catheterization, success of any therapeutic procedures performed, number and severity of procedure complications, amount of blood lost if complications arise, and an individual’s baseline medical, psychological, and nutritional status. A patient should be able to return to work and most normal activities 1 week after an angioplasty or stent procedure. Any limitations will usually depend on the healing of the insertion site. A symptom-limited exercise stress test the day after coronary stenting does
not increase the risk of clinical stent thrombosis or access site complications, indicating that patients may return to a full level of activity sooner rather than later. If the PCI was indicated as a treatment post–myocardial infarction, patients can begin exercise training 7–14 days after the procedure.

Post–Coronary Artery Bypass Grafting

Sternal precautions are almost universally given to patients following coronary artery bypass grafting (CABG) and other median sternotomy surgeries. The premise is that avoiding certain movements will avoid undue stress to the healing sternum and reduce the risk of sternal complications. This is especially true for individuals who have multiple risk factors for sternal wound dehiscence, including the use of the internal mammary artery in the bypass graft, women with pendulous breasts, morbid obesity, barrel chest, history of poorly controlled diabetes mellitus, osteoporosis, smoking, and redo operation for bleeding or repeat cardiothoracic surgery. Sternal precautions should be followed for 6–12 weeks, depending on the patient’s medical status and surgeon’s recommendations.

Current sternal precautions are listed in Table 23–5. Many patients remain functionally impaired long after cardiothoracic surgery, and it is possible that some sternal precautions contribute to this status by being overly restrictive and impeding ideal recovery. Recent literature does not support all the imposed restrictions, and clinical judgment should be incorporated in discussions between the medical team and the patient to allow safe and functional return to normal activities. More recently, patient-specific precautions focusing on function while not impeding recovery have been advocated.

| Table 23–5 Post-CABG sternal precautions. |
Off-pump CABG generally uses a median sternotomy approach and in some cases retraction techniques to elevate the heart, allowing access to vessels on the lateral and inferior surface of the heart. Off-pump surgery is performed on a beating heart after reduction of cardiac motion by a variety of pharmacologic and mechanical devices, such as β-blockers and calcium channel blockers and mechanical stabilizing devices to isolate the target vessel(s). Patients who undergo off-pump CABG should observe the same sternal precautions as described above.

Post–Minimally Invasive Direct Coronary Artery
Bypass

Minimally invasive direct coronary artery bypass (MIDCAB) is generally performed under direct visualization via a small left anterior thoracotomy that exposes the heart through the fourth intercostal interspace, allowing access to the diagonal branches of the left anterior descending artery, and possibly the anterior marginal vessels. This technique is performed primarily on patients with single vessel disease, because the number of anastomoses performed on the beating heart is limited to one or two. Advantages include shorter hospital stay, less infection, less pain, and quicker return to normal activities. Once at home, these patients have no specific physical restrictions, do not need to maintain sternal precautions, and can often return to usual activities and work within 2 weeks. This procedure is especially suited for patients who are at high risk of the traditional median sternotomy, including those who are elderly, on long-term corticosteroids, have severe chronic obstructive pulmonary disease, have severe deconditioning, or have advanced arthritis or orthopedic problems.


COGNITIVE DEFICITS IN THE CARDIAC REHABILITATION PATIENT

Ischemic heart disease and dementia are both associated with aging and may coexist. Cardiac rehabilitation patients may have preexisting cognitive deficits that have gone undetected until the cardiac incident. This is most notable in CABG patients, among whom 20–80% have cognitive impairment, including shortfalls in attention, concentration, short-term memory, fine motor function, and speed of mental and motor responses.

Post-CABG encephalopathy has a prevalence of 6.9% and is associated with increasing age, previous stroke, hypertension, diabetes, carotid bruit, and
cardiopulmonary bypass time. Each additional hour on cardiopulmonary bypass doubles the probability of postoperative encephalopathy. Stroke is estimated to occur in 1.6% of patients after CABG and is associated with increasing age, previous stroke, and hypertension. Abnormalities in magnetic resonance spectroscopy (an estimation of cerebral metabolism) correlate with postoperative neuropsychologic deficits, implying that transient metabolic neuronal disturbance is a cause of postoperative cognitive change.

Neuropsychologic deficits are most marked at day 3 after CABG, and these usually recover over the following week. Mechanisms responsible for neuropsychologic deficits include hypoperfusion during cardiopulmonary bypass; venous hypertension caused by manipulation of the heart during surgery; microemboli of air, atheroma, fat, and platelet aggregates originating from the cardiopulmonary bypass circuit and the ascending aorta; and a systemic inflammatory process that produces cerebral swelling. Randomized controlled trials of off-pump versus conventional on-pump CABG demonstrate that off-pump CABG is a safe and effective technique with no significant difference in the early composite outcome of death or complications. However, 1-year composite outcomes, completeness of revascularization, and graft patency are significantly worse with off-pump than with on-pump CABG. The early literature indicated that off-pump approaches had the potential to improve neuropsychological outcomes; however, current literature refutes this claim and shows no significant differences in neuropsychological outcomes between the groups.

Management

A neurocognitive evaluation should be part of the routine assessment of a comprehensive cardiac rehabilitation program. It is best completed before hospital discharge, and if cognitive decline is detected, followup serial evaluations should be scheduled. Patient and family education is essential, because cognitive deficits may influence safe functioning at home. Differentiating cognitive deficits from anxiety and depression disorders is important, because both can present with temporary cognitive changes or may mask underlying neurocognitive decline related to CABG.


**RISKS ASSOCIATED WITH CARDIAC REHABILITATION**

The AHA estimates the risk of any major adverse cardiac event during cardiac rehabilitation, including myocardial infarction, cardiac arrest, or death, as one event in 60,000–80,000 hours of supervised exercise. Patients at risk of an ischemic event during cardiac rehabilitation include those with postoperative angina or ventricular arrhythmias, excessive ventricular ectopy or myocardial ischemia with exercise, left ventricular ejection fraction less than 35%, New York Heart Association (NYHA) class III or IV congestive heart failure, or a systolic blood pressure drop of 10 points or more with exercise.

Arrhythmias are the most noteworthy adverse events during cardiac rehabilitation. Patients at risk include those within 6 weeks of an acute myocardial infarction as well as those with ischemia during exercise, a history of supraventricular or ventricular arrhythmias, a history of sudden cardiac arrest not yet stabilized on medical therapy, left ventricular ejection fraction less than 30%, and a newly implanted automatic implantable cardioverter defibrillator (AICD) or rate-responsive cardiac pacemaker. There is a 30% incidence of ventricular tachycardia with up to one urgent complication in 138 patient-hours of exercise, but only 1.3 fatalities per million patient-hours. The rate of arrhythmias is similar in men and women. Arrhythmias are induced by ischemia, which can alter depolarization, repolarization, and conduction velocity. Incorporating a warmup and cool-down period for each exercise session decreases the frequency of arrhythmias by promoting coronary perfusion.

**Management**
Cardiac rehabilitation should be delayed 6 weeks after implantation of an AICD to prevent dislodgement of the device leads. Peak heart rate during exercise should be 10 beats below the trigger to initiate anti-tachycardia pacing and AICD discharge. Smaller improvements in aerobic capacity may be achieved as compared to patients without AICD because of lower target heart rates, but these patients still obtain sizeable benefits from cardiac rehabilitation.


SPECIAL POPULATIONS

Elderly Patients

Elderly patients are more likely to have complications from myocardial infarctions and myocardial revascularization procedures and may have greater rehabilitation needs than their younger counterparts. More than half of cardiac rehabilitation candidates are over age 65. Elderly patients rarely drop out of cardiac rehabilitation and often achieve excellent outcomes (improved exercise tolerance and quality of life) with a low risk of adverse events. Despite these data, older individuals are less likely to be referred to and participate in cardiac rehabilitation.

Women

Mortality at all age groups is higher for women than men for myocardial infarction. Participation in cardiac rehabilitation affords both men and women equal benefit; however, women are less likely to be referred. When women are referred, they are less likely to attend due to limited financial resources, lack of social and emotional support, and transportation difficulties.

Racial Minorities
The prevalence of cardiovascular disease in black men (44.8%) and women (47.3%) far exceeds that in their white counterparts (37.4% and 33.8%, respectively). African Americans are less likely to be hospitalized for chest pain and have significantly fewer PCI and CABG surgeries as compared with whites despite data that support equivalent survival rates. Minorities are less likely to be referred to and participate in cardiac rehabilitation.

Heart Failure Patients

In the absence of particular contraindications to exercise, stable NYHA class II and III heart failure patients without complex arrhythmias should exercise as it improves quality of life and functional capacity, lessens symptoms, and increases exercise tolerance. Exercise training in heart failure patients is associated with significant reductions for all-cause and cardiovascular mortality and hospitalizations, and significant improvements in long-term health status compared with usual care. Patients with heart failure should execute a longer warmup period to increase skeletal muscle vasodilation. Exercise training should be gradual and start at 40–60% of the target heart rate for 2- to 6-minute intervals divided by 1- or 2-minute rest periods. Exercise duration is slowly lengthened until the patient can tolerate 30 minutes of uninterrupted exercise. Isometric exercises should be avoided due to resultant increased afterload.

As the incidence of end-stage heart failure rises in the United States, patients with an implanted left ventricular assist device (LVAD) are becoming more common among the cardiac rehabilitation population. Early initiation of exercise has been shown to be associated with improvements in exercise tolerance and survival rates in these patients. The most common limiting factors for exercise in LVAD patients are decrease in pump flow, infection, right heart failure, hemorrhage, and ventricular arrhythmia.

Cardiac Transplantation Patients

Cardiac transplant recipients experience exercise intolerance at workloads 40–50% below that of age-matched healthy individuals owing to residual skeletal muscle abnormalities developed prior to transplantation, decreased strength, and myocardial sympathetic denervation. The denervated myocardium and increased plasma norepinephrine seen in transplant patients cause elevated resting heart rate (often > 90 beats per minute), elevated systolic and diastolic blood
pressures, attenuated increase in submaximal heart rate, lower peak heart rate and stroke volume, and delayed slowing of heart rate in recovery. Exercise intensity relies on perceived exertion (Borg scale 11–13). Patients should begin walking up to 1 mile five times per week. To improve muscular strength and endurance, resistance exercise can be added after complete sternal wound stabilization, usually 12 weeks after surgery.

### Patients with Peripheral Artery Disease

More than half the patients with known peripheral artery disease have coexistent coronary artery disease. Additionally, at least one third of patients with known coronary artery disease have coexistent peripheral artery disease. Functional capacity is frequently less than 50% of the predicted value due to exercise limitation and claudication. To stimulate collateral formation, patients with symptomatic peripheral arterial disease should walk at least three times weekly and try to reach their claudication threshold within 3–5 minutes. They should then continue walking until claudication reaches moderate severity. This is followed by a short rest to permit symptom resolution. This exercise–rest–exercise cycle should be repeated several times during a 30- to 60-minute period.

More advanced peripheral artery disease may lead to dysvascular amputation. Rehabilitation must integrate cardiovascular training with preprosthetic and prosthetic training. Heart rate parameters for cardiac rehabilitation in amputees follow the same guidelines as for nonamputees. As a substitute for or enhancement of a walking program, arm ergometry is a safe and effective method for improving cardiovascular fitness in the patient with dysvascular amputation.

### Stroke Patients

One third of patients with documented coronary artery disease have a history of coexistent ischemic cerebrovascular accident and more than half of the patients with a known ischemic cerebrovascular accident have coexistent coronary artery disease. Although safe and comfortable walking speeds are often reduced in stroke patients, they are able to exercise at sufficient intensity to raise their heart rate and rate-pressure product to cardiovascular conditioning levels. The cardiac rehabilitation program should be individualized based on the degree of mobility dysfunction.


**CONTRAINDICATIONS TO EXERCISE TRAINING**

The American College of Sports Medicine has delineated several contraindications to exercise training. These include unstable angina, resting systolic blood pressure greater than 200 mm Hg or diastolic blood pressure greater than 110 mm Hg, new or uncontrolled tachycardia (> 120 beats per minute) or arrhythmias (atrial or ventricular), resting ST displacement (2 mm) on ECG, orthostatic hypotension, uncompensated heart failure, active myocarditis or pericarditis, significant aortic stenosis, third-degree heart block without a
pacemaker, acute systemic illness or fever, recent pulmonary or other embolism, thrombophlebitis, uncontrolled diabetes or metabolic disturbance, or severe comorbidity (physical or psychological) preventing participation. Nevertheless, most candidates for cardiac rehabilitation are eligible to participate fully in the program.


UNDERUTILIZATION OF CARDIAC REHABILITATION

Despite abundant evidence-based data supporting the safety and efficacy of cardiac rehabilitation, and its American College of Cardiology/AHA class I indication (evidence and general agreement that the treatment is effective and useful), the utilization of such services is low. Only 14–55% of patients who qualify for cardiac rehabilitation take part in such a program. Participation varies based on age (participation is inversely related to age), gender (men are more likely than women to participate, 22.1% versus 14.3%), race (whites are twice as likely to utilize cardiac rehabilitation as nonwhites), diagnosis (CABG surgery patients are more than twice as likely as myocardial infarction patients to participate), and median household income and level of education (highest levels are, respectively, 23% and 33% more likely to utilize cardiac rehabilitation).

Patient-related barriers include inadequate financial resources, lack of personal support system, and patient’s personal preference not to exercise or participate in risk intervention. Logistics (location of program, transportation, return to work, etc) can also pose a problem, which has led to an interest in home-based cardiac rehabilitation programs that have resulted in outcomes similar to hospital-based programs in terms of cardiac events, mortality, exercise capacity, health-related quality of life, modifiable risk factors, and health care costs. Preference may dictate the type of cardiac rehabilitation program a patient pursues.

Lack of physician referral is one of the principal barriers to cardiac rehabilitation participation. Older age, presence of most comorbidities, and non–ST-elevation myocardial infarction were associated with decreased odds of referral. In 2007, as a result of this ongoing lack of physician referral, the American Association of Cardiovascular and Pulmonary Rehabilitation,
American College of Cardiology, and AHA created performance measures for referral to and delivery of cardiac rehabilitation services. These measures serve as tools for determining the quality of care and for identifying opportunities to improve care. These performance measures are widely endorsed by the medical community.


Chronic pulmonary disease has a negative effect on several patient-centered outcomes. Among these, exercise capacity, dyspnea, and quality of life are the most relevant. Analysis of factors that contribute to exercise limitation, exercise-induced dyspnea, reduced quality of life, and pathophysiologic changes in patients with chronic pulmonary disease underlies research efforts to identify effective interventions for this patient population.

As defined by the American Thoracic Society and the European Respiratory Society, pulmonary rehabilitation is an evidence-based, multidisciplinary, and comprehensive intervention for patients with chronic respiratory diseases who are symptomatic and often have a decreased ability to participate in daily life activities. Integrated into the individualized treatment of such patients, pulmonary rehabilitation is designed to reduce symptoms, optimize functional status, increase participation, and reduce health care costs by stabilizing or reversing systemic manifestations of the disease. Comprehensive pulmonary rehabilitation programs include patient assessment, exercise training, education, nutritional support, and psychosocial support.

This treatment approach has clearly demonstrated favorable and long-lasting effects on all patient-centered outcomes. In addition, pulmonary rehabilitation appears to have positive effects on other important outcomes (eg, number and severity of exacerbations, health care resource utilization, and survival) in patients with chronic pulmonary conditions, particularly those with chronic obstructive pulmonary disease (COPD). Although the evidence for the efficacy of pulmonary rehabilitation is strong and it is highly recommended by current
guidelines, only a minority of eligible patients with chronic pulmonary disease are included in rehabilitation programs. This discrepancy may derive from lack of belief in the efficacy of such programs, lack of local access, or concerns about cost. The first of these impediments can be addressed by intensified promotion of the beneficial effects of pulmonary rehabilitation in the medical community; the other two, however, may require the design of simple and locally available programs using a minimum amount of resources that still produce clinically relevant effects.

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OVERVIEW OF THE NORMAL LUNG

Structural & Functional Anatomy

The primary function of the lung is ventilation–perfusion matching to maximize delivery of oxygen to and removal of carbon dioxide from the tissues. Air enters the respiratory system through the trachea, which then subdivides into the cartilaginous primary bronchi. These bronchi divide into secondary bronchi, supplying each lobe of the lung; the secondary bronchi branch into segmental bronchi (or tertiary bronchi); and these tertiary bronchi subdivide into terminal bronchioles.

At the level of the bronchioles, the lining has lost its cartilage and is now
predominantly epithelial tissue with smooth muscle. However, gas exchange does not occur until the terminal bronchiole subdivides again into respiratory bronchioles, containing the alveolar ducts and sacs. The predominant cell type in the alveoli is the type I alveolar cell, which allows for diffusion of oxygen and carbon dioxide. These cells comprise about 90% of the alveolar cells in the lung. The other 10%, type II cells, produce surfactant, which is needed to reduce the surface tension of the lung and allow for maximal gas exchange.

During normal inspiration, the diaphragm contracts and pulls down the contents of the abdominal cavity. At the same time, the external intercostal muscles pull the thoracic rib cage forward. These mechanisms combine to increase the volume of the thoracic cavity. According to Boyle’s law (which states that pressure is inversely related to volume), the increase in volume produced by these two maneuvers must be balanced by a decrease in pressure. It is this negative pressure that pulls air into the airways. In contrast, normal exhalation is by definition a passive process. Exhalation is determined by the elastic recoil of the lung and can be changed to forced exhalation with contraction of the internal intercostal muscles. The abdominal muscles (eg, external and internal obliques, rectus abdominis, and transversus abdominis) can also be used for forced exhalation. Accessory muscles that can be used for inhalation include the pectoralis, serratus anterior, and sternocleidomastoid.

Ventilation & Perfusion

Perfusion to the lungs is not uniform, given the pressure differences in the alveoli, arteries, and veins that exist from the apex to the base of the lung. A major reason for this difference is gravity, which leads to a higher-pressure blood supply at the base of the lung. Three perfusion zones, referred to as West’s zones, are differentiated (Figure 24–1). Zone 1, located at the apex of the lung, achieves the least amount of perfusion; the alveolar pressure ($P_a$) here exceeds the arterial ($P_A$) and venous pressures ($P_v$), causing the arteries and veins to collapse. This leads to a significant amount of dead space at the apex of the lung. In zone 2, located in the middle third of the lung, the $P_A$ is greater than the $P_a$, but $P_v$ is still less than $P_a$. This causes the venous vessels to collapse while the arteries remain open. Consequently, more perfusion occurs, but not as much as would occur were the venous system allowed to operate at full capacity without collapse. Zone 3, the point of maximal perfusion, is located at the base of the lung. In this zone, the $P_A$ and $P_v$ exceed the $P_a$; therefore, the vessels do not
collapse and maximal perfusion is achieved.

![Figure 24–1 West’s zones of the lung. $P_{a}$, alveolar pressure; $P_A$, arterial pressure; $P_v$, venous pressure.](image)

The ventilation capability of the lung is very similar to its perfusion capability. The alveoli at the apex of the lung are often overstretched and therefore receive less ventilation. However, perfusion is less than ventilation at the apex of the lung, despite both being reduced; therefore, the ratio of ventilation to perfusion ($V/Q$) is still greater than 1 (estimated at $\sim 3$). Moving downward through the lung, ventilation increases but perfusion increases to a greater extent, resulting in a $V/Q$ ratio less than 1. This functional difference has clinical relevance because often changes occurring in the base of the lung have a very large impact on overall gas exchange.

**Lung Compliance**

Lung compliance is defined as the change in volume of the lung in relation to the change in pressure. Lung compliance is dependent on elastic recoil; the less compliant the lung is, the smaller the volumes that can be attained with a given amount of pressure and the more dyspneic a patient can feel. Conversely, if lung compliance is too great, a large enough pressure for exhalation cannot be
generated given the overstretching of the lung that can occur. Restrictive lung
diseases are associated with reduced lung compliance and, therefore, reduced
total lung capacities. Obstructive lung diseases, however, often are caused by
destruction of the airway (eg, as in emphysema), resulting in increased
compliance. Age also causes an increase in airway compliance.

**EVALUATION OF LUNG FUNCTION**

- **Pulmonary Function Tests**

Pulmonary function testing is a well-established method of differentiating
between obstructive and restrictive lung diseases, following the progression of
such diseases, and differentiating lung disease from other possible causes of
dyspnea and other respiratory complaints. The key measurements and calculated
values that are extracted from pulmonary function testing ([Figure 24–2](#)) include
total lung capacity (TLC), vital capacity (VC), residual volume (RV), tidal
volume (VT), functional residual capacity (FRC), and inspiratory and expiratory
reserve volume (IRV and ERV, respectively). Other values that can be obtained
include forced expiratory volume in 1 second (FEV1), forced vital capacity
(FVC), and forced expiratory volume in 6 seconds (FEV6). These values are
obtained through spirometry, which provides a cost-effective method of
measuring and observing changes in specific components of lung function.
Pulmonary functions tests (PFTs) are noninvasive diagnostic tests that provide measurable feedback about the function of the lungs, evaluating and calculating values for total lung capacity (TLC), vital capacity (VC), residual volume (RV), tidal volume (VT), functional residual capacity (FRC), and inspiratory and expiratory reserve volume (IRV and ERV, respectively).

**Figure 24–2** Pulmonary functions tests (PFTs) are noninvasive diagnostic tests that provide measurable feedback about the function of the lungs, evaluating and calculating values for total lung capacity (TLC), vital capacity (VC), residual volume (RV), tidal volume (VT), functional residual capacity (FRC), and inspiratory and expiratory reserve volume (IRV and ERV, respectively).

**Spirometry**

Spirometry is performed using a calibrated instrument; patients are instructed to maximally inhale (from Vt to VC) and then maximally exhale (to RV) using all their force. This maneuver is repeated three times, and the highest values (assuming relative reproducibility) for FEV$_1$ and FVC (or FEV$_6$, depending on institutional preference) are documented. As defined, FEV$_1$ is the amount of air, and theoretically the largest proportion of air, that is exhaled within the first second. This is a reflection of the mechanics of the large and medium-sized airways. Today FEV$_6$ is increasingly being utilized, replacing FVC, as it makes it easier to identify the “end of expiration,” given a fixed value of 6 seconds, and additionally results in less participant fatigue and greater reproducibility. In patients with obstructive lung disease, the FEV$_1$ is significantly reduced compared with the FVC or FEV$_6$, and the ratio is reduced below the accepted
threshold of 0.7 or 70% of predicted value. In patients with restrictive lung
disease, however, both the FEV$_1$ and FEV$_6$ are reduced substantially in
proportion and the ratio remains normal (75–80%) or even elevated.

Measurement of Lung Volumes

Two methods are used to measure lung volumes: the gas dilution method and
body plethysmograph. The gas dilution method involves using either an inert gas
(eg, helium) or a method called nitrogen washout to calculate FRC. In the
helium method, the patient is connected to a spirometer attached to a fixed
concentration of helium in a fixed volume. The patient is allowed to breathe
normally at normal Vt respirations until equilibrium is reached between the
helium and the air in the patient’s lungs. Because the unit is a closed circuit, the
law of conservation of matter dictates that the volume multiplied by the fraction
of helium present beforehand must equal the volume multiplied by the
concentration of helium afterward. The volume present afterward is the total
volume of the lung’s FRC and the spirometer, and subtracting the volume of the
spirometer therefore gives the FRC value. This value can then be used to
calculate the TLC.

The other method, body plethysmography (Figure 24–3), involves placing
the patient in an enclosed box with a fixed, known volume. The patient is
instructed to pant against a closed shutter, which theoretically produces pressure
that is proportional to the FRC of the lungs. This value can then be used to
calculate the TLC and other lung volumes, as described previously. TLC is
generally most useful in calibrating treatment for patients with restrictive lung
disease. In patients with normal or even elevated FEV$_1$/FVC ratios,
determination of the TLC can help to confirm whether this is a normal variant or
represents restrictive physiology. TLC that is normal or elevated essentially rules
out restrictive lung disease, whereas reduced TLC points toward a restrictive
pattern.
Diffusion Lung Capacity for Carbon Monoxide

The diffusion lung capacity for carbon monoxide (DLCO) aids in further defining specific obstructive or restrictive patterns of disease. The test is performed by having the patient take in a single breath of 0.3% carbon monoxide and 10% helium, hold it for 10 seconds, and then exhale quickly for no more than 4 seconds. The subsequent concentration of carbon monoxide is measured and calculated. This is compared with standardized normal values; severe reduction in DLCO is defined as a value that is less than 40% of predicted.
The DLCO can help differentiate among likely causes in patients with obstructive lung disease. Significant reduction in DLCO is associated with emphysema. However, in chronic bronchitis-type COPD, patients often have a normal DLCO. Similarly, in patients with restrictive lung disease, the DLCO can help in differentiating among intrinsic and external causes. Intrinsic causes include lung diseases such as sarcoidosis and pulmonary fibrosis; external causes include restriction resulting from conditions such as obesity, kyphoscoliosis, and neuromuscular disease. If the patient’s DLCO is reduced, intrinsic lung disease is most likely causing the restrictive pattern. If normal, it points toward an external cause of the restrictive pattern, which should be investigated more thoroughly.

CLASSIFICATION OF LUNG DISEASE

1. Restrictive Lung Disease

**ESSENTIALS OF DIAGNOSIS**

- Decreased FEV\(_1\) and FVC are characteristic findings.
- DLCO is also decreased; however, the hallmark of restrictive disease is decreased TLC.
- Etiology includes musculoskeletal and neurologic disorders.

Restrictive lung disease can be grouped into three main categories based on etiology: intrinsic lung disease, chest wall abnormalities, and neuromuscular disease. All are associated with a restrictive pattern of disease that leads to reduced FEV\(_1\) and FVC (with a normal or elevated ratio) and decreased DLCO; however, the hallmark of this condition is decreased TLC. The TLC value is used to classify restrictive lung disease as mild, moderate, or severe. Mild disease is indicated by a TLC that is 65–80% of the predicted value; moderate, by a TLC that is 50–65% of the predicted value; and severe, by a TLC that is less than 50% of the predicted value.

Among the numerous causes of intrinsic lung diseases are asbestosis,
sarcoidosis, berylliosis, and pulmonary fibrosis. The inflammatory response associated with these disorders leads to V/Q mismatching that predominantly affects per-fusion, thereby promoting shunt physiology. Patients with these disorders are often tachypneic, with a compensatory decreased partial pressure of carbon dioxide (Pco₂). Chest wall abnormalities encompass a range of conditions that inhibit full expansion of the chest wall; these include spinal curvature disorders (eg, kyphoscoliosis), thoracic cage problems, and severe obesity. Neuromuscular diseases, such as polymyositis, myasthenia gravis, and muscular dystrophy, cause diffuse muscle weakness, decreased TLC, and a subsequent restrictive lung disease pattern.

2. Obstructive Lung Disease

**ESSENTIALS OF DIAGNOSIS**

- Low FEV₁/FVC ratio, with values at or below the fifth percentile of the lower limit of normal.
- DLCO and TLC aid in diagnosis of specific disorders: DLCO is low and TLC increased in patients with emphysema, whereas DLCO is normal in chronic bronchitis and preserved or elevated in asthma.

Obstructive lung diseases are identified by FEV₁/FVC ratios that are below predicted values. Formerly, an obstructive disease was diagnosed when these ratios were 70% of the predicted values or lower. However, because these ratios decline with age, using this method can result in an overestimate of the prevalence of these diseases. Consequently, using the fifth percentile of the lower limit of normal has been suggested as a more accurate method of defining obstructive lung diseases.

In contrast to restrictive lung disease, where disease severity is classified using TLC, obstructive lung disease severity is differentiated according to the FEV₁. Mild obstruction is defined as an FEV₁ that is 65–80% of the predicted value, moderate as 50–64% of the predicted value, and severe as less than 50% of the predicted value. Additionally, TLC and DCLO values help to identify the specific obstructive lung disease causing a patient’s symptoms, for example,
emphysema, chronic bronchitis, or asthma.

A. Emphysema
Emphysema is defined by the destruction of airways distal to the terminal bronchioles without evidence of fibrosis. In patients with emphysema, DLCO is decreased, TLC is increased, and symptoms are not reversible with bronchodilators. Emphysema can be further differentiated radio-graphically into proximal acinar, panacinar, and distal acinar disease. Proximal acinar (or centriacinar) disease is seen predominantly in the upper lobes and causes cystic lesions. Panacinar destruction is often seen in patients with α₁-antitrypsin deficiency. Paraseptal emphysema often leads to bullae that cause spontaneous pneumothoraces.

B. Chronic Bronchitis
Chronic bronchitis is defined on the spectrum of COPD by the presence of a chronic cough that occurs at least 3 months of the year for 2 consecutive years. In contrast to emphysema, there is minimal destruction of distal airways, which preserves the DLCO as normal. However, like emphysema, symptoms in chronic bronchitis are irreversible with bronchodilators.

C. Asthma
In patients with asthma, the FEV₁/FVC ratio is usually less than 70% of the predicted value or below the fifth percentile of the lower limit of normal. Bronchoconstriction and symptoms of impaired lung function occur during episodic attacks, but patients may seem unaffected between acute episodes. In order to induce findings characteristic of the underlying disease, a methacholine challenge test may be performed. This involves administering a potent bronchoconstrictor through a nebulizer to the patient, which activates the muscarinic M₃ receptor, thereby inducing bronchoconstriction. Spirometry can then be performed to analyze the FEV₁/FVC ratio. A bronchodilator is often given afterward to demonstrate reversibility. In asthmatic patients, the DLCO is preserved and sometimes even elevated.


**BENEFITS OF PULMONARY REHABILITATION**

A Cochrane review published in 2009 analyzed 31 randomized controlled trials that reported improvements in quality of life and in exercise capacity in patients receiving at least 4 weeks of pulmonary rehabilitation. Thirteen of the studies analyzed quality of life using two validated scales, the Chronic Respiratory Disease Questionnaire (CRQ) and St. George’s Respiratory Questionnaire (SGRQ). The CRQ repeatedly showed statistically significant improvements in fatigue, dyspnea, emotional function, and mastery. The SGRQ also showed statistically significant improvements in impact and activity and an improvement (not statistically significant) in symptoms. Maximal exercise capacity and functional exercise capacity also showed improvements but overall these were not statistically significant. The reason for this discrepancy (ie, statistically significant improvements in quality of life but lack of significant improvement in exercise capacity) remains to be determined.

The effectiveness of pulmonary rehabilitation in restrictive lung disease is much less well defined. The dropout/noncompliant rate reported in the preceding studies was high and few patients were followed to completion 1 year postrehabilitation. However, the results were positive, showing an improvement in exercise endurance and quality of life. This improvement was more pronounced in those who were already receiving long-term oxygen than in those who were not. Although the patients were screened using the CRQ and SGRQ
and showed a significant improvement, the large dropout rate was a concern, suggesting that the pulmonary rehabilitation program used for COPD patients may be too intense for those with restrictive lung disease. In another study, patients who received pulmonary rehabilitation showed improvement in the 6-minute walk test at 12 and 24 weeks. However, this study was neither controlled nor randomized.

Currently, and in contrast to the treatment of COPD, there are no solid guidelines to either recommend or refrain from performing pulmonary rehabilitation for patients with restrictive lung disease.

**COMPONENTS OF A PULMONARY REHABILITATION PROGRAM**

Pulmonary rehabilitation is a multidisciplinary program of care for patients with chronic respiratory impairment that is individually tailored and designed to optimize physical and social performance and autonomy.

**Areas of Emphasis in Pulmonary Rehabilitation**

**A. Education**

Education and training about pulmonary disease and self-care measures help patients to optimize living patterns and incorporate self-management into their lives. The aim is to teach methods that can be unobtrusively integrated into the patient’s lifestyle. The less obtrusive the training is, the more likely it is that patients will adhere to it. Training develops functional skills and habit patterns.

**B. Smoking Cessation**

Cigarette smoking has been identified as the most important source of preventable morbidity and premature mortality in North America. During the 1990s, tobacco was the largest single cause of premature death in the developed world. Smoking cessation is followed by immediate health benefits in terms of symptoms and organ function. Although many patients referred to a pulmonary rehabilitation program will have quit smoking prior to entering the program, others may still be smoking.

Whether or not patients should enter a pulmonary rehabilitation program if they are still smoking is a controversial topic. One study showed significantly
higher dropout and noncompliance rates among smokers than ex-smokers in such programs, leading to questions about the cost effectiveness of programs that include smokers. However, when the exercise tolerance and health-related quality of life in smokers and nonsmokers who underwent 7 weeks of pulmonary rehabilitation were compared, investigators found that the degree of improvement did not differ significantly among these two groups. Approximately 83% of rehabilitation programs in the United States enroll current smokers, and only 52% of them include a smoking cessation component that is overseen by the rehabilitation team. The number of such programs offering smoking cessation is lower in Canada.

Patients who continue to smoke may be advised to delay entrance into the program, or, alternately, to undergo parallel smoking cessation while proceeding with pulmonary rehabilitation. Smoking cessation is a vital part of the pulmonary rehabilitation process not only for smokers, but also for ex-smokers. Given that ex-smokers may be at risk of relapsing, reinforcing the important of smoking cessation in these programs may help ex-smokers maintain a smoke-free lifestyle.

C. Breathing Retraining

Patients with COPD have severe inefficiencies in lung function resulting from changes in chest wall shape, diminished muscle strength, lung hyperexpansion, reduction in gas exchange, and diaphragmatic malpositioning. In combination these changes cause the patient to develop dyspnea. Breathing retraining is one technique that may temporarily counter some of these changes. Controlled breathing techniques and chest physiotherapy are major components in the multidisciplinary rehabilitation of patients with COPD, bronchiectasis, and cystic fibrosis. Although only smoking cessation and long-term oxygen therapy prolong life in patients with COPD, it is likely that chest physiotherapy does the same for those with cystic fibrosis and diffuse bronchiectasis.

Diaphragmatic breathing is a method in which patients are taught to synchronize inspiration with abdominal expansion as they breathe slowly and deeply. On exhalation, the diaphragm is pushed upward by the abdominal muscles to create a more curved posture and a better length–tension relationship. This increases the effective force of the diaphragm as an inspiratory muscle. It also slows breathing and decreases the amount of accessory muscle use. This technique is used with pursed lip breathing as well as relaxation techniques. To date there have been few studies evaluating the effectiveness of diaphragmatic breathing, and those that were completed used different techniques. Depending
on the technique, some studies showed improvement in gas exchange, while others reached the opposite conclusion.

Pursed lip breathing (Figures 24–4 and 24–5) is a technique in which patients are taught to inhale slowly through the nose and exhale slowly through pursed lips. Pursing the lips retards exhalation and slows respiratory rate. Patients using this technique take deeper breathes and maintain their minute ventilation. This allows them to spend a greater portion of the breathing cycle at a higher lung volume. Slowing exhalation also reduces airway resistance by retarding airflow and turbulence. Pursed lip breathing also may improve V/Q matching by expanding lung volume. It has been shown to increase oxygen saturation for the period of time the maneuver is being performed. The technique is unique because many patients learn it spontaneously. Pursed lip breathing during exercise is an effective way to learn how to control dyspnea.

▲ Figure 24–4 Pursed lip breathing is one of the simplest ways to control shortness of breath. It is taught to patients with severe chronic obstructive pulmonary disease (COPD) to slow the pace of breathing, making each breath more effective. Pursed lip breathing may help to decrease dynamic hyperinflation, particularly during exercise.
D. Exercise

Exercise is the therapeutic engine that powers the benefits of rehabilitation. Exercise intolerance is one of the most troubling manifestations of COPD. Individuals with mild stages of COPD experience dyspnea and fatigue during significant exertion. Persons with moderate and severe COPD commonly have difficulty performing normal everyday tasks, including work activities, recreational exercise, and self-care. Dyspnea, leg fatigue, and discomfort are the three main factors that limit exercise in these patients. To avoid these symptoms, patients decrease their activity, leading many to become homebound and isolated.

1. Lower extremity training—Most pulmonary rehabilitation exercise programs involve training the lower extremities with cycling or walking, or both (Figure 24–6). Many studies have shown that skeletal muscle mass in the lower extremities is significantly decreased in a greater proportion than muscle mass in the upper extremities in patients with COPD. In one study, training on the cycle ergometer at 70–80% of the maximal workload ($W_{\text{max}}$) improved submaximal endurance time significantly more than training at 30% of $W_{\text{max}}$ (70% versus 9%). Other studies applying cycle ergometer training at 60% of $W_{\text{max}}$ showed
similar improvements in $W_{\text{max}}$ (−30%) and endurance time. The 6-minute walk distance also improved by 10–25%. In contrast, a study in a home-based environment at 60–75% of $W_{\text{max}}$ showed that $W_{\text{max}}$ improved by only 10%, and the 6-minute walk distance increased only marginally (2%). The less-significant results were believed to be secondary to the home-based environment, where supervision of training was likely much less strict.

▲ Figure 24–6 Lower body exercise.

2. Upper extremity training—Patients with pulmonary diseases report dyspnea and significant limitations when performing activities of daily living that involve the use of the upper extremities (Figure 24–7). Relatively few studies have examined the use of upper-limb exercise training for patients with COPD. Use of the arms is associated with high metabolic and ventilatory demand, and can lead to irregular, shallow, or dyssynchronous breathing. It is thought that the arm and shoulder muscles are recruited as accessory muscles of ventilation during exercise at higher workloads. When these muscles are also required to perform upper extremity work, they may not be able to manage the additional task of ventilatory support. Upper limb muscle training may involve endurance training (eg, arm ergometry [supported exercise], or unsupported, arm-lifting exercise) or strength training (eg, weight lifting).
Patients with moderate to severe COPD who participate in upper extremity exercise training have shown significant improvements in specific upper extremity performance tasks when compared with a control group. Reported benefits of upper limb training in COPD include improved arm muscle endurance and strength, reduced metabolic demand associated with arm exercise, and improved sense of well-being. However, these improvements did not result in significant changes in tests simulating activities of daily living. When arm training was combined with lower limb training, a group that received the combined training reported significant improvement in arm endurance compared with a group that received lower limb training only. In addition, the combined group had reduced Borg scores of perceived dyspnea when compared with the lower limb group. In a randomized controlled trial involving 26 patients with COPD that evaluated the results of upper arm versus respiratory muscle training, the outcome was similar: the endurance time of the upper extremities increased among those who received upper arm training when compared with the respiratory muscle training group. Unfortunately, although all of these studies have reported increases in patients’ upper extremity endurance, these changes have not been shown to translate into improvement in activities of daily living. Nonetheless, the current stance of the American College of Chest Physicians is to recommend unsupported endurance training in the upper
extremities.

3. **High-intensity exercise**—High-intensity exercise is considered to be that which takes place at greater than 60% of the patient’s Vo$_{2\text{max}}$ or W$_{\text{max}}$. High-intensity exercise must be undertaken for the patient to gain significant physiologic improvements in aerobic fitness. In one study, pulmonary rehabilitation in patients with moderate COPD led to reductions in lactate production and minute ventilation requirement for identical work rates. The magnitude of physiologic improvement was much greater in patients trained with a high-intensity workload versus those at a low-intensity workload. The cycle endurance time increased by 72% in the high-intensity group compared with only 9% in the low-intensity group. In a study assessing physiologic changes in patients with moderate to severe COPD after completing a 12-week exercise-training program, the aerobic capacity increased by 14% and there was also significant reduction in minute ventilation and lactic acid concentration post-training. Significant physiologic changes were noted in patients receiving high-intensity but not low-intensity exercise training.

It is important to recognize that not all patients are capable of high-intensity training at the onset of their exercise program. Patients should, however, be instructed to exercise to the maximum intensity they can tolerate to achieve the aerobic physiologic gains (Figure 24–8). Patients with severe COPD (mean FEV$_1$ of 38 ± 13% predicted) at baseline who participated in cycle ergometer endurance training for 12 weeks were able to tolerate, on average, exercising at 60 ± 22.7% of W$_{\text{max}}$. These patients also showed a significant increase in Vo$_{2\text{max}}$, as well as reductions in arterial oxygen consumption and minute ventilation.
Figure 24–8 Ambulation training with therapists. A regular walking schedule is an important component of pulmonary rehabilitation. Walking distance is increased progressively, and oxygen supplementation often is used in a patient who desaturates with exercise.

The potential detrimental aspects of high-intensity exercise training in patients with impaired lung function must also be considered. High-intensity exercise programs may lead to diaphragmatic fatigue in some participants. This was observed in a study that measured diaphragmatic pressure after completion of high-intensity cycling exercise. Of the 12 participants, 2 developed evidence of diaphragmatic contractile fatigue. Quadriceps fatigue has been shown to be a significant factor after a high-intensity exercise program. Another study showed reduced muscle redox capacity following a high-intensity training program.

Although high-intensity training programs in COPD patients clearly produce significant physiologic changes that are not seen in low-intensity training programs, studies have not correlated these changes with improvement in day-to-day function and activity. Further research is warranted to confirm the effects of high-intensity training on patient outcome.

4. Low-intensity exercise—Research has clearly demonstrated that lower intensity exercise programs lead to improved exercise tolerance in the absence of measured physiologic gains. In one randomized controlled trial, COPD patients
who underwent an 8-week rehabilitation program showed significant increases in treadmill endurance without a change in maximal oxygen uptake. Another randomized controlled trial analyzed the benefit of low-intensity peripheral muscle conditioning in patients with moderate to severe COPD. These patients showed significant improvement in both exercise tolerance and dyspnea reduction during the exercise program. Two recent studies directly compared higher intensity exercise with lower intensity exercise. Although gains in exercise endurance were noted following both types of training, patients in the higher intensity groups showed a higher magnitude of gains. However, the low-intensity groups had greater increases in arm endurance, and both groups achieved comparable reductions in overall dyspnea, functional performance, and health status as reported in responses to questionnaires.

5. Ventilatory muscle training—COPD patients have an inherent increase in ventilatory demand secondary to the dysfunctional pulmonary architecture. Hyperinflation, disadvantageous repositioning of the diaphragm, ventilatory muscle fatigability, and loss of ventilatory muscle strength are all factors contributing to progressive ventilatory dys-function. Ventilatory muscle training is a technique used in many pulmonary rehabilitation programs. It is believed that this technique may prevent or delay the onset of ventilatory muscle fatigue and failure, and eventually decrease symptoms dyspnea.

Two common forms of ventilatory muscle training are used in clinical practice. The first, called isocapnic hyper-pnea training, aims to increase ventilatory endurance. The patient is instructed to hyperventilate into a large tube to achieve supernormal target ventilation for approximately 15–20 minutes. This technique seems appropriate, when one considers that respiratory rate increases considerably during exercise. The second technique, inspiratory resistance training, focuses on building the strength of inspiratory muscles by having the patient inspire through small orifices. Patients are instructed to inspire through a sealed valve, which has an adjustable breaking pressure. Once the necessary pressure point is reached, the seal is broken and inspiratory flow starts.

Patients undergoing ventilatory muscle training demonstrate increases in strength and endurance in the ventilatory muscles; however, few studies have examined the clinical effects of ventilatory muscle training. In one study involving patients with moderate to severe COPD, higher $W_{\text{max}}$ was noted when patients received ventilatory muscle training with exercise compared with exercise only. The group that received ventilatory muscle training showed a 24% increase in $W_{\text{max}}$ compared with a 12% increase in $W_{\text{max}}$ in the exercise-only
group. It is not known if these improvements translate to an improvement in daily activities.

**E. Nutritional Support**

Patients with COPD are often underweight, and poor nutritional status frequently accompanies other chronic lung diseases. Weight loss results from negative net energy balance (ie, energy intake that is less than energy output). An individual’s daily energy expenditure includes three main components: resting energy expenditure (REE), which accounts for about 60% of the total expenditure; diet-induced thermo-genesis, accounting for less than 10%; and energy consumed for physical activity, which makes up the rest. REE is elevated (> 120% of normal) in patients with COPD. Most such patients who are malnourished demonstrate a significant increase in the oxygen cost of breathing compared with normally nourished patients. The increase in REE has been considered to result from the increased oxygen cost of breathing. The loss of fat-free mass is an independent predictor of worsening health status and mortality. Unfortunately, improving nutritional status in these patients can be extremely difficult.

About 25% of COPD patients are unable to maintain their nutritional status, as evidenced by weight loss. This percentage increases to 50% of COPD patients who are hospitalized for exacerbations of their disease. The cause of weight loss is not always known but is believed to be secondary to poor appetite, high energy cost of breathing, and desaturation during eating, which causes food avoidance. Loss of body mass leads to skeletal muscle and diaphragmatic weakness. The consequence is that patients with severe airflow obstruction who are also underweight have an increased mortality risk. Body mass index (BMI) is an important component of the BMI, Airflow Obstruction, Dyspnea, Exercise (BODE) index, a prognostic indicator of mortality.

Improving nutritional status is thus an important component of pulmonary rehabilitation. One study reported that COPD patients who gained more than 2 kg showed significant improvement in survival. In another study, a 1-point increase in BMI was also associated with improved survival in underweight patients. However, the use of the BMI can be misleading, as many COPD patients have a normal BMI but a low fat-free mass. Because of the complexities of nutritional planning for patients with COPD, nutritionists and dieticians should be involved in the rehabilitation program. When calculating calorie expenditure, nutritionists should take into account the increase that COPD patients have in energy expenditure when compared with normal patients of the same age. These professionals should also remember that undergoing weight
training may induce a negative protein balance, necessitating increased protein intake by patients.

Only one study has examined the use of supplements in COPD patients undergoing pulmonary rehabilitation. In this double-blind, randomized controlled trial, significant increases in work performance and health-related quality of life were noted primarily in the group of patients who received carbohydrate supplementation. With only one study for reference, it is difficult to make a strong recommendation for or against supplementation. Some opponents of this approach have suggested that supplementation may deter patients from consuming their regular meals, resulting in less than adequate calorie consumption. More research is needed before a consensus can be reached regarding the optimal caloric intake for COPD patients and the efficacy of specific supplements.

Optimal nutritional status in pulmonary rehabilitation should help to maximize the patient’s state of health, respiratory muscle function, and overall sense of well-being; it also may improve disease outcome. Obesity, which is defined as a body weight that is 30% greater than ideal body weight, may be detrimental to respiratory function. The large fat mass increases the work of the compromised respiratory system, particularly during weight-bearing activities. Obese patients with pulmonary disease should therefore be referred for weight loss education by a nutritionist. The goal of the intervention should be to decrease in fat while maintaining the patient’s fat-free mass.

**F. Psychosocial Support**

Physiologic impairment is only one aspect of the debilitating experience for patients with COPD or other pulmonary diseases. As patients become progressively more inactive and dependent on others, it is very common for them to develop depression, anxiety, panic attacks, and insomnia. Current prevalence estimates for clinically significant depression among pulmonary patients, particularly those with COPD, vary from 7% to 57%; prevalence for clinically significant anxiety is noted to be between 10% and 96%. To date no studies have shown a correlation between mortality and clinically significant depression or anxiety in such patients, although many studies have reported findings in patients with cardiac disease. While psychological distress is a major clinical feature in COPD that has implications for mortality, significant anxiety or depression has an even greater effect on patients’ daily lives, leading to impairment in quality of life and restricted activities.

Very few studies have been conducted in the past decade regarding
nonexercise-related psychosocial interventions in COPD patients. The literature includes reports that exercise is associated with enhanced psychological functioning, including decreased negative mood states. However, only one was a randomized controlled study. Participants in this study were assigned to one of three groups: one group received exercise with education and stress management; a second group, education and stress management without exercise; and a third group, no interventions. The group that received education and stress management without exercise showed significant increases in knowledge about and treatment of COPD, but no improvement in anxiety, depression, and quality of life. In fact, patients in this group actually showed increased distress. They also showed no changes in cognitive function. In contrast, the group that received exercise with education and stress management showed reductions in emotional impairment that were not seen in the other two groups, but there was no apparent difference in depression and anxiety, or quality of life. The only benefit noted was a 100% completion rate for the 12-week program among the group that received both education and exercise, compared with 64% in the exercise-only group. This marked improvement may indicate that educational intervention facilitates program adherence.

G. Family Training

Families of COPD patients play an essential role in the rehabilitation process. Their interactions with the patient and treatment team require that they have an understanding of the disease process, possible complications, plans of care, and prescribed medications. Family members must strike a fine balance between being supportive of and taking control of the patient’s life. The goal for the family is to provide support but leave the ultimate control of most aspects of life to the individual. Episodes of severe dyspnea and the progressive nature of the patient’s pulmonary disease can be extremely frightening for family members. Appropriate family members should be identified who can be trained in protocols for managing disease exacerbations, oxygen protocols (including when to use oxygen, and the correct procedure for doing so), medication administration, and when to seek the help of a health care provider. The family can also provide essential support for the patient in continued smoking cessation. It is imperative that all family members understand the importance of maintaining a smoke-free environment for their loved one.

H. Chest Physiotherapy

Chest physiotherapy, along with postural drainage, enhances mucus clearance
from central and peripheral airways of the lungs (Figure 24–9). The value of this therapy in stable patients with COPD and in acute COPD exacerbation is uncertain. Nonetheless, for patients who produce more than 30 mL of sputum every 24 hours or who have difficulty with sputum expectoration, chest physiotherapy combined with postural drainage and effective coughing techniques enhances sputum expectoration.

### Figure 24–9 Chest physiotherapy,

Chest physiotherapy is an essential component of therapy for patients with bronchiectasis and cystic fibrosis, as well as in the management of atelectasis in postoperative or seriously ill patients with COPD who are hospitalized. The frequency of treatments must be individualized based on the severity of disease and on the quantity of airway secretions that must be cleared. Standard chest physiotherapy performed in combination with postural drainage, coughing, and forced expiratory technique is the cornerstone of such treatment regimens. Newer modalities, such as mechanical chest percussion and positive airway pressure masks, have been introduced but require further clinical trials before they can be used routinely.


Ries AL: Pulmonary rehabilitation and COPD. Semin Respir Crit Care Med 2005;26:133–141.


Patient Selection for Pulmonary Rehabilitation

Evaluating patients for pulmonary rehabilitation is not, in theory, a complicated process. Two questions need to be answered: (1) Does the patient have a diagnosis that qualifies for pulmonary rehabilitation? (2) Is the program likely to help the patient? The team must determine whether the program will meet the patient’s goals effectively and safely. Essential to this determination is information about the patient’s diagnosis, stage of disease, comorbidities, and motivation.

For many years, patients with severe lung disease were not perceived to be good candidates for exercise training programs. However, recent studies utilizing high-intensity training stimuli clearly showed that patients with an FEV$_1$ less than 40% of the predicted value should not be excluded from exercise training. One large study found that patients with mild, moderate, and severe COPD demonstrated the same proportional improvement in exercise tolerance after pulmonary rehabilitation.

These findings confirm that patients at all stages of COPD can gain benefit from being part of a rehabilitation program; however, each case should be reviewed individually. Many patients can gain the same benefit from exercise programs that are self-directed. These factors needed to be taken into account when recommending the type of therapy that provides the greatest benefit to the patient.


Goals & Benefits of Pulmonary Rehabilitation

Achieving the highest possible level of independent function is the main goal of pulmonary rehabilitation, which is obtained by helping patients become more
physically active, by enabling them to learn more about their disease and treatment options, and by teaching them how to deal with their respiratory impairment. More specific goals of a pulmonary rehabilitation program are to reduce symptoms, improve activity, improve daily function, restore the highest level of independent function, and improve the overall quality of life of patients with chronic respiratory disease. These goals are achieved through patient and family education, exercise training, and psychosocial and behavioral interventions, as previously described. Rehabilitation interventions are geared to the problems and needs of each patient and are implemented by a multidisciplinary team of health care professionals. Success in achieving these goals has been studied and assessed by both physiologic and psychosocial outcomes.

Studies of patients who receive pulmonary rehabilitation have unanimously demonstrated no improvement in resting pulmonary mechanics or gas exchange values. There has also been no evidence of slowing in physiologic deterioration in the years following pulmonary rehabilitation. It is very important to understand that spirometry findings prior to the rehabilitation program do not predict how beneficial the program will be for that patient.

The largest and most consistent outcome in pulmonary rehabilitation is an improvement in exercise performance. A randomized controlled study by Ries and colleagues that compared exercise accompanied by education to education alone found a large improvement in exercise endurance and maximal exercise tolerance among patients receiving both education and exercise. In addition, a consistent reduction in muscle fatigue after exercise was demonstrated, along with improvement in quality of life and dyspnea scores on surveys. Other studies by Lacasse and associates and Wijkstra and coworkers noted similar improvement in exercise performance, dyspnea, and quality of life.

Improvement in functional status is a central goal of any rehabilitative effort. Functional status is divided into four dimensions: functional capacity, functional performance, functional reserve, and functional capacity utilization. Functional capacity describes the patient’s maximum work capacity. Functional performance is the level at which the patient is actually working. Functional reserve is defined as capacity minus performance. Functional utilization represents the percentage of capacity at which the patient is presently functioning. Pulmonary rehabilitation is designed to affect all four dimensions of functional status. In one survey administered to adult male patients with COPD who had completed hospital-based pulmonary rehabilitation programs, improvement in both functional ability and amount of dyspnea was reported for
Quality of life is a very important outcome that is measured after COPD patients have undergone rehabilitation. One investigator has differentiated quality of life into emotional functioning, social role functioning, ability to perform activities of daily living, and ability to participate in enjoyable activities. Two major scales used are the Quality of Well-Being Scale–Self-Administered (QWB-SA), and the Sickness Impact Profile to assess relief of symptoms. Another scale that has recently been used is the St George’s Respiratory Questionnaire (SGRQ, mentioned earlier in the chapter), which is more specific and sensitive to pulmonary disease state. Quality of life is very difficult to assess quantitatively and is very personal to the patient. Patients who have completed pulmonary rehabilitation programs have self-reported improvement in quality of life using established questionnaires. This is likely secondary to improvement in related fields, such as anxiety, depression, activity level, confidence, endurance, strength, dyspnea, and functional status.

Health care utilization has been investigated extensively in the past decade. Several studies have compared health care utilization in years following pulmonary rehabilitation with that in the year prior to the program. These studies have demonstrated reduction in hospital days over the succeeding years. One study showed a shift in activity from hospital-based care to phone-based monitoring. This likely reflected better communication between the physician and patient, as well as improved education about the disease process. Another group of investigators performed a randomized controlled trial in which hospitalization and outpatient visits were measured. The control group showed a significant increase in hospitalizations and outpatient visits, compared with a significant decrease in both inpatient and outpatient utilization in the rehabilitation group.

Survival is another outcome that has been the focus of much research. Several elements are known or believed to improve survival for patients with pulmonary disease, such as smoking cessation, improved nutrition, improved exercise and functional status, change to a more active lifestyle, and avoidance of exacerbations. However, studies to date have not reported improvement in morbidity or mortality in patients who participated in pulmonary rehabilitation programs. The reasons for this are unknown and research is ongoing in this area.

Program Setting & Staffing

The efficacy of pulmonary rehabilitation performed in inpatient, outpatient, or home settings has been well documented despite substantial variability in program structure. The effectiveness of pulmonary rehabilitation mainly depends on the structure and components of the program, rather than the setting itself. The choice of setting often depends on the variability and distance to the program, insurance payer coverage, patient preference, and the physical, functional, and psychosocial status of the patient. Inpatient rehabilitation is generally recommended for patients who are most affected by disease because with this option intensive rehabilitative services and specialized training for patients and families are available. Outpatient rehabilitation, which can be hospital or community based, has the potential to benefit patients but requires a certain level of functional ability. Although outcomes have not been well studied, home-based pulmonary rehabilitation is convenient for patients and family members and may provide sustained motivation for continued exercise training.

According to the well-accepted definition stated earlier, rehabilitation offers a holistic and comprehensive approach to medical care. For this reason, the combined expertise of an interdisciplinary team is highly desired. A multidisciplinary team whose structure varies according to patient population, program budget, reimbursement, and the availability of team members and resources delivers pulmonary rehabilitation. The rehabilitation team must be headed by a physician specialist (physiatrist or pulmonologist) who coordinates other physicians skilled in evaluating the neuromuscular, musculoskeletal, cognitive, and cardiopulmonary systems. Moreover, the team leader physician should be skilled in working with a team of professionals, because he or she is responsible for the medical treatment and rehabilitation program. The other members of the rehabilitation team include respiratory and physical therapists, an occupational therapist, a rehabilitation nurse, a vocational counselor, a social worker, a dietician, and a psychologist. Each member also needs to have
knowledge of the general principles of other members’ disciplines.

The environment in which the pulmonary rehabilitation program is carried out is of pivotal importance. Preferably it should be a large, quiet, and comfortable fitness facility in which all the people involved in the therapeutic process can be accommodated. Separate rooms should be available to offer discretion to patients who have undergone bronchial drainage procedures that may induce copious expectoration.


Outcome Assessment

Significant progress has been made in evaluating patient outcomes in pulmonary rehabilitation. However, outcome evaluations must be well-conceived to ensure that the most relevant information is collected for review. For example, programs should not only determine how much individual patients benefit from rehabilitation but should also identify what components of the process led to these benefits. Meaningful conclusions regarding the benefits of the program require robust evaluative instruments. Measurement of outcomes is essential in pulmonary rehabilitation as it directly relates to effectiveness. It is generally recommended that outcomes such as dyspnea, activity, and exercise capabilities be evaluated because these areas should improve with pulmonary rehabilitation.

Health care professionals involved in the care of pulmonary rehabilitation patients must consider which indicators of outcome are most useful to their clinical practice. Symptoms, exercise capacity, and health-related quality of life must be objectively measured before and after the pulmonary rehabilitation protocol and used as indicators of outcome. Symptoms can be measured with dyspnea and fatigue scales (eg, the Borg scale or Visual Analog Scale [VAS]).
Exercise capacity can be measured with field tests (eg, 6-minute walk distance), and health-related quality of life can be measured with self-administered disease specific questionnaires (eg, CRQ or SGRQ, described earlier).

A. Symptom Evaluation

The two major symptoms in patients referred for pulmonary rehabilitation are dyspnea and fatigue. These symptoms are complex, with multiple mechanisms of action. By nature, symptoms are subjective and require self-reporting. In the pulmonary rehabilitation setting, dyspnea or fatigue can be assessed in two ways: in “real time” or through recall. Each approach may yield differing results. Real-time evaluation of symptoms will only answer the question of how short of breath or fatigued the patient is at the moment of testing. The Borg scale and the VAS are most commonly used for this purpose and are useful in assessing dyspnea or fatigue during exercise testing or training. Recall of symptoms, such as dyspnea or fatigue, is usually accomplished through the use of questionnaires. Some questionnaires require patients to rate their overall dyspnea experience, whereas others ask about dyspnea related to activities. Although most have adequate psychometric properties, some were initially developed for research purposes and thus are not user friendly in the pulmonary rehabilitation setting. Other considerations should be the context in which the symptoms are measured, how the questions about symptoms are worded, and the timeframe over which the symptom is measured.

B. Performance Evaluation

Improvement of the patient’s ability to engage in activities of daily living is an important goal of pulmonary rehabilitation. Studies have shown that improvements in exercise capacity do not necessarily translate into increases in activities of daily living, and that assessment of functional performance is important. Improvement in performance can be accomplished by direct observation or by patient report. Most pulmonary rehabilitation programs rely on patient self-reports to assess activity levels using both the patient’s report of the intensity of dyspnea with activities and the degree to which a patient may perform activities in a real-life situation. An emerging method of evaluating activities in the nonlaboratory setting is the use of activity monitors or motion detectors. Activity monitors can be used in the rehabilitation setting to provide an objective measure of patients’ daily activities. Monitors range from simple, such as a pedometer, which evaluates the number of steps a patient takes, to more complex devices that measure movement in three planes, such as a triaxial
C. Exercise Capacity
Measurement of exercise capacity can be accomplished in several ways, including field tests, activity monitors, and cardiopulmonary exercise testing. Field tests have several advantages: they are simple to perform with little additional equipment, are conducted in a nonlaboratory setting, and are responsive to the pulmonary rehabilitation intervention. They are either self-paced, such as the 6-minute walk test, or externally paced, such as the incremental and endurance shuttle walk tests. Both tests measure distance walked. Although cardiopulmonary exercise testing can be of considerable help in the initial assessment of exercise limitation and formulating the exercise prescription, it can also be useful in outcome assessment. Physiologic measurements provide valuable insight into mechanisms of exercise intolerance. Cardiopulmonary exercise testing can be incremental to maximal symptom limitation or at a constant work rate.

D. Health-Related Quality of Life
A person’s satisfaction or happiness with life has been described as quality of life, and may be considered a balance between what is desired in life and what is achieved (although these indicators are difficult to measure). In rehabilitation, two types of instruments have been used to measure quality of life: general health questionnaires, such as the Sickness Impact Profile and short form; and disease-specific scales, such as the CRQ and the SGRQ (discussed earlier in the chapter). Disease-specific measures have demonstrated greater sensitivity to change from baseline after rehabilitation intervention.


Burn Rehabilitation

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Lisa Forbes, MSc, OT Reg(MB)
Michael Andreas Serghiou, OTR, MBA

Burn care and injury management continue to advance throughout the developed world, leading to improved survival and rehabilitation of individuals who suffer burn injuries. These individuals require a comprehensive approach to care to help them achieve full independence and maximum recovery from their injuries. Current estimates suggest that more than 1.2 million individuals in the United States require care for burn injuries annually, and this number is significantly higher in many other countries throughout the world. Successful rehabilitation of a person with a burn injury requires a team approach in which each member provides essential resources and skills to help the individual transition from being a “victim” of a burn to being a functionally independent “survivor.”

EPIDEMIOLOGY

Burns cause approximately 60,000 hospitalizations and over 6000 deaths annually in the United States. Burn injuries are most common among males (~70% of cases) between the ages of 16 and 40 years; however, both young and old are highly susceptible to burn injury. Burns are the second most common form of abuse among children aged 1–12 years, and the second leading cause of accidental death among adults older than 60 years.

Mortality from major burns has been significantly reduced in North America. Nonetheless, according to the American Burn Association National Burn Repository, the mortality rate for the median lethal dose (LD_{50}) for burns is 80%.
This median is even higher in the pediatric population between the ages of 1 and 15 years, where the rate reaches just over 95%. Several negative predictors influence burn survival, including the presence of inhalation injury, increased age, presence of other medical comorbidities (eg, diabetes mellitus or chronic obstructive pulmonary disease), and higher overall surface area injury.

The most common burn injuries affect approximately 10% of total body surface area (TBSA) and are the result of flame contact or scald. Of patients with these types of burns, approximately 95% survive their injuries and hospitalizations. Burns can also result from electrical contact, chemical contact, radiation exposure, and, to a lesser degree, various skin syndromes. Depending on the cause, patients with these injuries will require varying amounts of wound care, surgery, rehabilitation, and aftercare to recover.


**PATHOPHYSIOLOGY**

Human adult skin is the largest organ in the body, with a surface area of approximately 2 m², an overall depth of approximately 2 mm, and a weight of approximately 6 lb. The skin comprises three primary layers—epidermis, dermis, and hypodermis (subcutaneous tissue)—and serves many key functions,
including thermoregulatory, neurosensory, immunologic, evaporative, metabolic (eg, oxygen, nitrogen, and carbon dioxide diffusion), and protective. Equally as important is its role in physical identity.

The uppermost layer is the epidermis, composed primarily of five layers of thin, avascular, keratinized epithelium having as its primary role the protective (germ- and injury-resistant) function of the skin. The deeper dermal layer is composed of two layers: an upper papillary dermis, composed of loose areolar connective tissue, and a deeper reticular layer, composed of dense elastic and collagen fibers and housing many glands and vessels. This layer is connected to the underlying hypodermis and provides the mechanical strength that enables the dermis to attach to the underlying tissues. The hypodermis is the insulating layer to the underlying subcutaneous structures.

Burn injuries affect the skin in three histologic zones extending away from the center of injury in two directions (out and down). Injuries to each zone produce characteristic histologic changes, ranging from hyperemia to stasis to coagulation. The zone of hyperemia is the outermost zone of injury (the area least affected by direct injury). Injury to this zone results in vasodilation and minimal cellular death. In minor burns, this may be the only zone of injury. The zone of stasis is closer to the center of the burn site and experiences a deeper injury. Burns to this zone result in greater cell necrosis if concomitant factors such as vascular constriction, edema, and vascular thrombosis or ischemia are not addressed during the resuscitation process. The zone of coagulation is closest to the direct injury and experiences the deepest destruction. Burns to this zone produce cells that are completely necrotic and will require surgical intervention to effectively replace and repair the damaged tissues.


CLINICAL FINDINGS

Burn Classification
Burns are classified according to the depth of injury to the skin and the total surface area of the body that is affected. These two classifications are important because the greater the size, the more potential complications and contractures can arise, and the larger the depth of injury, the greater the mortality rate will be. Various methods have been promoted for calculation of the TBSA. A well-known method of estimation is the so-called Rule of Nines, which divides the body into various segments, each accounting for 9% of the surface area. Although widely used, this method has been found to undercalculate size differences based on age. (For example, the head represents a larger proportion of the body in children than in adults.) More routinely, burn size estimation is calculated using diagrams, such as the Lund and Browder chart (Figure 25–1). Additionally, calculation can be performed using the palm method, in which the palmer surface of the patient’s hand represents approximately 1% of TBSA.
Temple University Hospital
Temple University Health System

ADMISSION BURN RECORD
To be completed upon admission:

Date: __________________________
Height: ___________________ Weight: __________
2° ________ + 3° _________ = __________\%

FLUID RESUSCITATION GUIDELINE

_______ cc LR x ____% Burn x ________ kg/ft² = ________

24 Hour Total
1st 8 hr = ________ cc
2nd 8 hr = ________ cc
3rd 8 hr = ________ cc

Percent Surface Area Burned
(Berkow Formula)

<table>
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<th>AREA</th>
<th>1 YR.</th>
<th>1–4 YRS.</th>
<th>5–9 YRS.</th>
<th>10–14 YRS.</th>
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<th>ADULT</th>
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Burns are classified and size is estimated and documented using a form that shows the location of the injury on the body and the depth of the injury.

Computer modeling systems are currently being developed that will incorporate overall body mass into the calculation processes; however, these are not yet available. Thus, assessment is still routinely performed by direct visual inspection of the burn. Most importantly, it is the location of the burn injury, in addition to the depth, that tends to be most predictive of the overall rehabilitative needs of the patient. In areas with highly mobile joints or articular surfaces such as the face and hands, joint surfaces are more likely to heal with scar hypertrophy, leading to further contractures and loss of overall function.

Burn depth is classified as superficial, partial-thickness, full-thickness, and subdermal. These classifications are also referred to as first degree, second degree, third degree, and fourth degree, respectively. Partial-thickness burns (second degree) can be further subdivided into either superficial or deep categories. Depth of injury is important from a wound-healing perspective, because it correlates strongly with the formation of scar tissue and the potential for subsequent contracture development.

**Superficial Burns**

A superficial burn involves only the epidermal surface and leaves the epidermis intact (Figure 25–2). This type of burn is usually erythematous and painful, does not contain blisters, has characteristics similar to “sunburn,” and will heal spontaneously in less than a week, usually without any scarring.
A superficial burn is red and painful and usually heals without scarring.

**Partial-Thickness Burns**

A superficial partial-thickness burn involves both the epidermis and dermis. This type of burn is quite painful, appears moist and pink, has many blisters, and blanches with pressure (Figure 25–3). Superficial partial-thickness burns usually heal spontaneously in 2 weeks, with minimal scarring or loss of function; however, some pigment changes can occur.
thickness burn such as this leg burn is painful, pink, and moist; blanches; and has thick-walled blisters.

A deep partial-thickness burn also involves both the epidermis and dermis, and usually extends more deeply into the dermal structures. This type of burn is also painful but is not as tender as more superficial burns. The burn site may blister, but the overall appearance is red and less moist, with some areas that appear pale or slightly white. This type of burn should be monitored, because such burns, if allowed to heal spontaneously for more than 3 weeks, are much more likely to produce contractures and loss of function due to the prolonged inflammatory wound process. This type of injury may require a skin graft to improve the rate of healing and to limit the potential for contraction or loss of function.

**Full-Thickness Burns**

A full-thickness burn injury involves the loss of the epidermis and the entire dermis. These burns are pale and leathery in appearance, dry, insensate, and have taut and relatively inflexible skin (Figure 25–4. Full-thickness burns require skin grafting and cause scarring and higher loss of function if not effectively managed throughout the rehabilitation process. A subdermal burn involves destruction of the epidermis and dermis and extends more deeply into the fat, muscle, tendon, and bone (Figure 25–5. This type of injury usually results in significant loss of function and may necessitate amputation of appendages if tissue necrosis is significant.

![Figure 25–4](image) Full-thickness burn (third degree). This full-thickness burn of the
trunk and upper extremity is waxy white, dry, and insensate, with a taut skin envelope. This injury requires skin grafting and will heal with scarring.

Figure 25–5 Subdermal burn (fourth degree). This subdermal hand burn shows complete necrosis of the digits into flexion and complete loss of blood flow to underlying dermal structures, with tendon and nerve exposure.


Hawkins H, Finnerty CC: Pathophysiology of burn scar. In: Herndon DN
COMPLICATIONS IN BURN INJURY

Electrical Injuries

Burns that result from electrical injuries typically account for 5% of annual burn admissions in the United States and are divided into high-voltage (> 1000 V) and low-voltage (< 1000 V) injuries. Both types of injury pose unique challenges. In an electrical injury, the current travels the path of least resistance through the body, which can cause significant peripheral and central nervous system damage as well as potentially life-threatening arrhythmias and myocardial necrosis, with little apparent cutaneous injury. Moreover, the presentation of these symptoms may be delayed, which further impacts the rehabilitation process and may lead to permanent functional impairments. High-voltage injuries are also more readily associated with amputation and increased length of hospital stay. Lightning strikes are a rare type of high-voltage electrical injury that can pose significant complications both acutely and at a late stage; these include encephalopathy, myelopathy, ocular complications (macular holes and cataracts), and hearing loss.

Heterotopic Ossification

In heterotopic ossification, new bone forms in tissues that normally do not ossify. The incidence in the general burn population is relatively rare (between 1% and 3%). Heterotopic ossification can occur in burned or unburned areas and affect all major joints; however, most commonly the posterior elbow is affected, followed by the shoulder and hip. Pathogenesis is not completely understood.
Risk factors that have been suggested include delayed wound closure, long periods of immobilization with vigorous remobilization, resistance to physical therapy, and genetic predisposition. Recent reports indicate that the extent of burn area is not a determining factor; in fact, heterotopic ossification has been noted in patients with burns involving less than 10% of TBSA.

Early symptoms include pain and joint swelling, along with reluctance to move the affected joint. Clinical observation and bone scans can enable diagnosis at an earlier stage than radiographic changes, and ossification is often detected by physical and occupational therapists during rehabilitation.

Treatment and prevention of heterotopic ossification remains controversial. Current recommendations suggest early excision and grafting, and obtaining radiographs immediately upon observation of unusual swelling at a joint accompanied by a patient’s reluctance to move a joint freely. Once heterotopic bone or calcification is confirmed by radiographs, restricting joint exercise to gentle passive and assisted active range of motion (ROM), only, is recommended. Surgical resection is indicated when ROM is significantly decreased and should be performed only when scars are mature, the patient is well, and bone growth is mature or suspected neurovascular compromise is present. However, heterotopic ossification can recur even after surgical resection.


Peripheral Neuropathy
Peripheral neuropathy in burns has been found to seriously impair rehabilitation and tends to correlate highly with electrical burns, full-thickness burns, burn involvement of greater than 20% of TBSA, and individuals older than 20 years of age. There are several types of neuropathy in burns, including mononeuropathies, peripheral polyneuropathies, and patterns that resemble mononeuritis multiplex. In deeper and larger TBSA injuries, axonal injury is more prevalent than demyelinating neuropathies. Median sensory neuropathies are the most common peripheral nerve abnormality after burn injury. If electrodiagnostic testing is necessary in a burn patient, the practitioner should bear in mind that skin changes after burn injury (including thickened hypertrophic scars) can alter the results of both nerve conduction testing and electromyography; caution should therefore be exercised in the interpretation of test results.


Pruritus

Most patients (nearly 90%) develop severe itching after a burn injury. This itching, even with small burns, is very distressing to patients, often affects daily living activities, and can cause skin breakdown and loss of sleep. Itching can be worse in extreme heat, during physical activity, and in times of stress. The exact mechanism of itching is not understood but may be related to increased mast cell and histamine presence in the burn scar and increased nerve endings and substance P. The treatment for pruritus is based on trial and error and always includes the use of skin moisturizers several times per day. Trials of diphenhydramine hydrochloride and other antihistamines, gabapentin, doxepin, and analgesics, as well as cool compresses, loose clothing, and massage, may
also be tried to relieve burn itch. Additionally, advising that itching is part of the “normal” healing process may assist patients in coping with this complication.


Contractures

Burn scar contracture is a common complication of a burn injury that occurs when normal skin tissue is replaced with pathologic scar tissue that is shorter and less extensible than the tissue it replaces. The result is loss of motion, loss of tissue alignment, and, often, misalignment of an associated joint or anatomic structure.

Contractures may result from the effects of myofibro-blasts and free actin in the scar. Two types of contractures are distinguished: intrinsic (characterized by loss of tissue in the injured area, with subsequent distortion of the involved anatomic part) and extrinsic (characterized by loss of tissue at a distance from the affected area, without injury to the distorted structures themselves). To date, basic research on the mechanical effects of stretch has been difficult to translate into clinical practice. Small forces observed in the in vitro setting would suggest
that stretching can increase the risk of contracture, but this has yet to be proven clinically. A burn patient tends to prefer a position of comfort, folding joints into flexion, and it is this position that may influence the new collagen fibers to fuse together in a shortened length, becoming a solid mass of collagen. Prolonged immobility and the shortened, fused collagen across a joint crease may decrease ROM and result in contracture formation (Figure 25–6. Burn scar contractures are labeled opposite to the motion that is restricted. For example, if a patient cannot fully extend the elbow, the contracture is called an *elbow flexion contracture*. A contracture may develop in any skin crease overlying a joint, but the most common locations are shoulder, hand, elbow, and knee. Burns that involve a larger TBSA are associated with a greater number of contractures, and contractures are more likely with a full–thickness burn.

![Figure 25–6 Scar contracture. The contractile forces of the scar prevent any plantar flexion, significantly limiting this child’s ability to walk.](image)

Scar contracture can continue to occur throughout the scar remodeling phase. Splinting, positioning, serial casting, and ROM exercises are necessary to combat this relentless process until maturation is achieved. Contracture may still occur despite proper therapeutic modalities. In these cases, surgical releases may be necessary in which additional orthotic use and dedicated mobilization of the joint are required to counteract the potential redevelopment of hyper-trophic scar contracture.
TREATMENT

Initial Management of Acute Burns

Management of the initial burn injury involves a host of considerations stemming from review and assessment of the burn patient.
A. Wound Care
Burns are initially sterile but within 72 hours usually become colonized by endogenous bacteria. Initially gram-positive organisms predominate but by postburn day 5, gram-negative bacteria begin to colonize the burn wound. Daily wound care is most often performed, and silver sulfadiazine is the most common application owing to its widespread bactericidal properties against many gram-positive and gram-negative bacteria. Some data suggest a trend to better healing with the use of other dressings, such as biosynthetic materials, hydrocolloids, and other types of combination materials with long-acting antimicrobial properties. In either scenario, specialized wound care is required to maximize healing and minimize pain.

B. Escharotomy
In cases of circumferential burn, particularly around the chest or extremities, an escharotomy may need to be performed to relieve the pressure on the underlying soft tissue structures and prevent the development of compartment syndrome. In cases of emergent escharotomy to the limbs, elevation and a neutral position orthosis is recommended (Figure 25–7).

▲ Figure 25–7 An emergent escharotomy is performed in an attempt to preserve blood flow and relieve circumferential pressure. Note the clawing appearance of the hand.

C. Skin Grafting
Early tangential excision and autologous skin grafting is used to remove dead tissue, replant new tissue, and expedite wound closure. This early and aggressive wound management technique has significantly improved overall survival rates in patients with burn injuries. The devitalized tissue is removed and skin is harvested from a donor site typically located on the patient’s upper thigh. Once the donor skin is prepared, it is transplanted onto the newly excised wound bed and affixed to the skin with sutures, staples, or a dermal glue material. The skin is then held in place with a postoperative bolster dressing. If the wound bed covers a joint, a postoperative positioning orthosis is also applied. In areas of high aesthetics, a sheet graft is used to improve the cosmetic result and limit contracture potential. In other areas, or where more substantial skin is required, the skin can be meshed in various ratios, including 1:1, 2:1, or even 3:1; however, meshed grafts have greater potential for hypertrophic scarring (Figure 25–8).

▲ Figure 25–8 This close-up image shows a split-thickness skin graft covering a hand with meshed skin, demonstrating increased tissue expansion to cover the damaged tissue.

D. Inhalation Injury

Inhalation injury poses additional complications for the burn patient and is highly associated with greater rates of mortality in both children and older adults. Additional potential complications include the development of pneumonia, adult respiratory distress syndrome, multisystem organ failure,
hypoxia, and possible brain injury. Along with early debridement and excision, tracheotomies are performed in patients with larger burns; results have shown improved ease of oral hygiene and prevention of microstomia, especially in regard to facial burns.

**E. Nutritional Requirements**

Patients with burns covering more than 30% of TBSA have increased caloric and nutritional needs that should be addressed with early enteral feeding. Daily caloric requirements for an adult burn patient can be close to 25 kcal/kg plus 40 kcal per 1% of TBSA burn per day. Loss of lean muscle mass, loss of mineral bone density, and increased insulin resistance are all potential complications that should be addressed early in the acute care as well as later rehabilitation phases. Dysphagia can develop from inhalation injury and further impact the metabolic needs of the patients. Because the burn injury represents a hypermetabolic state, this cascade has been improved with the use of anabolic agents, β-blockers, and exercise. The use of a synthetic testosterone (oxandrolone) has demonstrated significant reduction in hospital length of stay and mortality, and improvement in lean body mass, in both adult and pediatric burn patients.


Positioning

Individualized custom patient positioning should begin immediately upon admission to the burn center and continue along the continuum of care as indicated to prevent and mitigate potential contracture development (Figure 25–9). Appropriate positioning during rest periods is invaluable in preserving function and counteracting all contractile forces of scarring as wounds heal. Antideformity positioning for the entire body can be achieved through splinting, mechanical traction, cut-out foam troughs and mattresses, pillows, strapping mechanisms, casting, and, in some cases, through surgical application of fixators or pins. When fabricating a splint or an orthosis, the clinician must be aware of all mechanical principles of splinting as they relate to pressure, mechanical advantage, torque, rotational forces, first-class levers, friction, reciprocal parallel forces, and material strength.

![Figure 25–9 Initial positioning of the burn patient. In this large pediatric burn, positioning devices are used to support axillary and neck position. Note how the neck splint accommodates the tracheotomy without compromising emergent access.](image)

Positioning and splinting must be designed in a way to:

• Aid in edema reduction.
• Maintain joint alignment.
• Support, protect, and immobilize joints.
• Maintain or increase ROM.
• Maintain tissues in an elongated state.
• Remodel joint and tendon adhesions.
• Promote wound healing.
• Relieve pressure points.
• Protect new surgical sites (grafts or flaps).
• Stabilize or position one or more joints, enabling other joints to function correctly.
• Assist weak muscles to counteract the effects of gravity and assist in functional activity.
• Strengthen weak muscles by exercising against springs or rubber bands.

Devices should:
• Not cause pain.
• Be designed with function in mind.
• Be cosmetically appealing.
• Be easy to apply and remove.
• Be lightweight and low profile.
• Be constructed out of appropriate materials.
• Allow for ventilation in preventing skin or wound maceration.

During the emergent and acute phases of recovery, the patient’s head should be positioned in midline and the head of the bed should be elevated at 30–45 degrees. If the patient’s hips are burned the entire bed should be elevated at the head of the bed by means of shock blocks (wooden blocks 12–16 inches high with recessed slots for bed legs). Modern hospital beds allow for this bed position to be achieved via integrated hydraulics. This position maintains the hips in neutral and may prevent future flexion contractures that might otherwise limit normal ambulating posture. Microstomia of the mouth can be prevented or managed though the use of mouth splints that stretch or open the mouth statically or dynamically in a horizontal, vertical, or circumferential fashion.

The patient’s neck should be positioned in neutral or in slight extension of
approximately 10–15 degrees. This position may be achieved through the use of rolls (towel, sheets, foam cushions) placed along the scapular line. Acutely, the injured shoulders and axillary complex should be positioned at 90 degrees of abduction and 15–20 degrees of horizontal adduction and external rotation. Positioning the shoulder solely in abduction for a prolonged period of time may place the glenohumeral joint at risk for anterior subluxation. Horizontal adduction reduces the potential for injury to the brachial plexus as a result of traction forces acting on the shoulder girdle if it is positioned in abduction and external rotation alone over a period of time. The glenohumeral joint is externally rotated to counteract the potential for deformity and to maintain balance of the soft tissue support of the shoulder complex (Figure 25–10). Positioning of the burned shoulder may be achieved with splints, gel cushions, bedside tables, sky hooks, foam or thermoplastic arm troughs, and commercially available abduction troughs that fit to the patient’s bed.

![Figure 25–10](image.png) Prone shoulder positioning. In addition to splints, use of prone positioning can facilitate improved posture and effective shoulder and trunk position to counteract contractile forces.

In the emergent and acute phase of recovery, the elbow should be positioned in extension and elevation with the goal of avoiding flexion contracture and the risk of posterior exposure of the joint. Full extension is the protected position for the elbow; however, the joint should not be locked in full extension for a prolonged period of time as this may lead to future joint capsular tightness.

The wrist should be positioned in neutral, the meta-carpophalangeal (MCP)
joints should be placed in the maximum degree of flexion permitted by the acute edema, and the interphalangeal (IP) joints should be in full extension. The thumb should be placed in a combination of palmar and radial abduction. This hand position, defined as the intrinsic plus position, places the collateral ligaments at the MCP level and the volar plate (collateral ligaments) at the IP level in maximum stretch, thus preventing future ligamentous tightness that might otherwise lead to chronic hand stiffness. Persistent edema on the dorsum of the hand may cause intrinsic ischemia which, in turn, may result in an intrinsic minus position of the hand (MCP joint hyper-extension, IP joint flexion, thumb adduction, and thumb IP joint flexion). This hand position is defined as a “claw” hand. Postoperatively and during the intermediate and rehabilitation phases of recovery, splinting and positioning of the hand focuses on preventing or correcting contractures. Splinting is considered to help position the hand in the antideformity position depending on the anatomic surface of the burn injury (Figure 25–11).

⚠️ Figure 25–11 Hand splint. Proper splinting is essential to ensure that the proximal interphalangeal joints are extended and the metacarpophalangeal joints are flexed, thus supporting the ligamentous structures in the lengthened position needed for proper hand function and to limit contracture potential.

The injured hips should be positioned in full extension, no rotation, and
approximately 15–20 degrees of symmetric abduction. Initially the knee should be positioned in full extension; however, care should be taken not to lock the joint in hyperextension for long periods of time as this may cause knee capsular tightness and issues with ambulating in the future. The ankle and foot should be positioned in neutral alignment while the patient is resting in bed (Figure 25–12). This position should be achieved in the supine or prone position.

Figure 25–12 Foot splint. Proper position of the ankle is critical to prevent contracture.


Burn Scar Management

A. Initial Treatment Concerns

The burn wound like any other wound heals by forming a scar—fibrous tissue that replaces normal tissues destroyed by injury or disease. Burn scars, if not managed appropriately, may become thick and elevated.

Two types of scar thickening are differentiated, hypertrophic and keloid; both are considered dermatoproliferative disorders of the skin. Hypertrophic scars are not cosmetically appealing, and if they cross joints can restrict skin mobility and function or distract joint position. Typically raised, red, rigid, painful, and pruritic, hypertrophic scars remain within the confines of the burn (Figure 25–13). Keloid scars share some of the same characteristics as hypertrophic scars but extend outside of the margin of the original injury, advancing into the surrounding soft tissue, and are somewhat less contractile in form (Figure 25–14).
Hypertrophic scars are red, raised, and rigid; can limit joint motion; and are painful and pruritic.
Keloid scars have a more pronounced appearance than hypertrophic scars and extend outside the wound margin.

Scar hypertrophy has a prevalence of over 65% in burn injury. Factors that may contribute to the development of hypertrophic scars include wound infection, the patient’s genetic makeup, repeatedly harvested donor sites, the patient’s age, chronic inflammatory processes, location of the injury, and skin tension. Generally, the deeper the burn injury, the longer the inflammatory wound process, and the longer a wound remains open, the higher the potential is for the formation of hypertrophic scar tissue.

As the wound begins the healing process, collagen fibers develop to bridge the wound, forming an immature (active) scar that appears as a red, raised, and rigid mass. Understanding of hypertrophic scar development is limited owing to the lack of well-defined randomized controlled trials and limited consensus on an adequate animal model of abnormal scarring. However, progress continues to be made in unlocking the scar development cascade, including the mechanisms mediating expression of signal mediator transforming growth factor–β (TGF-β),
the overproduction of extracellular matrix, and keratinocyte signal expression.

Many scar treatment options are available throughout the burn injury timeline; these manage only the appearance and effects of the scar and will not completely remove the scar itself. Initially, the focus is on wound closure and minimization of the inflammatory process, which are crucial to restoring the breached skin surface, promoting effective healing, and minimizing the appearance of the scar. Owing to the imbalance in skin hydration, moisturizers play a key role in the early management of scars. The application of moisturizers should be frequent to support damaged sebaceous gland functions and to mitigate the pruritic effect of the dry skin surface.

Pressure therapy during the scar maturation phase may contribute to a more linear, softer, and less vascularized scar. Patients with burn wounds that are either epidermal or superficial partial-thickness, which heal within 7–14 days, do not need pressure therapy. Those whose wounds heal within 14–21 days should be closely monitored so that pressure therapy may be initiated prophylactically if needed. In general, a burn wound that heals after 21 days will require the use of pressure garments.

Burn scars may take up to 2 years or longer to mature. Scars may begin to become thick and raised at approximately 8–12 weeks after the burn wounds close. The amount of pressure needed to suppress hypertrophic scar formation has not been determined. Pressure of as little as 10 mm Hg may be effective in remodeling scar tissue over time. Conversely, high pressures in excess of 40 mm Hg may be destructive to tissues and cause paresthesias. Clinically, custom therapeutic pressure for the prevention, control, and correction of scar hypertrophy averages 24–28 mm Hg, which is approximately equal and opposing to the capillary pressure (25 mm Hg). At this pressure level, many researchers believe that scars may be altered. For pressure therapy to be effective, pressure garments need to be worn at all times, day and night. They should be removed only for bathing and, on occasion, during rehabilitative exercises if they interfere with movements.

Silicone gel sheeting has been a mainstay of burn scar management since the early 1980s. The mechanism by which silicone affects the burn scar is unknown; however, recent research points to a cascade of action in the epidermal signaling mechanism whereby occlusion decreases transepidermal water loss and normalizes the hydration state of keratinocytes, which then signal dermal fibroblasts to downregulate extracellular matrix production. Clinically, silicone has been observed to hydrate burn scars, depress the height of hyper-trophic scars, prevent shrinkage of fresh skin grafts, and increase the pliability of scars,
thus allowing for increased ROM of affected joints.

Once the burn scars have matured enough to tolerate sheering forces, massage may be incorporated into the scar management regimen. Although the benefits of burn scar massage have yet to be scientifically demonstrated, those who use scar massage contend it is an effective modality for maintaining joint mobility. In patients with contractures, it may soften or remodel scar tissues by freeing adherent fibrous bands, thus allowing the scars to become more elastic and improving joint mobility.

Among other methods now being studied, intralesional corticosteroid injection may be of some limited benefit in the treatment of hypertrophic burn scars. The use of pulsed dye laser in the treatment of hypertrophic scars is controversial and not yet routine in most burn centers in North America. Some early preclinical evidence supports the effect of interferon-α as a promising treatment for hypertrophic burn scars, but larger human studies have yet to be done in this area.


**B. Scar Assessment**

No single comprehensive tool exists that objectively measures the volume, pliability, and color of a scar. The widely used Vancouver Burn Scar Assessment is a subjective tool that rates pigmentation, vascularity, pliability, and height of the burn scar. Other scales that have been developed to increase the reliability of subjective assessment and provide construct validity include the Modified Vancouver Scar Scale (MVSS; **Figure 25–15**), Patient and Observer Scar Assessment Scale (POSAS), Burn Specific Health Scale (BSHS), and Matching Assessment of Photographs and Scars (MAPS).
Although a comprehensive objective measure for rating burn scars remains elusive, many individual objective tools are currently available. Scar thickness and height can be measured with noninvasive transcutaneous ultrasound scanning devices (eg, Dermascan, Tissue Ultrasound Palpation System [TUPS]), as well as very simple noninvasive negative molding or 3D imaging. These methods compare the thickness of the traumatized dermal tissue with the dermal tissue of adjacent normal skin at regular intervals. Other devices analyze the color of scars. Some (eg, Minolta Chroma Meter, Labscan, Micro Color) evaluate pigment and vascular changes; others (eg, DermaSpectrometer, Mexameter) incorporate narrow-band reflectance meters. Computerized video camera images can assess the color of scars quantitatively by analyzing images using a custom-written computer program (eg, Image Tool system). Skin pliability, extensibility, and tension can be assessed objectively utilizing cicatrometers, pneumatonometers, or modified tonometers, or by using other dedicated systems (eg, Cutometer, Dermaflex, Dermal Torque Meter). More complex laser-based instruments (eg, laser Doppler flowmeter, laser Doppler imaging, laser speckle imaging) are being utilized for both live blood flow assessment and color assessment. These devices use a combination of red or near-infrared wavelengths and digital image-processing techniques to provide
detailed blood flow assessment in real-time processing.

Several methods currently being developed for objective scar assessment combine one or more high-tech instruments. For example, standardized digital imaging is being combined with spectral modeling to assess scars digitally. Techniques to assess discreet physiologic processes during the scar maturation process are also being pursued, including transcutaneous oxygen tension measurement via skin electrodes, transepidermal water loss assessment (eg, Dermalab, Tewameter, VapoMeter), and hydration of the skin surface (stratum corneum) via electrical conductance meters (eg, CorneoMeter, Skicon-200).


Psychosocial Recovery

Burn injury often causes emotional distress to burn patients and their families, and comprehensive rehabilitation must ensure that psychosocial needs are being addressed. Knowledge of preinjury level of adjustment is important, as it may influence recovery and the treatment plan. Stressors, premorbid conditions, substance abuse, coping skills, socioeconomic status, and family and social dynamics are important factors to consider. In particular, family and supportive social networks have been shown to positively affect recovery from a burn injury.

Post-traumatic stress disorder (PTSD) is the most common psychiatric disorder seen in survivors of major burns, with a prevalence as high as 45%. Nightmares and altered sleep are usually the first symptoms noted. Many burn survivors experience PTSD symptoms during the acute recovery phase, including intrusive memories of the injury. PTSD symptoms lead to greater physical and psychosocial disability, poorer social functioning, and less vitality. PTSD and depression are linked to poor long-term outcomes in adults. PTSD and pain together predict poor functional outcome and increased disability after major burns.

The experience of pain has been found to be a mediating risk factor for PTSD in both pediatric and adult burn patients. Symptoms of depression and agitation related to excessive pain usually subside with adequate pain management. It should be emphasized that the use of opioids and other pain medications will not cause dependence per se if adequately administered and tapered when pain levels decrease.

There is a reciprocal relationship between pain and depression or anxiety. Furthermore, acute pain at the time of discharge has been noted to produce long-term suicidal ideation. After discharge, many burn survivors have significant sleep problems that may be secondary to PTSD symptoms, depression, itching, or pain. Sleep can also affect pain level, and patients with more disturbed sleep tend to require more analgesics. Clinicians should bear in mind that the pain level experienced during the day may not predict a night of disturbed sleep. An individual’s ability to manage both the pain of daily therapy and loss of sleep often requires a comprehensive pharmacologic approach.

Burn injury can cause an alteration in body image, leading to psychological distress and challenges to the patient’s self-esteem. Regular screening at all phases of recovery is important. The greatest challenge for patients whose burns result in significant disfigurement is societal reaction to the disfigurement. The
burned individual must deal with a range of reactions from others, including overt and subtle aversion and prejudice, which can have damaging effects. Research shows that self-image, self-esteem, and self-confidence are threatened in a significant proportion of individuals with burn injuries. Living a solitary life—a form of social death—is a very real possibility for those with extensive disfigurement.

Body image, a person’s mental perception of his or her body, undergoes constant changes in response to input received from the senses as well as the perceptions and reactions of others. Although an individual’s expression of sexuality remains inseparable from body image and self-esteem, the effect of the burn injury on sexuality is often not adequately addressed with burn patients. Medical staff may avoid discussions of sexuality for various reasons: lack of knowledge and resources regarding the effects of burns on sexuality, discomfort with or uncertainty about how to raise the topic, the assumption that it is another team member’s job, time constraints, or lack of privacy. Burn patients should be offered the opportunity to discuss concerns related to sexuality. Ignoring sexuality because “the patient did not mention it” is not appropriate and may suggest that the patient can no longer express himself or herself sexually. Burns may affect sexuality in many ways, including decreased sensation and physical pain, joint stiffness, amputations, fragile skin, fatigue, concerns with sexual health and reproduction, anxiety and stress, lack of sexual drive, impotence or frigidity, medication side effects, distraction with changes in appearance, sense of sexual damage and withdrawal, not knowing how to share feelings about scarred areas, and dressing to cover injuries. Understanding the potential sexual problems of burn patients increases the comfort level for sexual discussions, and blending sexuality with other topics can make the discussion easier for both patient and staff.


Community Reintegration

The ultimate objective of rehabilitation is to reestablish the patient’s preinjury life (home, work, school, recreational, and community) as closely as possible. Burn survivors may feel ambivalent and anxious, fear social rejection, worry about being accepted and receiving social support, and often have significant problems in the areas of home integration, social integration, and productivity.

Return to work or school are essential first steps in community reintegration for most burn patients. Return to work may be affected by the size and severity
of the burn, duration of hospitalization, location of the burn, physical impairment, pain, prior employment history, lack of vocational training, work environment impediments, and psychosocial difficulties. Psychosocial support, positive thinking, and vocational training can assist with return to work. School is an integral part of every child’s normal life, encompassing their connection to friends, learning, and growing up. A child with burns may now look different and may need to wear pressure garments and other therapeutic devices. School reentry programs that educate peers help prevent teasing and potential social isolation by preparing the injured child, his or her family, school personnel, and peers for the child’s transition back to school. Formal programs have been developed, such as *The Journey Back*, by the Phoenix Society for Burn Survivors. The Phoenix Society is a nonprofit organization dedicated to providing support and resources for anyone affected by a burn injury. Many local burn units provide support programs as well (eg, peer support groups, burn camps). Burn camps provide an opportunity for pediatric burn survivors to socialize with other burn survivors and participate in recreational activities in a supportive environment, and may promote increased self-esteem.

Social skills training programs are available to facilitate the burn patient’s positive reintegration to society, improve social comfort, and increase confidence in social interactions. “Be Your Best,” developed by Barbara Kammerer Quayle and the Phoenix Society, and Changing Faces, organized by James Partridge, are two such programs. The strategies they promote prepare patients to answer questions related to the burn, and to deal with staring and stigmatizing behaviors.

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Oster C, Kildal M, Ekselius L: Return to work after burn injury: Burn-injured individuals’ perception of barriers and facilitators. J Burn Care Rehabil


Lower Limb Amputation, Rehabilitation, & Prosthetic Restoration

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The rehabilitative care of adults with acquired amputation of the lower extremity as a result of trauma or dysvascular disease is a challenging area of specialization within the field of physical medicine and rehabilitation. Management relies on careful evaluation of the patient, obtained through the history and physical examination, and presupposes a good understanding of the constantly evolving componentry for prosthetic fitting and rehabilitation.

EPIDEMIOLOGY

More than 100,000 major amputations occur each year in the United States. This number is steadily growing as a result of the increasing incidence of diabetes mellitus and dysvascular disease, and the steadily increasing number of individuals older than 65 years. Dysvascular disease accounts for 82% of all amputations, with 97% of these in the lower limb. Trauma accounts for 16% of amputations, with only 32% of these involving the lower limb. Cancer and infection together account for roughly 1% of amputations, with 75% of these involving the lower limb. The remaining 1% of amputations are the result of congenital deficiency or deformity.

There are more than 1.6 million amputee survivors in the United States, and this number is also steadily increasing. One third of these patients are survivors of dysvascular disease; the other two thirds represent traumatic amputation. The
long-term survival of the former group is dramatically lower than that of patients with trauma-related amputation.

The perioperative survival rate following lower limb amputation due to vascular disease is 94% for patients with amputation below the knee and 83% for patients with amputation above the knee. The 5-year survival rate for patients with amputation below the knee is 48%, and only 22% for patients with amputation above the knee. The long-term survival rate for patients with traumatic amputation is near normal. The rate of revision surgery for the vascular population is 18–25%, with nearly 10% converting from below-the-knee to above-the-knee amputation. Amputation of the remaining limb is also a significant concern. The risk of having amputation of the remaining limb is greater than 50% at 5 years. The rate of revision surgery for the traumatic population is approximately 14%, with a wound infection rate of 34%. Consequently, although many more amputations due to diabetes and dysvascular disease are performed annually compared with amputations from trauma, the long-term survival of the trauma patient is much better and much longer. The “50–50” rule remains unchanged for the dysvascular population: approximately 50% of vascular amputees die within 5 years of the first amputation, and of the survivors, 50% will lose a portion of the remaining lower limb.

The epidemiology of prosthetic fitting is more difficult to determine. Medicare data indicate that approximately 70,000 new prosthetic devices are fitted each year for new and existing lower limb amputees. This means that roughly 50–70% of new amputees are fitted with a prosthesis each year. The annual cost of prosthetic fit and fabrication exceeds $1 billion per year.


EVALUATION OF THE PATIENT

The opportunity to evaluate the patient before amputation is of great value as it allows for a more comfortable physical examination and offers an excellent opportunity to educate the patient about what to expect immediately after amputation and in the long term. Exercising critical muscle groups in preparation for amputation will make the postoperative physical therapy program much easier for the patient to understand and accomplish, and may shorten the rehabilitation phase. Unfortunately, most patients are evaluated after amputation surgery when pain and immobility make the evaluation more difficult for both the physician and the patient.

Once the history and physical examination have been completed, the medical information obtained should be consolidated and conveyed to the remaining team members to assist in the appropriate multidisciplinary management of the patient. Introduction of the team concept is the responsibility of the physician. Explaining the roles of the physical therapist, occupational therapist, prosthetist, psychologist, pedorthist, and case manager facilitates their interaction with the patient and improves patient adherence with the treatment program.

Medical History

A thorough history allows the physician to understand the cause of the amputation and the patient’s experience to date. Both the duration of injury or disability prior to amputation and the exact cause of amputation are important elements to be reviewed. The timeframe of disability helps predict the length of recovery and rehabilitation. Documenting repeated surgical procedures or complications (eg, bypass surgery, skin grafts, muscle flaps, and debridement) provides information important to the hospital course. Diagnostic testing, such as arterial, cardiac, pulmonary, and renal studies, provides relevant details about the patient’s underlying medical condition. Review of the past medical history helps the physician to understand the patient’s potential for rehabilitation and ambulation with a prosthesis. Patients with vascular disease commonly have multiple medical problems, including cardiac disease, diabetes mellitus, peripheral neuropathy, vision impairment, and renal failure. It is also important to review the pain history with the patient. Any pain prior to amputation should be assessed, in addition to postoperative surgical pain. Patients with acquired amputation commonly have phantom sensation of the limb but may also have
phantom pain of the limb that interferes with their ability to participate in therapy, self-care, or sleep. The treatment for each of these types of pain should also be reviewed carefully.

### Social History

Review of the social history should include assessment of the social support system at home and the direct involvement of that support system since surgery. Careful review of the patient’s previous level of activity and personal responsibilities should be conducted. Specific questions addressing the patient’s exact level of ambulation just before amputation are critical as they help predict the patient’s functional ambulation with a prosthesis. Examples include, “When was the last time you walked on two feet?” or “Could you walk one block outside?” The patient’s work history should be reviewed for potential return to work. Interest in hobbies and other activities at home should be elicited. The patient’s concerns regarding ability to return to these activities should be addressed early in the rehabilitation phase. Psychological status, including any specific concerns about cognitive status or prospective concern about depression related to the amputation, should also be addressed. The patient’s previous experience with prosthetic devices or conceptions about such devices should be elicited in this early phase to avoid any misunderstanding about the actual function or cosmesis of prosthetic devices. Finally, the history should include issues related to family responsibilities (eg, being a caretaker for younger children) or relationships (eg, the patient’s spouse or significant other). Often the presence or lack of a strong social support system is the factor determining whether a patient is able to return to the home environment and resume many of his or her previous activities.

### Physical Examination

Physical examination of the patient following amputation should include routine components involving overall hygiene and body habitus; head, eyes, ear, nose, and throat (HEENT); and cardiopulmonary, neurologic, and cognitive examinations. Careful attention should be given to the musculo-skeletal and sensory examinations as these will have a direct impact on prosthetic fitting and rehabilitation.

The uninvolved limbs should be examined first, before moving to the
involved limb, so the patient will understand the type of examination. Evaluation of the involved limb should clearly define the appropriate level of amputation, including bone length and soft tissue coverage. Skin integrity should be described carefully and a photograph taken, if possible. Overall limb shape, including measurements of the limb, should be noted. Healing at the surgical site or any remaining wounds should be carefully documented. Areas of skin grafting, scarring, or soft tissue adherence should be documented. Tenderness to palpation throughout the residual limb should be assessed as this will determine tolerance to prosthetic fit. Sensation throughout the soft tissue and skin should be recorded.

Manual muscle testing should include all four limbs, but critical areas of strength testing are the hip extensors and hip abductors, knee flexors and extensors, and ankle dorsiflexion–plantar flexion bilaterally, as appropriate. Strength of the upper limbs should also be tested, in anticipation of the need for upper limb strength to use an assistive device both before and after prosthetic fitting. The critical muscles in the upper limb include those involved in grip, elbow extensors, and shoulder depressors. Hand dexterity should be assessed, as peripheral neuropathy may impair sensation and fine motor skills in the hands.

Range of motion for hip extension, knee extension, and ankle dorsiflexion should be assessed because flexion contractures are common following amputation surgery and periods of immobilization. Careful examination of the remaining foot should include assessment of sensation, skin integrity, bony architecture, and vascular status. The remaining foot in the patient with dysvascular disease and major amputation is at high risk for compromised circulation and skin breakdown.

**CLASSIFICATION & LEVELS OF LOWER LIMB AMPUTATION**

A clear description and understanding of the level of amputation is necessary to understand the prosthetic restoration of the lost limb and the rehabilitation program that will accompany it. The biomechanical changes that occur at each level of limb loss influence the energy cost of ambulation and cardiac demand. The tolerance to prosthetic fit and long-term functional outcome are clearly determined by multiple factors, but most importantly by the level of amputation, soft tissue coverage, proximal muscle strength, and overall medical condition.
**Toe Amputation**

Loss of single or multiple toes often has little functional consequence for patients. From a biomechanical perspective, the architecture of the foot is well preserved and much of the weight-bearing surface remains. Use of a protective foot orthotic with a toe filler should be considered to prevent further complications to the remainder of the foot. However, amputation of a toe due to dysvascular disease clearly is a sign that the patient is at high risk for a higher level of amputation, and efforts should be made to monitor and preserve circulation to the remainder of the foot. Protection of both feet with appropriate footwear should be a lifetime management plan.

**Partial Foot Amputation**

Transmetatarsal amputation (TMA) is the most common level of amputation when disease or injury compromises the foot proximal to the metatarsal-phalangeal joint. TMA is performed through the midshaft of the metatarsals, using the plantar flap to cover the end of the metatarsal bones (Figure 26–1). Ideally, sensate tissue from the plantar surface will help protect the amputation site during future ambulation. The preservation of dorsiflexion, plantar flexion, inversion, and eversion leaves a very stable and mobile foot for indoor and outdoor ambulation. Prosthetic fitting with a TMA prosthesis includes a combination of foot orthotic and fore-foot filler inside an appropriate orthopedic-type shoe. A rigid plate under the prosthesis or a rigid sole on the shoe helps deflect any significant pressure at push-off to the end of the shoe rather than the end of the amputation site.
Other levels of partial foot amputation are less common and sometimes considered less desirable because they compromise the weight-bearing column of the skeleton or the balance of dorsiflexor and plantar flexor muscles. The Lisfranc level of amputation includes loss of all metatarsals and toes, while leaving some tarsals in place (Figure 26–2). This level of amputation compromises the arch of the foot and some of the insertion sites of the dorsiflexors, promoting a plantar-flexed position of the remainder of the foot. This deformity is even more exaggerated with the Chopart level of amputation, in which all tarsals and metatarsals are lost to amputation (Figure 26–3). The prosthetic fitting for these other levels of partial foot amputation requires some type of rigid boot or gauntlet to capture the remaining foot and restore the arch. Unopposed plantar flexion inevitably results in plantar-flexion contracture and weight bearing onto the distal calcaneus, with resultant skin breakdown. To prevent this problem, the Chopart-level prosthesis may incorporate offloading and immobilization of the remainder of the foot, using a bivalve prosthetic shell that transfers the load to the calf or to the patellar tendon area and immobilizes the calcaneus.

▲ Figure 26–1 Transmetatarsal amputation with a well-preserved arch and mobile ankle.
Figure 26–2 Lisfranc level of partial foot amputation with preserved tarsals, but compromised arch.

Figure 26–3 Chopart level of partial foot amputation with only the tibia, talus, and calcaneus preserved. Weight bearing is compromised and insertion of the dorsiflexors is lost, creating a plantar-flexed calcaneus.
Ankle Amputation

The most common ankle disarticulation procedure is Syme’s amputation. This procedure includes true disarticulation of the ankle and then removal of the medial and lateral malleolus to create a level weight-bearing surface on the tibial articular cartilage. The heel pad must remain intact because it is placed on the end of the tibia to obtain an end-bearing structure (Figure 26–4). The benefit of Syme’s amputation is that it permits partial end bearing and retains the full length of the tibia for excellent leverage and control of the prosthesis. The major disadvantage is the bulk of the distal end, which may lead to a cosmetically unappealing prosthetic socket design. There are also limitations in prosthetic foot availability due to the limited space and height available. The socket design commonly includes a removable medial or posterior window to allow the more bulbous distal end to reach the bottom of the socket (Figure 26–5). Once the window is closed, it captures the narrow portion of the distal tibia creating a self-suspending socket design. Commonly, 50% of weight is placed distally on the tibia and 50% is at the patellar tendon and the medial tibial flair. Migration of the heel pad off the end of the tibia is a common complication and may prevent end bearing.
Figure 26–4 Syme’s ankle disarticulation amputation with the medial and lateral malleoli removed, and the heel pad placed on the distal tibia for partial weight bearing.
**Transtibial Amputation**

The appropriate bone length in transtibial amputation depends on the quality of the soft tissue, extent of underlying disease, and length of the gastrocnemius–soleus muscle mass. The ideal length can be determined by identifying the musculotendinous junction of the gastrocnemius–soleus muscle and resecting the bone 2–3 cm proximal to this point. This approach should allow sufficient muscle for flap coverage of the distal tibia and fibula, which commonly occurs in the middle or proximal third of the tibia, but rarely in the distal third (Figure 26–6). The minimum length necessary for transtibial amputation is determined.
by the insertion of the patellar tendon and hamstring tendons. Compromising flexor or extensor mechanisms at the knee will leave a flail segment that may be impossible to fit with a prosthesis. Reconstructive techniques to maintain amputation below the level of the knee should be considered when appropriate. These may include soft tissue reconstruction using rotation flaps or free island muscle flaps to cover bony structures. Skin grafting is also fairly well tolerated at this level of amputation. Preservation of the amputation level below the knee at almost any cost should be considered, since the functional outcome is nearly always better compared with above-the-knee amputation.

![Figure 26–6](image)

**Figure 26–6** Ideal transtibial amputation, with long bone length and good soft tissue coverage using a gastrocnemius–soleus flap to cover distal tibia.

The socket design for transtibial amputation commonly includes total contact with more pressure at the patellar tendon, medial tibial flair, and gastrocnemius muscle. Some newer socket designs also capture the anterior tibial plateau and femoral condyles for additional weight-bearing surfaces. Pressure relief should be provided at the distal tibia and fibula, the fibula head, and the medial and lateral knee joint. The socket walls commonly rise higher on the medial and lateral sides to provide additional stability but are lower at the posterior wall to maximize knee flexion for sitting. The posterior brim of the socket should match the height of the patellar tendon–bearing (PTB) bar anteriorly. There is always a soft interface and rigid outer frame in the transtibial socket. The interface is
commonly a soft foam material for a temporary prosthesis, and a gel material for the permanent prosthesis. Initial use of the soft foam material allows for adjustments to be made with pads to accommodate for volume loss as the limb shrinks and atrophies. Once the patient’s limb shape has stabilized, the use of a gel liner with or without pin suspension provides a stable cushion interface for higher-level activity (Figure 26–7). The outer rigid socket may utilize a carbon fiber laminate or a thermoplastic design. The suspension options include a supracondylar wedge, elastic sleeve, suction system, thigh corset, and elevated vacuum system.

Figure 26–7 Gel liner with a pin (left) for soft interface and suspension. Rigid transtibial socket using a PTB weight-bearing design proximally and shuttle lock mechanism distally (right).

The remainder of the prosthesis can be endoskeletal or exoskeletal in design. The endoskeletal design incorporates modular components connecting the socket to the pylon and then to the foot. This design permits greater adjustability for length and angular alignment. It also facilitates replacement of components,
including socket, pylon, and foot. The exoskeletal design has a rigid outer shell extending from the socket to the foot. This design is touted as having greater durability and tolerance to aggressive outdoor activities; however, there is significant compromise of adjustability. Therefore, the exoskeletal design is reserved for specific populations such as active children, or adults involved in outdoor vocations such as construction or landscaping. The choice of prosthetic feet is reviewed later in the chapter.

### Knee Amputation

This level of amputation should be used exclusively for traumatic amputation and not in the dysvascular population. The primary advantage of knee disarticulation is end bearing, and dysvascular skin commonly will not tolerate the end-bearing socket design. If the full length of the femur can be preserved and good sensate skin can be used to cover the distal end, then the socket design can avoid any weight-bearing issues related to the groin or ischium, such as occur with transfemoral amputation. Knee disarticulation also preserves the full length of the femur, allowing for excellent control of the prosthesis and improved functional outcome compared with transfemoral amputation. However, retaining the full length of the femur and the bulk of the femoral condyles leads to a cosmetically unappealing socket design. It also limits the choice of prosthetic knee components, because most knee units require 3–5 cm of length for the mounting plate and body of the joint. This becomes an issue when the patient is sitting, causing a disproportionate length of the femur compared with the uninvolved side.

The socket design for a knee disarticulation prosthesis is similar to that for an ankle disarticulation, with an open medial window in the rigid socket to allow the wider distal portion of the residual limb to reach the base of the socket (Figure 26–8). The inner liner material can be elastic or have a split liner, enabling it to expand as the wider distal end passes by the midsection of the socket. Once the residual limb is properly seated into the socket base, the medial window is closed to create a self-suspending socket design. The proximal brim of a knee disarticulation socket typically stops at a point two-thirds of the way up the thigh, resulting in partial weight bearing on the thigh musculature and partial weight bearing on the end of the femur. This allows for very comfortable sitting and standing positions compared with transfemoral sockets. Either a flexible thermoplastic inner socket or a gel liner may be used for the interface material to cushion the limb inside a rigid outer frame of carbon fiber. In the
temporary prosthesis, the patient may apply socks to accommodate for volume loss, but once the patient is fitted into the permanent prosthesis a tight fit is expected, without the need for additional socks.

![Figure 26–8 Knee disarticulation socket with a medial window for ease of donning and self-suspending design. Weight bearing is shared between the distal femur and thigh muscles.](image)

**Figure 26–8** Knee disarticulation socket with a medial window for ease of donning and self-suspending design. Weight bearing is shared between the distal femur and thigh muscles.

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**Transfemoral Amputation**

In contrast to the transtibial amputation, the ideal length for transfemoral amputation should be in the distal third of the femur just above the flairs of the femoral condyles (Figure 26–9). In general, longer is better with this level of amputation. Sufficient muscle mass from the quadriceps and ham-string
musculature is usually available to cover the distal end of the femur. The muscles should be secured through myodesis or myoplasty to prevent retraction of the muscle and exposure of the distal bone structures. Ensuring that adductor muscles are properly secured also ensures proper position and control of the femur during prosthetic use. Unfortunately, the distal residual limb cannot tolerate end bearing; therefore, most of the weight-bearing characteristics of a transfemoral socket are in the proximal portion of the socket. Total contact is still recommended, but the primary weight-bearing structures are the ischium and the gluteal muscles, with some weight-bearing into the thigh muscles. Capturing the ischium with the medial posterior brim of the socket is critical to prevent lateral shift of the socket during stance phase (Figure 26–10).

▲ Figure 26–9 Long transfemoral amputation with good soft tissue coverage distally.
The contemporary socket design is referred to as ischial containment and narrow medial–lateral dimension. The interface is always a soft thermoplastic material and the outer frame is rigid. The soft thermoplastic interface provides some proximal control and protects the skin and soft tissue from the rigid frame. Another layer of interface material (eg, socks or a gel liner) can be used for additional protection. A semi-suction design with a suspension waist belt is most commonly used in the temporary prosthesis. A full suction design is more commonly used in a permanent prosthesis to avoid the necessity for a waist belt. The suction design can involve placement of bare skin directly against the thermoplastic inner socket to create the most intimate contact and control of the prosthesis. The patient dons the prosthesis using a lotion or a donning bag to prevent traction on the skin as the residual limb slides into the socket. The use of a gel liner is becoming more popular as it provides an additional layer of protection for the skin and soft tissue. The gel liner can be connected to the socket using a distal pin suspension, a strap suspension, or a sealing ring.
suspension mechanism (Figure 26–11). Active vacuum mechanisms using a mechanical or electrical pump to evacuate air from the space between the gel liner and the socket are becoming more popular in both transfemoral and transtibial socket designs. These designs require an air seal proximally to prevent air leaking in during ambulation.

▲ Figure 26–11 Transfemoral socket with a gel liner and plastic strap into a ratchet lock suspension.

The alignment of the socket relative to the position of the femur is of critical
importance. A neutral or adducted position of the femur is ideal to provide pelvic stability and normalize the gait pattern. The vigorous firing of hip abductor muscles to stabilize the pelvis in stance often promotes femoral abduction within the socket, causing distal femur pain (Figure 26–12). Proper contouring of the lateral wall of a transfemoral socket will help to promote the preferred adducted position of the femur.

![Figure 26–12](image.png)

**Figure 26–12** Radiographic image of a transfemoral residual limb within the socket. Poor ischial containment causes a lateral shift of the socket and abducted position of the femur.

With mid-length or shorter transfemoral amputation, the ability to maintain the femur in the ideal position becomes much more challenging. The patient’s ability to transfer force from the femur to the socket and, therefore, control the prosthesis also becomes more challenging. Patients with mid-length and shorter transfemoral amputation may require an assistive device for even moderate
ambulation. Once the femoral length approaches the level of the ischium, the movement of the remaining short segment of femur can no longer be captured. Therefore, patients with these very short levels of transfemoral amputation may be fitted with a socket design similar to that used for hip disarticulation. There is some advantage to leaving the short segment of femur in place to provide a greater weight-bearing surface within the socket.

### Hip Amputation

This level of amputation poses an extremely challenging situation for the physician, the patient, and the prosthetist. At this level, the prosthesis becomes so bulky and heavy that many patients abandon its use. Additionally, the challenge of controlling three joints may be too difficult for some patients. Because the control of the prosthesis comes from a pelvic tilt maneuver, if proximal muscle strength or pelvic mobility is severely compromised, use of a hip disarticulation prosthesis may be difficult or impossible. Furthermore, the prosthesis is often uncomfortable in both sitting and standing positions and interferes with toileting activities, necessitating its removal several times daily. Many patients report that they are able to ambulate faster and more easily using an assistive device (eg, crutches or a walker without a prosthesis) than they can with a prosthesis. Others with hip disarticulation find the energy cost required to use the prosthesis to be too great. Such patients may become wheelchair bound for primary mobility. As a result, fewer than 50% of patients with hip disarticulation use a prosthesis for functional ambulation. When hip disarticulation amputation is necessary, the gluteal muscles should be used as a muscle flap to cover the ischium and pelvic structures.

The traditional socket design is called a bucket design because the patient essentially sits within the rigid socket, which captures all of the involved side and a portion of the uninvolved side of the pelvis. The bucket design has a thermoplastic inner layer that may also incorporate a flexible segment that wraps around the contralateral side of the pelvis (Figure 26–13). Velcro straps or buckles are used to securely tighten the socket to the pelvis. The hip joint is commonly mounted in an anterior position on the socket to make sitting more comfortable.
▲ Figure 26–13 Hip disarticulation prosthesis with bucket-style socket and anterior mount hip joint.

Hemipelvectomy and Hemicorporectomy

These very proximal levels of amputation may be caused by significant trauma or invasive cancers. Prosthetic fitting becomes very challenging and most patients abandoned prosthetic use. Use of a wheelchair for primary mobility is common, with prosthetic use only for limited distance or for exercise purposes. The details of these very challenging levels of amputation are beyond the scope of this chapter.

REHABILITATION OF THE LOWER LIMB AMPUTEE
Successful rehabilitation following lower limb amputation involves several key areas, described below. The patient should be evaluated as early as possible following amputation surgery to minimize problems and complications. Ideally, the physiatrist should direct the rehabilitation care of the amputee patient, including ordering therapy services, writing the prescription for the prosthetic devices, and discharge planning. Incorporating all team members—patient, prosthetist, therapists, social worker, psychologist, and case manager—in the plan of care is essential to achieving a successful outcome.

Education

One of the primary roles of the physician is to educate the patient and other team members. In the preprosthetic phase, education includes explaining the need for early therapy to mobilize joints and maintain strength. Education during this phase should also address residual limb shaping and shrinking, pain control, and psychological issues. Conveying such information helps to promote independence in self-care, which in turn builds the patient’s confidence and self-esteem. Encouraging the patient to talk to other patients who have undergone a similar amputation can aid understanding of the process and the potential outcome. Explaining the clinical timeframe helps the patient to plan his or her life and activities.

Perioperative Phase

Following amputation surgery, the patient is likely to spend 1–5 days in an acute care hospital followed by transfer to another facility. Many patients are transferred to acute or subacute rehabilitation programs for 14–21 days to continue with the preprosthetic therapy program, counseling, and medical care. A small number of patients are able to manage this preprosthetic phase at home, using home care or out-patient therapy services. However, this population requires exceptional social support and an appropriate home situation. Commonly the family is unable to provide this level of care immediately after surgery. Once the patient shows sufficient healing (generally by week 4 or 5), the sutures or staples are removed and consideration of the temporary prosthesis can proceed.

Preprosthetic Phase
During the preprosthetic phase, shrinking and shaping of the residual limb is of critical concern. Appropriate elastic wrapping generally works best during the first 4–6 weeks, until sutures and staples are removed. Elastic wrapping must be done in a figure-of-eight fashion and the limb rewrapped several times daily, following inspection of the skin. A shrinker sock can be provided once the staples or sutures are removed, making it easier for the patient to perform self-care. Regardless of the shrinking device used, it should be kept in place 23–24 hours per day.

The preprosthetic therapy program should include strengthening the proximal muscles of both upper and lower limbs to prepare for ambulation. As mentioned previously, exercises to strengthen upper limb muscles should include grip, elbow extensors, and shoulder depressors to prepare the patient for use of assistive devices. The lower limb muscles should include hip extensors and abductors, knee extensors, and ankle muscles of the remaining limb. Core muscles of the trunk and cardiovascular conditioning should also be included, always respecting any cardiac precautions. The patient should be able to perform the exercises on his or her own, including wheelchair press-ups while in the wheelchair or bridging while in the bed.

Exercises that preserve essential range of motion for hip extension and knee extension should also be part of the therapy program. Cardiovascular conditioning with upper limb ergometer or lower limb resistance exercises is important to prepare the patient for the additional energy requirements of ambulation with a prosthesis. The therapy program should also include desensitization of the residual limb through gentle tapping, rubbing, or massaging. Modalities such as hot and cold packs, vibration, and transcutaneous electrical nerve stimulation (TENS) can be used to desensitize the limb.

Early mobilization from bed to wheelchair and to toilet is critical. Independence in dressing, bathing, toileting, and feeding should be an early goal for occupational therapy. Standing and hopping in parallel bars or using a walker may be appropriate for some patients prior to prosthetic fitting. Proper wheelchair use, both in and out of doors, should be reviewed with the patient and family.

Education about the prosthetic fitting and training process should be started early. Explaining the casting and fabrication process of the socket, as well as component selection, will help the patient understand some of the complexities of the prosthetic process. Cosmetic and functional considerations of various options should be reviewed to avoid misunderstandings and ensure adequate expectations relating to the completed prosthesis.
Prosthetic Rehabilitation

Insurance coverage for the prosthesis and training should be carefully investigated before fitting any device to avoid financial issues. Typically, fabrication of the temporary prosthesis begins 3–4 weeks after amputation. This step depends on appropriate healing and shaping of the residual limb. If healing is delayed because of wound issues, the pre-prosthetic therapy program should continue until these are resolved. If significant delay is anticipated, then alternative mobility devices should be considered, such as a kneeling scooter for a transtibial amputee. Once the temporary prosthesis is delivered to the patient, the prosthetic rehabilitation program should begin. This stage of rehabilitation can be accomplished through inpatient or outpatient programs. If outpatient therapy is appropriate, a minimum of 4–6 weeks of outpatient therapy will likely be necessary for patients with below-the-knee amputation. For those with above-the-knee amputation, 6–12 weeks of outpatient therapy may be necessary to achieve functional indoor and outdoor ambulation. Physician involvement throughout this time period is essential to monitor skin tolerance of the prosthesis, pain management, cardiovascular status, and psychological concerns.

Instructions for prosthetic use should include wearing time and weight-bearing activities. The wearing time should start with only 1–2 hours per day and advance by 1 hour daily if tolerated. All patients should start partial weight bearing in their prosthesis device until cleared by the physician or therapy services. Appropriate donning and doffing of the device should be reviewed early to ensure proper fitting of the prosthesis and to minimize skin problems. It is common for patients to require prosthetic socks to control proper fit of the socket as the residual limb shrinks in the first 3 months. Proper sock management should be addressed by the prosthetist and therapist on a regular basis. The patient should apply enough socks for a snug, intimate fit of the socket.

If the patient is unable to obtain sufficient fit of the socket, then modification by the prosthetist may be necessary. Issues such as pistoning or bottoming out within the socket can cause skin problems and pain. If significant volume loss occurs, replacement of the socket may be necessary, even in a temporary prosthesis, to achieve an appropriate fit. Therefore, both the physician and prosthetist must follow the patient on a regular basis to monitor these changes. Proper skin care and hygiene should also be included in the instructions to the patient. Daily skin checks by the patient or family will help to catch any problems related to pressure or irritation at an early stage. Excessive perspiration
can be controlled with topical application of antiperspirants or silver socks. Monitoring of the patient’s weight and volume of the residual limb should also be incorporated into a regular routine.


Therapy Program

The actual physical therapy prosthetic program should include progressive ambulation using assistive devices. Patients start on a level surface and progress to uneven outdoor terrain. The progression from walker to crutches to cane is common as the patient’s strength and mobility with the prosthesis improve. Car transfers, toilet transfers, and stair climbing are included as essential activities. Return to outdoor or recreational activities is accomplished as the patient achieves a higher level of skill. The return to driving should be considered on an individual basis. Loss of the right lower extremity significantly compromises driving ability, and use of the prosthesis to control a gas or brake pedal may be illegal in certain states.

Ultimately, the patient’s use of the prosthesis and functional outcome depend most on good socket fit and proper training. The proper selection of prosthetic components reflects the patient’s functional needs and limitations, and his or her performance during the temporary prosthetic phase.


Energy Expenditure with Prosthetic Ambulation

Most of the literature supports the concept that energy expenditure is increased when ambulating with a prosthetic device. Various methods of measuring energy cost at different speeds and under different circumstances have led to a plethora of confusing results. Most studies allow patients to ambulate at self-selected
speeds, making it difficult to compare results. Nonetheless, there is general consensus that transtibial amputation requires 20–40% additional energy over normal ambulation. Transfemoral amputation requires 60–80% additional energy for level surface ambulation. Hip disarticulation amputation may require as much as 100% additional energy cost for ambulation. Amputation of bilateral lower extremities is associated with variable results based on level of amputation and comorbidities. A few general conclusions can, however, be drawn. Below-the-knee amputation uses less energy than above-the-knee ambulation. Patients who have undergone amputation for traumatic reasons have lower energy costs for ambulation than those with underlying dysvascular disease. Amputee patients have a slower gait speed than normal controls, and the best energy efficiency is at self-selected speeds. Selection of componentry and prosthetic weight appear to have little or no impact on the energy cost of ambulation on a level surface.

### Functional Outcome

There is no consistent functional outcome measure for use in comparing various levels of amputation or prosthetic designs. Because of the variable population, it has been difficult to establish a consistent objective tool to assess the use of these prostheses. A survey of studies published in the past two decades indicates that approximately 80% of younger traumatic amputees ambulate with a prosthesis compared with only 56% of older dysvascular patients. Remember that morbidity and mortality are very high in the older dysvascular population. The only predictive factors for improved functional outcome appear to be age and amputation below the knee. Correlations between specific prosthetic componentry and functional outcome are lacking.

### Prescription Writing & Prosthetic Componentry

The goal in rehabilitation of a patient with lower limb amputation is to generate a treatment plan that maximizes the patient’s functional outcome with or without a prosthesis. The treatment plan should include the prosthetic prescription, if appropriate; proper footwear; therapy program; education; and long-term followup. The prosthetic prescription should be formulated and agreed to by the physician, prosthetist, and patient. Key elements of the prosthetic prescription include appropriate identifying information for the patient (eg, name, age, sex, date of birth, and medical record number), underlying medical conditions, appropriate level of amputation, long-term prognosis, and anticipated functional
level as defined by Medicare (Table 26–1; see also Prognosis, later). Details of the components and design of the prosthesis should be included and will help ensure that the appropriate device is fabricated and delivered to the patient. Appropriate justification for the product and duration of need should be included on the prescription. Finally, the prosthetic provider and prescribing physician should be clearly outlined on the prescription. For example, a prescription for a transtibial prosthesis should include the socket design with materials for the soft interface and rigid frame, in addition to suspension, pylon, and prosthetic foot details. The prescription for a transfemoral prosthesis would include all of the above along with the prosthetic knee component.

Table 26–1 Functional levels of ambulation according to Medicare guidelines.

<table>
<thead>
<tr>
<th>Functional Level</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level 1</td>
<td>Patient is nonambulatory</td>
</tr>
<tr>
<td>Level 2</td>
<td>Patient can tolerate limited community ambulation, but is restricted by distance or obstacles in the community</td>
</tr>
<tr>
<td>Level 3</td>
<td>Patient is considered an unlimited community ambulator for long distance at variable cadence</td>
</tr>
<tr>
<td>Level 4</td>
<td>Patient can tolerate high-energy activities related to work or sports</td>
</tr>
</tbody>
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LOWER LIMB PROSTHETICS

A. Prosthetic Foot

The selection of a prosthetic foot is based on the patient’s anticipated functional level of ambulation, including indoor and outdoor mobility, uneven terrain, distance, or any special needs of the patient. There are four basic categories of prosthetic feet.

1. SAFE and SACH—The simplest category includes SAFE (solid ankle
flexible endoskeletal) and SACH (solid ankle cushion heel) feet, which have essentially no true movement and no dynamic response (Figure 26–14). This category of foot is generally recommended for limited-distance household ambulators.

![SAFE and SACH prosthetic feet used for functional level 1 ambulators.](image)

2. **Single axis**—The single-axis foot allows some controlled dorsiflexion and plantar flexion. This type of foot works well on a level surface for moderate distances at a fixed cadence.

3. **Multi-axial**—The multi-axial foot is most appropriate for limited outdoor distances on uneven terrain, also at a fixed cadence.

4. **Energy storing**—Energy-storing feet are primarily fabricated of carbon fiber or similar material to assist in propulsion during the late stages of stance (Figure 26–15). This type of foot is most appropriate for longer distance ambulation at a variable cadence. Heel-height adjustable feet are available to accommodate for higher heeled shoes or boots for both men and women. In addition, supplemental ankle joints can be applied to an existing foot to increase motion, or absorb shock or torque. The next generation of prosthetic feet will include hydraulic or electronic-controlled ankle–foot mechanisms to provide true propulsion in the late stance phase. Specialized prosthetic feet are currently available for sprinting, swimming, rock climbing, and other recreational activities.
B. Prosthetic Knees

Prosthetic knees are divided into six categories based on functional activities.

1. Manual lock—The simplest category has a manual lock mechanism for patients who have limited musculature at the hip and, therefore, may have compromised control of the prosthesis.

2. Weight activated—The second category has a weight-activated locking mechanism for stance, but then unlocks for the swing phase (Figure 26–16). This type of prosthetic knee is very lightweight and very durable, but provides only fixed cadence.
3. **Polycentric**—Polycentric joints are primarily used for patients with knee disarticulation amputation or long transfemoral amputation, where space is limited. The main advantage of the polycentric knee joint is that the componentry is primarily in the shin section of the prosthesis and, therefore, a disproportionately long thigh section of the prosthesis is avoided (Figure 26–17). The polycentric knee joint has a migrating axis of rotation that provides inherent stability at full extension to approximately 20 degrees of knee flexion.
4. **Pneumatic**—Pneumatic piston–controlled knees provide variable cadence during the swing phase, but limited stance phase control.

5. **Hydraulic**—Hydraulic piston–controlled knees, which provide both swing and stance phase control with variable cadence (Figure 26–18), are commonly used for higher-level activity, including recreational sports. There is significant adjustability for both swing and stance phase resistance to accommodate for variable patient activities.
Hydraulic knee joint with mechanically adjustable flexion and extension resistance allowing for variable cadence, commonly used for functional level 3 and 4 ambulators.

6. Microprocessor—Microprocessor-controlled hydraulic knees (Figure 26–19) can be programmed by the prosthetist to customize multiple variables for each individual patient during the swing and stance phases. Some microprocessor units also have a self-learning capability, by which they adjust to the patient’s walking patterns. The microprocessor senses vertical loading and knee rotation to recalibrate the knee stability 60 times per second. The safety benefits of these advances would seem to be quite obvious, but clear-cut evidence is lacking. Additionally, there are some drawbacks to microprocessor-controlled knees, including increased cost and weight. Furthermore, the microprocessor knee unit needs regular recharging of the battery to maintain proper functioning, and patients must take care to avoid wet environments or impact.
Figure 26–19 Microprocessor-controlled hydraulic knee joint with computer-programmed flexion and extension resistance for patient-specific swing and stance phase control.
COMPLICATIONS

The medical complications of lower limb amputation include skin irritation or breakdown from pressure related to the prosthesis, bone-related issues, pain control, dysvascular issues, and issues related to the remaining foot.

Skin Issues

Excessive pressure from the prosthesis can cause skin irritation, pain, skin breakdown, and potentially deep infection. The residual limb must be appropriately maintained within the socket, with pressure applied to tolerant areas and little or no motion of the residual limb allowed within the socket. Soft tissue intolerance to pressure is heralded by redness and pain. Redistribution of pressure within the socket resolves this problem. If the socket cannot be modified or replaced to help redistribute pressure, then offloading with an assistive device may be the appropriate solution. The use of a cane, crutch, or walker may be appropriate for a period of time following skin irritation or breakdown to allow healing to occur. The selection of a soft interface is also critical to help protect underlying skin and bony prominences of the residual limb. The introduction of the gel liner material for both transtibial and transfemoral amputations has improved the ability of the skin to tolerate pressures. However, there are significant hygiene issues related to the use of gel liners. Daily cleansing of the skin and of the gel liner is essential to prevent maceration of the skin or development of bacterial and fungal infections.

Bone Growth

Careful attention to the bony architecture of the residual limb will help in designing an appropriate prosthetic socket for each patient. It is common for younger patients with traumatic amputation to develop bone spurs or heterotopic bone formation 3–6 months after surgery (Figure 26–20). This type of bone abnormality may continue to change for up to 2 years following amputation surgery. Older dysvascular patients may develop some bone changes following
surgery, as well. Radiographic imaging is indicated when the patient develops increasing pain in the residual limb despite a well-fitting socket.

![Figure 26–20](image) Radiographic image of a transtibial residual limb, showing heterotopic ossification and hardware used to stabilize a fracture of the tibial plateau.

**Pain**

Pain management following amputation requires an understanding of the several sources of pain. Surgical pain following amputation should be managed aggressively with narcotic and nonnarcotic medication, and then tapered aggressively within 2–4 weeks following surgery. Persistent pain within a well-healed residual limb may indicate neuroma or nerve irritation within the residual limb. If neuroma is identified by clinical examination or imaging studies, direct
injection of the neuroma may be successful in controlling pain. Application of topical medications, including lidocaine and antiinflammatory drugs, may also prove beneficial when the nerve is superficial. Soft tissue inflammation and even bursa formation within the residual limb can be treated with topical or oral antiinflammatory drugs, in addition to hot and cold modalities. However, it is most important to reassess the fit of the socket to minimize any shear forces or excessive pressure within the socket during ambulation.

Phantom pain should be addressed when it interferes with sleep or daily activities. Various oral medications have been used to block these pain signals, including tricyclic antidepressants, γ-aminobutyric acid (GABA) inhibitors, norepinephrine and serotonin blockers, antiepileptic medications, and even antiarrhythmic medications. Nonpharmacologic treatments have included TENS, vibration, massage, mirror therapy, acupuncture, hot and cold modalities, and hypnosis. There has been no consistent response from any of the abovementioned treatments, but all have produced some improvement over no treatment at all.

### Vascular Issues

Pain within the residual limb or the contralateral limb may represent ongoing dysvascular disease with resultant claudication. Vascular studies to assess the status of the arterial blood supply to the residual limb or to the remaining foot should be conducted when persistent pain is reported. Revascularization procedures should be considered, when appropriate, for the amputation limb or the remaining limb. Careful protection of the remaining contralateral foot is essential preventative care when the patient has underlying diabetes and peripheral vascular disease. A custom-molded foot orthotic and appropriate extra-depth orthopedic-type shoes should be part of the routine management of any patient who undergoes amputation as a complication of diabetes.


### PROGNOSIS
The decision-making process relating to selection of prosthetic components should reflect a team discussion led by the physician that considers the patient’s underlying medical status, previous functional level, level of amputation, and anticipated functional level following prosthetic fitting and training. The Medicare guidelines are the standard currently used to determine functional level (see Table 26–1) and encompass four categories of activity related to mobility. It is the physician’s responsibility to document the anticipated functional level in the patient’s medical chart and on the prescription for the prosthetic device. Because a level 0 patient is nonambulatory, no prosthesis should be ordered. The Medicare guidelines indicate that a level 1 patient would be appropriate for a SACH or single-axis foot, and manual lock or stance control knee. A level 2 ambulator can be fit with a multi-axis foot and a polycentric or pneumatic knee. A level 3 ambulator can be fit with an energy-storing carbon foot and a hydraulic- or microprocessor-controlled knee. For level 4 ambulators, the recommendations are the same as for level 3.

Following fit and delivery of any prosthetic device, the patient should be seen by the physician for checkout of the device and recommendation of therapy, as necessary. Appropriate prosthetic training in physical therapy should be based on the patient’s treatment goals and anticipated functional outcome. Regular followup by the physician during any therapy training program is appropriate and necessary. Long-term followup by the physician is necessary to reassess fit and function of the prosthesis and the patient’s response to the therapy program. Monitoring for complications related to amputation and prosthetic fit should also be conducted on a regular basis.

Most patients spend 3–6 months in the temporary prosthesis before plateauing in their functional activities. Once the limb shape has stabilized and functional activities have plateaued, the permanent prosthesis should be ordered. Sometimes additional physical therapy is necessary to achieve higher-level goals with the permanent prosthesis. The expected lifespan of the permanent prosthesis is 5 years; however, annual reassessment for fit and function is also necessary. Modification of the socket fit or repairs of prosthetic components are often necessary during this phase.


Recent decades have seen advances in the management and rehabilitation care of individuals with upper limb amputation. Prostheses for the person with upper limb amputation have changed greatly, with improvements in components, socket fabrication, fitting techniques, suspension system and sources of control, electronics, and power. Higher levels of limb amputation can now be fitted with functional prostheses, which allow more patients to achieve independent lifestyles. This is of particular importance for the multilimb amputee.

For the upper limb amputee, myoelectrically and proportionally controlled terminal devices and elbow joints are now used routinely in some rehabilitation programs. These devices have greatly improved the functional outcomes of patients with upper limb amputation. Progress in the areas of prosthetic fitting techniques and devices (eg, use of osseo-implantation for suspension of the prosthesis) and development of control systems is ongoing, and further developments are expected to take place as technology and the human–machine interface improves.

**EPIDEMIOLOGY**

The exact number of people around the world who have a major amputation is difficult to ascertain as many countries do not keep records of the number of people with limb amputation. Based on information available from the National...
Center for Health Statistics, there are approximately 100,000 new amputations every year in the United States. Extrapolating from these and other sources of health statistics worldwide, the major causes of amputation in order of incidence are trauma (including war-related injuries), diseases (eg, malignancies and arterial insufficiency), and congenital limb deficiencies. The causes of amputation vary from country to country. Because medical comorbidities leading to limb loss most often imperil the lower extremity, more lower limb than upper limb amputations occur, at a ratio of almost 5 to 1. Congenital limb deficiencies account for a small proportion of the total number of reported limb amputations, with a reported incidence of 4.1 per 10,000 live births.

Trauma-related amputations usually occur as a result of motor vehicle, military conflict, industrial, or farming accidents and may account for up to 30% of new major limb amputations. Traumatic amputations occur in a much younger, active, and economically productive population. Sixty percent of arm amputees are between the ages of 21 and 65 years, and 10% are younger than 21 years. Because of the higher risk of work-related accidents in men, there is a higher number of trauma-related amputations for this gender and, overall, a higher incidence of upper limb amputation.

Amputation of the distal segment of the upper limb is more common than proximal amputation and can occur at any age. Men between the second and fourth decades are most frequently affected, with involvement of the right more often than the left limb (related to dominance). The transradial level accounts for 65% and the transhumeral level for 25% of upper limb amputations. Shoulder, elbow, and wrist disarticulation levels together account for the remaining 10%.


CLASSIFICATION & LEVELS OF UPPER LIMB AMPUTATION

Amputations are best classified based on the anatomic level and site at which the amputation has taken place (Table 27–1). Thus, an amputation between the wrist and elbow is termed a transradial amputation. Other common levels of amputation in the upper limb include transhumeral; shoulder, elbow, and wrist disarticulation; and partial hand. Forequarter amputation involves the removal of the complete arm, including the clavicle, scapula, and portions of the chest wall. This type of extensive amputation is performed primarily in cases of malignancy or very severe trauma.

Table 27–1 Classification of common levels of upper limb (UL) amputation.
Nomenclature exists that describes a range of congenital limb deficiencies, from partial to complete omission of a limb or digit (*Table 27–2*). However, in the context of rehabilitation of the upper limb amputee, these deficiencies are best classified following the International Organization of Standards and the International Society of Prosthetics and Orthotics classifications, as modified from Frantz and O’Rahilly. The limb deficiencies can be transverse or longitudinal. The word *terminal* is used to describe the fact that the limb has developed normally to a particular level beyond which no skeletal element exists. In the intercalary limb deficiency, there is a reduction or absence of one or more elements within the long axis of the limb; in this case normal skeletal elements may be present distal to the affected segments. The incidence of congenital upper limb deficiency is approximately 4.1 per 10,000 live births.
Table 27–2 Nomenclature relating to congenital limb deficiency.

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acheiria</td>
<td>Absence of a hand or foot</td>
</tr>
<tr>
<td>Adactyly</td>
<td>Absence of fingers or toes including metacarpal or metatarsal bones</td>
</tr>
<tr>
<td>Amelia</td>
<td>Absence of a limb</td>
</tr>
<tr>
<td>Aphyangia</td>
<td>Absence of a finger or toe</td>
</tr>
<tr>
<td>Hemimelia</td>
<td>Absence of half of a limb</td>
</tr>
<tr>
<td>Meromelia</td>
<td>Partial absence of a limb</td>
</tr>
<tr>
<td>Phocomelia</td>
<td>Absence of the proximal portion of the limb with distal appendage attached to the trunk</td>
</tr>
</tbody>
</table>

Amputation of the distal segment of the upper limb is more common than proximal and can occur at any age, with more frequent involvement of the right limb (related to dominance). Amputations stemming from medical comorbidities result in greater numbers of lower limb than upper limb amputations, at a ratio of almost 5 to 1. Consequently, while most rehabilitation teams are prepared to deal with lower limb amputation, infrequently (in view of the limited exposure) will they have extensive experience in caring for the upper limb. Among upper limb amputations, the transradial level accounts for 65% and the transhumeral level for 25% of amputations. Shoulder, elbow, and wrist disarticulation levels together account for the remaining 10%.


REHABILITATION OF THE UPPER LIMB AMPUTEE

Optimal rehabilitation care of the upper limb amputee begins, if feasible, prior to the amputation. Ideally, this care should be provided by a specialized treatment
team. As previously noted, expertise and experience in caring for upper limb amputations is not as widespread as that relating to the lower limb because of the more limited exposure to patients with such injuries. Communication between the members of the team and with the patient and his or her family is essential and should provide the team with the necessary information to develop a treatment plan covering the phases of care from amputation until return to home.

Table 27–3 outlines treatment goals appropriate to the phases of rehabilitation for a patient undergoing amputation. Specific evaluation items, treatment goals, and objectives are of great help in designing a comprehensive rehabilitation program for the upper limb amputee.

Table 27–3 Phases of amputee rehabilitation
<table>
<thead>
<tr>
<th>Phase</th>
<th>Goal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative</td>
<td>Assessment of body condition; patient education; determination of surgical level of amputation; development of postoperative prosthetic plan</td>
</tr>
<tr>
<td>Amputation surgery and</td>
<td>Optimization of residual limb length, closure technique, soft tissue coverage, and nerve handling; application of rigid dressing</td>
</tr>
<tr>
<td>reconstruction</td>
<td></td>
</tr>
<tr>
<td>Acute postsurgical</td>
<td>Wound healing measures; pain control; confirmation of proximal body–joint motion; emotional support to patient and family</td>
</tr>
<tr>
<td>Preprosthetic</td>
<td>Soft tissue shaping; volume control; increased muscle strength; restoration of patient locus of control</td>
</tr>
<tr>
<td>Prosthetic prescription</td>
<td>Patient-centric focus; team consensus on prosthetic prescription and fabrication</td>
</tr>
<tr>
<td>Prosthetic training</td>
<td>Increased prosthetic functional utilization</td>
</tr>
<tr>
<td>Community integration</td>
<td>Resumption of roles in family and community, with focus on recreational activities, emotional equilibrium, and coping strategies</td>
</tr>
<tr>
<td>Vocational rehabilitation</td>
<td>Assessment and planning of vocational activities for the future. Patient may need further education, training, or job modification</td>
</tr>
<tr>
<td>Followup and preventive care</td>
<td>Lifelong prosthetic, functional, and medical assessment; emotional support; joint protection</td>
</tr>
</tbody>
</table>

**Preoperative Phase**

The team should review with the patient what to expect after surgery and during and after rehabilitation, taking into account physical status, level and cause of amputation, cognition, premorbid lifestyle, and socioeconomic level, and should prepare the patient with realistic short-term and long-term goals and expectations.

The viability of the soft tissues and availability of skin coverage with adequate sensation will usually determine the most distal possible functional level for amputation. Whenever possible, amputation at the transradial level is preferred to achieve optimal prosthetic fit (Figure 27–1). Preserving length of the residual limb to improve prosthetic suspension and force transmission from the residual limb to the socket is a principal responsibility and goal of the surgeon. The residual limb must be surgically constructed with care to optimize the intimacy of prosthetic fit, maintain muscle balance, and allow the residual limb to assume the stresses necessary to meet its new function. Bony prominences, skin scars, soft tissue traction, shear, and perspiration can complicate this function.
Postoperative Phase

After surgery the patient with an upper limb amputation should be able to use a prosthesis, be it body or externally powered, during most of the day through a newly created human–machine interface (the socket–residual limb). After limb amputation, fitting of the first prosthesis should be implemented as early as possible after wound healing (see Table 27–3). Sutures usually are removed after 14–21 days, at which point the skin can start to tolerate some degree of traction. Although infrequently used in the upper limb, the application of an immediate postoperative rigid dressing can help control swelling, expedite wound healing, and promote stump maturation. Elastic bandages can also be used for this purpose (Figure 27–2). This is of particular importance for the unilateral upper limb amputee, because there is a direct relationship between prosthetic fitting time and long-term prosthetic acceptance in this patient population. The window
of opportunity during which there is a significantly greater rate of acceptance and functional integration of a prosthesis for this patient population begins to close at approximately 6 months.

Figure 27–2 Transhumeral residual limb volume control using an elastic bandage.


Pain Management

Pain perceived by the patient with limb amputation can be divided into four possible categories: postsurgical pain, residual limb pain, prosthetic pain (caused primarily by the use of the prosthesis), and phantom pain (pain perceived as coming from the amputated body part). Although each one of these pain categories is described as a separate entity, the different categories may overlap.
It is important to recognize that pain may originate from other structures in the body (cardiogenic, articular, neuropathic, or radiculopathic) and be referred to the amputated limb. Systemic diseases such as diabetes mellitus, ischemia, or arthritis can produce pain and should be identified before attempting treatment of the pain complaints. With a wide variety of pain sources and treatment options available, management of pain in the amputee must begin with accurate diagnosis. Once the nature of the patient’s pain has been clarified, appropriate interventions can proceed to allow the patient to function comfortably. More in-depth discussion of the management of amputation-related pain is beyond the scope of this chapter and readers are referred to other sources for further discussion of the topic.


**UPPER LIMB PROSTHETICS**

Appropriate selection of componentry for prosthetic restoration of patients with upper limb amputation is an extremely important and challenging task in view of the variety and complexity of available prosthetic devices and the functional requirements of these patients. A survey of available components follows.

**Prosthetic Components**

Upper limb prosthetic choices have increased greatly over the past several years, with improvements in components such as hooks and hands, wrists, electronic elbows, and electric shoulder units. Improvements have also been made in socket fabrication materials (carbon graphite or high-temperature flexible thermoplastics), fitting techniques (miniframe sockets), suspension systems (silicone, osseointegration, etc), power sources, and electronic controls (Figure 27–3).
Figure 27–3 Top and side views of a transradial carbon graphite supracondylar suspension frame socket for a myoelectrically controlled prosthesis.

As a result of innovations in system control strategies using targeted reinnervation, multiple joints can now be simultaneously activated instead of the sequential control that has traditionally been used. These advances have greatly benefited patients with more traditional levels of amputation through the incorporation of implanted myoelectric electrodes and the availability of proportional controlled terminal devices that include slip sensors, multijoint finger articulation, and thumb placement for improved opposition and grip. Patients with higher levels of upper limb amputation, such as shoulder disarticulation, can now be fitted with functional prostheses, enabling more patients to achieve independent life styles. This is of particular importance for individuals with bilateral upper limb amputation, especially those with very high levels of amputation. Patients in such cases who were previously fitted with body-powered prostheses usually abandoned the devices because of perceived lack of function or high rate of complaints relating to socket fit or comfort.


Prosthetic Prescription

The prosthetic prescription should be carefully prepared to satisfy the needs and desires of the patient. A team approach to prescription writing should be used whenever possible in close communication with the patient. Appropriate training should be accomplished by a specialized team of professionals following the provision of a prosthetic device and implemented again after new components are prescribed. In general terms the prescription should include a terminal device, wrist, socket, suspension system, and, if appropriate, an elbow mechanism. Decisions relating to power options (body power, external power, or passive components) and their activation mode (myoelectric, switch, or cable) should be predetermined and frequently require assistance from an experienced rehabilitation team.

The selection of prosthetic system and components is determined by a number of factors, with residual limb length, condition of the soft tissues, joints, and muscle control as the main physiologic determinants and functional requirements. The patient’s vocational and avocational goals, cosmetic desires, and funding also factor into the decision-making process.

Body-powered devices tend to be less complex and more tolerant of physical demands, with lower maintenance requirements and overall cost (Figure 27–4). Body-powered devices require harnessing for activation and frequently for suspension. Externally powered devices are heavier, more expensive, and more complex; require less suspension; and have a more powerful grip with the potential for multijoint simultaneous use (eg, in a transhumeral prosthesis, the elbow and hand can be activated at the same time).
The harness functions to provide power to the terminal device by transferring motion from the shoulder and back (bicipital abduction or humeral flexion, or both) to open the hook, which is closed by rubber bands, or in a transhumeral prosthesis, to flex the elbow. Locking and unlocking of the elbow for a transhumeral prosthesis requires shoulder depression and extension motions. The harness can provide prosthetic suspension as well if no other means are in place.
Terminal Devices

The human hand is a complex anatomic and physiologic structure that cannot be fully replicated with current prosthetic technology. The functional activities of the hand are extensive but can be grouped into two major categories: nonprehensile (touching, feeling, tapping, typing, etc) and prehensile (three-jaw, lateral or key grip, power grip, hook grip, and spherical grip). Various prosthetic terminal devices are available, including passive, body-powered, and externally powered hooks and hands (Figure 27–5). Manipulators are used in less technologically developed environments. All lack sensory feedback and have limited mobility and dexterity even if they have multiarticulated fingers. Prosthetic hands provide a three-jaw chuck pinch and hooks provide the equivalent of lateral or tip pinch. Electric hands with multiarticulated fingers tend to be slower and less powerful in their grip.
Electric devices can have digital (on–off) or proportional (stronger signal equals faster action) control systems. In older terminal devices, gravity and weight of an object held could generate slippage from the gripping surface. Slip control is a recently introduced technology that can increase the security of prehension to prevent accidental dropping of objects.


**Prosthetic Elbows**

The prosthetic elbows available in the treatment of the transhumeral amputation can be passive, body powered, or externally powered. The mechanical elbows have a locking mechanism that is manually applied using the contralateral hand, or a remote lock activated by the chin or the ipsilateral shoulder via a cable system. Electric elbows have an electromechanical brake (Utah III, Boston Arm, or Otto Bock Dynamic Arm) or a switch-controlled locking mechanism to maintain the selected position.

**Sockets**

In the past sockets were carved out of wood but with the development of high-temperature rigid plastic materials such as polyester resin, sockets can be molded to have total contact with decreased weight and increased durability. Modern sockets are custom made by obtaining a negative impression of the residual limb (commonly achieved using a plaster of Paris wrap or digital scanning). More recently the availability of acrylic lamination, carbon graphite, and flexible thermoplastics have permitted the design of sockets with windows, lined with
flexible materials that are even more adaptable, comfortable, lighter, and more durable than in the past. Alternative suspension systems such as the constant suction socket, the silicon sleeve, and others are all useful in the appropriate clinical case.

Of more recent introduction, and considered experimental in many areas of the world, is the use of osseointegration as means of suspending a prosthesis directly from the bone. This type of suspension requires a titanium implant that is externalized and used to attach the prosthetic device directly without the need of a socket. Osseointegration allows shorter residual limbs to be interfaced with a prosthesis without the need of a socket, something that is impossible to achieve without this type of suspension.


Prosthetic Training & Community Reintegration

After prescription and fitting of the prosthetic device, training is indispensable and should include prosthetic management and functional integration with the goal of achieving community reintegration. Community reintegration should include recreational activities and sports, and, when appropriate, return to work or school as part of the rehabilitation program. Long-term follow-up and interventions for joint conservation and potential overuse syndrome are frequently ignored. A well-integrated and experienced team can better achieve the goal of returning patients to and maintaining them at the highest functional level. These are essential characteristics of a successful rehabilitation program for the person with an upper limb amputation.

Kohler F, Stucki G, Geertzen J, et al: Developing core sets for persons following amputation based on the International Classification of
Cosmetic Covers

The terminal device can be covered with a cosmetic glove that resembles a hand. Cosmetic covers range from off-the-shelf to highly detailed custom-made items designed as a mirror image of the opposite hand and covered with hair and human-like nails. In addition to improving cosmesis by providing a more realistic appearance, such covers can provide some degree of water protection. The cost of the custom covers can at times exceed the cost of the prosthetic components, and their life expectancy is limited due to wear and tear of the silicon materials and changes in coloration of the anatomic limb with exposure to sunlight that cannot be matched by the cosmetic device.

PROGNOSIS

Rehabilitation of the upper limb amputee is not simply concerned with the provision of a prosthetic device. Rather, it constitutes a process of restorative interventions necessary to return the individual to the highest possible level of function and to minimize the impact of limb loss on his or her life. In the past two decades, with the advent of comprehensive amputation rehabilitation, specialized treatment teams, and new suspension methods, prosthetic devices and materials, the outlook for the person with an upper limb amputation has substantially improved.
An orthosis, a mechanical device that is fitted and applied to the body, is used to modify structural or functional characteristics of the neuromusculoskeletal system. To that end, orthotic treatment is often employed to relieve pain, manage deformities, and attenuate abnormal neuromuscular function. More specifically, orthotic treatment may be used to (1) prevent, reduce, or stabilize a deformity; (2) modify the range of motion of a joint; (3) add to the length or alter the shape of a segment; (4) compensate for weak muscle activity or control muscle hyperactivity; and (5) reduce or redistribute the load on tissues. Orthoses may be commonly referred to as braces or splints.

Rehabilitation physicians often work with orthotists. In conjunction with the physician, the orthotist designs and fabricates an orthosis to meet the specific needs of the patient. Proper fit and alignment of an orthosis is paramount in order for a patient to benefit from its use. An orthosis that is incorrectly fitted or uncomfortable does not confer the desired effect and may be cosmetically unacceptable to the patient. This orthosis is less likely to be worn and represents a missed opportunity to help a patient. In such cases the ill-fitting orthosis may be either modified or replaced.

It is the joint responsibility of the physician and the orthotist to ensure that an orthosis is fitted properly and that the patient is instructed in its proper use. In circumstances where a custom fit is not essential, a variety of prefabricated orthoses may be used. However, prefabricated devices are often difficult, if not impossible, to modify due to the lack of plasticity of the materials and their inherent fragility once tampered with. It is therefore still absolutely essential for the physician to ensure that the orthosis is appropriate and fits correctly.
UPPER EXTREMITY ORTHOTICS

General Considerations

The function of the upper extremity, in maneuvering the hand so that it may access and manipulate objects in space, is achieved through prehension, which is the act of taking hold, seizing, or grasping an object. The shoulder, elbow, and wrist function together to place the hand in a desired locale for fine and gross motor tasks. The main goal of upper extremity orthoses, therefore, is to maintain the function of the hand, either by restoring or preserving prehension itself, or by allowing optimal positioning of the hand in space in order to manipulate objects in the environment. In this regard five common goals in upper extremity orthotics are (1) substituting for weak or absent muscles, (2) protecting damaged or diseased segments by limiting loads or motion, (3) preventing deformity, (4) correcting contracture, and (5) attaching to other assistive devices.

Management

A. Finger or Hand Orthoses

1. Swan-neck deformity—The swan-neck deformity is often seen in patients with rheumatoid arthritis and is characterized by hyperextension of the proximal interphalangeal (PIP) joint and flexion of the distal interphalangeal (DIP) joint. Finger–thumb orthoses such as the silver ring splint, the oval-eight splints, and thermoplastic splints can be used to correct the hyperextension of the PIP joint and prevent further progression of the deformity (Figure 28–1).
2. **Boutonnière deformity**—Like swan-neck deformity, the boutonnière deformity is also commonly seen in rheumatoid arthritis patients. But this deformity, conversely, produces flexion of the PIP joint and hyperextension of the DIP joint. The orthoses used are the same: finger–thumb orthoses such as the silver ring splint, the oval-eight splint, and thermoplastic splints. These correct the flexion of the PIP joint and prevent further progression of the deformity (Figure 28–2).
3. **Mallet finger**—Mallet finger results from a tear or avulsion of the extensor tendon as it cross the DIP joint. Examination reveals a finger that lags in extension at the DIP. Splinting of the DIP joint in extension is aimed at allowing for healing of the torn or avulsed tendon in its shortened state.

4. **Trigger finger**—Trigger finger results from inflammation of the flexor tendon as it courses through the first annular (A1) pulley. This produces a DIP joint that is stuck in flexion. A palpable nodule may be present at the point of inflammation. Splinting of the DIP joint in extension or the metacarpophalangeal (MCP) joint in slight flexion allows the inflammation to resolve and is part of the management of this ailment.

5. **Flexor tendon injury**—Patients with flexor tendon injury often have difficulty achieving full flexion at the PIP joint. Finger knuckle bender splints may be used to increase flexion in the affected joint. These splints have a point of pressure at the volar aspect of the PIP joint and points of counterpressure at the dorsal aspect of the proximal and middle phalanx, and utilize rubber bands to provide force to flex the joint. The goal of this orthosis is to increase range of motion with regard to flexion at the PIP joint.

6. **Median and ulnar nerve injury**—In median or ulnar nerve injuries the MCP joint has a tendency to become hyperextended due to lack of muscle strength in the hand intrinsics. Opposition often is lost in solitary median nerve injuries. Hand–finger orthoses can be used to control the MCP joint of the fingers and thumb. A C-bar is used to maintain the web space. An opponens bar is used so that the thumb opposes the other fingers, placing it in a position to maintain pinch grip. The goal of these orthoses is to prevent contracture formation and preserve hand function to perform gross motor tasks. In the event that the MCP joint becomes contracted, the hand–finger knuckle bender splint (a dynamic hand–finger orthosis) can be used to stretch the MCP joint. Like the finger knuckle bender, it uses either rubber bands or wires to provide flexion force.

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van der Giesen FJ, van Lankveld WJ, Kremers-Selten C, et al: Effectiveness of two finger splints for swan neck deformity in patients with rheumatoid...
B. Wrist or Thumb Orthoses

1. Radial nerve injury—Patients with radial nerve injury may lack wrist and finger extension. Lack of wrist extension puts the finger flexors on slack and compromises grasp and full finger flexion. Dorsal wrist extension dynamic finger splints and static wrist volar cock-up splints are used in radial nerve injury to extend the wrist and allow the fingers to flex more effectively for grasping activities.

2. Carpal tunnel syndrome—Carpal tunnel syndrome results from compression or inflammation of the median nerve as it courses under the transverse carpal ligament in the wrist. Wrist–hand–finger orthoses are used to control motion through the wrist and proximal hand, with the goal of providing symptom relief or conferring a mechanical advantage in order to improve prehension. In carpal tunnel syndrome, the wrist is placed into a few degrees of extension while allowing for freedom of thumb and finger movement (Figure 28–3). Immobilization of the wrist in carpal tunnel syndrome is thought to allow inflammation to resolve.

▲ Figure 28–3 Wrist orthosis for carpal tunnel syndrome.

3. Gamekeeper’s thumb—Gamekeeper’s thumb results from a tear of the ulnar
collateral ligament of the thumb. Patients present with pain and swelling on the posterior medial aspect of the thumb and instability, with a tendency for the thumb to go into hyperabduction. A thumb spica splint is used to protect and immobilize the injured segment and allow it to heal (Figure 28–4).

▲ Figure 28–4 Thumb spica splint for ulnar collateral ligament injury and De Quervain’s tenosynovitis.

4. De Quervain’s tenosynovitis—De Quervain’s tenosynovitis results in pain at the base of the thumb or wrist. It is caused by inflammation of the extensor pollicis brevis and abductor pollicis longus tendon sheaths. Patients often have pain at the base of the thumb with thumb extension or ulnar deviation of the wrist. A thumb spica splint is used to immobilize the inflamed segment and allow it to heal.

5. Unspecified thumb injury—A thumb spica splint may initially be used to manage fractures or ligamentous and neurovascular injuries of the thumb until the patient can be further evaluated by a hand specialist. In these cases the objective of the orthosis is to protect and immobilize the thumb in order to limit pain and allow use of the other digits. Dorsal wrist extension dynamic finger splints and static wrist volar cock-up splints are used in radial nerve injury to extend the wrist and allow the fingers to flex more effectively for grasping
activities.

6. Carpometacarpal arthritis—Arthritis of the first carpometacarpal joint may be treated with a long and short opponens splint. Resting wrist hand splints have been shown to give relief to symptomatic patients suffering from rheumatoid arthritis.

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C. Elbow Orthoses

1. Quadriplegia—Patients with C6 tetraplegia often have preserved elbow flexion and wrist extension but do not have use of their hand and finger muscles. The tenodesis prehension orthosis takes advantage of the finger flexion that normally occurs passively with wrist extension to allow these patients to grasp objects.

2. Elbow flexion contracture—Static progressive and dynamic elbow splints are often used to stretch flexion contracture of the elbow and improve range of motion after trauma.

3. Elbow flexor weakness—Dynamic spring-assisted orthoses are available for patients with elbow flexor weakness.

4. Cubital tunnel syndrome—Cubital tunnel syndrome results from compression of the ulnar nerve at the elbow. Patients often have sensory complaints and motor deficits in an ulnar distribution in the hand. Nocturnal splinting of the elbow at 30–35 degrees of flexion, the forearm in 10–20 degrees of pronation, and the wrist in neutral position has been shown to be effective in treating this condition. The objective of splinting is to reduce traction on the ulnar nerve and provide symptomatic relief.

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D. Upper Arm and Shoulder Orthoses

1. Humeral shaft fractures—The Sarmiento brace is a prefabricated nonarticular orthosis used for the conservative treatment of humeral shaft fractures. It remains the treatment of choice for nondisplaced or minimally displaced fractures, even in an era of newer intramedullary procedures and minimally invasive techniques. This orthosis is applied to the humerus and fastened with Velcro straps to maintain approximation of the distal and proximal aspects of the fracture. Bracing does not restore anatomic alignment to the fracture segment, but the end results are usually cosmetically and functionally acceptable. A benefit of this bracing method is that it allows for full range of motion at the elbow and shoulder and thus is effective in avoiding complications such as elbow flexion contracture and adhesive capsulitis.

2. Clavicular fractures—Clavicular fractures account for 2.5–5% of all fractures, and various classification schemes have been developed to describe them. Classically, they were divided into three types: type I, involving the middle third of the clavicle; type II, involving the distal clavicle; and type III, involving the proximal clavicle. Types I and III may be treated conservatively with a figure-of-eight splint or shoulder sling. Type II may be treated with surgical fixation as these fractures have a high rate of nonunion. The objective of splinting for clavicular fractures is to maintain glenohumeral integrity and also to limit motion across the acromioclavicular (AC) and glenohumeral joints.

3. Acromioclavicular (AC) joint separation—AC joint separation is a common injury in younger, physically active patients. These joint separations may be classified into six types. In type I, the AC ligaments are sprained; in type II, they are torn but the coracoclavicular (CC) ligaments are intact; in type III, the AC and CC ligaments are both torn; in type IV, the clavicle is displaced posteriorly; in type V, the AC and CC ligaments are torn and the deltrotrapezial fascia are also torn, causing the scapula to droop inferiorly; and in type VI, AC injury occurs when the clavicle dislocates inferiorly to the coracoid. Types I and II may be treated conservatively with a sling. Types IV, V, and VI are treated operatively as there is high risk of morbidity due to dislocation and soft tissue damage. The treatment of type III AC joint separations is not as clear. These injuries may be treated conservatively with a shoulder sling initially; however, if there is persistent pain or disability an operative course may be pursued. The objective of splinting in the patient with an AC joint separation is to maintain
glenohumeral integrity and also to limit motion across the AC and glenohumeral joints.

4. **Shoulder dislocations**—Glenohumeral dislocations may be treated with an adduction arm splint. Traditionally, the shoulder is also placed in internal rotation, but studies have shown that placement in neutral or even external rotation may be more beneficial. Several shoulder orthoses have been devised for stroke patients with post-stroke shoulder pain and issues with glenohumeral subluxation. These consist of various slings and arm trays that allow the shoulder to rest in a position of comfort and support the shoulder joint by compensating for muscles affected by the stroke. Patients may also neglect or have impaired sensation in the affected arm after a stroke, and these devices protect the upper limb from injury, keeping it in view and close to the body. The airplane splint and gunslinger orthosis can be used to immobilize the shoulder and keep the arm fixed in relation to the torso. These may be employed postoperatively after soft tissue repairs, fractures, and brachial plexus injuries. The balanced forearm orthosis is a shoulder–elbow–wrist–hand orthosis that supports the upper limb in a gravity-eliminated position so that patients with weakness in their shoulder flexors and elbow flexors (eg, high-level tetraplegia) can gain access to the face and mouth for feeding.

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LOWER EXTREMITY ORTHOTICS

General Considerations

Orthoses in the lower extremity, as in other parts of the body, are used to modify the structural and functional characteristics of a particular body segment. They are central components of treatment for a variety of conditions and, when employed properly, often have a dramatic impact on the gait pattern of the patient. Because safe, functional, pain-free, and energy-efficient ambulation is often the goal of lower limb bracing, a general conceptual knowledge of the gait cycle, biomechanical forces during gait, and functional anatomy of the lower limbs is crucial. Discussion of these topics appears elsewhere in this book (see Chapters 4 and 11).

Management

A. Foot Orthoses

1. Pes planus—Pes planus (“flat feet”) is a common cause of foot pain. Several footwear modifications may be made to help alleviate the pain experienced by patients with this condition. The first of these is medial counter-reinforcement, which involves building up the medial compartment of the shoe to prevent medial collapse of the longitudinal arch of the foot. A second option (the “split and widen” method) involves widening the midsole of the shoe to allow more space to accommodate the collapsed medial prominence. Finally, a rocker-sole shoe may be used. This type of shoe reduces bending stresses in the sagittal plane and can reduce forefoot pressures by loading the midfoot during midstance, further serving to support the longitudinal arch and reducing pain.

When choosing a foot orthosis to treat pes planus, the goal is to provide support to the longitudinal arch of the foot by placing the orthosis in the medial
compartment. This may be accomplished by a medially posted foot orthosis. Another option is to use the University of California Berkeley Laboratory (UCBL) orthosis, which is designed to control the tri-planar motion of the hindfoot and midfoot and provide support to the longitudinal arch. Like a medially posted foot orthosis, the UCBL orthosis has a medial buildup that supports the longitudinal arch. The posterior aspect of the orthosis is constructed in a way that prevents calcaneal eversion and further flattening of the longitudinal arch.

2. **Pes cavus**—Pes cavus is so named for the high arches observed in this condition. Owing to the decreased weight-bearing surface area of the cavus foot, force is preferentially distributed over the metatarsals and lateral cuneiforms, leading to pain, contusions, and osteophyte formation. Treatment of this condition is aimed at arch support, pressure redistribution, structural alignment, and shock absorption. This is accomplished by building up the medial compartment so that weight is distributed more evenly over the medial and lateral compartments.

3. **Leg-length discrepancy**—There are many causes of leg-length discrepancies; among the most commonly encountered are total hip arthroplasty, total knee arthroplasty, neuromuscular disorders, and trauma. Conservative treatment is accomplished through the use of an internal shoe lift (as with a shoe insert) or by building up the sole of the shoe. Discrepancies of approximately 20–60 mm can be corrected with a lift; however, it is recommended that the shorter leg be corrected to only 10 mm of the longer leg. The goal of the shoe lift is not to correct the discrepancy completely but rather to improve the overall gait pattern so that ambulation is more comfortable and energy efficient.

4. **Metatarsalgia**—Metatarsalgia is a common foot problem. Patients experience pain in the plantar foot from the second through fourth metatarsal heads. The goal of the orthosis in metatarsalgia is pain reduction. This is achieved by shifting the weight-bearing force and plantar pressures off of the metatarsal heads. Common methods employed are the use of an excavated insole; a metatarsal bar (built into either the insole or the outsole), which adds padding proximal to the metatarsal heads; and a rocker-bottom sole.

5. **Plantar fasciitis**—Plantar fasciitis is a common cause of heel pain in adults. It is thought to be an overuse syndrome resulting from prolonged running or standing. Affected patients develop a calcaneal enthesopathy that can make it difficult to walk or stand for more than short periods of time. The goal of the
orthosis in plantar fasciitis is to reduce pain and allow the inflammation in the plantar fascia to resolve. Part of the treatment regimen includes using heel cups, longitudinal arch supports, and custom-made full-length insoles. Heel cups help absorb shock and cushion against tenderness at the heel. Longitudinal arch supports and insoles act to support and reduce stretch on the plantar fascia, allowing the inflammation to resolve. Studies have shown that the use of these orthotic devices can reduce foot pain and improve function.

6. Achilles tendonitis—When treating Achilles tendonitis, it is important to relieve tension on the Achilles tendon by plantar flexing the foot and reducing any excessive internal or external rotation of the hindfoot. A deep heel cup that controls valgus or varus posting helps to maintain the subtalar joint in a neutral position, which is essential for treatment. Relief of tension to the Achilles tendon can be achieved by heel pads placed in the shoes or by using shoes with elevated heel height (eg, running shoes or clogs). These measures allow the inflammation in the area of the tendon to resolve and protect the tendon from undue strain. Patients should be counseled to avoid walking barefoot, wearing flat-soled shoes, or walking on soft surfaces as doing so allows the heel to sink below the forefoot, placing undue tension on the tendon. The goal of the orthosis is to reduce inflammation and strain on the Achilles tendon and reduce pain associated with ambulation.

Goff JD, Crawford R: Diagnosis and treatment of plantar fasciitis. Am Fam Physician 2011;84:676–682.

B. Knee Orthoses

1. Medial compartment osteoarthritis of the knee—Patients with this condition may be treated with a lateral heel wedge. This type of foot orthosis has been shown to alleviate knee pain by shifting the tibiofemoral angle, causing more weight to be born on the lateral aspect of the knee joint. This achieves the goal of relieving forces on the medial joint—the source of pain in these patients.

2. Knee instability—A key concept to remember with regard to lower limb
orthoses is that positional changes distally can confer biomechanical changes in the proximal joints. If, for example, the ground reaction force is anterior to the knee, then a hyperextension moment will be created at the knee. If the ground reaction force is posterior to the knee, then a flexion moment will be created at the knee. (Chapter 4 discusses biomechanical concepts in depth.) Changes in the relative positioning of the ground reaction force can be accomplished by altering the amount of dorsiflexion or plantar flexion allowed at the ankle through the use of dorsiflexion and plantar flexion stops.

These stops are created differently, depending on the type of orthosis. In a dual-channel, metal, double upright ankle–foot orthosis (AFO), posterior pins are used to produce a plantar flexion stop, while anterior pins are used to produce a dorsiflexion stop. In an articulated plastic AFO, a control strap attached to the foot and the calf components posteriorly acts as a dorsiflexion stop, while a posterior buildup or an adjustable pin between the posterior aspect of the articulating surfaces of the orthosis serves as a plantar flexion stop (Figure 28–5).

▲ Figure 28–5 Articulated plastic ankle–foot orthosis (AFO) with a posterior control strap to limit dorsiflexion.

Control at the knee is achieved by altering the ground reaction force relative
to the knee. If stability is lacking and the knee is buckling with each step, the AFO should be adjusted to place the ankle in greater plantar flexion to reduce the external flexion moment at the knee or even create a slight knee extension moment, thereby conferring more stability. If, however, the knee is being forced into hyperextension (genu recurvatum), the AFO may be adjusted to place the ankle into greater dorsiflexion to reduce the amount of hyperextension. The goal of using an AFO in individuals with knee instability is to manage the amount of knee flexion or hyperextension that occurs to prevent knee buckling or genu recurvatum, respectively.


C. Ankle Orthoses

1. Mediolateral ankle instability and deformity—Custom-fitted molded AFOs can provide mediolateral ankle stability during stance (Figure 28–6). This allows for control of ankle eversion and inversion and may be used to treat and accommodate for equinovarus and equinovalgus deformity. Equinovarus and equinovalgus are caused by muscle imbalances that can occur in several neurologic and musculoskeletal conditions. Bracing may be used either alone or in conjunction with surgery or chemodenervation to restore balance to the muscles and correct the deformity. The goal of bracing in equinovarus and equinovalgus is to prevent further deformity of the joint and allow for it to be placed in an anatomically functional position.
2. **Heel decubiti and plantar flexion contracture**—A special type of AFO called a pressure relief ankle–foot orthosis (PRAFO) can be used to both offload the heel when ulceration or other forms of injury are present and prevent plantar flexion contracture. The goal in a patient with heel decubiti is to offload the heel and allow sufficient time for healing to occur. The goal in a plantar flexion contracture is to stretch the contracture, prevent worsening of a preexisting contracture, or maintain and increase ankle range of motion postoperatively.

Malas B: What variables influence the ability of an AFO to improve function and when are they indicated. Clin Orthop Res 2011;469:1308–1314.

**D. Orthoses for Complex Pathology of the Knee, Ankle, and Foot**
1. **Charcot foot**—Charcot foot is a deformity seen in patients with neuropathic arthropathy. Its etiology remains unclear. Current thinking suggests that impaired neurogenic joint control leads to repetitive joint trauma, ultimately leading to inflammation and subsequent destruction of bone, cartilage, and ligaments. The patient is left with a deformed and poorly functioning foot.

   The treatment of Charcot foot includes use of an orthosis to unload the foot, either partially or completely, and to limit joint motion in order to prevent the aggregate trauma that initiated the inflammation and joint destruction. Orthoses may be used initially to quell the inflammatory cascade but do little to reverse deformity, which is usually accomplished with surgery. Postoperatively, orthoses are employed to provide some degree of unloading as well as limitation of motion at the foot and ankle. In cases where more unloading is required, an AFO is fabricated with an anterior component that allows for weight bearing through the proximal tibia. A rocker-bottom sole may be implemented to further limit motion at the midfoot. The goal of orthotic management of Charcot foot is to reduce pressure and motion through the ankle joint, and to increase the contact surface area of the foot.

2. **Foot drop**—Foot drop is a manifestation of many neurologic and musculoskeletal conditions, including post-stroke hemiplegia, cerebral palsy, myopathy, spinal cord injury, radiculopathy, lumbosacral plexopathy, and direct injury to the peripheral nerves, muscles, or tendons. AFOs may be used temporarily to assist with gait while the cause of foot drop is treated, or indefinitely in patients whose underlying condition is not reversible. In the patient with foot drop, the purpose of the orthosis is to allow for clearance of the foot during swing phase and prevent foot slap during the loading phase. This may be accomplished through a plantar flexion stop, using a pin or posterior strap, and dorsiflexion assist, using a spring. However, the effect of the AFO on the knee must be considered. Specifically, limiting plantar flexion and assisting dorsiflexion may lead to knee instability and cause buckling. A delicate balance must be achieved; once the brace is fabricated, the patient’s gait must be monitored for buckling or hyperextension, after which adjustments may be made as necessary. The goal of the orthosis in foot drop is twofold: to assist with dorsiflexion to prevent catching of the foot during the swing phase of gait and, simultaneously, to maintain stability of the knee to prevent buckling or recurvatum.

3. **Neuromuscular disease or spinal cord injury**—Knee–ankle–foot orthoses (KAFOs) are often used in patients with spinal cord injury, neurologic disorders,
or muscular disorders such Duchenne muscular dystrophy to facilitate ambulation and activities of daily living (Figure 28–7). In these disorders, weakness of the quadriceps muscles (used in knee extension) causes the knee to collapse. Patients using KAFOs are instructed to adopt a C-shaped posture in which they hyperextend the lumbar spine and hips to maintain balance. Various knee designs allow the orthosis to lock at the knee during specific actions, thus providing stability during gait. Ambulation may be achieved with or without the use of an assistive device, such as forearm crutches. Battery-powered models, with knee extension assist, have shown promise and may offer further advantages, so long as their size, bulk, and cost do not preclude utility. The goal of the orthosis is to provide knee stability and allow ambulation without excessive energy expenditure.

Figure 28–7 A knee–ankle–foot orthosis (KAFO) can stabilize the knee and the ankle.

4. Knee injury—The Swedish knee cage is a bracing device that uses a three-point pressure system to prevent genu recurvatum by applying a posterior-directed force above and below the knee and an anterior-directed force at the knee. It may be used separately or in conjunction with an AFO to improve knee control during gait.
The use of knee bracing in sports is controversial. Recently, the use of prophylactic knee bracing to protect the knee in high-risk sports has come under scrutiny. Multiple variables must be taken into account when considering using an orthosis in an athlete. Player position and the impact the orthosis may have on performance are important factors. Evidence appears to support the use of prophylactic bracing to prevent medial collateral injury from a lateral blow in contact athletes. Functional bracing, which is often practiced after anterior cruciate ligament (ACL) injury, has also been criticized. The usefulness of this bracing may derive from increased proprioceptive feedback and increased patient comfort and confidence in the knee. However, trials have failed to demonstrate the ability of bracing to affect pain, range of motion, graft stability, or protection from subsequent injury. Nonetheless, the goal of bracing in knee injury is to protect the knee during activity, provide proprioceptive feedback on knee position, and instill confidence in the user.

5. Spinal cord injury with hip weakness—Reciprocating gait orthoses, a type of hip–knee–ankle–foot orthosis (HKAFO), are used in patients with thoracic-level spinal cord injury and require bracing that includes the hip and extends to the ankle. The patient’s knee and ankle joints are fixed and the hips are allowed to move through available ranges of flexion and extension. Ambulation is achieved with the use of an assistive device, such as crutches. With the help of the crutches, the patient shifts his or her body weight over a predetermined stance leg. As the center of mass passes over the stance leg, which is in extension, the design of the reciprocating gait orthosis brings the contralateral hip of the swing limb into flexion, thus allowing for limb advancement. This cycle is then repeated, enabling the patient to ambulate.

Although the reciprocating gait orthosis may confer a sense of independence and some of the benefits of weight bearing, an extremely high metabolic energy cost is associated with walking in this manner. Studies have reported the oxygen cost of walking with a reciprocating gait orthosis to be 1.0 mL/kg per meter at user-selected walking speeds ranging from 0.2 to 0.3 m/s compared with 0.176 mL/kg per meter at 1.28 m/s for nondisabled persons. Consequently, this orthosis is primarily used for household ambulation and activities of daily living that require the patient to navigate in tight spaces. The goal of this orthosis is to provide the psychological and physical benefits of ambulation, which include increased independence, maintenance of bone mass, and prevention of venous thromboembolism; however, it is not an ideal device for prolonged ambulation owing to its energy-intensive nature.


**SPINAL ORTHOSES**

### General Considerations

As in other parts of the body, orthotic devices may be employed throughout the spine to improve structural and functional deficits. Spinal orthoses have five primary functions; they serve as a kinesthetic reminder and offer total contact, three-point pressure, end-point control, or elevated pressure with the goal of supporting or immobilizing a specific region of the spine that would otherwise have motion. Prescription of the proper spinal orthosis requires knowledge of spinal anatomy and the biomechanics of the spine as a whole and of its individual segments. This allows the physician to choose the most appropriate orthosis to protect an injured or weakened area, and correct or prevent further deformity.

### Spinal Anatomy
The vertebral column consists of 33 vertebrae. Starting cephalad and moving caudally, there are 7 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 5 coccygeal vertebrae. The spinal column serves several functions, among them, bearing body weight, protecting the spinal cord from injury, and allowing movement between one part of the body and another. Approximately one third of the spinal column height can be attributed to the intervertebral discs that lie between the vertebral bodies. These discs are made of a gelatinous material and are important shock absorbers in the spine. In the thoracic spine, the vertebral bodies are heart shaped and the zygapophyseal joints are composed of superior articular processes that face posteriorly and laterally and inferior articular processes that face medially and anteriorly. In the lumbar spine, the vertebral bodies have a kidney bean shape and the zygapophyseal joints have superior articular processes that face medially and slightly posteriorly and inferior articular processes that face laterally and slightly anteriorly. The sacral and coccygeal vertebrae are fused segments and have no intervertebral discs. Movement within the different regions of the spine is closely related to the orientation of the superior and inferior articular processes within the different spinal regions.

Management

A. Orthoses for Problems of the Cervical Spine

1. Unstable fractures of the cervical spine—Head cervicothoracic orthoses are used to confer stability in a patient with an unstable cervical spine. An unstable spine is one in which movement between vertebrae or movement of bony fragments could result in neurologic compromise as a result of damage to the spinal contents (ie, spinal cord and nerve roots), which are ordinarily protected. As the highest degree of stability is conferred by completely limiting motion in all planes, orthoses for unstable cervical fractures often do not allow for any cervical motion.

   The halo orthosis consists of a halo ring, uprights, body vest, and pins. When the orthosis is donned, the halo is surgically fixed to the skull with pins, which are screwed into the cranium. This type of orthosis maximally limits the amount of movement in all planes of motion and is the most restrictive of all cervical braces. Complications associated with its use include pneumonia, pin site infections, and dysphagia.

   The Minerva brace is a cervicothoracic orthosis that consists of a body jacket, applied to the patient’s torso; a chin piece, to which it connects anteriorly;
and a posterior support for the occiput. Evidence suggests that this orthosis may afford a similar level of support to that achieved using a halo, with a much lower complication rate. The Minerva brace does not require surgical fixation to the skull and is lighter in weight than the halo. It is effective in providing significant support in flexion, extension, side bending, and rotation of the cervical spine.

The sternal–occipital–mandibular immobilizer (SOMI) brace is an effective orthosis for patients with atlantoaxial instability, in particular, those with rheumatoid arthritis. Like most cervicothoracic orthoses, it provides more control of flexion than extension, with relatively less control of side bending than devices such as the Miami J and Philadelphia collars (described below). However, the SOMI controls flexion at C1–C3 better than any other cervicothoracic orthosis. This makes it suitable for rheumatoid arthritis patients with atlantoaxial instability, for whom instability is far greater with flexion due to ligamentous insufficiency, while the intact dense bone provides intrinsic resistance to extension. For the same reason, the SOMI may also be used in patients with neural arch fractures of C2, as flexion prevents opposition of healing bony segments and must be restricted.

The SOMI may be donned while the patient is supine, facilitating application by those involved in patient care. The brace consists of mandibular and occipital supports that connect to a body vest at a fixed angle. The mandibular piece may cause intersegmental movement of the cervical spine while eating, and may be removed for this activity. The SOMI provides more stability than the Miami J and Philadelphia collars in the sagittal plane but is less restrictive than a halo apparatus.

**2. Stable fractures of the cervical spine**—Fractures that do not have an immediate risk of compromising the spinal components or have been stabilized surgically still require restriction of cervical motion to promote bony and ligamentous healing and avoid worsening the injury. Various cervical orthoses may be used for this purpose.

The Philadelphia and Miami J collars are two commonly used cervical orthoses. These orthoses provide control of flexion, extension, side bending, and rotation in the cervical spine. Compared with the Philadelphia collar, the Miami J has a more prominent sternal component, which may provide more effective support and may be better tolerated by patients. These braces are also used in the treatment of cervical sprains and strains.

Another option is a two- or four-poster brace. These braces are rigid orthoses consisting of anterior and posterior chest pads connected to molded occipital and mandibular support pieces by adjustable struts (one anterior and one posterior in
the case of a two-poster brace; two anterior and two posterior in a four-poster brace). This type of brace controls flexion and extension, and the spine can be positioned in either flexion or extension by adjusting the length of the posts. However, lateral bending and axial rotation are relatively less well controlled, and the mandibular plate can interfere with eating. This brace uses shoulder straps for suspension, and the open design allows for heat loss from the neck. The brace is as effective as a cervicothoracic brace and is better than the Philadelphia collar at controlling flexion in the midcervical area, but it is also bulkier and potentially less well tolerated by the user because of its size.

3. **Neck pain without bony or ligamentous instability**—Soft cervical collars are used for minor injuries to the cervical spine. They are largely ineffective at limiting cervical range of motion but may have value as a proprioceptive reminder to the patient in the avoidance of provocative maneuvers. The indications for this collar are whiplash injuries and neck pain without unstable bony or ligamentous injury.

4. **Cervicothoracic junction injuries**—Injuries at this level require not only cervical stabilization at the lower cervical levels, but also significant stability distal to the cervicothoracic junction. The Yale orthosis is a modified version of the Philadelphia collar. It has fiberglass thoracic extensions that extend anteriorly and posteriorly, with midthoracic straps on the sides connecting them. The thoracic extensions are useful in providing support for injuries at the cervicothoracic junction. The occipital piece extends higher posteriorly and adds stability to the brace. It limits flexion most effectively in the middle to lower cervical spinal levels and provides adequate extension control but is largely ineffective at controlling motion in the upper cervical spine, particularly at the atlantoaxial joint.


B. Orthoses for Problems of the Thoracolumbar Spine and Pelvis

1. Stable compression fracture—Osteoporosis is defined as a bone mineral density greater than or equal to 2.5 standard deviations below the mean for normal, healthy, gender-matched 25 year olds. A major complication in patients with osteoporosis is compression fracture of the spine, which occurs when there is loss of vertebral body height. Often vertebral height loss occurs preferentially in the anterior portion of the vertebral body (wedge fracture) owing to the asymmetric shape of the vertebral bodies. Fracture can occur both with and without pain, and once present, postural mechanics can worsen existing fractures or precipitate the occurrence of new compression fractures, as increased kyphotic posture produces additional anterior compressive forces.

Nonoperative management of compression fractures may employ thoracolumbosacral orthoses (TLSO; Figure 28–8). The TLSO provides control of flexion, extension, and side bending and can confer more control in different planes of motion, depending on the design. Several designs help to provide the flexion limitation beneficial in compression fracture management.
A thoracolumbar spinal orthosis (TLSO) can control trunk motion in all directions but is not very comfortable to wear.

The cruciform anterior spinal hyperextension (CASH) TLSO primarily provides control of flexion in the sagittal plane for the lower thoracic and lumbar regions. This orthosis consists of a sternal pad, a suprapubic pad, and two lateral pads in the form of a cross anteriorly, with a thoracolumbar pad posteriorly. The CASH is best suited for mild compression fractures of the lower thoracic and thoracolumbar regions.

The Jewett hyperextension TLSO is used to control flexion of the lower thoracic and lumbar spine. This orthosis is similar to the CASH; however, the anterior frame has sternal, suprapubic, and lateral pads in a boxlike design. The Jewett hyperextension TLSO is used for mild compression fractures of the lower thoracic and thoracolumbar regions and provides more lateral support than the CASH.

2. **Unstable thoracolumbar fracture**—An unstable spinal fracture is one in which there is risk of damage to the spinal contents (or there has already been damage to the contents) from the injury, either from excessive intersegmental motion or expulsion of bony fragments into the spinal canal. This type of injury can occur from fracture dislocation, Chance fracture, or ligamentous injury or deformity with dislocation.

For the purpose of evaluating spinal injury, the spine is viewed as having three columns: anterior, middle, and posterior. The anterior column consists of the anterior longitudinal ligament and the anterior two thirds of the vertebral body; the middle column includes the posterior third of the vertebral body and extends to the posterior longitudinal ligament (PLL); and the posterior column includes the pedicles, facets, laminae, spinous processes, and posterior ligaments. When two contiguous columns (ie, anterior and middle, or middle and posterior) are disrupted, the spine is considered to be unstable and at increased risk of damage to the spinal contents.

As an example, an unstable compression fracture can occur in the lumbar vertebrae due to excessive axial loading pressures from a fall. In this type of compression fracture a central crush pattern is seen, in which there is involvement of both the anterior and posterior cortex. In this case, because two contiguous vertebral columns are involved and bony fragments may be expelled into the spinal canal the fracture is considered unstable. A Chance fracture is typically cause by rapid deceleration such as occurs with a restrained passenger in a motor vehicle accident. High shearing forces produce a fracture line through
the entire vertebra; this type of fracture is therefore also considered to be unstable.

Unstable fractures are generally managed surgically with fixation. Given the high level of instability, a TLSO conferring a high level of stability in all planes is employed postoperatively. A total contact TLSO, which is a custom-molded brace fabricated from molded thermoplastics, provides a high level of stability by restricting the greatest amount of motion. The circumferential design encompasses the body, from the sternum to the pubic symphysis anteriorly and from the superior border of the spine of the scapula to the coccyx posteriorly. The typical design is a clamshell, in which anterior and posterior components of the brace are held together by Velcro straps. This facilitates removal for hygiene and when the patient is in bed. This orthosis has been used for traumatic or pathologic fractures of the mid to lower thoracic or lumbar region and is also commonly used in the postsurgical spine (eg, for patients with lumbar decompression and fusion) to protect weakened or damaged segments as they heal.

3. Stable postsurgical and nonsurgical thoracolumbar fracture—TLSOs are used for nonsurgical management of stable spinal fractures or in the postsurgical spine (Figure 28–9). Several different designs exist, which confer varying amounts of motion restriction and stability in each plane. Motion restriction is dependent upon the design of the orthosis and positioning of supportive struts or rigid components, or both.
Restriction of spinal extension is an important feature in bracing for patients with pars interarticularis fracture (spondylolysis), as well as in spinous process fractures. The Taylor brace is a flexion–extension control TLSO with paraspinal bars attached to a pelvic band. The inclusion of a lumbar corset in the design increases intraabdominal pressure, providing further support to the spine in all positions. This brace provides sufficient limitation to extension to allow for healing of spinous process fractures. The Knight–Taylor brace is a Taylor brace with lateral struts, which add further control of lateral flexion.

4. Low back pain without spinal instability—Lumbosacral orthoses may be used in patients with acute low back pain caused by degenerative joint disease, herniated discs, and lumbar muscle strain. They also may be used in patients with stable spinal fracture, where there is no risk of instability or damage to the
spinal contents (eg, transverse process fractures). Studies have shown that these devices affect motion by increasing trunk stiffness and may have value as proprioceptive and kinesthetic reminders to maintain appropriate protective posture. Additionally, by elevating intraabdominal pressure they may reduce the force applied to the spine and lower intradiscal pressure.

The lumbosacral corset helps to contain the lateral and anterior trunk and elevate intraabdominal pressure. The addition of steel uprights may afford some control of flexion and extension. This device is usually made of cloth, and closure is achieved via laces or Velcro straps.

The lumbosacral chair-back brace can be used to limit flexion, extension, and side bending of the lumbar spine as well as to increase intraabdominal pressure. The brace resembles the back of a chair, resting on the iliac bones bilaterally and extending to the T10 level superiorly. It may be fabricated from thermoplastics or aluminum and fastens anteriorly.

The use of bracing for mechanical low back pain beyond the acute phase may cause atrophy of spinal stabilizing muscles; hence, prolonged use of bracing for this indication is generally not recommended. In the case of a transverse process fracture, the brace is generally recommended only for symptomatic use in the acute phase, and often this fracture is managed without bracing.

5. Pelvic fractures and sprains—Sacroiliac (SI) joint pain and instability is a common and disabling problem for many women during pregnancy. An SI orthosis is effective in the treatment of pelvic girdle pain in pregnancy as it decreases pain and joint laxity. The devices also can be used for pelvic fractures and sprains. The SI orthosis, which resembles a belt, provides anterior and lateral trunk support and can also provide some restriction to flexion and extension. It is usually made of cloth and closure is achieved via hooks, Velcro, or metal fasteners.

6. Scoliosis—Scoliosis, defined as lateral curvature of the spine greater than 10 degrees, can be congenital, idiopathic, or neurogenic in origin. The degree of scoliotic curvature is determined using the Cobb angle measured on an anterior–posterior radiographic view of the spine. To obtain the Cobb angle, lines are drawn extending from the endplate of the vertebrae with the most tilt relative to horizontal from the superior and inferior aspect of the curve. The Cobb angle is measured at the point where perpendicular lines drawn from these lines intersect.

Treatment for scoliosis varies, depending on etiology, but generally bracing is reserved until the Cobb angle reaches 25–30 degrees. Patients with lesser degrees of curvature are monitored with serial radiographs. Bracing is used in
the skeletally immature patient with idiopathic scoliosis for Cobb angles between 25 and 45 degrees. The goal of bracing is to slow or halt curve progression. It should be understood by the practitioner and explained to the patient or caregiver that bracing will not correct the existing curve. Curve progression is defined as an increase of 5 degrees or more in Cobb angle during the treatment period. Curves greater than 50 degrees, those limiting diaphragm excursion and causing pulmonary compromise, or those that are progressing despite conservative treatment warrant evaluation for surgical management.

The Milwaukee orthosis is a cervicothoracolumbosacral orthosis that is used to manage scoliosis. It provides control of flexion, extension, side bending, and rotation and is primarily effective for those patients with high thoracic curves (T7 and above) and double curves. The Boston brace is a TLSO that is used for the management of scoliosis in lower thoracic and lumbar curves in the spine. The Boston brace is smaller, low profile, and much better tolerated by patients than the Milwaukee orthosis. If treatment of curves higher than T10 with a Boston brace is desired, a Milwaukee brace superstructure can be added to the Boston apparatus.

The clinical field of sports medicine involves the care and treatment of those injured during sporting activities. Physiatrists were granted the ability to become subspecialty certified in sports medicine in 2006. In this chapter, common sport-specific injuries are discussed, with the injuries organized by sport rather than by body region (the more usual format). The chapter describes the most prevalent injuries in the most common sports, and while many of these injuries can occur among participants in various sports, to avoid repetition they are discussed once, under the most relevant sport.

**CONTACT SPORTS**

Contact sports are those in which participants strike or crash into one another with external force. Examples include football, rugby, and ice hockey. Athletes involved in collision sports are at a high risk for injury due to the violent nature of the competitions.

**COMMON INJURIES IN FOOTBALL & RUGBY**

1. Concussion
ESSENTIALS OF DIAGNOSIS

- Direct or indirect force transmitted to the head.
- Rapid onset of transient neurologic disturbance that resolves spontaneously.
- Functional rather than structural impairment, with normal neuroimaging.

General Considerations

Concussions are considered mild traumatic brain injuries caused by biomechanical forces affecting the head, with symptoms improving spontaneously over time. All athletes suspected of having a concussion must be immediately removed from competition and evaluated onsite.

Clinical Findings

Immediate symptoms may include but are not limited to headaches, neck pain, dizziness, visual or auditory disturbances, loss of balance, post-traumatic or retrograde amnesia, confusion, drowsiness, difficulty concentrating, or fatigue. More objective measures, such as the Sport Concussion Assessment Tool 2 (SCAT2), aim to determine the presence of a concussion and its severity by compiling a number of acute measures.

Computed tomography (CT) imaging should be performed if loss of consciousness (LOC) lasts more than 60 seconds, with suspected skull fracture, or in the presence of any focal neurologic deficits to rule out acute subdural or epidural hematomas.

Treatment

Managing concussion begins with baseline neurocognitive testing. The SCAT2 may be used to this end, though computer-based programs such as the Immediate Post-Concussion Assessment and Cognitive Testing (ImPACT) have been found to be valid and reliable and are popular on all levels of play. Once a concussion has been diagnosed, the player must be monitored for cognitive or functional decline. High school and college athletes who suffer a concussion are not allowed to return to game action on the same day; this applies to all athletes, not
only football and rugby players. Physical and cognitive rest is the mainstay of initial postconcussive management. Athletes of all ages should refrain from activity until symptoms have cleared. Student athletes should refrain from activities that challenge cognition or concentration, such as texting, video game play, and test taking. ImPACT continues to be administered at predefined intervals until the athlete has returned to baseline.

There are no standardized return-to-play (RTP) guidelines. Following a period of rest and once all postconcussive symptoms have completely abated, athletes are placed under a five-phase RTP protocol; if each phase lasts the expected 24 hours, athletes can be game ready by the next weekend. However, progression may only occur if the athlete remains symptom-free. If not, he or she must return to the previous day’s activity until asymptomatic and may have to rest and begin the cycle again if symptoms persist. Phase 1 includes light aerobic activity, phase 2 entails sports-specific coordination exercises, phase 3 involves higher level training drills, phase 4 is when full-contact practice may ensue, and phase 5 is return to competition given that all previous days were free of any concussive symptoms.


2. Anterior Cruciate Ligament Tears

ESSENTIALS OF DIAGNOSIS
Valgus stress to an externally rotated tibia (typically on a planted foot).
Positive Lachman test.

General Considerations

The typical mechanism of an anterior cruciate ligament (ACL) injury is a valgus force applied to an externally rotated femur on a planted foot. More than twice as many injuries occur during games than in practice and more take place on turf than on natural grass. In the English Professional Rugby Union, ACL injuries accounted for 29% of all days lost to knee injury. Women sustain ACL tears at a much higher rate than men, and their tears are more often due to noncontact injuries.

Clinical Findings

Players often hear a “pop” and suffer immediate pain with full-thickness tears. They may also complain of the knee “giving way.” Hemarthrosis usually forms within hours of the injury, so the optimal time to examine the injured knee is within the first hour postinjury or after swelling has somewhat abated and the athlete no longer guards the injured knee. The Lachman test is the most sensitive physical examination maneuver to assess the integrity of the ACL.

Plain radiographs should be taken to look for a Segond fracture, which is an avulsion of the lateral tibial plateau. Magnetic resonance imaging (MRI) can be used not only to evaluate the ACL itself, but also to determine the presence of concomitant injuries, which are common.

Treatment

Initial management of ACL tears involves rest, ice, hinged bracing, crutch ambulation until weight bearing is tolerable, and early range of motion (ROM). Isometric strengthening exercises are important to prevent contracture and to lessen arthrogenic muscle inhibition of the quadriceps. In the athletic population, surgery is the suggested intervention. Grafts range from bone–patellar–bone and hamstring autografts to patellar tendon allografts. Aggressive rehabilitation has been advocated for all athletes before and after surgery. The most important
aspect of the preoperative phase of rehabilitation is restoration of full extension to maximize long-term functional outcome.

In the immediate postoperative phase, from 0 to 2 weeks, athletes are transitioned to partial or full weight bearing, depending on the presence of additional injuries and repairs. Bracing is optional but generally used. In this initial phase, extension to 0 degrees is again critical, and leg strengthening should reach at least 4+/5 in the quadriceps and 5/5 in the hamstrings. Phase 2 lasts 2–12 weeks and should focus on knee hyperextension, flexion to 130 degrees, proprioception and balance control, and unhindered walking. Phase 3, from 3–6 months postsurgery, maximizes balance, flexibility, proprioception, and begins targeting running and sport-specific drills. The final phase is RTP. Athletes should not return to competition until they have reached 90% strength in the injured leg compared with the uninjured leg and can perform all full-speed and full-intensity maneuvers necessary for their position.

The most common complications of ACL reconstruction include quadriceps weakness, knee flexion contracture, and anterior knee pain due to patellofemoral instability. There is also a 5% risk of graft failure (Figure 29–1).
Figure 29–1 T2-weighted sagittal image of a failed anterior cruciate ligament reconstruction. Narrow arrow represents complete graft disruption, and thick arrow demonstrates tibial tunnel.


3. Medial Collateral & Lateral Collateral Ligament Tears

**ESSENTIALS OF DIAGNOSIS**

*Medial Collateral Ligament Tear*
- Force to the lateral knee, usually when in flexion.
- Pain on valgus stress test.

*Lateral Collateral Ligament Tear*
- Force to the medial knee.
- Pain on varus stress testing.

**General Considerations**

Tears to the medial collateral ligament (MCL) occur most often with a direct valgus blow to a flexed knee when the foot is planted. In the English Professional Rugby Union, 25% of all days lost to knee injury were a result of MCL tears. The lateral collateral ligament (LCL) of the knee is very rarely injured in isolation, and tears are more often associated with other structural damage.

**Clinical Findings**

Using the valgus stress test, the examiner elicits pain but finds no laxity in a grade I MCL injury; finds some laxity with a clear end point in grade II; and notes gapping of the medial joint without a clear end point (indicating complete disruption of the ligament) in grade III. LCL tears are graded similarly to those of the MCL, but diagnosis is made by testing for varus laxity.

Radiographs should be obtained to rule out epiphyseal or occult fractures.
The use of MRI for evaluation of MCL sprain is only indicated when other injuries are suspected. However, in LCL sprains, an MRI scan is indicated for confirmation because an avulsion of the biceps femoris tendon or ACL tear can cloud the diagnosis.

**Treatment**

Isolated MCL tears very rarely require surgery; rather, they are treated with a hinged brace for up to 6 weeks, depending on the grade. Conservative treatment for MCL injuries requires a stepwise, multiphase rehabilitation program over 4–6 weeks. It is imperative that all therapy be prescribed and tailored on an individual basis, as no universal algorithm can apply to all athletes.

Treatment of isolated LCL tears consists of conservative management in a manner similar to that for MCL injuries. Although high-level athletes may opt for early surgical repair, one cohort study of isolated grade III LCL tears in National Football League (NFL) players showed faster RTP and more optimal long-term outcomes in those treated nonoperatively. In LCL injuries, surgery is the treatment choice in athletes with multiligament or complex structural injury.

Flexion–extension braces can be worn during competition to help minimize the incidence of recurrent MCL and LCL sprains and tears; however, there is no evidence for their overall efficacy.

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**4. Meniscal Tears**
The menisci are most commonly injured in football players, acutely, when a rotational shear force is applied to a planted foot. Tears are also caused by long-term repetitive stress on the knee. In a study of NFL players, 17.3% of participants were found to have sustained a full-thickness meniscal tear (Figure 29–2). Further information regarding meniscal injuries can be found in Chapter 30.
posterior horn medial meniscus tear with both oblique and longitudinal components (arrow).


5. Hip Pointer

ESSENTIALS OF DIAGNOSIS

- Trauma to and pain over the iliac crest.
- No evidence of injury on radiographs.

Any blow to the iliac crest or fall on a hard surface can cause a contusion to the bone, known as a “hip pointer.” The primary symptom is isolated pain over the iliac crest, with no evidence of osseous injury on radiographs. Rest, ice, nonsteroidal antiinflammatory drugs (NSAIDs), ROM exercises, and strengthening of the muscles of the hip and pelvic girdle are mainstays of treatment. Padding of the iliac crest and gluteus muscles during competition is the only preventative measure. RTP is allowed when the athlete can tolerate the discomfort and can perform all sport-specific activities under full competitive conditions.


6. Quadriceps Contusions
Direct trauma to anterior thigh.

Pain and swelling over the quadriceps.

General Considerations

Quadriceps contusions can occur when a player sustains direct trauma to the anterior thigh, as occurs in blocking and full-contact collisions.

Clinical Findings

Pain and swelling over the anterior thigh and decreased ROM are the most common presenting symptoms of quadriceps contusion. Using the Jackson and Feigin classification, quadriceps contusions can be differentiated as mild, moderate, or severe based on ROM and gait patterns (Table 29–1). Imaging is not usually necessary.

Table 29–1 Jackson and Feigin classification of quadriceps contusions.

<table>
<thead>
<tr>
<th>Grade</th>
<th>Knee Flexion (degrees)</th>
<th>Gait</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>&lt; 90</td>
<td>Normal</td>
</tr>
<tr>
<td>Moderate</td>
<td>45–90</td>
<td>Antalgic</td>
</tr>
<tr>
<td>Severe</td>
<td>&lt; 45</td>
<td>Severely antalgic</td>
</tr>
</tbody>
</table>

Treatment

Immediate management of a quadriceps contusion requires immobilization of the knee joint in flexion, with ice and compression for 24 hours postinjury to
decrease hematoma formation. Functional rehabilitation begins once pain-free ROM has returned and can be advanced based on the athlete’s tolerance and overall improvement. RTP is dependent on the player attaining a fully functional knee with ROM to at least 120 degrees and the ability to perform all sport-specific activities. The use of NSAIDs has been advocated to help prevent myositis ossificans, which is a potential long-term complication of hematoma formation from a contusion.


7. Anterior Glenohumeral Dislocations

ESSENTIALS OF DIAGNOSIS

▶ Appropriate mechanism of injury (most commonly, collision with the shoulder abducted and externally rotated, fall on a hyperflexed arm, or blow to the shoulder in slight adduction).
▶ Prominent humeral head and loss of normal shoulder contour.

General Considerations

The shoulder accounts for a large proportion of injuries, with dislocations being the most severe in terms of time missed from competition. Dislocations occur most often in the anterior direction (Table 29–2). Three mechanisms of anterior dislocation have been described in rugby and football players. The most prevalent mechanism involves a tackle with the shoulder abducted and externally rotated. Other mechanisms, in order of frequency, are a fall on a hyperflexed
arm, and a blow to the shoulder in slight adduction.

### Table 29–2 Evaluation of glenohumeral joint dislocations.

<table>
<thead>
<tr>
<th>Direction of Dislocation</th>
<th>Mechanism of Injury</th>
<th>Clinical Findings</th>
<th>Associated Injuries</th>
</tr>
</thead>
</table>
| Anterior                 | Tackle or fall on an extended, abducted, and externally rotated arm  
                          | Fall on a hyperflexed arm                                   | Loss of shoulder contour          | Bankart lesion                   |
|                          | Blow to the shoulder with the arm in slight adduction     | Prominent humeral head                                      | Bony Bankart lesion              |                                  |
|                          |                                                          | Sulcus below the acromion                                   | Hill–Sachs lesion                |                                  |
|                          |                                                          | Possible loss of sensation over the lateral shoulder (axillary nerve) | +/− Pain                        |                                  |
| Posterior                | Direct blow to or fall on an internally rotated and adducted arm | Pain with loss of external rotation                          | Reverse Bankart lesion           |                                  |
| Inferior                 | Fall on an abducted and externally rotated arm            | Shoulder locked at 90 degrees of abduction                   | Same as anterior dislocations     |                                  |
|                          |                                                          | Severe pain                                                 |                                  |                                  |

### Clinical Findings

Athletes who suffer traumatic anterior dislocations on the field instantly manifest loss of normal shoulder contour, a prominent humeral head, and a sulcus below the acromion. Pain may or may not be present. Testing of sensation to light touch over the lateral arm and of deltoid strength must be done immediately to check for injury to the axillary nerve, which is a potential serious complication. Radiographs of a dislocated shoulder should be obtained prior to reduction, as a fracture may be present and reduction could aggravate the break. However, given that most reductions occur on the field rather than in an emergency department setting, postreduction films are indicated.

### Treatment

There is no standard technique for shoulder reduction. The athlete may be able to self-reduce; alternately, the medical staff may use counter-traction or weight-
assisted reduction on the sideline or in the emergency department, with conscious sedation if needed. A postreduction sling can be used for patient comfort, if needed. Recent evidence supports primary surgical stabilization in players younger than 25 years of age owing to the high rate of recurrence. Prior to surgery, MRI is performed to evaluate the glenohumeral ligaments and for secondary associated injuries. Detachment of the labrum from the anterior glenoid margin, called a Bankart lesion, is normally repaired during arthroscopic stabilization of the shoulder (Figure 29–3). A fracture of the glenoid rim is termed a bony Bankart, and a Hill–Sachs lesion is a compression fracture of the posterior humeral head that occurs as it is displaced from the glenoid fossa. The most common surgical procedure is a labral Bankart repair. Full RTP can take up to 3 months.

![Figure 29–3](image)

▲ **Figure 29–3** Anteroposterior (AP radiograph reveals a bony Bankart fracture of the inferior glenoid rim (arrow) following glenohumeral dislocation.

8. **Posterior & Inferior Glenohumeral Dislocations**
ESSENTIALS OF DIAGNOSIS

- Posterior dislocation: internal rotation and adduction of the shoulder.
- Inferior dislocation: external rotation and abduction of the arm to 90 degrees.

## General Considerations

Glenohumeral dislocations are much less common in the posterior direction than in the anterior direction, and inferior dislocations occur in only about 1% of all traumatic dislocations (see Table 29–2). A posterior dislocation results from a direct blow to or fall on an internally rotated and adducted arm. An inferior dislocation, also termed *luxatio erecta*, occurs with a fall on an abducted and externally rotated arm.

## Clinical Findings

An athlete with a posterior dislocation has loss of contour of the anterior shoulder, with pain and loss of external rotation. An athlete with an inferior dislocation has extreme pain because the humeral head is locked beneath the glenoid, preventing both adduction and internal rotation. Radiographs should include anterior–posterior, lateral, Y, and axillary views, although the latter may not be possible because of patient discomfort.

## Treatment

Forward traction and pressure on the humerus is necessary to relocate posteriorly dislocated shoulders. Reduction of inferiorly dislocated shoulders is a two-step process that should be done in an emergency or clinical setting under conscious sedation because of severe pain during reduction. Guidelines for MRI and surgical management are similar to those for anterior glenohumeral dislocations (see earlier discussion).
9. Acromioclavicular Joint Separations (see Chapter 30)

ESSENTIALS OF DIAGNOSIS

- Pain over the acromioclavicular joint.
- Physical findings consistent with type I–VI injury, using the Rockwood classification.

The acromioclavicular joint (ACJ) is a synovial joint formed by the acromion of the scapula and the distal end of the clavicle. Injuries to the ACJ have been reported to account for 41% of shoulder injuries in football and 32% of shoulder injuries in rugby, the highest percentage of any shoulder pathology. The usual mechanism of injury during football or rugby is a direct hit from one player to another.

The joint has dynamic and static stabilizers, the most important in terms of separations being the joint capsule, the coracoclavicular (CC) ligament, and the acromioclavicular (AC) ligament. The Rockwood classification uses the disruption of these structures as the basis for six different types of ACJ injury. Type I involves sprain of the joint capsule and the AC ligament, with tenderness over the joint itself; type II involves complete tearing of the capsule and AC ligament, sprain of the CC ligament, and a palpable step-off from the clavicle to the acromion; type III involves complete tears of the capsule, AC ligament, and CC ligament and a visual step-off, with elevation of the clavicle up to 100%; type IV involves posterior dislocation of the clavicle; type V involves greater than 100% elevation of the clavicle; and type VI involves inferior dislocation of the clavicle (Figure 29–4). In one study of National Collegiate Association of America (NCAA) football players, 96.4% of ACJ injuries were type I or II.
Weighted radiographs show the level of clavicular displacement and aid in grading of the injury.

Figure 29–4 AP radiograph of the right shoulder demonstrates a type V acromioclavicular separation, indicating complete disruption of the joint capsule and all ligaments, which allows for greater than 100% displacement of the clavicle relative to the acromion.

Management depends on the type of injury and is described in detail in Chapter 30. Conservative treatment is indicated for type I, II, and III separations; and surgical repair for types IV, V, and VI, and type III separations that do not respond to therapy. The phases of therapy include acute, recovery, and RTP. Return to competition often occurs after 2–4 weeks for type I separations, 4–6 weeks for type II, and 6–8 weeks for type III.

10. Mallet Finger

**ESSENTIALS OF DIAGNOSIS**

- Sudden forced flexion of an extended distal interphalangeal (DIP) joint.
- Pain and swelling in the finger with inability to extend the DIP joint.

**General Considerations**

The terminal extensor mechanism of the finger crosses the dorsal aspect of the DIP joint and is essential for full active extension of the digit. In ball sports, the ball can strike and forcefully flex an extended fingertip, causing the extensor tendon to snap or an avulsion fracture of the tendinous attachment at the distal phalangeal base. This disruption of the extensor mechanism is called a “mallet finger.” In football, wide receivers are at particular risk for this injury when attempting to catch a pass.

**Clinical Findings**

Most mallet injuries involve the middle finger, although the injury can occur in any digit, including the thumb. Athletes present with pain and swelling, tenderness over the distal phalanx, and an inability to actively extend the DIP joint. Radiographs may show a small avulsion fracture, which is only significant if more than one third of the joint is involved.

**Treatment**
Treatment consists of splinting of the injured digit in slight hyperextension for 6–8 weeks, and compliance is essential. Any flexion of the DIP joint will disrupt healing and put the athlete at risk for recurrent injury or longer absence from play. Rapid RTP is only possible if there is no risk of disrupting the splinted finger. However, because of the importance of all finger movements in football and rugby, especially among the skill positions and in tacklers, cessation of competition until full healing is achieved is often indicated. RTP may occur once pain-free active extension of the DIP joint has been achieved after a full course of splinting. The splint must continue to be worn during all sporting activity and at night for an additional 6–8 weeks. Throughout the course of treatment and RTP, the proximal interphalangeal joint should remain mobile, and skin breakdown should be prevented.


11. Jersey Finger

**ESSENTIALS OF DIAGNOSIS**

- Pain and swelling in the finger with an inability to flex the DIP joint.
- History of a tackling or grabbing injury.

When football or rugby players tackle or grab an opponent’s jersey, they are at risk for disruption of the flexor digitorum profundus from the distal phalangeal base, an injury termed a “jersey finger.” Seventy-five percent of these injuries involve the fourth digit. The athlete presents with pain and swelling over the volar aspect of the finger, often with the digit in extension. There may be tenderness over the full length of the flexor digitorum profundus, and the athlete will not be able to actively flex the DIP joint with isolation of the tendon. The
flexor digitorum superficialis should also be evaluated.

Surgery is indicated to repair the disrupted flexor digitorum profundus tendon, and early intervention within 7–10 days improves outcome. Postoperative hand rehabilitation with a focus on flexor stretching to decrease long-term stiffness is essential for full recovery. Return to tackling sports is not usually allowed until at least 3 months after surgery.


12. Gamekeeper’s Thumb

**ESSENTIALS OF DIAGNOSIS**

- Pain at the base of the first digit.
- Radiographs may show a fracture.

**General Considerations**

Partial or complete ulnar collateral ligament (UCL) tears account for 86% of all traumatic thumb injuries. A fall on an outstretched arm with an abducted thumb or clutching a ball while falling to the ground may produce a disruption of the UCL of the thumb. This injury is also commonly seen in skiers who suffer a valgus stress to an abducted thumb while holding a pole during a fall.

**Clinical Findings**

The athlete presents with pain, swelling, and tenderness over the ulnar aspect of the base of the thumb and the first MCP joint. There may also be ulnar deviation
or subluxation of the proximal phalanx. Radiographs must be obtained to
determine whether an avulsion fracture is present, but should be obtained prior
to physical examination to decrease the risk of worsening an unknown break.
When no end point is felt on valgus testing, a complete tear is deemed present. If
the UCL displaces proximally, the adductor aponeurosis of the adductor pollicis
may slip between the torn ligament and the bone, thereby preventing healing.
This abnormality, termed a Stener lesion, should be suspected in any athlete with
a displaced avulsion fracture visible on plain radiographs.

**Treatment**

Conservative management with a thumb spica splint or thermoplastic casting for
4 weeks is the treatment of choice for partial tears of the UCL. Complete tears
with no end point on valgus stress or displaced avulsion fractures necessitate
surgical repair. Regardless of management, athletes may return to competition if
their position allows use of a splint or cast, such as offensive or defensive
linemen in football. Once the thumb has regained full strength with normal pain-
free ROM, athletes may return to full competition without splinting or casting.
For partial tears this generally occurs after about 4 weeks; complete tears take
about 3 months, regardless of whether surgery was performed.

Peterson JJ, Bancroft LW: Injuries of the fingers and thumb in the athlete. Clin

Ritting AW, Baldwin PC, Rodner CM: Ulnar collateral ligament injury of the

**COMMON INJURIES IN ICE HOCKEY**

Frequent accelerations and decelerations, changes in direction, and the overall
force produced by full-body hitting and checking at high velocities place ice
hockey players at a high risk for injury. Many of the injuries discussed earlier in
the football and rugby section, particularly concussions, glenohumeral
dislocations, and AC joint separation, are also prevalent in hockey. In this
section, additional injuries not specific to but seen quite often in the sport are
examined.
1. Athletic Pubalgia (Sports Hernia)

**ESSENTIALS OF DIAGNOSIS**

- Nonspecific groin or lower abdominal pain, or both, with tenderness over the pubic tubercle.
- Positive MRI findings involving the pubic bone and soft tissues.

**General Considerations**

The differential diagnosis for groin and lower abdominal pain is vast. It includes but is not limited to femoroacetabular impingement, arthritis, and labral tears; lumbar stenosis or disc herniation; adductor tendinopathy or rupture; muscle strains or contusions; osteitis pubis; pelvic stress fractures; iliopsoas tendinopathy; and athletic pubalgia, often referred to as a “sports hernia.”

Some physicians argue that *athletic pubalgia* is an umbrella term referring to myriad potential diagnoses, with sports hernia as only one subset. Meyers and colleagues characterized the injury as a hyperextension injury, which results when the pubis serves as a pivot point of insertion for the rectus abdominis and adductor longus muscles. The injury results in disruption of the conjoined tendon of the adductor longus and rectus abdominis, leading to degeneration arthropathy of the pubic symphysis.

For the purposes of this discussion we have opted to view athletic pubalgia and sports hernia as the same entity: a spectrum of potential muscular, fascial, tendinous, and aponeurotic disruptions—including those of the rectus abdominis and adductor longus aponeurosis and insertions, the external obliques, the interface between conjoined tendon and rectus abdominis, and the muscles that form the conjoined tendon (internal obliques and transversus abdominis)—that presents as nonspecific lower abdominal or groin pain, or both. Athletic pubalgia is seen most often in athletes who perform repetitive bending and twisting, such as soccer and hockey players (“hockey groin”), which leads to either one larger tear or many microtears in the structures listed.
Clinical Findings

A. Symptoms and Signs
Athletes present with lower abdominal and groin pain that worsens with exercise and improves with rest. Most players can pinpoint a specific inciting event, although onset is often insidious. The pain is typically, though not always, unilateral; may radiate to the perineum, thigh, or scrotum; and is worse with cutting, twisting, turning, kicking, sit-ups, and activities involving a sudden Valsalva maneuver.

Examination should include evaluation of adductor-, iliopsoas-, and abdominal muscle–related pain and strength at the pubic symphysis. Patients usually report tenderness at or above the pubic bone on or near the insertion of the rectus abdominis or adductor longus muscle. A subset of patients may have point tenderness at the external inguinal ring or a dilated superficial inguinal ring. Patients may also have pain at the pubic tubercle and symphysis. Unlike an abdominal or inguinal hernia, a sports hernia has no palpable defect or bulge on palpation.

Provocative testing with resisted sit-ups and resisted adduction with the hips in external rotation may reproduce groin pain. Additionally, the following five signs and symptoms have been identified as clues in diagnosing sports hernia: (1) deep groin or lower abdominal pain, (2) pain with activity such as kicking, (3) palpable tenderness over the pubic ramus at the insertion of the rectus abdominis and conjoined tendon, (4) pain with resisted hip adduction at 0, 45, or 90 degrees of hip flexion, and (5) pain with resisted abdominal curl-ups. The pain may be elicited with a resisted sit-up or resisted hip flexion when supine or with bending forward when standing. A full evaluation of the hip joint, lumbosacral spine, abdomen, adductor and iliopsoas tendons, and pelvis must be done to rule out other conditions.

B. Imaging Studies
Radiographs of the hip and pelvis are standard to rule out stress fractures, impingement, or other bony pathology. If a sports hernia is suspected, an MRI with a special protocol is indicated. MRI findings can diagnose sports hernia is 68% of cases. Findings can vary based on the injury, but there is typically tearing of the rectus abdominis–adductor aponeurosis and bony edema of the pubic symphysis. The presence of a T2 hyperintense area at the anterior subcortical region of the bone, 1–2 cm lateral to the symphysis, is indicative of rectus
abdominis insertional pathology. Other findings on MRI may include a more intense and linear configuration, reflecting a stress response or early stress fracture 5–15 mm lateral to the pubic symphysis. Often a fluid or near-fluid signal is present within the pubic symphysis. The combination of these and previously mentioned findings represents injury at the lateral edge of the rectus abdominis and adductor aponeurosis. MRI findings can diagnose sports hernia in 68% of cases.

Dynamic ultrasound can also be used to visualize posterior inguinal wall deficiency. Ultrasound studies will show abnormal ballooning of the posterior wall when the patient is asked to strain.

**Treatment**

Management for in-season athletes with a sports hernia begins with a period of rest, ice, NSAIDs, and possible corticosteroid or platelet-rich-plasma injections to the insertion of the rectus abdominis or adductor longus muscle. In some instances, the injury is self-limited.

After 4 weeks in which only closed-chain lower limb exercises are allowed, the athlete is put through a sport-specific functional assessment to determine whether pain-free RTP is possible. If the pain has not dissipated, the player is allowed to return to competition if he or she can tolerate the discomfort and perform at a high level. There is no evidence that competing through pain worsens the abdominal wall defect or surgical outcome. A full course of physical therapy is attempted prior to surgical referral, which is often sought many months after initial onset of symptoms.

If pain persists after a period of rest, surgical treatment should be considered; this consists of unilateral or bilateral reattachment of the rectus. Surgical techniques include both laparoscopic and open procedures and are dependent on the surgeon’s preference and on the exact nature of the MRI findings. Patient satisfaction after surgery ranges from 77% to 100%.


2. Adductor Muscle Strain

ESSENTIALS OF DIAGNOSIS

- Groin pain.
- Local tenderness over the adductor muscle.

General Considerations

Acute “groin pulls” are among the most common injuries in ice hockey, soccer, and other sports involving frequent cutting and sudden changes of direction. The proposed mechanism for injury is that of an excessive eccentric force on the hip adductors as they work to decelerate the leg during a stride. Risk factors for adductor strain include adductor-to-abductor strength ratios of less than 80%, core weakness, lack of preseason training or pre–sport-specific training, and previous adductor strain.

Clinical Findings
Athletes can usually pinpoint the moment the adductor strain occurred. Pain usually localizes to the respective muscle belly, the musculotendinous junction, or the origin of the adductor tendon on the inferior pubic ramus. Palpation along the entire length of the muscle and corresponding tendon is an essential part of the examination. The adductor squeeze test, in which the athlete lies supine with the legs extended and squeezes the examiner’s fist between his or her knees, is another provocative maneuver that reproduces pain. An adductor strain is a clinical diagnosis, and imaging is not necessary unless a more complex injury is suspected, such as an adductor enthesisopathy or athletic pubalgia.

**Treatment**

After the initial injury, rest, ice, compression, and elevation (RICE) are initiated for 48 hours. However, active rehabilitation should be started as soon as acute symptoms have settled. The initial phase of therapy utilizes gentle stretching, massage, and, possibly, dry needling to release tight iliopsoas and gluteal muscles and manual techniques to release myofascial tension in the adductors. This release is especially important for chronic groin pain. To prevent aggravation an injured adductor should not be overstretched. Core and lumbopelvic stability is a critical component of the rehabilitation program to decrease load on the adductor compartment.

Once muscles have been released, a pain-free exercise regimen is implemented and progresses over 8–12 weeks, with the goal of gradually increasing the load on the pubic bones and surrounding soft tissues. Bike riding is acceptable for the first 6 weeks, after which pain-free jogging is allowed. Hockey players may advance to sport-specific exercises when adductor strength is at least 75% that of the ipsilateral abductors and may attempt pain-free skating when that level reaches 90–100%. It has been shown that preseason adductor strengthening in players with an adductor-to-abductor strength ratio of less than 80% helps prevent in-season groin pulls.


Maffey L, Emery C: What are the risk factors for groin strain injury in sport?

LIMITED CONTACT SPORTS

This category encompasses sports such as basketball, soccer, lacrosse, and field hockey, which have in common dynamic movement, limited-contact physical blocking actions, and explosive bursts of speed during play. Players of such sports are prone to injuries of the foot, ankle, knee, and leg. Although full-body collisions are not central elements of play, as in football, rugby, and ice hockey, player-on-player contact that occurs while advancing the ball or blocking another player may cause injuries such as those described earlier for contact sports.

COMMON INJURIES IN BASKETBALL

Basketball is a sport that demands high-intensity dynamic movement in all planes; thus it is no surprise that it carries a higher risk of injury than most other sports. The most common and serious injuries in basketball involve the ankle, leg, and knee.

1. Ankle Sprain (see also Chapter 30)

ESSENTIALS OF DIAGNOSIS

► Lateral ankle tenderness, with edema, ecchymosis, and inability to bear weight.
► Pain on passive inversion with dorsiflexion, or plantar flexion, or both.
► Positive anterior drawer and (occasionally) talar tilt tests, showing laxity.
General Considerations

Ankle injuries are the most common injury experienced by basketball players. The greatest risk factor is a previous recent ankle injury. The history is that of a forced inversion with dorsiflexion of the ankle, with or without an audible “pop,” immediate pain, swelling, and ecchymosis. Lateral ligament injuries are the most common, with the anterior talofibular ligament most frequently involved, followed by the posterior talofibular ligament. Lateral ankle sprain is discussed in detail in Chapter 30.

A high ankle sprain is an injury to the syndesmosis between the distal tibia and fibula, involving the anterior inferior tibiofibular ligament, and represents only 1% of ankle sprains. The mechanism of a high ankle sprain is that of forced eversion and pronation in a plantar-flexed position. Although more rare, it is clinically associated with prolonged recovery and an increased likelihood of requiring surgery.

Clinical Findings

Immediately after the injury, the ligaments of the lateral ankle are tender to palpation, with pain on ankle inversion. Laxity may be noted during the anterior drawer test or the talar tilt test but may not be obvious if there is edema and guarding. A syndesmotic injury presents with swelling and tenderness between the tibia and fibula and a positive squeeze test (in which squeezing of the distal calf causes pain to shoot into the distal ankle). Although rare, fractures can occur—more often in children and most often involving the fifth metatarsal and distal fibula. Radiographs of the ankle should be obtained if there is a high suspicion for fracture based on the Ottawa ankle rules (Table 29–3). An injury to the syndesmosis produces separation of greater than 6 mm between the medial malleolus and the talus on a mortise views, but if this is absent on radiographs and clinical suspicion is high, stress views or a CT scan should be obtained.

Table 29–3 Ottawa ankle rules for radiographic imaging of ankle and midfoot pain.
Once the diagnosis of an ankle sprain has been established, rehabilitation of acute ankle injuries is typically divided into four phases. Phase 1 focuses on reducing pain and edema and protecting ligamentous structures; phase 2, on normalization of gait pattern; phase 3, on return to pain-free daily activities; and phase 4, on return to sports (for details, refer to Chapter 30). Typical RTP is 4–6 weeks, although this varies based on severity, and supportive taping or bracing is often used to promote RTP. Prior to RTP, the athlete should be able to participate in a graded progression of pain-free running, lateral movement, cutting, and jumping, and eventually sport-specific movements. (For additional discussion, see Chapter 30.)

Treatment of high ankle sprains is more prolonged. Athletes with clinical symptoms of a high ankle sprain without significant radiographic findings can be treated conservatively with a non–weight-bearing cast for 4 weeks, while those with radiographic findings should be treated surgically to repair the syndesmotic injury. RTP is approximately 3 months.

2. Patellar Tendinopathy

ESSENTIALS OF DIAGNOSIS

- Anterior knee pain, exacerbated by jumping.

General Considerations

Patellar tendinopathy, also known as “jumper’s knee,” is a common basketball injury and is also seen in other jumping sports, such as volleyball, and explosive running sports.

Clinical Findings

The athlete reports pain at the inferior pole of the patella that is exacerbated by jumping or explosive movements. Pain is typically absent with passive ROM of the knee, and swelling is usually not present. Patellar tendinopathy is rarely confused with other knee disorders, but it is important to differentiate it from patella femoral pain syndrome, fat pad impingement syndrome, and Osgood–Schlatter disease (in adolescents). Patellar tendon rupture is rare, but can occur if an athlete continues to play sports with a thickened, inflamed tendon.
Treatment

Similar to many tendinopathies, treatment includes relative rest as well as rehabilitation with an eccentric exercise program, core stabilization, and ankle stability. Athletes completing a rehabilitation program consisting of eccentric loading of the patella tendon in conjunction with rest from sport showed good recovery at 12 weeks. Corticosteroid injections have not resulted in long-term benefit in the treatment of patellar tendinopathy and increase the risk of patella tendon rupture. Platelet-rich plasma has been used in the treatment of various tendinopathies, but long-term outcomes in patellar tendinopathy remain to be determined. In recalcitrant cases, surgical debridement can be considered.


COMMON INJURIES IN SOCCER, LACROSSE, & FIELD HOCKEY

Soccer, lacrosse, and field hockey are similar sports, all played on a field with limited stoppage in time and with a common theme of attempting to get a ball into a goal. The most common injuries in soccer include ankle, knee, and upper leg injuries. Although there is a fair amount of contact in lacrosse and field hockey, the majority of injuries are minor.

1. First Metatarsophalangeal Joint Sprain

ESSENTIALS OF DIAGNOSIS
Pain and swelling at the first metatarsophalangeal (MTP) joint.

Pain with weight bearing, and passive flexion–extension of the first MTP joint.

General Considerations

The incidence of this injury, also known as “turf toe,” has risen because of the increased popularity of artificial ground surfaces. Athletes who wear flexible shoes with limited support at the toe box are also at risk. This injury typically occurs when the first digit of the foot is planted on the ground while a simultaneous force is directed to the back of the foot. Turf toe is an injury to the plantar capsuloligamentous complex and, like sprain, has three grades (Table 29–4).

Table 29–4 Severity of first metatarsophalangeal joint sprain injury.

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Stretch of the plantar capsuloligamentous complex</td>
</tr>
<tr>
<td>II</td>
<td>Partial tear of the plantar capsuloligamentous complex</td>
</tr>
<tr>
<td>III</td>
<td>Complete tear with avulsion of the plantar plate from the metatarsal head</td>
</tr>
</tbody>
</table>


Clinical Findings

The athlete often has pain and swelling around the first MTP joint. Weight bearing, and any flexion or extension of the first MTP joint, causes pain. Radiographs of the foot are obtained to observe for complications, such as sesamoid injury, avulsion injury, or joint subluxation.
Treatment

Initial treatment consists of RICE and limited weight bearing with a surgical shoe for 72 hours. Most players respond well with nonoperative treatment. Grade I injuries are treated by taping the great toe to the digits; RTP is typically immediate. Grade II injuries are treated and respond similarly to grade I but may require up to 2 weeks before RTP. Grade III injuries require long-term immobilization in a boot or cast, with RTP ranging from 2 to 6 weeks. The presence of any of the complications listed earlier may require surgical intervention. Following surgical repair, recovery may take 6–12 months before RTP. Although most athletes return to full function, those with grade III injuries are at increased risk for hallux valgus or rigidus and failure to achieve full push-off strength.


2. Tarsometatarsal (Lisfranc) Joint Injury

ESSENTIALS OF DIAGNOSIS

Marked swelling and tenderness of the midfoot.
Inability to bear weight, especially with toe walking.
Weight-bearing radiographs may show greater than 2 mm of displacement between the first and second metatarsals.

General Considerations

The Lisfranc joint consists of the medial articulation between the first and second metatarsals and the medial and middle cuneiforms. Although rare, injury
to this joint may occur, most often from low-impact trauma (either direct force or trauma related to a rotational injury).

Clinical Findings

Athletes with this injury may present with marked swelling and inability to bear weight, especially with toe walking. Some athletes may describe a “pop” during the time of the injury. Physical examination reveals tenderness along the Lisfranc joint, pain with compression of the midfoot, and pain when applying a dorsal–plantar stress to the first metatarsal while stabilizing the second metatarsal head. It is important to evaluate the pedal pulse as the dorsalis pedis artery courses over the second metatarsal.

Diagnosis is confirmed radiographically; weight-bearing views of the foot show greater than 2 mm of displacement between the first and second metatarsal. A small avulsion fragment, known as the “fleck sign,” may also be seen arising from the medial cuneiform or the medial aspect of the second metatarsal base (Figure 29–5). If initial radiographs are negative and clinical suspicion remains high, CT scan is the study of choice.
Nonsurgical treatment is indicated for nondisplaced, stable Lisfranc sprains. Athletes are immobilized with a short-leg walking cast or a non–weight-bearing cast for 4–6 weeks or until asymptomatic. Afterward, the athlete should begin with progressive weight bearing and ambulation. Surgical intervention is indicated if the weight-bearing radiographs show displacement of greater than 2
mm compared with the contralateral side.

After surgical fixation, RTP is possible within 12–16 weeks, depending on symptoms. However, if the sport involves high-impact, explosive activity or frequent cutting and turning, this may be a career-ending injury. In addition to the poor rate of RTP, common problems include post-traumatic arthrosis, malunion, and complex regional pain syndrome.


NONCONTACT SPORTS

Sports that are traditionally included in this category include baseball, golf, racquet sports (eg, tennis, racquetball), and extreme sports such as skiing, snowboarding, and skateboarding. Coverage in this chapter is limited to injuries sustained in baseball; readers are referred to other sources for information on additional noncontact sports injuries.

COMMON INJURIES IN BASEBALL

1. Superior Labrum Anterior–Posterior (SLAP) Lesions

ESSENTIALS OF DIAGNOSIS

- Posterior shoulder pain, especially in abduction and exacerbated by overhead activity.
- Tear seen on magnetic resonance arthrography.
General Considerations

The glenoid labrum is a ring-shaped fibrocartilaginous cushion that adds depth and stability to the glenohumeral joint. It serves as a primary attachment for the glenohumeral ligaments and the long head of the biceps tendon. Superior labrum anterior-to-posterior (SLAP) lesions are tears that extend from a point anterior to the biceps tendon to one posterior to the tendon. They are classified into four types. In type 1 lesions, there is fraying and degeneration of the labrum, but its attachment to the glenoid remains intact. In type 2 lesions, the superior labrum and the biceps tendon attachment is detached from the glenoid rim. In type 3 lesions, the superior labrum is detached from the glenoid, but the biceps tendon attachment to the labral rim is intact. Finally, in type 4 lesions, the superior labrum is torn with extension into the biceps tendon and displacement of both the labrum and the tendon into the joint.

When the throwing shoulder is in the cocked position—abducted and externally rotated—the biceps tendon transfers a torsional force to the glenoid labrum. This force can lead to a “peel back” of the posterior superior labrum off the glenoid, resulting in a tear. Risk factors for SLAP lesions include excessive scapular protraction and a glenohumeral internal rotation deficit (GIRD). In GIRD, there is a difference of 20 degrees or more of internal rotation in the throwing shoulder compared with the contralateral side. GIRD has been shown to place pitchers at risk for shoulder injuries such as SLAP tears and posterior impingement of the rotator cuff due to abnormal humeral head translation, although it has been argued that posterior impingement is a normal finding in pitchers.

Clinical Findings

A. Symptoms and Signs

The athlete may present with pain when throwing or may describe pain over the posterior shoulder or posterior–superior joint line. A catch, click, or pop is often experienced. Pitchers may also complain of a “dead arm” (SLAP lesions are among the injuries most commonly associated with this complaint). Examination of the shoulder should include inspection for asymmetry, full ROM, general strength testing, and isolation of the rotator cuff muscles. Special tests may give more information about the function of the glenohumeral joint.

O’Brien’s active compression test involves placing the arm at 90 degrees of
flexion with the elbow extended, the shoulder in full internal rotation, and the arm adducted 10–15 degrees past midline. Maximum force is placed in a downward direction and then repeated with the arm in supination or fully externally rotated. If pain is elicited with the first maneuver and disappears with the second, then the test is a positive and a labral tear is suspected.

The crank test is performed with the patient supine or seated and the arm in 160 degrees of flexion in the scapular plane. An axial load is placed on the humeral head while the arm is externally and internally rotated. Reproduction of pain with or without a click represents a positive test.

The resisted supination external rotation test is more specific and sensitive than either of the preceding tests. It is performed with the patient supine and the injured shoulder at the edge of the examining table. The limb is positioned with the shoulder abducted to 90 degrees, the elbow flexed to about 70 degrees, and the forearm in neutral or slight pronation. The patient is then asked to supinate fully against resistance while the shoulder is externally rotated to its maximum point. The test is positive with reported anterior or deep shoulder pain, a catch or click, or reproduction of symptoms.

**B. Imaging Studies**

Plain films should be obtained to rule out any bony abnormality, although if a labral tear is suspected, magnetic resonance arthrography is usually indicated (Figure 29–6). The dye allows for detection of small tears in the labrum and can assess for loose bodies or additional cartilaginous defects.
Figure 29–6 T1-weighted coronal image of the shoulder reveals a large superior labral tear (arrow).

Treatment

Surgery is the primary form of management for type 2 and 4 SLAP lesions, which are considered unstable because of the detachment of the biceps tendon. About 70% of major league baseball pitchers who undergo labral repair are able to return at the same level of competition. However, overhead athletes who undergo surgery return to competition at the same level less frequently than do non–overhead athletes. Therefore, a trial of conservative treatment with aggressive physical therapy may be warranted in all athletes with suspected SLAP tears, but especially in pitchers and other throwers. Scapular stabilization, rotator cuff stretching (eg, with the sleeper stretch and band work), ROM, proprioceptive control, and dynamic stability are all key components of the early stages of rehabilitation, with progression to full strength, power, and sport-
specific exercises. When rehabilitating the shoulder it is essential that the entire kinetic chain, including the lower limbs and core, be incorporated into the regimen.

Following labral debridement or repair, the athlete is placed in a sling for comfort for 7–10 days and told to follow strict precautions for 6 weeks. Any movement that tenses the biceps tendon, such as shoulder extension, lifting with the elbow extended, or behind the back motions, should be avoided. Abduction and external rotation of the shoulder must also be limited while pain and swelling are controlled. By 6 weeks, fully passive forward flexion of the shoulder should be achieved, at which time internal rotation, extension, and cross-body adduction stretching can begin while introducing strengthening for the rotator cuff, deltoid, internal and external rotators, and extensors. Between 12 and 24 weeks, athletes should focus on advanced scapular strengthening, biceps work with very light weight, and overall neuromuscular control and proprioception. Lifting biomechanics can be introduced, and the athlete should be prepared for sport-specific exercise. Before progressing to a controlled throwing program, upper and lower limb ROM, strength, the kinetic chain, scapular stability, and proprioception should all be assessed. Once the athlete is deemed ready to throw, he or she should be brought along gradually, keeping attention to the distance, duration, intensity, and number of throws at each step.


2. Little League Shoulder
Pain over the proximal humerus during throwing.
Classically an overuse injury.

General Considerations
“Little league shoulder” is an epiphysiolysis of the proximal humerus and is most commonly seen in young athletes between 11 and 16 years of age. It is an overuse injury and is thought to result from constant traction on and torsional overload of the shoulder during maximal external rotation in the cocking phase of throwing. The stress causes microfractures in the epiphysis of the proximal humerus and may resemble a Salter–Harris type I fracture.

Clinical Findings
Young pitchers present with the complaint of progressive shoulder pain when throwing. The pain can usually be localized to the proximal humerus, and the area is often tender. Examination of the shoulder should include inspection for asymmetry, palpation, strength testing, ROM, and any special tests to evaluate for other potential causes of pain. The examiner should also assess for GIRD, which has been shown to be a cause of shoulder pain in the pediatric population, although not specifically associated with little league shoulder.

Plain films show widening of the proximal humeral epiphysis (Figure 29–7). An anterior–posterior view with the arm in external rotation may be necessary to visualize the widening. If radiographs are negative and clinical suspicion remains high, MRI is indicated.
Figure 29–7 AP external rotation film with the arm extended demonstrates widening of the proximal humeral epiphysis consistent with little league shoulder.

**Treatment**

The mainstay of management is rest for at least 3 months followed by a progressive throwing program. Physical therapy may be initiated at any point during the rest or throwing phases of recovery, focusing on pain-free ROM, strengthening, stability, and neuromuscular control. In response to research conducted at the American Sports Medicine Institute in Birmingham, Alabama, the USA Baseball Medical & Safety Advisory Committee has established recommendations for pitch count limits and type of pitches that youth baseball pitchers should be allowed to throw. As with all injuries in the pediatric and adult populations, prevention is critical.


3. Ulnar Collateral Ligament Sprains & Tears

**ESSENTIALS OF DIAGNOSIS**

- Medial elbow pain.
- Valgus overload.

**General Considerations**

The ulnar collateral ligament (UCL) is the primary stabilizer of the elbow when valgus stress is placed on the joint. During the late cocking and acceleration phases of the throwing motion, the UCL is subject to particularly high tensile forces that over time may cause repetitive microtrauma and subsequent fraying or complete rupture of the ligament.

**Clinical Findings**

Overhead throwers with UCL injuries usually present with medial elbow pain. Pitchers often report a loss of velocity and throwing command, instability, and muscular weakness. Coaches may notice a “dropped elbow” during the throwing motion, which predisposes the player to injury. On examination, there is tenderness over the medial aspect of the elbow and reproduction of pain with valgus stress at 30 degrees of elbow flexion. Valgus testing should also be performed at 0 degrees of elbow flexion. Musculoskeletal ultrasound may be used to evaluate the ligament, although MRI is commonly used to confirm a tear.

**Treatment**

Surgical correction of UCL injuries is common in overhead athletes. Conservative management of partial tears may be indicated, especially in position players who are able to manage the pain. Most tears in pitchers,
however, are repaired. Named after the first pitcher to undergo the procedure, “Tommy John” surgery aims to reestablish elbow stability and allow for eventual RTP. Postoperative rehabilitation lasts 6–9 months, with more than 80% of major league pitchers returning to play the following season after injury with no loss of performance level.


4. Little League Elbow

ESSENTIALS OF DIAGNOSIS

- Medial elbow pain in an adolescent athlete.
- Overuse injury.

General Considerations

“Little league elbow” refers to apophysitis of the medial epicondylar growth plate of skeletally immature athletes, though this term has expanded over the years to include most medial elbow pain in adolescent athletes. Repetitive unnatural valgus stress placed on the elbow during throwing, overuse, high pitch counts, and inadequate recovery times are the primary risk factors. In adults, these factors often lead to UCL injuries. However, because pitch speeds and overall forces are not as high as in children and adolescents as they are in adults, the microtrauma incurred in younger players results in damage to the apophysis near the UCL attachment. It has been estimated that 20–40% of baseball players aged 9–12 years experience medial elbow pain.
### Clinical Findings

Adolescents present with medial elbow pain and tenderness to palpation over the medial epicondyle. They may have elbow laxity on valgus stress testing. It is essential to check for any neurovascular changes or flexion contractures, as young athletes may compensate for injury by bending the elbow. Most radiographs are negative, and diagnosis is primarily clinical in nature. MRI may be helpful in identifying early bony edema and evaluating for ligamentous injury.

### Treatment

Cessation of throwing for 4–6 weeks is the mainstay of management. Ice and NSAIDs are also helpful. During the rest phase general conditioning, including lower body strengthening and kinetic-chain stability exercises, are important to maintain conditioning and help prepare for a structured throwing program. If contractures are present, an elbow extension brace may be used.

Multiple organizations including USA Baseball, Little League Baseball, and the American Sports Medicine Institute in Birmingham, Alabama, have published guidelines for the prevention of pitching injuries in skeletally immature throwers. Proper biomechanics, rest, adherence to evidence-based pitch count limits, response to fatigue or early signs of potential injury, and medical followup for any complaint are all necessary to lower the injury rate in youth baseball players.

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### ENDURANCE SPORTS

Endurance sports have gained popularity over the years with both athletes and the general public. While beneficial to the population for health and wellness, these sports, owing to their nature, carry an increased risk for injury.

### COMMON INJURIES IN RUNNING
1. Patellofemoral Pain Syndrome

**ESSENTIALS OF DIAGNOSIS**

- Anterior knee pain, worsened with prolonged sitting (theatre sign) and stairs.
- Retropatellar crepitus, pain with knee squat, and positive J-sign.

**General Considerations**

Patellofemoral pain syndrome (PFPS), also known as “runner’s knee” accounts for 25% of running injuries and is more common in women. Runners typically present with progressive onset of achy and occasionally sharp peripatellar pain. At first the ache or pain is noticed several hours after a run, most notably with prolonged sitting or descending stairs, but eventually it progresses to pain while running. Overtraining and biomechanical malalignment tend to play a prominent role in the development of PFPS. The possible factors that contribute to dynamic patellar instability are many, with the most common being femoral anteversion; knee valgus due to weak hip abductors and external rotators; trochlear dysplasia; overpronation of the foot; muscular imbalance of the vastus medialis oblique and vastus lateralis; tightness of the iliobibial band, hamstrings, quadriceps, and lateral retinaculum; and laxity of the medial patellofemoral ligament.

**Clinical Findings**

Physical examination focuses on biomechanical and dynamic examination, as well as a focused knee examination evaluating ROM, joint stability, and evidence of effusion. Dynamic patellar stability should be examined during active knee flexion and extension to observe for a J-sign (excessive lateral patellar movement at terminal extension). The fat pad should be evaluated for hypertrophy and tenderness. Imaging studies begin with lateral radiographs in 30 degrees of knee flexion and a sunrise view to evaluate patellar height and for
patella tracking.

**Treatment**

Treatment for PFPS is largely conservative. A comprehensive rehabilitation program is the mainstay of treatment and should be tailored to the individual based on information obtained from the history, examination, and biomechanical assessment. There is no role for NSAIDs in the treatment of PFPS. Once pain and inflammation have been reduced through rest and therapeutic modalities, therapy progresses to core strengthening, flexibility, and neuromuscular control of the hip abductors, hip external rotators, quadriceps, and hamstrings. Patellar mobility is also assessed and treated with mobilization. Dynamic stabilization braces have been shown to decrease pain in 50% of women. Correction of training errors is essential. Common surgical procedures include chondroplasty and lateral retinacular release, but there are no scientific data to support these interventions and some studies report poor surgical outcomes with worsening of symptoms after surgery.


2. Iliotibial Band Friction Syndrome

**ESSENTIALS OF DIAGNOSIS**

- Lateral knee pain at the same point in every run, worsened on hills or uneven surfaces.
- Positive Ober test revealing a tight iliotibial band.

**General Considerations**
Iliotibial band friction syndrome (ITBFS) is another common running injury and is the most common cause of lateral knee pain in runners. While it more often affects distance runners, it can affect sprinters and cyclists as well. The iliotibial band (ITB) begins proximally as the tensor fascia latae and continues as a thick band down the lateral thigh, crossing the knee to insert onto the patella and Gerdy’s tubercle of the tibia.

Current literature points to biomechanical abnormalities as the cause of ITBFS, particularly tibial internal rotation, decreased subtalar motion, and weak hip abductors. Pain is thought to be caused by compression of richly innervated fatty and connective tissue.

Clinical Findings

A runner with ITBFS often presents with acute sharp pain in the lateral knee that begins at the same point in every run. Questioning usually reveals a change in the training program within the past month. Tenderness over the lateral femoral epicondyle is worsened with repetitive knee flexion and extension, and the Ober test reveals a tight ITB. Imaging studies are typically not necessary.

Treatment

Initial treatment focuses on pain control and reduction of inflammation through relative rest, ice, and NSAIDs. Corticosteroid injection can be considered for immediate relief, but studies do not show a long-term benefit. The wear pattern of shoes should be examined, old shoes replaced, and suggestions given for the proper choice of shoe (motion-control, stability, neutral), with or without orthotics. Comprehensive physical therapy is often beneficial for stretching, sustained myofascial tension, and core stabilization, and a home exercise program should include use of a foam roller. Soft tissue mobilization with active release therapy or Graston-type techniques has not been subjected to randomized controlled trials but may be of benefit. There are currently no studies to support the use of an ITB compression strap. Surgery is reserved for refractory cases and typically involves partial resection of the ITB over the lateral femoral epicondyle or a Z-lengthening procedure.

3. Medial Tibial Stress Syndrome

ESSENTIALS OF DIAGNOSIS

- Diffuse pain along the mid-distal posteromedial tibia.
- The presence of bony tenderness should prompt evaluation for stress fracture.

General Considerations

The hallmark of medial tibial stress syndrome (MTSS), commonly known as “shin splints,” is pain over the posterior medial tibial border that develops at the start of a training program or season. It is a classic overuse injury due to “too much, too soon.” Both intrinsic and extrinsic risk factors have been identified. Intrinsic factors include female gender, body mass index above 20, overpronation, dropped navicular, and above-average plantar flexion strength. Extrinsic factors include being a novice runner (less than 5 years), previous history of MTSS, training errors, and running on a different type of terrain.

Clinical Findings

The runner typically presents early in the season with diffuse distal posteromedial calf pain that is often bilateral in nature. The pain is sharp and severe at the beginning of a run and slowly dissipates once the muscles are warmed. As the condition worsens pain lasts longer into a workout and eventually develops into an achy pain that persists hours after the workout. On
physical examination, diffuse tenderness of the soft tissue is noted immediately posterior to the middle and distal medial tibial border. Mild swelling may be present, as well as pain with passive ankle plantarflexion and eversion of the ankle and a flexible pes planus. An area of increased point tenderness should be appropriately evaluated for stress fracture.

Imaging studies are not needed unless there is a need to rule out stress fracture. If examination findings are questionable, bone scintography or MRI can be ordered. However, MRI is usually preferred because it can visualize both bony injuries and soft tissue edema. In addition, bone scintography has a low sensitivity and higher false-positive rate that MRI.

**Treatment**

Treatment of MTTS is conservative. Typically RICE therapy is instituted. Extracorporeal shock wave (ECSW) therapy has shown long-term benefit for recalcitrant cases of MTSS. Surgery is reserved for severe refractory cases and while good to excellent outcomes were reported in 69–92% of runners in one study, return to preinjury level was not as favorable.


**4. Stress Fracture**

**ESSENTIALS OF DIAGNOSIS**

- Point bony tenderness exacerbated by weight-bearing activities.
- Plain radiographs often are inconclusive; MRI is the next study of choice.
- Determination of cause is key to prevention of future stress fractures.
General Considerations

Stress fracture is one of the most debilitating injuries for a runner because the diagnosis is an absolute indication for rest from running. A stress fracture is a bony injury that results from repetitive microtrauma without adequate recovery time. Several risk factors for stress fracture have been identified. Intrinsic risk factors include female gender, low bone mineral density, menstrual disorders, decreased body muscle mass, leg-length discrepancy, and poor preparticipation physical conditioning. Extrinsic risk factors include rapid increase in training regimen, running on angled surfaces, running more than 20 miles per week, improper footwear, wearing a pair of running shoes for more than 6 months, poor nutrition, and smoking.

The most common sites for development of stress fractures in runners are the tibia, metatarsal, fibula, navicular bone, femur, and pelvis (Table 29–5). Differentiation must be made between high-risk and low-risk stress fractures, as the distinction will have an effect on treatment and return to activity. A high-risk stress fracture is located on the tension side of a bone or involves cancellous bone and is commonly seen at the femoral neck, navicular bone, fifth metatarsal diaphysis, and tibial diaphysis. A low-risk stress fracture involves the compression side of a bone or involves cortical bone, and is most commonly seen at the distal posterior medial tibia and second to fourth metatarsals. Injuries involving compressive forces allow for osteogenesis and usually produce a favorable response to conservative treatment, whereas those resulting from tension forces can lead to displacement, instability, and possible fracture gap.

Table 29–5 Severity, location, and treatment of common stress fractures.
Clinical Findings

The runner presents with pain at a bony prominence, initially absent or minimal at the start of a run but becoming progressively worse with continued running. This is an important distinction between stress fracture and MTSS, as in the latter condition the pain is severe at the onset of the activity but improves as the activity progresses. The study of choice to evaluate for stress fracture is MRI. Plain radiographs have poor sensitivity because the periosteal reaction and cortical irregularities that occur with stress fracture may not be visible for several weeks.

Treatment

<table>
<thead>
<tr>
<th>Severity</th>
<th>Location</th>
<th>Treatment Suggestions</th>
</tr>
</thead>
<tbody>
<tr>
<td>High risk</td>
<td>Fifth metatarsal</td>
<td>Non-weight-bearing (NWB) immobilization for 6–8 weeks</td>
</tr>
<tr>
<td></td>
<td>Navicular bone</td>
<td><em>No cortical disruption</em>: NWB immobilization for 6 weeks</td>
</tr>
<tr>
<td></td>
<td>Proximal to middle third of tibial diaphysis</td>
<td><em>Cortical disruption</em>: surgical fixation</td>
</tr>
<tr>
<td></td>
<td>Femoral neck</td>
<td>Limited weight bearing or surgical fixation, depending on clinical scenario</td>
</tr>
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<td></td>
<td></td>
<td><em>Compression side</em>: NWB for 6 weeks</td>
</tr>
<tr>
<td></td>
<td></td>
<td><em>Tension side</em>: surgical fixation</td>
</tr>
<tr>
<td>Low risk</td>
<td>Calcaneus</td>
<td>Activity modification with allowance of pain-free progression of weight bearing and activity. Treatment and progression should be tailored to the individual athlete.</td>
</tr>
<tr>
<td></td>
<td>Second to fourth metatarsals</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Distal third of tibia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lateral malleolus</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pubic rami and sacrum</td>
<td></td>
</tr>
</tbody>
</table>
Current treatment protocols are based on the location of the stress fracture and whether it is high risk or low risk. Low-risk stress fractures can be treated conservatively by limiting activity based on the runner’s pain, thus requiring a tailored treatment. Essentially, the runner is able to participate in activities that are pain free and gradually progress to additional activities as tolerated. Treatment of high-risk stress fractures differs, depending on location. Compression-side femoral neck stress fractures require a period of non–weight bearing with crutches, whereas tension-side femoral neck stress fractures require surgical fixation. Anterior tibial stress fractures can be treated either operatively or nonoperatively. Surgery involves fixation with an intramedullary nail and immediate weight bearing and allows for RTP 5 months earlier than nonoperative management with non–weight bearing. Navicular and fifth metatarsal diaphyseal stress fractures are generally managed with a relatively prolonged period of immobilization and non–weight bearing in the range of 8–12 weeks. A low threshold for early surgical considerations is advised for fifth metatarsal stress fractures, with closed reduction and percutaneous screw fixation if there is lack of response to non–weight bearing or a desire for expedited return to sport.


5. Achilles Tendinopathy & Rupture

ESSENTIALS OF DIAGNOSIS

► Most often presents as midsubstance tenderness, swelling, and thickening of the tendon.
► Pain begins insidiously after activity, becoming progressively worse and more constant.
► A positive Thompson test is pathognomonic for Achilles tendon rupture.
General Considerations

Achilles tendinopathy tends to occur in runners when logging longer distances or adding hill workouts. Athletes predisposed to this condition tend to overpronate when planting the foot, with hindfoot valgus and decreased talocrural ROM.

Clinical Findings

The runner typically presents with insidious onset of pain and thickening of the Achilles tendon. A palpable nodule may be noted about 3 cm proximal to the insertion in runners with midsubstance tendinopathy. An insertional tendinopathy causes symptoms at the insertion of the Achilles onto the calcaneus and is often refractory to treatment.

Treatment

Treatment consists of a short period of rest, possibly with a walking boot for 2–4 weeks, a heel lift, and standard RICE treatment. Corticosteroid injections provide short-term but not long-term control of symptoms; they are associated with a high rate of tissue atrophy and thus should be avoided. The most effective treatment is an eccentric strengthening program, but the runner must be compliant in resting from running during the 8-week program, otherwise there is little likelihood of success. Surgical options lend a poor outcome and a longer recovery than an eccentric program (eg, McLauchlan).

The risk of Achilles tendon rupture is 30% if a runner continues to train or compete with a tendinopathy. Sprinters are at the highest risk for acute rupture, owing to sudden onset of an extreme force. An Achilles tendon rupture can be diagnosed clinically with a positive Thompson test and is treated surgically in the athletic population with an Achilles tendon repair. Recovery after surgery requires a prolonged course of rehabilitation for several months, but surgical outcomes are good to excellent.

6. Plantar Fasciitis

ESSENTIALS OF DIAGNOSIS

- Plantar foot pain with first steps of the morning, and tenderness over the calcaneal tuberosity.
- Initially pain improves during activity, but becomes more consistent with chronicity.
- Tight gastrocnemius and soleus muscles.

General Considerations

Plantar fasciitis is an overuse injury to the plantar fascia that produces microscopic and occasionally frank tearing at the calcaneal enthesis. More than one million people seek treatment annually for this condition, signifying that it affects more than just runners. Risk factors include pes planus deformity, overpronation, obesity, sudden increase in training program, and prolonged standing.

Clinical Findings

The runner reports severe pain for the first few steps in the morning. There is tenderness over the medial calcaneal tubercle and midfoot. Initially, the pain resolves with activity, but it becomes constant with weight bearing as the condition worsens. On focused examination, there is tenderness to palpation over the medial calcaneal tuberosity. There is often a lack of ROM at the ankle, particularly in dorsiflexion due to a tight heel cord. A pes planus deformity may be noted. Radiographs can be ordered and often show a heel spur, but this finding has not been shown to correlate with plantar fasciitis.

Treatment
Plantar fascitis is generally a self-limiting condition that improves after 1 year. Rest in a walking boot, frequent icing, and stretching of the heel cord and intrinsic foot muscles is started immediately. Formal physical therapy can be initiated, in which an eccentric tendon loading program is the most important aspect. ECSW therapy has come back into favor in recalcitrant cases of plantar fasciitis, as a means of promoting neovascularization and aiding in tissue repair. Platelet-rich plasma injections and prolotherapy (regenerative injection therapy) have been used in treatment, but to date there are no randomized controlled clinical trials to support their efficacy. Further research is underway in this area. Surgical treatment, consisting of plantar fasciotomy, is used for severe recalcitrant cases and produces satisfactory results in 80–85% of cases.

Goff J, Crawford R: Diagnosis and treatment of plantar fasciitis. Am Fam Physician 2011;84:676–682.

COMMON INJURIES IN CYCLING

1. Overuse Injuries in the Cyclist

ESSENTIALS OF DIAGNOSIS

- Proper bike size and positioning are the most important factors in preventing overuse injury.
- Slower cadence (< 80 rpm) places undue stress on the body and is an independent risk factor for overuse injury.
- The most common areas of pain are the knees, hips, low back, and neck.

Depending on the training regimen, a cyclist may log 25–35 weekly training hours or more on his or her bike. This alone places a burden on the body and predisposes the cyclist to overuse injury. However, with the correct bike position and following of basic rules, injury can be prevented.

The most common complaint from a cyclist is anterior knee pain, almost
always caused by a seat that is too low (causing excessive knee flexion) or a seat that is too far in the fore position (causing excessive anterior force on the knee). Adjusting the bike seat tends to relieve the symptoms, but if symptoms persist, the cyclist should be further evaluated for causes of anterior knee pain. Alternatively, a seat that is too high can cause hip pain or hamstring strain. Repetitive hip flexion can cause iliopsoas bursitis with anterior groin pain or greater trochanteric bursitis with lateral hip pain.

Cyclists often have problems with neck pain due to positioning in slight hyperextension, or low back pain from slight lumbar flexion. Both problems can be addressed by shortening the handle-bar reach or tilting the seat angle upward slightly. Stretching and strengthening exercises in the “on-bike” position should also be emphasized.

Ulnar neuropathy is often seen from compression of the deep palmar branch of the ulnar nerve, which can usually be prevented by frequent hand repositioning and use of padded gloves. The seat angle, height and firmness can cause compression neuropathy of the pudendal nerve, traumatic urethritis, and saddle sores. In the male cyclist, a pudendal neuropathy with numbness in the penis and scrotum warrants a break from cycling until symptoms resolve, so as not to cause impotence. Saddles sores tend to occur in novice riders, with symptoms usually resolving once a callous is formed, but painful fibrous lesions can develop over the area of pressure. Any of the above problems can be corrected by adjusting seat position and experimenting with different saddles to the individual cyclist’s comfort.


2. Traumatic Injuries in the Cyclist

- Bicycle accidents are a major cause of serious injury related to sports and recreation.
- Risk factors for sustaining injury include lack of helmet, motor vehicle...
involvement, an unsafe riding environment, and the male gender.

Most traumatic bike injuries are minor abrasions, lacerations, and contusions. The most common sites of fracture are the wrist and ribs. Up to 47% of cycling injuries involve head trauma, and head injury is responsible for the majority of cycling-related deaths. The use of a helmet decreases injury to the head and face by 85% when worn correctly. Other preventative measures include campaigns to promote helmet use and separate bicycle paths away from traffic.


COMMON INJURIES IN SWIMMING

1. Swimmer’s Shoulder

**ESSENTIALS OF DIAGNOSIS**

- First sign is usually a dropped elbow during the recovery phase of freestyle swimming.
- Signs of impingement are universally positive.
- Evidence of scapular winging suggests a long duration of symptoms.

**General Considerations**

“Swimmer’s shoulder” is a general term used to describe an overuse injury that results in shoulder pain in a competitive swimmer. Evaluation requires a thorough history, with particular emphasis on details of the swimmer’s training history. Relevant information includes how many years the athlete has been swimming, what strokes are used, how many days a week the athlete trains in the
pool, how many laps are swum done in a typical session, whether the intensity or duration of workouts has recently changed, and what dry land exercises and stretches are done. Changes in technique associated with shoulder pain include a dropped elbow during the recovery phase of the swimming stroke, asymmetric pull, early hand exit out of the water, and excessive body roll.

**Clinical Findings**

The vast majority of swimmers present for evaluation with diffuse pain, but report that the pain began at the anterior or anterolateral aspect of the shoulder. Stress is particularly concentrated on the anterior structures during humeral hyperextension at mid recovery, increasing the risk for labral or biceps injury. The scapulae should be examined for winging, and ROM of the shoulder, cervical spine, and thoracic spine should be evaluated, as well as rotator cuff strength.

The following special tests should be performed in swimmers: apprehension and relocation test and sulcus sign test, to assess for laxity; Neer or Hawkins test, or both, to assess for signs of impingement (see Shoulder Impingement, in Chapter 30); and O’Brien test (described earlier, under SLAP lesions), to assess for possible labral tear. There is little role for imaging studies in swimmer’s shoulder unless another diagnosis is suspected.

**Treatment**

Treatment begins with rest from swimming and involves comprehensive formal physical therapy in three phases (acute, recovery, and functional). Stretching of the anterior capsule should be avoided due to the increased risk in swimmers of permanent laxity from overstretching. Thoracic and cervical hypomobility should also be addressed.

The serratus anterior and subscapularis have been identified as the muscles most likely to fatigue in swimmers because of their continuous activity throughout the stroke cycle, and should therefore be emphasized. Swimming form should be evaluated at regular intervals to assess for errors that can lead to injury. The thumb-down hand entry is associated with excessive shoulder internal rotation and impingement and therefore is no longer a recommended technique. Two common mistakes during the pull-through phase that can provoke symptoms are crossing the arm past midline and inadequate body roll.
During the recovery phase of the swimming stroke, a low elbow can lead to increased drag, placing undue force on the shoulder complex. Finally, unilateral breathing patterns seen in novice and intermediate swimmers carry an increased risk of ipsilateral shoulder pain; all swimmers should therefore be encouraged to adopt a bilateral breathing pattern.

In the recovery phase of rehabilitation, progressive resistive exercises can be added for the rotator cuff and scapular stabilizers. The functional phase of rehabilitation focuses on RTP.


2. Low Back Injuries

**ESSENTIALS OF DIAGNOSIS**

- Injury stems from the repetitive nature of swimming in a position of lordosis.
- Any adolescent swimmer with low back pain that is worsened in extension should be evaluated for spondylolysis.

**General Considerations**

The repetitive nature of swimming can place increased stress on the low back. All four competitive strokes, but particularly the breast stroke, place the back in a lordotic position. Divers are also at increased risk for low back pain due to the excessive vertical and gravitational forces placed on the spine.

**Clinical Findings**

The typical findings seen in low back strain are noted, without neurologic abnormality. Any pain in the lumbosacral spine that is worsened with extension should be further evaluated with radiographs. If these are negative, a bone scan or CT scan is the gold standard. However, MRI is gaining popularity owing to
the excellent visualization of soft tissue anatomy, ability to see bony edema, and lack of radiation exposure.

## Treatment

The swimmer diagnosed with low back strain will likely respond favorably to a conservative course of physical therapy involving core stabilization and flexibility with a short period of rest from swimming. A diagnosis of spondylolysis requires a longer period of rest, typically of 6–12 weeks. Bracing may be needed if symptoms fail to resolve with rest alone, and therapy alone. A slow progression back to sport is recommended.


### 3. Knee Injury

#### ESSENTIALS OF DIAGNOSIS

- The second most common complaint in a swimmer, usually seen in breast stokers.
- The most common site of pain is the medial joint line.

#### General Considerations

Knee pain is a common complaint among swimmers; in one study, 75% of swimmers reported experiencing knee pain at least three times a year. Extrinsic risk factors for knee pain include length of time swimming, volume of training, and increased age. The highest association with knee pain is seen in competitive breast stroke swimmers, who show evidence of repetitive stress on the medial
collateral ligament (MCL), the medial patellar facet, and synovial plica.

Clinical Findings

An adequate swim and pain history must be obtained. Reproducible pain and thickening over the medial joint line suggests synovial plica syndrome. Valgus stress testing may reproduce pain with MCL involvement. Radiographs should be ordered only if clinically necessary. MRI is reserved for those who do not respond to conservative treatment.

Treatment

A painful knee in a swimmer often responds well to a period of rest and activity modification. Antiinflammatory medications may be helpful. For swimmers with PFPS, treatment is the same as that outlined earlier for runners. For any of the causes of medial knee pain, a course of formal physical therapy is recommended if rest alone fails to relieve the pain. For recalcitrant cases, surgery may be warranted. Prevention of injury is essential and centers on addressing common errors in breast stroke form, including excessive external rotation of the hip, leg abduction rather than adduction during the recovery phase of the stroke, and flexion of the hips and knees, rather than extension, during the recovery phase.


COMMON INJURIES IN ATHLETIC PERFORMING ARTISTS

This section highlights the most common injuries in a select group of performing artists: dancers, gymnasts, cheerleaders, and figure skaters (Table 29–6). These athletes are subject to unique physical and emotional stressors owing to the extremes placed on their bodies and therefore require a specialized approach to diagnosis and treatment. Other performing artists may also develop injuries in response to stresses associated with their particular art form (eg, rib injuries in singers, hand injuries in instrumentalists, etc); discussion of these latter injuries is beyond the scope of the chapter.
**Table 29–6** Most common injuries by location in athletic performing artists.

<table>
<thead>
<tr>
<th>Location</th>
<th>Bony Injury</th>
<th>Soft Tissue Injury</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foot and ankle</td>
<td>Sesamoiditis <strong>“Dancer’s fracture”</strong> (fifth metatarsal)</td>
<td>Ankle sprain</td>
</tr>
<tr>
<td></td>
<td>Bunion hallux valgus or rigidus</td>
<td>Plantar fasciitis</td>
</tr>
<tr>
<td></td>
<td>Metatarsalgia</td>
<td>Flexor hallucis longus tendinitis</td>
</tr>
<tr>
<td></td>
<td>Ankle impingement</td>
<td>Posterior tibial tendon dysfunction (PTTD)</td>
</tr>
<tr>
<td></td>
<td>Haglund’s deformity</td>
<td>Achilles tendinopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lace bite (extensor hallucis longus or extensor digitorum longus tenosynovitis)</td>
</tr>
<tr>
<td>Knee</td>
<td>Patellofemoral pain syndrome (PFPS)</td>
<td>Patellar tendinopathy</td>
</tr>
<tr>
<td></td>
<td>Plica syndrome</td>
<td>Anterior or medial cruciate ligament sprain</td>
</tr>
<tr>
<td></td>
<td>Meniscal tear</td>
<td>Osteoarthritis</td>
</tr>
<tr>
<td>Hip</td>
<td>Iliopsoas bursitis</td>
<td>Piriformis syndrome</td>
</tr>
<tr>
<td></td>
<td>Snapping hip syndrome</td>
<td>Labral injury</td>
</tr>
<tr>
<td>Back</td>
<td>Low back strain</td>
<td>Spondylolisthesis</td>
</tr>
<tr>
<td></td>
<td>Sacroiliac joint dysfunction</td>
<td>Spondylolysis</td>
</tr>
<tr>
<td></td>
<td>Discogenic back pain</td>
<td></td>
</tr>
<tr>
<td>Wrist</td>
<td>Distal radius stress injury</td>
<td>Avascular necrosis of the capitate</td>
</tr>
<tr>
<td></td>
<td>Dorsal impingement syndrome</td>
<td>Lunotriquetral impingement</td>
</tr>
<tr>
<td></td>
<td>Ulnar impaction syndrome</td>
<td>Ganglion cyst</td>
</tr>
<tr>
<td></td>
<td>Scaphoid impaction syndrome</td>
<td>Carpal instability</td>
</tr>
<tr>
<td></td>
<td>Scaphoid fracture</td>
<td></td>
</tr>
</tbody>
</table>
Knowledge of the mechanism of injury as it pertains to the performing artist is essential.

A high index of suspicion for underlying stress fractures, especially when any bony tenderness is present.

1. Tendinopathies

In no sport is the foot and ankle complex placed into such strenuous conditions as in dance, especially classical ballet. The demi-pointe and en-pointe positions that are central to the technique of the ballerina place tremendous load through the first MTP, as well as excessive strain on the tendons as they work to stabilize the foot–ankle complex in extremes of motion. The most commonly injured tendon is the flexor hallucis longus (FHL), leading to the condition known as “dancer’s tendinitis,” followed by the posterior tibialis and the peroneus longus. The FHL tendon lies in a fibro-osseous tunnel on the medial aspect of the ankle dorsal to the talus. The most common site of injury is at the posteromedial ankle during extremes of repetitive plantar flexion, such as occurs during frequent changes of position from plié to relevé.

Pain is reported in the posteromedial ankle and worsened with resisted toe flexion or passive extension. The dancer also experiences pain with moves involving forefoot weight bearing, such as in demi-pointe, or with push-off in jumps. On physical examination, pain is present diffusely at the posteromedial ankle and under the toe. Passive dorsiflexion of the toe aggravates the pain. If the dancer fails to obtain treatment in a timely manner, the condition can progress to hallux saltans, a triggering of the great toe due to nodule formation within the fibro-osseous tunnel. The posterior tibialis is injured in a similar manner, and both tendons are at increased risk for injury in dancers who overpronate, practice on a hard surface, or have a “dead” (ie, worn out) pointe shoe. The peroneus longus tendon acts a stabilizer in the foot and ankle, and is therefore easily injured when the demands of repetitive plantar flexion exceed
the tensile strength of the tendon. The dancer reports pain on the outer ankle that is worsened with passive inversion and resisted eversion. Achilles tendinopathy is also common in dancers and frequently leads to rupture of the tendon.

Treatment for these tendon injuries is similar and includes ice, avoidance of aggravating activities, evaluation of shoe wear, and physical therapy. Therapy should include manual mobilization of the subtalar joint, eccentric strengthening, and stretching of the appropriate tendon. In recalcitrant cases of FHL tendinopathy, surgery may be indicated. This generally entails decompression of the FHL with medial retinaculum release and debridement of any nodules.

2. Bony Injuries

The extreme ROMs and repetitive motion at the foot and ankle required in dance lay the framework for bony injuries: sesamoiditis, metatarsalgia, hallux valgus (bunion) or rigidus, and acute or stress fracture, to name a few.

A. Sesamoiditis—This painful inflammatory condition involves the medial or lateral sesamoid bones, or both, on the plantar surface of the great toe. Injury occurs from repetitive activities on the ball of the foot or with excessive heel wear. The condition is sometimes confused with FHL tendinitis but can be distinguished clinically during physical examination due to point bony tenderness. Imaging studies should be obtained to rule out fracture. Treatment consists of rest, use of a J-pad to offload the area, taping in slight plantar flexion, and use of a rigid-soled rocker-bottom shoe with steel shank insert. Surgery to remove the sesamoid is a last resort and can result in imbalance at the joint and eventual deviation.

B. Metatarsalgia—This is an imprecise term that describes pain at the metatarsals in the absence of other pathology. The general underlying cause is a synovitis of the MTP joint from excessive pressure, most often involving the second MTP. The dancer complains of forefoot pain worsened with push-off into pointe or demi-pointe. On examination, there is local tenderness. Metatarsal squeeze (squeezing all five metatarsals) may or may not cause pain, but a positive test should make the clinician suspicious for fracture. Radiographs and MRI may be warranted if a stress fracture is suspected. Treatment involves rest and padding proximal to the area of pain in order to offload pressure. In rare cases an MTP injection with corticosteroid is needed.

C. Hallux valgus or rigidus—Over time fixed deformities can occur in the feet
of dancers, most commonly in the form of bunion deformity (hallux valgus) and hallux rigidus. Hallux valgus is seen when the first metatarsal head deviates medially, producing a painful bump and causing the hallux to compensate with a lateral deviation. The dancer has pain with weight-bearing and push-off activities. Examination and radiographs reveal the diagnosis. Treatment consists of use of a toe spacer between the first and second toes to aid in alignment, padding over the prominent bone, and use of a custom orthotic to stabilize the medial column of the foot and slow the progression of the deformity. Once conservative measures fail to relieve discomfort, surgery is recommended.

Hallux rigidus results from early arthritic changes and bone spur formation and is characterized by restricted movements at the great toe, limiting a dancer’s ability to perform the full 90 degrees of MTP dorsiflexion needed for full demi-pointe. Consequently, the dancer goes into half demi-pointe or into a “sickle” position (ankle inversion during en-pointe). Treatment consists of ice, NSAIDs, taping the great toe in slight plantar flexion to prevent full demi-pointe, stretching in the demi-pointe position while non–weight bearing, and physical therapy to mobilize the joint.

D. Fractures—Fractures of the foot are not uncommon in performing artists. The classic “dancer’s fracture” is an acute fracture of the fifth metatarsal that results from improper landing on an inverted foot and is often seen at the end of a training session when the athlete is fatigued (Figure 29–8). More often, stress fractures occur, most commonly in the second and fifth metatarsals, fibula, navicular, calcaneus, and sesamoid bones. The cause of stress fractures in dancers is multifactorial and requires a thorough investigation by the clinician in order to prevent future fractures. Factors to consider include lack of rest, repetitive impacts, training methods, floor surfaces, shoe wear, biomechanical alignment, nutritional status, and hormonal factors. The need to maintain a thin body habitus for aesthetic purposes and the external pressures of the sport can lead to inadequate nutrition, in turn leading to altered bone mineral density and significantly increased risk for stress fracture. Any athlete who presents with point tenderness over a bone requires thorough workup for a stress fracture, starting with radiographs and moving on to either bone scan or MRI as warranted.
Treatment consists of an initial period of immobilization, and use of casting is suggested owing to the high rate of noncompliance to activity restrictions among dancers. The length of time for rest is tailored to the individual athlete based on the location of the fracture, as well as resolution of pain on bony palpation and with weight bearing. Return to training and activity should be a slow progression and monitored closely by the clinician, as most performing artists return too quickly without guidance.

3. Impingement Syndromes

Anterior or anterolateral impingement results from repetitive contact of the tibia
with the talus and causes anterior and slightly lateral ankle pain with activities that involve dorsiflexion. It is commonly seen in dancers who perform repetitive pliés and in gymnasts who land short when dismounting from apparatus. Posterior ankle impingement is caused by repetitive contact of the posterior tibia against a prominent posterior process of the talus or an os trigonum (an accessory bone present in 10% of the population). It is seen in ballet dancers as a result of hyper plantar flexion in pointe or demi-pointe.

Diagnosis is generally straightforward given the history (of the position causing symptoms) in conjunction with physical examination reproducing the pain (with either forced dorsiflexion or plantar flexion, respectively). In prolonged cases bone spurs and bony cysts may be seen on radiographs. Bony impingement may be seen if radiographs are taken in the dance position (plié or demi-pointe).

Treatment consists of a trial of rest and therapy for manual subtalar, talocrural, and midfoot joint mobilization. Athletes with anterior impingement may benefit from a ¼-inch heel. In a dancer, evaluation of the ballerina’s form is also useful, as those who force turnout or “sickle” the foot are at increased risk. Gymnasts can be assessed for a short landing, which causes the tibia to move anteriorly. Therapy in gymnasts should emphasize engaging the gluteus and hamstring muscles during landing and providing adequate bend at the knee without sacrificing style points.


**SPINAL INJURIES: SPONDYLOLYSIS & SPONDYLOLISTHESIS (see also Chapter 31)**

There is a fourfold increase in spondylolysis (fracture of the pars interarticularis) and spondylolisthesis (forward slippage of one vertebra on another) in athletic
performing artists when compared with the general population. It is principally
the hyperlordotic position adopted in performing artists that places them at risk,
commingled with heavy jumping and axial loads on landing and lifting.

The athlete typically presents with low back pain, worsened with lumbar
extension. A step-off deformity may be palpable in the presence of
spondylolisthesis. Any point tenderness on extension in this population warrants
radiographs. A pars defect or spondylolysis is seen on oblique films only a small
percentage of the time, so a negative result requires further investigation. Bone
scan, MRI, or CT scan can all be used, with CT scan being the most sensitive
and specific. However, we prefer to begin with an MRI in most circumstances
because of decreased radiation exposure and the ability to see bony edema.

An athlete with spondylolysis needs to rest from any activity that causes
lumbar extension and begin a neutral core strengthening program. If pain
continues with daily activity or if there is concern for compliance, a custom
lumbosacral orthosis can be used. A typical period of rest is 6 weeks but may be
longer based on clinical response. Return to activity should be graded and pain
free. Athletes with grade I and II spondylolisthesis are generally treated
symptomatically, but a symptomatic grade III typically requires surgical
stabilization.

D’Hemecourt P, Luke A: Sport-specific biomechanics of spinal injuries in
aesthetic athletes (dancers, gymnasts and figure skaters). Clin Sports Med
2012;31:397–408.

WRIST INJURIES

ESSENTIALS OF DIAGNOSIS

► Presentation is often delayed, increasing the likelihood of an advanced injury.
► In gymnasts, the three most common wrist diagnoses are distal radius stress
injury, dorsal impingement syndrome, and ulnar impaction syndrome.
1. **Distal Radius Stress Injury**

Wrist injuries are commonly seen in gymnasts and cheerleaders. Chronic repetitive compression combined with 90 degrees or greater of dorsiflexion and ulnar or radial deviation places a tremendous strain on the wrist structures. A stress injury to the distal radius is the most common diagnosis and is due to repetitive subthreshold injury with impaction and torsional forces. This injury predominantly affects female athletes aged 12–14 years, with increased risk in those practicing more than 35 hours weekly.

The athlete presents with palpable dorsal wrist pain that is worsened with extreme dorsiflexion. Grip strength may be diminished. A stage I injury is diagnosed clinically on the basis of negative radiographs, and the athlete can typically return to sport once the symptoms resolve. Stage II and III injuries show radiographic evidence of physial injury and require prolonged time off to allow healing and rehabilitation. If compliance with treatment recommendations is of concern, a cast can be applied. Once an athlete returns to sport, a Gibson brace may be used, which provides palmar padding and decreases radial load. In advanced stage III injuries, a positive ulnar variance occurs as the body attempts to redistribute the stresses on the radius; this can lead to ulnar impaction syndrome.

2. **Dorsal Impingement Syndrome**

Athletes with dorsal wrist pain may have an impingement syndrome, most often seen as thickening of the extensor retinaculum or synovitis of one or more of the wrist and digit extensor tendons. Those who participate in the vault and pommel horse are at highest risk. The hallmark of this injury is dorsal wrist pain, worsened with dorsiflexion of the wrist as well as with resisted digit extension. Radiographs may show osteophyte formation of the distal radius in chronic cases. If the impingement syndrome is diagnosed early enough, conservative treatment with rest, NSAIDs, and splINTing is typically sufficient. If this approach fails, corticosteroid injections may be of benefit. Recalcitrant cases may require surgical exploration; common interventions include retinaculum release, tendon debridement, and synovectomy.

3. **Ulnar Impaction Syndrome**

Also known as *ulnar abutment syndrome*, this injury is caused by a positive
ulnar variance and is the third most common wrist injury in gymnasts. Although a positive ulnar variance can be acquired, more often it is due to repetitive strain involving preferential ulnar loading on a pronated wrist, such as in the pommel horse event. The athlete presents with tenderness in the ulnar snuff box and with passive ulnar deviation of the wrist. Radiographs show a positive ulnar variance with possible cystic or sclerotic formation of the ulnar head. Varying types of soft tissue injury can also be incurred, such as tear of the triangular fibrocartilage complex or lunotriquetral ligament, which requires magnetic resonance arthrography for diagnosis if there is clinical concern. Treatment includes rest, NSAIDs, and corticosteroid injections. If these measures fail to relieve pain, surgery involving ulnar shortening via osteotomy or arthroscopic wafer resection can be performed.

Acromioclavicular Joint Sprain

General Considerations
Acromioclavicular joint (ACJ) sprains are common among young adults who engage in sporting activities, and usually result from falling directly on the acromion or onto an outstretched hand. The forces involved in such a fall drive the acromion inferiorly and superiorly, respectively.

Pathogenesis
The ACJ is a synovial joint located between the distal clavicle and the acromion. The posterolaterally facing medial facet of the distal clavicle articulates with the anteromedially facing acromion. Static stabilizers of the ACJ are the
acromioclavicular (AC) and coracoclavicular (CC) ligaments. The superior, inferior, anterior, and posterior AC ligaments become part of the AC joint capsule. The CC ligaments, trapezoid and conoid, extend from the coracoid process to the inferior surface of the clavicle. The trapezoid is lateral and has a wide insertion on the clavicle, whereas the conoid is medial and has a thin insertion on the posterior tubercle of the clavicle. Dynamic stabilizers of the ACJ are the deltoid and trapezius, also known as the deltotrapezial complex. The deltoid prevents superior and posterior migration of the clavicle. Contraction of the trapezius compresses the ACJ.

Five to 8 degrees of motion at the ACJ facilitates clavicular rotation along the long axis and elevation and retraction of the distal clavicle. This motion aids in maintaining the subacromial space during terminal arm elevation.

Clinical Findings

ACJ sprain or separation can be diagnosed clinically by localized pain and deformity. The horizontal adduction test compresses the ACJ and causes pain in the presence of injury. The Zanca view radiograph allows for optimal visualization of the joint. ACJ sprains and separations are graded on a scale from I to VI, using the Rockwood classification.

• **Type I** injury results in a sprain of the capsule and AC ligaments, but the AC and CC ligaments remain intact. No clavicular instability can be detected on examination, and radiographs are normal.

• **Type II** injury results in rupture of the capsule and AC ligaments, but the CC ligaments remain intact. The clavicle is unstable under direct stress. Stress radiographic views are negative; however, the lateral clavicle can be slightly elevated.

• **Type III** injury results in complete rupture of the AC and CC ligaments without disruption of the deltotrapezial fascia. On examination, the lateral clavicle appears elevated, with the acromion depressed. The clavicle is unstable vertically and horizontally. ACJ separation is evident on stress radiographs.

• **Type IV** injury results in rupture of the AC and CC ligaments, with posterior displacement of the clavicle into the trapezius. This is evident both clinically and radiographically on axillary view.

• **Type V** injury results in a type III injury with disruption of the deltotrapezial
fascia. The lateral clavicle is elevated and the scapula is downwardly displaced. Radiographs demonstrate a 100–300% increase in the clavicle-to-acromion distance (CC distance).

• **Type VI** injury results in inferior displacement of the clavicle into the subacromial or subcoracoid spaces. This usually occurs in the setting of severe trauma and accompanies other injuries.

### Complications

Patients with type II or III sprains can develop subacromial impingement, rotator cuff pathology, and ACJ osteoarthritis from scapular dyskinesis. If symptoms persist more than 6 months after injury, then corticosteroid injections or surgical repair may be needed.

### Treatment

**A. Type I and II Sprains**

The acute phase of treatment for type I and II sprains begins at the time of injury and continues through 3 weeks. Therapies include pain-free range of motion (ROM) exercises, soft tissue and scapular mobilization, and isometric strengthening of scapular stabilizers using closed kinetic chain exercises without arm elevation (ie, internal and external rotation exercises at 0 degrees of abduction). Trunk and leg strengthening begins using a wobble board. The patient may progress to arm elevation as tolerated, wear an arm sling for the first 3–10 days, and use analgesics for pain. The recovery phase occurs over the next 3–8 weeks postinjury. Therapies during this period include increasing loads with isotonic closed kinetic chain exercises. The patient may begin active arm elevations. The maintenance phase begins when the patient has full, pain-free ROM and approximately 75% strength compared with the contralateral side. Approximately 12% of young athletes with low-grade sprains undergo surgical repair because of persistent symptoms.

**B. Type III Sprain**

The acute phase of treatment for type III sprains begins with shoulder rest in a sling for 7–14 days. The patient may slowly incorporate active, assisted ROM isometric exercises with internal and external rotation at 0 degrees, shoulder
shrugs, and pendulum exercises. After 7–14 days, the patient may remove the sling, beginning the recovery phase of treatment, which includes active ROM isotonic exercises with flexion greater than 160 degrees and abduction to 90 degrees. The maintenance phase begins when the patient has full, painless ROM and approximately 75% strength compared with the contralateral side. Patients with type III sprains may undergo surgical repair if pain or instability persist. Patients with type III separations have alterations in scapular motor planning and scapular dyskinesis that must be addressed through physical and occupational therapy.

C. Type IV–VI Sprains

Type IV–VI sprains are severe injuries that often occur in combination with other traumatic injuries. For patients with such injuries, noninvasive treatment measures are generally insufficient. Consequently, approximately 75% of young athletes with high-grade sprains undergo CC ligament reconstruction. Most patients, nonsurgical and surgical, are capable of returning to full function with adequate rehabilitation.


SHOULDER IMPINGEMENT

1. External Impingement of the Shoulder

General Considerations

Primary external impingement of the shoulder is caused by subacromial or subcoracoid impingement of the rotator cuff. Secondary impingement of the shoulder is caused by abnormal glenohumeral and scapulothoracic motion. Static stabilizers of the shoulder include the glenoid labrum and glenohumeral ligaments. Dynamic stabilizers of the shoulder include the rotator cuff muscles
(supraspinatus, infraspinatus, teres minor, and subscapularis). Typically the rotator cuff tendons run along the longitudinal axis of the muscle; however, there are medial and deep components of the tendons that course transversely. They are known as the rotator cable. The rotator interval is a gap in the rotator cuff through which the long head of the biceps tendon passes.

The undersurface of the acromion can be a cause of impingement. Grade I acromion impingement is flat, grade II is concave, and grade III is hooked.

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**Pathogenesis**

The deltoid and supraspinatus muscles provide superior translational forces to the humeral head, while the infraspinatus, teres minor, and subscapularis provide inferior translational forces. Normally, these opposing forces are balanced. Supraspinatus impingement syndrome occurs when there are unopposed superior translational forces of the humeral head in 30–60 degrees of shoulder abduction.

Partial-thickness tears of the rotator cuff tendons decrease the overall power of the shoulder musculature. This leads to superior migration of the humerus under the acromion, which then further impinges the rotator cuff. The impingement typically occurs in the tendon’s hypovascular zone, which is located 2–6 cm proximal to the insertion, and ultimately leads to full-thickness tears.

In younger patients, supraspinatus tendon degeneration may be induced by repetitive overhead activity, which causes tendon thickening and erosion under the coracoacromial ligament. Secondary impingement can occur in association with humeral instability caused by microtrauma to the static stabilizers of the glenohumeral joint. The weakness of the static stabilizers leads to increased demand on the dynamic stabilizers, causing muscle fatigue. Subluxation of the humeral head occurs anteriorly and superiorly, with impingement on the coracoacromial arch. Upward and anterior scapular tilting can cause increased subacromial contact with the greater tuberosity.

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**Clinical Findings**

**A. Symptoms and Signs**

The clinical examination finding most often used to diagnose impingement is resisted initial shoulder abduction causing pain in the deltoid muscle. A positive
finding (ie, pain or resisted motion) in any of the following maneuvers is also indicative of impingement.

1. **Neer sign**—The examiner passively induces 160 degrees of shoulder flexion while stabilizing the scapula (Figure 30-1).

![Figure 30-1](image)

▲ **Figure 30–1** Neer sign can detect shoulder impingement.

2. **Hawkins sign**—The examiner passively induces 90 degrees of shoulder flexion with maximal internal rotation (Figure 30-2).
3. **Empty can test**—The examiner resists shoulder abduction with the patient’s shoulder flexed to 90 degrees in the plane of the scapula with the forearms maximally pronated.

4. **Speed’s test**—With the patient’s shoulder flexed to 90 degrees, the examiner resists shoulder flexion while the patient’s elbow is extended and forearm is supinated (Figure 30-3).
**Figure 30–3** Speed’s Test can be indicative of bicipital tendonitis.

**B. Imaging Studies**

Anteroposterior and outlet-view radiographs are useful in diagnosis. The height of the subacromial space can range from 10 to 15 mm; narrowing of the space to 6–7 mm may indicate impingement. Cross-sectional views using magnetic resonance imaging (MRI) with arthrography and ultrasonography can aid in diagnosis. MRI demonstrates both bony and soft tissue abnormalities. Magnetic resonance arthrography is the gold standard in diagnosing full-thickness tears.

Partial-thickness tears should be graded by the percentage of tendon thickness involved (grade I, < 25%; grade II, 25–50%; grade III, 50–75%). Muscle atrophy with fatty infiltration can be seen on computed tomography (CT) scans (**Figure 30–4**). The diagnosis of supraspinatus tendinosis secondary to subacromial impingement and bicipital tendinosis can be confirmed using ultrasonography. (See the discussion of musculoskeletal ultrasound in Chapter 39 for more details.) Up to 34% of patients with full-thickness tears are asymptomatic.

**Figure 30–4** Coronal oblique fat-saturated T1-weighted magnetic resonance
arthrogram of a patient with a partial-thickness supraspinatus tear (*arrows*).

## Differential Diagnosis

The differential diagnosis of external shoulder impingement includes osteoarthritis of the acromioclavicular joint and subacromial subdeltoid bursitis.

## Treatment

Treatment includes nonsteroidal antiinflammatory drugs (NSAIDs) and physical therapy. The latter focuses on strengthening the inferior rotator cuff muscles, excluding the deltoid, which as previously noted is thought to cause the upward humeral subluxation leading to subacromial impingement. Subacromial subdeltoid steroid injection may be beneficial in relieving pain in order to allow greater gains with therapy; however, a significant rotator cuff tear should first be excluded, as an intratendinous injection can cause tendon rupture. Full-thickness tears may require surgical repair.


### 2. Internal Impingement of the Shoulder (SLAP Lesion)

Internal impingement of the shoulder occurs when soft tissues, such as the joint capsule and labrum, become impinged between the surface articulations of the humerus and the glenoid. Repetitive shoulder abduction and external rotation leads to posterosuperior impingement of the labrum and undersurface of the rotator cuff. This can manifest as labral fraying and partial tears of the rotator cuff. Excessive external rotation loosens the inferior glenohumeral ligament, promoting anterior translation and posterosuperior impingement. Glenohumeral rotation deficit (GIRD) may also be the cause of internal impingement. Repetitive throwing causes a posteroinferior joint capsule contracture, which creates “pseudolaxity” with increased external rotation and decreased internal rotation. This imbalance predisposes the individual to a lesion of the superior
glenoid labrum, also known as a superior labrum anterior–posterior (SLAP) lesion. SLAP lesions are most often reported in throwing athletes, and are described in detail in Chapter 29.

The throwing athlete with internal impingement of the shoulder typically reports posterior shoulder pain in the late-cocking phase of pitching, or when the arm is abducted and externally rotated. The affected arm may feel weak and unstable after throwing. Signs of external impingement are usually negative (eg, Hawkins and Neer signs, described earlier). The following radiographic views are used to detect internal impingement: anterior–posterior views (in internal and external rotation), West point, Stryker notch, axillary, and scapular Y views. Common radiographic signs of internal impingement are Bennett lesions (exostosis or calcification in the region of the posterior–inferior glenoid rim), sclerotic changes in the greater tuberosity, an osteochondral lesion or cystic changes in the posterior humeral head, and rounding or remodeling of the posterior glenoid rim. MRI remains the gold standard as the aforementioned radiographic findings are often difficult to detect. Conservative treatment includes shoulder rest, antiinflammatory medications, and cryotherapy. Rest should continue for 4–6 weeks, or until the pain has resolved. Surgical treatment ranges from debridement of the frayed labrum or rotator cuff to complete repair of rotator cuff tendons having 50–75% tears.

Most patients are able to meet goals for recovery after 3 months of physical therapy, and return to work or sport within 6 months.

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**ELBOW DISLOCATIONS**

**General Considerations**

The elbow is the most commonly dislocated joint in the pediatric population and is second to the shoulder in the adult population. Elbow dislocation is more common in males than females and is usually associated with sports such as gymnastics, wrestling, basketball, and football. The most common mechanism is a fall on an outstretched hand, causing axial compression, external rotation of the
forearm, or supination, and a valgus moment.

## Pathogenesis

O’Driscoll described a “ring of instability” involving progressive disruption of the elbow, differentiating three stages of disruption. In stage 1, the lateral portion of the ulnar collateral ligament is disrupted, causing posterolateral rotatory subluxation with spontaneous reduction. Stage 2 occurs with continued force, causing incomplete subluxation that leaves the coronoid perched on the trochlea. Stage 3A includes complete disruption of the soft tissues and the posterior part of the medial collateral ligament. Stage 3B includes complete disruption of the medial collateral ligament, causing varus, valgus, and rotatory instability.

Elbow dislocations can likewise be classified into three categories: posterior, divergent, and anterior. Posterior elbow dislocations are subdivided into posterolateral, posteromedial, and pure lateral dislocations. Divergent dislocations are associated with high-impact trauma and radioulnar displacement with damage to the interosseous membrane. Anterior dislocations are rare and are seen in younger patients.

## Clinical Findings

Patients with an acute elbow dislocation present with severe pain and a deformed upper limb. There is usually a history of trauma to the arm. Before attempting reduction maneuvers, a neurovascular assessment should be performed. The wrist and shoulder should also be examined for coinciding injuries, and anteroposterior and lateral radiographs obtained.

## Complications

Elbow dislocations may be associated with radial head and neck fractures, avulsion fractures of the medial and lateral epicondyles, and coronoid fractures. Neurovascular complications include brachial artery injuries, median nerve entrapment, and compartment syndrome. The median nerve may be displaced posteriorly and become entrapped by an avulsion fracture of the medial epicondyle. The tension placed on the median nerve may “notch” the epicondylar flare of the humerus, producing a Matev sign on radiographs (sclerotic edges of a bony tunnel at the medial epicondylar ridge where the
median nerve has been entrapped).

**Treatment**

Rapid closed reduction is preferred before soft tissue swelling occurs. Conscious sedation may be necessary to allow muscle relaxation. Closed reduction is performed in prone traction with the elbow extended, using the physician’s thumb to guide the coronoid around the trochlear. Irreducible elbow dislocations may be caused by radial head entrapment within the soft tissues. Postreduction, the elbow should be evaluated for instability. Posterolateral rotatory instability is best assessed using the pivot–shift test, which is positive when a “clunk” is heard or felt as the radius and ulna reduce on the humerus. Radiographs should be obtained postreduction. Widening of the joint space may indicate entrapped intraarticular osteochondral fragments. Surgical intervention is required for unstable reductions, fractures, compartment syndrome, and open dislocations.

Rehabilitation with early, supervised ROM exercise has been shown to be most beneficial. Static flexion and extension splints may be used to aid patients in regaining ROM. Patients with “perched” dislocations have less severe injuries and can expect a full and rapid recovery. Those with more severe injuries may not recover complete ROM of the elbow, lacking full extension. With inadequate bone alignment there is an increased risk for arthritis of the elbow joint.


**OSTEOCHONDRITIS DISSECANS OF THE CAPITELLUM**

**General Considerations**

Osteochondritis dissecans of the capitellum is most commonly seen in teenage boys who participate in sports activities that involve repetitive overhand throwing. Contributing factors may be repetitive valgus stress to the elbow, as in throwing, and immature articular cartilage over the capitellum. These cause local injury to subchondral bone and can lead to avascular necrosis and subchondral osseous changes affecting the humeral capitellum of the dominant hand.
Clinical Findings

The typical presentation is a 10–15-year old male pitcher with lateral elbow pain and swelling. Patients with stable osteochondritis dissecans present with normal elbow ROM. Radiographs reveal flattening of the capitellum with articular defects; loose bodies may also be present. Three radiographic stages are differentiated. In stage 1, a cystic shadow is visible at the capitellum. In stage 2, the lesion and subchondral bone separate. In stage 3, loose bodies are present (Figure 30–5). The gold standard for diagnosis is MRI, which can aid in staging. CT scans are helpful in detecting loose bodies.

▲ Figure 30–5 Three-dimensional computed tomography scan of the left elbow, showing a loose body in the anterior compartment and a lesion characteristic of osteochondritis dissecans in the capitellum.
Differential Diagnosis

Osteochondritis dissecans of the capitellum should be distinguished from osteochondrosis of the capitellum, which is characterized by capitellar epiphyseal necrosis, regeneration, and calcification in children from 7 to 12 years of age.

Treatment & Prognosis

Stable lesions are treated with avoidance of repetitive stress, muscle strengthening, and occasionally splinting. Unstable lesions require surgical intervention, including debridement, removal of loose bodies, and, potentially, internal fixation of larger fragments or autologous chondrocyte graft transplantation. Approximately 90% of stage 1 lesions and 52% of stage 2 lesions heal with conservative management consisting of avoidance of heavy elbow use on the affected side for 6 months.


KIENBÖCK DISEASE

General Considerations

The lunate is the central bone in the proximal carpal row, articulating with the radius, triangular fibrocartilage complex, hamate, and capitate. Multiple etiologic factors can contribute to osteonecrosis of the lunate, also known as Kienböck disease. Mechanical extrinsic factors include a short or long ulna, causing uneven distribution of axial forces; altered radial head inclination; and repetitive trauma. Mechanical intrinsic factors may be related to variations in the shape of the lunate. Vascular factors include interruption of the dorsal and
palmar arterial supply; decreased venous outflow; and transient synovitis, compromising vascular supply. Sickle cell disease and thrombocytosis may also compromise vascular supply. Low-grade osteomyelitis has also been linked with this condition.

Clinical Findings

Osteonecrosis of the lunate is commonly seen in men between 20 and 40 years of age. Patients present with pain and weakness of the affected wrist, but without a history of trauma. Findings are usually unilateral and exacerbated by wrist extension and axial loading. Physical examination reveals dorsal wrist swelling and tenderness over the lunate. Although forearm pronation and supination is preserved, wrist flexion and extension is limited. Four stages of disease are differentiated: stage 1 is similar to wrist sprain, with minimal MRI abnormalities; stage 2 is characterized by lunate hyperdensity that is visible on MRI; stage 3A, by lunate collapse with carpal stability; stage 3B, by lunate collapse with carpal instability; and stage 4, by disruption of the surrounding bones (Figure 30–6).
Radiograph of the wrist of a patient with Kienböck disease, revealing articular collapse and rotatory deformity of the scaphoid.

**Treatment**

Treatment varies according to the stage of the disease. Patients with stage 1 disease are generally treated with cast immobilization for 3 months. Those with stage 2 or 3A disease, but ulnar-negative variance, benefit from radial shortening, ulnar lengthening, and capitate shortening. Stage 2 or 3A ulnar-positive variance requires more complex surgical procedures, including vascularized bone graft and external fixation, radial wedge osteotomy, and
capitate shortening. In stage 3B disease (lunate collapse with carpal instability), scaphocapitate or scaphotrapezium–trapezoid fusion or proximal row carpectomy are required. Treatment options for patients with stage 4 disease include wrist arthrodesis, arthroplasty, proximal row carpectomy, and wrist denervation. Most patients experience pain relief and increased ROM with revascularization; however, nearly one quarter show disease progression.


LOWER EXTREMITY INJURIES

HIP OSTEOARTHRITIS

General Considerations

Osteoarthritis of the hip is the most common pathologic condition of the hip. It is a common complaint in older adults but may also affect younger patients. The prevalence is about 3.1% in the general population. Symptoms are bilateral in about 42% of patients, and genetics appear to play a role in disease progression.

Pathogenesis

Osteoarthritis occurs when cartilage breakdown leads to release of proinflammatory cytokines, matrix metalloproteinase, and prostaglandins. The inflammatory response promotes subchondral bone remodeling, resorption, neovascularization of synovial fluid, and calcification of the joint cartilage. Over time, these processes lead to the formation of osteophytes, joint space narrowing, and bone sclerosis. Risk factors for premature development of hip osteoarthritis
include abnormal hip joint architecture (eg, of the femoral head or acetabulum) and alignment.

Clinical Findings

A. Symptoms and Signs

Hip pain is usually associated with decreased ROM, made worse with activity, and relieved with rest. Patients with hip osteoarthritis walk with an antalgic gait, characterized as decreased single-limb stance on the painful limb, with short stride length on the contralateral limb. ROM should be evaluated in all planes, including flexion, extension, adduction, abduction, and internal and external rotation. Examination maneuvers should include the FABER test (performed by hip flexion, abduction, and external rotation); a positive test produces groin pain suggestive of intraarticular hip pain, such as hip osteoarthritis.

B. Imaging Studies

Plain radiographs are usually the first diagnostic imaging study ordered (Figure 30–7). The shortest distance on the radiograph between the femoral head margin and the acetabulum can be used as a measure of the severity of hip osteoarthritis. This is referred to as the minimal joint space (MJS) and is measured medially, laterally, superiorly, and axially. A study by Chu and colleagues found a weak association between joint space width and symptoms.
The Kellgren–Lawrence Grading Scale (KLGS) is another system used to grade hip osteoarthritis. The KLGS takes into account the presence of osteophytes, subchondral sclerosis, and cysts in grading osteoarthritis. Grade I is characterized by possible osteophytes; grade II by small osteophytes and possible narrowing of the joint; grade III by multiple, moderately sized osteophytes, definite joint space narrowing, some sclerotic areas, and possible deformation of bone end; and grade IV by multiple large osteophytes, severe joint space narrowing, marked sclerosis, and definite bony end deformity.

MRI is not necessary but can be used to rule out other conditions in the differential diagnosis of hip pain, including avascular necrosis, labral tear, and fracture. There is a poor relationship between radiographic findings and the need for surgical management.

**Differential Diagnosis**

The differential diagnosis of hip osteoarthritis includes the conditions previously stated above, as well as low back pain, lumbar radiculopathy, sacroiliac joint disorder, meralgia paresthetica, and greater trochanteric pain syndrome.
Complications

Patients with progressive osteoarthritis develop joint stiffness, chronic joint pain, and compensatory postural changes that can lead to kyphosis, lumbar radiculopathy, and low back pain. Synovitis and fracture about the hip joint are other causes of concern.

Treatment

A. Conservative Measures

1. Exercise and weight loss—Several nonpharmacologic modalities for treatment of hip osteoarthritis are strongly supported in the literature. These include aerobic, aquatic, and resistance exercises and weight loss. An exercise regimen focusing on ROM, stretching, strengthening, and aerobic conditioning is recommended. Water-based exercises for hip osteoarthritis can decrease pain and increase function in the short term to up to 1 year. Many patients with hip osteoarthritis tolerate cycling and aquatic activities better than high-impact exercises such as stair climbing. However, even with the use of the preceding therapies, a Cochrane review found only a small treatment effect of exercise on hip osteoarthritis pain.

Weight loss is important in the treatment of hip osteoarthritis because body weight is an independent risk factor for the condition. Three to five times the body weight is exerted on the hip while at rest. This increases to eightfold with jogging.

2. Heat and cold therapy, assistive devices, and orthoses—Modalities such as thermotherapy and cryotherapy can be utilized. Additionally, the use of a cane on the contralateral hand will decrease the amount of the force on the arthritic hip. A shoe lift is helpful in correcting leg-length discrepancy.

B. Pharmacotherapy

Pharmacologic modalities include acetaminophen, NSAIDs, tramadol, and intraarticular corticosteroid injections. Acetaminophen is the initial medication choice in the treatment for hip osteoarthritis (maximum daily dose, 4 g). If the patient’s symptoms are not relieved with acetaminophen, NSAIDs should be considered.

Fluoroscopically guided intraarticular cortisone hip injection has been shown
to decrease pain and improve function. Limited studies have shown the use of hyaluronic acid to be superior to placebo; however, there is no evidence supporting hyaluronic acid injection as more effective than corticosteroid or other conservative therapies in the treatment of hip osteoarthritis.

C. Surgical Treatment

Surgical management of end-stage osteoarthritis of the hip includes a total hip replacement with implantation of a prosthetic femoral head and acetabulum. While surgical replacement is usually well tolerated, complications can include infection, prosthetic loosening, and dislocation. Intense rehabilitation is required postoperatively to regain function and strength. (See Chapter 33 for further discussion.)


FEMORAL ACETABULAR IMPINGEMENT

General Considerations
Femoral acetabular impingement (FAI) occurs when there is noncongruent articulation of the acetabular rim, femoral head, and neck junction, or both. Osseous deformity leads to degenerative arthritis and injuries to the acetabular labrum and cartilage. FAI is differentiated according to the mechanism of deformity into cam-type, pincer-type, or a combination of both. The cam-type (from the Dutch word *cam*, meaning “cog”) results in a nonspherical femoral head; in the pincer-type, there is overcoverage of the femoral head by the anterior acetabulum.

**Pathogenesis**

FAI is usually caused by repetitive microtrauma. The cam-type deformity results in an abnormal head and neck junction, with increased waist radius. The anterior femoral neck loses its concavity and develops a “bump” that impinges on the anterosuperior labrum. In pincer-type FAI, excessive acetabular coverage extends from the anterior acetabular rim prominence over the posterior rim. This overcoverage causes the anterior wall to be displaced in a more lateral direction compared with the posterior wall.

**Clinical Findings**

**A. Symptoms and Signs**

Pain associated with FAI is usually insidious and becomes more severe with time. It is described as hip stiffness, decreased ROM, or pain with weight-bearing activities. It is often associated with groin pain, but lateral thigh pain has also been described. Associated symptoms include clicking, catching, and locking. The pain is worsened with prolonged sitting, stair climbing, putting on shoes, and getting out of a car. On examination, patients have decreased ROM compared with the contralateral limb or pain with the hip in flexion and internal rotation. The classic maneuver for evaluating FAI is the FADDIR test, in which the hip is placed in flexion, adduction, and internal rotation.

**B. Imaging Studies**

Plain radiographs, especially in anteroposterior and true lateral views, typically show the cross-over sign, an abnormal finding that is characteristic of FAI (Figure 30–8). Measurement of the α angle can help refine the diagnosis. The
α angle is the angle that exists between lines drawn from the neck–shaft axis and head center to the aspheric point of the femoral head. An angle greater than 55 degrees is indicative of a cam lesion.

▲ Figure 30–8 Radiograph of the hip in a patient with femoroacetabular impingement (FAI) showing the classic cross-over sign in which the anterior and posterior walls of the acetabulum overlap (arrows).

MRI can be performed to evaluate the labrum and articular cartilage. Magnetic resonance arthrography is the most accurate study for diagnosing labral tears and osseous abnormalities.

► Differential Diagnosis

Numerous conditions can produce pain in the hip, including muscular, ligamentous, tendinous, and bursal inflammation around the hip. Pain radiation from the lumbar spine can also masquerade as hip pain. Patients should be evaluated for stress fractures, as well as inflammatory and infections processes.
Complications

If left untreated, FAI can progress, resulting in hip osteoarthritis, as well as chondral and labral damage.

Treatment

Conservative treatment should be attempted initially, including the use of acetaminophen, NSAIDs, and activity modification. Fluoroscopically guided intraarticular injection with local anesthetic and corticosteroid can be diagnostic and therapeutic. Patients should undergo physical therapy to improve ROM, with emphasis on strengthening the pelvic girdle and restoring pelvic muscle balance. Conservative measures are sometimes ineffective because the underlying problem is mechanical in nature, requiring surgical correction. Patients with FAI who have advanced osteoarthritis have a poor prognosis without joint replacement.


AVASCULAR NECROSIS OF THE FEMORAL HEAD

General Considerations

Avascular necrosis (AVN) of the femoral head develops when the vascular supply to the femoral head is disrupted. The resulting areas of necrotic trabecular bone and bone marrow extend to the subchondral plate. Risk factors for AVN include hip dislocation, femoral neck fracture, history of corticosteroid use, alcohol abuse, sickle cell disease, rheumatoid arthritis, lupus erythematosus, chemotherapy, and radiation agents. Corticosteroid use as a risk factor for AVN
is related to the strength of the dose and chronicity.

Clinical Findings

A. Symptoms and Signs
Patients usually describe pain that is dull, achy, and throbbing in nature in the groin, buttocks, or lateral hip. On examination, there is decreased active and passive ROM of the hip. Limitations of internal rotation are common in these patients. Patients tend to walk with an abnormal antalgic or Trendelenburg gait. The Trendelenburg sign is positive when weight bearing on the affected side results in contralateral hip drop.

B. Imaging Studies
Anteroposterior and frog’s leg–lateral views of the hip should be obtained. In the early stages of AVN, radiographic studies will show patchy areas of sclerosis and lucency. As the disease progress, a sclerotic area beneath the articular surface, indicative of a subchondral fracture, may be noted. This is termed a crescent sign. As AVN worsens, degenerative changes occur on the acetabulum (Figure 30–9).

▲ Figure 30–9 Bilateral avascular necrosis of the hip (arrows indicate areas of necrotic destruction).

The Ficat classification, which relies on a combination of radiologic and clinical findings, can be used to grade the degree of osteonecrosis. In grade 1, radiographic features are absent but the patient has some clinical symptoms.
Grade 2 is characterized by diffuse sclerosis and cyst development, with additional clinical manifestations. The transition from grade 2 to grade 3 reveals the classic crescent sign. In grade 3 there is a normal joint space with broken contour of the head sequestrum. Grade 4 is characterized by decreased joint space and collapse of the femoral head.

MRI is quite sensitive to AVN and likely will reveal pathology before plain films. Typically, a focal serpiginous low signal line with fatty center is seen with diffuse edema and possible osteochondral fragmentation. Bone scan is sensitive as well, often revealing a cold area with uptake.

### Treatment

The treatment of AVN encompasses both conservative (joint protective) and surgical approaches. Patients may be managed initially with conservative measures, including avoidance of weight-bearing activities. Recently alendronate, statins, and enoxaparin have each been shown to prevent the collapse of the femoral head. Mont and colleagues, in an older study, reported a 22% success rate using conservative treatment consisting of limited weight bearing and analgesics. Nonoperative management of AVN of the hip has largely been unsuccessful in halting progression of the disease and the majority of patients require surgical treatment. Surgical interventions include core decompression, bone grafting, osteotomy, resurfacing arthroplasty, total hip arthroplasty, and bipolar arthroplasty.


### SLIPPED CAPITAL FEMORAL EPIPHYSIS

#### General Considerations
A slipped capital femoral epiphysis (SCFE) occurs when the femoral head displaces, most commonly posteriorly, on the femoral neck at the level of the physis. It is the most common adolescent hip disorder, with an incidence of 10.8 cases per 100,000 children, and typically occurs bilaterally. The rate is 2.5–4 times higher in white compared with nonwhite children.

Slippage of the femoral head is caused by weakness of and mechanical stress across the growth plate. Panhypopituitarism, hypogonadism, hypothyroidism, and renal osteodystrophy have all been implicated as contributing factors. Mechanical factors such as retroversion of the femur, increased physial obliquity, and obesity place abnormal stress on the physis. About 90% of patients with SCFE are able to bear weight on the affected leg.

Clinical Findings

A. Symptoms and Signs

Patients usually present with an atraumatic painless limp. When the limp is associated with pain, they complain of knee, hip, groin, or thigh pain. The most important physical finding is limited internal rotation. The affected leg is often held in external rotation and restricted internal rotation. External rotation is noted in the involved hip when the hip is flexed to 90 degrees.

B. Imaging Studies

Plain radiographs are usually effective in diagnosing SCFE (Figures 30–10 and 30–11). Wilson’s radiologic classification of SCFE is based on degree of slippage. Slippage of less than one third is considered mild, one third to one half is considered moderate, and greater than one half is considered severe.
Figure 30–10 Slipped capital femoral epiphysis. A: Klein’s lines, revealing a left slipped epiphysis. B: Slipped epiphysis with widening of the growth plate.
Figure 30–11 Slipped capital femoral epiphysis of the left hip that is moderately displaced and unstable.

The Southwick slip angle is another method used to differentiate severity of SCFE; it measures the epiphyseseal–shaft angle. Mild slippage using this method is defined as less than 30 degrees; moderate, between 30 and 50 degrees; and severe, greater than 50 degrees.

A third method uses Klein’s line, drawn on the anteroposterior or frog’s leg–lateral radiograph, to evaluate for SCFE. The frog’s leg–lateral position is more sensitive in diagnosing SCFE. The line is drawn across the superior femoral neck and normally intersects with some part of the femoral head. In SCFE, Klein’s line does not intersect the femoral head (see Figure 30–10A).

**Differential Diagnosis**

Conditions to be considered in the differential diagnosis include fractures, AVN, osteomyelitis, septic arthritis, groin pull, and stress fractures.

**Complications**

Complications of untreated SCFE include AVN and cartilage loss.
**Treatment**

Patients with SCFE should be referred to an orthopedic surgeon once the diagnosis is made. Initially the child should be fitted with crutches or a wheelchair to limit weight-bearing stress on the hips. Definitive treatment requires surgical stabilization of the epiphysis with pins or a screw. Most patients are treated with a single-screw procedure. Some surgeons perform prophylactic pinning of the contralateral pelvis in patients with endocrinologic or metabolic diagnosis. Partial weight bearing with crutches is allowed after surgery for the first 6 weeks. Exercises focus on improving ROM and strength. After 6 weeks if pain is controlled full weight bearing is permitted and any assistive device may be discontinued.

Peck D: Slipped capital femoral epiphysis: Diagnosis and management. Am Fam Physician 2010;82:258–262.

**LEGG-CALVÉ-PERTHES DISEASE**

**General Considerations**

In Legg-Calvé-Perthes disease (LCPD), osteonecrosis of the femoral head epiphysis in a skeletally immature patient results in contact of the large femoral head with the acetabulum. The disease process is not well understood but appears to involve interruption of the blood supply to the femoral head. The cause of hip pain in patients with LCPD is likewise unclear. Some of the proposed generators of pain are femoral acetabular impingement, instability, labral disease, and early osteoarthritis.

**Clinical Findings**

**A. Symptoms and Signs**

A thorough history and physical examination should identify characteristic
features of LCPD. Physical examination includes ROM testing, as well as the femoral acetabular impingement test and observation for presence of the Trendelenburg sign. The Trendelenburg sign is positive when weight bearing on the affected side results in contralateral hip drop, indicating weakness of the gluteus medius muscle. The anterior impingement sign is positive when pain is elicited with hip flexion to 90 degrees. On examination, the hip tends to go into external rotation with significant limitation in internal rotation. The lateral impingement sign is considered positive if pain is elicited with internal rotation in a supine position with the pelvis stabilized. The posterior impingement sign is considered positive if the patient has pain with extension and external rotation.

**B. Imaging Studies**

Plain radiographs are essential in the evaluation of LCPD. Abnormalities of the head size ratio and articular–trochanteric distance should be screened for and assessed. The head size ratio is calculated as the size of the unaffected femoral head divided by the affected femoral head. The larger the head size ratio that is calculated, the better the prognosis. The articular–trochanteric distance is defined as the distance between two lines, one at the top of the femoral head and the other at the top of the trochanter. The smaller the articular trochanteric distance that is measured, the more severe the disease progression.

**Differential Diagnosis**

Other conditions to consider in the differential diagnosis include transient synovitis, septic arthritic, and sickle cell crisis.

**Treatment**

Treatment can involve nonsurgical or surgical management. Nonsurgical treatment includes bracing and hip ROM exercises. ROM exercises, hip adductor tendon release, traction, and abduction plaster casting are used to maintain greater than 30 degrees of hip abduction. Bracing involves the use of the Atlanta Scottish Rite orthosis to position the legs in greater than 30 degrees of abduction. The most important factor for healing is age and amount of femoral head involvement.
HIP DISLOCATION

General Considerations

Traumatic hip dislocation is categorized as simple or complex based on the presence of associated fractures: simple dislocations are not associated with fracture; complex dislocations involve a fracture. Factors such as depth of the acetabulum and labrum, thickness of the joint capsule, and strength of the muscular structure are important considerations when evaluating a patient with hip dislocation.

Anterior hip dislocation is much less common than posterior dislocation (10% versus 90%). Anterior dislocation can be differentiated according to the mechanism of dislocation into superior and the inferior types. The superior type is caused by abduction, external rotation, and extension of the hip. The inferior type is caused by forced abduction, external rotation, and flexion of the hip. Posterior hip dislocation occurs as a result of forced adduction, internal rotation, and flexion of the hip.

Femoral anteversion is a risk factor for hip dislocation. Most hip dislocations occur during a motor vehicle collision as a result of the impact of the knee against the dashboard.

Clinical Findings

A. Symptoms and Signs
The classical appearance of a posterior hip dislocation is a flexed, internally rotated, and adducted leg. In contrast, the leg of a patient with an anterior hip dislocation appears in a position of external rotation, extension, and abduction. The femoral head may be palpable in the buttock region.

B. Imaging Studies
Plain radiographs with anteroposterior and lateral views are needed to diagnose hip dislocation. With posterior dislocation, an anteroposterior view shows a small and superiorly located femoral head that is incongruent with the acetabulum. An anteroposterior view of an anterior dislocation will show an abducted femur with the femoral head inferior to the acetabulum. If there is a high index of suspicion for a fracture and plain radiographs are negative, a multidetector CT scan should be used in the diagnosis of hip dislocation.

**Differential Diagnosis**

It may be difficult to differentiate hip dislocation from intertrochanteric, femoral neck, or acetabular fracture. Sacroiliac joint and hip subluxation can also mimic hip dislocation.

**Complications**

Recurrent hip dislocation, AVN, heterotrophic ossification, early degenerative osteoarthritis, and nerve injuries are some of the possible complications of untreated hip dislocation.

**Treatment**

A dislocated hip should be reduced within 6 hours to decrease the likelihood of AVN. After relocation of the hip, posterior hip dislocation precautions should be followed. Patients should be restricted to touch-down weight bearing for 4 weeks, and then advanced to “weight bearing as tolerated” by 8 weeks. Treatment of hip dislocation includes closed reduction followed by hip arthroscopy to resect or repair labral pathology. Physical therapy should be initiated after the relocation to strengthen the musculature around the hip. Therapy sessions should focus on gait training and passive hip pendulums.


GREATER TROCHANTERIC PAIN SYNDROME

General Considerations

Trochanteric bursitis is described as tenderness to palpation over the greater trochanter when the patient is in a side-lying position. It usually radiates down the lateral aspect of the thigh or into the buttocks. Symptoms develop in response to repetitive friction between the greater trochanter and the iliotibial band when the hip is in flexion and extension.

Greater trochanteric pain syndrome encompasses several disorders affecting the lateral, peritrochanteric space of the hip, including trochanteric bursitis and tears of the gluteus medius and minimus. It is reported in 10–25% of the population. A higher prevalence has been reported in patients with low back pain, and the condition is also more common in women than men, a circumstance that has been linked to the higher incidence of gluteus medius tears in women (20%) because of the wider female pelvis. Coxa saltans, also known as snapping hip syndrome, is described as an audible and snapping of hip during activities that require repetitive flexion, extension, and abduction. This finding most often represents the iliotibial band and anterior border of the gluteus maximus snapping over the greater trochanter.

Clinical Findings

A. Symptoms and Signs

Typically patients have tenderness to palpation of the symptomatic trochanteric bursa. Other criteria for diagnosis include pain with hip abduction against resistance and a positive FABERE test (pain elicited with flexion, abduction, external rotation, and extension of the hip).

Ober’s test for iliotibial band tightness may also be positive. To perform this test, the patient is placed in a lateral decubitus position with the affected side up. The symptomatic hip is extended and abducted. The test is positive when it elicits lateral hip pain and limitations in hip adduction past the midline.

On physical examination, patients with gluteus medius and minimus tear display weakness and pain in response to active, resisted abduction in extension and external rotation while the hip is flexed to 90 degrees. Other examination findings include pain lasting 30 seconds or longer during single-leg stance.
B. Imaging Studies

Although not required for the diagnosis of trochanteric bursitis, plain radiographs may be obtained to evaluate for enthesopathy and intraarticular sources of pain. Calcifications are sometimes seen around the bursa space in trochanteric bursitis. Ultrasound study can show calcific tendinopathy. MRI studies can show evidence of gluteus muscle tendonitis or tear. Edema is the earliest sign of gluteal tendinopathy. Other signs of tendinopathy include fatty atrophy, bony irregularity, tendon calcification, thickening or increased intrasubstance sign intensity on T2-weighted scan, partial tear with focal tendon disruptions, and complete tear with tendon discontinuity. Elongation of the gluteal medius tendon greater than 2 cm in the myotendinous junction is indicative of a tear. Loose bodies or synovial chondromatosis can also be seen on MRI.

Differential Diagnosis

Several conditions should be considered in the differential diagnosis, including low back pain, lumbar radiculopathy, sacroiliac joint dysfunction, hip osteoarthritis, and hip fracture.

Complications

If left untreated, greater trochanteric pain syndrome can lead to gait abnormalities such as a Trendelenburg gait. The compensatory actions involved in altered stance and gait can predispose a patient to hip osteoarthritis.

Treatment

Greater trochanteric pain syndrome is usually a self-limiting condition that responds to conservative measures such as rest, activity modification, ice, anti-inflammatory agents, and physical therapy focused on stretching, flexibility, strengthening, and gait mechanics. In one study, local anesthetic injections with corticosteroid had an efficacy rate of 77.1% for 1 week and 61.3% for up to 6 months. A report by Cohen and colleagues noted no difference between fluoroscopically guided injection and bedside injection.

Among other approaches, low-energy shockwave therapy has been found to
be superior to other nonoperative modalities. In one report, shockwave therapy allowed 64–76% of patients to return to normal activities. For recurring or refractory cases, surgical intervention such as bursectomy and iliotibial band release have been found to be effective.


OSGOOD–SCHLATTER DISEASE

▶ General Considerations

Osgood–Schlatter disease results from repetitive injury and small avulsion injuries at the bone–tendon junction where the patellar tendon inserts into the secondary ossification center of the tibial tuberosity. It usually manifests during early adolescence, is more common in boys than in girls, and affects athletic more than nonathletic teens (21% versus 4.5%). The disease is found bilaterally in 20–30% of patients.

▶ Clinical Findings

A. Symptoms and Signs

Patients report pain that is worse with running, jumping, and kneeling activities. On examination, tenderness and swelling may be noted at the insertion of the patellar tendon into the tibial tubercle. Patients may have an antalgic gait, and extension lag may be observed during the examination. This is associated with tightness of the hamstring and quadriceps. Bony irregularities also may be palpated in patients with symptoms of long duration.

B. Imaging Studies

Plain radiographs can be used to diagnose Osgood–Schlatter disease.
Anteroposterior and lateral views of the knee often show soft tissue swelling or small spicules of heterotrophic ossification anterior to the tibial tuberosity. Enlargement of the tibial tubercle also may be noted. Ultrasound and MRI can also be used in making the diagnosis.

**Differential Diagnosis**

The physician should rule out avulsion fracture of the tibial tuberosity, patellofemoral stress syndrome, pes anserinus bursitis, and Sinding-Larsen-Johansson disease (discussed next) as the cause of symptoms.

**Complications**

Osgood–Schlatter disease may lead to persistent hamstring and quadriceps tightness.

**Treatment**

Conservative measures, including ice and NSAIDs, can be used to alleviate symptoms. Decreasing activities allows for healing of the microscopic avulsion fractures. Immobilization is sometimes needed for patients with severe symptoms. Surgical treatment, which is rarely utilized, consists of a tibial tuberosity osteotomy using an anterior approach. Most patients treated for Osgood–Schlatter disease who are evaluated in adulthood have no symptoms.

**SINDING-LARSEN-JOHANSSON DISEASE**

Sinding-Larsen-Johansson disease is a disorder of the knee that affects the junction of the patellar tendon and the distal pole of the patella. Similar to Osgood–Schlatter disease, it occurs in maturing adolescents. Patients report anterior knee pain that is worse when climbing and descending steps. The pain usually improves with rest.

Physical examination usually reveals tenderness to palpation at the patellar tendon–patella junction. Palpation of this junction should not find evidence of a palpable gap. Such a gap is more consistent with patellar fracture.

Radiographs, including anteroposterior, lateral, and merchant views, should
be ordered for use in staging the disease. Medlar and Lyne classified radiographic staging into five stages. Stage 1 is defined by normal radiographs, stage 2 by irregular calcifications at the inferior patella, and stage 3 by coalescence of calcifications. Stage 4 is further divided into two subcategories: stage 4a, showing incorporation of calcium into the patella, and stage 4b, showing a calcific mass separate from the patella.

Differential diagnosis encompasses other conditions of the patella, including Osgood–Schlatter disease, bipartite patella, patella alta, patella baja, patellar tendinitis, and patellar stress fracture. Activity modification, ice, antiinflammatory agents, and other conservative modalities are mainstays of treatment. The condition is self-limiting and resolves after full maturation, generally within 12–18 months. However, symptoms may continue until full maturation of the inferior pole of the patella has occurred.

ANKEle SPRaIN

1. Lateral Ankle Sprain

General Considerations

Lateral ankle sprains comprise up to 21% of sports-related injuries. Owing to the complexity of the ankle, trauma to the structures that make up this joint may have serious consequences for both athletes and nonathletes. When evaluating a patient with a possible ankle sprain, the physician must first rule out fracture and the need for radiologic examination. The Ottawa ankle rules, reproduced in Table 29–3 and described below, aid in determining the need for radiographs. Careful assessment, management, and followup are key to averting persistent complications of these injuries.

Pathogenesis

Eighty-five percent of ankle sprain injuries involve the lateral ligaments and occur with forced ankle plantar flexion, supination, and inversion. The lateral ankle ligaments consist of the anterior talofibular ligament, calcaneofibular ligament, and posterior talofibular ligament. The anterior talofibular ligament
inhibits anterior translation of the talus in the mortise and excessive inversion and internal rotation of the talus in the mortise joint. In ankle plantar flexion, the talus moves anteriorly, leaving the more narrow posterior aspect of the talus in the mortise joint. The anterior talofibular ligament is prone to injury following excessive ankle inversion. The calcaneofibular ligament runs from the inferior tip of the lateral malleolus to the lateral calcaneus and primarily restricts adduction and inversion. The anterior talofibular ligament is injured first, and the calcaneofibular ligament is injured second if there is a greater force of injury. The posterior talofibular ligament, the strongest of the lateral ligaments, inhibits eversion and runs from the posterior aspect of the lateral malleolus to the lateral calcaneus.

Clinical Findings

A. Symptoms and Signs

Assessment of any ankle injury should begin with inspection for swelling, ecchymosis, and gross deformity. ROM of the ankle should be evaluated, and vascular, motor, and sensory examinations should be performed. Two maneuvers are used to test the stability of the anterior talofibular ligament: the anterior drawer test and the talar tilt test.

In performing the anterior drawer test, the physician stabilizes the tibia and fibula anteriorly while the patient is supine (Figure 30-12). The opposite hand cups the calcaneus, inducing 30 degrees of plantar flexion, and translates the foot anteriorly while holding the tibia and fibula in place. A positive test is indicated by a dimple sign at the anterior talocrural joint.
Anterior drawer test assesses anterior talofibular ligament stability.

The talar tilt test assesses both the anterior talofibular and the calcaneofibular ligaments (Figure 30-13). The physician stabilizes the tibia and fibula and induces ankle inversion, with some supination, while cupping the calcaneus. The test is positive when the affected ankle translates into greater inversion when compared with the unaffected side; this signifies a tear of the calcaneofibular ligament.
Sprains are classified into three grades, representing mild, moderate, and severe injury. A grade I ankle sprain does not demonstrate ligamentous laxity using the previously mentioned tests. Grade II sprains are unstable with increased laxity caused by a tear of anterior talofibular ligament. Grade III sprains are unstable, with increased laxity as a result of tears of the anterior talofibular calcaneofibular ligaments.

The syndesmosis is the interosseous membrane that transmits force between the tibia and fibula. It is often injured in ankle sprains. The three most commonly used maneuvers to assess the syndesmosis for injury are the external rotation test, squeeze test, and crossed leg test. The physician performs the external rotation test by stabilizing the tibia and inducing external rotation of the ankle, or abduction and eversion. In the squeeze test, the tibia and fibula are compressed just above the midpoint of the calf. Pain elicited by these maneuvers suggests injury to the syndesmosis. The crossed leg test can be performed by placing the patient’s affected leg on the opposite thigh. The physician then presses down on the medial side of the knee on the patient’s affected leg. A positive test elicits pain because the area of the injured syndesmosis is being used as a fulcrum over the unaffected thigh.

B. Imaging Studies

The Ottawa ankle rules are guidelines that aid in determining the need for radiologic imaging in patients with ankle trauma (ie, to rule out fracture). The examiner inspects for bony tenderness at the posterior lateral and posterior medial malleolus, as well as the base of the fifth metatarsal and navicular bones, and observes the patient for inability to bear weight for more than four steps. The guidelines have a sensitivity of 99.6% when used within 48 hours of injury.

MRI can provide insight into osteochondral defects, bone bruises, and stress or occult fracture if the diagnosis is questionable.

Differential Diagnosis

Differential diagnosis includes fracture of the ankle bones, which can be aided by examination using the Ottawa rules.

Complications
Ankle sprains, despite their frequent occurrence, are potentially serious injuries that even with appropriate treatment, can lead to scar tissue, persistent ankle instability, and recurrent sprains.

**Treatment**

No difference in subjective instability has been reported between patients receiving conservative and surgical treatment. Early mobilization within the first week, with weight bearing as tolerated and active ROM exercises, expedites the patient’s functional outcome and prevents atrophy related to immobilization. Taping protects the ankle from reinjury by restricting ROM, rather than enhancing proprioception. Ankle–foot orthoses (AFOs) are essential adjuncts in the treatment of patients with ankle sprains that result in foot drop from peroneal nerve traction injury. AFO use can be weaned as neurologic recovery ensues. Ankle sprain rehabilitation involves four phases: (1) pain control, including reduction of edema and protection of ligamentous structures, (2) gait normalization, (3) return to daily activities, and (4) return to sport.

Osteopathic manipulative treatment of the acutely sprained ankle in the emergency department has resulted in statistically significant reductions in edema and pain. Edema increases the likelihood of adhesion formation and necessitates the redirection of fluid back into the lymphatic system. The peroneal muscles are also addressed in osteopathic manipulation, as they undergo strain and benefit from muscle energy or counterstrain techniques to restore proper resting length.

**Prognosis**

The risk of ankle sprain recurrence ranges from 3% to 34%. There is good evidence supporting the use of taping, a semirigid orthosis, or an ankle brace to prevent subsequent sprains in patients participating in high-risk sporting activities. There is no conclusive evidence supporting the use of high-top shoes in the prevention of ankle sprain. The ability to walk again within 24 hours after trauma is indicative of good prognosis.

Among patients with accompanying peroneal nerve injury, those with neurapraxic lesions have the best prognosis; those with axonal loss have a favorable, but less predictable recovery.

The return-to-play decision for the patient who is an athlete must be made on
an individual basis and sometimes can be arbitrary. According to the American Academy of Orthopedic Surgeons, ankle sprains typically heal in 4–6 weeks. However, the athlete can usually resume light activity with swimming or cycling if tolerated within the first week. Anderson and colleagues developed a chart and checklist to help patients and therapists estimate the return to play (see full citation below).


2. Medial Ankle Sprain

General Considerations
Medial ankle and syndesmotic sprains account for 10–15% of ankle sprains and are commonly seen in male football, soccer, and basketball players, and female volleyball players. A common mechanism of injury in these sports is planting the foot and cutting.

**Pathogenesis**

Medial ankle sprains occur with excessive ankle eversion and dorsiflexion, causing injury to the deltoid ligament. Eversion and dorsiflexion result in widening of the ankle mortise joint. Often, medial ankle sprains are associated with ankle syndesmosis injuries. In addition to the deltoid ligament, other structures often injured with medial ankle sprains are the anterior inferior tibiofibular ligament, posterior tibiofibular ligament, and syndesmosis (distal one third). Fracture of the proximal fibular (Maisonneuve fracture) must be also ruled out.

**Clinical Findings**

**A. Symptoms and Signs**

Tenderness to palpation may be elicited at the anterior inferior talofibular ligament. The patient may walk with a heel-raise gait pattern to avoid dorsiflexion needed during pushoff. Dorsiflexion drives the wider, anterior talus into the mortise joint, stressing the tibiofibular ligaments and syndesmosis.

Diagnostic maneuvers include the external rotation test, squeeze test, and crossed leg test (described earlier under Lateral Ankle Sprain). Other maneuvers include the point test, during which the examiner applies pressure over the anterior aspect of the distal tibiofibular syndesmosis, and the one-legged hop test, which involves unilateral hopping on the affected limb (the latter should be administered with caution). Pain with either test indicates a positive result. In the heel thump test, performed with the patient seated, the examiner plantar flexes the affected ankle, applying a firm thump on the heel in an attempt to impose a separation of the distal tibia and fibula. This test may also be used to diagnose tibial stress fractures.

**B. Imaging Studies**

Ankle anteroposterior and mortise views are used to assess the tibiofibular clear
space, which is the area between the lateral border of the posterior tibia and the medial border of the fibula. The tibiofibular clear space should be less than 6 mm. Stress radiographs may not be tolerated by the patient. CT scan and MRI are helpful in diagnosing syndesmotic injuries and secondary ligamentous injuries.

Complications

Anterior impingement syndrome of the ankle can be a sequela of the syndesmotic injury. Heterotopic ossification near the syndesmosis may develop and require surgical excision.

Treatment

Conservative treatment consisting of rest, ice, compression, and elevation with immediate non–weight bearing (ie, use of a rolling walker or crutches) is recommended. Ankle taping and semirigid bracing are helpful. As pain and swelling subside, the patient can begin partial weight bearing with low-level standing balance training and strength training. Syndesmotic screw fixation is only considered if there is persistent widening of the mortise joint and lateral displacement of the fibula.


SPORTS HERNIA (ATHLETIC PUBALGIA; see also Chapter 29)

“Sports hernia” is the term given to a controversial and difficult-to-diagnose syndrome that has also been called *athletic pubalgia*, Gilmore’s groin, pubic inguinal hernia syndrome, and athletic hernia. It refers to weakness or disruption of the musculotendinous junction of the rectus abdominis insertion to the
superior pubic ramus. Detailed discussion of this condition appears in Chapter 29.

Sports hernia occurs more frequently in soccer and ice hockey players, accounting for 10–13% of injuries in soccer annually, and is more common in men than in women. Patients complain of groin pain that is usually relieved with rest and resumes with activities such as kicking, sudden acceleration, twisting, turning, cutting, sit-ups, coughing, and sneezing. There is a significant correlation with hip disease, especially FAI. Physical examination, provocative testing, and MRI are useful in identifying the source and location of the pain. MRI findings include bone marrow edema at the pubic symphysis. The differential diagnosis is wide and includes inflammatory conditions (eg, inflammatory bowel disease, osteoarthritis, appendicitis), musculoskeletal conditions (eg, stress fracture, bursitis, muscle strain, tendon rupture, osteitis pubis), and infectious diseases (eg, prostatitis, osteomyelitis, urinary tract infections). Neurologic diseases, such as nerve entrapments and neoplastic origins, should also be considered.

Conservative treatment includes rest, ice, and antiinflammatory agents. Physical therapy is an important component of rehabilitation. Surgical referral may be considered in patients who do not respond to these measures. For additional details, see Chapter 29.


COMPARTMENT SYNDROME

1. Acute Compartment Syndrome

General Considerations

Compartment syndrome can be acute or chronic. Acute compartment syndrome is a complication following fractures, soft tissue trauma, and reperfusion injury after acute arterial obstruction. It is caused by bleeding or edema in a nonelastic
muscle compartment surrounded by fascia or bone. Risk factors for the development of acute compartment syndrome include anticoagulant therapy and bleeding disorders. Risk factors for foot compartment syndrome include joint dislocations, especially Chopart and Lisfranc fracture–dislocation. Most cases of acute compartment syndrome in the foot are caused by high-energy deceleration trauma.

### Pathogenesis

The lower leg has four compartments: anterior, lateral, superficial posterior, and deep posterior. Perfusion within a compartment is compromised when the difference between the diastolic pressure and the intracompartmental pressure is less than 30 mm Hg. When the pressure balance is altered, ischemia causes capillary wall damage that eventually results in permanent nerve or muscle damage. The incidence of acute compartment syndrome is about 6% in patients with foot injuries and 1.2% in patients with closed diaphyseal fractures of the tibia. Studies have shown that there is a temporal–pressure relationship for conduction block and muscle necrosis, with altered pressures noted as early as 3 hours after a precipitating event or injury.

### Clinical Findings

Pain, paraesthesia, paresis, pulse, and skin color should be evaluated during physical examination. The most important factor in diagnosing acute compartment syndrome is clinician suspicion. Pain is the earliest and most sensitive symptom. The threshold at which the compartment should be decompressed is debatable. Intracompartmental pressure of 30 mm Hg, mean arterial pressure of 30 mm Hg, or pressure of 20 mm Hg below diastolic blood pressure are used in diagnosing acute compartment pressure.

### Complications

Permanent injury to the nerves and muscles is a potential concern in all cases of compartment syndrome.

### Treatment
Management of compartment syndrome centers on adequate and prompt fasciotomy of all compartments involved. The earlier the compartment pressure is reduced, the less severe will be the sequelae. Fasciotomy can be carried out by single lateral incision or by combined anterior lateral and posteromedial incisions.


2. Chronic Exertional Compartment Syndrome

▶ General Considerations

Chronic external compartment syndrome is a condition of pain induced by exercise, swelling, and impaired muscle function. It is common in runners and individuals undergoing military training. Risk factors include muscular hypertrophy, increased intracranial pressure, venous hypertension, post-traumatic soft tissue inflammation, and decreased fascial elasticity

▶ Pathogenesis

Symptoms are usually bilateral and can result in an increase of 20% in muscle volume due to the nonconfined space of the fascia. Chronic external compartment syndrome results in increased muscle compartment pressure, which affects the tissue circulation, leading in turn to ischemia. The anterior and deep posterior compartments are most often affected at a rate of 25% each.

▶ Clinical Findings

Patients usually complain of anterolateral pain radiating to the dorsum of the foot and feelings of calf tightness. The definitive diagnosis is made on the basis of elevated dynamic intramuscular compartment pressure. Widely accepted parameters during compartment measuring include basal pressure greater than 15 mm Hg, greater than 30 mm Hg after 1 minute postexercise, and greater than 20 mm Hg at 5 minutes postexercise.
Differential Diagnosis

Medial stress fracture syndrome, popliteal artery entrapment syndrome, myopathy, and sural nerve entrapment syndrome should be considered in the differential diagnosis.

Complications

As in acute compartment syndrome, described earlier, avoidance of permanent injury to the nerves and muscles is the primary concern.

Treatment

Conservative measures such as rest and cessation of activity should be the first line of treatment. If these fail, surgical release should be considered, as studies show it to be effective in 81–100% patients. Partial fasciotomy is associated with a high recurrence rate. Pain relief after fasciotomy does not correlate with postcompartment measurements.


FRACTURES

A fracture is a severe musculoskeletal injury. Any trauma or pain associated with injury requires that fracture be ruled out first. Fracture rehabilitation is essential for a successful functional recovery and should be started as soon as the fracture is stabilized. There are four main goals in fracture rehabilitation: (1) to maintain the ROM of related joints; (2) to preserve muscle strength; (3) to facilitate the fracture healing by activity; and (4) to return the patient to function and employment at the earliest possible time.
Classification

Numerous classification systems have been developed that enable fractures to be differentiated for various purposes. For instance, fractures may be classified by age of the patient, as adult and childhood fractures heal quite differently and have different potentials for remodeling. Fractures may be described on the basis of etiology (eg, traumatic, stress, pathologic), associated soft tissue injury (eg, open or closed), fracture direction (eg, transverse, oblique, spiral), anatomic location (eg, proximal, middle, distal, supracondylar, subtrochanteric, diaphysis, or metaphyseal), displacement, and degree of fragmentation (eg, linear, comminuted, or segmental).

Fracture-specific classification systems have also been generated that classify a single fracture in a single location. For example, the Garden classification system for femoral neck fracture and the Neer classification system for proximal humeral fracture have both been long and widely used.

The AO Foundation (Arbeitsgemeinschaft fur Osteosynthesefragen) and Orthopaedic Trauma Association (AO/OTA) Müller fracture classification system is a universal system that has been internationally accepted by many orthopedists and other physicians. It is an alphanumeric system that can be used with fractures. The coding system includes the bone, location of the bone, type of fracture, group, and subgroup of the fracture. For example, a proximal humeral fracture that is impacted with marked displacement would be coded using this system as follows: 1.1-C2. The first numeral, “1”, represents the affected bone, in this case the humerus; the second “1” specifies the location as proximal; the “C” indicates that the fracture is intraarticular; and the “2” specifies that it is impacted with marked displacement.

Bone Healing of Fractures

Fracture healing follows a characteristic course, which can be divided into three overlapping phases: inflammation, repair, and remodeling. The inflammation phase starts immediately after injury and lasts approximately 5–14 days. During this phase, hematoma formation and recruitment of inflammatory cells are major events of fracture healing. Polymorphonuclear leukocytes followed by macrophages and lymphocytes migrate to the fracture area and release multiple cytokines and growth factors that lead to angiogenesis and stimulate the fracture repair process.

The repair phase lasts weeks to months. This phase is characterized by
mesenchymal cell differentiation, cell proliferation, and synthesis of reparative matrix. The mechanical stability of the fracture site determines the different repair processes through which the fracture healing will progress. When the fracture ends are rigidly fixed in direct and intimate contact, fracture healing will go through the primary bone healing process. Primary bone healing is characterized by bone healing without the formation and replacement of visible callus. The primary healing of cortical bone is very slow and not superior to secondary healing. Secondary bone healing is a much more common way for bone to heal. Secondary healing is characterized by callus formation and bridging across the fracture gap. It begins with soft callus, which is a cartilage callus with minimal mineralization. This converts to hard callus with further mineralization, increasing mechanical strength. The greater the motion at the fracture site, the greater will be the quantity of callus. The bridge callus across the fracture gap indicates the successful healing of the fracture and is taken as the point at which the patient can resume loading of the bone.

The total fracture healing process may last years. The longest component of the total process is the remodeling phase. It lasts months to years and is characterized by replacement of callus formed in the repair phase with more organized and mechanically efficient bone tissue. Woven bone formed in the cortical fracture gap is remodeled to lamellar bone by osteon formation. Remodeling and resorption of the periosteal and medullary calluses leads to reshaping of diaphyseal bone. The three main fracture healing phases overlap each other, and activities that occur mainly in one stage may have begun in an earlier stage.

Multiple factors adversely influence fracture healing. These factors include poor regional blood supply, open fracture and subsequent infection, severe soft tissue damage, segment fracture, pathologic fracture, systemic disease, osteoporosis, and corticosteroid use, among others.

Clinical Evaluation & Diagnosis

It is critical for physiatrists to be vigilant about the possibility of fracture in patients with even minor trauma. This is particularly so when evaluating osteoporotic patients, or those with sensory deficits, cognitive impairment, or expressive aphasia. Whenever a patient presents with musculoskeletal pain, with or even without significant trauma history, a fracture needs to be excluded. A misdiagnosed nondisplaced fracture can easily become a displaced fracture, thereby complicating healing and potentially delaying and prolonging the
rehabilitative process. Physiatrists should never assume all fractures have been previously detected when receiving transferred patients with trauma. This is particularly so if notified by the therapist that a patient had regression of function or increased pain or swelling.

A. Symptoms and Signs

Pain and tenderness to palpation are noted at the fracture site in most fractures. However, patients with deep fractures sometimes have a referred pain pattern. Femoral neck fracture can manifest as knee pain, and patients with spine fracture can present with referred symptoms to the buttocks or legs. Other symptoms and clinical signs of fracture include loss of motion, loss of function, swelling, bruising, deformity, and abnormal mobility. Fusiform ecchymosis is characteristic of common finger and toe fractures.

B. Imaging Studies

In most but not all instances, radiographic examination should be sufficient to diagnose a fracture. Radiographic signs of fracture should correlate to local tenderness. Fracture diagnosis cannot be made by radiographic findings without clinical supporting evidence. Comparison radiographic study of the opposite extremity is a very useful method for diagnosis of minor fractures or epiphyseal fracture in children.

CT and bone scans are both helpful in diagnosing small subtle fractures and those of the spine, wrists, and feet, where overlapping bones may hide a subtle fracture. Bone scan and MRI can help to identify stress fractures and differentiate new fracture sites from old.

C. Determination of Fracture Healing

The healing status of a fracture is key to the initiation of rehabilitation. The judgment involved in determining the status of fracture healing depends on knowledge of expected healing times specific to the bones injured, radiographic evidence, and clinical evaluation. Most fractures follow the timelines outlined in Table 30–1. These predicted healing times give the physician a basic time frame to use in evaluating the fracture.

Table 30–1 Time frame for healing of common fractures.
Radiographic evaluation is the major method to test fracture healing. From initial fracture to clinical healing, serial radiographs may be needed depending on the severity and stability of the fracture. Radiographic changes indicative of healing include widening or blurring, disappearance of fracture line, and callus formation. Bridging callus formation is not necessary but, if present, indicates completion of the fracture healing process.

The clinical evaluation is based on the patient’s symptoms and physical examination. Pain and tenderness to palpation should be absent or greatly diminished at the fracture site. The following discussion reviews a sampling of

<table>
<thead>
<tr>
<th>Fracture Location</th>
<th>Predicted Healing Time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clavicle</td>
<td>3–8 weeks</td>
</tr>
<tr>
<td>Scapula</td>
<td>6 weeks</td>
</tr>
<tr>
<td>Ribs</td>
<td>4 weeks</td>
</tr>
<tr>
<td>Humerus (upper arm)</td>
<td>4–10 weeks</td>
</tr>
<tr>
<td>Olecranon (elbow)</td>
<td>4–6 weeks</td>
</tr>
<tr>
<td>Radius and ulna (lower arm)</td>
<td>6 weeks</td>
</tr>
<tr>
<td>Scaphoid (wrist)</td>
<td>6–24 weeks</td>
</tr>
<tr>
<td>Distal</td>
<td>6–8 weeks</td>
</tr>
<tr>
<td>Middle</td>
<td>8–12 weeks</td>
</tr>
<tr>
<td>Proximal</td>
<td>12–24 weeks</td>
</tr>
<tr>
<td>Fingers</td>
<td>4–6 weeks</td>
</tr>
<tr>
<td>Pelvis</td>
<td>4–6 weeks</td>
</tr>
<tr>
<td>Femur (upper leg)</td>
<td>12 weeks</td>
</tr>
<tr>
<td>Patella (knee)</td>
<td>4–6 weeks</td>
</tr>
<tr>
<td>Tibia and fibula (lower leg)</td>
<td>10–24 weeks</td>
</tr>
<tr>
<td>Ankle</td>
<td>6 weeks</td>
</tr>
<tr>
<td>Foot</td>
<td>3–12 weeks</td>
</tr>
<tr>
<td>Toes</td>
<td>3 weeks</td>
</tr>
</tbody>
</table>
fractures that are likely to be seen by physiatrists.

1. Fracture of the Clavicle

General Considerations

Clavicular fractures account for 2.6–5% of all fractures and occur most often in children and young adults. In fact, fracture of the clavicle is the most common pediatric and newborn fracture. These fractures may also occur in children victimized by child abuse.

Most clavicular fractures result from a fall onto the shoulder (87%) or a direct blow to the clavicle (7%). The clavicle serves as a strut linking the axial skeleton to the upper extremity. Huge amounts of energy can be applied to the clavicle during a fall. The middle one third of clavicle is the most vulnerable part. Common incidents that may lead to fracture include automobile accidents, horizontal falls on the shoulder joint, or athletic injuries incurred in activities such as football, gymnastics, and cycling. Most of these sports injuries may be preventable with the use of protective equipment and adequate padding. Muscles that may affect clavicular fractures include the deltoid, trapezius, subclavius, sternocleidomastoid, sternohyoid, and pectoralis major muscles. The ligaments involved include the conoid and trapezoid ligaments.

Classification

Clavicular fractures are broadly categorized by their anatomic location: medial third, middle third, and distal third (Allman classification). Most clavicular fractures occur at the middle third (80%), followed by distal third (15%), and then medial third (3–5%). Fracture of the middle third is further classified by displacement and fragmentation. The most commonly used classification for fracture of the distal clavicle is the Dameron and Rockwood classification, which is similar to that used for ACJ dislocation.

Clinical Findings

A. Symptoms and Signs
Clavicular pain with a history of trauma makes the diagnosis straightforward. However, in newborns and very young toddlers, the fracture is often challenging to identify. The young child may present with a pseudoparalysis or paucity of use on the affected side.

B. Imaging Studies
Fractures in the lateral aspect of the clavicle may require a stress anteroposterior view with the patient holding a 5–10 lb weight to accentuate the fracture deformity and reveal subtle injury to the coracoclavicular ligament. Cephalad-directed views are helpful in illustrating the displacement of fractures of the middle third of the clavicle.

Treatment

A. Nonoperative Management
Because the clavicle has an excellent blood supply, most clavicular fractures can be effectively treated nonoperatively. The goal of treatment is to maintain the length and curvature of the clavicle. Figure-of-eight harness braces and simple slings are the two most common methods used for immobilization. The exact immobilization method does not appear to change the outcome.

One of the advantages of the figure-of-eight harness is that it can free the hand on the affected side for non–weight-bearing work, including writing, typing, and personal hygiene. Therefore, if the fracture is on the dominant side, a figure-of-eight brace may be the better choice. In addition, the figure-of-eight brace may be more effective in preventing shortening of the clavicle. However, a simple sling is more comfortable and better tolerated by patients. In addition, care must be taken when using a figure-of-eight brace to avoid compression of the axillary vessels and brachial plexus.

Immobilization is normally required for 4–8 weeks. The sling or brace is removed when there is no pain or tenderness at the fracture site.

B. Operative Management
Indications for operative management of a middle third clavicular fracture include severely displaced fracture, shortening of more than 20 mm, open injury, vascular compromise, progressive neurologic loss, scapulothoracic dislocation, and irreducible fracture. The relative indications include multiple trauma,
bilateral fractures, intolerance to immobilization, and cosmesis.

Rehabilitation

Weight bearing is not permitted until fracture healing is established. However, passive ROM of the glenohumeral joint is encouraged to avoid joint contracture. Codman pendulum exercises may cause displacing moment to the fracture site and should be avoided. Contact sports should be avoided for 3 months after injury. In most patients, if fracture healing occurs within the normal time frame, light lifting can be allowed by 6 weeks postinjury and heavy lifting by 12 weeks.


2. Radial & Ulnar Fractures

General Considerations

Injury to the lower arm may result in fracture of the radius, the ulna, or both bones. Because the two bones function as a unit, they usually break simultaneously, or one bone fractures with distal or proximal radioulnar joint dislocation or subluxation. The exception is direct blunt-force type injuries, which can cause a single break to either bone. An ulnar fracture with radial head dislocation is termed a Monteggia fracture; a radial fracture with distal
radioulnar joint dislocation is termed a *Galeazzi fracture* (Figure 30–14).

![Figure 30–14](image)

**Figure 30–14** Galeazzi fracture showing radial midshaft fracture causing distal radioulnar joint disruption and dislocation.

### Classification

Fractures of the radius and ulna are classified according to the location of the fracture, degree of displacement and angulations, involvement of the radioulnar joints, fracture pattern, and degree of soft tissue injury in both closed and open fractures. The most commonly used classification system is that of the AO/OTA, described earlier, which includes 27 types of fracture, from A1.1 to C3.3. In this system, type A1.1 is the simplest fracture, involving one bone, and type C3.3 is the most severe, representing an irregular or comminuted fracture of both bones.
Treatment

Open reduction with internal fixation is the treatment of choice for almost all forearm fractures, including minimally displaced fractures. Even when closed reduction appears satisfactory on radiographs, this treatment approach for displaced forearm fractures is associated with a high incidence of malunion and nonunion, and a 71% unsatisfactory result.

Cast immobilization is generally appropriate only for a nondisplaced forearm fracture, unless the potential for remodeling is quite good. Ten degrees of angulation deformity of either bone individually results in little or no motion loss; however, the structural result may be cosmetically unacceptable. Significantly greater losses of motion may occur when one or both bones are angulated 20 degrees or more. When casting is an option, a long arm cast is used to immobilize both the elbow and wrist. Six to 8 weeks of cast immobilization is usually required to achieve adequate healing.

Complications

Neurovascular and musculoskeletal injury often accompanies forearm fracture. However, some neurovascular complications are iatrogenic, caused by the surgical repair or by prolonged tourniquet use. The most common procedure-related nerve injury is injury to the posterior interosseus nerve, which results in wrist and digit extensor injury without sensory loss.

With any extremity fracture the examiner should be vigilant for the possibility of a compartment syndrome (see earlier discussion). The forearm has both a volar and a dorsal compartment, and weakness, sensory deficits, weak or absent pulse, pallor, pain, or swelling in either compartment should warrant prompt compartmental pressure readings and consideration for fasciotomy.

Radioulnar synostosis is uncommon but is a serious complication when it does occur as it will severely affect forearm rotation. Risk factors include open fracture, infection, multiple injuries with traumatic brain injury, and delayed internal fixation. Surgery is the only treatment for radioulnar synostosis.

Rehabilitation

Patients with fractures of the radius, ulna, or both bones who are treated with open reduction and internal fixation are often placed in a wrist splint for first 7
days after surgery. The fingers, elbow, and shoulder should be passively mobilized several times a day to maintain ROM and prevent stiffness. Forearm rotation and full ROM exercises may start after the splint removed at postoperative day 7. Weight bearing, weight lifting, traction, and sports activity should be avoided until bone union has been achieved. Bone union usually takes 6 to 8 weeks.

The forearm is a predominantly nonsynovial joint with high-amplitude motion. The most common long-term complication of forearm shaft fracture is significant forearm stiffness, with loss of pronation occurring twice as frequently as loss of supination. Loss of forearm rotation sometimes occurs despite a perfectly normal-appearing radiogram. Forearm rotation plays a key role in upper extremity function, and is involved in most activities of daily living (eg, eating, grooming, dressing, personal hygiene, etc). Even subtle loss of pronation and supination will cause functional loss of activities, especially in sports and music instrument performance. Consequently, early forearm rotation exercises are mandatory to prevent rotation deficits.

3. Olecranon Fracture

General Considerations

The olecranon is the site of attachment of the triceps, which is the major elbow extensor. Olecranon fracture may also occur in combination with elbow dislocation or coronoid fracture. An olecranon fracture can be intraarticular or extraarticular, displaced or nondisplaced. The majority of olecranon fractures are intraarticular and displaced.

Classification

The Mayo classification is a commonly used to differentiate olecranon fractures. It classifies the fractures according to displacement, comminution, and ulnohumeral instability. Type I is an undisplaced fracture; type II is a displaced but ulnohumeral-stable fracture; type III is a displaced and ulnohumeral-unstable fracture. The letter A indicates a noncomminuted and, B, a comminuted fracture.

Treatment
Most olecranon fractures are intraarticular and displaced, requiring surgery with open reduction and internal fixation. Tension band wiring is the classic procedure and is appropriate for most simple fractures. A plate-and-screw fixation is used for comminuted fractures.

Nondisplaced olecranon fractures are rare. They can be treated with a long arm cylinder cast in 90 degrees of elbow flexion without immobilization to the wrist and hand. Cast immobilization for 4–6 weeks is usually required.

### Rehabilitation

The goal in rehabilitation of a patient with an olecranon fracture is to maintain ROM of the elbow, shoulder, and wrist, as well as extension strength of the triceps and other elbow extensors.

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### 4. Scaphoid Fracture

#### General Considerations

Scaphoid fractures comprise almost 70% of carpal bone fractures. The scaphoid is the largest bone of the proximal row of carpal bones (Figure 30–15). More than 80% of the surface of scaphoid is covered by articular cartilage. This characteristic reduces its capacity for periosteal healing and increases the rate of nonunion. The scaphoid is the major bony block to extension of the wrist and is prone to fracture when forced into a position of excessive extension.
The scaphoid is located at the radial aspect of the wrist and transfers the longitudinal force from the thumb to the radius. A longitudinal direct impact through the thumb can result in a stable nondisplaced scaphoid fracture. The only blood supply to the middle and proximal scaphoid comes from the dorsal (scaphoid) branches of the radial artery in a retrograde flow. Therefore, middle
and especially proximal scaphoid fractures have very high risk of avascular necrosis and nonunion. The distal scaphoid receives blood supply from volar branch of the radial artery and branches of the anterior interosseous artery. For this reason, fractures of the distal scaphoid generally heal with less difficulty.

**Classification**

The classification of scaphoid fracture is based on anatomic location. The most common scaphoid fracture is through the waist (65%), followed by proximal fracture (15%), distal fracture (10%), and tubercle fracture (2%). An occult scaphoid fracture refers to an undisplaced fissure fracture and should always be considered when initial radiographs are negative but clinical suspicion is high.

**Clinical Findings**

**A. Symptoms and Signs**

The patient with a scaphoid injury usually complains of wrist pain on the radial side after a fall onto the hyperextended wrist or an injury resulting in direct axial loading from the thumb to the wrist. Anatomic snuff box tenderness, swelling, and loss of wrist extension are the typical signs of acute scaphoid fracture. Diffuse wrist pain and swelling develop after 24 hours. A carpal metacarpal grind test with the thumb often helps in diagnosing this injury.

**B. Imaging Studies**

Four-view radiographs are required for evaluation of scaphoid fracture, including posteroanterior, lateral, radial (supinate) oblique, and ulnar (pronate) oblique views. The radial oblique view is often best for diagnosis of waist scaphoid fractures. The scapholunate angle is measured on the lateral view; an angle of 45 degrees is considered to be normal, and greater than 60 degrees indicates carpal instability and fracture displacement.

CT scan and bone scan can be helpful in evaluating fracture malunion and nonunion, but MRI is the gold standard to detect occult scaphoid fracture and associated ligamentous injuries.

**Treatment**
A nondisplaced occult scaphoid fracture can be very challenging to diagnose. A delayed or failed diagnose of an occult scaphoid fracture often carries a much poorer prognosis. Therefore, the patient with wrist hyperextension injury and anatomic snuff box tenderness should be suspected of having a scaphoid fracture even if initial radiographs show no evidence of fracture. Immobilization of the affected wrist with a short arm thumb spica cast for 2 weeks and repeated radiographs are recommended, or alternatively, an MRI can be obtained if the initial radiographs are negative and clinical suspicion remains high.

Nondisplaced scaphoid fractures can be treated with nonsurgical or surgical methods. Surgical treatment is associated with better functional outcome, patient satisfaction, and grip strength, and earlier return to work than nonsurgical treatment, but the rate of nonunion, ROM limitation, and pain are similar in both treatments. There is some controversy about cast immobilization; most orthopedists believe a short arm thumb spica cast is sufficient for stabilization, but several studies have shown that use of an above-elbow cast may increase the rate of union. The immobilization time for distal scaphoid fracture is about 6–8 weeks; for middle scaphoid fracture, about 8–12 weeks; and for proximal fracture, about 12–24 weeks.

A displaced scaphoid fracture occurs when there is more than 1 mm step-off between fracture segments or greater than 60 degrees of scapholunate angle. Surgical intervention is the treatment of choice, especially for middle or proximal scaphoid fracture, because the nonunion rate is very high without surgery. Surgical treatment includes closed reduction with percutaneous screw fixation and open reduction with screw internal fixation.

### Complications

Delayed union, malunion, nonunion, and avascular necrosis are common complications of scaphoid fractures. Surgical treatment includes internal fixation along with various bone graft and vascularized bone grafts procedures.

### Rehabilitation

Scaphoid fractures usually require long-term immobilization, especially for middle and proximal fractures. Stiffness of the wrist and thumb is common after cast removal. Early aggressive stretch exercises of the thumb and wrist are
necessary to restore wrist and thumb function. Grip strengthening exercises with a squeeze ball or therapeutic putty are commonly used.

After open reduction and internal fixation, a short arm thumb spica cast or splint is usually needed for 8–12 more weeks until the bone is radiographically and clinically healed. ROM exercises of all digits, elbow, and shoulder are important during the immobilization period with the cast.

Electromagnetic stimulation has proved helpful for delayed union and nonunion of the scaphoid.


LOW BACK & NECK PAIN

ESSENTIALS OF DIAGNOSIS

► Neck and back pain are the most common physical conditions for which patients seek medical attention.
► History and physical examination are paramount in diagnosis, as imaging is often falsely positive or negative.
► Finding the root cause of the symptoms is essential.

General Considerations

The chief complaint of spinal pain dominates in primary care offices and emergency departments across the United States. Once thought of as nuisance conditions associated with a speedy recovery, back and neck pain are now
known to produce lingering problems for many patients, with few returning to preinjury activity. Despite increasing awareness of this problem, the incidence of spinal pain remains at about 80%, with 60% of adults reporting neck or low back pain within the previous 3 months.

**Clinical Findings**

Pain is merely a symptom; therefore, clinicians must evaluate for anatomic and physiologic causes of the pain, many of which are discussed elsewhere in this chapter. Assessing for so-called “red flags” is essential (Table 31–1). These well-known and established indicators may require careful assessment, changes in treatment plan, or emergency care or referral to another health care specialist. Ignoring these “red flag” indicators increases the likelihood of patient harm. “Yellow flags” are psychosocial factors that have been shown to be indicative of long-term disability and can become a barrier to treatment (Table 31–2).

Table 31–1 “Red flags” in back pain.
<table>
<thead>
<tr>
<th>Cancer related</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of cancer</td>
</tr>
<tr>
<td>Pain at rest or nighttime</td>
</tr>
<tr>
<td>Weight loss</td>
</tr>
<tr>
<td>Age &gt; 50 or &lt; 17 years</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Infection related</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistent fever</td>
</tr>
<tr>
<td>Recent infection</td>
</tr>
<tr>
<td>Pain out of proportion to examination</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
</tr>
<tr>
<td>Immunocompromised state</td>
</tr>
<tr>
<td>Intravenous drug abuse</td>
</tr>
<tr>
<td>Recent spinal procedure</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cardiovascular related</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of abdominal aortic aneurysm</td>
</tr>
<tr>
<td>Pulsating abdominal mass</td>
</tr>
<tr>
<td>Atherosclerotic vascular disease</td>
</tr>
<tr>
<td>Age &gt; 60 years</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Related to spinal instability</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma (motor vehicle accident or fall from height)</td>
</tr>
<tr>
<td>If age &gt; 70 years, history of osteoporosis or fall from any height</td>
</tr>
<tr>
<td>Prolonged corticosteroid use</td>
</tr>
</tbody>
</table>

<table>
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<tr>
<th>Signs of neurologic compromise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cauda equina syndrome (saddle anesthesia, bowel or bladder incontinence)</td>
</tr>
<tr>
<td>Sensory or motor loss</td>
</tr>
<tr>
<td>Myelopathy</td>
</tr>
</tbody>
</table>

**Table 31–2** “Yellow flags” in back pain.
Treatment

The treatment of back and neck pain is dependent on etiology. The physiatrist must have a thorough knowledge of spinal disease in order to provide the best treatment for these patients. Although most acute exacerbations can be treated conservatively, a steady and steep rise in spinal procedures and surgeries has occurred.

Prognosis

The overall prognosis for spinal pain is poor. The pain may subside initially; however, the annual rate of recurrence is about 40%. Loss of function plays a large role in the poor prognosis. Up to 26% of patients with spinal pain report limitations in the amount or type work they can perform because of back pain. About 5% of the population state they cannot work at all because of health limitations resulting from back pain. Limitations seem to be most severe in senior citizens, leading to involuntary or early retirement. Work-related injuries present a particular challenge. Factors such as work restrictions, job satisfaction, and the presence of litigation affect the patient’s ability to return to work, and less than 1% of patients who are out of work for more than 1 year ever return.


| Negative attitude (ie, that back pain is harmful or potentially severely disabling) |
| Fear or avoidance behavior and reduction in activity levels |
| Expectation that passive, rather than active, treatment will be beneficial |
| Tendency to depression, low morale, and social withdrawal |
| Social or financial problems |
Spondylolisthesis & Spondylolysis

ESSENTIALS OF DIAGNOSIS

- Spondylolysis is a defect in the pars interarticularis.
- Spondylolisthesis involves the translation of one vertebra over another.
- They often occur together, commonly at the lumbosacral junction, producing sensory, motor, and reflex changes.

General Considerations

Spondylolysis is a defect in the pars interarticularis that is associated with spondylolisthesis in approximately 50% of cases. Spondylolisthesis is the slippage of one vertebral body with respect to the one beneath it. This most commonly occurs at the lumbosacral junction but can occur anywhere in the spine. Spondylolisthesis is classified based on etiology into six types (Table 31–3) and can be further graded based on the amount of vertebral subluxation in the sagittal plane (Figure 31–1).

Table 31–3 Classification of spondylolisthesis.
<table>
<thead>
<tr>
<th>Class</th>
<th>Type</th>
<th>History</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Dysplastic</td>
<td>Results from a congenital defect in the neural arch of L5 or S1. Symptoms first present in childhood, usually with radicular pain that worsens with extension and activity. Commonly seen at L5–S1.</td>
</tr>
<tr>
<td>IIa</td>
<td>Isthmic–spondylolysis</td>
<td>Results from a defect in the pars interarticularis either through repeated stress (IIa) or congenitally (IIb). Presents in childhood or young adulthood with axial or radicular pain, or both, often with a history of repeated flexion–extension–rotational stress (eg, gymnastics). Commonly seen at L5–S1.</td>
</tr>
<tr>
<td>IIb</td>
<td>Isthmic–pars elongation</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>Degenerative</td>
<td>Manifests later in life with a chronic and progressive pattern of axial pain and neurogenic claudication. Commonly seen at L4–5.</td>
</tr>
<tr>
<td>IV</td>
<td>Traumatic</td>
<td>Occurs with acute axial and or limb pain following trauma.</td>
</tr>
<tr>
<td>V</td>
<td>Pathologic</td>
<td>Insidious onset caused by generalized bone disease (cancer, infection, metabolic disorder) that leads to attenuation of the posterior elements.</td>
</tr>
<tr>
<td>VI</td>
<td>Postsurgical</td>
<td>Onset may be gradual or acute. Caused by prior surgical resection of the lamina or facet without fusion.</td>
</tr>
</tbody>
</table>
Table 31–3 summarizes key aspects of the disease process for various types of spondylolisthesis. Depending on the cause, onset of symptoms may be chronic or acute. Adequate workup must be completed to determine the pathologic causes of spondylolisthesis prior to initiating treatment with conservative methods.

Clinical Findings

A. Symptoms and Signs

In cases of severe slippage, neurologic compromise often produces motor, sensory, and reflex changes. Extension often exacerbates pain. Hyperlordosis of the lumbar spine along with hyperkyphosis of the thoracic spine may occur in compensation as the center of gravity shifts. A palpable step-off of the spinous process may be felt as one vertebra slides past another. Paraspinal muscle
hypertonicity, restricted range of motion, and root tension signs may be common.

B. Imaging Studies
Standing radiographs are used to evaluate and grade lumbar spondylolisthesis. Lateral and oblique radiographs may show a fracture of the pars interarticularis, a classic fractured collar of the “Scotty dog.” On flexion and extension views, sagittal translation of greater than 4.5 mm in the lumbar spine and 2 mm in the cervical spine indicates instability, while sagittal rotation of greater than 11 degrees on the cervical spine and 15 degrees in the lumbar spine indicates instability. Thin-slice computed tomography (CT) scan without contrast can be helpful to visualize the bony anatomy and isthmic defects, as well as the spinal canal. Magnetic resonance imaging (MRI) may be helpful in viewing the neural structures, and bone scan may be helpful in demonstrating acute fractures. Younger patients have a higher risk for progression of isthmic or congenital spondylolisthesis. Serial radiographic studies (standing lateral films, only) should be performed every 6 months to follow these patients. Progression rarely occurs after adolescence.

Differential Diagnosis
Once the diagnosis of spondylolysis or spondylolisthesis is made, emphasis should be placed on finding the underlying cause, especially if pathologic fracture is suspected.

Treatment
Most patients can be treated conservatively. If an isthmic lesion is acute, the patient should be restricted from provocative activities or sports until he or she is asymptomatic. Medications such as acetaminophen and nonsteroidal antiinflammatory drugs (NSAIDs) are often used to control the symptoms of pain associated with spondylolisthesis.

Physical therapy is an integral part of the patient’s rehabilitation process. The most accepted protocol includes activity and exercise that reduces extension stress. The goals of exercise are to improve abdominal strength and increase flexibility. Since tight hamstrings are almost always part of the clinical picture, appropriate hamstring stretching is important. Instruction in pelvic tilt exercises
may help reduce any postural component causing increased lumbar lordosis.

Bracing for acute spondylolisthesis is controversial, but it has been shown to reduce symptoms and to facilitate healing. A thoracolumbosacral spinal orthosis or modified Boston brace can be used for low-grade slips and is recommended for 3–6 months.

Interventions such as epidural steroid injections may be helpful in treating the radicular symptoms. The medial branch provides innervation to the facet joint as well as the lamina, so a medial branch block or ablation may be helpful in treating axial pain.

Surgery is indicated in patients with intractable pain or progressive neurologic deficits, or those in whom symptoms are resistant to nonoperative measures. Decompression laminectomy, with or without fusion, or interspinous spacers have been shown to reduce pain associated with degenerative spondylolisthesis. Traumatic spondylolisthesis often requires surgery. The goal of surgery is to stabilize the spinal segment and decompress the neural elements.

**Prognosis**

The degree of recovery is dependent on the type and degree of spondylolisthesis. Young patients with low-grade slips tend to do well with conservative care. Patients with degenerative disease and those who undergo surgery often continue to have symptoms.


Panjabi M: Clinical spinal instability and low back pain. J Electromyogr
SPINAL STENOSIS

ESSENTIALS OF DIAGNOSIS

- Back, buttock, or limb pain on standing or walking that is relieved with sitting.
- Cross-sectional imaging shows narrowing of the spinal canal.

General Considerations

Spinal stenosis is a degenerative process that is one of the most common painful conditions of the elderly, causing symptoms in more than 1.2 million Americans. Stenosis (a term derived from the Latin for “narrow”) occurs in the spine when the central canal (neural foramen) decreases in size, causing compression of the neurovascular structures. The natural curvature of the spine plays an important role in the signs and symptoms of spinal stenosis. Any tube will naturally narrow when it is bent. In the spine, the lordosis of the cervical and lumbar segments is reduced in flexion, and increased with extension. Thus, activities that promote extension of the spine at these levels, such as standing, produce symptoms.

Pathogenesis

Degenerative disc disease results in posterior disc bulging, which may compress the ventral and lateral aspects of the thecal sac. Ligamentum flavum hypertrophy causes compression dorsally, whereas facet joint hypertrophy causes lateral recess and neuroforaminal stenosis. Spinal stenosis is often acquired, although congenital variations, such as shortened pedicles, may cause symptoms in younger patients (Table 31–4).
Clinical Findings

A. Symptoms and Signs

Neurogenic claudication is the quintessential chief complaint of the patient with lumbar spinal stenosis and is described as pain in the buttocks and lower limbs with standing and walking that is relieved with sitting. The classic “shopping cart sign,” in which the patient leans forward on a cart while in the market, is often reported. Patients may be able to differentiate the symptoms with certain activities. Walking uphill is often better tolerated than walking downhill or on flat surfaces. Likewise, patients may be able to tolerate riding an exercise bike better than walking on a treadmill. Patient with cervical spinal stenosis often have pain in the neck and upper limbs. In more severe cases they become myelopathic, reporting spasticity, weakness, loss of balance, or even bowel or bladder incontinence. Careful note of previous surgery should be made, as degeneration adjacent to the level of previous spinal fusion may lead to iatrogenic spinal stenosis.

Physical examination findings are dependent on the severity of disease, with many patients having unremarkable examinations. Changes such as a forward-flexed posture while standing and walking may be observed in the office. Range of motion in the spine may be diminished with extension. In patients with more severe disease, neurologic symptoms can be seen, such as motor weakness, sensory loss, and hyperreflexia in the case of cervical stenosis, and hyporeflexia in stenosis of the lumbar spine.

Table 31–4 Classification of spinal stenosis.

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Idiopathic</th>
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</thead>
<tbody>
<tr>
<td>Achondroplastic</td>
<td>Osteopetrosis</td>
</tr>
<tr>
<td>Acquired</td>
<td>Degenerative</td>
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<tr>
<td></td>
<td>Iatrogenic</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Combined</td>
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</tbody>
</table>
**B. Imaging Studies**

Imaging of the spinal canal is essential for diagnosis of spinal stenosis. MRI has become the gold standard as the ability to visualize soft tissue plays a major role in spinal stenosis. The availability of standing and seated MRI allows examiners to see how stenosis is related to posture (Figures 31–2 through 31–5). When MRI is unavailable or contraindicated, CT may be used, preferably with myelography. It has been suggested that an anteroposterior canal diameter of less than 10 mm, or a cross-sectional area of less than 100 mm$^2$ defines spinal stenosis. A 50% reduction in canal diameter compared with adjacent levels also indicates spinal stenosis. Plain films offer little in terms of visualizing the central canal, although they may visualize the neural foramen. Flexion–extension views should be ordered to assess for dynamic instability.

▲ **Figure 31–2** Seated sagittal MRI scan of the lumbar spine showing a patent lumbar spinal canal.
\textbf{Figure 31–3} Seated axial MRI scan of the lumbar spine showing patent lumbar central canal and lateral recess.

\textbf{Figure 31–4} Standing sagittal MRI scan of the lumbar spine showing L4–5 disc bulging and ligamentum flavum hypertrophy resulting in spinal stenosis.
Figure 31–5 Standing axial MRI scan of the lumbar spine showing L4–5 posterior disc bulging and ligamentum flavum buckling resulting in severe central and lateral recess stenosis.

C. Other Tests

Electrodiagnostic testing is not part of the routine workup for spinal stenosis, although paraspinal mapping electromyography (EMG) may be predictive.

Selective spinal nerve block may help differentiate neurogenic claudication from other causes of limb pain in patients with spinal stenosis. This may also help to isolate the most significant lesion in patients with multilevel disease on imaging.

Treatment

For mild stenosis, over-the-counter pain medications (eg, acetaminophen and NSAIDs) may be useful. Muscle relaxants have little utility in treating neurogenic claudication. Neuropathic agents may improve symptoms in neuropathic pain, and gabapentin has been shown to improve pain and walking distance in patients with lumbar spinal stenosis. For patients with severe symptoms, opiates may provide the only means for relief, although special consideration is needed when prescribing potentially sedating medication to these patients, as they are already at increased risk for falls.
Physical therapy should be used in conjunction with other therapies as part of the comprehensive management of spinal stenosis. Attention should be given to stretching the tight ventral muscles of the core. A balanced strength program helps patients to maintain good posture and stand erect. Cardiovascular fitness provides the patient with endurance. Because spinal stenosis causes pain on walking, the use of a treadmill should be limited; however, the use of a stationary bike as well as a hand bike can be encouraged.

Rigid bracing may improve symptoms of lumbar spinal stenosis but is not recommended because it impairs mobility. Use of a flexible corset has been shown to be effective in increasing walking distance while it is worn, but there are no lasting benefits to the therapy once discontinued. Epidural steroid injections are useful in the treatment of neurogenic claudication. The use of a series (performed at regular intervals irrespective of the patient’s symptoms) of injections is not recommended, but the use of multiple injections throughout the course of treatment has been shown to provide durable and significant relief.

Once the only surgical option, lumbar laminectomy without instrumented fusion is still the recommended surgical treatment of choice, unless hardware is needed to prevent instability of the spine. The Spine Patient Outcome Research Trial (SPORT) found that patients with moderate to severe lumbosacral stenosis who underwent earlier surgical decompression had better outcomes than those who only underwent conservative treatment, although a complication rate of 9–13% and prolonged hospital course were noted. Recent advances, such as interspinous spacers and image-guided percutaneous laminotomy, are effective in increasing standing and walking tolerance and can be performed by interventional pain physicians as well as spine surgeons, often in an outpatient setting. Spinal cord stimulation is indicated for intractable pain of the trunk or limb and is a promising therapy for patients with stenosis. In patients with cervical and thoracic stenosis and signs of myelopathy, decompression should be considered early in the treatment course as permanent injury may occur.

**Prognosis**

Because spinal stenosis is a degenerative process, the overall prognosis is poor. However, with optimal treatment, many patients can lead full and productive lives.

Brown L: A double-blind, randomized, prospective study of epidural steroid


SPINAL INFECTIONS

ESSENTIALS OF DIAGNOSIS

- Rare but potentially life-threatening cause of severe spinal pain.
- May result in discitis, osteomyelitis, or epidural abscess.
Often caused by hematogenous seeding, but may occur following spinal procedures.

Pain is usually out of proportion to the physical examination.

Prompt MRI should be ordered if spinal infection is suspected.

General Considerations

Spinal epidural abscess has an estimated incidence of 0.2–2.8 cases per 10,000 per year, with the peak incidence occurring in people who are in their 60s and 70s. The most common pathogen is *Staphylococcus aureus*. Discitis can be defined as an inflammation of the vertebral disc space that is usually related to underlying infection. It should be considered in patients with vertebral osteomyelitis as these conditions are almost always present together. Discitis/osteomyelitis infections are usually categorized into pyogenic and nonpyogenic forms. *S aureus* is also the most common pyogenic organism, followed by *Staphylococcus epidermidis*, gram-negative bacilli, and *Mycobacterium tuberculosis* (Pott’s disease). Postoperative discitis accounts for approximately 30% of all cases of pyogenic discitis. Fungal, yeast, and parasitic infections are less common.

Pathogenesis

Spinal infections are normally a consequence of bacteremia, although often the patient does not have any prior signs of sepsis. The most common risk factor is diabetes, followed by trauma and intravenous drug abuse. Immunocompromised patients are at high risk. Although most cases involve patients without a history of spinal procedure, a recent spinal injection or surgery may be the cause. Surgical implants present a special risk, as bacteria tend to seed there. Radicular symptoms mark the progression of the abscess or infections, which could lead to neurologic deficit or paralysis.

Clinical Findings

Pain is usually out of proportion to physical examination. The open and continuous structure of the spinal canal allows the epidural abscess to spread as
it grows. Subsequently neurologic deficits may not be present initially, and symptoms may spread to other areas of the spine. Patients with discitis/osteomyelitis can present with back or neck pain varying from mild and insidious, to acute and severe. Unfortunately, adult discitis usually has a slow, insidious onset and it may take a few months or even years before a final diagnosis is confirmed. Patients may present with fever, although the nature of the infection (ie, being “walled off”) can prevent that. Tenderness at the posterior midline and paramidline region of the spine is the typical physical finding.

Imaging of the spinal canal is essential to the diagnosis and should not be delayed if infection is suspected. MRI with contrast is the gold standard, showing enhancement of the lesion. In the event an MRI is contraindicated or not available, a contrast-enhanced CT scan should be obtained. CT myelography is discouraged as this could spread infection into the subdural space. A tagged white blood cell scan is often not readily available but may be helpful if the other modalities cannot be utilized.

Laboratory studies that indicate infection, such as C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are usually elevated while the complete blood count may be normal; however, none of these findings are specific to spinal infection. The patient should be “pan”-cultured (ie, should have every culture run on every fluid) to evaluate for the suspected pathogen. Aspiration of an epidural abscess is not recommended as this might introduce infection into the subarachnoid space. If blood cultures are negative, culture of the abscess via laminectomy is the route of choice. In patients with discitis/osteomyelitis, it is often technically safer to perform a disc or vertebral body biopsy.

**Differential Diagnosis**

The midline pain associated with epidural abscess is usually severe and may mimic vertebral compression fracture. Facet joint or discogenic pain may also be considered.

**Complications**

Spinal infections lead to paralysis in as many as 34% of patients. Persistent pain or even death can occur if infection is not recognized early.
### Treatment

Spinal infections without neurologic sequelae may be treated medically. The patient should not be started on antibiotics until cultures are drawn or a biopsy is performed. Thereafter, they should be treated symptomatically for pain and sepsis. A rigid or semirigid orthosis may help control pain, and may be prescribed following surgery. Spinal injections are contraindicated. The majority of patients with epidural abscess will need to undergo surgery. Decompression laminectomy is indicated in the case of neurologic deficit or to obtain biopsy results. If decompression is performed over multiple levels, or in the cervical or thoracic spine, instrumented fusion is often indicated, adding to the morbidity of the procedure. Many factors, including the patient’s clinical picture, response to antibiotics, and extensiveness of the surgery, must be taken into account.

### Prognosis

Mortality from epidural abscess or discitis/osteomyelitis is approximately 15%. Patients who receive appropriate, early treatment usually make a full recovery.

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ESSENTIALS OF DIAGNOSIS

- Results from damage to the intervertebral disc with associated morphologic changes.
- Produces axial low back pain with or without radicular pain.
- Pain is provoked with activities that increase intradiscal pressure.

General Considerations

Discogenic low back pain is the cause of pain for almost 40% of chronic low back pain sufferers. The term discogenic means that the disc is the source of the patient’s pain. In some cases, anatomic changes occur that cause the disc to compress adjacent neural structures, causing pain (radiculitis) or loss of function (radiculopathy). In other cases, the anatomic changes allow for ingrowth of the neurovascular supply, causing the disc itself to be painful and results in nonradiating low back pain.

Pathogenesis

Discogenic pain usually falls into one of the following categories: (1) degenerative disc disease, (2) intervertebral disc displacement or internal disc disruption, and (3) disc herniation.

A. Degenerative Disc Disease

Degenerative disc disease starts as a result of the normal aging process, or by excessive or repetitive trauma, leading to disc dehydration and an increase in the collagen concentration. Disc degeneration can begin as early as the third decade of life. Aging, obesity, smoking, and excessive axial loads can accelerate the degeneration of the intervertebral discs, with age being the strongest association. Degenerative changes in the lumbar intervertebral discs are a common finding on imaging and may have little correlation with back pain. However, in patients with chronic low back pain, studies have found that disc degeneration was one of
the main reasons for their pain.

**B. Internal Disc Disruption**

Internal disc disruption is breakdown of the internal architecture of the disc that can lead to back as well as limb pain without signs of disc degeneration and disc protrusion, and without nerve root compression on imaging. Internal disc disruption is characterized by the presence of isolated radial fissures penetrating from the nucleus pulposus into the annulus fibrosis. The morphologic and biophysical features of internal disc disruption correlate strongly with back pain, as do certain MRI features. The presence of a single fissure distinguishes an affected disc from a normal disc. These fissures can be graded according to the extent to which they penetrate the annulus.

**C. Disc Herniation**

A *disc herniation*, also known as slipped disc, prolapsed disc, bulging disc, or herniation of the nucleus pulposus, is a broad term that includes three specific types of disc abnormalities. These abnormalities are differentiated based on the integrity of the posterior longitudinal ligament that reinforces the back of the disc as follows: protrusion, extrusion, and sequestration.

In a disc protrusion the posterior part of the disc is focally or eccentrically pushing backward into the anterior epidural space and has contacted, and even somewhat compressed, the traversing nerve root and front of the thecal sac. The posterior longitudinal ligament is still intact. Disc protrusions without nerve root compression are seen in about 30% of the normal nonsymptomatic population (*Figure 31–6*).
Figure 31–6 Sagittal MRI scan of the lumbar spine showing degenerative disc disease at L4–5, as demonstrated by loss of signal intensity within the nucleus pulposus, creating the “dark disc.” At L3–4 a contained disc protrusion is seen.

A disc extrusion is defined by the rupture of the posterior longitudinal ligament, allowing for further migration of the nucleus pulposus into the anterior epidural space. A disc extrusion is not typically seen in the asymptomatic person (Figure 31–7).
The final type of disc herniation, known as a sequestration, occurs when a fragment of nuclear material detaches itself from the main body of the disc and resides loosely in the epidural space. Sequestration-type disc herniations can be excruciatingly painful and, if centrally located, occasionally cause neurologic deficits requiring prompt intervention.

**Clinical Findings**

Most patients present with a complaint of axial low back pain with or without radiation to the limb related to the level of the affected disc. The pain may be progressive and longstanding, in the case of degenerative disc disease, or acute, in the case of internal disc displacement. Occupational factors, such as exposure
to vibrational energy or heavy lifting, may be present. Particular positions or activities can trigger the pain by increasing intradiscal pressure, particularly jumping, coughing, twisting, or flexing the spine.

Physical hallmarks include tenderness over the spinous processes and paraspinal muscles. Pain is often exacerbated with trunk flexion and relieved with extension of the spine. Maneuvers such as sustained hip flexion and the pelvic rock test are often provocative; however, neurologic examination is often normal unless there is compression of the neural structures.

Plain radiographs cannot specifically evaluate the disc, but they can show structural changes, such as loss of disc height, that could be suggestive of a herniated or degenerative disc. MRI is the test of choice in evaluating intervertebral disc pathology and also provides excellent imaging of the surrounding soft tissues. CT scan with or without myelography is used when MRI is not available or is contraindicated. Imaging must be carefully correlated with history and physical examination, as studies can often be falsely positive or falsely negative. MRI and CT scans detect disc herniation 20–36% of the time within an asymptomatic patient population.

Provocation discography with postdiscography CT remains the gold standard for confirming the diagnosis of discogenic low back pain. In this procedure, pain is reproduced by pressurizing the affected disc with a contrast material. Unfortunately, provocation discography may actually produce more damage to the disc or trigger a degenerative disc disease; therefore, it should be performed only in cases when further intervention is being seriously contemplated. The presence of “red flags” (see Table 31–1) in the patient’s history or physical examination should prompt an early imaging study.

**Treatment**

Bed rest should be limited to acute and severe cases and last no longer than 2–3 days as inactivity can potentiate prolonged disability and produce even more pain. Treatment goals focus on restoring strength, flexibility, and function previously lost because of immobility. The physical therapy prescription should include stabilization of core muscles, emphasizing neutral to extension bias. Physical modalities may be helpful in alleviating localized back pain.

Proper spinal bracing can help relieve pain by preventing painful movement or further spinal deformities but does not alter the natural course of the disease. Traction is commonly recommended to increase the foraminal dimensions, reduce intradiscal pressure, and, thus, relieve radicular pain caused by a
herniated disc. Manipulative therapy is designed to maximize motion and decrease pain, but long-term studies of efficacy are lacking.

NSAIDs are first-line agents in the treatment of discogenic low back pain but may be contraindicated in some cases because of their side effects (ie, gastrointestinal bleeding, allergic reactions, and renal or liver impairment). Opiates and muscles relaxants are frequently prescribed for severe pain but often have concomitant sedatives effects, and their long-term efficacy for discogenic back pain has not been established.

Epidural steroids injections are effective in providing symptomatic relief of radicular pain and are offered to patients who have not responded to conservative measures. Surgery is indicated for progressive or worsening neurologic symptoms. Standard open discectomy is the most common surgical approach unless there are signs of instability, for which fusion would be necessary.

Regenerative treatment strategies designed to reverse or inhibit disc degeneration include the administration of growth factors, autologous or allogenic cells, gene therapy, and the introduction of biomaterials, and are undergoing clinical trials.

Anderson D, Tannoury C: Molecular pathogenic factors in symptomatic disc degeneration. Spine J 2005;5:260S–266S.


SPINAL FRACTURES

ESSENTIALS OF DIAGNOSIS

► May occur anywhere in the spine (cervical, thoracic, lumbar, sacral, coccygeal), with thoracic fractures being most common.
► Diagnosed by imaging studies (radiographs, CT, or MRI).
► Patients should be evaluated for osteoporosis.

General Considerations

Of the three major categories of spinal fractures—fragility, traumatic, and pathologic—fragility fractures account for the vast majority. Fragility fractures are defined as fractures that occur when the elastic resistance of bone is inadequate to withstand the stresses of normal activity. A fracture that occurs following a minor fall (ie, less than standing height) is not considered traumatic, and osteoporosis should be suspected in patients with risk factors. Likewise, a metabolic disorder or malignancy should be suspected in those without risk factors for osteoporosis.

Clinical Findings

The vertebral levels most prone to osteoporotic compression fractures are T7 and T8, followed by T12 and L1. Most osteoporotic vertebral compression fractures (VCFs) are asymptomatic and tend to be found incidentally when imaging is performed for other reasons. Symptomatic VCFs tend to be characterized by localized pain in the midline of the spine, although pain can be referred to more caudal levels. A loss in total body height, or the development of a stooped-over posture or kyphosis (also called a “dowager’s hump”) may be reported. Altered biomechanics and a shift in the center of gravity may result in increased loads across the posterior elements, causing facet joint pain. The most likely finding on physical examination is tenderness over the spinous process of the involved vertebra. Neurologic deficits are unlikely, unless there is retropulsion of bony
fragments into the canal, which is more common in traumatic and pathologic fractures.

Plain radiographs of the thoracolumbar spine (anteroposterior and lateral) should be ordered initially and are generally sufficient in diagnosing and characterizing the fracture (ie, wedge, concave, crush, burst, etc). Axial imaging such as CT scan provides superior detail for visualizing the bony anatomy, and is helpful in evaluating for retropulsed fragments and assessing the pedicles (Figure 31–8). MRI is also helpful in differentiating between acute and chronic compression fractures (Figure 31–9). As part of the workup for osteoporosis, dual-energy X-ray absorptiometry (DEXA) should be ordered.

▲ Figure 31–8 Axial CT scan of the lumbar spine showing intact pedicles with retropulsion of the posterior cortex.
Figure 31–9 Sagittal MRI scan of the thoracic spine. Stir images contrast acute (bright signal) and chronic (low signal) compression fractures.

Sacral insufficiency fractures are often painful over the sacral ala, sacroiliac joint, and buttock and are aggravated by weight-bearing activity and relieved by rest. MRI is the best imaging modality for detecting bone marrow changes and edema, which will be present in acute fractures. Bone scintigraphy may demonstrate the classic “H-shaped” pattern, which is pathognomonic for
bilateral sacral insufficiency fracture, but this test may not be as sensitive or specific as MRI.

Laboratory studies are important for diagnosing osteoporosis and ruling out more sinister etiologies, such as osteomyelitis, metabolic disorders (eg, hyperparathyroidism), or malignancy. In cases of suspected malignancy or metabolic disease, a vertebral body bone biopsy should be performed.

**Differential Diagnosis**

Other sources of pain include sacroiliac joint dysfunction, spinal tumors, vertebral osteomyelitis, fracture or contusion of the spinous process, and intervertebral disc pathology.

**Treatment**

The treatment of underlying osteoporosis is paramount. Bisphosphonates inhibit bone turnover, and therefore should be discontinued following an acute VCF. A 4-week course of calcitonin, which may also have an analgesic effect during the acute phase following a VCF, is recommended in recent guidelines by the American Academy of Orthopedic Surgeons (AAOS). To treat the pain associated with the fracture, medications such as NSAIDs, acetaminophen, muscles relaxants, and opiates can be utilized. Caution must be taken with sedating muscle relaxants and opiates in the geriatric population as there is a high risk of falls.

Manual therapies and modalities help to reduce pain during the acute phase. A core strengthening program with a neutral or extension bias should be initiated. Bracing options include corsets and hyperextension braces, such as the cruciform anterior spinal hyperextension (CASH) or Jewett brace. However, caution must be used in cases of severe osteoporosis as there may be an increased risk of posterior element fracture.

Transforaminal epidural steroid injections may be helpful in the treatment of pain associated with VCFs, especially if there is a radicular component. Because biomechanical changes often affect the posterior elements, facet joint injections may be indicated.

The development of improved vertebral augmentation systems has enhanced patients’ access to these procedures, as they can be performed by orthopedic surgeons as well as interventional pain physicians. Both vertebroplasty and
kyphoplasty are deemed safe and efficacious, and may result in noticeable improvements in pain, disability, and quality of life. The most recent guidelines from the AAOS recommend strongly against the use of vertebroplasty, but weakly support the use of kyphoplasty, in the treatment of osteoporotic VCFs. The evidence supporting vertebroplasty and kyphoplasty is somewhat stronger in the setting of tumor-related VCFs.

Physical therapy is commonly employed in the treatment of sacral insufficiency fractures. During the acute phase of the fracture, weight bearing may not be tolerated. Emphasis should be given to transfers, bed mobility, wheelchair mobility, and use of assistive devices. As the patient progresses, gait training should be employed. Sacroplasty has been shown to be effective in patients who do not respond to the preceding measures.

In cases of unstable fractures, spinal cord compression, or neurologic deficits, the patient should be referred to the neurosurgery or orthopedic-spinal surgery department for open decompression and fusion.

Prognosis

Most patients have significant improvement in pain within 3 months, but up to 40% continue to have disabling pain 1 year later. For this reason, percutaneous vertebral augmentation is increasingly being offered as an early treatment option. Prevention remains the best treatment for spinal fractures, and behaviors that maximize bone density and minimize fracture risk are therefore important.


ESSENTIALS OF DIAGNOSIS

► Most spinal neoplasms are extradural and metastatic.
► Signs, symptoms, and risk factors for malignancy should be investigated.
► MRI is the imaging modality of choice, but definitive diagnosis is by biopsy.

General Considerations

Spinal tumors are classified by their location as extradural, intradural, or intramedullary. Extradural tumors are the most common, comprising about 60% of all spinal tumors, and the majority of these are metastatic (25 times more common than primary), usually originating from the lung, breast, prostate, kidney, colon, or thyroid.
Metastasis typically occurs hematogenously via Batson’s venous plexus into the vertebral body. Most metastatic lesions are osteolytic in nature, but metastasis from the breast, lung, and prostate may be osteoblastic. The most common primary extradural neoplasm is multiple myeloma, which causes lytic lesions. Rarer entities include plasmacytoma, osteoblastoma, and chordoma. Primary benign neoplasms are also rare, the most common being osteoid osteoma, which affects the pedicles. Others include eosinophilic granuloma, aneurismal bone cyst, giant cell tumor, ependymoma, neurofibroma, lipoma, and meningioma.

Clinical Findings

The patient complains of pain localized to the affected vertebrae. Classic “red flag” symptoms of malignancy include pain that is worse at night, interfering with sleep. The physician should inquire about a prior personal and family history of cancer, as well as history of smoking or known exposure to other carcinogens. The review of systems should include questions about night sweats, unintentional weight loss, cough, hemoptysis, breast discharge or mass, constipation, and rectal bleeding.

Examination of the spine may reveal tenderness over the affected vertebrae. Neurologic deficits or myelopathy may be present, depending on the size and location of the tumor. In an effort to identify a possible source of metastasis, the physician should also complete a pulmonary, thyroid, breast, prostate, and rectal examination (including Hemoccult testing).

Tumor invasion may cause elevations in alkaline phosphatase as well as inflammatory markers, such as ESR and CRP. However, the primary purpose in laboratory testing is to diagnose and characterize the malignancy present. Biopsy is ultimately required to definitively diagnose and characterize the tumor.

Plain radiographs will not be diagnostic until bony destruction involves 50% of the vertebrae. CT scan is excellent for detecting small bony lesions, and contrast can be used to differentiate fatty infiltration and hemangioma from malignant soft tissue. However, MRI with gadolinium contrast is the imaging modality of choice for assessing spinal tumors, as it provides superior visualization of the vertebrae and soft tissues (Figure 31–10). In cases of neurologic compromise or cord compression resulting from suspected epidural involvement, MRI with contrast should be ordered immediately. If MRI is contraindicated or unavailable, then a CT myelogram is the next best option. A bone scan can also be helpful but may be falsely negative in cases of osteolytic lesions, such as multiple myeloma. Further imaging may be required to locate
the primary tumor or identify additional areas of malignancy.

**Figure 31–10** Sagittal MRI scan of the thoracic spine showing a pathologic wedge compression fracture with lytic lesions in the adjacent vertebrae.

### Differential Diagnosis

The differential diagnosis for a space-occupying lesion within the spine includes osteomyelitis, epidural abscess, hematoma, syrinx, Schmorl’s node, and reactive edema resulting from trauma or degenerative processes.
Treatment

The approach to the management of spinal tumors is two-pronged, consisting of pain relief and treatment of the cancer itself, and requires a comprehensive approach across specialties. Pain can initially be addressed with medication management and may require significant doses of opioid medications while radiation and chemotherapy options are considered.

Referral should be made to physical and occupational therapies for a preventative, supportive, or palliative rehabilitation program, depending on the patient’s prognosis. Emphasis should be placed on improving endurance, work simplification, and energy conservation. Bracing may be indicated before or after surgery for patients with instability, and may also provide additional comfort.

Epidural steroid injections may be considered in cases of associated radiculitis. Vertebral body fractures can be treated with kyphoplasty or vertebroplasty to restore vertebral height and reduce pain. Intrathecal opiate delivery is indicated for malignant pain in patients who cannot tolerate the adverse effects of oral opiate medications.

Surgery is indicated for spinal instability or neurologic compromise, when decompression and stabilization are required.

Prognosis

The prognosis for survival is widely variable, depending on the type and staging of the neoplasm. In addition, the prognosis for pain management can vary based on the size and location of the tumor.


ZYGAPOPHYSEAL JOINT ARTHROPATHY

ESSENTIALS OF DIAGNOSIS

- Paraspinal neck or back pain, with or without referral to the limb girdles or proximal limbs.
- Tenderness to joint palpation; pain with extension maneuvers.
- Joint injections or medial branch blocks are useful in diagnosis.

General Considerations

The zygapophyseal (or facet) joints are a frequent generator of pain throughout the spine. These joints comprise the superior articulating process from below, and the inferior articulating process from above. Depending on their orientation, they function to restrict and facilitate certain planes of motion throughout the spine.

Pathogenesis

Spondylosis is a general term referring to a degenerative process that can affect any area of the spine. It typically begins with intervertebral disc degeneration, which then leads to increased loads being placed on the facet joint, causing pain.

Clinical Findings

The patient typically complains of sharp or aching pain in the axial neck or back. On physical examination, range of motion may be reduced and painful within the affected area of the spine. Spinal extension, which increases the load bearing through the facet joint, is often provocative.
Facet arthropathy can be demonstrated on plain radiographs, CT, and MRI by visualization of joint space narrowing, sclerosis, or hypertrophy of the superior or inferior articulating processes, or both. These details are most clearly seen on axial CT or MRI images (Figure 31–11).

▲ Figure 31–11 Axial MRI scan of the lumbar spine showing significant facet joint arthropathy.

Fluoroscopically guided intraarticular facet joint injections and medial branch blocks may be diagnostic as well as therapeutic. The interventionalist must remember the innervation of the facet joints when planning out medial branch blocks.

▶ Differential Diagnosis

The differential diagnosis includes spondylolysis, pedicle fracture, paraspinal muscle strain, degenerative disc disease, and intervertebral disc displacement. In the thoracic and lumbar spine, it also includes costovertebral joint pathology and sacroiliac joint dysfunction.
Treatment

Initial treatment should include NSAIDs or acetaminophen. A short course of muscle relaxants or opioid analgesics may be indicated during acute pain flares. Physical therapy can be helpful in improving range of motion throughout the affected segments, with emphasis on flexion-biased core stabilization exercises. Bracing and orthoses are not indicated in the treatment of facet arthropathy.

Image-guided controlled facet joint blocks are the gold standard for the diagnosis of facet joint–mediated pain. For patients who obtain relief with facet joint blocks, radiofrequency thermal ablation of the medial branches may provide up to 12 months of relief.

Prognosis

Although facet arthropathy is a chronic, degenerative process, with available treatment the patient’s pain and function can often improve.


Sacroiliac Joint Dysfunction

Essentials of Diagnosis

- Pain localized to the sacroiliac joint or to the region of the posterior superior iliac spine.
- No physical maneuver or motion test accurately makes the diagnosis.
Diagnosed by fluoroscopically guided sacroiliac joint injection and not radiographically.

General Considerations

Patients frequently present with pain localized to the low back and buttocks. Sacroiliac joint dysfunction accounts for 10–30% of all cases of chronic mechanical low back pain.

Pathogenesis

Sacroiliac joint dysfunction may occur due to excessively high acute forces or chronic repetitive loads that lead to stress reaction between the sacrum and ilium. The sacroiliac joint may be affected in various rheumatologic disorders, including seronegative arthropathies. A lumbosacral transitional vertebra, defined as total or partial, unilateral or bilateral fusion of the transverse process of the lowest lumbar vertebra to the sacrum, may be present. Six to 31% of cases may be painful (Bertolotti syndrome); however, most patients are asymptomatic.

Clinical Findings

The classic presentation of sacroiliac joint dysfunction is pain localized to the region of the sacroiliac joint or to the region of the posterior superior iliac spine (PSIS). Sacroiliac joint pain is typically dull and aching in quality, with occasional radiation to the buttocks, groin, or proximal posterior thigh. Less frequently patients report pain in the hip or calf. It is often unilateral but presents bilaterally in approximately 20% of patients. Prolonged sitting or bending is provocative, whereas standing and walking is palliative. Patients may describe worsening pain while stair climbing or rising from a seated position. The complaint of pain in the morning that improves with exercise is suggestive of inflammatory disease.

Neurologic signs are absent in sacroiliac joint dysfunction. Inspection of the low back, pelvis, and lower limbs should be performed to help identify the source of pain. Attention should be paid to a leg-length discrepancy. Patients with sacroiliac joint dysfunction may demonstrate tenderness over the sacral
sulcus and posterior sacroiliac joint. Provocative maneuvers, including Gaenslen’s test, Patrick’s test, pelvic distraction, iliac compression, or sacral thrust, may be positive although no single maneuver accurately makes the diagnosis. Sacroiliac joint dysfunction is not diagnosed radiographically, and imaging studies may be considered to rule out other pathologies. A fluoroscopically guided joint injection is the gold standard in diagnosing sacroiliac joint dysfunction.

**Treatment**

NSAIDs and acetaminophen are the first-line medications of choice. Muscle relaxants or opioid medications may help ameliorate the symptoms of acute spasm pain. Physical therapy should include joint mobilization and manipulation, as well as exercises to address flexibility and strength in the quadratus lumborum, quadriceps, adductors, and hamstrings. Biomechanics and postural training are emphasized, while avoiding those activities that are found to exacerbate symptoms. A sacroiliac joint belt helps to reduce pain by decreasing joint motion and providing proprioceptive feedback. A shoe lift or modification can be fitted to address a possible leg-length discrepancy (see Chapter 28).

Image-guided intraarticular joint injections with local anesthetic and corticosteroid may be both diagnostic and therapeutic. If sacroiliac joint injections are diagnostic but fail to provide long-term therapeutic relief, the patient may be a candidate for radiofrequency ablation of the nerve supply to the sacroiliac joint via the dorsal ramus of L5–S4. Because the sacroiliac joint receives innervation from the both the dorsal and ventral rami, significant improvement can be expected in about 60% of patients.

In cases refractory to more conservative treatment, sacroiliac joint fusion can be considered (Figure 31–12). Instrumentation with or without bone grafting has been utilized. The goal of fusion surgery is to stabilize the joint in its intended position. Success rates are favorable only in carefully selected patients whose pain is confirmed, through diagnostic blocks, to originate at the sacroiliac joint. Postintervention, patients will have partial weight-bearing restrictions, necessitating use of walker or crutches.
Prognosis

The prognosis for patients receiving available treatment for sacroiliac joint dysfunction is favorable.


TARLOV CYSTS

ESSENTIALS OF DIAGNOSIS

- Often asymptomatic, although patients may present with low back pain, buttock pain, tailbone pain, or radicular pain.
Physical examination findings depend on the size of the cyst.
Diagnosed radiographically by MRI.

General Considerations

Tarlov cysts are perineural cysts that are frequently located in the sacrum at the S1–4 region of the spinal cord. Between 15% and 30% of Tarlov cysts are symptomatic.

Pathogenesis

The cysts are filled with cerebrospinal fluid (CSF) and may be differentiated from other meningeal cysts by their nerve fiber–filled walls. The cysts arise from the junction of the dorsal ganglion and nerve root between the endoneurium and perineurium.

Clinical Findings

Patients may present with low back pain, buttock pain, tailbone pain, or radicular pain. Headache or abdominal pain have also been reported. Symptoms may be progressive as Tarlov cysts can increase in size over time, potentially causing complications and eroding the surrounding bone tissue. Weakness, paresthesias, and urologic or sexual dysfunction may occur as a result of larger cysts impinging on nearby structures.

MRI demonstrates isointense CSF on pulsed sequence. Cysts larger than 1.5 cm in diameter will usually be symptomatic. However, most cysts are asymptomatic and are found incidentally during CT or MRI examinations for other reasons (Figure 31–13). On myelography and postmyelography CT there is delayed filling of the cyst with contrast, reflecting the absence of a direct connection with the subarachnoid space.
Figure 31–13 Sagittal MRI scan showing an incidental Tarlov cyst posterior to the S2 vertebral body.

**Treatment**

Interventions such as medications, epidural steroid injections, and sacral dorsal ramus blocks may be used to control symptoms of pain associated with Tarlov cysts. Surgical intervention should be reserved only for cases in which there is intractable pain or neurologic deficits. Cyst aspiration may be carried out, although the success is often temporary because of recurrence. Direct surgical excision can also be performed and is often accompanied by laminectomy.

**Prognosis**
The prognosis is favorable for patients with smaller cysts. Those with larger, symptomatic cysts requiring intervention often have recurrence of the cyst with persistent symptoms.


Trauma Rehabilitation

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Trauma is a major cause of disability and death for individuals in the United States. In fact, trauma from unintentional injuries, homicide, and suicide is the leading cause of death for Americans aged 1 to 44 years. Injuries associated with trauma account for approximately one third of all emergency department visits and 8% of all hospital stays in trauma care systems. The top four mechanisms of traumatic injuries involve falls, motor vehicle accidents (struck by or against another individual), and other transportation accidents. The mechanism of injury varies in different age groups. For those younger than 7 years or 75 and older, falls account for 40% of traumatic injuries; but in those aged 15–33 years, motor vehicle injuries account for 27% of the injuries, peaking around age 19.

Based on these numbers and the burden on the U.S. health care system, one can understand the need for integrated trauma care programs. In the U.S. civilian health care system, there has been and continues to be a growth of trauma care systems. A trauma center is a hospital that has the resources and equipment to help care for severely injured patients. The American College of Surgeons (ACS) Committee on Trauma classifies levels of trauma centers from 1 to 4, with a level 1 trauma center providing the highest level of trauma care. The designation of trauma facilities is a geopolitical process that only the government is empowered to delegate; however, the ACS verifies the presence of the resources listed in the Resources for Optimal Care of the Injured Patient. ACS certification is a voluntarily process undertaken by participating hospitals. Having access to specialized resources and equipment is critical for severely injured patients. Research supported by the Centers for Disease Control and
Prevention shows that there is a 25% reduction in the death rate for severely injured patients who received care at a level 1 trauma center rather than a nontrauma center.

**THE ROLE OF THE REHABILITATION PHYSICIAN IN TRAUMA CARE**

With more people today surviving severe traumatic injuries, it is essential that rehabilitative services be made available throughout the acute and postacute hospital care system. The rehabilitation process for acute trauma patients should be initiated as early as possible in the hospital setting. Many of these patients have specific medical and rehabilitative needs that can benefit from the expertise of rehabilitation physicians in an acute care hospital setting.

The role of rehabilitation physicians in an acute care hospital can differ somewhat from their role in an inpatient rehabilitation facility. In the inpatient facility, the rehabilitation physician serves as the team leader; in the acute hospital setting, the rehabilitation physician is a valuable team member whose importance and input may vary based on the patient’s specific injury, time since injury, and clinical course. For patients with recent multitrauma injury who are being treated in an intensive care unit, the primary role of the rehabilitation physician may consist of an initial evaluation, documentation of the patient’s injury, and deciding what, if any, therapy can be initiated based on the patient’s status (Tables 32–1 and 32–2). This information is useful for future rehabilitation planning, prognosis, and care.

**Table 32–1** History of present illness: information of special relevance for trauma rehabilitation.
### Table 32–2 Focused review of systems: information of special relevance for trauma rehabilitation.

- **Mechanism of injury**
- **Type of injury**
- **Treatment in the field (prehospital care)**
- **Hospital course**
  - Emergency department
  - Intensive care unit
  - Step-down unit
- **Interventions**
  - Surgical
  - Medical
- **Testing**
  - Radiographic
  - Laboratory studies (eg, drug and alcohol screen)
  - Physiologic
- **Rehabilitation status**
  - Spinal clearance
  - Neurologic clearance
  - Fracture status (eg, weight bearing; range of motion)
  - Skin precautions
- **Pertinent medications**
In the acute care hospital, the team of rehabilitation providers may consist of members who spend variable amounts of treatment time with each patient. Physical, occupational, and speech therapists, social workers, and psychologists also may have varying degrees of expertise with certain types of injuries. It is important that the rehabilitation physician recognize and understand these team variables and potential limitations as certain therapists may require greater instruction on specific care precautions than others. The initial team focus is often preventive in nature, centered on preventing morbidity associated with immobility, positioning, nutrition, or other specific diagnostic issues. Monitoring
of a patient's changing status is essential, as adjustment of therapy orders and medical intervention may be required.

Often the rehabilitation physician serves to educate patients, their families, and other team members about the patient’s injury and future functional course. At discharge from the acute care hospital, the most important role of the rehabilitation physician may be to assist with discharge placement, whether in a postacute care facility or the home setting. This requires matching the patient’s medical and functional needs with available patient and community resources (see later discussion).

American College of Surgeons: Trauma Programs. Verified trauma centers. 

Centers for Disease Control and Prevention: Access to trauma care: Getting the right care, at the right place, at the right time. Available at: 

National Trauma Data Bank: 2013 Annual Report. 

**EVALUATION OF THE PATIENT REQUIRING TRAUMA REHABILITATION**

The initial evaluation of the trauma patient focuses on rapidly assessing key systems and organs necessary for sustaining life and maintaining function. Once the patient’s condition is stabilized, ongoing evaluation seeks to clarify the extent of an individual’s injuries, as well as their current and future functional status.

**Spinal Clearance**

If there is concern that a patient may have spinal trauma, a process for documenting spinal clearance is implemented. At many centers this is protocol driven, and patients undergo a spinal series with review and clearance by a specified physician. The spinal clearance protocol usually begins with plain radiographs to identify fractures by looking at the alignment of the bones and at
adjacent soft tissue outlines. When indicated, computed tomography (CT) scanning may also be used. CT provides better imaging of bones when evaluating for fractures and is more sensitive than plain radiographs. Magnetic resonance imaging (MRI) scanning can be used for evaluation of soft tissues and potential ligament injuries. Although the task of giving final spinal clearance usually rests with the primary team, it is important that the rehabilitation physician understand which films have been completed. If there are questions or concerns about spinal stability, the physician should communicate directly with the primary team. Spinal clearance must be obtained before initiating patient mobilization or therapy services.


### Head

Cerebral imaging results for the acute patient should be reviewed initially and followed over time. Ongoing studies may assist in tracking patient status, presence of edema or blood, or other intracranial processes. Care providers may need to be reminded that a negative cerebral imaging study does not rule out an underlying head injury. (See Chapter 13 for further details.)

### Pulmonary Function

Many trauma patients have some injury that affects their respiratory system either directly or indirectly. These patients may require mechanical ventilation as a result of severe head trauma, high cervical injuries, or multiple other injuries requiring medical management. Patients with multiple rib fractures or other direct trauma often require insertion of chest tubes. Once the chest tube is in place, negative pressure is maintained either by wall unit suction or by placing the tube into a water seal. The negative-pressure suction system draws fluid out of the pleural space. The chest tube and its water seal container are a closed system and allow for one-way movement of air and liquid out of the chest. Proper functioning of the latter system relies on having the appropriate amount of fluid in the water base chamber and keeping the chamber below the level of the tube insertion site. If the container tips over or is placed above the level of
the tube (eg, during patient care or mobilization therapy), the negative-pressure system can be disrupted, which may cause backflow into the pleural space. Clamping of the tube can also disrupt the negative pressure and should be avoided when possible. Therapists need to be aware of this precaution when mobilization is initiated.


Gastrointestinal Function & Nutritional Status

Patients with severe trauma develop hypermetabolic rates. For example, individuals with spinal cord or head injuries have marked increased nutritional needs in the acute phase of their recovery. Often complicating this increased nutritional requirement is the fact that patients have difficulties with oral intake owing to reduced level of consciousness, medications, mechanical ventilation, direct facial and neck injury, or other problems. This issue must be addressed early in the patient’s care to ensure that nutritional needs are met. Consequently, the rehabilitation team should be familiar with the different options for nutritional delivery, as well as any treatment-related precautions. Special caution must be taken with patients who receive non-oral feedings, as they may still be at risk for aspiration. Patients should be positioned with the head of the bed elevated during feedings to reduce this risk.


Genitourinary Function
Intravenous fluids and indwelling Foley catheters are often required during the acute phase of trauma care. Whenever medically possible these catheters should be discontinued; however, prior to catheter removal, the clinician must understand why the catheter was inserted. In patients with penetrating or urethral injuries, it is often appropriate to insert either a urethral or a suprapubic catheter and leave it in place during the acute recovery phase. The Foley catheter is often maintained in patients with spinal cord or head injuries during their acute hospitalization. After catheter discontinuation, the patient is monitored for any problems with urgency, dysuria, or voiding. Urinalysis, postvoid residuals, or urologic evaluation may be required for bladder management. These principles can be applied to trauma patients with prolonged hospital stays.


**Skin Integrity**

All trauma patients are immobilized for a period of time after the initial injury. It is important to understand the complications associated with loss of mobility in the trauma patient, particularly those with severe injuries. Excessive time and pressure on any skin surface can lead to the development of pressure ulcers. Any prolonged pressure to the skin that exceeds tissue capillary pressures of 32 mm Hg creates a critical pressure interface. Although the amount of pressure is a critical factor, so, too, is the length of time the pressure is applied. When pressure far exceeds systolic blood pressures, the development of necrosis is hastened. However, even pressures below the level of systolic blood pressure, if applied for long enough periods, can result in skin breakdown. The need to avoid excessive capillary pressure and nonrelief of pressure underlies the principle of turning patients every 2 hours. Other factors that place the trauma patient at risk for developing pressure ulcers include chronic hypoxia; edema, resulting in impaired oxygen and nutrition exchange; and neurologic issues.

Nursing staff must pay particular attention to proper patient positioning and bed movement in immobilized patients. A patient who is placed in a reclining position in a hospital bed with the head elevated can slide, creating sheer forces across the sacrum that result in skin breakdown. An initial evaluation of the patient’s bony prominences and sacral areas is imperative, along with periodic
Functional Changes Associated with Prolonged Hospitalization

Multiple changes in organ system physiology are induced by inactivity; these are collectively termed deconditioning. Studies have shown that during bed rest, muscle strength declines by 10–20% per week. Loss of strength occurs to a greater extent in the lower limbs than in the upper limbs. Other concerns in the bedridden patient include osteoporosis, joint contractures, increased heart rate (by 0.4 beats/min per day), orthostatic hypotension, anxiety and depression, skin breakdown, and shallow breathing pattern. (See Chapter 5 for additional discussion.)

To minimize the effect of these changes, it is important to start physical and occupational therapy as soon as the patient is medically stable and able to participate. Studies have shown that patients who received early physical therapy were out of bed earlier, had a shorter stay in an intensive care unit, shorter hospital length of stay, and increased survival-to-discharge rate than patients in whom physical therapy was initiated later. Furthermore, patients with acute respiratory failure who received early mobilization in the intensive care unit were less likely to die or to be readmitted in the first year following discharge.

In addition to the muscle weakness and atrophy that occur with bed rest, approximately one third of patients in intensive care units develop neuromuscular disorders, predominantly critical illness polyneuropathy (CIP) and critical illness myopathy (CIM). Both disorders tend to manifest as weakness associated with difficulty in weaning the patient from the ventilator. Major risk factors for CIM include severity of illness and use of intravenous corticosteroids. The major risk factor for CIP is sepsis. Although symptoms of both disorders are similar, involving diffuse weakness, CIP tends to cause primarily distal weakness, whereas CIM generally causes more proximal weakness. Additionally, there is a sensory component to CIP that is absent from pure CIM. Electrodiagnostic findings may be apparent as early as 1–2 weeks
after injury in patients with CIM, and by 2 weeks in those with CIP.


**COMMON TRAUMATIC INJURIES REQUIRING REHABILITATION**

The disorders discussed in this section are covered in depth elsewhere in this book. The discussion here focuses on the role of the rehabilitation physician in the acute care hospital setting. Readers are also referred to Chapters 12 and 31 for additional discussion of spinal cord injury and rehabilitation; 13 and 20, for traumatic brain injury in adults and children; 26, for lower extremity amputation; 27, for upper extremity amputation; and 30, for musculoskeletal disorders.

**1. Spinal Cord Injury**

The acute and long-term care of patients with a spinal cord injury is detailed in Chapters 12 (spinal cord injury) and 31 (spinal rehabilitation) of this book. The primary issue for any patient after a spinal cord injury is spinal and medical stabilization. Once this is achieved, evaluation of potential associated injuries, such as head injuries, long bone fractures, and other trauma, is addressed. Specific issues considered by the acute and trauma rehabilitation consultant are
tailored to the injury level and completeness of injury. For example, patients with complete high cervical injuries are at a greater risk for pulmonary complications and cardiac arrhythmias than those who have incomplete lower lumbar injuries. As with all patient care, treatment must be individualized based on the patient’s injury.

Clinical Evaluation Concerns

It is essential to perform a full examination in accordance with the International Standards for Neurological Classification of Spinal Cord Injury (ISCSCI). The patient’s spinal cord injury should be classified as either complete or incomplete based on the presence or absence of function of the lower sacral cord segments. Patients with incomplete lesions usually have clinical findings that are consistent with previously documented incomplete injury syndromes (see Chapter 31). Patients with complete cervical thoracic injuries may lose all sympathetically mediated reflexes below the level of the injury, a condition known as spinal shock. This condition may last a variable number of days postinjury and presents as areflexia and loss of tone below the injury.

Complications & Trauma Care Issues

A. Respiratory Function

All patients with a cervical or high thoracic injury must be monitored for respiratory complications. Patients with complete cervical injuries (C5–7) have decreases in tidal volumes, vital capacity, and negative initiatory force to less than one third of normal following injury. Pulmonary function may also be decreased in those with thoracic injuries due to loss of innervation of the intercostal musculature. The rehabilitation team must pay close attention to issues of pulmonary toilet and secretion management. The use of intermittent positive-pressure breathing (IPPB), bronchodilators, assisted coughing techniques, and abdominal binders, along with close monitoring of pulmonary function, is recommended (see Chapter 31).

B. Cardiovascular Function

Bradycardia is a risk in patients who have unopposed vagal tone. Patients must also be monitored for the development of cardiac arrhythmias and symptoms
secondary to bradycardia. Rarely, asystole occurs, requiring a temporary pacemaker. However, care must be taken when performing tracheal suctioning or passing feeding tubes, as both can increase vagal tone and exacerbate bradycardia.

C. Gastrointestinal Function

Monitoring of bowel function is essential, as patients with spinal cord injury can develop decreased bowel motility acutely that may lead to paralytic ileus. If this occurs, nasogastric suctioning should be started for decompression. Subsequently, a bowel program, including stool softeners, oral agents, and digital stimulation, should be initiated.

Prognosis

Many spinal cord–injured patients and their families are eager for any type of prognostic indicator that relates to future functioning. It is important during the acute phase to balance issues relating to patient and family hopes and fears stemming from the traumatic event with the need to convey truthful—not wishful—information. Some generalities (eg, expressing cautious optimism about potential improvement of incomplete injuries versus complete injuries; or relating that patients with lower level cord injuries tend to have better functional outcomes than those with upper level injuries) can be made; however, both overly optimistic and overly discouraging or negative comments should be avoided.


Kirshblum SC, Burns SP, Waring W: International standards for neurological
2. Traumatic Brain Injury

Traumatic brain injury (TBI) is discussed in detail elsewhere in this book. Chapter 13 describes the assessment, care, and rehabilitation of adults with TBI; Chapter 20 reviews TBI in pediatric patients. Readers are referred to those chapters for additional discussion beyond that provided here.

Clinical Evaluation Concerns

Important information when evaluating a patient with a traumatic brain injury (TBI) includes the mechanism of injury, associated injuries, and the Glasgow Coma Scale (GCS) score. The GCS is the gold standard for measurement of initial TBI severity (see Table 13–1). This scale, which provides a score ranging from 3 to 15 points, allows the examiner to rate the patient’s motor, verbal, and eye-opening responses. A score lower than 8 is defined as coma. The Rancho Los Amigos Levels of Cognitive Functioning Scale is another descriptive tool that allows the rehabilitation team to follow changes in the patient’s cognitive and behavioral functioning.

CT scanning is the preferred initial imaging method for evaluation of TBI. MRI is often used to assess for small petechial hemorrhages, diffuse axonal injury, and contusions of the frontal and brainstem regions. It is important to remember that a patient can have a TBI without radiographic evidence of injury.

Complications & Trauma Care Issues

Dysautonomia, or paroxysmal sympathetic hyperactivity, occurs most commonly in younger patients and in those with lower GCS scores. Symptoms include tachycardia, hypertension, elevated respiratory rate, hyperthermia, and diaphoresis. When a patient presents with these symptoms, it is important to rule out other causes, such as infection, deep vein thrombosis, or pulmonary
embolism before initiating treatment.

Agitation can occur after a TBI secondary to pain, confusion, fatigue, frustration, hypoxia, fear, discomfort, or a combination of these factors. Management of agitation should initially focus on environmental changes; then, if needed, pharmacologic measures can be implemented. Involvement of the rehabilitation team is important, because some medications that are typically used to treat agitation can interfere with neurologic recovery.

Neuropharmacologic therapies can be initiated in the acute hospital setting. In particular, these may be necessary to address issues relating to arousal, alertness, sleep, and agitation. It is also important to review the patient’s current medications for any that might impede neurologic recovery.

**Prognosis**

It is often the job of the rehabilitation physician to discuss prognosis with the patient or family. Several prognostic factors are assessed in patients with TBI. Among these, length of coma has been shown to correlate most closely with prognosis. Severe disability is unlikely when length of coma is less than 2 weeks; however, if coma persists for more than 4 weeks, the likelihood of a good recovery is low. Other prognostic factors include length of post-traumatic amnesia, patient age, and GCS score.


Teasdale G, Jennett B: Assessment of coma and impaired consciousness.
3. Fractures

Clinical Evaluation Concerns

Many patients with multiple trauma have bony fractures of one or more bones of their extremities. In providing fracture care for these patients, orthopedic specialists often determine corrective interventions and weight-bearing status across specific skeletal segments. As patients increase their mobilization, any precautions for weight bearing or range of motion need to be clarified. Typical weight-bearing instructions include “Weight Bearing As Tolerated (WBAT),” “Partial Weight Bearing (PWB [often delineated by a percentage]),” “Toe Touch Weight Bearing (TTWB),” and “Non–Weight Bearing (NWB).”

A patient with a distal extremity fracture may be able to bear weight through a more proximal skeletal structure. For example, a man with a distal radial fracture might be allowed weight bearing through his elbow (humerus). Similarly, a woman with a metatarsal fracture might be able to bear weight through her femur. The utility of this added weight-bearing surface can have large impact on a patient’s functional ability and postacute placement options. Additionally, as patients become more mobile, they may develop discomfort in areas where it was not previously noted. These areas should be evaluated with the same diligence that would be provided for a painful joint or muscular complaint.

Complications & Trauma Care Issues

Patients with multiple fractures run the risk of developing a compartment syndrome. Compartment syndrome refers to a cluster of symptoms that arise as a result of increasing pressure in a compartment. There are approximately 30 compartments in the upper and lower arm, leg, hand, and foot. An increase in the volume of the contents of these compartments or a decrease in their size or compliance can result in acute compartment syndrome, which is characterized by the 6 Ps (pain, pallor, paresthesia, pulselessness, poikilothermia, and paralysis). Detailed discussion of acute compartment syndrome appears in Chapter 30.
Fractures account for 69% of the cases of compartment syndromes. Fractures of the tibia are the most common cause, followed by fractures of the radius and ulna. Not all symptoms need be present for diagnosis as they represent a continuum of symptoms throughout the disease evolution. Once suspected, diagnosis is confirmed by compartment pressure testing. Normally, compartment pressure is 10 mm Hg; a pressure greater than 30–45 mm Hg is an indication for emergent fasciotomy. Treatment must be performed as quickly as possible, as irreversible muscle damage can occur with high pressures in as little as 3 hours. All dressings, splints, and casts are removed, followed by fasciotomy.

Certain types of fractures place patients at risk for peripheral nerve injuries of either a complete or an incomplete nature. When performing motor and sensory physical examinations of affected limbs, the physician should take care to perform dermatomal, myotomal, and peripheral nerve testing. For example, patients with mid shaft humeral fractures have a 12–19% incidence of radial nerve injuries. Nerve injury with associated fractures occurs more commonly in the upper extremity. Recovery can be evaluated and prognosis estimated on the basis of followup clinical examinations and electrodiagnostic studies.


4. Amputation

Amputation is discussed in detail in Chapters 26 (lower extremity) and 27 (upper extremity). It is often the responsibility of the rehabilitation physician to discuss future rehabilitative courses, pain management, and symptoms with patients who undergo acute amputation as a result of traumatic injury. When possible, this should be initiated prior to the surgical procedure. Patients undergoing amputation may have concerns regarding phantom limb sensation or phantom limb pain.

Complications & Trauma Care Issues
A. Phantom Limb Pain

Most amputees (51–80%) report some presence of sensation of the lost limb within days of amputation. Pain in a lost limb, commonly referred to as *phantom limb pain*, is reported in up to 75% of those who undergo amputations. In a busy trauma center, patients need to be educated about these issues, their frequency, and treatment.

Identification and treatment of phantom limb pain should begin early and include multiple pharmacologic and nonpharmacologic approaches. Among the nonpharmacologic options are sensory discrimination, mirror therapy, mental imagery, transcutaneous electrical nerve stimulation, central stimulation, and prosthetic use. Pharmacologic approaches include tricyclic antidepressants, sodium channel blockers (carbamazepine), lamotrigine, gabapentin, lidocaine, and opioids.

B. Edema

Edema control is an important aspect of postsurgical care that should be addressed prior to discharge from the acute care hospital. Edema can cause increased pain, interfere with healing, and result in a bulbous-shaped residual limb, which ultimately can slow the patient’s functional recovery. Several edema control systems are available, including elastic wraps (Ace bandages), socks, stockinets, prefabricated residual limb shrinkers, nonremovable rigid dressings, and rigid removable dressings.

C. Contractures

Patients should be taught proper positioning and range-of-motion exercises in the early postsurgical period. Prevention of hip and knee contractures is essential in those with lower limb amputation to ensure proper fitting of the prosthesis once the residual limb has healed. Education of family and nursing staff is also important. In particular, pillows under the patient’s knee and between the legs should be avoided. A knee extension board can be fitted under the wheelchair to promote knee extension in lower-limb amputees. Some surgeons also use a knee immobilizer in the early postoperative period if there is concern for knee flexion contractures. Lying prone can aid in the prevention of hip flexion contractures. If patients are transitioning back home after the amputation, instructions about these techniques should be provided so they can be utilized at home.

D. Prosthetics
Realistic goals regarding use of a prosthesis should be outlined early on so the patient will be aware of what to expect from a functional standpoint. In patients who are appropriate prosthetic candidates and who are not subsequently transitioned to an inpatient rehabilitation center, it is important to review preprosthetic training and provide close followup supervision and a plan of care for the patient at the time of discharge.


5. Gunshot Injuries

▶ Clinical Evaluation Concerns

Gunshot injuries cause massive tissue damage relative to the small size of the missile. When a missile travels through the body it causes large tissue deformation, or cavitation, which can injure organs not directly contacted by the projectile. The bullet does not necessarily travel in a straight line, and it can spin end over end during penetration. Therefore, one should be vigilant regarding all structures in the region of a gunshot wound.

▶ Complications & Trauma Care Issues

Treatment is generally surgical for severe injuries; however, minor surface wounds may be managed conservatively, with standard wound care and monitoring for signs of infection. As with all traumatic injuries, tetanus prophylaxis should be updated. It is important to remember that gunshot wounds can result in musculoskeletal injury, spinal cord injury, visceral injury, or TBI and can affect any, and often multiple, organ systems.

Gunshot wound is the third leading cause of spinal cord injury in the United States and a common cause of TBI. Gunshot wounds to the head confer a high risk for infection as the majority of bullets are retained in the skull. Bone fragments also may be present in the area of the bullet, and focal necrosis of
tissue in the bullet’s path can lead to abscess; therefore, surgical debridement is often performed. An important consideration in treatment of patients with self-inflicted gunshot wounds is involvement of a psychologist.


POSTACUTE CARE OF THE TRAUMA REHABILITATION PATIENT

The rehabilitation physician’s role as a consultant providing input to the medical team increases in preparation for patient discharge from the acute care hospital to the postacute care setting. Whenever a patient is transitioned from one level of care to another, his or her medical status, whether considered stable or unstable, must be evaluated and documented accurately to ensure proper management at the next level of care. This can involve difficult decisions, as the definition of “medical stability” may differ relative to each care setting. Thus, although it may be appropriate to transfer a patient from an intensive care setting to a regular acute hospital floor, this patient may not be medically stable enough to transfer from the intensive care unit setting to a skilled nursing facility.

The trauma patient’s medical status is one of many factors considered in postdischarge planning. Other issues that need to be considered when determining the most appropriate level of care for a patient include functional, social, financial, and geographic factors; eligibility; patient choice; and specific concerns that may be unique to each patient. These issues must be identified and various options for care explained and reviewed with the patient and the primary care team, so that the patient and team can make an informed decision about the best option (Table 32–3). Examples of some key issues follows.
**Table 32–3** Postacute trauma considerations.

| **Medical**: intensity of services needed (eg, ventilator, daily physician visits, wound care, nursing intensity) |
| **Functional**: level of assistance needed, prognosis to progress with therapy, specialized equipment, tolerance to therapy |
| **Social**: family support, structural barriers, available resources |
| **Financial**: governmental payor, private insurance, worker compensation, automobile insurance, self-pay |
| **Geographic**: types of facilities available (eg, long-term acute care hospital, skilled nursing facility, inpatient rehabilitation facility, home health care) |
| **Eligibility**: patient condition |
| **Choice**: patient or family may accept or refuse presented options |

aRefers to services in the local area.

**Social Issues**

A patient with a moderate TBI may not be able to return home if there is no social support structure in place. However, if the same individual has a strong social support system and resources, he or she may be able to return to the community setting after discharge from a rehabilitation center.

**Financial Issues**

Patients who meet the medical, functional, and social criteria for the inpatient setting may not have access to certain postacute settings if they are uninsured or their insurance does not cover that level of care.

**Geographic Issues**

Patients who live in a rural area may not have a local long-term acute care hospital, inpatient rehabilitation facility, or skilled nursing facility close to their
home. Some of these patients may not wish to be transferred to a center that is geographically distant from their family.
General Considerations

Approximately 330,000 total hip arthroplasties (THAs) were performed in the United States in 2010. The number of THA surgeries in this country has increased dramatically over the past four decades, and these procedures are expected to increase significantly by 2030.

A. Epidemiology

The majority of hip replacements are performed in individuals between 60 and 80 years of age. Women account for 62% of such surgeries in the United States and typically undergo the procedure between ages 75 and 84 years (compared with ages 65 and 74 years for men). However, procedures are increasingly being performed in patients who are younger or older than this cohort, and advanced age is not a contraindication to THA. Differences in access to care and insurance coverage, as well as socioeconomic level, influence the demographic profile of patients who undergo the procedure. White Americans are more likely than African Americans to undergo THA (at 4.2 versus 1.7 per 1000 individuals), and individuals with higher incomes are 22% more likely to have the surgery than those with low incomes.
B. Indications and Contraindications

Indications for THA are listed in Table 33–1. The main goals of THA are to improve pain and restore function. Candidates for THA should have moderate to severe pain or disability and radiographic evidence of joint damage, which has not been relieved by conservative treatment. This includes a trial of nonsteroidal antiinflammatory drugs (NSAIDs), physical therapy, weight loss, activity modification, walking aids, and disease-specific treatments where appropriate. Absolute and relative contraindications to THA are listed in Table 33–2.

Table 33–1 Indications for total hip arthroplasty.

<table>
<thead>
<tr>
<th>Indications</th>
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<tbody>
<tr>
<td>Hip pain that requires medication stronger than aspirin, is worse with activity; night pain</td>
</tr>
<tr>
<td>Severe osteoarthritis</td>
</tr>
<tr>
<td>Severe inflammatory arthritis</td>
</tr>
<tr>
<td>Traumatic arthritis</td>
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<tr>
<td>Hip fractures, in particular, Garden stages III and IV</td>
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<tr>
<td>Avascular necrosis</td>
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<tr>
<td>Benign and malignant bone tumors</td>
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<tr>
<td>Arthritis associated with Paget’s disease</td>
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<tr>
<td>Juvenile rheumatoid arthritis</td>
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<tr>
<td>Ankylosing spondylitis</td>
</tr>
<tr>
<td>Functional limitations, eg, inability to walk more than several blocks without stopping, difficulty with stair climbing, difficulty performing lower extremity activities of daily living</td>
</tr>
</tbody>
</table>

Table 33–2 Contraindications to total hip arthroplasty.
Clinical Findings

A. Symptoms and Signs

The patient with chronic hip pain may complain of pain in the groin or buttocks that often radiates into the thigh. This pain is often described as a dull ache that is worse with activity and improves with rest. Examination may reveal decreased range of motion, groin or anterior thigh pain on straight-leg raise testing, limp, positive Trendelenburg test, and pain at extremes of range of motion.

Injury to the hip that produce Garden type III or IV fracture requires THA as definitive treatment. In these patients, the hip joint capsule along with the blood supply is disrupted and the patient is at high risk for necrosis of the femoral head. This finding is classically noted in the older individual with acute onset of hip pain after a fall. On examination, the limb appears to be externally rotated and shortened on the affected side.

B. Imaging Studies

Radiographic findings of the osteoarthritic hip include asymmetric joint space narrowing, superior lateral joint space narrowing, subchondral bony sclerosis, osseous cysts, and absence of erosive changes (Figure 33–1). The rheumatoid hip will show symmetric joint space narrowing and, possibly, bone erosions.
Treatment

The femoral head and acetabulum are replaced during THA surgery, in contrast to hemiarthroplasty, in which only the femoral component is replaced. Hemiarthroplasty of the hip is indicated for displaced femoral neck fractures, osteonecrosis of the femoral head with sparing of the acetabulum, and certain cases of pathologic fractures and tumors. Hip arthroplasty varies in the type of prosthesis used (ie, unipolar versus bipolar), fixation of the prosthesis (cement versus cementless), formulation of the prosthesis (ceramic, plastic, or metal), and type of surgical approach (Figure 33–2).
**A. Unipolar versus Bipolar Prostheses**

In hemiarthroplasties, a unipolar or bipolar femoral prosthesis may be used. A unipolar prosthesis has only one large head that articulates with the acetabular cartilage. A bipolar prosthesis has two heads, allowing motion between one head and the acetabular cartilage and between the two heads. The theoretical advantage of this configuration is to reduce acetabular wear and increase range of motion. Bipolar implants are more expensive than unipolar; however, there appears to be no clinical difference in outcomes between patients receiving unipolar implants and those receiving bipolar implants in terms of acetabular wear and hip motion.

**B. Cement versus Cementless Fixation**
Attachment of the prosthetic components to bone can be achieved with cement or cementless fixation. When used, cement serves as the interface between the implant and bone; in the cementless approach, a porous implant surface is used to obtain a press fit to the bone.

Uncemented component fixation is nearly universally indicated. The advantages of a cementless fixation are long-lasting fixation and significant pain-free results. Cemented components have been shown to have a higher rate of loosening and have therefore fallen out of favor. However, cemented acetabular components may be indicated for patients with a life expectancy of 10 years or less and poor bone stock.

C. Surgical Approach

1. Posterolateral approach—This is the most common surgical approach when performing THA. During this approach, the hip is internally rotated, adducted, and flexed. This approach is associated with the highest risk of hip instability postoperatively. This technique also places the sciatic nerve at risk for injury during dissection of the posterior hip capsule. However, the advantage is ease of anatomy, exposure, and avoidance of abductor musculature.

2. Anterior approach—The anterior approach provides good exposure to the hip without requiring a trochanteric osteotomy. During this approach, the hip is extended, externally rotated, and abducted. This approach is associated with notable risks of injury to the femoral nerve, artery, or vein from prolonged anterior retraction. However, the advantages include excellent visualization of the femur and acetabulum as well as decreased postoperative risk of dislocation.

Complications

Complications from THA include, but are not limited to, infection, deep vein thrombosis and pulmonary embolism, nerve palsies, dislocation, loosening of hardware, leg-length discrepancies, heterotopic ossification, trochanteric nonunion, and fracture. Patients are at greatest risk for these events from the acute postoperative period until 6 months after the operation.

A. Infection

Infection occurs in 0.4–1.5% of patients who undergo THAs. Unfortunately, this complication requires removal of the prosthesis, antibiotic therapy, and
subsequent reimplantation of the components 1.5–6 months later. Consequently, sterile technique and prophylactic antibiotic therapy are essential to prevention.

**B. Deep Vein Thrombosis and Pulmonary Embolism**

THA surgery places patients at high risk for deep vein thrombosis (DVT) and subsequent pulmonary embolism (PE). This is secondary to femoral vein damage, stasis, immobility, and hypercoagulability as a result of the operation. A previous history of DVT/PE, hormone replacement surgery, history of cancer, advanced age, female sex, obesity, and prolonged duration of surgery place patients at increased risk of these complications. PE is a potentially fatal complication, most often occurring in the first 2 weeks after the operation. Fortunately, the incidence of fatal PE is low, at 0.3%. An estimated 40–60% of THA patients will develop DVTs if they do not receive anticoagulation postoperatively. There is no consensus on appropriate anticoagulation status post-THA. However, low-molecular-weight heparin, warfarin, and fondaparinux are safe and effective options. The American College of Chest Physicians recommends anticoagulation for DVT prophylaxis for at least 10 and preferably 35 days after THA.

**C. Nerve Palsies**

The incidence of nerve injury in THA surgery ranges from 0 to 3%. Based on the surgical approach, certain nerves are more vulnerable to injury than others. The posterolateral approach is associated with risk of injury to the sciatic nerve, the most commonly injured nerve in THA surgery. Eighty percent of these injuries involve the peroneal distribution of the sciatic nerve, often leading to foot drop. The anterior approach risks damage to the femoral nerve, a complication in 1.7% of cases. In addition, the lateral femoral cutaneous, obturator, and superior gluteal nerves are susceptible to injury. Nerve injuries range from neurapraxia to neurotmesis. Nerve injury may be secondary to compression injury (hematoma), direct trauma, transection, or ischemia. Electromyography may aid in diagnosis of nerve injuries but typically shows no changes until 3 weeks postinjury. Treatment is typically conservative unless a large hematoma or nerve injury is suspected, in which case surgical exploration is indicated. Placing the hip and knee in flexion may decrease tension on the sciatic and femoral nerves. In addition, ankle–foot orthoses may help patients during the rehabilitation process. Prognosis is dependent on the extent of nerve injury, with isolated peroneal injury having a better outcome than complete sciatic palsy.
D. Dislocation

Hip dislocation rates vary by patient, surgeon, and surgical technique between 1% and 8%. Most hip dislocations occur within the first 4–6 weeks postoperatively and are associated with nonadherence to postoperative precautions and instructions. Higher rates of dislocation are associated with previous dislocation (the most important risk factor), female sex, revision surgery, use of the posterior surgical approach, poor muscular tone, femoral neck fractures, intellectual impairment, and advanced age.

Hip precautions must be upheld following THA surgery to avoid dislocation (Table 33–3). The risk of hip dislocation after THA decreases as time passes without dislocation. Once hip dislocation has been discovered, the source should be determined. Dislocation may be a result of patient nonadherence or improper component alignment. The first dislocation may be treated with closed reduction in the simple case of nonadherence or may require adjustment of the components. Abductor braces are helpful in the case of dislocation to assist in healing of lax tissue and help patients comply with their hip precautions. Revision surgery may be required in patients who sustain more than three dislocations.

Table 33–3 Hip dislocation precautions based on surgical approach in total hip arthroplasty.
### E. Leg-Length Discrepancies

This complication is one of the major reasons for lawsuits after THA. Leg-length discrepancy may lead to a limp, cause low back pain, and require the use of assistive devices (canes). Consequently, patients require shoe lifts to compensate for leg-length differences. It is recommended that surgeons measure leg lengths preoperatively, account for preoperative leg-length differences (scoliosis, etc), express to patients the possibility of this outcome, and make surgical attempts if possible to avoid discrepancies that may be caused by laxity of soft tissues around the hip.

### F. Heterotopic Ossification

Heterotopic ossification (HO) is the formation of bone in inappropriate places (Figure 33–3). Severe cases of HO occur in 5–10% of THA surgeries. Risk factors include male sex (HO is twice as common in men as in women), prior occurrence of HO, age older than 65 years, ankylosing spondylitis, head injury, and diffuse idiopathic skeletal hyperostosis. The development of HO over weeks to months may be marked by hip pain or stiffness, decreased range of motion of the joint, fever, swelling, and tenderness to palpation. Diagnostic studies may
reveal an elevated erythrocyte sedimentation rate (ESR), elevated alkaline phosphatase level, and increased uptake on bone scan. Routine prophylaxis is not indicated for HO unless the patient has risk factors, as discussed above. Prophylactic therapies include NSAIDs (indomethacin is the most commonly used agent) or external beam radiation. These treatments are most beneficial when initiated 24–48 hours postoperatively.

Figure 33–3 Heterotopic ossification.

Rehabilitation

For patients to make functional gains post-THA, postoperative rehabilitation is essential. During rehabilitation, patients should be monitored for adequate pain control. Inadequate pain control may result in poor or slow functional progress. Elderly patients must be monitored for sedation resulting from pain medications.
Medications should be titrated so pain is adequately controlled without causing impaired mentation.

Bowel and bladder care are also integral to successful rehabilitation. Postoperative constipation is common secondary to pain medications and decreased mobility. To avoid postoperative ileus, early mobilization is key. Patients should also be placed on a bowel regimen as soon as possible postoperatively consisting of stool softeners, laxatives, and enemas if needed. If there are no issues with urinary retention, catheters should be discontinued on admission to the rehabilitation hospital. Management of urinary retention may necessitate urologic evaluation. Various strategies, which are usually surgeon dependent, are used to prevent thromboembolic complications; these include low-molecular-weight heparin and warfarin. If warfarin is administered, the international normalized ratio (INR) should be maintained between 2.0 and 3.0. Weight-bearing restrictions also vary by surgeon. If a cemented prosthesis is used, the patient is usually able to bear weight as tolerated. However, when using a cementless prosthesis, partial or toe-touch weight bearing is usually recommended to allow for bony ingrowth.

To prevent hip dislocation, the patient must work with the therapy team to learn and abide by total hip precautions (as listed in Table 33–3). Abductor pillows should be provided to all THA patients to assist in compliance with posterior hip dislocation precautions. The rehabilitation team should also evaluate the patient to determine appropriate assistive devices, which will enable patients to comply with orthopedic weight-bearing recommendations and achieve functional gains. Inpatient rehabilitation should focus on transfers, ambulation, stair training, and activities of daily living (while maintaining total hip precautions). In addition, strengthening of the quadriceps and gluteal muscles is essential.

### Prognosis

Overall the procedure is well tolerated, with a perioperative mortality rate of 0.7%. Sweden, which has the largest joint registry, demonstrated a 10-year survival rate of 91–94% in 93,000 implants, and similar results have been reported in published studies. Significant functional improvement on the Harris Hip score (from 51 preoperatively to 94 postoperatively) is seen in 86% of patients. Population-based studies revealed that 90% of THA patients were satisfied with the procedure and demonstrated functional improvement. In addition, 90% of prostheses were noted to be functioning without pain or
complications 10–15 years postsurgery.


KNEE REPLACEMENT

General Considerations

A. Epidemiology
Total knee arthroplasty (TKA) is one of the most common orthopedic procedures in the United States, with over 400,000 performed annually, and this number is expected to increase by as much as 673% by 2030. The average age of a patient undergoing TKA is 70 years. Two thirds of these patients are women, and one third are obese individuals (defined as those with a body mass index [BMI] > 30). Nearly 90% of patients undergoing TKA have a diagnosis of osteoarthritis. As with THA, there are sociodemographic differences in the patient population, with nonwhites receiving the operation half as often as their white counterparts.

**B. Indications and Contraindications**

Indications for TKA are listed in Table 33–4. The goals of TKA, similar to those of THA, are to improve pain and restore function. Candidates for TKA have knee pain that is unrelieved with conservative treatments such as medications, including NSAIDs and other pain relievers; physical therapy; weight loss; activity modification; bracing; and use of assistive devices. Absolute and relative contraindications to TKA are listed in Table 33–5.

### Table 33–4 Indications for total knee arthroplasty.

<table>
<thead>
<tr>
<th>Indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee pain</td>
</tr>
<tr>
<td>Severe osteoarthritis</td>
</tr>
<tr>
<td>Severe inflammatory arthritis</td>
</tr>
<tr>
<td>Juvenile rheumatoid arthritis</td>
</tr>
<tr>
<td>Osteonecrosis</td>
</tr>
<tr>
<td>Post-traumatic injury</td>
</tr>
<tr>
<td>Valgus or varus deformity</td>
</tr>
<tr>
<td>Fracture</td>
</tr>
<tr>
<td>Infection</td>
</tr>
</tbody>
</table>

### Table 33–5 Contraindications to total knee arthroplasty.
Clinical Findings

A. Symptoms and Signs
Patients often report a history of knee pain or stiffness that is worse with activity (eg, climbing stairs, walking long distances) and improved with rest. Physical examination of the knee may reveal medial or lateral joint line tenderness, antalgic gait, and decreased range of motion.

B. Imaging Studies
Osteoarthritis manifests most often in the medial compartment of the knee. Consequently, medial joint space narrowing on radiographs is a finding consistent with osteoarthritis (Figure 33–4). Absence of erosive changes, subchondral sclerosis, and osseous cysts may also be seen. The rheumatoid knee will show symmetric joint space narrowing, with or without bone erosions.
▲ Figure 33–4 Osteoarthritis of the knee.

**Treatment**

TKA procedures have femoral, tibial, and patellar components (Figure 33–5). They vary in terms of prosthesis used and type of fixation. Currently, there is no consensus on choice of prosthesis, and the decision is surgeon dependent. The TKA prosthesis may be posterior cruciate ligament (PCL) retaining or sacrificing. PCL-retaining and -sacrificing surgeries each offer theoretical advantages that have not been proven in clinical studies. PCL retention may have the advantage of increased range of motion secondary to shifting the center of rotation posteriorly during knee flexion, which improves the lever arm of the quadriceps. It may also result in improved stair-climbing capacity, decreased
stress on the implant, and improved proprioception. In contrast, PCL-sacrificing surgery may more reliably correct the deformity and make it easier to balance the knee.

\textbf{Figure 33–5} Total knee replacement.

TKA procedures also differ in terms of fixation: cemented versus cementless versus hybrid (femur component uncemented, tibial component cemented). Most TKAs are performed with cemented components because cementless implants have a higher incidence of tibial loosening and osteolysis.

\section*{Complications}

Surgical complications are often associated with three main factors: the patient, the surgery, and the materials used (Table 33–6). Lack of adherence to
postoperative instructions and preexisting medical conditions account for the majority of patient factors causing complications. The overall complication rate of TKA is 8% within the first 6 months after surgery. Increased risk of complications has been associated with age older than 65 years, greater number and severity of medical comorbidities, and low numbers of procedures performed at the institution. Complications include, but are not limited to, knee stiffness, infection, DVT, poor wound healing, loosening of components, periprosthetic fracture, and nerve injury.

**Table 33–6** Complications of total knee arthroplasty.

<table>
<thead>
<tr>
<th>Patient Factors</th>
<th>Surgical Factors</th>
<th>Material Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee stiffness</td>
<td>Knee stiffness</td>
<td>Loosening of implants</td>
</tr>
<tr>
<td>Infection</td>
<td>Infection</td>
<td>Patellar instability</td>
</tr>
<tr>
<td>Deep vein thrombosis (DVT)</td>
<td>DVT</td>
<td></td>
</tr>
<tr>
<td>Poor wound healing</td>
<td>Anemia</td>
<td></td>
</tr>
<tr>
<td>Nerve injury</td>
<td>Nerve injury</td>
<td></td>
</tr>
<tr>
<td>Fracture</td>
<td>Fracture</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Vascular injury</td>
<td></td>
</tr>
</tbody>
</table>

**A. Knee Stiffness**

The incidence of knee stiffness post-TKA ranges from 1.3% to 12%. Stiffness may be secondary to patient nonadherence, inappropriate alignment or size of hardware, inadequate bone resection, instability, infection, HO, or poor pain management. Factors that place patients at higher risk for developing this complication are female sex, prior knee surgery, higher BMI, diabetes mellitus, depression, pulmonary disorders, and disability. The best predictor of postoperative stiffness is preoperative range of motion. Treatment of stiffness differs by surgeon and patient. Options include TKA revision and closed or arthroscopic manipulation.

**B. Infection**

Infection is a very serious complication seen in 0.5–12% of patients
postoperatively. When prophylactic antibiotics are used, this decreases to 1.6%. Rheumatoid arthritis, diabetes mellitus, and previous knee surgery increase the risk of infection. TKA patients who develop postoperative infection of the knee will require prolonged duration of antibiotics, removal of hardware, and reimplantation of a new prosthesis in the future when deemed appropriate by the surgeon.

C. Deep Vein Thrombosis
DVT is a commonly complication after TKA. Incidences of 40–88% have been reported in patients who did not receive anticoagulation compared with 1% in those who received prophylaxis. There is no uniform consensus regarding type and duration of anticoagulation therapy after TKA. However, both medications (warfarin, heparin, aspirin, or enoxaparin) and pneumatic compression devices are often utilized.

D. Poor Wound Healing
Obesity, rheumatoid arthritis, and diabetes mellitus place the patient at increased risk of poor wound healing. Superficial wound complications include infection, erythema, and excessive drainage. Prompt recognition and treatment is necessary to prevention progression to deep infection and possible loss of the implant. In the case of multiple surgeries, consultation with a plastic surgeon may be of assistance with wound closure.

E. Periprosthetic Fracture
Although most often occurring in the patella, this complication may also affect the supracondylar femur or tibia. Rheumatoid arthritis, osteopenia, and poor knee flexion all increase the risk of periprosthetic fracture. The rate of periprosthetic patellar fracture is 0.68%, and treatment options range from conservative to surgical depending on severity. Similarly, treatment of femur and tibial fractures depends on the severity of fracture and status of the implant.

F. Nerve Injury
The incidence of nerve injury after TKA is 0.9–1.3%. The peroneal nerve is most commonly injured; however, cases of brachial plexus, sacral plexus, and sciatic injury have also been reported. Flexion contracture, valgus deformity, increased tourniquet time, postoperative hematoma, external leg compression, epidural anesthesia, and history of nerve compression all place the peroneal nerve at
higher risk of injury.

**Rehabilitation**

Rehabilitation should begin prior to surgery with information about the surgical process and expected outcomes. Postoperative day 1 rehabilitation should consist of bedside exercises, such as ankle pumps, and initiating transfer training. Progression to active and active-assisted range of motion and quadriceps strengthening can begin as early as day 2. Ambulation with the least restrictive assistive device can begin on day 2, as well. The use of a continuous passive motion (CPM) machine is often dependent on the surgeon. If a CPM machine is used, range of motion movement usually begins on postoperative day 1 at full extension (0 degrees) and 30 degrees of flexion and should be advanced 5–10 degrees per day until 90 degrees of flexion is achieved at approximately 6 weeks postoperatively. The progression of rehabilitation is dependent on the patient’s tolerance, but milestones should be advanced as quickly as possible to include advanced strengthening exercises, such as leg extensions and knee bends; ambulation on uneven surfaces and stairs; and independence in activities of daily living, such as lower body grooming and dressing.

Several other factors must be considered during the rehabilitation process. Adequate analgesia must be provided and is usually accomplished with a combination of short- and long-acting opiates. A variety of strategies are used to prevent thromboembolic complications; these choices are usually surgeon dependent and include low-molecular-weight heparin and warfarin. If warfarin is administered, the INR should be maintained between 2.0 and 3.0. Weight-bearing restrictions also vary by surgeon. If a cemented prosthesis is used, the patient is usually able to bear weight as tolerated. However, when using a cementless prosthesis, partial or toe-touch weight bearing is usually recommended to allow for bony ingrowth.

**Prognosis**

TKA is a well-tolerated procedure with a perioperative mortality of 0.6%. Several studies have shown knee implant survival rates to be greater than 90% at 10 years, and 85–95% at 15 years. Most patients (85%) indicate that they are satisfied with the results of surgery.


SHOULDER REPLACEMENT

General Considerations

A. Epidemiology

The shoulder is the third most common joint for which arthroplasty is performed. Initially developed to treat humeral fractures, the treatment has broadened to include severe osteoarthritis, currently the leading indication for shoulder arthroplasty.

Although significantly fewer shoulder arthroplasties are performed than THA or TKA, the number of procedures has increased tenfold in the past 25 years. Approximately 30,000 shoulder arthroplasties are performed in the United States each year; half of these are total and half are hemiarthroplasties. Of all patients who undergo joint arthroplasties, those with shoulder replacement have the lowest total mean age.

B. Indications and Contraindications

In general, shoulder arthroplasty is indicated in patients who have severe shoulder pain or loss of function that is unresponsive to nonoperative treatments. Additional indications and contraindications for shoulder arthroplasty are listed in Tables 33–7 and 33–8.

Table 33–7 Indications for shoulder arthroplasty.

<table>
<thead>
<tr>
<th>Indications</th>
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<tbody>
<tr>
<td>Severe glenohumeral arthritis</td>
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<tr>
<td>Severe rheumatoid arthritis</td>
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<tr>
<td>Post-traumatic arthritis</td>
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<tr>
<td>Dislocation arthropathy</td>
</tr>
<tr>
<td>Ankylosing spondylitis</td>
</tr>
<tr>
<td>Proximal humeral fractures</td>
</tr>
<tr>
<td>Rotator cuff arthropathy (controversial)</td>
</tr>
</tbody>
</table>
Table 33–8 Contraindications to shoulder arthroplasty.

<table>
<thead>
<tr>
<th>Contraindications</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Active infection (absolute contraindication)</td>
</tr>
<tr>
<td>• Neuropathic joint</td>
</tr>
<tr>
<td>• Complete paralysis of deltoid or rotator cuff muscles</td>
</tr>
<tr>
<td>• Severe brachial plexopathy</td>
</tr>
<tr>
<td>• Intractable shoulder instability</td>
</tr>
<tr>
<td>• Poor surgical candidates (secondary to current medical status)</td>
</tr>
</tbody>
</table>

Clinical Findings

The patient with osteoarthritis of the shoulder may complain of deep shoulder pain that is worse with activity and improved with rest and at night as the disease progresses, as well as shoulder stiffness and decreased functional abilities. Examination may reveal decreased active and passive range of motion, crepitus, and joint line tenderness to palpation. On imaging, the osteoarthritic shoulder may show osteophytes, subchondral sclerosis, humeral head flattening, subchondral cysts, and joint space narrowing (Figure 33–6). In contrast, patients with rheumatoid arthritis present at an earlier age and with more joint destruction. These patients may complain of morning stiffness and inflammation. Examination may reveal erythema, warmth, crepitus, effusions, and decreased range of motion. Radiographs of the rheumatoid shoulder typically show joint erosions.
**Figure 33–6** Osteoarthritis of the shoulder.

### Treatment

In patients needing shoulder replacement, treatment ranges from hemiarthroplasty to total arthroplasty to reverse total shoulder arthroplasty. In a shoulder hemiarthroplasty, the humeral head is replaced with a prosthesis, without replacement of the glenoid. In TSA, a prosthesis is used to replace the humeral head and a plastic socket replaces the glenohumeral cavity (**Figure 33–7**). However, in a reverse TSA, the metal ball is “reversed” and secured to the glenoid instead of the humerus. The socket is then added to the humerus. This shifts the center of rotation of the shoulder medially, thereby lengthening the deltoid and enabling the deltoid to provide overhead motion in the absence of rotator cuff musculature.
Reported survival rates of the prostheses are lower after TSA in patients younger than 50 years of age. Therefore, TSA is typically only considered in patients older than 50. In those younger than 50 years of age with glenohumeral arthritis, hemiarthroplasty is usually recommended. Hemiarthroplasty is also commonly indicated for inadequate glenoid bone stock, irreparable rotator cuff tears with fixed upward displacement of humeral head, and proximal humeral fractures in elderly patients. TSAs are generally preferred over hemiarthroplasty in the treatment of glenohumeral osteoarthritis.

Patients with rotator cuff arthropathy, severe glenohumeral joint arthritis, or failed prior surgery may benefit from a reverse TSA. This prosthesis provides a fixed fulcrum for the shoulder joint, allowing the arm to be raised overhead even when the rotator cuff muscles are absent.

Complications
The average orthopedic surgeon performs only two TSAs per year. Consequently, decreased complication rates are seen in surgeons and hospitals with higher volumes of procedures. Complications include, but are not limited to, infection, loosening, glenohumeral instability, periprosthetic fracture, rotator cuff tear, deltoid muscle dysfunction, and neural injuries.

**A. Infection**
The overall prevalence of infection post-TSA is 0.7%. An increased risk of infection is seen in patients with diabetes mellitus, rheumatoid arthritis, systemic lupus erythematosus, previous surgery, history of chemotherapy, systemic corticosteroid therapy, and repeated intraarticular steroid injections. Pain along with an elevated erythrocyte sedimentation rate, C-reactive protein level, and white blood cell count may be indicative of infection. Treatment strategies are similar to those discussed earlier for patients with post-THA and post-TKA infection.

**B. Prosthetic Loosening**
Loosening of the prosthesis accounts for 39% of complications in TSA and occurs in 6.3% of patients. Loosening occurs in the glenoid and humeral components, more often affecting the glenoid.

**C. Glenohumeral Instability**
Instability of the glenohumeral joint is the second leading cause of post-TSA complications, with a prevalence of 4%, accounting for 30% of all TSA complications. Instability is most often present in the anterior and superior aspects. If instability is related to subscapularis tendon rupture, repair of the rupture may be undertaken. Some patients may require Achilles tendon grafting to stabilize the shoulder.

**D. Periprosthetic Fracture**
Periprosthetic fracture has a prevalence of 1.5–3% post-TSA and is most often seen in rheumatoid arthritis patients with fragile and osteopenic bones. The majority of fractures occur intraoperatively as a result of technical error and may involve the glenoid or proximal humerus. Postoperative fractures may be treated nonoperatively or with open reduction and internal fixation surgery.

**E. Rotator Cuff Tear**
Rotator cuff tear is the fourth most complication after TSA, with a prevalence of 1.3%. The majority of these events results from tears of the subscapularis tendon. Subscapularis tears are associated with multiple operations, overly aggressive postoperative therapy involving external rotation, and tendon compromise from lengthening techniques.

**F. Deltoid Muscle Dysfunction**

This complication often occurs secondary to axillary nerve injury or deltoïd muscle detachment. Patients with problems involving the deltoïd muscle have severe loss of shoulder function.

**G. Neural Injuries**

Most commonly these injuries involve the axillary nerve, but the brachial plexus may also be compromised after TSA.

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**Rehabilitation**

Rehabilitation should begin prior to surgery with information about the surgical process and expected outcomes. Postoperative rehabilitation varies based on the preferences of the surgeon and the surgical approach taken. Proper donning and doffing of the shoulder sling should be reviewed with the patient. The arm is positioned in adduction, internal rotation, and slight forward flexion. For the first few days after surgery, ice is applied to reduce pain and swelling. Heat may also be applied before exercise to encourage relaxation of the shoulder muscles.

Typically, active range of motion of the fingers, wrist, and elbows is started the day after surgery. Codman exercises, in which the involved extremity hangs free and is moved in a pendulum fashion through the range of motion without contraction of the shoulder muscles, can begin as early as postoperative day 1. The use of a CPM machine is controversial and often dependent on the surgeon’s preference. If a CPM machine is used, the range of motion should be advanced 5–10 degrees per day until 60 degrees of external rotation and 140 degrees of forward flexion is achieved. Rehabilitation is progressed, based on the patient’s tolerance, to include scapular strengthening, active shoulder range of motion, and rotator cuff strengthening.

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**Prognosis**
Shoulder pain relief is achieved in 90–95% of shoulder arthroplasty patients. Perioperative mortality is low, at approximately 0.005%. The short-term complication rate is higher in patients undergoing total or hemiarthroplasty for fracture as compared with nonfracture indications. Patients at lower risk for revision are advanced in age, have had a prior fracture, or have rheumatoid arthritis, all of which are associated with lower functional demands on the prosthesis. Preoperative factors associated with worse postoperative outcomes after TSA included glenoid erosion, humeral head subluxation, and preoperative loss of range of motion. Patients with TSA have better scores for pain, mobility, and activity at 2-year followup compared with hemiarthroplasty patients. In addition, a multicenter trial found a 94% rate of good or excellent results after TSA compared with an 86% good or excellent rate after hemiarthroplasty.


WRIST REPLACEMENT

General Considerations

Many factors should be considered before deciding to perform a total wrist arthroplasty (TWA). These include the patient’s age, specific disease process, functional and occupational requirements, and tolerance of potential complications. TWA had been considered a salvage procedure for patients with rheumatoid arthritis. The procedure should be thought of as a treatment option
whose goal is to alter the progression of the disease and improve functional outcomes. There are no good data describing the annual incidence of TWA in the United States. Absolute and relative contraindications to TWA are listed in Table 33–9.

Table 33–9 Contraindications to total wrist arthroplasty.

<table>
<thead>
<tr>
<th>Absolute Contraindications</th>
<th>Relative Contraindications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Active infection</td>
<td>Immunosuppression</td>
</tr>
<tr>
<td>Uncooperative patient</td>
<td>Multiple ruptures of extensor digitorum communis</td>
</tr>
<tr>
<td>Patient unwilling to accept potential arthrodesis</td>
<td>Use of assistive device in affected hand</td>
</tr>
<tr>
<td>Presence of nonfunctional hand due to neurologic disease</td>
<td>Poor bone stock</td>
</tr>
<tr>
<td>Rupture of radial wrist extensors</td>
<td>Active inflammatory disease</td>
</tr>
<tr>
<td></td>
<td>Systemic lupus erythematosus with ligamentous laxity</td>
</tr>
<tr>
<td></td>
<td>Vascular insufficiency</td>
</tr>
<tr>
<td></td>
<td>Younger age</td>
</tr>
<tr>
<td></td>
<td>Heavy, manual labor</td>
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</tbody>
</table>

Clinical Findings

The most common indication for TWA in the United States is rheumatoid arthritis (Figure 33–8). Other indications include advanced osteoarthritis and post-traumatic arthritis of the wrist. Success of the procedure depends on good bone stock and adequate soft tissue balance. The patient’s occupational and recreational goals should also be considered. Depending on the prosthetic device, a number of restrictions may be placed on the patient, including limitations on weight carried and avoidance of activities that increase the risk of falling.
Treatment

Traditional surgical treatment of severe wrist arthritis has ranged from removal of carpal bones (carpectomy) to partial carpal fusions to total wrist fusions (arthrodesis). Total wrist arthrodesis sacrifices movement at the wrist for pain control. In an attempt to preserve range of motion at the wrist, designs for different total wrist arthroplasty systems have been developed and tested since the early 1970s with varying results. Figure 33–9 shows one example. A 2008 systematic review compared arthrodesis to arthroplasty; its findings are summarized in Table 33–10.
**Figure 33–9** Wrist arthroplasty.

**Table 33–10** Comparison of outcomes for patients undergoing wrist arthrodesis versus arthroplasty.
### Complications

The most common complications following TWA are infection and soft tissue imbalance. Following surgery, the patient may be unable to move the wrist into a neutral position. Dislocation can occur soon after joint replacement, or months to years later. Dislocation occurring early (within 4 weeks) is usually caused by inadequate soft tissue protection. Midterm (at 4 weeks or more) and late dislocation (occurring months to years later) occurs when disease progression causes loosening of components. Long-term complications include implant failure and loosening.

### Rehabilitation

Rehabilitation following TWA varies considerably based on surgical factors, the patient’s prior level of function, and the specific implant chosen. Initially, the wrist is immobilized in the position that provides the most stability to the implant. The period of immobilization can last from a few days to a few weeks based on the intraoperative stability achieved. During the period of immobilization, the primary focus of rehabilitation should be pain control, wound care, and maintaining range of motion of the elbow and fingers. Mobilization of the wrist varies with different implants, but usually begins with flexion–extension and is advanced to include radial–ulnar deviation, and, lastly, pronation–supination. Rehabilitation is progressed to include strengthening of the wrist flexors and extensors and functional use of the wrist to accomplish...
activities of daily living. It is important to be aware of the limitations of the implant at all times throughout the rehabilitation process. Early loosening and failure of the implant will occur by exceeding the weight restrictions.

### Prognosis

Patient satisfaction with both total wrist arthrodesis and total wrist arthroplasty remain above 90%. The rate of complications and need for further surgical intervention is higher in patients undergoing arthroplasty. Despite its perceived benefit of improved range of motion, only 3 of 14 studies surveyed in a recent review demonstrated functional range of motion with TWA.


### ANKLE REPLACEMENT

#### General Considerations

Total ankle arthroplasty (TAA) is recommended in patients with chronic ankle pain associated with a limited number of conditions (see below) that is unresponsive to conservative, nonoperative treatments. Compared with arthrodesis (surgical fixation of the joint), TAA has the benefit of maintained
foot and ankle motion, with similar improvements in pain relief. The preservation of ankle movement provides a near-normal gait pattern in contrast to arthrodesis, where loss of ankle range can lead to abnormal gait. There are no good data concerning the number of TAA procedures performed annually in the United States.

Indications for TAA are listed in Table 33–11. The ideal candidate for TAA is a mobile, middle- to older-aged adult, with a normal or low BMI and no signs of neurovascular comprise. These criteria are much more restrictive when compared with ankle arthrodesis, which can be performed in patients with deformities, paralysis, neuropathy, avascular necrosis of the talus, and every age and body type. Absolute and relative contraindications to TAA are listed in Table 33–12.

Table 33–11 Indications for total ankle arthroplasty.

| • End-stage osteoarthritis |
| • Severe inflammatory arthritis |
| • Post-traumatic arthritis (most common indication) |

Table 33–12 Contraindications to total ankle arthroplasty.

<table>
<thead>
<tr>
<th>Absolute Contraindications</th>
<th>Relative Contraindications</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Active infection</td>
<td>• Younger age</td>
</tr>
<tr>
<td>• Charcot arthropathy</td>
<td>• High body mass index (BMI)</td>
</tr>
<tr>
<td>• Insensate foot</td>
<td>• Diabetes mellitus</td>
</tr>
<tr>
<td>• Avascular necrosis of talus</td>
<td>• Smoking</td>
</tr>
<tr>
<td>• Inadequate leg musculature</td>
<td>• Heavy work demands</td>
</tr>
<tr>
<td>• Lower limb deformities</td>
<td>• Osteopenia</td>
</tr>
<tr>
<td>• Severe tibiotalar malposition</td>
<td></td>
</tr>
</tbody>
</table>

Clinical Findings
A. Symptoms and Signs

Patients may relate a history of trauma antecedent to the onset of ankle pain and commonly report ankle stiffness, pain, and swelling. They may have a history of bone spurs or other joint deformity, as well as decreased tolerance for standing and walking.

B. Imaging Studies

Radiographs may reveal asymmetric joint space narrowing, osteophytes, and sclerosis of subchondral bone (Figure 33–10).

![Figure 33–10 Osteoarthritis of the ankle.](image)

Treatment

Approximately 20 different TAA prostheses are available worldwide (Figure 33–11). The first generation of devices had a two-component design (tibial...
component, talar component) and a high rate of failure. This led surgeons to recommend arthrodesis over TAA in patients who met criteria for these procedures. The high rate of failure was attributed to the inability to accommodate biomechanical forces at the ankle joint. However, the newer three-component design includes an additional intermediate meniscal-bearing interface between the tibial and talar components. This helps to absorb forces through the tibia and fibula and distribute them to the talus. The three-component design is currently the gold standard for TAA in Europe and will likely spread to the United States. In most TAA procedures, the implant is performed without cement. TAA has a significant advantage over arthrodesis; namely, arthrodesis is associated with a high incidence of pseudoarthrosis (10–20%) despite extended cast immobilization to achieve fusion.

![Figure 33–11 Total ankle arthroplasty.](image)

**Complications**

The most common complications of ankle replacement surgery are infection,
impaired wound healing, residual pain, component loosening, stress fractures of the medial malleolus, and periprosthetic fracture.

### Rehabilitation

For the first 4–6 weeks following TAA, the patient is usually in a cast or wearing a walking boot and limited to non-weight bearing status. During the first few weeks, rehabilitation focuses on maintaining range of motion at the knee and hip and education on proper use of assistive devices (crutches, walker) for ambulation. When the cast is removed or when the surgeon has given clearance to remove the walking boot, the focus shifts to active range of motion at the ankle. Typically, the patient can progress to weight bearing as tolerated by 4–6 weeks. At that time, gait training should be initiated. As rehabilitation advances, emphasis is shifted to improving balance and proprioception of the affected ankle.

### Prognosis

Outcomes for TAA have been reported as excellent or good in 82% of patients who received a newer generation ankle device compared with 72% who underwent fusion. Meta-analysis of meniscal-bearing devices has shown improved ankle range of motion, a prosthesis survival rate of 90.6% at 5 years, and a complication rate ranging from 1.6% to 14.7% among patients who underwent TAA with these devices.

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OVERVIEW OF WORKPLACE INJURIES

As a society, we place great significance on the value of work and productivity. This value system is reflected in the amount of time the average adult spends in the workplace. Similarly, the expectation of safety in the workplace is recognized as a good business model. A healthy workforce decreases the rate of employee turnover and costs associated with training and hiring. Healthy workers also minimize costs associated with work absenteeism and loss of productivity. Many workers are able to sustain their level of productivity without interruption, but as with any system upset situations can develop, resulting in injury and impaired function.

The field of occupational and environmental medicine is particularly geared to manage these issues. Health care providers in this field are trained as advocates and champions of safer work environments but also aim to effectively and safely manage work injuries. The nature of work itself and the potential risks associated with specific work settings have changed significantly since the industrial revolution. The field of occupational medicine has evolved with these changes. Specialists in this field function as change agents who can often spearhead adaptation of best practice standards and laws affecting a larger population of workers and their workplace.

Health care providers can also have an impact on individual workers by interacting with stakeholders such as the injured worker, the employer, case managers, third-party administrators, union representatives, and lawyers in managing the medical claims.
**Workers’ Compensation & Workplace Safety Regulation**

Most people have some familiarity with the workers’ compensation system. In the United States, many employers are required to obtain insurance for workers’ compensation, and this coverage secondarily provides wage replacement and medical benefits to employees injured while performing their work duties. The general premise of this coverage also holds that employees are covered for all work-related injuries in this “no-fault” system. As such, employees do then have limited recourse to seek damages through the traditional tort system.

Many employers have developed active systems of minimizing costs associated with workers’ compensation coverage. Ideally this should involve development of safer processes aimed at reducing the risk of injury and illness. Some adaptations have been industry driven; others have emerged in response to regulatory mandates.

**A. Regulation of Workplace Safety**

The most recognizable regulatory body driving workplace changes in the United States is the Occupational Safety and Health Administration (OSHA). OSHA was created in 1970 by a congressional mandate (the Occupational Safety and Health Act of 1970) as a federal program and was tasked with the mission of improving workplace safety and enforcement of standards. Today, there are approximately 27 state-based OSHA programs, but these entities must still meet or exceed federal OSHA standards.

**B. Risk Factors for Workplace Injury**

OSHA estimates that annually approximately 3.3 million workers sustain serious injury in their workplaces. Some observers view this as a public health issue and have extensively studied the risk factors associated with work injuries. Such an analysis can include consideration of system failure, process failure, or human error.

In one study, three broad categories affecting work injuries were identified: human factors, job content, and environment. Human factors that influence the risk of work injury include work experience, baseline physical impairments, stress, and job satisfaction. More recently the issue of fatigue has been recognized as a predisposing factor in injury for various industries. Job-related factors can involve task order and scheduling. Environmental factors can involve
physical hazards and stressors such as noise.

The study of injury prevention utilizes findings from all of these factors in addressing potential areas of harm, but absent full and self-sustaining automation, injuries to workers cannot be completely eliminated. Even with the best practices and systems, there remains a risk of injury.

C. Reporting of Workplace Injuries and Occupational Illness

In the workers’ compensation system, the time course of work injury management starts with reporting of the event by the injured worker. The time constraints under which a worker must report an injury are further defined by specific state requirements. The reporting timeline for an injury typically differs from that for an occupational-related disease, with longer time being allowed for the worker’s recognition of a disease or illness attributed to work exposures. To be compensable, an injury must arise out of or occur in the course of employment. For example, an employee who slipped while rushing down a ladder would be eligible for coverage of her injuries. In contrast, an employee who had a seizure episode while at work, fell, and fractured his wrist might not be eligible for coverage.

▶ Evaluation of Workplace Injuries

In addition to performing a medical assessment, health care providers who manage workplace injuries should have some awareness that they likely will also be expected to supply an opinion about work-relatedness; that is, did the work task or work injury result in whatever diagnosis has been obtained? A stepwise approach to injury management should first and foremost revolve around establishing a medical diagnosis and determining the immediate needs of the worker. Escalation of care, when medically indicated, should not be delayed. Thereafter, the provider needs to address work-relatedness, as this determination will further inform the course of injury management. The health care provider will continue to treat the worker if the injury if deemed to be work related. Alternatively, the provider may refer the worker to a primary provider if the medical condition is not work related. In this case, as occurs in some primary care practices where the provider also functions as the treating physician (for general medical care) of record, this separation can be challenging.

The American Medical Association’s (AMA) Guidelines to the Evaluation of Disease and Injury Causation highlight a stepwise approach to this process
starting with initial establishment and verification of a diagnosis. Thereafter, the provider should attempt to determine if a cause-and-effect relationship exists. In some cases, there is a very clear and easily determined relationship, as occurs when a worker falls after tripping on equipment in the workplaces and fractures a wrist. The determination is more difficult in the worker who attributes long-term changes or illness to work exposure, particularly if the condition is fairly common. This could be the case with occupationally related asthma, various degenerative musculoskeletal conditions, or neurocognitive conditions.

Although determination of work-relatedness can be challenging, the provider should take into consideration several variables, including the proposed length and severity of exposure; should explore the presence of similar conditions in coworkers or cohorts; and should also apply best practice with regard to what has been established in the literature about similar associations. Confounding factors, such as tobacco use, concurrent employment, and obesity, should be considered, particularly for degenerative conditions.

OSHA defines an injury or illness to be a work-related matter when the event or exposure occurs within the work environment and when work caused or contributed to the resulting condition or significantly aggravated a preexisting illness. Exceptions to this rule could include injuries or illness that surface at work but are solely due to non–work-related issues. An example would be the worker who has a stroke or heart attack—medical conditions that would not typically be attributable to work. Additional caveats to these examples depend on specific work settings and occupational exposure. In the case of firefighters, sudden cardiac death has increasingly been recognized as a significant risk associated with this profession, and, as such, an acute cardiac event could be attributed to work. The increased risk is linked to a combination of commonly identified factors, including age, family history, diabetes, and hypertension. Occupational exposures to gases such as carbon monoxide and hydrogen cyanide during active fire suppression have more recently been recognized as an additional risk factor in this occupation. As well, various studies have shown a strong enough association between cardiovascular disease and work activities in the fire service that this health problem typically is now recognized as a work-related issue for this job category.

Other exceptions when considering work-related injuries include injuries or illness sustained as a result of an employee eating, drinking, or preparing meals, assuming the illness is not due to food poisoning from food supplied by the employer or food otherwise contaminated in the workplace. Injuries sustained while the employee is engaged in personal tasks unrelated to employment, such
as self-grooming or self-inflicted injuries, also are excluded. Mental illness is typically exempted but OSHA may allow for further consideration based on the opinions obtained from a physician or other licensed health care professional with appropriate training.

Osteoarthritic changes in the absence of acute radiographic findings present one of the more challenging scenarios for a provider who is asked to supply an opinion about causality. This is especially true when the injured worker either was not aware of this condition prior to injury or did not appreciate the extent of such findings. Osteoarthritis (OA) itself is a fairly common condition in the general population irrespective of occupational factors. Predisposing factors for OA include age (with an increased incidence in older persons), obesity, physical deconditioning, smoking, and certain metabolic syndromes. In the context of work injuries, research on the development of post-traumatic OA has included analyses of knee and ankle injuries, specifically fractures. In early reporting, some subjects were found to have radiographic changes on followup 2–4 years after a fracture injury, but these presented as localized changes. Future research into the role of trauma in localized OA may necessitate reconsideration of work-relatedness and OA; however, overwhelmingly the literature still requires that specific factors be present, such as a clearly defined injury. Moreover, the area of injury should correlate with the area of degenerative change. The presentation of OA may also appear prematurely, ahead of the normally anticipated onset, further minimizing the role of age and other non–work-related factors commonly identified in the general population.

In some state systems, the injured worker’s choice of initial health care provider is directed by a panel of medical providers selected by the employer or by the employer’s third-party administrator. In other states, an injured worker may be able to receive treatment through his or her private physician. In all instances, the assessment completed for a work injury should mirror the typical assessment completed in any medical setting. Pertinent differences include a focus on factors such as the injured worker’s mechanism of injury and prior medical history.

The goal in management of an injured worker’s care is restoration of prior function. In many cases, this can be achieved through conservative measures involving medication, therapy, and other modalities. During this period, the employee can either be released to resume work in a modified work capacity or placed out of work, depending on the opinion of the examining physician. Ideally, return to work in some capacity should be advised unless a provider truly feels that the injured worker is disabled and cannot function in a manner
that allows completion of activities of daily living.

This decision can create tension in the provider’s interaction with some injured workers, who may have anticipated complete removal from work. Consider the example of a worker who sustains a lumbar sprain without signs of neurologic urgency. The American College of Occupational and Environmental Medicine (ACOEM) Practice Guidelines advise that persons with low back pain tend to improve with some form of aerobic exercise and maintenance of preinjury activity. Recognizing the importance of activity in the rehabilitative process, many leading organizations, including ACOEM, stress safe but early return to work. Should the injury require a need for escalation of care (eg, surgery or hospitalization), providers would need to consider other factors in making a recommendation for resumption of work. Various guidelines are available in these scenarios.

Approximately 60–80% of the general population will experience at least one episode of low back pain in their lifetime. This frequency remains true in the workplace. Within the context of workplace injuries, back pain represents one of the most commonly reported work-related complaints. Treatment for low back disorders also tends to be more expensive than other types of claims. Direct costs attributed to management of occupationally related back pain are estimated at $10.8 billion annually and represent an area of significant impact. The literature further demonstrates that at least 90% of back pain episodes have a mechanical cause, meaning there has been some injury to muscles, ligaments, bones, or discs. In most cases, resolution of symptoms is expected. Studies show that in 50% of these cases, patients report resolution of pain within a period of 1 week. By 8 weeks, more than 90% of patients are asymptomatic. Less than 5% of patients reporting an acute episode of pain progress to having persistent chronic symptoms. These parameters would exclude situations where a provider notes “red flags” or areas of clinical concern, such as weight loss, fevers, worsening pain, or bowel or bladder incontinence. Even in the setting of a workplace injury, clinicians should always consider the possibility of other nonmechanical pathology, which would significantly alter the treatment course.

Treatment options for work-related injuries should be consistent with best practice standards. Many providers utilize rehabilitative services in returning the injured worker to his or her preinjury baseline level of function. The duration of treatment can be further defined by various guidelines, such as those of ACOEM. Resolution of pain and an improvement in function are the initial goals of treatment. In cases of delayed recovery, defined in this setting as the presence of symptoms after an appropriate healing period, use of work conditioning
programs should be considered.


REPETITIVE MOVEMENT INJURY

Trends in Repetitive Injuries in the Workplace

Data from the Bureau of Labor Statistics (BLS) show that 3,063,400 nonfatal work injuries were reported in 2010. Approximately 933,200 cases reported from private industry involved time away from work. Of these injuries, 40% were categorized as sprains or strains, and 20% involved injury to the back. Ergonomic injuries or musculoskeletal disorders accounted for an additional 29% of injuries that required time away from work. Injuries defined as repetitive in nature accounted for approximately 3% of the total sustained in private industry. These types of injuries tended to result in a longer time away from work, with a median of 24 days. (For reference, the median time away from work for fractures is 30 days.) The data further suggest a correlation between factors such as age and gender and reported repetitive injuries. Although repetitive-type injuries represent a small percentage of private industry injuries,
they disproportionately accounted for more lost time from work. Consequently, many employers actively attempt to minimize the risk of injury by implementing controls on repetitive actions and engineering ergonomic solutions.

#### Defining Repetitive Work

Activities such as typing and keyboarding, which are commonly associated with repetitive-type injuries, are not always isolated to the work setting. With the popularity and availability of social networks and technological devices, more people are spending time online, texting, or playing game systems. In the setting of a work injury, the clinician is often asked to comment on work-relatedness and as such needs to have an understanding of what constitutes repetitive work. One definition put forth by the BLS is that repetitive injuries involve stress or sprain brought on by repetitive tasks such as grasping and moving objects. As examples, the Bureau lists scanning groceries at a checkout counter or typing.

Determination of work-relatedness in repetitive type injuries involves more than simply confirming that an employee types as part of his or her work responsibilities or uses the hands in completion of work tasks. Using such a broad definition, having almost any type of job would place a person at risk for repetitive injury, as nearly all work requires use of the hands. Instead, providers should consider contributory factors, such as frequency of tasks or prior medical history. Any discussion of work-relatedness and repetitive-type injuries should utilize the framework definition set by the research on cumulative trauma or musculoskeletal disorders.

#### Carpal Tunnel Syndrome & Work-Relatedness

Carpal tunnel syndrome (CTS) is one of the most common conditions associated with repetitive work but is also a highly prevalent condition in the general population. With regard to work-relatedness, health care providers should recognize the occupational factors that have been associated with CTS but also address nonoccupational risk factors, such as age, gender, family history, body mass index, diabetes mellitus, and thyroid disease. Key factors that can guide the provider in making a determination about work-relatedness include the presence of work tasks that require constant application of dynamic pinching or gripping. There is a higher association when the pinch forces exceed 10 N or 1 kg. Providers should also have some familiarity with the research on CTS, which
has shown that the strongest association between work and CTS occurs in industries such as manufacturing and meat packing or processing.

Likewise, the National Institute for Occupational Safety and Health (NIOSH) guidance document on repetitive injuries found that the strongest association between CTS and work occurred in occupations categorized as highly repetitive. The studies reviewed by NIOSH typically found repetitive work to include a task cycle time of less than 30 seconds. Research on repetitive injuries in workers using instruments such as scissors found an even higher task cycle of 2–26 seconds. In addition to repetitive tasks, force has also been identified as a risk factor for both CTS and tendonitis. The force cited in some of these studies was defined as force greater than 6 kg. Studies examining shoulder injuries in the workplace have also demonstrated a strong relationship with short task cycles of less than 30 seconds, but arm positioning was also a factor. More injuries were noted for work performed with repetitive flexion or abduction of the shoulder.

American College of Occupational and Environmental Medicine: 

**BARRIERS IN THE RETURN-TO-WORK PROCESS**

Most workers are able to return to work after a work injury. For workers who experience prolonged periods of work stoppage, various barriers have been identified in their return-to-work process, including factors such as lack of accommodation in the workplace, union issues, lack of trust or confidence in the employer on the part of the injured worker, and lack of job satisfaction. Some health care providers also identified their lack of occupational health training as an additional barrier. This perhaps reflects lack of comfort with initiating a return-to-work process for an injured worker who remains in treatment. Ideally the return-to-work process should not be delayed in lieu of the worker’s report of full recovery. The psychological benefit of being able to maintain some level of productivity often has a positive impact on recovery, decreasing the length of disability. Further, the process of returning to work itself can provide physical benefits that help counteract the general deconditioning that can occur upon complete work stoppage. Such an approach would be consistent with ACOEM’s position, which encourages early and safe provider-facilitated return to work. For injured workers who are unable to return to work for nonmedical reasons, ACOEM advises the provider to take an active role in communicating with the
employer about the worker’s readiness for work.

The provider should clearly state his or her recommendation concerning the injured worker’s ability to complete tasks relative to examination findings, with a secondary awareness of what the worker’s job may entail. As an example, consider an injured crossing guard who cannot stand as a result of a foot fracture. The provider might advise restrictions for standing, walking, climbing, or other weight-loading activities but the worker would be capable of using his or her upper extremities in various other tasks and could be returned to work in a seated position. Some workers may report lack of accommodations in the workplace, but this should not be the basis for determining appropriate work restrictions. Larger employers typically have more aggressive return-to-work programs, as well as a process through which to return the injured worker to some level of productivity within the workplace.

**Comorbid Conditions**

The impact of comorbid conditions such as diabetes and hypertension are increasingly recognized as additional barriers in return to work. Data collected by the National Council on Compensation Insurance suggests that the costs of managing work injury claims increases with the degree of obesity. These data further suggest that obese workers have a higher risk of becoming permanently disabled as a result of a work injury.

This finding is consistent with conclusions from a Duke University Medical Center–sponsored retrospective cohort study of 11,728 health care and university employees who completed at least one type of health appraisal between January 1, 1997, and December 31, 2004. Researchers examining the relationship between body mass index (BMI) and number of workers’ compensation claims found a strong association between BMI and injury claims as well as costs associated with treatment. Obese workers were twice as likely to file a work injury claim, and the number of lost workdays was 13 times higher in obese workers. Care for this population also resulted in medical claims that were seven times higher than those for nonobese workers. The average workers’ compensation medical claim cost per 100 employees was $51,019 for obese workers and $7,503 for nonobese workers. The strongest associations between BMI and claims were found in injuries to the lower extremity, wrist or hand injuries, back injuries, and slips and falls.

These studies not only highlight barriers to recovery in work injury management but also offer opportunities for employers who wish to address the
overall health of their employee population ahead of an injury. Similar to the systems in place for active surveillance of potential chemical or biological hazards, employers today are utilizing wellness programs to more effectively monitor their worker population for general health concerns that typically would have been handled outside the workplace. These wellness-focused programs provide additional opportunities for health care providers to facilitate both injury management and health promotion as complementary elements in health management.

Costs Associated with Delayed Return to Work

Health care providers should utilize diagnostic testing and pharmaceutical therapy appropriately in managing the employee’s return to work. Overtesting can result in increased anxiety on the part of the injured worker and confers some degree of harm. Overprescribing, particularly of pain medications such as opioids, can create additional barriers to safe and timely return to work and function. One study of workers’ compensation claims in Michigan found that factors such as age of the injured worker, length of time out of work, involvement of an attorney, and use of long-acting opioids were associated with higher total claim cost. Although the medication cost represented a small portion of the total expenditure, claims for injured workers receiving long-acting opioids were more than nine times more expensive than similar claims for workers treated with nonopioid medications.


FUNCTIONAL CAPACITY EVALUATION (FCE) & WORK DISABILITY

Injured workers may not recover from their injuries or may be left with some degree of functional loss after undergoing prescribed treatment. In such cases, a provider typically needs to consider what functional deficits might result, and further determine the impact of such deficits on the worker’s ability to perform his or her job. Functional capacity evaluations (FCEs) can be used in addressing these questions. The ACOEM guidelines are neutral with respect to the use of FCEs, particularly in management of chronic low back pain. However, many providers utilize these tests in defining a worker’s physical capacities; the findings can then be applied as an objective measure of the person’s ability to complete activities of daily living or, alternatively, to perform the functions of a specific job. FCEs are not always necessary, particularly if a provider feels comfortable in setting a worker’s physical limitations or if a worker has a job that is sedentary or requires only light physical demands. The utility of these tests is generally highest in situations where a patient is not progressing as anticipated or where there is some ambiguity regarding the reported symptoms and examination findings.

Providers who utilize FCEs should have an appreciation of the strengths and weaknesses of this tool. The test should be completed by a trained and qualified therapist and the formulation of testing should be based on standardized psychometric measures. Guidelines developed by the American Physical Therapy Association (APTA) emphasize the utility of FCEs in decisions regarding return to work and also in settings such as job placement, disability evaluation, and treatment planning. The guidelines further outline two types of FCEs: general purpose testing and job-specific testing. Both forms address a person’s ability to perform basic tasks such as pushing, pulling, lifting, kneeling, and reaching. Provision of a job description better enables the examiner to organize a series of tests that can address the patient’s ability to complete specific work functions. Results of the FCE are typically then organized in a manner that correlates with the Department of Labor’s physical demand definitions from the Dictionary of Occupational Titles. This information includes categories of strength, as outlined in Table 34–1.
Table 34–1 Categories of occupational physical demand used in evaluating return to work.

<table>
<thead>
<tr>
<th>Category of Work</th>
<th>Physical Demands</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sedentary</td>
<td>Entails an ability to occasionally exert up to 10 lb of force or alternatively to frequently exert a negligible amount of force to lift, carry, push, pull, or otherwise move objects. This work capacity would involve a significant amount of time spent seated but may also require brief or occasional periods of walking or standing.</td>
</tr>
<tr>
<td>Light</td>
<td>Entails occasionally exerting up to 20 lb of force or frequently exerting up to 10 lb of force in lifting or carrying objects. This category may require a significant amount of walking or standing relative to sedentary work. Can also involve positions in which one sits for most of the work function but pushes or pulls in operating arm or leg controls.</td>
</tr>
<tr>
<td>Medium</td>
<td>Entails occasionally exerting up to 20–50 lb of force, frequently exerting up to 10–25 lb of force, or constantly exerting up to 10 lb of force in lifting or carrying objects.</td>
</tr>
<tr>
<td>Heavy</td>
<td>Entails occasionally exerting up to 50–100 lb of force, frequently exerting up to 25–50 lb of force, or constantly exerting between 10 and 20 lb of force in lifting or carrying objects.</td>
</tr>
<tr>
<td>Very heavy</td>
<td>Entails demand capacities occasionally in excess of 100 lb of force, or frequent use of 50 lb of force, or constant use of force in excess of 20 lb to lift or carry objects.</td>
</tr>
</tbody>
</table>
Standard assessments are based on an 8-hour work day and the terms *occasionally* (up to one third of the time), *frequently* (from one third to two thirds of the time), or *constantly* (two thirds or more of the time) are used in defining a person’s tolerance for specific activities. In addition to the use of standardized terms, the APTA identifies other elements that should be included in an FCE. Among these are a review of the patient’s history; the purpose of the study, with identification of the referral source; a review of the patient’s job description; and, when present, identification of behaviors that may have an impact on the patient’s physical performance.

Most examiners will note whether the patient appears to provide “full effort” in completing the test. Lack of full effort does not always indicate a specific intent to circumvent testing and can be related to other conditions, such as test anxiety, fear avoidance behavior, or fatigue. On the other hand, some patients may purposefully attempt to perform at a capacity that is below their functional level. Various tools can be used to aid in this determination, but examiners should use good judgment in stating their conclusions regarding a patient’s perceived effort. Examiners may utilize tests for Waddell’s signs and measurement of grip strength to measure patient effort, when assessing for consistency of effort. Grip strength can be measured three times with a hand dynamometer. A schematic review of these values should show a bell-shaped curve when maximal effort has been put forth. Some examiners consider a non-bell-shaped curve to be a possible indicator of submaximal effort.

**Strengths & Limitations of FCE Testing**

Use of FCEs in disability assessments and in determining the ability of an injured worker to resume his or her preinjury position has come under scrutiny. Although various testing methodologies and systems are available, no gold standard exists. Among the available testing systems in common use, some have been shown to have better reliability and validity than others. These considerations are important because, increasingly, the findings from these studies are applied in clinical decision making. In the case of work injuries, these finding may result in the injured worker being released back to work or, alternately, recommended for continued removal from work. The provider must have a sufficient level of confidence in the findings of the FCE, in light of the risk and liability associated with inappropriately returning someone back to work or unnecessarily furthering disability and work stoppage. Some studies have noted that disability benefits are suspended an average of 32 days after the
completion of the FCE.

Studies evaluating the ability of FCEs to predict actual return to work have been inconclusive. This in part reflects the fact that some injured persons subsequently report additional injuries in the recovery period for an initial claim. When this occurs, it is difficult to separate the instances in which delayed return to work was attributed to the initial injury from those in which the delay may have been affected by other claims or reported injuries. Providers should also understand the limitations of an FCE. The ACOEM guidelines emphasize that this study tends to measure voluntary performance, and individuals with lower pain tolerance or other behavioral barriers, such as fear avoidance, may be more likely to underperform during testing. FCEs have not been shown to accurately predict return to work nor do they accurately predict the risk for future recurrence. In light of such caveats, providers would be well advised to adopt the findings from an FCE as another measure or tool in making their determination about a worker’s capabilities, but not to rely solely on the findings of these tests.


**IMPAIRMENT RATING**

The timeline for most work injuries should result in resolution or return to a baseline level of function that allows the injured worker to resume his or her preinjury job. If the injured worker reports ongoing symptoms of pain or functional deficits, the provider must develop a treatment plan that allows for the maximal level of medical treatment. Eventually the provider may determine that the injured worker has reached the level of maximum medical improvement (MMI). At this stage, the provider is essentially recommending that the injured worker may not experience any significant changes from additional treatments such as surgeries, therapy, or further allowance of time.

Such a concept typically precipitates subsequent impairment evaluation.
Treatment may continue after this date, but further improvement is not reasonably expected. The designation of MMI does not indicate that the injured worker has been released to resume work, although sometimes the worker may have regained enough function to allow a return to a modified employment position. For example, an injured worker may have an impairment after sustaining a rotator cuff injury of the nondominant arm but would not be considered disabled if he or she remained capable of completing a significant portion of work and nonwork activities.

An impairment evaluation may be requested for the injured worker who is unable to resume his or her preinjury job or has a decreased level of function that significantly impacts the ability to seek alternate employment. Most state systems and providers use the American Medical Association’s (AMA) *Guides to the Evaluation of Permanent Impairment* as the standard text for evaluating impairment. The first edition of the *Guides* was published in 1971, and the most recent (sixth) edition in 2008.

### Functionality: Impairment, Disability, & Handicap

The terms *impairment*, *disability*, and *handicap* tend to be used interchangeably but each carries a different meaning. Impairment relates to anatomic or psychological loss or abnormality. Disability reflects the impact of a respective impairment on an individual’s social and work life. Handicap is a less frequently used term that refers to the overall impact of that disability within a social context of normative roles.

The content of a disability or impairment examination is similar to that of a standard medical examination but key differences exist. Histories obtained in both settings are applied differently. Because the diagnosis is well defined at the time of an impairment examination, the impairment history stresses documentation of the person’s functional status. Providers may inquire about a person’s ability to complete activities of daily living. They may also inquire about the use of assistive devices or other aids required to maintain the person’s level of function. Issues with self-care, sleep, and daily activities should be clearly addressed and documented. In comparison, a standard medical examination focuses on obtaining a diagnosis and proceeding with treatment. Other differences include the added goal, in an impairment examination, of rating an individual’s impairment and apportionment. Providers completing impairment assessments also establish a different relationship and should explicitly state to the individual that the evaluation does not constitute the
development of a patient-to-provider relationship. AMA guidelines suggest that the report generated should include the information listed in Table 34–2.

Table 34–2 Information that should be included in an impairment evaluation.

<table>
<thead>
<tr>
<th>Information that should be included in an impairment evaluation.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Purpose of the evaluation and identification of the referral source</td>
</tr>
<tr>
<td>History of current illness or injury</td>
</tr>
<tr>
<td>Individual's ability to perform activities of daily living</td>
</tr>
<tr>
<td>Medical record review</td>
</tr>
<tr>
<td>Physical examination findings</td>
</tr>
<tr>
<td>Laboratory and radiographic findings</td>
</tr>
<tr>
<td>Diagnosis, with explanation of basis for diagnosis and causation, if appropriate</td>
</tr>
<tr>
<td>Continuing treatment needs</td>
</tr>
<tr>
<td>Maximum medical improvement status</td>
</tr>
<tr>
<td>Impairment rating, with charts used and explanation of the final impairment rating</td>
</tr>
</tbody>
</table>

The Guides have been found to be a reliable tool for rating impairment. An analysis by Forst and colleagues concluded that both the current (sixth) and previous editions provide a systematic and reliable rating system for objectively measuring and compensating injured workers. However, questions have been raised about differences between the fifth and sixth editions. Specifically, the sixth edition may utilize a more inclusive definition of disability and impairment. Furthermore, although both editions were found to have high interrater reliability, use of the sixth edition could potentially result in a decrease in compensation, because the rating assigned takes into consideration any physiologic abilities on examination. The examiner should bear this in mind, and retain a heightened awareness of the potential implications for workers with regard to the final rating assigned.

Whenever possible, ACOEM encourages safe and early return to work as a fundamental element of effective injury management. This philosophy can prove difficult for providers who are not comfortable with managing the expectations of various stakeholders (injured worker, employer, etc). However, providers must maintain an awareness of the potential harm that the worker can sustain
from prolonged work absence. Some providers develop a narrow focus on injury management, perceiving time away from work as a period for healing, without maintaining an appropriate appreciation for the disruptive impact of prolonged work stoppage on a worker’s daily life. This method of injury management can result in what ACOEM defines as “system-induced disability.” An ideal process should address the worker’s acute needs in injury or illness management but stress early and safe return to work whenever possible.

Similarly, providers often struggle with the return-to-work release, but there are, in fact, limited situations in which a person requires complete, total, and prolonged work stoppage. One such example would be an individual with a severe psychiatric or cognitive disorder. Postsurgical patients receiving acute rehabilitation may also require some period of work stoppage. Excluding these cases, most people are never incapacitated to an extent that precludes the ability to work at a sedentary physical demand level. Moreover, if patients are limited to an extent that makes sedentary work impossible, objective evidence should be readily attainable, on examination or through diagnostic testing. Absent such objective findings, providers should not support disability.


Cancer is the uncontrolled growth of cells, which damages healthy tissue and causes disease. The American Cancer Society differentiates more than 100 types of cancers that manifest in diverse ways throughout the human body. Treatment of cancer can vary from local to systemic and from mildly invasive to radical depending on the type of cancer and the extent of disease.

With nearly 12 million cancer survivors in the United States, cancer rehabilitation is a growing field. The goal of this chapter is to highlight the most common issues in the care of these patients that are pertinent to physiatrists. Physiatrists are experts in restoring function and improving quality of life for patients, making them well trained to address the rehabilitative needs of the growing cancer survivor population. As in other settings for rehabilitation, a multidisciplinary team approach is ideal throughout the diagnosis, treatment, survivorship, and palliative care of patients.

OVERVIEW OF CANCER REHABILITATION

More than 1.6 million new cases of cancer were diagnosed in the United States in 2012. With advancements in health care, people are living longer, and as they age, their risk of developing cancer increases. The greatest incidence of cancer is
between the ages of 65 and 74 years. The three most common cancers diagnosed in the United States are those of the prostate, female breast, and lung; these unfortunately also have the highest mortality rates.

Cancer rehabilitation was first documented by Drs Howard Rusk and Eugene Taylor in 1949. Funding for cancer rehabilitation was established in 1965 by the Rehabilitation Act, which provided 75% of cost by federal dollars. In 1973 the National Rehabilitation Act provided protection from discrimination for people with handicaps, now defined as disabilities; this was the precursor to the Americans with Disabilities Act, enacted in 1990. More recently, the requirements for the American College of Surgeons’ Commission on Cancer accreditation state that hospitals and cancer centers must provide rehabilitation services to cancer survivors either at their primary facility or by referral.

Two programs that were in the forefront of cancer rehabilitation in the 1960s have maintained their status as leaders in the field of cancer rehabilitation today. They are the University of Texas MD Anderson Cancer Center and a cooperative program with Drs Howard Rusk and J Herbert Dietz, in New York, that has become Memorial Sloan Kettering Cancer Center. These two centers are the only locations in the United States that offer a fellowship dedicated to training physicians in cancer rehabilitation.

Dietz created the first definition of cancer rehabilitation derived from evidence-based medicine. He also created a classification system for the goals of rehabilitation therapy, differentiating among four phases of care: prevention, restoration, supportive care, and palliative care. Prevention is treatment provided to a patient before development of a potential disability that is expected to lessen the severity of disability or its duration. Restoration is the return of the patient to a premorbid state without handicap or known residual disease, including return to gainful occupation. Supportive care is control of ongoing disease while the patient remains active and productive but with known residual disease and possibly a slowly progressive handicap. In this stage, increased tolerance and circumvention of the residual disability can be expected from adequate supportive training and care. Palliative care addresses increasing disability expected from relentless progression of disease, with a focus on provision of an appropriate program that will prevent or reduce complications that might otherwise develop. These complications include, but are not limited to, bedsores, pain, contractures, problems with personal hygiene, weakness, and emotional deterioration secondary to inactivity and depression.

The number of cancer survivors in the United States has increased more than threefold in the past 30 years. With nearly two thirds of newly diagnosed cancer
patients expected to survive at least 5 years or more, it is essential for physiatrists to understand the disease processes, treatment options, and complications that confront cancer survivors.


CANCER-RELATED FATIGUE

General Considerations

Cancer-related fatigue (CRF) is a highly prevalent and distressing symptom affecting cancer patients. The National Comprehensive Cancer Network (NCCN) defines CRF as a “distressing, persistent, subjective sense of physical, emotional and/or cognitive tiredness or exhaustion related to cancer or cancer treatment that is not proportional to recent activity and interferes with usual functioning.” Clinical features include loss of interest; exhaustion; lack of energy; impaired sleep, memory, and cognition; pain; anxiety; and depression. Thus, CRF can cause further physical, psychological, emotional, and economic sequelae for the patient, caregivers, and family. Estimates of prevalence range
from 50–90%. It may occur at any time during the disease state from diagnosis through treatment, and including the years following treatment. To adequately treat CRF, underlying mechanisms must be elucidated.

Pathogenesis

The pathophysiology of CRF is complicated, multifactorial, and not well understood. It remains uncertain whether CRF is a result of the tumor itself, treatment of the tumor, behavioral or environmental factors, or has a genetic disposition. Proposed pathophysiologic factors include dysregulation of inflammatory cytokines, gene polymorphisms, changes in the central nervous system (CNS) serotonergic system, disturbances of the hypothalamic regulatory circuit, and disturbances of circadian melatonin secretion. Factors that may exacerbate or precipitate CRF include pain, nutritional status, anemia, deconditioning, sleep disturbances, and existing comorbidities.

Several theories have been proposed to explain the mechanism of CRF. Dysregulation of the serotonin system is often implicated in the pathogenesis of CRF. Serotonin (5-HT) has many functions, including control of appetite, sleep, memory, mood, behavior, cardiovascular function, endocrine regulation, and depression. It is believed that 5-HT levels or 5-HT receptors may be upregulated in patients with cancer, causing an overall decrease in somatomotor drive. The metabolism of serotonin may be affected by any of the proinflammatory cytokines, specifically tumor necrosis factor-alpha (TNF-α) and cytokines such as interleukin-1b.

The hypothalamic–pituitary–adrenal axis is another pathway affecting fatigue. Theories that focus on this mechanism propose that any cancer treatment or the cancer itself may affect the axis system, thus causing endocrine changes contributing to fatigue. Cortisol and corticotropin-releasing hormone appear to be implicated in this pathway.

Treatment

Just as the causes of CRF are multifactorial, so are treatment interventions. The NCCN identifies four categories of CRF intervention: education and counseling, management of fatigue, nonpharmacologic measures, and pharmacologic measures. Treatment should be comprehensive to include all categories, and should begin as early as time of diagnosis. Energy conservation techniques help
to manage fatigue, and strengthening helps to alleviate deconditioning and inactivity. Exercise is known to improve functional capacity, thereby reducing energy expenditure for activities of daily living. Moderate aerobic exercise seems to provide the most benefit. Additional nonpharmacologic interventions include stress management, yoga, acupuncture, mindfulness-based stress reduction, and cognitive-behavioral therapy. Pharmacologic interventions include treatment of anemia, psychostimulants, and proper pain management. Methylphenidate and modafinil are recognized to improve fatigue. Use of corticosteroids has also been studied in the treatment of CRF.


General Considerations

Pain in the cancer patient is a challenge to both patient and provider. It is estimated to affect between 30% and 50% of those actively undergoing treatment, and upwards of 70% of those with advanced disease, thus having major implications on quality of life. While pain may be the first diagnostic sign of malignancy, it may also be present at any time, and the frequency and intensity of pain increase with advanced stages of cancer.

Pathogenesis & Clinical Findings

Pain may be divided into three categories: somatic, visceral, and neuropathic. The first two of these are considered nociceptive. Pain may be caused by tumor infiltration or occur as a result of additional treatments (eg, radiation, surgery, or chemotherapy). Examples of peripheral neuropathic pain include radiation neuropathy, chemotherapy-associated neuropathy, surgery-related nerve injury, and plexopathy.

Primary afferent sensory neurons are the pathway by which sensory information from the periphery is referred to the spinal cord and brain. The cell bodies of sensory fibers are located in the trigeminal and dorsal root ganglion, consisting of large-diameter myelinated Aβ fibers and small-diameter thinly myelinated Aδ and unmyelinated C fibers. It is the smaller diameter C fibers and Aδ fibers—known as nociceptors—that generate chronic pain in cancer patients.

Once a malignancy has caused local or systemic tissue injury, neurotransmission of these nociceptors is altered. Hyperalgesia, the perception of mildly noxious stimuli as highly noxious, and allodynia, the perception of normally nonnoxious sensory information as noxious, alter the body’s normal pain interpretation. This pathway plays a role in both somatic as well as visceral pain.

Visceral pain is described as diffuse and poorly localized; it is often referred to other locations and may be accompanied by motor and autonomic reflexes. It is typically described as dull, aching, deep, vague, or diffuse. In contrast, neuropathic pain is commonly described as burning, shooting, or lancinating. Associated negative motor phenomena (weakness, fatigue) or positive motor phenomena (tremors, ataxia, dystonia, and dyskinesia) may occur with neuropathic pain. Pain sensitization may occur peripherally, centrally, or through both routes, which adds to the complexity of treatment.
Treatment

Once the type of pain has been diagnosed, management is paramount. Options include pharmacologic as well as nonpharmacologic measures and should be used in combination, as this has been established by the World Health Organization (WHO) as the standard of care. The WHO developed the pain control ladder (Figure 35–1), a stepwise approach to initiation of analgesic control. For mild pain, nonopioids with or without an adjuvant are recommended. Adjuvant medications include nonsteroidal antiinflammatory drugs, topical analgesics, antidepressants, anticonvulsants, corticosteroids, and anxiolytics. Additional agents such as pregabalin have also been shown to be efficacious in treating neuropathic cancer pain. Once a moderate level of pain is reached, opioids may be introduced. To maintain adequate pain control, analgesics should be given around the clock every 3–6 hours rather than as needed.
For patients with severe, refractory pain or chronic pain, interventional techniques offer another treatment option. Neurolysis, trigger point injections, sympathetic blocks, vertebroplasty, and spinal cord stimulation are examples of available interventional options as adjuncts to pharmacologic therapy to improve neuropathic pain.

Other nonpharmacologic options exist and have been studied in the cancer patient. Modalities such as exercise, yoga, acupuncture, use of transcutaneous electrical nerve stimulation (TENS) units, biofeedback, and psychosocial interventions all have been shown to be efficacious for the management of cancer pain. In addition to ameliorating pain, exercise and yoga may also reduce
fatigue, thereby improving quality of life. Bracing and orthotics may help to offload joints, provide stability, and help with weakness thereby decreasing pain.


RADIATION TOXICITY

General Considerations

Approximately 50% of all patients diagnosed with cancer will require radiation therapy at some point during the course of their disease. Radiation can be used with intent to cure, or palliatively to control pain, prolong life, or preserve function. Radiation therapy can be delivered by means of external beams (external beam radiation therapy) or radioactive material placed internally (brachytherapy). The dose of radiation is determined by tissue tolerance (radiosensitivity) of the target tissue; Table 35–1 lists examples of organ-specific
tolerance doses. The therapeutic intent of radiation is to kill fast-dividing cancer cells while sparing the relatively slower growing somatic cells. Patient-related factors, including cancer size, location, and tissue radiosensitivity, play an integral role in dosing. Patient-related factors that are incorporated into the treatment plan include age, obesity, prior surgery, trauma, and the presence of microvascular diseases such as diabetes, hypertension, or collagen vascular disease.

![Table 35–1](image)

**Table 35–1** Tolerance doses (TD$_{5/5}$-TD$_{50/5}$) to whole-organ irradiation.

<table>
<thead>
<tr>
<th>Organ</th>
<th>Single Dose (Gy)</th>
<th>Fractionated Dose (Gy)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brain</td>
<td>15–25</td>
<td>60–70</td>
</tr>
<tr>
<td>Eye (lens)</td>
<td>2–10</td>
<td>6–12</td>
</tr>
<tr>
<td>Skin</td>
<td>15–20</td>
<td>30–40</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>15–20</td>
<td>50–60</td>
</tr>
<tr>
<td>Vasculoconnective tissue system</td>
<td>10–20</td>
<td>50–60</td>
</tr>
<tr>
<td>Mucosa</td>
<td>5–20</td>
<td>65–77</td>
</tr>
<tr>
<td>Peripheral nerve</td>
<td>15–20</td>
<td>65–77</td>
</tr>
<tr>
<td>Muscle</td>
<td>&gt; 30</td>
<td>&gt; 70</td>
</tr>
<tr>
<td>Bone and cartilage</td>
<td>&gt; 30</td>
<td>&gt; 70</td>
</tr>
<tr>
<td>Thyroid</td>
<td>—</td>
<td>30–40</td>
</tr>
</tbody>
</table>


Radiation doses are currently measured in grays (Gy) or centigrays (cGy). Radiation was previously measured in rads (1 Gy = 100 cGy = 100 rads). Fraction refers to the amount of radiation delivered in one treatment session. Total dose radiation takes into account the fractions repeated over a period of
time and boosts the dose if it was delivered. Hyperfractionated regimens deliver smaller radiation doses more than once a day and therefore decrease associated radiation complications. Hypofractionated radiation regimens deliver higher radiation doses in fewer treatment sessions. Dose-sculpting techniques such as image-guided radiation therapy (IGRT) and intensity-modulated radiation therapy (IMRT) allow for very tight confirmation of the radiation beam so that tumors near vital structures (eg, the spinal cord) can be radiated to within a few millimeters. Such techniques allow for radiation to be given in a single fraction in certain instances, which allows tumors that were previously thought radioresistant (eg, melanoma) to be effectively treated.

Complications

The complications associated with radiation toxicity are numerous and can be categorized as acute and late effects (Table 35–2). Acute effects occur during treatment or shortly afterward, while late effects occur months to years later. Acute complications damage rapidly proliferating cells; therefore, the effects are typically temporary and often resolve during the course of treatment. Late effects can appear decades after treatment and progress for the duration of the patient’s life. There is a risk for secondary carcinomas as well as toxic effects on organs within the radiation field, including cardiomyopathies, pulmonary fibrosis, and thyroid dysfunction. Approximately 10% of people who receive radiotherapy ultimately develop a secondary cancer. Children treated before the age of 15 years are at the highest risk.

Table 35–2 Effects of radiation therapy.
RADIATION FIBROSIS SYNDROME

Radiation has damaging effects on multiple structures, including soft tissue, ligament, muscle, nerve, blood vessels, and the lymphatic system. The term radiation fibrosis syndrome is used to describe the numerous neuromuscular, musculoskeletal, and organ complications that occur as a direct result of radiation-induced fibrosis. It is a common phenomenon; approximately 60% of all patients receiving radiation therapy eventually develop radiation-induced fibrosis.

Microvascular injury appears to be the critical feature in both acute and chronic radiation toxicity of normal tissue. The pathologic effect of radiation is the formation of a rigid interstitial fibrinous exudate (fibrin) preceding progressive fibrotic encasement of blood vessels, thus distorting surrounding
tissue (Figure 35–2). The effects of radiation can cause injury to the nervous system at every level, including encephalopathy, myelopathy, radiculopathy, plexopathy and mononeuropathies, and myopathy. Nerve damage may be secondary to compromise of the vasa nervorum, the vascular supply of the nervous tissue.

▲ Figure 35–2 Radiation-induced fibrosis. Sections of parietal pericardium at equal magnifications. **Left:** Normal pericardium showing an (upper) layer of fibrous tissue and a (lower) thick layer of adipose tissue. **Right:** Extensive fibrosis of irradiated pericardium replacing adipose tissue. Hematoxylin and eosin stain. (Reproduced with permission from Luis F Fajardo, MD, Stanford Medical School.)

Acute radiation encephalopathy can occur with a single fraction greater than 300 Gy due to increased intracranial pressure from whole-brain radiation therapy. Symptoms include somnolence, headache, and progressive focal neurologic deficits. The combination of imaging, laboratory studies, and clinical presentation can distinguish encephalopathy from recurrence or infection. It is typically self-limiting, and corticosteroids may be utilized to decrease edema. Chronic radiation encephalopathy associated with cerebral atrophy manifests with dementia, cognitive deficits, ataxia, and urinary incontinence. Encephalopathy from radiation necrosis typically occurs 1–2 years after treatment, with a 3–5% occurrence rate, and is associated with doses greater than 5000 Gy. It can be difficult to distinguish necrosis from tumor recurrence by magnetic resonance imaging (MRI); hence, positron emission tomography (PET) is the study of choice. Treatment is supportive, consisting of corticosteroids or resection. Surgical decompression may not reverse the functional decline in the majority of patients and serves as more of a palliative measure. Bevacizumab
has shown promising results clinically and radiologically for reversal of cerebral radionecrosis.

Myelopathy is associated with radiation levels in excess of 5000 Gy, with an average of 14 months latency. Presenting symptoms are typically sensory abnormalities in the lower extremities with ascending weakness up to the site of radiation or a Brown-Séquard syndrome presentation. Hyperreflexia with Babinski’s and Lhermitte’s signs are commonly present. Painful paresthesias may be experienced in the dermatomal distribution correlating with the spinal level of irradiation.

Radiation-induced brachial plexopathies occur more commonly than lumbosacral plexopathies and are associated with fractions to the brachial plexus greater than 2 Gy. Clinically, radiation-induced plexopathy is less likely to be painful than infiltrative neoplastic plexopathy and is more likely to involve the upper trunk. Diagnosis is made based on imaging studies, and the finding of myokymia on electrodiagnostic study is highly suggestive of a radiation-induced cause but does not exclude tumor. Peripheral neuropathies are less common and occur with cumulative doses greater than 6000 Gy but can be seen with lower doses. The phrenic and recurrent laryngeal nerves can be affected, and patients present with dysphagia, hoarseness, and respiratory distress. Radiation fibrosis management has been attempted with pentoxifylline, superoxide dismutase, hyperbaric oxygen therapy, and a combination of pentoxifylline and tocopherol, with mixed results.


NERVOUS SYSTEM CHEMOTHERAPY-INDUCED TOXICITY

Cancer patients experience numerous side effects from both the disease and the antineoplastic agents used to treat it. The CNS and peripheral nervous system
(PNS) are vulnerable targets of chemotherapy agents; however, the CNS is less vulnerable than the PNS to toxic exogenous agents owing to the blood–brain barrier. Therefore, with lower dose antineoplastic agents, changes will be seen in the PNS before the CNS.

1. Central Nervous System Toxicity

► General Considerations

Cognitive dysfunction occurs in non-CNS cancer patients, and it is unknown whether it is due to the disease, chemotherapy, or both. Often called “chemo-brain” or “chemo-fog,” this dysfunction varies greatly between patients and can affect executive function, attention, concentration, processing speed, reaction time, motor speed, and dexterity. These deficits affect the daily lives of cancer survivors.

► Pathogenesis

Numerous mechanisms have been proposed for cognitive dysfunction in cancer patients; these include direct neurotoxic effects of therapy, oxidative damage, immune dysregulation, microemboli, and genetic predisposition. The underlying mechanism of injury is still not fully understood, but risk factors include exposure to higher doses, synergistic effects of multiagent chemotherapy regimens, radiation, and intraarterial or intrathecal administration of chemotherapy with blood–brain barrier disruption. Studies have reported evidence for all of the mechanisms stated, but definitive proof of a single process is lacking, and discovery of one common pathway is unlikely. Furthermore, additional factors, such as depression, anxiety, fatigue, and sleep disturbances, have a direct impact on cognitive function. Cognitive deficits have also been seen prior to treatment in patients with non-CNS cancers.

► Clinical Findings

Assessment of cognitive function can be completed in numerous ways, including neuropsychological evaluation, neuroimaging, and subjectively through use of questionnaires. Brain imaging can be used to correlate cognitive deficits with
neurochemical mechanisms.

Functional magnetic resonance (fMRI) imaging evaluates in vivo brain function by detecting blood flow changes. One fMRI study in breast cancer survivors 3 years after chemotherapy found decreased memory encoding–related activity in the prefrontal cortex. Currently, findings from the functional imaging literature are inconsistent, making it difficult to apply this research to clinical practice. Although more studies are needed to determine clinical relevance, in the future this modality might be able to identify cancer patients at risk for cognitive decline.

Subjective evaluations are used frequently in medicine to identify patient symptomatology and quality-of-life measures. These are easy to score and many have been validated. The Functional Assessment of Cancer Therapy–Cognitive Function (FACT-cog) scale is a measure of cognitive complaints and is composed of behaviorally based items in an effort to minimize the impact of distress unrelated to cognitive abilities. Scores have been correlated with depression, anxiety, and fatigue, but not with neuropsychologic performance.

**Treatment**

Cognitive dysfunction experienced by cancer survivors is not well understood, but it is important to address the overall disabilities that are associated with these deficits. Treatment can be as simple as addressing increased levels of stress or consulting a neuropsychologist to create a plan to cope with the deficits. With the wide range of cognitive deficits, it is unlikely that a specific pharmacologic intervention will be identified to address this multifactorial dysfunction.


2. Peripheral Nervous System Toxicity

▶ General Considerations

Chemotherapy-induced peripheral neuropathy (CIPN) occurs as a result of the accumulation of neurotoxins in the neuronal cell bodies. Neurologic manifestations vary based on the class of antineoplastic agents and dose. Platinum compounds are widely used antineoplastic agents that can accumulate in the dorsal root ganglion, causing a sensory neuronopathy. Two cellular mechanisms in platinum-induced neuropathy have been proposed. The first suggests that apoptosis occurs as a result of a change in the DNA structure that reenters the dorsal root ganglion neurons; the second, that oxidative stress and mitochondrial dysfunction cause apoptosis.

▶ Pathogenesis & Clinical Findings

CIPN causes distal loss of vibratory sensation, loss of muscle stretch reflex, and paresthesias in the distal upper and lower limbs. Prolonged treatment can progress to proximal vibratory sensitivity loss and generalized loss of the muscle stress reflex. Severe loss of proprioception can lead to profound incoordination. Nerve conduction studies (NCS) reveal sensory axonal damage with reduced amplitude of sensory action potentials (SNAPs). A “coasting” phenomenon—progression of symptoms for weeks to months after cessation of treatment—also may be seen.

Vinca alkaloids are broad-spectrum agents used for many cancers. These antineoplastic agents have an affinity for tubulin, which causes a loss of axonal microtubules and alteration of their length that leads to axonal swelling in
myelinated and unmyelinated fibers. This dose-limiting toxicity is responsible for the development of distal axonal neuropathy affecting sensory more than motor fibers. Presentation is similar to that seen in diabetic neuropathy and is characterized by loss or decrease of the muscle stress reflex followed by paresthesias. Without cessation of therapy, muscle weakness can occur. This is most evident in the ankle dorsiflexors or toe and finger extensors and, to a lesser extent, in intrinsic hand muscles.

Autonomic neuropathy can occur prior to the development of the aforementioned symptoms of peripheral neuropathy. NCS show sensorimotor axonal neuropathy with a reduction in amplitude of SNAPs and compound muscle action potentials (CMAPs). Patients who have hereditary peripheral neuropathy, Charcot-Marie-Tooth type I, or a family history of this condition should not be given vincristine as it can cause an acute profound neuropathy. This vincristine-induced neuropathy is usually reversible upon discontinuation of therapy, but coasting phenomenon can occur in 30% of patients treated with high-dose regimens.

Taxanes (cabazitaxel, docetaxel, and paclitaxel) are microtubule-stabilizing agents that are effective in treating various solid tumors. Paclitaxel most commonly causes a sensory axon neuropathy that is dose dependent, but it can also cause a motor neuropathy affecting primarily proximal muscles. Taxanes promote assembly of disorganized microtubules. This process starts distally and progresses proximally, similar to the dying-back neuropathies that may have their origin in the cell body or in axonal transport. Clinically, this produces paresthesias in a stocking-and-glove distribution, affecting both the large (proprioception, vibration) and small (temperature, pinprick) fibers, with loss of the muscle stress reflex.

Similar to the mechanism of paclitaxel, epothilones are also microtubule-stabilizing antineoplastic agents. They also cause sensory peripheral neuropathy, but there are no studies that show this electrodiagnostically.

Bortezomib is a proteasome inhibitor that is used to treat refractory progressive myeloma. It may cause a distal, painful sensory neuropathy in a stocking-and-glove distribution, with decreased or absent muscle stress reflex and loss of proprioception. Several small studies in humans and rats demonstrated an increased risk of neuropathy when bortezomib was used in patients with preexisting neuropathies. On NCS, a predominantly sensory, axonal polyneuropathy is seen, affecting the sural SNAP more than the radial SNAP in a length-dependent pattern.

Thalidomide is notorious for its teratogenic effects but was approved by the
U.S. Food and Drug Administration (FDA) for treatment of multiple myeloma and chronic graft-versus-host disease, among others. The symmetric distal sensory neuropathy it produces is dose limiting and affects small and large fibers, with minor motor and posterior column involvement. In one study NCS showed a sensory axonal neuropathy in 40% of subjects, with reduced SNAP amplitude in a length-dependent pattern, and a sensorimotor peripheral neuropathy in 26.6%, with CMAP amplitude significantly reduced.

Nelarabine is a purine analog used to treat T-cell acute lymphoblastic leukemia. It is associated with development of a poorly characterized peripheral neuropathy that results in leg weakness and paresthesias. On rare occasions effects are severe, mimicking Guillain-Barré syndrome; treatment cessation does not always result in full recovery.

## Treatment

While CIPN is an acceptable side effect for a lifesaving treatment, it may result in reduced function and quality of life for patients. Patients must be educated about side effects of chemotherapy agents so they can monitor their symptoms and be accountable for their health. Screening for side effects of antineoplastic agent is facilitated by asking patient-specific questions to rule out CIPN. Over the course of chemotherapy, patients are seen by numerous health care providers. It is important for physiatrists to ask quality-of-life questions that not only screen for CIPN, but also are vital in promoting return to function for cancer survivors.


VELASCO R, PETIT J, CLAPÉS V, ET AL: NEUROLOGICAL MONITORING REDUCES THE

OTHER CENTRAL NERVOUS SYSTEM COMPLICATIONS

Tumors involving the CNS may arise from the spine or from the brain as primary tumors or metastatic lesions. Gliomas and lymphomas are the two most common primary CNS tumors in adults, while cerebellar astrocytomas are the most common in children. Lung, breast, skin (melanoma), and gastrointestinal tumors account for the majority of brain metastasis. Headache, cognitive impairment, weakness, gait disturbance, seizures, and speech and swallowing difficulties are common presentations. Treatment options may include use of corticosteroids to reduce edema, whole-brain radiation, and surgical resection.

Tumors arising from the spine are most commonly metastatic and epidural. Commonly metastasizing to the spine are lung, breast, prostate, and kidney tumors. Pain is the most common presenting symptom of epidural spinal cord compression (ESCC). ESCC is a serious complication of spinal tumors and represents a medical emergency. It is associated with rapid neurologic decline and may progress to paraplegia or tetraplegia. High-dose intravenous corticosteroids are appropriate acute management of ESCC while surgical decompression is being arranged. Radiotherapy has become an integral part of the management of most epidural tumors. Single and hypofractionated radiotherapeutic techniques are showing promise in better controlling tumor growth, particularly with radioresistant disease.

The peripheral nervous system is also affected by tumor cells, causing radiculopathy, plexopathy, and peripheral neuropathies. Radiculopathy occurs through hematogenous spread and by invasion of the paravertebral space. This type of cancer pain produces dysesthetic, burning pain in the affected dermatome. Horner syndrome may also be seen if the lower cervical or upper thoracic roots are involved. MRI and electromyography are used in diagnosis, and radiation may effectively alleviate some pain.

Tumors at the apex of the lung or breast cancers are causes of malignant brachial plexopathy, affecting the inferior trunk and medial cord of the brachial plexus. In comparison, radiation-induced plexopathy typically affects the upper plexus. Pain in the shoulder is the most common presenting symptom of malignant plexopathy and helps to distinguish it from painless radiation-induced plexopathy. Patients with radiation plexopathies typically present with weakness in the C5–6 myotomes. Lumbosacral plexopathies may occur in conjunction
with colorectal cancers, retroperitoneal sarcomas, or metastatic lesions. Back, buttock and leg pain are common presenting signs. For diagnosis, chest radiographs are useful in assessing the lung apex, whereas computed tomography (CT) or MRI of the abdomen can assess the retroperitoneal space for compressive masses. MRI with gadolinium is the best method to visualize the plexuses. Electromyography can help distinguish plexopathy from radiculopathy. The presence of myokymia on needle examination is frequently associated with radiation plexopathy but can also be seen in root and peripheral nerve lesions. Treatment consists of corticosteroid administration and radiation.

Peripheral neuropathies usually occur as a result of nerve compression by bony metastasis or as a side effect of chemotherapy. Some sites of compression include the radial nerve at the humerus, the sciatic nerve in the pelvis, and the intercostal nerves.


METASTATIC BONE DISEASE

General Considerations & Pathogenesis

A frequent complication of visceral tumors is bone metastasis, which is a common occurrence in advanced breast, prostate, lung, renal, and thyroid carcinomas. Most solid tumors preferentially metastasize to the axial skeleton with predilection for the pelvis, femur, vertebrae, and ribs. Metastatic bone lesions may cause pain, neurologic compromise, pathologic fractures, and limitations in function and mobility. Lesions can be classified as osteolytic, osteoblastic, or mixed, with osteolytic lesions being the most common and possessing the greatest danger of causing pathologic fracture. In addition to metastatic bone lesions, bone integrity may be further reduced from long-term glucocorticoid use or hormonal therapy (selective estrogen receptor modulators, aromatase inhibitors, androgen deprivation therapy). Vertebral compression fractures arise at an escalating risk when radiotherapy fractions exceed 20 Gy.
Clinical Findings

Pain is the most common presenting sign. Intensified bone pain elicited with activity is commonly the first sign of an impending fracture. Pain may be described as deep, piercing, and worse at night (in contrast to degenerative joint pain, which is worse with activity). Pain emanating from the vertebrae occurs in a predictable pattern. Lesions affecting C7–T1 cause referred pain to the interscapular region, and lesions from T12 to L1 refer pain to the sacroiliac joint or iliac crest.

The most sensitive imaging modality for diagnosing skeletal metastasis is radionuclide bone scan. PET scans offer valuable information; however, conventional radiographs are of limited value because involvement of 30–50% of the cortex must be present for visualization. CT scan can be beneficial in early bone metastasis, particularly in patients with PET-positive and radiographic-negative lesions.

Treatment

The treatment algorithm for metastatic bone disease provides multiple options. Bisphosphonates act by inducing apoptosis of osteoclasts and thereby restrict growth of osteolytic lesions. They have been shown to reduce bone pain and prevent fractures. Spinal orthotics can be used to moderate pain and protect neurovascular structures during the healing process. The primary goal of palliative radiotherapy is pain control, restoration of function, and arrest of malignant growth.

Prophylactic surgical resection and stabilization confers superior results in functional outcomes compared with surgical repair after pathologic fracture has already occurred. Given the associated risks of morbidity and mortality with surgery, it is imperative to appropriately select patients in whom the risk for impending fracture is significant. This determination should be made in conjunction with the physiatrist and surgeon. Among the several scoring systems that have been used to predict risk of impending fracture, the Mirels’ and Spine Instability Neoplastic (SINS) scores demonstrate strong validity and sensitivity.

The Mirels’ score (Table 35–3) is a composite weighted scoring system based on tumor location, pain intensity, size, and radiologic appearance used for evaluation of long bone lesions. A total score of 9 or higher indicates a significant risk for fracture of the femur whereas a cutoff score of 7 is optimal for the humerus. Spinal instability from traumatic injuries and neoplastic
invasion behave in different patterns; therefore, standard measures of spinal stability should not be utilized in the neoplastic spine. The SINS quantifies risk of vertebral compression fractures based on tumor location, pain, bone lesion type, spinal alignment, vertebral collapse, and posterolateral spine involvement. A SINS score of 0 to 6 indicates spinal stability; intermediate stability is scored from 7 to 12; and a score ranging from 13 to 18 denotes frank instability. A surgical consultation should be made when a patient has a SINS score greater than 7, Mirels’ score greater than 9, persistent pain following irradiation, or any signs of neurologic compromise.

Table 35–3 Mirels’ scoring system for predicting risk of impending fracture.

<table>
<thead>
<tr>
<th>Variable</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>Mild</td>
<td>Moderate</td>
<td>Functional</td>
</tr>
<tr>
<td>Location</td>
<td>Upper extremity</td>
<td>Lower extremity</td>
<td>Peritrochanteric</td>
</tr>
<tr>
<td>Size</td>
<td>&lt; 1/3</td>
<td>1/3–2/3</td>
<td>&gt; 2/3</td>
</tr>
<tr>
<td>Nature</td>
<td>Blastic</td>
<td>Mixed</td>
<td>Lytic</td>
</tr>
</tbody>
</table>

There is increased risk of fractures with metastatic disease. An orthopedic consultation is important for patients with bone metastasis to determine the stability of the involved bones. Fractures are common in lytic lesions and in weight-bearing bones, with damage to cortical and trabecular bone. When less than one third of the diameter of a long bone is affected, pathologic fracture is relatively unusual, but above this amount and especially when more than 50% of the cortex is destroyed, the fracture rate increases markedly to approximately 80%. New back pain in a cancer patient warrants radiographic evaluation, and if there is an abnormality an MRI should be obtained. More than 60% of these patients will be myelopathic or have evidence of epidural lesions. Spinal instability causing back pain affects approximately 10% of this population.


**LYMPHEDEMA**

Detailed discussion of lymphedema appears earlier in this text, in Chapter 3. Lymphedema affects an estimated 3 million Americans and 30% of all breast cancer survivors. The most common cause worldwide is infectious (filariasis), but in developed countries, cancer treatment is the leading cause. Lymphedema is most frequently associated with breast cancer but it may also be seen with melanoma, lymphoma, and cancers affecting the pelvis, head, or neck regions.

By definition, lymphedema is edema caused by an imbalance between lymphatic fluid production and transport capacity within a low-pressure system (Figure 35–3). In patients with cancer, it is commonly caused by lymph node destruction from lymph node dissection or radiation. Patients who undergo axillary lymph node dissection in addition to radiation have significantly higher rates of secondary lymphedema than those undergoing sentinel lymph node biopsy alone. Additional factors predisposing to lymphedema are trauma, infection, obesity, and location of a tumor in the upper outer quadrant of the breast. In addition to presenting in any of the four limbs, lymphedema can occur in the genital area, torso, and head and neck region. (The latter is discussed separately, following this discussion.) Treatment fundamentals are essentially the same as those described earlier (see Chapter 3), with individualized
modifications to compression bandaging and garments for appropriate fit.

The swelling typically begins insidiously, and the great majority of cases
(77%) are diagnosed within 3 years of cancer treatment. Patients present with unilateral edema in the ipsilateral limb. Advanced stages are defined by progressive cutaneous and subcutaneous thickening, the hallmarks of which include Stemmer sign, peau d’orange, fibrosis, and pitting edema. Patients with early subclinical disease present with a sensation of heaviness, tightness, discomfort, or ache without any visible signs of swelling. Diagnostic workup must include ruling out edema secondary to deep vein thrombus, infection, tumor obstruction, heart failure, and renal insufficiency. Severe limb swelling occurring years after cancer treatment outside of any recent limb trauma or infection is highly suspicious for tumor recurrence.

Lymphedema can be quantified by various grading criteria, which measure circumferential differences between extremities (see Chapter 3). The management of lymphedema can be initiated at any stage, although the most favorable response occurs with early and multimodal treatment. As described in Chapter 3, complete decongestive therapy (CDT) is the gold standard treatment for lymphedema and comprises two phases: treatment and maintenance.


McLaughlin S: Prevalence of lymphedema in women with breast cancer 5 years after sentinel lymph node biopsy or axillary dissection J Clin Oncol 2008;26:5213–5219.


HEAD & NECK LYMPHEDEMA

General Considerations

Face and neck lymphedema is a common sequela of treatment modalities in cancers of the larynx, pharynx, thyroid, tonsils, oral cavity, lips, and salivary glands. The risk of lymphedema increases with the number of lymph nodes damaged or removed during surgery or radiation.

Clinical Findings

Head and neck lymphedema typically develops 2–6 months after treatment and is most severe in patients who receive a combination of surgery and radiotherapy. It is worse in the morning, because fluid pools during nighttime recumbent positioning. During the daytime, head and neck lymphedema is minimal because gravity promotes lymph node drainage when upright or seated. Therefore, it can be difficult to assess for the presence of head and neck lymphedema on examination.

Patients should be specifically asked if they have experienced face or neck swelling, a sensation of fullness, or difficulty with breathing or swallowing. Clinical symptoms include tightness; difficulty moving the neck or jaw; ptosis from eyelid swelling; dysphagia; dysarthria; respiratory compromise; sialorrhea; nasal congestion and chronic middle ear infection; and swelling of the eyes, face, lips, neck, and submandibular area. Internal lymphedema affects structures within the aerodigestive tract, causing difficulties with breathing, swallowing, and eating. Confirmation of internal lymphedema requires visualization with a flexible endoscope (Figure 35–4).
**Figure 35–4** Internal lymphedema. **A:** Normal endoscopic view of the upper aerodigestive tract. **B:** Internal lymphedema in a patient with head and neck cancer. (Reproduced with permission from American College of Surgeons, Chicago, Illinois.)

**Treatment**

A multidisciplinary approach to care that includes a speech and swallow therapist and otolaryngologist is imperative. Treatment makes use of the standard lymphedema management techniques (described in Chapter 3), as well as compensatory speech and swallowing techniques (described in Chapter 38). Use of higher doses of corticosteroids may be considered to limit edema. Management should include CDT, self-manual lymphatic drainage (self-MLD), and the use of a nocturnal compressive mask. Refer to Lymphedema, earlier, for more details.


**BREAST CANCER COMPLICATIONS**

Many complications can occur following treatment for breast cancer. This section highlights several of the most common.

Following axillary lymph node dissection or sentinel node biopsy, axillary web syndrome is a frequent occurrence. It usually occurs in the first several weeks following surgical treatment for breast cancer. Presenting symptoms and signs include pain in the medial arm, limited shoulder range of motion (especially abduction), and palpable cords of subcutaneous tissue, extending from the axilla into the medial arm and, in severe cases, down to the base of the thumb. It is unclear if the palpable cords represent pathologic changes in the lymphatic or superficial venous system. In most patients, symptoms resolve after 2–3 months, and treatment may consist of manual soft tissue techniques, range of motion exercises, and lymphatic drainage.

Postmastectomy pain syndrome is another common occurrence following breast surgery. The syndrome includes a number of disorders related to mastectomy and reconstruction, including phantom breast pain, incisional
allodynia, neuroma formation, pectoralis muscle pain, and intercostobrachial neuropathy. Many of the disorders included in postmastectomy pain syndrome are believed to be neuropathic in origin. A somatic component, such as spasm of the pectoralis major and serratus anterior muscles following breast reconstruction, is often present. Pain may be present in the axilla, chest wall, shoulder, or arm. Phantom breast pain is common after mastectomy and may interfere with activities of daily living as well as worsen the pain cycle. Intercostal neuromas are potential complications following mastectomy and breast reconstruction. If treated surgically with resection, pain control can be achieved.

Arthralgias, particularly of the shoulder, are another common complaint following chemotherapeutic and other endocrine treatments (i.e., aromatase inhibitors) for breast cancer. Shoulder pain may manifest as frozen shoulder, rotator cuff dysfunction, or myofascial pain. In addition to being painful, these conditions affect the quality of life of breast cancer survivors. A common sequela of endocrine treatments is bone loss, contributing to osteoporosis and fractures, further impeding quality of life. Thus, early intervention is warranted. Therapeutic exercise, supplementation with vitamin D, dual-energy X-ray absorptiometry (DEXA) scans, and treatment with nonsteroidal antiinflammatory drugs can ameliorate these musculoskeletal complaints. A secondary type of arthralgia in breast cancer patients results from use of aromatase inhibitors. Although these agents have been shown to be more beneficial than tamoxifen, complaints of arthralgias are common. Switching the patient to a different type of aromatase inhibitor may alleviate some of this pain.


Head and neck cancer accounts for 640,000 cases per year worldwide, making it the sixth most common cancer. Greater than 95% of all cases are squamous cell by pathology. Major risk factors are tobacco use and heavy alcohol use. The risk of death increases significantly with each additional pack year of tobacco use. Overall prognosis is poor with a 5-year survival rate of 30–40%. A small percentage of these cancers are attributable to infection with human papillomavirus (HPV), predominantly type 16. The HPV-associated head and neck cancers are associated with a better overall prognosis and a significantly increased 3-year survival rate.

Current recommendations for treatment are chemoradiation with possible dissection. To minimize toxicities, IMRT and modified or selective neck dissections are increasingly being used. Complications are abundant and can occur long after treatment. Severe late toxicity after chemoradiation is common and associated with older age, advanced T stage, and primary cancers of the larynx or hypopharynx. Given the high risk for complications in this population, it is optimal to have a physiatrist assess baseline maximal interincisal distance (MID), and neck and shoulder range of motion and strength, to enable earlier intervention to prevent debilitating complications.


MUCOSITIS & XEROSTOMIA

Complications within the oral cavity are common among patients undergoing cancer treatment. Mucositis, an acute effect of radiation, is ulceration of the oral mucosa. It can lead to pain and vomiting from thick secretions and infections. This should be managed with meticulous dental care, use of lubricants, use of humidifiers, and pain medication.
Xerostomia, decreased salivation, is the most common late side effect of chemotherapy and radiotherapy. Risk of damage is dose dependent, with detectable parotid dysfunction at 10–15 Gy and doses between 40 and 50 Gy causing greater than 75% of cases. The use of conformal radiotherapy techniques to minimize parotid gland exposure and administration of amifostine before each fractionated dose have been shown to reduce the incidence of xerostomia. However, the drug’s major side effects of hypotension, nausea, vomiting, and allergic reactions limit its use clinically.

Xerostomia increases a patient’s risk for infection, dental caries, poor nutritional intake, and speech difficulties. Treatment is often multimodal and may include saliva substitutes, frequent sips of water, hard sugarless candy, gum, and avoidance of food with a dry, hard consistency. Gustatory stimulation of saliva can be induced by use of acidic or bitter lozenges. In addition, surgical transfer of the submandibular gland out of the radiation portal has served to preserve salivary gland function. Pilocarpine, a cholinergic parasympathomimetic, is effective in half of treated patients but side effects include dizziness, vasodilation, headache, nausea, diarrhea, dyspepsia, and sweating. The drug requires lifelong treatment and must be used cautiously in patients with cardiac disease or asthma. Cevimeline and bethanechol are other cholinergic agonists for which long-term studies documenting adverse effects are limited. Hyperbaric oxygen has shown modest effects, but further confirmatory studies are needed. Acupuncture offers an effective and long-lasting treatment modality to stimulate increased saliva flow rates in patients with residual gland function and has a favorable side effect profile.


OSTEORADIONECROSIS
Osteoradionecrosis (ORN) is a late complication of high-dose radiotherapy, with a reported incidence of 5–15%. The most common site is the mandible. ORN typically occurs 6 months to 5 years after irradiation. The mechanism has been described as a primarily metabolic event rather than infectious. Radiation induces a matrix that is hypoxic, hypovascular, and hypocellular (the 3 H’s), leading to tissue breakdown and impaired tissue response to injury. This, in turn, allows for a chronic nonhealing wound.

ORN is associated with location of the primary tumor near the mandible, T stage, dentition, surgical complications, concomitant chemotherapy, nutritional status, and the radiation dose and delivery method by brachytherapy. The inciting factor in most cases (88%) is trauma from tooth extraction. Consequently, prior to radiotherapy, all unhealthy teeth should be extracted. A delay of 2–3 weeks is then necessary to allow for complete healing before beginning the radiation regimen. Unnecessary postradiation tooth extractions and trauma should be avoided.

The presentation is variable, ranging from asymptomatic to severe necrosis with pathologic fracture, orocutaneous fistula formation, severe pain, and osteomyelitis. Diagnosis is made through imaging studies, using panoramic radiographs, MRI, or CT scan.

Patients with mild asymptomatic cases may be managed conservatively with antibiotics, pentoxifylline–vitamin E, and local debridement. Refractory ORN is treated with segmental mandibulectomy and reconstructive surgery. Hyperbaric oxygen has been used in combination with debridement, although studies are limited and results are conflicting.


TRISMUS

Trismus is the tonic contraction, spasm, or fibrosis of muscles of mastication as a result of radiotherapy, leading to limitations in mouth opening. There is no
absolute consensus on the distance of mouth opening that constitutes trismus; however, a maximal interincisal distance of less than 20–40 mm is considered to be indicative. Overall prevalence is between 5% and 38%, with a 25% prevalence using older radiotherapy techniques and 5% using IMRT. The risk of trismus is increased in patients who have received prior radiotherapy treatment and in those receiving doses greater than 60 Gy to the temporomandibular joint, medial pterygoids, or masseter muscles. Radiation-induced trismus may occur anytime within 24 months of therapy, and symptoms may become progressive. Complications, which include nutritional deficits, weight loss, risk of dental caries, ORN, decreased phonation, social isolation, and aspiration risk, have a significant impact on patient quality of life.

Management should be multimodal, including physical therapy with a focus on formation of a daily home exercise plan, pain medication, passive motion devices, and aggressive dental care. Passive motion devices should be avoiding during radiotherapy but can be used early in the postoperative period. Botulinum toxin injection to the masseter and medial pterygoids is effective in relieving pain and alleviating muscle spasms but has not been associated with significant improvement in jaw opening. Dynamic jaw-opening devices are likely effective in improving jaw range of motion even in patients with chronic trismus related to radiation. Microcurrent electrotherapy and oral pentoxifylline have been found to be effective modalities, as well.


SPEECH & SWALLOW DYSFUNCTION

General Considerations & Pathogenesis
Speech dysfunction is primarily caused by cancers of the oral cavity, tongue, and pharynx, in contrast to swallowing dysfunction, which can occur with cancers anywhere within the aerodigestive tract. Furthermore, radiotherapy has a more significant effect on swallowing than speech production, which can be exacerbated by concomitant chemotherapy. Dysphagia following chemoradiation therapy results directly from fibrosis of soft tissue structures and cranial neuropathies, causing denervation of suprahyoid musculature. The radiation-induced alteration of neutral pH saliva to acidic and thick saliva further complicates swallowing. Aspiration risk is highly correlated to prior smoking history, radiation doses greater than 50 Gy to the larynx, and greater than 54 Gy to the inferior pharyngeal constrictor muscles.

### Clinical Findings

Speech dysfunction can be mild (eg, dysarthria and hypophonation) or severe (eg, aphonia) and may be caused by surgical resection, radiation injury, or nerve damage affecting the vocal cords. Partial laryngectomy may have mild effects on voice quality as compared with total laryngectomy, which always results in complete loss of voice.

### Treatment

Multiple studies have documented that quality of life after head and neck cancer treatment was directly related to swallowing functions, highlighting the importance of aggressive and early management. Early intervention by speech and swallow therapists with swallowing exercises can have restorative effects on function. Feeding tubes as a temporizing or long-term measure are needed in only 3% of cases.

Mild speech dysfunction may be managed with speech therapy and the use of a palatal augmentation prosthesis for partial glossectomies. A total laryngectomy can be managed with an artificial larynx, tracheoesophageal prosthesis (TEP), or training in esophageal speech. Speech with an artificial larynx produces a computerized voice that is unnatural in sound. Esophageal speech calls for the coordination of inhaling and releasing air into the esophagus to create vibration and hence sound. This technique is challenging to learn and only a quarter of the patients master it. The preferred method is TEP, because it allows for early restoration of speech. TEP is a surgical procedure that creates a connection
between a stoma and the esophagus and is used with a prosthesis inside the tract. Speech is produced during exhalation while covering the stoma to allow vibrating airflow into the pharynx and esophagus. Irrespective of the method of speech production chosen, speech therapy is a critical component of management. (Chapter 38 discusses this topic in detail.)


SPINAL ACCESSORY NEUROPATHY

Spinal accessory neuropathy is a commonly encountered complication in patients with head and neck cancer. Although modified and functional neck dissections by definition spare the accessory nerve, multiple studies document the development of spinal accessory neuropathy in patients despite undergoing these nerve-sparing procedures. Radiation-induced injury to this nerve is also a common cause. The spinal accessory nerve innervates the sternocleidomastoid and trapezius muscles.

Individuals affected with spinal accessory neuropathy assume a resting posture of ipsilateral shoulder depression and protraction. The shoulder retractors become overly stretched with overactivity of the pectoral musculature, creating a protracted posture (Figure 35–5). Functional limitations include inability to fully abduct the ipsilateral arm and lateral winging of the scapula with shoulder abduction. Rehabilitation should focus on stretching the pectoral muscles and strengthening the rhomboids and levator scapula, with an emphasis on scapular stabilization. Biofeedback, TENS, and Kinesio tape can aid in the rehabilitation process. In severe cases, a shoulder stabilization orthotic maybe used to aid in stabilizing the shoulder (for prevention of shoulder subluxation) or the scapula (for improved function).
**Figure 35–5** Accessory neuropathy. This male patient underwent tonsillectomy and radiation therapy for treatment of tonsillar carcinoma 13 years earlier. He presented with new onset of right upper extremity radiation-induced accessory neuropathy as evidenced by myokymia on electrodiagnostic study. Notice the depressed and protracted resting posture of the right shoulder and the inability to fully abduct the right shoulder. Although not seen in this image, the patient also had lateral scapular winging.


**CERVICAL DYSTONIA**

**General Considerations & Pathogenesis**

The sternocleidomastoid, scalenes, and trapezius muscles are superficial neck muscles that are frequently injured during mantle radiation. These muscles can become fibrotic, thereby limiting range of motion, and, if untreated, can become contracted. They can also develop spasms, which can progress to a dystonic posture. Unilateral neck radiation often results in a torticollic posture characterized by contralateral neck rotation and ipsilateral lateral neck flexion from ipsilateral sternocleidomastoid and anterior scalene contracture or spasm.

**Clinical Findings**

Patients often present with significant neck and shoulder pain, headaches, and restricted range of motion. Management should include physical therapy, with a focus on stretching exercises, myofascial tissue release, and soft tissue mobilization, and consideration of heat or TENS modalities. Muscle relaxants are rarely helpful but treatment with nerve-stabilizing medications, trigger point injections, or botulinum toxin injection can provide considerable relief.

A common late complication following the use of mantle field radiation to treat Hodgkin’s lymphoma is neck extensor weakness (dropped head syndrome). Patients present with severe atrophy and weakness of the shoulder and
cervicothoracic paraspinal muscles. Although, the exact mechanism of injury is not clear, dropped head syndrome has been attributed to various degrees of radiation-induced damage to the entire neuromuscular axis, including the spinal cord, nerve roots, plexus (cervical plexus), local peripheral nerves, and muscles within the radiation field. The term “myelo-radiculo-plexo-neuro-myopathy” has been coined to describe this phenomenon. On average, it typically occurs 19.7 years (range, 5–30 years) after irradiation.

**Treatment**

While it remains an irreversible complication, patients with progressive head drop can benefit cosmetically and functionally from use of a Headmaster neck orthotic.

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**CANCER REHABILITATION SERVICES**

Rehabilitation is an essential aspect of the cancer treatment process. Unfortunately, there is often a lack of awareness of the dysfunctions involving ambulation and activities of daily living that impede the lives of cancer patients, as well as survivors. It is not only health care practitioners who lack awareness of the role that rehabilitation can play in improving the lives of these patients. Third party payers and the Centers for Medicare and Medicaid Services do not recognize the need for acute rehabilitation programs for cancer patients. Furthermore, cancer diagnoses are not considered to be compliant with admission criteria for inpatient rehabilitation facilities (IRF).

Nonetheless, cancer patients can make significant, functional gains when provided with acute inpatient rehabilitation services, as shown by improvements in their Functional Independence Measure (FIM) scores. Cancer patients who undergo chemotherapy or radiation during an acute inpatient rehabilitation stay also have improved functional outcomes relative to their counterparts who receive cancer treatment prior to rehabilitation, or no rehabilitation at all. Restoration of function in cancer patients is vital to their quality of life, resulting
in improvement in fatigue, physical functioning, and mental health. Although cancer patients have a higher transfer rate back to acute care than do noncancer patients, the reason for readmission is typically not related to their cancer diagnosis.

Outpatient rehabilitation for cancer patients addresses a broad spectrum of issues related to cancer treatment and complications, such as lymphedema, contractures, pain, ambulation, speech and swallow problems, and dysfunctions in activities of daily living. Outpatient rehabilitation programs are successful in improving physical and psychosocial functioning and reducing symptoms. Communication between physiatrist and therapist is vital for ensuring patients’ continued progress. Rehabilitation requires an interdisciplinary approach that utilizes physical, occupational, speech, and recreational therapies. All disciplines play a vital role in the restoration of function to these patients.

The American College of Sports Medicine (ACSM) has developed exercise guidelines for cancer patients, including recommendations for pre-exercise assessment and medical evaluations. Additionally, the U.S. Department of Health and Human Services Physical Activities Guidelines for Americans include adjustments for cancer survivors. ACSM recommends evaluation of peripheral neuropathies and musculoskeletal morbidities, determination of fracture risk in patients treated with hormonal therapy, skeletal evaluation for patients with known metastatic disease, and a medical examination that includes stress testing for individuals with preexisting cardiac conditions. There are additional recommendations for specific cancers. For example, patients with a history of breast cancer should have the upper extremity evaluated; prostate cancer patients should be evaluated for muscle weakness and wasting; colon cancer patients with an ostomy require consistent and proactive infection prevention; and gynecologic cancer patients should be evaluated for lymphedema.

Among cancer survivors, contraindications to exercise include adequate time to heal after surgery, extreme fatigue, anemia, ataxia, deep venous thrombosis, and severe thrombocytopenia. Additionally, exercise should be temporarily discontinued in the following instances: in breast cancer patients if changes in arm or shoulder pain, inflammation, or swelling occur; in colon cancer patients if hernia or ostomy-related systemic infection develops; and in gynecologic cancer patients if changes in swelling or inflammation are noted in the abdomen, groin, or lower limbs.

Adamsen L, Quist M, Andersen C, et al: Effect of a multimodal high intensity


Medical Emergencies in Rehabilitation Medicine

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As the role of physiatrists continues to evolve, the practice of rehabilitation medicine may pose additional challenges to physicians who manage patients with acute and chronic conditions that affect overall function. Medical emergencies, although infrequent, are reported in the rehabilitation setting, and staff must be trained in appropriate responses. The physiatrist must also know when to request early consultation to prevent minor medical problems from developing into major medical catastrophes. Physiatrists are frequently the “gatekeepers” of medical and nonmedical services when patients are participating in rehabilitation. The idea of gatekeeping, as promoted in other specialties such as family medicine, internal medicine, and pediatrics, requires that physiatrists be involved in the general coordination and medical care of patients with disability.

Health insurers, particularly Medicare, have continued to promote financial incentives for inpatient rehabilitation for acutely ill patients and patients with several comorbidities. Patients are admitted to an acute inpatient rehabilitation unit with the expectation that they will be able to tolerate or participate in at least 3 hours of rehabilitation services per day. The broader interpretation of the 3-hour rule supports the treatment of medical problems that previously required transfer to an acute medical floor. Acute medical events, such as deep vein thrombosis, pneumonia, chest pain, or high blood pressure, are treated in the inpatient rehabilitation unit—unless these conditions suggest hemodynamic
inability—with the expectation that the patient’s progress and functional recovery may be interrupted many times during the acute rehabilitation program. In addition, many managed health care providers, as well as Medicare, will not pay for hospital-based inpatient rehabilitation programs unless medical justification is explicitly defined to keep the patient in a hospital setting. Patients are expected to have active comorbidities that require medical management by a physiatrist or a consultant during their progression through functionally oriented programs, and merit a hospital-based rehabilitation stay. Otherwise, less-expensive settings for participating in rehabilitation programs are sought.

Several factors have contributed to changes in the types of patients who utilize acute rehabilitation settings, among them:

- Older patients are participating in rehabilitation. More people are living beyond the age of 65 years. Aging baby boomers are becoming more involved in their health care management.
- Sicker patients are participating in rehabilitation (eg, those with traumatic brain injury, spinal cord injury, and multiple traumas).
- More people with chronic diseases that are treated with highly technical surgical and medical interventions are participating in rehabilitation.

Thus, changes in the overall demographic distribution, advancing medical and surgical knowledge and methods, and social policies directing cost-containment are creating a rehabilitation population at greater risk for developing medical emergencies that may interfere with traditional rehabilitation programs. Rehabilitation professionals are expected to prevent emergencies by anticipating the anatomic and pathophysiologic impact of rehabilitation interventions in such high-risk patients.


AUTONOMIC DYSREFLEXIA

General Considerations

Autonomic dysreflexia has been described as the classic rehabilitation emergency. This life-threatening condition occurs in patients with a spinal cord injury (SCI) at the level of T6 or above as a consequence of unchecked sympathetic tone. (Complications of SCI are described in detail in Chapter 12.)

Clinical Findings

Symptoms include a sudden increase in systolic blood pressure that is 20 mm Hg above baseline, pounding headache, flushing or sweating above the lesion (ie, face, neck, and shoulders), nasal congestion, blurry vision, piloerection, and meiosis. Bradycardia is classically described, although not always present, and tachycardia may even occur. In evaluating the increase in blood pressure, it is important to note that the normal systolic blood pressure for a patient with an SCI above T6 is 90–110 mm Hg. The differential diagnosis is limited and includes increased intracranial pressure, with resultant hypertension and bradycardia (particularly if the patient has a known concomitant traumatic brain injury), and pheochromocytoma.

Immediate treatment must ensue to prevent potentially life-threatening complications, such as cerebrovascular accident or intracranial hemorrhage, retinal hemorrhage, seizure, myocardial infarction, cardiac arrhythmia, and pulmonary edema.

Treatment

The first step in management is to raise the head of the bed or sit the patient up, thus causing blood to pool in the lower limbs and abdomen. Next, all
constricting clothing and external devices, such as compression stockings, abdominal binders, and Foley leg bags, should be removed or loosened. The key component in management is then to quickly survey the patient and alleviate any noxious stimuli below the level of the SCI. The urinary system is the most common source, stemming from bladder distention or irritation. Other considerations are fecal impaction, in-grown toenails, pain, menstruation, preeclampsia (if pregnant), and abdominal emergencies, such as appendicitis, cholecystitis, or pancreatitis.

Evaluation should begin with the urinary system. If the patient has an indwelling catheter, the entire length should be checked for the presence of kinks or obstructions, proper placement should be verified, and the tube should be flushed. If the patient does not have a Foley catheter, a straight catheter can be inserted with lidocaine jelly. Attention should then be given to the bowels and fecal impaction, although the physician can consider waiting until after medications have been given, as disimpacting with lidocaine jelly can initially worsen autonomic dysreflexia. When turning the patient to check for stool, recall that the more acute and more cephalad the injury, the greater the risk of vagal-mediated bradycardia, which can compound bradycardia associated with autonomic dysreflexia. Throughout these steps, blood pressure should be checked every 2–5 minutes until the patient is stabilized.

If the systolic blood pressure remains greater than 150 mm Hg, medications should be started while further causes are investigated. First-line treatments are nifedipine or nitroglycerin. The advantage of topical nitroglycerin, in patch or paste form, is that it can be removed if the condition resolves or blood pressure drops too low. If nitroglycerin patch or paste is used, 1–2 inches is placed above the lesion; alternately, a 0.4-mg sublingual nitroglycerin tablet may be given. Nifedipine, 10 mg, can be provided in capsule form, which can be chewed or crushed and swallowed for rapid absorption. This dose can be repeated in 30 minutes if necessary. Caution is advised when using nitroglycerin as many individuals with SCI take phosphodiesterase inhibitors for erectile dysfunction and interaction of these drugs could result in drastic hypotension. Because blood pressure tends to normalize rapidly after the inciting event of autonomic dysreflexia is removed, long-acting antihypertensive medication is not recommended as significant hypotension may ensue. β blockers should be avoided as they may cause excessive α-adrenergic activity.

Blood pressure should continue to be monitored for at least 2 hours after initial improvement. Persistence of autonomic dysreflexia necessitates transfer to an intensive care unit (ICU) or emergency department for close hemodynamic
and telemetry monitoring and titration of more aggressive intravenous blood pressure medication, such as hydralazine, nitroglycerine, or nitroprusside.


**SYMPATHETIC STORM IN TRAUMATIC BRAIN INJURY**

### General Considerations

Sympathetic storm describes a disturbance in autonomic control that goes by many names, with some preferring the term *paroxysmal autonomic instability with dystonia (PAID)*. It occurs in 15–35% of patients with severe traumatic brain injury. When uncontrolled it can result in arrhythmias, myocardial infarction, neurogenic pulmonary edema, and secondary brain injury as a result of intracranial hemorrhage, increased intracranial pressure, increased temperature, or some combination of these.
Clinical Findings

Clinical manifestations may include any combination of paroxysmal episodes of tachycardia, hypertension, tachypnea, hyperpyrexia, agitation, diaphoresis, and dystonia. The differential diagnosis must initially include infection, especially when fever is a predominant feature, although other entities should be considered. These include delirium tremens, thyroid storm, neuroleptic malignant and serotonin syndromes (if causative medications have been utilized), and intrathecal baclofen withdrawal.

Treatment

The management of PAID begins with an evaluation for any noxious stimuli, as described earlier in the initial treatment of autonomic dysreflexia in SCI. A calm and quiet environment should be provided. An infectious source of fever should also be sought if there has not been an extensive recent workup. Additional tests to rule out alternative diagnoses may include electrocardiography (ECG), chest radiography, thyroid panel, basic metabolic panel, and creatine kinase level.

Medication should be initiated if PAID persists despite environmental control and alleviation of potential noxious stimuli. First-line medications are propranolol or opioids, with the choice depending on the predominant symptoms. For hypertension and tachycardia, propranolol may be started at 10 mg every 12 hours and titrated up in 10-mg intervals. Oxycodone or morphine can be effective to decrease the sympathetic outflow, as well as treat unresolved pain that may be triggering the storm. Oxycodone can be given at a dosage of 5 mg every 4 hours and titrated up in 5-mg increments or, if enteral access is not possible, morphine, 2 mg intravenously, can be given every 4 hours and increased by 1 or 2 mg with successive doses.

Alternative, less frequently used medications include gabapentin and bromocriptine. Gabapentin has been found to be effective at starting dosages of 300 mg three times daily, with titration up to 600-mg doses. Caution should be used in patients with renal dysfunction (maximum dosage, 300–400 mg/day). For PAID that consists primarily of fever and diaphoresis, bromocriptine can be tried at dosages of 2.5–5 mg every 8 hours and may be titrated up to 30 or 40 mg daily. A cooling blanket or ice packs may also be helpful for patients with persistent high fevers.


SEIZURES

General Considerations

A primary rehabilitation diagnosis of traumatic brain injury, cerebrovascular accident, or other intracranial pathology places patients at a higher risk for seizures than the general population and is associated with more serious underlying causes of initial or breakthrough seizures, such as recurrent bleeding, infarction, and infectious foci. Uncontrolled seizures are medical emergencies that require rapid and aggressive treatment to prevent further neurologic damage and systemic complications. They can result in pronounced systemic decompensation, including respiratory failure, cardiac arrhythmias, hyperthermia, lactic acidosis, aspiration pneumonitis, and rhabdomyolysis.

Treatment

Initial management must consist of closely monitoring the airway and oxygenation, checking the blood glucose level, and obtaining intravenous access. Transfer to the emergency department or ICU should be initiated while further stabilization is being carried out, as described below.

Lorazepam is the first-line medication and should be given at 2–4 mg (0.1 mg/kg) intravenously and may be repeated at 2 mg/min, up to a maximum dose of 4 mg per dose and 8 mg in 12 hours. If intravenous access is not obtainable,
lorazepam may be given intramuscularly. If lorazepam is not available, a reasonable alternative is diazepam, 10 mg rectally.

If the patient is still seizing, phenytoin, 15–20 mg/kg intravenously, can be considered and repeated at 10–15 mg/kg 20 minutes later if necessary. Phenytoin should be given in a monitored setting, if possible, because of its proarrhythmic effects. Once the situation permits, several laboratory values should be obtained, including electrolytes, blood glucose, and seizure medication levels. Scheduling the patient for a computed tomography (CT) scan of the head without contrast should be considered when seizures are under better control.


FALLS

General Considerations

Falls are unfortunately common among patients with the functionally impairing diagnoses that require inpatient rehabilitation. Between 14% and 65% of stroke survivors experience falls while inpatients at an acute care or rehabilitation hospital. Falls in this population can result in serious injuries, such as fracture and intracranial hemorrhage. Hip fractures occur at a four times higher rate in patients with stroke than in the general population.

Treatment

The management of a fall, as with all emergencies, begins with verification of hemodynamic stability. Once the patient is assured to be stable, an extremely careful history and physical examination must ensue, both for patient safety and potential medicolegal purposes. The examiner should determine if the fall was
witnessed, and if so, speak directly to the witness and the patient. It is essential to document clearly what the witness and patient state has occurred, particularly regarding the mechanism of the fall, including whether the head was struck and if an alteration of consciousness occurred. Further questioning of the patient should focus on any specific complaints and should include a thorough neurologic and cardiopulmonary review of systems to help rule out a fall due to syncope or other acute medical condition. The physical examination should include evaluation for gross deformity, head laceration or hematoma, hip logroll, heel strike, and spinal step-off, as well as a complete neurologic examination, for comparison to baseline. Further attention should be paid to any area or system that has been potentially injured once hip, spine, and neurologic stability are verified.

Based on the clinical findings, imaging may be necessary. The physician should consider obtaining an emergent CT scan of the head without contrast if a change in mental status, head trauma, or an unwitnessed fall in a patient with a craniectomy has occurred. If CT scan is not indicated, it should be clearly documented that no head trauma or change in mental status has occurred. Plain radiographs of other injured body parts should be considered based on clinical suspicion of injury. Frequent neurologic checks, as often as every 1–2 hours, should be performed on any patient who hit his or her head and in whom a CT scan was performed that was negative for acute injury, or not done at all.

The focus should then be directed at preventing future falls through reinforcement of existing safety measures (eg, encouraging patients to ask for assistance with getting out of bed), as well as consideration of additional means. For example, less-aggressive physical barriers, including elevated side rails, a low bed with floor mats, and a bed-enclosure, might be appropriate, depending on the patient’s condition. Changing the room environment (eg, by minimizing the noise level or bright lights) should also be considered. If a patient continues to be at a high risk for falling or has actually repeatedly fallen, chemical or physical restraints may be necessary to prevent harm. Options include low-dose quetiapine (12.5–25 mg) or risperidone (0.5 mg) and, lastly, a Posey vest while in bed and a lap belt when in a chair for safety reasons. The occurrence of major patient events, such as a fall, should always be communicated directly to the patient’s primary contact person in a timely fashion.

ACCIDENTAL GASTROSTOMY (PEG) TUBE DISLODGEMENT

General Considerations

The accidental dislodgement of percutaneous endoscopic gastrostomy (PEG) tubes is estimated to occur in 1.6–20% of patients with the device. The paramount management consideration is the time from PEG placement. If a PEG tube is dislodged less than 2–4 weeks after the procedure, it should not be reinserted blindly as the gastrocutaneous stomal tract is not mature. Attempting to reinsert the tube can lead to an increased risk of separation from the abdominal wall, intraperitoneal tube insertion, and possible peritonitis from the leakage of gastric contents or tube feedings into the peritoneum. Adequate healing may require 4 weeks or longer in patients with factors that impair wound healing, such as malnourishment, steroid or other immunosuppressant therapy, and diabetes mellitus, which are not uncommon in the rehabilitation population. An additional factor is the traumatic injury often caused by a traction removal of the PEG, which increases the risk of tract disruption.

Treatment

If the PEG tube is dislodged soon after insertion (ie, within 2–4 weeks), an urgent general surgery or gastroenterology consultation should be requested if available; otherwise the patient should be transferred to the emergency department or ICU as acute care may be warranted. These services may be able to facilitate image-guided tube reinsertion, although open surgical intervention or intravenous antibiotic therapy may also be necessary.

If more than 2–4 weeks have elapsed from insertion of the PEG tube to its dislodgement, reinsertion should be attempted as soon possible after the dislodgement. This will help to avoid the need for a more invasive procedure, as mature stomas may close within minutes to hours. If the PEG tube is intact, the balloon should be deflated and a gentle attempt made to reinsert the tube into a
mature tract that has been lubricated with lidocaine jelly. The tube should never be forced, as a false tract or separation of the stomach from the external stoma can occur. If the initial tube is not intact or easy to reinsert, a Foley catheter (16–20 French) can be used. Once the tube is placed in the stomach, the balloon is inflated with saline and gentle traction is used to draw the balloon to the stomach wall. An urgent supine abdominal radiograph should be obtained after injecting 20–30 mL of water-soluble contrast solution (diatrizoate meglumine diatrizoate sodium [Gastrografin]) into the tube to confirm placement and rule out extravasation. Feedings should never be restarted or medications given until proper placement is confirmed. If the replacement tube is internally leaking, an emergent surgery consultation should be requested, if available.


ACCIDENTAL TRACHEOSTOMY TUBE DECANNULATION

General Considerations

The management of accidentally dislodged tracheostomy tubes is similar to that
of PEG tubes, in that emergent nature and management vary according to the time frame after placement. Inadvertent decannulation has the potential to become life threatening, especially if it occurs before the tract between the skin and the trachea has matured.

**Treatment**

If decannulation occurs less than 7 days after insertion, the tube should not be replaced blindly, as the stomal tract is not mature and a false passage into the mediastinum and subsequent respiratory arrest can occur. In this early time frame, the patient should be immediately ventilated using a bag–valve–mask and 100% fraction of inspired oxygen (FiO₂), while pulse oximetry is continuously monitored. If available, emergent anesthesiology (and possibly ear–nose–throat or general surgery) consultation should be requested for possible reinsertion using fiberoptic guidance or orotracheal intubation. The patient should be transferred immediately to an emergency department if the preceding services are not available.

In the period between 1 week and 1 month after initial insertion, the risk of tube misplacement progressively decreases. During this time frame, the tracheostomy tube may be reinserted at the bedside using a careful and delicate technique; however, consideration should still be given to obtaining an urgent consultation for fiberoptic evaluation of placement. Once the stomal tract has matured, safe reinsertion may be accomplished without image guidance; however, oxygenation should still be closely monitored. When replacing the tube, the inner cannula is first removed, then the obturator is inserted and the cuff is deflated. Next, the tracheostomy tube is lubricated and an attempt is made to reinsert it. If minor difficulty is encountered, a smaller sized tube can be tried. If adequate ventilation cannot being provided or the patient is otherwise in distress, he or she should be transferred immediately to emergent services.


General Considerations

Chest pain, pressure, or discomfort is a symptom often encountered on rehabilitation units and can occur as a result of cardiovascular, pulmonary, gastrointestinal, musculoskeletal, or psychological disorders. Life-threatening causes of chest symptoms, such as acute coronary syndrome (ACS), pericarditis, aortic dissection, pulmonary embolism (PE), pneumonia, and esophageal rupture, must first be ruled out before considering other causes. To make matters more complicated, certain patients with ACS, especially the elderly, women, and diabetics, may not present with chest complaints but with atypical symptoms such as left shoulder or back pain, dyspnea, and nausea.

Rehabilitation patients, particularly those who have experienced a stroke or amputation as a result of peripheral vascular disease, are at a higher risk of having ACS as they often have coronary artery disease as well. Recent surgery, especially orthopedic procedures, cancer, trauma, immobilization, presence of deep vein thrombosis, and hypercoagulable states, also predispose them to developing PE. They are also at high risk of pneumonia, which may manifest with chest pain, although often there are associated pulmonary symptoms. Possible causes of chest pain are included in Table 36–1.

Table 36–1 Possible causes of chest pain.
Clinical Findings

A. Symptoms and Signs
Classic symptoms of ACS are crushing, substernal chest pain with radiation to the left upper extremity, dyspnea, and diaphoresis. Stable angina is usually worse with physical exertion and relieved with rest, whereas unstable angina continues regardless. Patients with PE classically present with pleuritic chest pain, dyspnea, and tachycardia. Aortic dissections are characterized by tearing back pain or crushing chest pain. Pneumothorax manifests with sudden chest pain and dyspnea and is mostly present in patients with chest trauma, chronic obstructive pulmonary disease, or a history of prior pneumothorax but can also be a complication of certain procedures, such as placement of lines and needle electromyography. Pericarditis is usually pleuritic in nature and typically relieved by leaning forward. Congestive heart failure (CHF) and bronchospasm
can be confused with ACS as patients with either may also present with chest discomfort. Gastrointestinal complaints can often be correlated with meals. Neural causes may follow a dermatomal pattern around the chest. Anxiety is a common cause of chest pain, but is a diagnosis of exclusion, and the practitioner must clarify whether the patient became anxious before or after the onset of chest pain. If anxiety is suspected, relaxation techniques and deep breathing should be encouraged while the patient is worked up for other causes.

Vital signs should be checked and a physical examination focusing on the cardiovascular and pulmonary systems performed on every patient who complains of chest pain. In patients with an aortic dissection, differences in blood pressure measurements between extremities may be noted as a result of perfusion abnormalities. A tension pneumothorax causes hypotension, jugular venous distention, and rapid clinical deterioration requiring emergent needle decompression. Careful auscultation of heart sounds may help with diagnosis. An S\textsubscript{3} or S\textsubscript{4} heart sound on auscultation may be a sign of CHF or PE. Pneumonia often manifests with evidence of consolidation in the lungs, such as decreased breath sounds, rales, egophony, or abnormal fremitus. A friction rub is often heard with auscultation in pericarditis. Musculoskeletal-based chest pain is often reproducible with palpation or range of motion.

**B. Diagnostic Tests**

1. **Electrocardiography**—If possible, an ECG should be obtained in every patient with a complaint of chest pain. It should be compared with prior or baseline ECGs, when available, to ensure that any findings noted are new.

2. **Laboratory tests**—Typically three sets of cardiac enzymes (troponin and creatine kinase–MB isoenzyme) are obtained in patients at risk for cardiac ischemia. Although cardiac enzyme tests can be initiated in the rehabilitation unit, if there is a high suspicion of a cardiac event, the patient should probably be transferred to a higher level of care for cardiac monitoring and further evaluation, especially if he or she is hemodynamically unstable. Other laboratory studies to consider obtaining are a complete blood count, electrolytes, liver function tests, coagulation studies, and levels of brain natriuretic peptide, lipase, and amylase.

3. **Imaging studies**—Chest radiographs (posterior–anterior and lateral views) should be obtained whenever possible in all patients with complaints of chest pain and compared with prior or baseline images, if available. Chest radiographs
can be used to evaluate for aortic dissection (mediastinal widening), CHF, pneumothorax, and pneumonia. A CT scan of the chest with contrast is indicated if a PE or aortic dissection is suspected. If the patient has renal disease or a dye allergy, a ventilation/perfusion (V/Q) scan can be used to assess for PE, along with chest magnetic resonance imaging (MRI). An abdominal CT or ultrasound study can be performed to assess for intraabdominal pathologies.

**Treatment**

Supplemental oxygen should be administered and intravenous access established as soon as possible in patients with chest pain. If ACS is suspected, 325 mg of aspirin should be administered immediately along with nitrates for chest pain while simultaneously preparing the patient for transfer to an acute medical care unit. Patients with possible aortic dissection should be transferred to an acute care unit for monitoring, intravenous blood pressure control, and potential emergent surgical intervention. If available, consultation with cardiothoracic surgery should be arranged as early as possible. Patients with a PE should be started on anticoagulation therapy unless contraindicated once the diagnosis is established.

Exacerbation of CHF is usually managed with diuretics and restriction of fluid and sodium. Management of other pulmonary causes of chest pain is described in the next section. Fluid intake, output, and weight should be monitored closely. Intravenous diuretics may be necessary if oral diuretics are not effective.
Patients in the rehabilitation unit are at high risk of developing life-threatening pulmonary complications as a result of prior intubations and prolonged hospitalizations. Their respiratory muscles can also become debilitated secondary to general debility or neurologic impairment. They also have an increased chance of experiencing a thromboembolic event or aspiration.

Assessment of a patient with dyspnea or hypoxia is similar to that of the patient with chest pain as life-threatening cardiovascular, pulmonary, and neurologic causes must be ruled out first. Airway obstruction can be caused by tracheal stenosis, anaphylaxis, bronchospasms, and mucus plugs. Restricted lung expansion can be a consequence of decreased lung compliance, morbid obesity, chest wall deformities, injuries, or scoliosis, resulting in a decreased vital capacity. It has been reported that for every 10 degrees of thoracic scoliosis, there is a nearly 4% reduction in vital capacity. However, the cause of chest pain in rehabilitation patients is often mixed because of their medically complex conditions. The differential diagnosis of dyspnea and hypoxia is included in Table 36–2.

Table 36–2 Differential diagnosis of dyspnea and hypoxia.
<table>
<thead>
<tr>
<th>Emergencies</th>
<th>Pulmonary embolus, acute myocardial infarction, foreign body, nosocomial and aspiration pneumonia, acute pneumothorax, cardiac tamponades, anaphylaxis, tracheal stenosis or edema, stroke, Guillain-Barré syndrome, botulism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary</td>
<td>Acute bronchospasm secondary to asthma or chronic obstructive pulmonary disease, restrictive lung disease, pleural effusion, allergy, sleep apnea, chemical pneumonitis, pulmonary hypertension, bronchitis, emphysema</td>
</tr>
<tr>
<td>Cardiac</td>
<td>Acute coronary syndrome, congestive heart failure, arrhythmias, valvular disorders</td>
</tr>
<tr>
<td>Hematologic/Oncologic</td>
<td>Anemia, malignancy, radiation- or chemotherapy-induced lung fibrosis</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Spinal cord disease or injury, amyotrophic lateral sclerosis, phrenic neuropathy</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Scoliosis, chest wall deformities, rib fractures, muscular dystrophies, glycogen storage diseases</td>
</tr>
<tr>
<td>Rheumatologic</td>
<td>Sarcoidosis, vasculitis, and other connective tissue disorders</td>
</tr>
<tr>
<td>Medications</td>
<td>Opioid- or benzodiazepine-induced respiratory depression</td>
</tr>
<tr>
<td>Psychological</td>
<td>Anxiety attack</td>
</tr>
</tbody>
</table>

**Clinical Findings**

**A. Symptoms and Signs**
The classic symptoms of PE, described earlier, are pleuritic chest pain, dyspnea,
and tachycardia; however, the neurologically compromised patient with PE may be asymptomatic. Hypoxia and tachycardia may be noted incidentally in therapy or during routine assessment of vital signs and patients later found to have a large PE on a subsequent imaging study. Therefore, the physician must always have a high index of suspicion for a PE in a rehabilitation patient. Pneumonia usually manifests with a productive cough, fever, and chills. With aspiration pneumonia, choking on food or vomiting may be witnessed; however, many cases are silent and caused by reflux. Dyspnea in patients with Guillain-Barré syndrome is usually accompanied by neurologic manifestations. Patients having an exacerbation of CHF may complain of orthopnea, wheezing, and leg swelling. Opioid- or benzodiazepine-induced respiratory depression is usually preceded by lethargy or somnolence.

Decreased breath sounds or rales, or both, on auscultation may represent a consolidation or edema in the lungs secondary to pneumonia or CHF exacerbation. Wheezing is typically associated with acute bronchospasm and CHF exacerbation. The classic presentation of cardiac tamponade, known as Beck’s triad, consists of hypotension, muffled heart sounds, and jugular venous distention. Neurologic examination of patients with Guillain-Barré syndrome typically reveals decreased reflexes and weakness in the lower more so than the upper limbs.

B. Diagnostic Tests

1. **Laboratory tests**—If ACS is strongly suspected the patient’s cardiac enzymes should be checked. Arterial blood gas analysis can be helpful if the patient is not improving rapidly with supplemental oxygen. As a general guideline, a partial pressure of arterial oxygen (Pao₂) less than 60 mm Hg, partial pressure of carbon dioxide (Pco₂) greater than 45 mm Hg, and pH less than 7.3 should warrant intubation. A complete blood count with differential should be checked to assess for an infectious etiology or anemia. Sputum and blood cultures are useful in patients with suspected pneumonia. Chemical pneumonitis resulting from aspiration of sterile gastric contents may also be confused with pneumonia but the sputum culture should be relatively clean. Lumbar puncture is indicated if Guillain-Barré syndrome is suspected.

2. **Imaging studies**—Chest radiographs (posterior–anterior and lateral views, if possible) are almost always warranted in the evaluation of acute dyspnea or hypoxia. For patients with pneumonia diagnosed radiographically, a followup radiograph is recommended within at least 6 weeks to document resolution and
to rule out underlying neoplasm. Spiral CT scan of the chest or a V/Q scan should be obtained in all patients suspected of having a PE.

**Treatment**

All patients who complain of dyspnea should be given supplemental oxygen even if their oxygen saturation levels are within normal limits. Oxygen should be delivered initially at high flow rates (ie, 6 L/min via nasal cannula, 40% FiO₂) and titrated down as necessary. If the patient continues to be hypoxic despite this treatment, a 100% nonrebreather face mask (100% FiO₂) may be more effective. In patients with respiratory weakness or hypercapnia, biphasic positive airway pressure should be used instead to provide assistance with ventilation. Patients with signs of impending respiratory failure (eg, altered mental status, respiratory muscle fatigue, severe hypoxia) require intubation. Intravenous fluids should be administered if hypotension is present. Any possible reasons for airway obstruction should be addressed using suctioning or bronchodilators, or both. Suctioning or the use of an exsufflator–insufflator device (eg, Coughlator) may be helpful in removing mucus plugs in patients with tracheostomy tubes, although more severe obstructions may require bronchoscopy. Bronchodilators administered via nebulizer should be used if the patient’s bronchospasms result from exacerbation of asthma or chronic obstructive pulmonary disease. Anaphylaxis can be treated with intramuscular epinephrine. Patients who are receiving β blockers may not respond to epinephrine and thus require intravenous glucagon.

In patients with CHF exacerbation and pneumonia, oxygen diffusion across the alveoli is reduced because of pulmonary edema or consolidations, or both. CHF exacerbations should be treated with intravenous or oral diuretics while monitoring for renal and electrolyte disturbances. Although wheezing may be present in CHF patients, they usually respond poorly to bronchodilator therapy. Current recommended treatment for hospital-acquired pneumonia is vancomycin plus an antipseudomonal penicillin or third-generation cephalosporin or fluoroquinolone or carbapenem. If aspiration pneumonia is suspected, a third-generation cephalosporin or fluoroquinolone and clindamycin or metronidazole should be initiated. Patients suspected of having new onset or recurrence of Guillain-Barré syndrome should be transferred off the rehabilitation unit for respiratory support and treatment with intravenous immunoglobulin or plasmapheresis. Opioids or benzodiazepines act on the respiratory control centers in the medulla, causing depression. Naloxone is a potent reversal agent
FEVER

General Considerations

Fever is a very nonspecific sign commonly detected when collecting vital signs and is defined as an elevation in body temperature above the normal range 36.5–37.5°C (98–100°F). It can be caused by the release of pyrogens such as prostaglandin or the lipopolysaccharides in the cell wall of some bacteria, which then act on the hypothalamus to increase the temperature set point (analogous to turning up a thermostat). This generates a systemic response (ie, vasoconstriction or shivering) back to the rest of the body that then causes temperature to rise until it reaches this set point. The term hyperthermia is commonly used interchangeably with fever but is different in that it is an elevation in body temperature above the hypothalamic set point. Therefore, the body will respond to hyperthermia differently by vasodilation and sweating, but this can also occur when the fever resolves and the set point is lowered. Fever is generally associated with an inflammatory process in the body and in theory is thought to be a protective mechanism, aiding host defense and accelerating healing. Processes other than infection can cause fever, hence the differential diagnosis is vast (Table 36–3).

Table 36–3 Differential diagnosis of disturbance in body temperature
<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Immediate life threatening</td>
<td>Sepsis, bacteremia, meningitis, encephalitis, endocarditis, pneumonia pulmonary embolus, gas gangrene, necrotizing fasciitis, neuroleptic malignant syndrome, serotonin syndrome, toxic epidermal necrolysis</td>
</tr>
<tr>
<td>Infectious</td>
<td>Hardware infection, surgical site or line infection, osteomyelitis, cellulitis, urinary tract infection, cystitis, pyelonephritis, septic arthritis, abscess, gastroenteritis, colitis, wet gangrene, viruses (Epstein-Barr, cytomegalovirus, HIV, influenza, etc)</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>Atelectasis, pneumonitis</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Inflammatory bowel disease, cholecystitis, pancreatitis, appendicitis</td>
</tr>
<tr>
<td>Rheumatologic</td>
<td>Sarcoidosis, crystalline arthropathy, vasculitis, connective tissue disorders</td>
</tr>
<tr>
<td>Endocrine</td>
<td>Thyroid storm, acute adrenal insufficiency</td>
</tr>
<tr>
<td>Hematologic/Oncologic</td>
<td>Thrombus, cancer (especially leukemia and lymphoma), transfusion reaction</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Central dysautonomia, poikilothermia, intracranial hemorrhage</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Hematoma, heterotopic ossification, rhabdomyolysis, gangrene</td>
</tr>
<tr>
<td>Dermatologic</td>
<td>Wound healing, pressure ulcer</td>
</tr>
<tr>
<td>Iatrogenic</td>
<td>Surgery, drug-induced reactions (eg, antiepileptics, sulfonamides, antibiotics, stimulants), exercise, heat stroke, dehydration</td>
</tr>
</tbody>
</table>

Life-threatening infections, PE, and drug reactions must be ruled out before assessing for other causes. Noninfectious causes of fever, such as dysautonomia (central fevers), poikilothermia, heterotopic ossification, deep vein thrombosis,
and drug-induced reactions must also be considered in rehabilitation patients with central nervous system involvement and medically complex courses. In addition, elevations in body temperature resulting from exercise and menstrual cycle fluctuations can be mistaken for fever in rehabilitation patients (see Table 36–3).

Clinical Findings

A. Symptoms and Signs

Although the patient has an elevated body temperature, he or she typically complains of feeling cold or “having the chills” secondary to vasoconstriction in the extremities. As a first step, the medication list should be reviewed with attention to drugs that may cause fever (acetaminophen and antiinflammatory medications) or increase risk of infection (immunomodulating drugs). Other symptoms and signs that may help focus the evaluation should be assessed. All intravenous lines, catheters, surgical drains, tubes, and wounds should be identified and sites inspected carefully for signs of possible infection.

Prosthetic joint, hardware, and surgical site infections are a common concern encountered on the rehabilitation unit and in long-term care facilities. The signs and symptoms may include wound drainage that persists several days after surgery or begins in a wound that had been dry, implant site erythema, induration, increased joint pain unrelated to activity, effusion, and edema. Patients with a cranial or spinal implant or cranial surgery infection may present with signs and symptoms of meningitis or encephalitis, such as neck or head pain and change in mental status. A normal erythrocyte sedimentation rate and C-reactive protein level greatly reduce the likelihood of an early prosthetic joint infection. A plain radiograph may show lucency at the bone–cement interface, periosteal reaction, or motion of the components on stress views. Establishing the diagnosis of prosthetic joint infection requires a joint aspiration and analysis of the joint fluid for cell count, Gram stain, and culture. Antibiotic therapy should be withheld until the sample has been obtained, unless the patient is septic or otherwise too unstable. If there is suspicion of a prosthetic joint infection based on a clinical and noninvasive workup, the patients’ surgeon should be contacted immediately.

Several life-threatening drug reactions can occur with the use of pharmacologic agents that act on the central nervous system, such as antiepileptics, neuroleptics, and serotonergic agents. Toxic epidermal necrolysis
and Stevens-Johnson syndrome are life-threatening dermatologic conditions that are usually induced by a reaction to medications such as anticonvulsants and antibiotics, leading to confluent epidermal necrosis.

Serotonin syndrome produces signs and symptoms of hyperthermia, blood pressure instability, diaphoresis, change in mental status, hyperactive muscle stretch reflexes, and myoclonus when overdosage or interactions between drugs lead to excess serotonin activity. Neuroleptic malignant syndrome can be confused with serotonin syndrome, but it is an adverse reaction to antipsychotic medications leading to decreased dopaminergic activity that typically consists of fever, muscle rigidity, tremors, elevated blood pressure, and changes in mental status.

**B. Diagnostic Tests**

1. **Laboratory tests**—A complete blood count with differential, basic metabolic panel, and hepatic function panel should be obtained for all rehabilitation patients with new-onset fevers. The C-reactive protein level or erythrocyte sedimentation rate can be helpful, if negative, to rule out an infectious or inflammatory process. If an infectious etiology is suspected, a urinalysis or two sets of blood cultures, or both, should be collected especially during the peak of fever.

2. **Imaging studies**—Chest radiographs are commonly ordered in the fever workup to rule out pneumonia and atelectasis. A plain radiograph of a joint arthroplasty should be obtained. An MRI or bone scan can be used to diagnose osteomyelitis.

**Treatment**

Treatment of the fever itself should be comfort based as it will very rarely rise high enough (41°C or 107.6°F) to cause damage to the brain. The first steps can include loosening of clothing and use of fans before resorting to antipyretics. Infectious causes should be treated with appropriate antibiotics according to infectious disease guidelines. When infectious causes have been ruled out and drug-induced fevers are in the differential, a trial cessation of the suspected medication can be performed to see if fevers cease.

Management of all adverse drug reactions starts with cessation of the suspected agents and cooling blankets and ice packs to treat the hyperthermia. Serotonin antagonists such as cyproheptadine may be administered along with
benzodiazepines to help manage agitation and myoclonus. Pharmacologic management of neuroleptic malignant syndrome has not been well established; however, bromocriptine may have some benefit. Dantrolene and benzodiazepines have also been used as needed to reduce muscle rigidity and control agitation. Patients with these drug reactions, as well as Stevens-Johnson syndrome and toxic epidermal necrolysis, may require circulatory and ventilator support.

Fevers are commonly detected after surgery, especially when hardware has been implanted into the body, as in a total joint arthroplasty. Most are a result of the normal postoperative course, hemarthrosis, crystalline-induced arthropathy, dislocation or metallic debris–induced synovitis, or osteolysis. Principles of treatment are similar to those for other surgical-site infections, particularly intracranial, spinal, and abdominal.


**CHANGE IN MENTAL STATUS**

### General Considerations

A change in mental status is a divergence from a clearly defined baseline that includes disturbances in one or more of the following: cognition, perception, mood, and personality. It is especially important to determine the patient’s baseline status as he or she may already have deficits in these areas due to preexisting central nervous system lesions or dementia. Cognition includes attention, language, memory, visual–spatial perception, and executive
functioning. Changes in perception may manifest as auditory or visual hallucinations or as a delusion. Mood changes include not only depression, anxiety, and anger but also mania and euphoria. Agitation is a primary example of a change in personality; however, one must also be alert for other changes such as increased passivity or obsession. Often, these changes are first detected by family members, nurses, or therapists. Hence, any concern brought to the clinician’s attention should be carefully assessed as it may be an initial presenting sign of a more serious underlying condition. However, mental status changes are a nonspecific finding for which the differential diagnosis is broad, ranging from central nervous system lesions, infections, and cardiopulmonary disease to fecal impaction or sleep deprivation (Table 36–4).

**Table 36–4** Differential diagnosis of change in mental status.
Clinical Findings

A. Symptoms and Signs

The timing of the incident is crucial to see if there is a correlation to medications, sleep deprivation, or underlying dementia. Patients with dementia typically have increased confusion and restlessness in the evening or as the sun
is setting (ie, “sundowning”), likely due to disruption in their circadian rhythm.

B. Diagnostic Tests

1. Laboratory tests—A fingerstick blood glucose level should be obtained immediately to rule out hyperglycemia or hypoglycemia. Among the screening laboratory tests that might be ordered are a complete blood count; basic metabolic panel; hepatic function panel; ammonium level; thyroid profile; calcium, magnesium, phosphorus, vitamin B$_{12}$, and folate levels; and a urine drug screen, if warranted. The physician should maintain a high degree of suspicion for infectious causes as patients may not initially present with a fever, especially with urinary tract infections. Hence, a urinalysis is recommended in almost all rehabilitation patients. Further infectious workup may include sputum, blood, or stool cultures; HIV tests; and syphilis studies.

2. Imaging studies—If new focal neurologic deficits are detected or are questionable on examination, a CT or MRI scan of the head to rule out new or worsening lesions is indicated. Chest radiographs are indicated if pneumonia is suspected or hypoxia is also present. Abdominal imaging can help assess for fecal impaction or bowel obstruction.

3. Other studies—An ECG should be performed if possible to check for myocardial ischemia. An overnight pulse oximetry test may be useful to assess for sleep-disordered breathing.

Treatment

The safety of others as well as the patient should first be ensured before and while the diagnostic workup is underway. If the patient’s behavior is potentially harmful to self or others, then all potentially injurious objects should be removed from the vicinity, the patient should be secluded with one-to-one supervision, and his or her attention diverted if possible. Intramuscular benzodiazepines (eg, lorazepam, 1–2 mg) or neuroleptics (eg, haloperidol, 2–5 mg) should be administered if the patient threatens or resorts to violence or assault. Physical restraints to the extremities should be used as a last resort because of the risk of self-injury.

A thorough medication review should be performed; this includes looking into medications that patients may have received before admission to the unit, as toxic metabolites may have accumulated in those with impaired clearance.
mechanisms. Medication dosage errors and polypharmacy can be minimized by simplifying dosing regimens and discontinuing unnecessary medications. Standing doses of nonnarcotic medications can be used to control pain, thus avoiding the need for opioids. Identifiable causes should be treated based on results of clinical evaluation and diagnostic studies. Mental status should be monitored and documented serially to assess for treatment efficacy.

If the patient continues to have episodes of agitation, it may be beneficial to initiate a trial of low-dose benzodiazepines every 6–8 hours or low-dose bedtime neuroleptics to prevent escalation to aggressive behavior. There is no Food and Drug Administration–approved medication for the treatment and prevention of agitation, and all pharmacologic measures are off label. Therefore, regular assessment of medication efficacy is required, and agents should be subsequently discontinued if no benefit is noted.

UNCONTROLLED HYPERTENSION

General Considerations

Hypertension is a common comorbidity in rehabilitation patients and predisposes them to complications of vascular disease such as heart disease and cerebrovascular accidents. Approximately 95% of these patients have essential hypertension for which there is often no single identifiable cause. Medications (eg, cyclooxygenase-2 inhibitors, cyclosporine, tacrolimus, oral contraceptives, anticholinergics, and sympathomimetics) and drugs of abuse (eg, alcohol and stimulants) can cause hypertension.

Hypertensive urgency—an episode of systolic blood pressure greater than 180 mm Hg or diastolic blood pressure greater than 120 mm Hg with minimal or no target-organ damage—can be treated with oral agents. Hypertensive emergency, in contrast, requires immediate reduction in blood pressure using intravenous agents due to acute or progressive end-organ damage. The primary organs targeted in hypertensive emergencies are the brain, heart, large blood vessels, kidney, and eye. Nonadherence to medication regimens, especially clonidine, can result in rebound hypertension. However, in rehabilitation patients, uncontrolled hypertension may signify a more severe underlying problem, such as increased intracranial pressure due to worsening brain injury, hemorrhagic conversion of stroke, or a new stroke. Other causes to consider are elevations resulting from acute pain or stress, new medications, drug withdrawal,
physical exertion, and autonomic instability.

Clinical Findings

A. Symptoms and Signs

The patient with a hypertensive emergency may complain of blurry vision, chest pain, and shortness of breath or headache. Other symptoms include dizziness, change in mental status, focal neurologic deficit, chest pain, and palpitations. An $S_3$ heart sound auscultated during cardiac examination may be a sign of a myocardial infarction or CHF. An abdominal aortic aneurysm may be identified by a pulsatile mass on palpation and a bruit on auscultation. A neurologic examination with a cognitive component should also be conducted to assess for transient ischemic attack or stroke. Hypertensive emergency may also result in renovascular injury leading to oliguria or hematuria.

Patients with increased intracranial pressure due to brain mass or hemorrhage will also present with extreme elevations in blood pressure. The classic presentation is referred to as Cushing’s triad and includes widening of pulse pressure, bradycardia, and irregular respirations. This triad is an ominous sign as the irregular respirations are caused by pressure on the brainstem as it is pushed down into the foramen magnum, which may ultimately herniate. Initially there is tachycardia as the body increases sympathetic response in an attempt to raise blood pressure to overcome intracranial pressure and provide cerebral perfusion; however, the extreme hypertension triggers a parasympathetic response in the carotid baroreceptors via the vagus nerve, resulting in bradycardia. Hypertensive encephalopathy is different in that the rising blood pressure initially causes vasoconstriction until the mean arterial pressure reaches about 180 mm Hg; then breakthrough cerebral vasodilation occurs, leading to hyperperfusion followed by cerebral edema and mental status changes.

B. Diagnostic Tests

1. Laboratory tests—The blood urea nitrogen and creatinine levels should be checked in all patients to rule out renal failure. A urinalysis, including sediment analysis, should also be checked for hematuria and proteinuria.

2. Imaging studies—A chest radiograph is useful in assessing for CHF, cardiomegaly, and aortic dissection. A chest CT scan with contrast or chest MRI can also be obtained to diagnose an aortic dissection. A low threshold should be
used in evaluating a head CT scan to rule out intracranial bleeding in these patients, especially when focal neurologic deficits or changes in mental status are present. An MRI scan to rule out new or worsening strokes may also be warranted.

3. **Other studies**—ECG should be performed in all patients to check for myocardial ischemia, left ventricular hypertrophy, and conduction abnormalities.

## Treatment

The management of uncontrolled hypertension in patients on an inpatient rehabilitation unit differs from that provided to patients in an acute care setting. This reflects the lack of available telemetry monitoring and subsequent limited use of intravenous medications. Additionally, cerebrovascular accident is a very common rehabilitation diagnosis and raises unique considerations for blood pressure management in the acute and subacute periods. Before taking action in any patient with an elevated blood pressure reading, the pressure should be manually rechecked and the patient’s recent blood pressure trends should be evaluated. Additionally, one must assess for signs and symptoms of a hypertensive emergency, as described earlier.

The patient who is having a hypertensive emergency is placed on cardiac monitoring immediately. Intravenous access is established and treatment with intravenous antihypertensive agents, such as nitroglycerin, hydralazine, or labetalol, is initiated. The mean arterial pressure should not be lowered more than 25% in the first 24 hours to prevent cerebral ischemia. Hypertensive episodes in patients who have not sustained end-organ damage can be treated using oral agents, such as hydralazine and clonidine or topical nitroglycerine.

If the patient has recently had a stroke, one must be extremely careful to avoid aggressively lowering the blood pressure, as this may further compromise cerebral perfusion and neurologic function. The guidelines for treatment with permissive hypertension in the first 24 hours after stroke and chronic management with a normotensive goal are well defined; however, definitive data are lacking to help guide the management of elevated blood pressure in the days to weeks following a stroke.

A patient with a resting systolic blood pressure of 180 mm Hg measured more than 24 hours after a stroke should be considered for prompt treatment; however, cutoffs vary based on individual medical problems and are beyond the scope of this chapter. A reasonable goal would be to lower blood pressure
HYPOTENSION

General Considerations

Hypotension is primarily defined as systolic blood pressure less than 90 mm Hg or a drop in blood pressure greater than 40 mm Hg from the baseline. However, in rehabilitation, any patient with a drop in blood pressure that is symptomatic should be worked up and treated to allow for maximum participation in therapies as physical activity itself will cause increased blood flow to muscles and may lower blood pressure even more. More severe underlying causes, such as septic shock, anaphylaxis, myocardial ischemia, cardiac tamponade, and adrenal insufficiency, should be ruled out first. More common causes are hypovolemia, medications, and orthostatic hypotension secondary to autonomic dysfunction. Patients with myocardial infarctions, particularly in the right ventricular or inferior wall, may present with hypotension secondary to cardiac pump failure and have a poor prognosis. Those with adrenal insufficiency that is more likely of secondary than primary etiology may also present with hypotension. Secondary adrenal insufficiency is usually caused by long-term steroid use that results in atrophy of the adrenal glands, and it may also be potentiated by prolonged stress on the body due to lengthy hospitalizations, especially in
intensive care units. An infectious etiology, especially bacteremia leading to sepsis, can cause blood pressure instabilities.

Hypovolemia is common in patients who have burns and other causes of elevated metabolism (eg, traumatic brain injury, wound healing, infections), are on modified liquid diets (ie, with poor intake), or have any of the following problems: diarrhea or emesis, end-stage renal failure after dialysis, or anemia or active bleeding. Orthostatic hypotension occurs when blood vessels are unable to vasoconstrict as an individual stands abruptly, thus causing a postural drop in blood pressure. It can occur as a result of hypovolemia, central nervous system dysfunction (eg, hypothalamus and multiple system atrophy, previously called Shy-Drager syndrome), carotid baroreceptor dysfunction (eg, cervical spine surgery, carotid endarterectomy), autonomic neuropathy (eg, diabetes mellitus), inability to vasoconstrict peripheral vasculature effectively (eg, spinal cord injury, calcified vessels in the elderly), or increased heart contractility (eg, β-blocker administration, dilated cardiomyopathy). Medications such as opiates, benzodiazepines, antispasticity agents, dopaminergic compounds, tricyclic antidepressants, peripherally acting α blockers, and histamine blockers may cause hypotension as a side effect. Excess administration of antihypertensives is also commonly implicated, especially calcium channel blockers and angiotensin-converting enzyme inhibitors, which have typically stronger antihypertensive effects than β blockers and diuretics.

Clinical Findings

A. Symptoms and Signs
Orthostasis can be assessed by measuring blood pressure in the supine, sitting, and standing positions. Orthostasis is defined as a decrease in systolic blood pressure of more than 20 mm Hg, a decrease in diastolic blood pressure of more than 10 mm Hg, or an increase in heart rate of more than 20 beats per minute when transitioning from supine to sitting or sitting to standing position, usually within 2 minutes. Sometimes the patient may complain of lightheadedness from the test, if positive. The pulse is likely to be unreliable in patients taking β blockers because of a blunted autonomic response. The physical examination should focus on neurologic status (including cognition), cardiovascular system (assessing for cardiac hypertrophy and carotid bruits, and checking pulses), and extremities (evaluating for edema versus poor skin turgor).
B. Diagnostic Tests

Usually an ECG is sufficient to assess cardiac function; if it shows evidence of ischemia or if the patient reports chest pain, however, cardiac enzymes should be checked or an echocardiogram ordered. Blood urea nitrogen and creatinine levels should be checked to assess renal function while keeping a close eye on urine output. A complete blood count with differential and blood cultures is helpful in ruling out sepsis or anemia.

Treatment

If the patient is sitting when a hypotensive event occurs, have the patient lie flat on the bed or in the therapy gym to increase perfusion to the brain, then administer supplemental oxygen. If there is hemodynamic instability, suspicion of end-organ damage, or hypotension that cannot be reversed rapidly, intravenous lines should be inserted, fluids begun, and the patient transferred to an intensive care unit for cardiac monitoring and acute interventions. Intravenous empiric antibiotics should be initiated after cultures have been drawn. In cases of suspected β-blocker overdosage, 3.5–5 mg of glucagon can be administered once by means of intravenous push and repeated 10 minutes afterward if there is no response. Adrenal insufficiency can be treated with oral prednisone, hydrocortisone, or fludrocortisone supplementation.

In patients who have sufficient urine output (> 0.5 mL/kg per hour) and no end-organ damage, fluids can be pushed orally or using PEG. Caution is needed in treating patients with end-stage renal disease or dilated cardiomyopathy as they may, in fact, be in a state of fluid overload and may derive more benefit from hemodialysis or diuresis. If a patient is indeed volume depleted, then small fluid boluses (250–500 mL) may be appropriate to prevent fluid overload. If the hypotension is a result of anemia, the patient should be transfused with packed red blood cells while being worked up for etiology.

ARRHYTHMIA

General Considerations

Tachycardias can range from a benign response (e.g., to stress or exertion) to deadly arrhythmias requiring immediate intervention. They can be generally
divided into narrow-complex supraventricular tachycardias (SVTs) and wide-complex tachycardias (WCTs) based on the duration of the QRS complex. Sinus tachycardia is the most commonly encountered SVT. It is usually not pathologic but rather a response to exertion, stress, illness, pain, and prolonged bed rest. Atrial fibrillation is the most frequently encountered pathologic SVT, followed by atrial flutter. Ventricular tachycardia (VT) is a WCT that can become life threatening if sustained or if it converts to ventricular fibrillation.

Sinus bradycardia can be a normal physiologic finding in physically fit individuals or can be caused by medications, severe hypoxia, sepsis, increased intracranial pressure, myxedema, or hypothermia. Other causes of bradycardia include sick sinus syndrome and atrioventricular (AV) block.

Clinical Findings

A. Symptoms and Signs

Patient complaints of chest pain, hypotension, and altered mental status are signs of instability and warrant urgent action (eg, electrical cardioversion or transcutaneous pacing). All patients presenting with arrhythmia should have vital signs, pulse oximetry, blood glucose, and a 12-lead ECG checked. Hypoxia, hypothermia, hypoglycemia, and hypovolemia can contribute to arrhythmias. A focused physical examination should also be performed once the patient is stable to assess for underlying pulmonary, structural heart, or thyroid disease.

B. Diagnostic Tests

A 12-lead ECG is required to differentiate between arrhythmias and evaluate for underlying acute myocardial ischemia, infarction, and other conduction abnormalities. SVTs and WCTs can be differentiated by the length of the QRS complex (≤ 120 ms in SVTs; ≥ 120 ms in WCTs). SVTs can result from atrial or AV junction dysfunction; Table 36–5 summarizes their associated ECG findings.

Table 36–5 Supraventricular tachycardias and their associated electrocardiographic findings.
In patients with bradycardia, AV block can be differentiated from sinus bradycardia by prolongation of the PR interval (> 200 ms). In first-degree AV block all impulses are conducted; therefore, each P wave is followed by a QRS complex. In second-degree AV block, not all impulses are conducted and this abnormality can be further divided into Mobitz type I and II classifications. Mobitz I AV block is characterized by a progressively increasing PR interval.
until the impulse is not conducted. In Mobitz II AV block, a consistent PR interval is present that improves with vagal maneuvers. In third-degree AV block, there is no conduction of impulses, resulting in complete dissociation of P wave from QRS waves.

Once the arrhythmia has been identified, possible contributing causes or factors are identified. Acidosis, low serum potassium, and low serum magnesium levels can increase excitability of the myocardium and predispose the heart to developing tachycardia. A complete blood count with differential can help rule out an underlying anemia or infectious etiology. Other laboratory studies that are commonly performed include a thyroid function panel, toxicology screen, cardiac enzymes, and digoxin level. Digoxin can initially affect the baseline ECG, causing downward scooping of the ST segment and inverted T waves, but this does not necessarily signify toxicity. A chest radiograph or CT scan, V/Q scan, or CT angiography can help rule out underlying pulmonary disease. A CT scan of the head can assess for a new or worsening disease process. A cardiac echocardiogram, stress test, or cardiac catheterization can be performed at a later date to assess for structural heart disease or myocardial ischemia.

### Treatment

Most patients with sudden-onset tachycardia and stable pulses should initially be provided with supplemental oxygen and placed on cardiac monitoring for observation and treatment. If the patient is unstable but conscious, sedation should be provided followed by synchronized cardioversion. Those with stable SVTs and a regular rhythm should initially be treated with vagal maneuvers, such as the Valsalva maneuver, carotid massage, bearing down, and immersion of the face in ice water. If vagal maneuvers fail, 6 mg of adenosine can be administered, but only under monitoring and with a defibrillator on hand. Adenosine is contraindicated for patients with bronchospastic lung disease. Irregular SVTs such as atrial fibrillation or flutter with a heart rate of 150 beats/min or higher, should be rate controlled using verapamil, diltiazem, or β blockers. The use of calcium channel blockers can increase mortality in patients with VT and should be avoided.

Bradycardia and signs of poor perfusion (altered mental status, ongoing chest pain, and hypotension) necessitate cardiac monitoring, supplemental oxygen, and intravenous access; patients should be prepared for possible transcutaneous pacing. Patients with symptomatic sinus bradycardia and Mobitz type I AV block
can be treated initially with atropine, 0.5 mg intravenously; up to 3 mg total may be given. Those with Mobitz type II and third-degree AV block should be placed on transcutaneous pacing immediately. A Mobitz type II block can progress to a third-degree block without warning; hence pacing is warranted even in asymptomatic patients.

Digoxin, β-blocker, and calcium channel blocker toxicity are pharmacologic causes of arrhythmias for which specific treatments are available. Digoxin toxicity warrants immediate infusion of digoxin- and digitoxin-specific antibodies (DigiFab), along with treatment of the resultant arrhythmia. In cases of severe sinus bradycardia resulting from an overdosage of a β blocker or calcium channel blocker, 3.5–5 mg of glucagon can be administered once by intravenous push and repeated 10 minutes afterward if there is no response.


CHANGES IN VISION

General Considerations

The prevalence of prior traumas, myopia, cerebrovascular disease, and diabetes mellitus puts rehabilitation patients at high risk of developing visual dysfunction. Acute transient vision loss is a sudden deficit in monocular or binocular vision lasting less than 24 hours. It can be caused by temporary vascular insufficiency to the eye or visual cortex (amaurosis fugax or hypotension with vertebrobasilar artery insufficiency), compression of the optic nerve (papilledema), or neuronal
depression after a seizure or migraine.

Acute persistent visual loss lasts at least 24 hours and can be further divided into painless and painful etiologies. Causes of painless persistent visual loss include central retinal vein and artery occlusion, temporal arteritis, vitreous hemorrhage, maculopathy, ischemic optic neuropathy, lens dislocation, and retinal detachment. Intracranial masses and hemorrhages that compress the optic chiasm, optic radiations, and occipital lobes can cause stereotypical visual field abnormalities (anopias) or cortical blindness; however, these tend to be more gradual in onset. Causes of painful persistent visual loss are acute angle-closure glaucoma, optic neuritis, corneal abrasion or ulcer, uveitis, endophthalmitis, and keratitis.

Clinical Findings

Unfortunately most rehabilitation settings have neither the equipment nor the expertise to conduct a comprehensive ophthalmic examination. Therefore, the history and physical examination must be used in determining possible causes, and whether the patient requires an emergent or urgent referral to an ophthalmologist.

A distinction should be made between sudden-onset visual loss and a preexisting visual deficit that has just been discovered. Because of compensation by the contralateral eye, patients sometimes do not realize they have a chronic disease process in the other eye. Bilateral visual changes suggest a pathologic process at the optic chiasm, a generalized process (eg, hyperglycemia, papilledema resulting from increased cranial pressure), or an injury caused by bilateral exposure. The presence of pain suggests an inflammatory cause such as keratitis, uveitis, endophthalmitis, and optic neuritis or acute glaucoma. Pain in acute glaucoma typically is characterized as a deep eyebrow pain, with associated nausea and vomiting; patients also report seeing colored halos around lights as a result of corneal edema. Temporal arteritis manifests with headache on the same side as the visual loss. The patient history can also elicit whether the visual loss affects peripheral more than central vision. Peripheral monocular vision loss suggests retinal detachment prior to macular detachment, ischemic optic neuropathy, or retinal vessel occlusion. Retinal detachment usually presents with painless, unilateral photopsia (perceived flashes of light), an increasing number of floaters, and decreased visual acuity.

The physical examination should include appraisal of general appearance and testing of visual acuity (with glasses and one eye at a time), extraocular
movement, visual fields, symmetry of pupils, and reactivity to light. Peripheral vision and central vision can be further assessed by performing visual field testing or having the patient read letters or identify objects on paper while focusing on the center of the page. Erythema, photosensitivity, and tearing may be noted in patients with uveitis and endophthalmitis. Defects in visual fields and extraocular movements suggest intracranial pathologies affecting the visual pathways or causing cranial nerve palsy. Further workup may include fluorescein application, intraocular pressure testing, slit-lamp examination, or an ophthalmoscopic examination.

### Treatment

Immediate treatment of central retinal artery occlusion and angle-closure glaucoma is directed at lowering intraocular pressure. To effect this, 500 mg of oral acetazolamide and one drop of 0.5% timolol maleate or 2% pilocarpine can be administered as soon as possible. Giant cell arteritis with ocular involvement and optic neuritis can be treated with high-dose intravenous steroids.

Psychological disorders are highly prevalent among patients admitted to acute rehabilitation units. Soon after their medical event, patients are forced to quickly wrestle with myriad emotions, such as powerlessness, shock, demoralization, and loss, while often concomitantly experiencing a variety of physical discomforts. Many patients must also suddenly grapple with the reality of alterations in lifestyle and relationships, cope with stigma that may accompany their limitations or injury, and bear significant financial burdens. Particularly during the acute phase, physical and cognitive deficits, as well as the inability to perform basic activities of daily living, serve as a significant source of distress and irritation. Unanswered questions related to prognosis and quality-of-life issues fuel feelings of worry and unrest about the future, for both patients and caregivers.

Although many patients adjust well to their disability, others develop significant psychological distress, in a manner that goes beyond expected reactions to loss. Estimates of the prevalence of mood disorders among rehabilitation patients vary, ranging from 20% to a striking 64%. However, these disorders are common among a host of diagnostic categories, including spinal cord injury (SCI), burns, stroke, amputation, brain neoplasms, and traumatic brain injury (TBI). Identification and treatment of psychological disorders in patients with injuries such as SCI is particularly important, as psychological and
biopsychosocial factors are often better predictors of adjustment or quality of life than biomedical variables, such as severity of injury. Emotional disturbances following acquired brain injury may be the result of a structural brain lesion or psychological reaction to the trauma, or some combination thereof. A psychologist skilled in assessment can be invaluable in determining the relative contributions of neurocognitive and emotional factors to a patient’s presentation. Discrimination of these factors can advance treatment strategies and facilitate successful postacute transition and care.

**COMMON PSYCHOLOGICAL REACTIONS & DISORDERS AMONG REHABILITATION PATIENTS**

Psychological reactions or disorders commonly occurring in rehabilitation patients are summarized below.

### Adjustment Disorders

Many patients admitted to rehabilitation units are confronted with medical events that may result in physical disfigurement, pain, psychological distress, cognitive dysfunction, and reduced functional independence; these events can be abrupt, unexpected, and have implications for change in life roles or plans (e.g., inability to fulfill domestic responsibilities or return to work). Successful adjustment to a new disability requires the patient to acknowledge the functional consequences of the injury and incorporate these changes into his or her personal identity. Significant emotional reactions can occur that can lead to poor adjustment or more severe psychological disorders, such as depression or anxiety. According to the *Diagnostic and Statistical Manual, Fourth Edition Text Revision (DSM-IV-TR)*, an adjustment disorder is present if the patient’s reaction to an identified stressor results in marked and excessive distress, or the reaction to this stressor causes significant impairment in functioning. An inability to adapt to injury can have an impact on quality of life, self-esteem, life satisfaction, or self-concept, and lead to increased subsequent hospitalization rates, longer hospital stays, and poorer functional outcomes.

### Depression

Depression is perhaps the most common neuropsychiatric manifestation in the
rehabilitation setting. In addition to the typical features associated with depressed mood (e.g., dysthymia, melancholia, anhedonia, decreased energy, perturbed sleep and appetite), depression can also result in diminished attention, memory, motor skills, processing speed, executive functions, initiation, and motivation, all of which serve as potential barriers for rehabilitation progress. Among rehabilitation patients, depression has also been linked to excess disability, slow physical recovery, poor quality of life, and increased mortality. A history of psychiatric illness, dysphasia, functional limitations, and social isolation are risk factors for depression in the rehabilitation setting.

The prevalence of depression varies somewhat for different diagnostic categories of rehabilitation patients, with some clinical populations being at very high risk. According to most studies, depression affects 30–50% of stroke patients within the first year and has been found to be associated with poorer functional scores up to 6 months following acute rehabilitation. Depression is estimated to occur in 11–40% of SCI patients and 33% of patients with lower limb amputations. Depression may be the most common psychological sequela following TBI, and depression in TBI has been associated with greater functional disability, poorer recovery and quality of life, and greater health care costs. A variety of somatic and cognitive indicators included in the DSM-IV-TR diagnostic criteria for depression overlap with symptoms found in nondepressed stroke, dementia, and TBI patients (e.g., apathy, decreased motivation, fatigue, poor concentration); therefore, a thorough neurocognitive examination is recommended to evaluate the nature of these symptoms in such patients.

Anxiety among patients in rehabilitation settings is well documented in the literature. Reactions to disabling events are often marked by significant worry, tension, and feelings of loss of control. Patients diagnosed with inoperable cancers, as well as survivors of stroke, TBI, SCI, and burns, are sometimes confronted with the inevitability of death for the first time, resulting in increased rates of anxiety disorders, including generalized anxiety or post-traumatic stress disorder (PTSD). Rates of anxiety for various diagnostic groups can run very high; for example, some studies estimate up to 30% of stroke patients, 27% of TBI patients, 35% of burn patients, and up to 17% of SCI patients experience significant anxiety. Risk factors for anxiety disorders such as PTSD following a disabling event include preinjury psychiatric history, mal-adaptive coping strategies, perceived lack of social support, and high emotional distress.
Guilt

Guilt is a painful emotion commonly experienced in rehabilitation settings and may result in heightened levels of depression. Guilt occurs when one imposes self-blame for his or her injury or disability. Stroke survivors, for example, may blame themselves for not immediately calling 911, abruptly discontinuing their hypertensive medications, or not taking adequate care of their health. Survivors of traumatic incidents that resulted in TBI or SCI may ruminate over choices or behaviors that led up to their injury, such as, “I could have prevented the accident by driving more slowly,” or “If only I did not go out that night.” Similarly, patients who have undergone limb amputations as a result of poorly controlled diabetes or other medical conditions may experience guilt over their disability.

Denial of Illness

Denial of illness is a psychological coping mechanism that may emerge following a threat to one’s identity and self-preservation, and is not uncommon among rehabilitation patients. Individuals utilizing this defense mechanism may deny the existence of the illness or minimize its nature, severity, and implications. In different phases of illness, denial may have an adaptive benefit, in that it serves to psychologically protect the patient, preserving optimism and hope, thereby allowing the individual to cope in the midst of a crisis. However, prolonged or excessive denial may be maladaptive, impeding rehabilitation activities and precluding the patient from adopting a more problem-focused orientation. Studies suggest that TBI patients evidencing denial are more likely to refuse rehabilitation therapies, are perceived as more difficult to work with, and are less likely to ask for help when needed. Denial has been linked to delays in seeking treatment and to negative treatment outcomes and survival.

A subtype of denial, anosognosia, is neurologically based and can serve as a significant barrier to rehabilitation. An individual with this neurologically acquired denial of illness may refuse to engage in therapies out of the belief that he or she is unimpaired. These patients may also attempt to leave the hospital or engage in unsupervised activities and thus pose a serious safety risk.

Preexisting Psychiatric & Personality Disorders
Personality disorders comprise a constellation of longstanding, persistent, maladaptive traits that are characteristic of the way an individual experiences and interacts with his or her environment. Several of the cluster B personality disorders described in the DSM-IV-TR involve impulsivity, unstable mood, suicidal gestures, and risky behavior as core criteria. Personality-disordered patients with limited judgment and impulse control at baseline are prone to higher rates of injury as a result of suicide attempts, assaults, and dangerous, sensation-seeking behaviors, and thus may constitute a higher percentage of rehabilitation patients. Individuals with personality disorders may pose a unique challenge in the rehabilitation setting by refusing to participate in therapies, smuggling illegal substances into the facility, or verbally abusing staff. Patients with personality disorders unfortunately show a tendency toward poor discharge outcome.

Caregiver Distress

Acquired injury affects the family system in varying ways and degrees, as changes in family functioning and roles are almost inevitable. Emotional support, personal hygiene, ambulation, and feeding often become the primary responsibility of loved ones. The implications of acquired injury, including changes in the spouses’ perceived romantic relationship, sexual life, and changing role from partner to caregiver, may become more apparent in the weeks and months following the life-altering medical event. Owing to the demands involved in tending to their loved ones, caregivers are prone to experience a range of long-term difficulties, including depression, anxiety, decreased life satisfaction, and deterioration of health and social life. Identifying and treating emotional dys-function in caregivers can contribute to improved outcomes for patients following discharge from postacute rehabilitation settings.


PSYCHOLOGICAL ASSESSMENT TECHNIQUES

Psychological evaluations are frequently requested in rehabilitation settings to identify emotional dysfunction. As noted earlier, psychological disorders have been associated with poorer functional recovery and social outcomes, reduced quality of life, and increased frequency of cognitive impairment and mortality. Psychological distress can include feelings of helplessness, inertia, demoralization, and lack of motivation, which may reduce therapeutic compliance and, as a result, rehabilitation treatment efficacy. Thus, early identification of patients with emotional dysfunction is essential, as it can have a positive effect on recovery and long-term outcome.

Clinical Interview

Assessment of an individual’s psychological functioning requires a clinical interview with the patient and other informants, if possible. Information is gathered regarding the patient’s developmental, educational, and vocational history; social and medical history; prior psychiatric or psychological treatment; behavioral health issues (eg, substance abuse); and existing coping skills (eg, support network). For elderly patients, it is also important to inquire about preinjury functioning and to look for signs suggestive of premorbid cognitive decline or dementing conditions, which can complicate rehabilitation and disposition outcome. In these instances, neuropsychological assessment can be helpful in distinguishing dementia symptoms from other etiologies (eg, TBI,
Questionnaires & Inventories

Psychological assessment of emotional functioning is often supplemented by questionnaires and inventories. Assessment tools used in the screening, diagnosis, and assessment of psychiatric disorders can be classified into two basic types: self-report measures and observer-rating scales. Repeatable self-report measures for depression and anxiety are the Beck Depression Inventory—Second Edition (BDI-II) and the State Trait Anxiety Inventory (STAI), respectively. Brief questionnaires with a “yes–no” format, such as the Geriatric Depression Scale (GDS), are preferable for elderly patients with cognitive limitations. Assessment of coping strategies can yield important information regarding how an individual may adapt to a new injury or disability; instruments such as the Ways of Coping—Revised (WOC-R) or the Acceptance and Action Questionnaire (AAQ) can identify maladaptive coping mechanisms that can interfere with treatment and recovery. Lengthy inventories assessing personality and emotional functioning, such as the Personality Assessment Inventory (PAI) or Minnesota Multiphasic Personality Inventory, Second Edition (MMPI-2) are occasionally used with disabled patients who are largely cognitively intact.

Although the use of self-report measures to identify the presence and level of emotional distress has several advantages, it is not without limitations. Fatigue, pain, cognitive limitations, medications, and environmental noise can affect patients’ ability to participate in testing and the validity of the results. Moreover, some questionnaires contain several test items related to health, stamina, and physical functioning, which can be wrongly attributed to depression. Self-report measures also assume that patients have awareness into the nature and magnitude of their illness, which is not always the case.

NEUROPSYCHOLOGICAL ASSESSMENT

The inclusion of neuropsychologists within a multidisciplinary rehabilitation team provides a valuable complement to other disciplines, as they are uniquely trained in the quantification of cognitive impairment following neurologic insult. Cognitive disability is present in a large proportion of rehabilitation patients, including individuals with TBI, stroke, and dementia. Acute and chronic health conditions can also have a negative impact on cognition (eg, cardiovascular disease, obesity, diabetes, cancer). Neuropsychological assessment offers an
objective, valid, and reliable method for detecting and tracking brain impairment. It is often the only means of detecting impairment in higher cortical abilities and provides valuable information above and beyond neuroimaging techniques. Advances in structural and functional neuroimaging have provided a window into regions affected by brain injury but cannot quantify the magnitude of cognitive impairment nor predict the degree of functional disability experienced by the individual. Neuropsychological assessment identifies cognitive and behavioral barriers to patient care and predicts functional capacities postdischarge, such as the ability to live independently, resume driving, or return to work. Neuropsychological assessment can facilitate successful treatment strategies, assist with maintaining patient safety and compliance, and provide critical information related to discharge planning and outcomes.

Neuropsychological Tests

Neuropsychological tests have strong psychometric properties that allow them to be used to assist with the diagnosis of brain pathology and to inform treatment. Evidence for brain dysfunction is determined by referencing a comparison standard. Specifically, neuropsychological tests allow for quantification of deficit by utilizing collected test data from reference groups. These data serve as a benchmark against which patient performance is statistically compared with that of other groups to determine if the respective cognitive domain signifies an area of strength or deficit. Patient performance can be compared against a healthy normative sample from the population, other clinical groups, or the patient’s previous test performance.

The raw scores undergo statistical analyses to provide a standardized score and percentile, with the 50th percentile marking the population average. Qualitative descriptors (eg, “mildly to moderately impaired,” “low average,” “very superior”) are often used in written reports to facilitate communication with other disciplines. For many neuro-psychological tests, normative sample groups are available with specific demographically based corrections (eg, age, education, gender), so that the clinician may best match the patient to samples with similar characteristics, thereby providing the optimal interpretation of test data. When using the individual comparison method, neuropsychologists often estimate premorbid ability using demographic variables (education, occupational history) or screening measures typically resistant to the effects of brain dysfunction (eg, word recognition). This estimate provides a baseline by which
current test performance is compared. The neuropsychologist may often be called upon to track improvements over the course of admission and uses the initial testing for detection of change.

Clinical neuropsychologists rely on a multidimensional approach to assessment, integrating results of cognitive measures with behavioral observations, clinical interview, medical history and other background data, current emotional state, and biopsychosocial contributors. Cognitive measures span a multitude of functional domains, and the degree to which each domain is assessed can vary based on setting, relevance to referral question, patient limitations, or other barriers (eg, time limitations). Broadly, these cognitive domains include attention, processing speed, motor skills (strength, speed, dexterity), sensory acuity, working memory, learning and memory, language skills, visuospatial perception and constructional ability, executive functions (eg, abstract reasoning, problem solving, behavioral planning, judgment, task switching, inhibition), intellect, and academic aptitude. Assessment of these cognitive domains can be achieved using a wide variety of measures. Table 37–1 lists neuropsychological instruments that are commonly used in inpatient rehabilitation settings, grouped by cognitive domain. Because many of these measures capture more than one cognitive ability, they are listed according to the primary domain that each test was designed to assess. The cognitive domains most relevant to neuropsychological assessment within inpatient rehabilitation settings are described briefly here.

### Table 37–1

Common neuropsychological instruments used in rehabilitation settings.
<table>
<thead>
<tr>
<th><strong>Brief Screening Batteries</strong></th>
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<tbody>
<tr>
<td>Repeatable Battery for Assessment of Neuropsychological Status (RBANS)*</td>
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<tr>
<td>Brief Cognitive Neuropsychological Examination (BCNE)*</td>
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<tr>
<td>Neurobehavioral Cognitive Status Examination (Cognistat)*</td>
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<tr>
<td>Neuropsychological Assessment Battery (NAB), Screening Module*</td>
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<tr>
<th><strong>Arousal, Orientation, and Mental Status</strong></th>
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<tbody>
<tr>
<td>Galveston Orientation and Amnesia Test (GOAT)*</td>
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<tr>
<td>Confusion Assessment Method (CAM)*</td>
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<tr>
<td>Montreal Cognitive Assessment (MoCA)*</td>
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<tr>
<td>General Cognitive Screener from Wechsler Memory Scale—4th Edition (WMS-IV)*</td>
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<tr>
<th><strong>Attention and Concentration</strong></th>
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<tbody>
<tr>
<td>Digit Span from Wechsler Adult Intelligence Scale—4th Edition (WAIS-IV)*</td>
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<tr>
<td>Symbol Digit Modalities Test (SDMT)*</td>
</tr>
<tr>
<td>Mental Control from the Wechsler Memory Scale—3rd Edition (WMS-3)*</td>
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<tr>
<td>Subtests from NAB Attention Module*</td>
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<tr>
<td>Cancellation tasks, if suspected hemi-attention (eg, letter or line cancellation)</td>
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<tr>
<th><strong>Speech and Language</strong></th>
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<tbody>
<tr>
<td>Boston Naming Test*</td>
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<tr>
<td>Subtests from NAB Language Module*</td>
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<tr>
<td>Complex Ideational Material from Boston Diagnostic Aphasia Examination*</td>
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<tr>
<td>Token Test—Short Version*</td>
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<tr>
<th><strong>Visual Perception and Construction</strong></th>
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<tr>
<td>Judgment of Line Orientation (JLO)*</td>
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<tr>
<td>Clock Drawing Test*</td>
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<tr>
<td>Facial Recognition Test (FRT)*</td>
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<tr>
<td>Rey-Osterrieth Complex Figure Test (ROCF)*</td>
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<tr>
<th><strong>Somatosensory and Motor Abilities</strong></th>
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<tbody>
<tr>
<td>Finger Localization*</td>
</tr>
<tr>
<td>Right–Left Orientation*</td>
</tr>
<tr>
<td>Grooved Pegboard*</td>
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<tr>
<td>Finger Tapping Test (FTT)*</td>
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<tr>
<th><strong>Learning and Memory</strong></th>
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<tbody>
<tr>
<td>California Verbal Learning Test, 2nd Edition—Short Form (CVLT-II-SE)*</td>
</tr>
<tr>
<td>Hopkins Verbal Learning Test-Revised (HVLT-R)*</td>
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<tr>
<td>Subtests from the Wechsler Memory Scale IV (WMS-IV)*</td>
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<tr>
<td>Subtests from NAB Memory Module*</td>
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<tr>
<td>Brief Visual Memory Test—Revised (BVMAT-R)*</td>
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<tr>
<th><strong>Executive Functioning</strong></th>
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<tbody>
<tr>
<td>Fluency (semantic, phonemic, design)</td>
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<tr>
<td>Wisconsin Card Sorting Test—64 (WCST-64)*</td>
</tr>
<tr>
<td>Trail Making Test*</td>
</tr>
<tr>
<td>Subtests from the NAB Executive Functions Module*</td>
</tr>
<tr>
<td>Subtests from Delis–Kaplan Executive Function System (DKEFS)*</td>
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</tbody>
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A. Attention and Working Memory

At its most basic level, attention begins with arousal and alertness. Successively more complex levels of attention include sustained, selective, alternating, and divided attention. Working memory is often described as a facet of complex attention and involves the ability to store and mentally manipulate information on a temporary basis. Tests that assess working memory include reciting digits backward, calculating serial seven subtractions, and performing mental math. Attention underlies most cognitive skills and attentional deficits can compromise other domains, such as memory and executive functioning. Attentional impairments are the most common manifestation of brain damage and occur following insult to a variety of cortical and subcortical brain regions or disturbances in neurophysiologic systems or metabolic state. Impairment in this domain can compromise safety (eg, medication errors due to distraction, leaving appliances turned on) and skills contributing to functional independence (eg, driving).

B. Language

Typical language assessments performed by the neuropsychologist include measures that evaluate processing (“receptive”) and communicative (“expressive”) aspects of language ability. Observations of spontaneous speech and formal assessment of auditory comprehension, sentence repetition, confrontation naming, reading, and writing may be included in the evaluation. Aphasia typically occurs following injury to the left hemisphere, which is language dominant for most individuals (ie, 95% of right-handed individuals and 70% of left-handed people). The most common injury causing aphasia is a left middle cerebral artery stroke; other sources include cerebral hemorrhage, TBI, brain tumors, and dementias. Language deficits can preclude return to functional independence or compromise safety in a variety of ways. For example, a patient with an expressive aphasia may be unable to voice basic needs to others, call for help in times of emergency, or relay critical medical information to others. Receptive aphasias hinder a patient’s ability to comprehend spoken language, including medical or therapeutic instruction, and can prohibit the patient from participating in medical decision making and future planning.

C. Visual–Spatial Skills

This broad cognitive domain includes visual attention, visuoperceptual ability,
localization of objects, visual construction, visual organization, and visual synthesis. Posterior right hemisphere lesions following stroke, TBI, or brain tumors are the most common cause of visual–spatial deficits. Acquired deficits or syndromes include visual agnosia, visual neglect, topographic disorientation, and constructional dyspraxia. Deficits in this cognitive domain are common among patients in rehabilitation settings and can be quite responsive to therapeutic intervention. Unresolved visual–spatial impairment can imperil patient safety and may indicate the need for assistance or supervision for the patient at discharge. For instance, visual neglect can impede performance of household tasks (eg, cooking), financial or medication management due to reading errors, driving, or safe community navigation. Topographic disorientation can leave a patient prone to getting lost, even in familiar environments.

D. Memory

Memory functions include the ability to encode, store, and retrieve sensory information; in the context of a neuropsychological assessment, memory typically refers to declarative memory systems (ie, semantic and episodic memory). Effective assessment of this domain should include multiple learning trials with delayed free, cued, and recognition memory trials. Memory dysfunction can occur following insult to a variety of cortical and subcortical regions, as this ability relies on intact underlying processes, such as attention and sensory registration. Amnestic syndromes typically occur following damage to medial temporal lobe regions or diencephalic lesions. Frontal lobe damage can adversely affect encoding and retrieval memory processes. Deficits in memory have serious implications for return to functional independence and can affect safety, medication adherence, performance of instrumental activities of daily living (IADLs), and decision-making capacity.

E. Executive Functioning

Executive functioning is a heterogeneous domain of higher order cognitive abilities, which include strategy formation, organization, sequencing, task persistence, motivation, self-monitoring, inhibition of responses, mental flexibility, and abstract reasoning. This broad cognitive domain strongly interacts with working memory and attention, and executive deficits are often observed if impairment occurs in these areas. Executive functioning deficits can occur from damage to a variety of neural regions; generally speaking, they occur following lesions or insults to the prefrontal cortex, associated frontal–subcortical circuitry,
anterior cingulate cortex, or cerebellum. Given its heterogeneity, executive functioning is the most complex domain to evaluate and quantify. Functionally, executive dysfunction can affect a variety of IADLs, including driving and medication adherence, and can also compromise overall safety. It is considered to be the strongest predictor of limitations to functional independence and successful performance of IADLs.

Goals of the Neuropsychological Evaluation in Acute Rehabilitation Settings

Relative to the comprehensive nature of outpatient neuro-psychological evaluations, neuropsychological assessment in acute rehabilitation units is generally limited in scope. Patient time is at a premium and the psychologist must compete with other disciplines to complete his or her work. Quantification of a patient’s deficits is further compromised by factors such as fatigue, sedation, pain, medication effects, language impairment, sensory and motor changes, and psychological distress. Despite these limitations, the neuropsychologist practicing in an acute rehabilitation setting can generate a rich evaluation using relatively brief screening measures, providing vital data relevant to effective treatment and discharge planning for cognitively impaired patients. Specifically, the neuropsychological evaluation can provide data that can quantify the magnitude of cognitive impairment, assist with effective treatment strategies, clarify aspects of the patient’s psychosocial environment relevant to discharge planning, and generate predictions regarding level of functioning and care postdischarge (eg, return to work, driving ability, level of supervision), as well as long-term outcomes.

A. Information about Premorbid Conditions

In the course of the neuropsychological evaluation, the clinician can acquire information regarding premorbid experiences with learning, such as attained level of education, attitudes about school or other types of instruction, preexisting attention deficit disorder or learning disabilities, and knowledge about how skills are best acquired (eg, “hands-on” versus verbal instruction). This knowledge can help the psychologist anticipate how a patient will adjust to the rehabilitation setting and can also indicate the preferred method(s) of instruction. For example, individuals who frequently failed in school and struggled in lecture-based, verbal, or written instruction may find a didactic
approach to rehabilitation uncomfortable or threatening. For these patients, a more visually based instruction that emphasizes demonstration and practice may be most effective at sustaining their engagement and participation. In contrast, patients with a history of positive experiences with education who demonstrate continued pursuit of learning may be more amenable to verbal methods of instruction and willing to engage in written material and “homework” assignments. This information can help tailor interventions in a form that is most comfortable to the patient, facilitating participation in the rehabilitation process.

B. Identification of Cognitive Deficits

Cognitive deficits are common in patients admitted to acute rehabilitation units, and these deficits can significantly disrupt the rehabilitation process. For example, memory dysfunction can hamper patients’ ability to learn new skills or compensatory techniques due to poor carryover on a daily basis. Difficulty with sequencing or self-monitoring can limit the patient’s ability to follow complex or multistep instruction from rehabilitation professionals. Early identification of deficits through neuropsychological testing can facilitate individualized rehabilitation regimens that simplify or otherwise accommodate instruction for patients’ identified areas of weakness. Knowledge of a patient’s specific cognitive deficits can allow for greater integration of cognitive strategies within the standard therapeutic retraining regimen. In addition, patients’ cognitive status can change rapidly over the course of an admission, and relatively brief, serial assessments can document areas in need of continued intervention.

Cognitive impairment in rehabilitation patients can stem from a primary central nervous system condition (eg, stroke, TBI, brain tumor) or develop secondary to non–central nervous system disease (eg, pulmonary disorders, chronic kidney disease, cardiac disorders, hypertension, obstructive sleep apnea, or type 2 diabetes). Delirium secondary to metabolic disturbance or infection (eg, UTI) is also seen in rehabilitation settings. Moreover, it is not uncommon for patients to be admitted following a mechanical fall or other physical injury resulting from insidious cognitive decline not previously identified. Neuropsychological assessment is invaluable in these circumstances to quantify cognitive deficits that both inform functional limitations and indicate the necessary level of care required postdischarge.

C. Guidance for Postdischarge Care and Outcomes

The onset of neurocognitive deficits following a disabling event can have important implications for immediate and long-term outcomes in a variety of
disorders. Cognitive dysfunction in patients admitted to acute rehabilitation can hinder treatment, increase length of stay, and complicate discharge. Deficits in cognition may necessitate alterations in treatment planning and postacute care and should be identified as early as possible. A thorough review of the literature regarding the benefit of neuropsychological testing for outcome prediction is beyond the scope of this chapter. The summary below highlights some of the benefits of neuropsychological assessment in acute rehabilitation in the prediction of postacute outcomes such as return to functional independence, driving ability, and vocational functioning.

1. **Independent living**—In regard to everyday tasks that contribute to functional independence, information garnered from the neuropsychological evaluation can generate prediction of IADL performance, such as medication and financial management, functional communication skills, cooking, shopping, and other household and community activities. Basic tasks such as bathing and grooming are simple, over-learned, motor-based tasks that may be more associated with physical dysfunction than with cognitive impairment and are less frequently addressed in the neuropsychological report. Numerous reports have demonstrated an association between neuropsychological test performance and IADL dysfunction, including studies of older adults, post–acute head injury patients, heart transplantation patients, vascular dementia patients, individuals with HIV, and stroke patients. Executive dysfunction is often at the core of IADL failures and may be the most predictive of impairment for these everyday skills.

2. **Driving ability**—Findings from several studies indicate that neuropsychological test performance is predictive of driving ability, leading to the general conclusion that cognitively impaired drivers have a higher numbers of crashes and show poor performances on simulated driving measures. Rizzo and Kellison provide an excellent review of current research into the neuropsychology of driving ability (see reference below). Although tests of executive functioning appear to be the strongest predictors, multiple cognitive domains have been implicated in impaired driving, including measures of information-processing speed, complex attention, and working memory), and visuoperceptual skills. No single test is sufficiently reliable to determine driving fitness, and a broad assessment approach is indicated.

3. **Vocational functioning**—When a patient is discharged from an acute rehabilitation unit, the team often renders an opinion regarding the patient’s ability to return to work. Although neuropsychological tests cannot simulate
vocational skills, they can provide a valid estimate of the cognitive abilities that are applicable to a broad range of vocations, greatly informing vocational decision making. Much of the literature examining neuropsychological predictors of vocational outcome are derived from TBI samples. There have been numerous published reviews of research spanning several decades, which are nicely summarized by Sadek and van Gorp (see reference below). They conclude that neuropsychological test results are associated with employment status and vocational outcome. A range of cognitive domains have been identified as having predictive value for vocational outcome, including executive functioning, memory, and general intelligence. However, use of a global impairment score (ie, average of scores across all domains) may be more appropriate than using specific cognitive domains. When making specific predictions about the ability to return to work, neuropsychological test performance should be considered in the context of current and premorbid factors, such as work history and medical or psychological issues, as these can enhance predictive accuracy.


PSYCHOLOGICAL INTERVENTIONS IN ACUTE REHABILITATION

Because of the life-altering changes that accompany disability, psychological interventions for patients exhibiting emotional distress are critical to fostering healthy adjustment, boosting coping skills, and improving mood. A complete
return to the patient’s preinjury baseline following a life-altering medical event is atypical. Among stroke survivors, for example, the majority are unable to return to their previous jobs and approximately half are unable to engage in any type of gainful employment. For patients facing these uncertainties, psychological interventions can be helpful in preventing onset of depression or anxiety disorders, and improving overall adjustment and outcome.

**BARRIERS TO PSYCHOLOGICAL INTERVENTIONS**

Psychological interventions on rehabilitation units are adapted to overcome a variety of obstacles encountered in these settings. The psychologist may need to compete for patient time with clinicians from other disciplines, may have suboptimal session time because of patient fatigue or illness, or may encounter reluctance from the patient to engage owing to preexisting biases or misconceptions about psychotherapy. Patients may harbor suspicion or mistrust of the psychotherapeutic process or regard psychotherapy as reserved for the mentally ill; the presence of a psychologist may therefore have a stigmatizing effect. It is the authors’ experience that presenting psychological services as “routine” for all inpatient consumers may help minimize stigma and alleviate worry or mistrust. Inpatient contact with a psychotherapist can alleviate patient biases and encourage the individual to seek assistance postdischarge if problems arise. Interventions on inpatient units are necessarily brief and problem focused, when compared with traditional psychotherapy modalities offered in outpatient settings. A multimodal, biopsychosocial approach to assessment and treatment is indicated, given the complexity of issues confronted by patients in these settings.

**GOALS OF PSYCHOTHERAPY**

Psychotherapy involves an approach based on a particular theory or paradigm with the goal of relieving distress and maximizing functioning and quality of life. Studies have found that the magnitude of distress following a life-altering medical event is usually better accounted for by one’s coping resources, rather than the actual injury. For this reason, a major goal for rehabilitation psychologists is to boost patients’ coping resources and help them adapt and manage the new medical, physical, cognitive, and psychological challenges that may besiege them.

Using interview and assessment data, as well as input from the rehabilitation team, psychologists can help the patient and family foster hope and motivation,
develop realistic expectations, grieve over losses, overcome frustration, accommodate to ongoing limitations, and establish connections with outside sources necessary for aftercare, such as case managers, social workers, and vocational specialists. Common goals in rehabilitation psychology are summarized in Table 37–2, and several of these are described in more detail below.

### Table 37–2 Addressing psychological distress: common goals for individuals in rehabilitation.

<table>
<thead>
<tr>
<th>Goal</th>
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<tbody>
<tr>
<td>Accept grief, loss, and functional limitations</td>
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<tr>
<td>Normalize emotional reactions to trauma or illness</td>
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<tr>
<td>Instill hope</td>
</tr>
<tr>
<td>Overcome frustration</td>
</tr>
<tr>
<td>Maintain realistic goals and expectations</td>
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<tr>
<td>Accept help from others</td>
</tr>
<tr>
<td>Avoid toxic thoughts</td>
</tr>
<tr>
<td>Address changes in work and relationships</td>
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<tr>
<td>Reconstruct self-image</td>
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<tr>
<td>Redefine life purpose</td>
</tr>
<tr>
<td>Use social support system</td>
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<tr>
<td>Engaging in problem solving</td>
</tr>
<tr>
<td>Replace negative with positive self-talk</td>
</tr>
<tr>
<td>Allow time for relaxation and diversion</td>
</tr>
<tr>
<td>Use humor</td>
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<tr>
<td>Focus on spirituality</td>
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**Facilitation of Acceptance**

A major goal for rehabilitation psychologists is to facilitate patients’ acceptance of their disability. Patients are taught to acknowledge their feelings of grief, while also allowing space and time for reflection and contemplation. Information regarding the patient’s psychiatric history and coping methods can assist the rehabilitation team in predicting the patient’s ability to accept and adjust to his or her disability.
Instillation of Hope

In dealing with the effects of a life-altering medical event, it is essential that patients have hope. The experience of hope can help patients manage the demands of their disability and promote a sense of well-being. Patients can hope, for example, that they maintain a positive sense of self, preserve basic functional abilities, and continue to achieve specific goals. Patients can learn to maintain hope by repeating self-statements such as, “I am lucky to have a good family and treatment team.”

Use of Techniques for Overcoming Frustration

Learning to manage the sequelae of acquired injury or illness can be exceedingly frustrating for patients and caregivers. Rehabilitation patients frequently feel frustration when progress in therapies seems slow or they fail to achieve their goals. This response sometimes reflects patients’ or caregivers’ unrealistic expectations about the length of time necessary to complete a task. Rehabilitation patients are taught to make self-statements such as, “I need to take it one day at a time,” or “I am doing my best.” Psychologists also teach patients to break down goals into simple, realistic, achievable steps, because mastery of one skill helps bolster the confidence needed to tackle a new skill. Patients are also advised to recognize and reward themselves for their accomplishments, regardless of how small or insignificant. To decrease frustration, caregivers are also instructed to remind patients of any gains made.

Use of Relaxation & Distractions

Patients and family members can use meditation, soothing audiotapes, and guided imagery to induce relaxation and manage pain. Distractions, such as surfing the Internet, listening to music, or engaging in a hobby, are also helpful. In rehabilitation settings, patients capable of reading are encouraged to read the works of inspiring authors, which can help instill positive expectations. Many patients also find comfort in faith, prayer, and scripture, which can dampen postinjury depression.

Identification of Social Support
Of all the resources thought to help patients cope with stress, social support is perhaps the most commonly studied. Social support has been repeatedly linked with outcomes, including postinjury employment following SCI and TBI, mortality following cardiac rehabilitation, and post-stroke health status. Caregivers shield against stress by reducing unnecessary demands, managing the environment to increase comfort and control, planning pleasant activities, and recognizing and responding to early signs of frustration. Social isolation and limited social support are risk factors for poor outcomes; if identified, interventions should be targeted toward social reintegration utilizing the community’s available resources.

**PSYCHOLOGICAL INTERVENTION STRATEGIES**

Rehabilitation psychologists assist patients with psychological issues using a variety of intervention modalities. In inpatient settings, psychological interventions are usually direct and problem focused; sessions can take place daily or on an infrequent basis, depending on the patient’s psychological condition and needs. Treatment is influenced by patient factors (eg, psychological strengths and weaknesses, mental health history, capacity and willingness to engage in treatment) and institutional factors (eg, length of hospitalization, resource allocation, and demands on the patient’s time and stamina in a therapeutic day). Commonly used intervention strategies are psychoeducation, motivational interviewing, behavior modification, cognitive–behavioral therapy, and support groups.

**Psychoeducation**

Psychoeducation is the provision of didactic information related to a patient’s condition; it allows patients and care-givers to better understand the patient’s condition and cope with changes and lingering deficits. Psychoeducation promotes a sense of control by enhancing knowledge and facilitating informed treatment choices and, as a result, may also decrease psychological distress.

**Support Groups**

Support groups can be immensely helpful for rehabilitation patients and caregivers. They can facilitate peer support, enhance knowledge regarding the illness or injury and prognosis, provide a context for normalizing feelings of
frustration and disappointment, and reduce the sense of isolation that many rehabilitation patients experience following a disabling event.

Cognitive–Behavioral Therapy

Cognitive–behavioral therapy (CBT) is an evidence-based therapeutic technique that emphasizes an educational and skill-building approach, stressing collaboration between the therapist and patient in identifying and modifying factors that contribute to emotional disorders or problematic behaviors. Psychopathology is viewed as arising from distorted, incorrect, or maladaptive cognitions concerning the self, others, and the world. Treatment aims to modify the thoughts, feelings, and behaviors causing distress.

CBT is structured, yet flexible, and provides the psychologist with a plethora of specific, concrete tools with which to work. Where patients have suffered a life-altering medical event, CBT provides psychoeducation and teaches coping strategies, problem-solving skills, stress management, and cognitive restructuring to battle toxic, negative thoughts. Some common examples of negative thoughts targeted in treatment include catastrophizing (“I will never be able to walk again”), mislabeling (“That person had to have been staring at my prosthetic leg”), and polarized thinking (“Today has been a complete waste because I did not accomplish everything I intended”). CBT has substantial empirical support in the treatment of depression, anxiety, eating disorders, substance abuse, and chronic pain conditions. Given the frequent maladaptive thoughts, behaviors, and emotions among rehabilitation patients, CBT is quite applicable to individuals with chronic health conditions and acquired injury.

Motivational Interviewing

Motivational interviewing is an empirically based, therapeutic technique that involves a semidirective, client-centered counseling style for eliciting behavior change by helping clients to explore and resolve ambivalence. This technique is nonjudgmental, nonconfrontational, and nonadversarial; it recognizes that not all individuals are ready to change and uses inquiry and reflection to elicit talk of change. Motivational interviewing has evolved from its original use as a treatment for substance abuse to one having a wide variety of medical applications, including management of chronic health conditions such as diabetes, hypertension, heart failure, and pain.
Behavior modification is a treatment approach based on the principles of operant conditioning, in which undesirable behaviors are replaced with more desirable behaviors, often using positive or negative reinforcement. The chief goal is to increase or decrease the frequency of specific behaviors. Behavior modification approaches are highly structured and generally incorporate task analysis, goal setting, and continuous behavioral assessment and monitoring, adapting treatment strategies as necessary. Behavior modification has wide application in rehabilitation and has been used in treating problematic behaviors following acquired brain injury, reducing the impact of chronic pain, and promoting participation in therapies.


Communication is the process by which people exchange information. Language is the process by which thoughts, feelings, and emotions are generated using a recognizable verbal or written system. It includes both receptive and expressive skills. Speech is the process of creating sounds and combining them to form words that are understood by a listener. It requires the integration of cognitive and neurologic abilities along with the musculoskeletal skills that support respiration, phonation, and articulation. Cognition refers to mental processes, including attention, memory, auditory processing, problem solving, and executive function. Some of the cognitive skills that are important for communication include processing speed, concentration, self-monitoring, thought organization, and memory. Verbal communication, the most common form, involves the use of speech, language, and cognitive skills, augmented by facial expressions and gestures.

GOALS OF SPEECH, LANGUAGE, & SWALLOWING THERAPY IN ACUTE REHABILITATION

Speech, language, cognitive, voice, and swallowing problems are key issues for patient management in acute rehabilitation settings. Speech–language pathologists (SLPs) assess and treat impairments in each of these domains. During the initial stages of a patient’s diagnostic workup, a speech and swallow evaluation is warranted, especially if caregivers and family members report
changes in communication, cognitive, or swallowing ability. During the rehabilitation process, the SLP, along with other members of the multidisciplinary team, provides quality care to help the patient reach his or her fullest physical, psychological, social, vocational, and educational potential.

The main areas to be tested in patients undergoing evaluation include speech, language, voice, cognition, and swallowing. All parameters are assessed or at least screened using formal tests or informal measures. The primary objective is to determine the presence of impairment, classify the type, and determine the level of severity. Based on the results, recommendations and functional prognoses are made, a treatment plan is formulated, and measurable therapy goals are developed. The information from the initial assessment provides an important baseline for measuring change.

The goal of speech, language, and cognitive-based therapy is to improve communicative effectiveness, efficiency, and naturalness. Three main therapeutic approaches, outlined by Duffy, can be used to achieve these goals. The first is to restore lost function. This effort aims to reduce impairment and targets deficits directly. Achieving success with this approach depends on the etiology and course of the disease, as well as the type and severity of the disorder. The second approach is to promote compensatory abilities and the use of residual function. Compensation can take many forms; for example, a patient might be instructed in speech-enhancing strategies or the use of augmentative devices or alternative means of communication (gestures, functional communication board, etc). A third approach encompasses strategies to reduce a patient’s need for lost function by modifying the environment and facilitating more effective speaker–listener interactions. Through education and counseling, SLPs also help patients and their families adjust and cope.

The primary objective for swallow rehabilitation is to achieve the least restrictive dietary level while maintaining swallowing safety. Swallowing safety can be described as the maintenance of nutrition and hydration without medical complications. Specifically, SLPs aim to have a patient tolerate the least restrictive diet without aspiration. Dietary modifications focusing on liquid consistencies or food textures may be made. Dysphagia therapy entails teaching a patient compensatory swallowing strategies and targeting deficits directly through various exercises to change swallowing performance. Patient and family education is important to ensure compliance with diet modifications, aspiration precautions, and carryover of swallowing exercises and strategies.

When determining a treatment plan, all clinicians should consider patient, family, and caregiver needs and desires while simultaneously considering the
patient’s level of impairment and functional abilities. Treatment can focus on functional goals, which may be tailored to a patient’s specific needs (eg, a personalized functional communication board for a patient with expressive aphasia) or traditional goals, which target specific deficits (eg, word recall for the patient with expressive aphasia). Therapy can be direct (eg, drills, structured tasks) or indirect (eg, informal therapy tasks such as board games). In all cases, treatment within the inpatient rehabilitation setting should aim to make the patient as independent as possible. Therapeutic change achieved in the medical setting that is effectively carried over to the patient’s daily life is considered to be a successful treatment outcome.


**LANGUAGE**

The human nervous system, comprising the central nervous system (the brain and spinal cord) and the peripheral nervous system (the sensory and motor nerves), is integral to our ability to formulate and express language. The brain initiates, controls, and regulates sensorimotor and cognitive functions through the functions of its various parts: the cerebral cortex, cerebellum, brainstem, and subcortical structures. Language and speech disturbances can be indicators of structural and physiologic impairment in the brain. A review of basic brain physiology is therefore essential to understanding the language disorders that derive from disease and injury affecting these structures. With knowledge of the area of neurologic insult, the SLP can predict what type of impairment to expect upon initial evaluation. An understanding of the neurologic bases of communication also contributes to a more effectively structured and realistic rehabilitation program.

The cerebral cortex is divided into two hemispheres, each comprising four lobes that regulate complex functions (*Table 38–1*). The cerebellum, located inferior to the occipital lobes and posterior to the brainstem, is crucial for
maintaining balance in space and executing coordinated movements. A critical function of the cerebellum for speech and swallow is the integration of sensory input from other regions of the brain, allowing it to coordinate muscle groups. Through modifications of muscle tone, speed, and range of motion, the cerebellum allows movements to be executed smoothly and helps to make sequenced motor skills automatic. It does not initiate motor activity, but instead controls and performs online correction of planned movement.

Table 38–1 Key functions of the lobes of the brain.
<table>
<thead>
<tr>
<th>Lobe</th>
<th>Location</th>
<th>Function</th>
<th>Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frontal</td>
<td>Most anterior part of the brain</td>
<td>Language production, Planning and initiation, Judgment and reasoning, Concentration, Emotional range, Inhibition, Adaption to change, Insight</td>
<td>Poor pragmatics, Loss of flexible thinking, Perseverations, Poor focus or attention, Emotional lability, Inability to express language (Broca's aphasia), Difficulty in problem solving</td>
</tr>
<tr>
<td>Temporal</td>
<td>Laterally in cerebral hemispheres</td>
<td>Auditory sensation and perception, Auditory comprehension, Memory acquisition, Visual perception, Categorization of objects</td>
<td>Difficulty comprehending language (Wernicke's aphasia), Prosopagnosia, Disturbance with selective attention to what is seen and heard, Memory loss</td>
</tr>
<tr>
<td>Parietal</td>
<td>Between occipital lobe and central sulcus</td>
<td>Sensory integration—touch, pressure, pain, temperature, taste</td>
<td>Poor divided attention, Anomia, Alexia, Agraphia, Dyscalculia, Poor kinesthetic awareness, Unilateral neglect</td>
</tr>
<tr>
<td>Occipital</td>
<td>Most posterior part of the brain</td>
<td>Visual processing, Visual discrimination, Visual association</td>
<td>Visual field cuts, Difficulty locating objects in environment, Difficulty reading or writing</td>
</tr>
</tbody>
</table>

The brainstem, which consists of the medulla oblongata, the pons, and the midbrain, connects the brain to the spinal cord and regulates primary life functions such as respiration, swallowing, blood pressure, eye movements, and heart rate. It also mediates functions such as coughing, gagging, hearing, balance, and body temperature. The medulla oblongata is important for speech and motor control, as it contains nerve fibers that control phonation, articulation, velopharyngeal closure, and swallowing. It is also crucial for overall arousal, and important for sleep. The pons bridges the brainstem to the cerebellum, helps control breathing and sleep, and is also important for alertness and arousal. The
midbrain contains structures important for vision and hearing. The sub-cortical structures (thalamus, hypothalamus, hippocampus, amygdala, and basal ganglia) are located above the midbrain (Table 38–2); each of these structures plays a role in supporting speech, language, and swallowing.

**Table 38–2** Key functions of the subcortical structures of the brain.
In the peripheral nervous system, sensory (afferent) and motor (efferent) nerves are connected to the spinal cord (spinal nerves) and to the brainstem (cranial nerves). The spinal nerves extend to the organs, muscles, joints, blood vessels, and skin surface. The cranial nerves originate from the brainstem and innervate the muscles of the head, neck, face, larynx, tongue, pharynx, and glands. The cranial nerves are essential for speech, resonance, and phonation. In
addition, these nerves transmit signals that serve the special senses: vision, audition, smell, and taste. Their motor and sensory functions are summarized in Table 38–3.

**Table 38–3** Cranial nerves and their functions.
The two major systems of the peripheral nervous system are the autonomic nervous system and the somatic nervous system. The somatic nerve fibers mediate the skeletal muscle reflexes. Through its sympathetic and parasympathetic divisions, the autonomic nervous system regulates the activity of organs, such as the salivary glands, heart, lung, blood vessels, stomach,
intestines, kidneys, and bladder. Regulation and monitoring of these functions are essential to survival.

The brain’s functions are regulated by a set of general organizational principles. An understanding of these principles allows further appreciation of how impaired neurologic function can impede communication and swallowing (Table 38–4).

**Table 38–4** Functional organization of the brain.
<table>
<thead>
<tr>
<th>Principle</th>
<th>Organization</th>
<th>Implications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interconnectivity of the brain</td>
<td>All functions (sensory and motor) in the cerebral cortex are connected through association and commissural fibers, which allow for constant interaction within each hemisphere and between the two hemispheres.</td>
<td>Helps to explain how messages from multiple sources are rapidly integrated for an appropriate response to given stimuli.</td>
</tr>
<tr>
<td>Centrality of the central nervous system (CNS)</td>
<td>The CNS integrates all incoming and outgoing information in order to generate appropriate responses to the information received. The response can be volitive or reflexive.</td>
<td>Allows for the ability to analyze and synthesize multiple sources of information and to generate distinct responses.</td>
</tr>
<tr>
<td>Laterality of brain organization</td>
<td>Bilateral anatomic symmetry</td>
<td>The two cerebral hemispheres are essentially similar in structure. Left hemisphere has language dominance. Right hemisphere dominates emotions, musical skills, metaphors, humor, and stress. Left motor cortex controls movements in the right half of the body. Sensory information from the left half of the body projects to the right sensory cortex, and vice versa.</td>
</tr>
<tr>
<td>Unilateral functional differences—each hemisphere acquires an advantage over the other for different specialized functions</td>
<td>Contralateral sensorimotor control—all sensory and motor fibers decussate at the body's midline</td>
<td></td>
</tr>
<tr>
<td>Functionally specialized networking</td>
<td>Neuronal systems are functionally specialized; sensory and motor systems consist of various pathways that transmit differentiated information to different limbs.</td>
<td>A specialized pathway controls speech muscles in the face and neck through cranial nerves in the brainstem.</td>
</tr>
<tr>
<td>Topographic organization in cortical pathways</td>
<td>The spatial organization of neurons, tracts, and terminals reflects the spatial relationships of the body's surface and functionally related muscle groups</td>
<td>Topographic maps of functions helps physicians precisely locate lesions in the CNS.</td>
</tr>
<tr>
<td>Brain plasticity</td>
<td>The brain is able to recognize and modify tissue functions and adapt to internal and external changes. This permits regeneration of nerves and reorganization of cellular functions.</td>
<td>Can permit new learning and adaptability following a neurologic insult.</td>
</tr>
</tbody>
</table>
Familiarity with lesion localization and causes of neurologic impairment is required to conduct a thorough evaluation of speech and language abilities and interpret the results. Speech and language disorders commonly result from cerebrovascular accidents (stroke), brain tumors, or traumatic brain injuries.

**A. Signs and Symptoms of Impaired Speech and Language**

Initial examination can identify spared versus disrupted functions. Any abnormal sign in a patient presumably reflects a breakdown in neural circuitry and a specific pathway. Lesion localization can assist in providing differential diagnoses and can also provide a rationale for specific probing during an evaluation. For example, a patient with a left cortical lesion and right hemiplegia warrants testing for aphasia or cognitive communication disorders.

**B. Assessment of Speech and Language Function**

Initial assessment of receptive and expressive language abilities and differential diagnoses occurs in the acute care setting. Upon admittance to the acute rehabilitative setting, the patient is reevaluated to determine a level of functional communication, provide a prognosis, establish a baseline for measuring progress, and create and implement a treatment plan. Ongoing assessment of recovery and efficacy of treatment is provided throughout the rehabilitation process.

Formal diagnostic testing as well as informal observational findings are used to evaluate speech and language. Receptive and expressive communication skills are probed when assessing for aphasia, an impairment in language ability that may result from a head injury or stroke. Several areas of communication are evaluated (Table 38–5).

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**Table 38–5** Language areas assessed in a standard speech–language evaluation.
General Considerations

Cognitive impairment and language impairment are unique entities that influence one another. Although the person with aphasia has a primary disorder in the area of language, cognitive functions must also be evaluated as the ability to compensate for language deficits reflects cognitive functioning. Evaluation and treatment of cognitive deficits is discussed later in the chapter.
Clinical Findings

Features that confirm the presence of aphasia include difficulty with language formulation and comprehension. Virtually all patients with aphasia have some degree of anomia (ie, difficulty recalling names or words), and varying abilities in comprehension, repetition, conversation, reading, and writing. Specific types of aphasia can be determined through examination of a patient’s language presentation and lesion localization. Figure 38–1 provides a visual representation of a screening tool to classify the major aphasias.

![Classification of The Aphasias](image)

Figure 38–1 Classification of aphasias.

Aphasic speech often contains recognizable errors that can assist in differential diagnosis. Agrammatism occurs in individuals with nonfluent aphasia and is characterized by the absence of functor words (ie, words that perform a function), thereby creating sentences that include primarily nouns and verbs. The speech is telegraphic and there are omissions of grammatical morphemes (small units of grammar). Paragrammatism occurs in fluent aphasias and is characterized by inaccurate application of syntactic rules. Paraphasias are errors in word recall. Phonemic paraphasias are characterized by a single phoneme substitution or similar word substitution and reflect a disruption at the
phonologic level (eg, saying “ocoput” for “octopus”). Semantic paraphasias reflect a disruption at the lexical level; errors may be related to the target word (eg, saying “jellyfish” for “octopus”) or unrelated (saying “chicken” for “octopus”). A neologism is not a real word (eg, saying “ertup133” for “octopus”). Perseveration is the inappropriate repetition of a previous response that continues after the task requirements have changed and the response is no longer needed. A patient can perseverate on single words, concepts and ideas, or specific behaviors. Individuals may or may not be aware of their errors.

Each of the aphasic syndromes has both shared and unique characteristics; however, in reality, very few of them are pure, and people with aphasia have overlapping features. The subtypes of aphasia and their clinical characteristics are outlined in Table 38–6.

Table 38–6 Subtypes of aphasia.
Treatment

Aphasia rehabilitation is better understood as the process of achieving a closer connection between an individual’s language deficit and his or her ability to communicate functional needs and desires. The SLP must consider both what the patient and caregivers want to do, and what the patient can do in terms of communication ability. Many factors affect communicative needs and wants,
such as living environment, independence level, social interactions, and employment. The goals of treatment in therapy should always meet the following criteria, as outlined by Klein and Mancinelli:

- Promote function.
- Promote an effective communicative environment.
- Provide compensatory strategies to communicate.
- Provide education and counseling for adjustment of patient and family.
- Reduce interfering behaviors.
- Provide a relevant home program for patient and family.

**A. Direct Therapy**

Direct therapy for aphasia often involves targeting the deficit directly. For example, the tasks relating to lexical retrieval and sentence production therapy aim to improve functional communication by facilitating word retrieval and then producing the most grammatically correct utterance possible. Therapy exercises may include object identification, convergent and divergent naming, picture description, or conversational exchanges. Cueing hierarchies are used to elicit a response from the patient with the least amount of help from the clinician. As therapy continues, cues become less necessary. The patient is ultimately trained to use self-cuing and internal strategies for expressive language.

**B. Evidence-Based Treatment Programs**

Among the evidence-based treatment programs available to clinicians are melodic intonation therapy and compensatory communication strategies, such as visual action therapy.

1. **Melodic intonation therapy**—Melodic intonation therapy is a popular approach that involves musical and rhythmic intonations of phrases and sentences as the tool of language rehabilitation. The rationale for using melodic intonation is that the right hemisphere mediates musical stimuli and intonation. It is speculated that an intact right hemisphere may have the ability to improve the language functions of the left hemisphere when that hemisphere has sustained damage. The goal of this therapy is to stimulate the recovery of speech skills in severely nonfluent aphasic patients by using phrases that are intoned and paced syllable by syllable based on a melodic pattern, rhythm, and points of stress. The patient repeats melodic utterances of increasing length amid fading cues from
the clinician. For example, the clinician may begin by saying the word “Hello” in a particular melody while simultaneously separating the syllables (“Hel-lo”). The clinician then encourages the patient to join him or her in producing the target word together with the same melody. Once the patient achieves this step in the process with the maximum number of cues, the clinician begins to fade cues, for instance by producing the first sound of the target word and humming the rest, then humming only the tune of the target word, and eventually guiding the patient to independently produce the target word or phrase.

2. Compensatory strategies—Patients with severe aphasia may require compensatory communication strategies that can provide alternative ways to communicate wants and needs. An example of such a strategy is the functional communication board, which allows patients with poor verbal output to identify pictures, symbols, or written stimuli as a means of communicating more effectively with others. Patients can be taught to use an alternative communication device, such as speech-generating computer programs that build graphic symbols and pictures into phrases and sentences that are spoken aloud by a computer-generated voice. Gestures can also be taught, through visual action therapy, as a nonspeech method for the purpose of functional communication.


COGNITION
Cognitive communication impairments are changes in a person’s ability to talk, listen, read, or write because of changes related to cognitive, or thinking, skills. These skills include arousal, attention, executive function, memory, processing speed, visuospatial function, affect, and pragmatics. The major causes of
cognitive dysfunction include stroke, traumatic brain injury, and brain tumors. Traumatic brain injury can be caused by a closed or penetrating head injury, or a deceleration injury, leading to either focal or diffuse damage to the brain. In addition to the damage caused directly to the brain by the impact of the injury, secondary effects on brain function result from vascular disruptions, such as subdural hematomas and subarachnoid hemorrhages.

In most people, the left side of the brain contains the language centers while the right side plays a crucial role in cognitive functioning. Thus, right hemisphere brain damage (damage to the right cortical and subcortical structures) often leads to cognitive communication impairments. The frontal lobes are important for planning and initiation, judgment and reasoning, inhibition of behavior, and adaptation to change. These lobes also play a role in mediating memories for habits, motor activities, and emotional responses. Subcortical structures important for memory include the hippocampus and the amygdala. The hippocampus is important for recent working memory, while the amygdala’s primary role is in forming and storing memories associated with emotional events. The contributions of these and other brain structures involved in cognition was reviewed earlier in the chapter (see also Tables 38–1 and 38–2).

Clinical Evaluation

A. Symptoms and Signs of Impaired Cognition

The patient with cognitive–linguistic deficits may often be improperly referred to as having aphasia by other medical staff; however, aphasia is a primary disorder in the area of language. As previously noted, language impairments and cognitive impairments are unique entities that influence each other. Depending on the severity of the aphasia, it may be difficult to determine the extent to which aspects of cognition may be impaired. Major distinguishing features of cognitive communication disorders without aphasia include the absence of significant problems in naming, fluency, comprehension, and reading. Assessment and treatment of cognitive skills in the presence of aphasia is a challenge, as many tests and therapy plans depend on language skills to determine communication effectiveness.

B. Assessment of Cognitive Ability

Examination of all domains of cognition is of great important for successful diagnosis and rehabilitation of speech–language disorders. Attention, memory,
executive function, auditory processing, and visual processing are major cognitive functions, which are discussed below and summarized in **Table 38–7**.

**Table 38–7** Cognitive functions.
<table>
<thead>
<tr>
<th>Domain</th>
<th>Definition</th>
<th>Skills</th>
<th>Examples of Assessment Tasks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Attention</td>
<td>Selectively concentrating on one aspect of the environment while ignoring other things</td>
<td>Sustained attention—maintaining attention to tasks over time&lt;br&gt;Selective attention—disengaging, moving, and engaging attention to a different target&lt;br&gt;Alternating attention—moving between tasks that have different cognitive requirements&lt;br&gt;Divided attention—planning and coordinating multiple task demands simultaneously</td>
<td>Listen to a randomized list of spoken letters and raise a finger whenever a designated target is heard&lt;br&gt;Count to 10 against background noise&lt;br&gt;Shift attention between a writing task and a listening task&lt;br&gt;Watch television while answering yes–no questions</td>
</tr>
<tr>
<td>Memory</td>
<td>Registering, retaining, processing, storing, and retrieving information from the recent or distant past</td>
<td>Procedural memory—performing automatic tasks (e.g., driving a car)&lt;br&gt;Working memory—short-term storage for new or recently learned information&lt;br&gt;Semantic memory—factual and conceptual knowledge of the world&lt;br&gt;Episodic memory—memory of one’s own experiences and daily events</td>
<td>Repeat sentences or strings of numbers&lt;br&gt;Recall 3 words after a time lapse&lt;br&gt;Answer questions or retell the story from a paragraph&lt;br&gt;Answer orientation questions&lt;br&gt;Recall autobiographical information (answer questions about birth date and place, family, occupation, etc.)</td>
</tr>
<tr>
<td>Executive function</td>
<td>Planning, sequencing, and accomplishing goal-directed activities in a flexible manner as demanded by situational and environmental changes</td>
<td>Thought organization&lt;br&gt;Sequencing, planning&lt;br&gt;Problem-solving, initiation&lt;br&gt;Orientation, insight&lt;br&gt;Reasoning, judgment&lt;br&gt;Inhibition&lt;br&gt;Goal-oriented behavior</td>
<td>Demonstrated by patient behavior and discourse during clinical examination (Is the patient tangential, distractible, perseverative, withdrawn, disinhibited, etc?)&lt;br&gt;Verbally solve problems&lt;br&gt;Perform in vivo problem-solving tasks&lt;br&gt;Solve mazes or puzzles</td>
</tr>
<tr>
<td>Auditory processing</td>
<td>Taking in, interpreting, and understanding spoken communication</td>
<td>Processing speed—how slowly or quickly a person is able to listen, think, and respond&lt;br&gt;Auditory discrimination—gross and fine differences in sounds&lt;br&gt;Sound localization</td>
<td>Answer yes–no or open-ended questions&lt;br&gt;Follow multistep directions&lt;br&gt;Raise hand when the word “the” is spoken&lt;br&gt;Turn head toward sound&lt;br&gt;Assess for slow rise time, overstimulation, information capacity deficits, retention deficits</td>
</tr>
<tr>
<td>Visual processing</td>
<td>Taking in, interpreting, and understanding visual information</td>
<td>Visual perception—the ability to discriminate, analyze, and recognize familiar stimuli&lt;br&gt;Visual construction—combines visual perception skills with motor responses</td>
<td>Visually track items&lt;br&gt;Point to objects in response to questions&lt;br&gt;Complete cancellation tasks&lt;br&gt;Draw a clock&lt;br&gt;Assess for: visual inattention, visual neglect, visual field cut, gaze preference</td>
</tr>
</tbody>
</table>
During the evaluation, the patient’s emotional behaviors and social interactions are monitored. Affect describes a patient’s emotional state or the quality of his or her mood. Traumatic brain injury often causes changes in affective expression (how one expresses one’s feelings), resulting in personality changes, and changes in emotional capacity, tolerance for stress, or emotional behavior. Flat affect is the phrase that is used to describe a patient who loses motivation and drive, or seems to lack any emotional response. Emotional lability describes the patient who may laugh or cry inappropriately in response to minor events. Reactions of anger, frustration, anxiety, and depression may also occur.

Pragmatic skills are those needed to use language appropriately in social contexts. Examples include initiating communication, establishing appropriate eye contact, taking turns in conversation, and staying on topic. Impairments in these social skills are common after a brain injury. When assessing pragmatic skills and affect, careful observation of a patient’s discourse ability, nonverbal language (such as facial expression and eye contact), amount and appropriateness of gesturing and spontaneous movement, as well as direct questioning regarding his or her mood state are required. Reports from family or caregivers regarding the patient’s ability to express feelings help to supplement the information gathered.

Metacognitive processes such as self-awareness, insight, motivation, self-monitoring, and self-initiation are also crucial to assess, as these processes can influence responses to therapy and generalization of favorable responses to functional activities. Self-awareness and insight reflect the extent to which individuals possess an accurate, conscious representation of their functional disability. A patient’s level of self-awareness and insight can best be judged by his or her actions or what is said about the disability (eg, persistently trying to get up to use the bathroom despite physical limitations). Attempts to self-correct or compensate for errors is an indication of self-monitoring. Self-initiation refers to the ability to start or begin an intended action as compared with responding reflexively to stimuli. Sometimes patients acknowledge what they should be doing, yet fail to initiate the action.

**Implications for Treatment**

The extent to which cognitive skills in all domains are spared or impaired can significantly influence response to therapy and functional outcome. At the most
basic level, therapy requires individuals to attend and concentrate. Purposeful learning relies on memory skills, and executive functions are crucial to independent implementation of compensatory measures to ensure successful communication. The integrity of visuospatial skills is also important for the use of compensatory communicative techniques such as writing, drawing, and gesturing. Cognitive–linguistic therapy involves targeting deficits directly to relearn skills, teaching compensatory strategies, modifying the environment and caregiver interactions, as well as providing family education and counseling.

Physicians, residents, medical students, nurses, and personal care assistants interact with patients who have cognitive–linguistic deficits on a daily basis. The SLP can provide these personnel with a description of the specific areas affected as well as strategies to aid in communication. General suggestions in relating to this type of patient include the following:

• Speak to the patient as an adult.
• Keep the patient updated to time, place, and events in the patient’s routine.
• Ask simple, direct questions (eg, “Where are you right now?” instead of “Do you know where you are right now?”).
• Remember that sometimes the patient has slower processing speed; allow extra response time before repeating questions or directions.
• Support or encourage speech efforts.

**ATTENTION DEFICITS**

### Clinical Findings

Individuals with impairments in attention may be unable to sustain attention long enough to complete a sentence or task, or to comprehend lengthy amounts of information. Patients may be easily distracted by surrounding noise in their environment. The patient with impairment in alternating attention has difficulty moving from one task to another and easily forgets where he or she left off on a previous task.

### Treatment

Treatment for these deficits involves attention training exercises, in which
patients practice such skills as sustaining attention over time or shifting attention between different tasks. SLPs may teach patients how to set up their environment so that their attention problems are least likely to interfere with their daily functioning. For example, a patient may be trained to utilize checklists, eliminate extraneous noises, or say the steps of a task out loud as they are completed.

Family and caregivers are counseled on ways to help the patient cope. Suggestions may include helping the patient pace himself or herself during an activity, eliminating unnecessary distractions (such as the television or radio), and reducing demands by utilizing organizational devices (eg, calendars, message boards). Families are also educated on effective communication strategies such as making sure the patient is visibly paying attention by looking at the speaker, or allowing the patient to finish what he or she is concentrating on before interrupting or switching to something different.

MEMORY DEFICITS

Clinical Findings

Patients with deficits in short-term memory might be repetitive in their output, forget task instructions, or forget something that was said a few minutes, hours, or several days ago. Severe deficits in long-term memory or autobiographical recall can cause a patient to forget personal likes and dislikes, or fail to recognize family members. Memory impairments often lead to behaviors that can endanger a patient, such as forgetting to lock the door upon leaving the apartment.

Treatment

Therapy involves mental exercises to improve recall of salient information directly, and instruction on utilizing compensatory strategies with either internal cueing or external aids. Internal cueing strategies include associations (making associations between names and related information); rehearsal (silently repeating new information several times); visual imagery ( picturing what one wants to remember); mnemonic devices (such as using the first letter of each word in a phrase to form an acronym that can be easily remembered); chunking (creating meaningful groups for details); and saliency (focusing on the most important information in a message).
External memory aids are highly recommended. Examples include keeping a schedule of a daily routine, setting a timer for activities, making lists of groceries or things to do, keeping phone number/address books, using cue cards to remember names, and using calendars for orientation and tracking of appointments. Patients are also encouraged to keep a pen or pencil and notepad at the bedside or in a pocket to write down information. Family members often play a large role in helping the patient comply with these strategies.

**EXECUTIVE FUNCTION DEFICITS**

**Clinical Findings**

Executive functions, such as organizing, planning, sequencing, problem solving, reasoning, and judgment, are critical skills necessary for completing activities of daily living. Self-monitoring and impulse control contribute to successful performances. If these functions fail, a person may be tangential, distractible, disinhibited, or perseverative. Patients might have impaired ability in organizing thoughts to tell a cohesive story or sequencing steps to a task. Patients may also have difficulty understanding cause and effect, display decreased awareness of unsafe situations, or show inflexibility in solving problems.

**Treatment**

Therapy focuses on relearning these skills through verbal and in vivo problem solving, heightening deficit awareness, and compensating for deficits through the use of strategies to maximize the patient’s ability to complete daily activities. Compensatory strategies may include writing out goals and plans for the day, utilizing checklists, setting timers to finish tasks in a timely fashion, and completing tasks with a step-by-step approach. Family counseling may focus on teaching caregivers to recognize these problems as the result of a traumatic brain injury, as often caregivers can become frustrated and annoyed with a patient who is no longer is able to function independently in activities of daily living.

**AUDITORY PROCESSING DEFICITS**
Clinical Findings

Patients with deficits in comprehension of auditory information are likely to experience difficulty listening to a lively conversation, following multistep directions, understanding stories, or answering complex questions.

Treatment

Therapy is focused on teaching the patient listening strategies, which may include making sure he or she can see the speaker’s mouth; paying attention to the speaker’s facial expressions and body language; asking the speaker to repeat, rephrase, or simplify information; verifying information by paraphrasing what has been said; when entering a group in the middle of a conversation, asking someone to summarize what was talked about; and, if a conversation is occurring in a noisy environment, moving into a quieter environment.

VISUAL PROCESSING DEFICITS

Clinical Findings

Visual perceptual problems, such as field cuts, right-sided or left-sided neglect, or inattention to one side of space, can have serious consequences for everyday functions such as reading and writing, eating a meal, shaving, or driving safely. Spatial disorientation can lead to loss of navigational ability; thus, a patient might lose his or her way easily, even in familiar environments. Spatial inflexibility results in the loss of the ability to mentally manipulate objects in space. In-depth evaluation of visuoperceptual deficits should be conducted to discern what type of deficit is present.

Treatment

Interventions focus on relearning visual processing skills by completing therapy tasks involving visual discrimination, visual memory, scanning, tracking, or focusing. Heightening the patient’s awareness of his or her own visual deficits is crucial to making progress. Compensatory visual strategies might include covering parts of a page and looking at exposed areas systematically, using a
finger drag to assist in scanning, using large print or audio-books, or turning one’s head to compensate for a field cut.


SPEECH & VOICE

Speech and voice manifest through movements triggered by cranial and spinal nerves that innervate respiratory, phona-tory, resonatory, and articulatory muscles. Motor (efferent) fibers transmit commands from the central nervous system, through direct and indirect pathways, to activate muscles and glands throughout the body. The basal ganglia and cerebellum play key roles in regulating their movements.

Respiration, phonation, resonance, and articulation make up the bases of motor speech and voice production. Speech and voice processes rely heavily on particular structures and muscles, including the lips, teeth, tongue, hard palate, velum, velopharyngeal wall, larynx, vocal cords, and diaphragm. The larynx (commonly referred to as the “voice box”) houses the vocal folds, which are essential for phonation. It functions as a biologic valve for regulation of sound production and respiration. The larynx is innervated by the vagus nerve, in particular the recurrent laryngeal nerve and superior laryngeal nerve branches, which contains both motor and sensory fibers.

The power sourcing for voicing is respiration. The muscles of the chest wall (the rib cage, diaphragm, and abdomen) regulate breathing to provide a stream of exhaled air. The regulation of exhaled air affects the loudness of speech, number of words spoken on a single breath, and duration of pauses between breaths. The exhaled air is then passed through the larynx and causes the vocal folds to
vibrate to produce voice (phonation). Muscles in the larynx adjust the length and tension of the vocal folds to regulate the pitch and loudness of the voice. Muscles in the throat alter the quality of the voice (resonance). The jaw, lips, tongue, and palate change the shape of the mouth to produce sounds of speech (articulation).

▶ Clinical Evaluation

A. Symptoms and Signs of Speech and Voice Disorders

A stroke, brain tumor, or traumatic brain injury can interrupt the neurologic pathways for normal phonation and articulation. Deficits in the dominant hemisphere’s speech planning and programming areas, as well as in the motor system’s control and neuromuscular execution, can lead to motor speech deficits. Two such deficits, dysarthria and apraxia of speech, are contrasted below. Voice disorders can result from nerve damage, but can also have nonorganic causes.

B. Assessment of Impaired Speech and Voice

When evaluating a patient with a suspected motor or speech disorder, as much diagnostic information as possible is gathered to discern the underlying mechanisms involved and resulting in impairment. Respiratory, phonatory, resonatory, and articulatory systems work together to produce voicing and speech, and breakdowns can occur in one or more of these systems. If a patient presents with a deficit, each system must be evaluated to determine where the deficit originates in order to properly diagnose and provide treatment.

Evaluations typically begin with an oral peripheral mechanism examination to assess oral–motor symmetry, strength, range of motion, tone, accuracy and coordination of movements, steadiness, and speed. Muscles of the face and speech structures are examined at rest, during movement, and during sustained postures. Asking the patient to perform nonspeech tasks, like blowing a kiss, also helps to assess oral–motor function. Respiration for speech may be assessed by asking the patient to produce sustained phonation of an open vowel for as long as possible. During phonation tasks, speech intelligibility is rated at syllable, word, sentence, and conversational level. Rate of speech and articulatory precision are noted during diadochokinetic tasks, which assess how quickly a person can accurately repeat a series of rapid, alternating phonetic sounds. Voice is assessed by noting various perceptual characteristics (as perceived by the clinician), which include pitch, quality, resonance, and vocal...
intensity. The SLP may then judge communication effectiveness, or the ability to understand speech in one-on-one situations, versus noisy crowds, versus known and unknown contexts.

Motor speech disorders are speech production deficits that result from impairments in the neuromuscular or motor control system, or both. The signals that control speech musculature are affected, or muscle weakness or rigidity is present, thereby altering the speech signal. A voice disorder is a persistent abnormality in the sound of a voice. It is a condition in which the voice is problematic to the user in social, professional, and other contexts. Voice disorders are classified as organic, psychogenic, or functional.

**DYSARTHRIA**

**Clinical Findings**

Dysarthria is a collective name for a group of speech disorders caused by disturbances in muscular control over the speech mechanism and resulting from damage of the central or peripheral nervous system. In patients with these disorders, problems in oral communication stem from paralysis, weakness, or incoordination of speech musculature. Dysarthria may affect the ability to vocalize sufficiently, or to properly move the muscles of the respiratory system, velum, or the articulators quickly and accurately. These impairments may result in slurred speech sounds, uneven, harsh, quiet, or slow output. Several different types of dysarthrias are differentiated, based on the underlying neuropathology and the nature of the motor disturbance (Table 38–8).

| Table 38–8 | Types of dysarthrias. |
Treatment

Therapy for dysarthria involves targeting the deficits directly and teaching compensatory strategies. The clinician may target one or more of the previously mentioned subsystems to increase overall speech intelligibility and increase communicative effectiveness.

Working on placement of articulators and facilitating muscle strength through oral motor exercises may improve articulatory precision. For example, lingual elevation and lateralization exercises against resistance may improve strength and range of motion necessary for particular sound productions. It
should be noted that oral motor exercises are a controversial treatment method, as critics argue that movements elicited are not necessarily the ones used for speech production. There is no conclusive evidence confirming the efficacy of muscle strengthening in treating dysarthria, but it appears to have positive effects for at least some patients.

Compensatory speech strategies are taught for immediate modifications to speech that may improve overall intelligibility. Some patients benefit from elaborating or overarticulating utterances on a syllable-by-syllable or word-by-word level. Other patients who have rapid speech benefit from pacing techniques to slow their rate. Gestures and writing may also help to enhance a patient’s verbal message.


APRAXIA

General Considerations

Whereas dysarthria involves disturbances in speech stemming from impaired muscular control, apraxia of speech reflects disturbances in the planning or programming involved in the movements of speech. There is a distinct difference between the two. Apraxia is a neurologic speech disorder that reflects an impaired capacity to plan or program sensorimotor commands necessary for
directing movements that result in phonetically and prosodically normal speech. It can occur in the absence of physiologic disturbances associated with the dysarthrias and in the absence of disturbance in any component of language.

**Clinical Findings**

Clinical findings associated with apraxia include nonverbal and verbal characteristics. Nonverbal oral apraxia is characterized by the inability to imitate or follow volitional oral commands (eg, whistling or blowing). Verbal apraxia manifests as difficulties with tasks that place demands on the sequencing of various sounds and syllables with varying patterns of stress. Consonant and vowel distortions, slow overall rate, syllable segregation, successful and unsuccessful attempts to self-correct articulatory errors, and effortful groping for articulatory postures are common characteristics seen in patients with apraxia. Speech sequential motion rates and imitation of complex multisyllabic words and sentences are generally the most difficult tasks for these patients to perform.

**Treatment**

Treatment for apraxia includes motor-learning techniques to help the patient regain and relearn motor speech skills. Repetitive drills in which the patient repeats commons sounds, words, and phrases are often crucial in order for the brain to relearn accurate motoric programming for various speech productions. Integral stimulation is an approach that includes “watch me, listen to me, and say it with me” in order to increase self-awareness of sound placement and auditory feedback.

Another technique involves articulatory cueing. This technique increases awareness and actualization of articulatory postures or movements, or both, and may take the form of phonetic placement or phonetic derivation. Phonetic placement cues use descriptions of what (which articulators), where (positioning or location), and how (manner, voicing) sounds are made, using descriptions from verbal explanations, visual modeling, drawings, and physical cueing by the clinician of the orofacial musculature. When working with patients with apraxia, it is of particular importance to guide treatment in a hierarchical format. Specificity and consistency within treatment sessions are important.

Important strategies that can be implemented to assist patients in optimizing speech output include limiting the number of conversational partners, reducing
background noise, and facilitating closer speaker–listener proximity. As always, the SLP must educate the patient, family members, and caregivers on these goals of care.


### VOICE DISORDERS

#### General Considerations

Aphonia is defined as loss of phonation. Dysphonia is the term used to describe a disorder of voice. It is an impairment in the ability to produce voice sounds using the vocal organs. Voice disorders can result from multiple conditions or adverse events. Surgical and medical procedures can result in detrimental effects on voice production through direct injury to laryngeal structures or through damage to peripheral nerves (ie, during extubation). Atypical or functional voice disorders (paradoxical vocal cord movement, hyperfunctional vocal use, or increased muscle tension) may occur as reactions to illness, medical treatment, or traumatic life events. Neurogenic causes of dysphonia include paresis, paralysis, or spasmodic dysphonia. Organic causes may include vocal nodules, polyps, cysts, or other mass lesions.

#### Clinical Findings

Common overall symptoms include hoarseness, breathiness, reduced loudness, vocal fatigue, pitch breaks or inappropriate pitch, and strained or strangled voice. By assessing vocal characteristics and determining the underlying cause of the dysphonia, proper treatment can be implemented.

#### Treatment
It is recommended that otolaryngology services perform a laryngoscopy to properly assess the larynx and vocal cords before initiating therapy. Voice therapy is usually the first line of treatment for hyperfunctional dysphonia that has resulted in vocal fold lesions such as vocal nodules, polyps, or cysts. A program may be designed to reduce vocal abuses through guided change in vocal behaviors and lifestyle changes. Therapy ultimately aims to eliminate harmful vocal behavior, shape healthy vocal behavior, and teach appropriate or modified vocal use in order to alleviate symptoms, eliminate or reduce size of lesions, or prevent further trauma from occurring. Recommendations for improving vocal hygiene may include increased hydration; decreased shouting, overuse, and straining; and elimination of nonpurposeful voicing. Patients are also instructed to modify their intake of food and liquids that exacerbate reflux (which may reach the level of the vocal cords and irritate the mucosa).

Another target of voice therapy is coordination of respiration and phonation. For phonation to occur, respiratory support must be strong enough to create subglottic pressure to assist in movement of the vocal cords (mucosal vibration). If a patient has poor respiratory drive for voicing; the clinician may begin teaching patients techniques to improve breath support and promote efficient use of the airstream. Using an incentive spirometer may help to improve overall lung function. Diaphragmatic breathing exercises help with relaxation and decrease shallow breathing patterns so that a patient will not speak on residual air.

If impaired vocal cord mobility resulting from paresis or paralysis is the problem, clinicians may teach patients to improve medial glottal closure through push–pull exercises. These tasks aim to have the healthy vocal cord move more toward midline, to compensate for the immobile cord, or attempt to get the immobile cord moving again.


Johnson AF, Jacobson BH: Medical Speech–Language Pathology: A

TRACHEOSTOMIES

Within the hospital or rehabilitation setting, many patients with tracheostomies are evaluated and treated for voice and swallowing deficits. A tracheotomy (surgical opening in the trachea) may be performed because of an obstruction, blockage, or swelling that prevents normal airflow between the mouth, nose, and lungs. It can also be performed when prolonged connection to an artificial ventilator is required. Air is inhaled through the tube in the neck, not through the
mouth or nose. The purpose of the tracheostomy tube is to provide an adequate airway and provide an easy method of removing secretions from the trachea and lungs.

Speech and voice may be difficult for patients with tracheostomies. The ability to vocalize will depend on how open the airway is and the health of the vocal cords. Although the tracheostomy may be temporary, during that time patients will need some way to communicate or vocalize needs and wants throughout daily activities. If a patient is an appropriate candidate for a Passy-Muir valve (PMV) this one-way speaking valve can be utilized to help the patient speak more clearly, allowing him or her to direct personal care.

Prosthetic Treatment: Use of the Passy-Muir Speaking Valve

The PMV is a small, lightweight, one-way valve attached to tracheostomy tubes that allows the inflow and outflow of air so users can speak without manual occlusion. The valve is always in a closed position until the patient inhales, which helps restore the patient to a more “normal” closed respiratory system. The valve opens easily with less-than-normal inspiratory pressures and closes automatically at the end of the inspiratory cycle without air leak and without patient expiratory effort. It then redirects air flow up and through the upper airway and vocal folds, allowing phonation to be produced.

A. Clinical Indications

Various clinical indications are assessed before medical placement of a PMV. Patients who meet the following criteria may be considered for PMV use: (1) minimum of 48 hours post-tracheostomy placement; (2) expressive communication attempts; (3) alert and responsive; (4) stable vital signs (heart rate, respirations, blood pressure, and oxygen saturation); and (5) able to tolerate full cuff deflation. Contraindications to PMV use include severe aspiration risk, upper airway obstructions (eg, tracheal or laryngeal stenosis), and thick or excessive secretions.

B. Placement and Monitoring Considerations

Prior to attachment of the valve, the tracheostomy cuff must be completely deflated and the patient assessed for signs and symptoms of respiratory insufficiency. Tracheal and oral suction is performed and the patient is instructed
to inhale through the tracheostomy tube and exhale through the mouth or nose while the tracheostomy is manually occluded. Following digital occlusion, voicing may or may not be achieved due to lack of use of the vocal cords, damage, or paralysis. Although this may not preclude placement, if air trapping is present, indicating airway obstruction or an oversized tracheostomy tube, after exhalation the valve should not be attached. If the patient is able to exhale or voice adequately, the PMV can be attached and the patient subsequently monitored for tolerance. While the valve is in place, clinicians should be conscious of changes in vital signs (eg, heart rate, respiratory rate, and oxygen saturation), breath sounds, color and responsiveness, and work of breathing.


**SWALLOWING**

Swallowing of food and liquid is the process by which humans receive nutrition, which is essential to the rebuilding and maintenance of good health. Dysphagia can be defined as a disorder of swallowing. It can result from mechanical interference or obstruction (head and neck cancer), neurologic insult (stroke or traumatic brain injury), or pulmonary complications. It can exist at any or all of the following stages: oral–preparatory, oral, pharyngeal, and esophageal. Although broken down into stages, swallowing performance should be thought of as one behavior with four components that act in an integrated manner if swallow success is to be achieved. A swallowing disorder differs from a feeding disorder, which is an impairment in the process of gathering food outside of the nutritional system.

**Physiology of Swallowing**

The initial stage (oral–preparatory) of swallowing begins with the acceptance and containment of the food or liquids. The food is manipulated and prepared into a cohesive bolus in the oral cavity. Crushing, repetitive movement of the jaw to help reduce bolus size is known as mastication (chewing). This activity is not required for a liquid bolus. Chewing activity stimulates the salivary glands,
allowing moisture to form and lubricate the bolus. The oral stage of swallow begins after the bolus is prepared. The tongue moves the bolus posteriorly in the mouth and when it reaches the faucial pillars, a swallow response is initiated. The oral–preparatory and oral stages are volitional, as these actions can be easily altered by the patient.

When the bolus begins its transfer to the pharynx and touches the posterior wall, several physiologic events are initiated that comprise the pharyngeal stage of swallowing (Figure 38–2). The velum elevates to seal the nasopharynx. The base of the tongue retracts and moves posteriorly to make contact with the posterior pharyngeal wall, which helps propel the bolus into the hypopharynx. Posterior retraction of the tongue lifts the hyoid bone, which causes the larynx to be pulled up and forward. The true vocal cords adduct, the larynx elevates approximately 1 inch, and the epiglottis inverts to seal or close the airway, preventing penetration or aspiration of food or liquids into the trachea. Pharyngeal constriction helps to propel the bolus through the pharynx into the esophagus, the channel to the stomach.
Figure 38–2  The swallowing mechanism. **A:** Lateral view of the oropharynx.  **B:** Bolus transferred from the oral cavity to the pharynx.  **C:** Bolus transit through the pharynx (note that the larynx is elevated and the epiglottis inverted).  **D:** Bolus transit through the pharynx.  **E:** Bolus passage to the proximal esophagus.

At the beginning of the esophagus lies the upper esophageal sphincter (cricopharyngeus), which is normally contracted at rest. Upon initiation of swallowing, the cricopharyngeus muscle relaxes in anticipation of the bolus and opens when the food passes. It then immediately closes, preventing backflow or reflux from the esophagus into the pharynx. Swallowing continues automatically as the bolus is swept downward to the stomach by peristalsis in waves of synchronized muscle movements. Both the pharyngeal and esophageal stages normally are depicted as more reflexive events. However, the act of swallowing does not qualify as a true reflex, as variations in the stimulus, such as different bolus textures and volume, can cause physiologic changes.

Swallowing is best understood as a programmed response to sensory stimuli with underlying neural circuitry that can adapt to change if needed. The key integrator of the swallow sequence is the medulla, lying in the brainstem. Six cranial nerves (CNs) provide the peripheral afferent and efferent component of the muscles involved in swallowing. The trigeminal nerve (CN V) carries sensory information from the oral cavity, and sends efferent fibers to the muscles of mastication. The hypoglossal nerve (CN XIII) is the motor nerve to most of the tongue musculature. The facial nerve (CN IV) innervates the facial muscles, including the lips, and mediates taste for the anterior tongue. The mediator of taste for the posterior part of the tongue is the glossopharyngeal nerve (CN IX), which also innervates muscles involved in bolus propulsion.

Muscles involved in bolus propulsion are also innervated by the vagus nerve (CN X), which plays a large role in swallowing performance. The pharyngeal branch innervates the upper esophageal sphincter, the superior laryngeal nerve provides sensory innervation to the epiglottis and structures in and around the airway, the inferior laryngeal nerve supplies most of the motor components to the larynx and muscles involved in airway closure, and autonomic fibers innervate the smooth muscle portion of the esophagus. The vagus nerve also innervates organs such as the lungs, which may be linked to normal swallowing performance because of their role in respiration. The muscles involved with swallowing, and their innervation, are described more fully in Table 38–9.
Table 38–9 Neurologic and anatomic basis of swallowing.
<table>
<thead>
<tr>
<th>Action</th>
<th>Muscle</th>
<th>Function</th>
<th>Cranial Nerve Innervation</th>
</tr>
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<tbody>
<tr>
<td><strong>ORAL-PREPARATORY PHASE</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prepares food for swallowing (~ 15-30 chews)</td>
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<tr>
<td>Mouth opens</td>
<td>Mylohyoid</td>
<td>Depresses jaw</td>
<td>Trigeminal (CN V)</td>
</tr>
<tr>
<td></td>
<td>Geniohyoid</td>
<td>Depresses jaw</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lateral pterygoid</td>
<td>Protrudes jaw</td>
<td></td>
</tr>
<tr>
<td>Bolus enters; labial seal on cup or utensil</td>
<td>Medial pterygoid</td>
<td>Elevates jaw</td>
<td>Facial (CN VII)</td>
</tr>
<tr>
<td></td>
<td>Orbicularis oris</td>
<td>Closes mouth</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Zygomatic</td>
<td>Draws angles of mouth laterally</td>
<td></td>
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<tr>
<td>Tongue is cupped to hold liquid; tongue moves solids to molars for chewing</td>
<td>Genioglossus</td>
<td>Retracts and protrudes tongue</td>
<td>Hypoglossal (CN XII)</td>
</tr>
<tr>
<td></td>
<td>Styloglossus</td>
<td>Pulls tongue up and back</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Palatoglossus</td>
<td>Elevates tongue</td>
<td></td>
</tr>
<tr>
<td>Jaws open and close to grind food</td>
<td>Masseter</td>
<td>Closes, lifts, and raises jaw</td>
<td>Trigeminal (CN V)</td>
</tr>
<tr>
<td></td>
<td>Temporalis</td>
<td>Elevates and protrudes jaw</td>
<td></td>
</tr>
<tr>
<td>Saliva helps bolus form</td>
<td>(Salivary glands)</td>
<td></td>
<td>(IX) Glossopharyngeal</td>
</tr>
<tr>
<td>Cheeks flatten to keep food in</td>
<td>Risorius</td>
<td></td>
<td>Facial (CN VII)</td>
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<td></td>
<td>Buccinator</td>
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<tr>
<td>Taste and sensation</td>
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<tr>
<td><strong>ORAL PHASE</strong></td>
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<tr>
<td>Posterior Oral Transit of Bolus to Pharynx (~ 8 sec)</td>
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<tr>
<td>Posterior tongue drops</td>
<td>Mylohyoid</td>
<td>Depresses jaw</td>
<td>Trigeminal (CN V)</td>
</tr>
<tr>
<td></td>
<td>Geniohyoid</td>
<td>Depresses tongue</td>
<td>Vagus (CN X)</td>
</tr>
<tr>
<td></td>
<td>Lateral pterygoid</td>
<td>Pulls sides of tongue down</td>
<td>Hypoglossal (CN XII)</td>
</tr>
<tr>
<td>Tongue tip elevates</td>
<td>Superior longitudinal</td>
<td>Pulls tongue tip upward</td>
<td>Vagus (CN X)</td>
</tr>
<tr>
<td>Tongue squeezes bolus against hard palate in a sweeping motion</td>
<td>Palatoglossus</td>
<td>Elevates tongue</td>
<td>Hypoglossal (CN XII)</td>
</tr>
<tr>
<td></td>
<td>Hypoglossus</td>
<td>Pulls sides of tongue down</td>
<td></td>
</tr>
<tr>
<td>Velum begins to elevate</td>
<td>Levator veli palatini</td>
<td>Elevates velum</td>
<td>Vagus (CN X)</td>
</tr>
<tr>
<td></td>
<td>Uvular</td>
<td>Shortens and lifts velum</td>
<td></td>
</tr>
<tr>
<td>Pharyngeal walls constrict</td>
<td>Superior, middle, and inferior constrictors</td>
<td>Constrict pharynx</td>
<td>Glossopharyngeal (CN IX) and vaug (CN X)</td>
</tr>
<tr>
<td>Lips seal tightly</td>
<td>Orbicularis oris</td>
<td>Pull lips together</td>
<td>Facial (CN XII)</td>
</tr>
<tr>
<td><strong>PHARYNGEAL STAGE</strong></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Reflexive Swallow Begins (~ 1-2 sec)</td>
<td></td>
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<tr>
<td>Bolus contacts faacial pillars, soft palate, and posterior tongue base</td>
<td></td>
<td>Reflexive swallow</td>
<td></td>
</tr>
<tr>
<td>Velem continues to elevate</td>
<td></td>
<td></td>
<td>Vagus (CN X)</td>
</tr>
<tr>
<td>Vocal folds adduct</td>
<td>Lateral cricoarytenoid</td>
<td>Close vocal folds</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Circular cricoarytenoid</td>
<td>Close vocal folds</td>
<td></td>
</tr>
<tr>
<td>Hyoid bone and larynx move up and forward</td>
<td>Digastric</td>
<td>Raises hyoid bone up and forward</td>
<td>Glossopharyngeal (CN IX)</td>
</tr>
<tr>
<td></td>
<td>Geniohyoid</td>
<td>Elevates larynx</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hypoglossus</td>
<td>Elevates hyoid bone</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Stylopharyngeus</td>
<td>Elevates pharynx</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Geniohyoid</td>
<td>Elevates larynx</td>
<td>Trigeminal (CN X)</td>
</tr>
</tbody>
</table>
Clinical Evaluation

A. Physical Examination

The patient’s mental status and posture are first assessed to determine appropriateness for oral swallowing trials. Also noted is the patient’s respiratory status and coordination of respiration and swallowing. The action of swallowing interrupts the exhalation phase of the respiratory cycle (to swallow) and return to exhalation (after the swallow). Patients with pulmonary conditions may be unable to tolerate this “swallowing apnea,” which may further compromise their respiratory status.

The clinical assessment typically proceeds with an examination of the anatomic structure of the oral cavity, including its symmetry and structural integrity. Observation of lingual function, lip closure, and velar elevation can be done through speech and nonspeech tasks. The presence and status of any oral
secretions should be noted, especially the pooling of secretions or excessive, drooling, or dried secretions. The frequency of spontaneous swallowing, the ability to initiate a dry swallow without any stimuli, the ability to cough, and the presence of a gag reflex, are other variables to consider. The patient undergoes trials with various food and liquid consistencies to determine the presence of dysphagia and risk or signs of aspiration. Base of the tongue retraction, laryngeal elevation, and timeliness of swallow initiation are assessed using digital laryngeal palpitation.

**B. Symptoms and Signs of Impaired Swallow**

Dysphagia can result from a mechanical interference or obstruction (head and neck cancer), neurologic insult (stroke or traumatic brain injury), or pulmonary complications. Various signs and symptoms of dysphagia may be observed in the oral phase. If the patient has significant labial weakness, he or she may be unable to accept oral nutrition because of difficulty securing a labial seal on a spoon or straw. Food leaking from the mouth may result from weakness of the muscles needed to contain the bolus in the oral cavity. Difficulty chewing or manipulating the food into a cohesive bolus and moving it posteriorly may result in oral or buccal retention (referred to as “pocketing”). The patient may be observed holding the food in the mouth. If velar elevation cannot be maintained during the swallow, nasal regurgitation can occur. Poor cognition may also affect this volitional stage of swallowing; a patient may demonstrate poor awareness of food substances (failure to chew) or decreased attention to the act of swallowing (eg, talking while chewing).

Pharyngeal dysphagia may present as difficulty initiating a swallow, resulting in latency in the swallow reflex. Laryngeal elevation may be decreased (normal elevation is 1 inch) and muscle excursion may be sluggish owing to weakness. Nasal regurgitation and complaints of stasis (pharyngeal retention, or “food sticking in the throat”) are other indicators of a pharyngeal dysphagia. Obvious signs of aspiration (food or liquids entering the airway) include coughing, choking, or throat clearing. Changes in pulmonary status (eg, oxygen saturation level dropping, changes in work of breathing), sweating, tearing of the eyes, or skin color changes are other signs of possible aspiration.

**C. Additional Tests**

The pharyngeal and esophageal stages of swallowing are sometimes difficult to evaluate in bedside examinations. Furthermore, some patients aspirate “silently” or may demonstrate no sensory response after an aspiration event. In such cases,
Oropharyngeal dysphagia can be further evaluated using a modified barium swallow study via videofluoroscopy, which provides continuous radiographic imaging of a patient during oral intake. Videofluoroscopy can provide valuable information about the adequacy of airway closure and the coordination of respiration and swallowing. It is also helpful in evaluating a patient’s ability to swallow various materials, including the sensory or behavioral reactions that occur, and can be useful in evaluating the impact of compensatory therapy maneuvers on swallowing function and airway protection.

D. Post-evaluation Monitoring

Following the evaluation, if a patient is approved to begin an oral diet, he or she is closely monitored for signs of aspiration pneumonia. A patient who develops spiking fevers, an increased white blood count, increased secretions or congestion after eating, or new infiltrates (primarily of the right lower lobe) on a chest radiograph may not be tolerating the diet.

Treatment of Impaired Swallowing

When managing a patient with dysphagia, clinicians must consider three important variables: airway protection, nutrition, and hydration. If there is a risk that food or liquid may enter the lungs, resulting in aspiration pneumonia, nonoral feeding by means of a nasogastric tube may be necessary. Crary and Groher group the behavioral interventions for dysphagia that are effective in facilitating a return to eating into three categories: modifying the food, modifying the patient and feeding activity, and modifying the swallowing mechanism.

In the rehabilitation setting, the SLP will work closely with the dietician, whose role is to monitor caloric intake versus caloric needs, wean the patient off tube feedings, and provide food consistent with the patient’s preferences and cultural limitations. Upon discharge, staff should provide extensive family or caregiver education and training. Instructions may include how to thicken liquids, comply with aspiration precautions, ensure the most effective communication with the patient, and utilize external aids.

A. Dietary Modifications

The first treatment avenue pursued for a patient with dysphagia is diet modifications to allow immediate return to eating. Foods and liquids may be
prepared in various textures or consistencies that are appropriate for the patient’s current swallowing ability. Foods may be pureed, chopped, mixed, or blended to reduce the need for chewing. Patients with oral motor weakness or patients with compromised respiratory systems who become out of breath from chewing may benefit from these changes. Thickening of liquids is often done in an attempt to slow liquid bolus transit and form a slightly more cohesive bolus. These changes can give patients with a delayed swallowing reflex, or with poor breathing and swallowing coordination, a better opportunity to swallow without (or with less) risk for aspiration. The thickness of liquids varies and is described as having a pudding, honey-thick, or nectar-thick consistency. As the patient improves, diets are advanced to higher levels with the goal of progressing to a regular diet with thin liquids.

Modifications may also include altering the size, temperature, taste, or smell of a food or liquid bolus to increase sensory stimulation or decrease aspiration risk. Cold materials are thought to enhance awareness of the bolus. Hot materials typically are ingested in smaller amounts. Taste and smell alterations may contribute to changes in appetite, motivation, and enjoyment of meals. Modifications to feeding activity are beneficial to patients who are unable to safely and adequately feed themselves. Patients who experience poor oropharyngeal clearance may benefit from alternating bites of food and sips of liquid, in the manner of a “liquid wash” or cleansing mechanism.

**B. Compensatory Strategies and Postural Adjustments**

For certain patients, compensatory swallowing strategies and postural adjustments can allow for safer oral intake or improved swallow function. Changes in head posture may include extension, flexion, or rotation. These changes in posture can redirect the bolus and may change the speed of bolus flow, giving the patient time to adjust the swallow. For example, the “chin tuck” has been shown to facilitate improved airway protection as it has the anatomic effect of narrowing the oropharynx, widening the valleculae, and reducing the distance between the hyoid and the larynx. The supraglottic swallow and super-supraglottic swallow maneuvers are techniques designed to protect the airway from aspiration of food or liquid by closing the airway before swallowing (with an effortful breath hold) and then coughing immediately after the swallow to clear any residue that may be on the vocal folds. These therapy techniques are not ideal for patients who are at risk for noncompliance because of cognitive or behavioral limitations.
C. Oral Motor Exercises

During dysphagia therapy, the specific swallowing deficit is targeted directly and attempts are made to modify the swallowing mechanism through motor exercises or sensory stimulation, or both. Oral motor exercises against resistance are used to improve strength. Range of motion may be increased by stretching exercises. Some common pharyngeal strengthening exercises are described in Table 38–10.

Table 38–10 Pharyngeal strengthening exercises.

<table>
<thead>
<tr>
<th>Exercises</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Masako maneuver</td>
<td>Patient holds the anterior tongue between the teeth while swallowing to increase strength of base of tongue and its ability to retract and touch the posterior pharyngeal wall</td>
</tr>
<tr>
<td>Mendelsohn maneuver</td>
<td>Patient is asked to suspend the swallow at the peak of hyolaryngeal elevation and pharyngeal contraction and to prolong this posture for a couple of seconds before relaxing and allowing the swallowing mechanism to return to the pre-swellow position</td>
</tr>
<tr>
<td>Effortful swallow</td>
<td>Patient attempts to increase the force applied to the bolus from structures within the swallowing mechanism by swallowing “hard” or “forcefully”</td>
</tr>
<tr>
<td>Thermal–tactile stimulation</td>
<td>Provision of a sensory stimulus before a swallow attempt may serve as an alerting mechanism to the nervous system to help prepare the swallowing mechanism for the subsequent swallow; achieved by presenting cold, tactile stimuli to the anterior faucial pillars</td>
</tr>
<tr>
<td>Shaker exercise</td>
<td>An activity intended to improve opening of the upper esophageal sphincter by increasing the strength of certain muscle groups that contribute to sphincter opening; the patient lies supine and raises the head (not the shoulders) sufficiently to see the toes</td>
</tr>
</tbody>
</table>

D. Behavioral Adjustments

Behavioral adjustments are often implemented in swallowing therapy. Certain patients may need to eat at a slower rate, take smaller bites or sips, or be prohibited from using a straw in order to eat and drink without aspirating. Patients and caregivers are always educated about aspiration precautions and risks of noncompliance. For patients who present with a severe aspiration risk, an alternative source of nutrition may be necessary. A nasogastric tube provides
a temporary source of nonoral nutrition, while a percutaneous endoscopic gastrostomy tube is indicated for patients who are to receive nothing by mouth and have a poor prognosis for resuming eating in the near future.


Musculoskeletal Ultrasound

Paul Lento, MD
Edward Rosero, DO

Since the development of high-frequency transducers in the 1980s, physicians and ultrasonographers have been utilizing musculoskeletal ultrasound to provide detailed imaging of anatomic structures. Today, many practitioners are incorporating musculoskeletal ultrasound as a useful tool to help evaluate and treat their patients. As the popularity of musculoskeletal ultrasound continues to increase, a better understanding of its capabilities will be required.

This chapter surveys essential knowledge about musculoskeletal ultrasound, including the indications for its use; advantages and disadvantages of this technique over other imaging modalities; and the basic physics of ultrasound. The appearance of normal musculoskeletal tissues is reviewed and illustrated using numerous images of normal and pathologic structures evaluated through diagnostic musculoskeletal ultrasound examination.

INDICATIONS

After obtaining a detailed history and performing a comprehensive physical examination, the physician may determine that further diagnostic workup is required to identify the source of a patient’s dysfunction. A musculoskeletal ultra-sound examination can provide high-resolution scans of various anatomic structures, including tendons, ligaments, nerves, joint capsules and muscles. Consequently ultrasound can be used to diagnose tendon pathology, muscle injury, ligament damage, and joint effusions. In addition, ultrasound can be utilized to evaluate other structures throughout the musculoskeletal system while helping to guide interventional procedures in real time.

ADVANTAGES
ADVANTAGES

Currently a vast array of modalities is available that can provide imaging of the human body. Musculoskeletal ultrasound offers certain advantages over other forms of imaging such as radiography, computed tomography (CT), and magnetic resonance imaging (MRI).

Ultrasound can be used to provide real-time, high-resolution images that require no preparation aside from body positioning. Unlike MRI or CT, ultrasound can be utilized to provide a dynamic and interactive examination. Apart from its ability to provide static imaging, ultrasound allows the physician to investigate a patient’s dynamic complaints such as “snapping” or “popping,” which can be evaluated while performing provocative maneuvers.

In addition, in response to patient feedback, the ultrasound beam can be targeted over involved areas of tenderness, which may help correlate patient symptoms with abnormalities identified on the ultrasound scan. In comparison, MRI and CT may identify many abnormalities that have no clinical relevance to the patient’s symptoms. Furthermore, utilizing diagnostic musculoskeletal ultrasound, structures can be compared with the contralateral limb. Musculoskeletal ultrasound images of many soft tissues typically have greater resolution than those provided by MRI or CT.

Musculoskeletal ultrasound is also relatively safe and inexpensive to use on children, individuals with pacemakers, and pregnant women, since it emits no radiation and lacks a high-powered magnet. In addition, in comparison with CT, MRI, and radiography, ultrasound units can be portable, allowing utilization outside a hospital or radiology suite. Finally, because ultrasound can identify structures such as vessels and nerves, it can be used to safely and accurately guide musculoskeletal interventional procedures.


DISADVANTAGES

Although musculoskeletal ultrasound is a highly valuable diagnostic tool, it does
have limitations. The quality of the ultrasound examination is ultimately dependent on the skill of the examiner. The high-resolution images obtained from ultrasound provide a limited field of view, which is not ideal for a larger area of study. Ultrasound does not penetrate through bone and cannot fully evaluate some intraarticular structures. The utility of diagnostic ultrasound in obese or very muscular individuals may be limited as tissue resolution is sacrificed at greater tissue depth.


**ULTRASOUND BASICS**

An ultrasound machine has three components: a transducer, the connecting cord, and the main apparatus. The transducer contains an array of thin crystals. An ultrasound wave is produced when an electric current is applied to the crystals, which causes them to vibrate, creating a sinusoidal sound wave. This transformation of electrical energy to mechanical energy is known as the *piezoelectric effect*. Because these waves are not propagated well through air, a medium, such as water or gel, is necessary to enable them to penetrate tissue. The sound wave then travels until it encounters a change in the stiffness or density of an adjacent tissue. This difference is known as an *acoustic interface*. The acoustic interface will reflect a portion of the sound wave, while allowing some of the wave to pass through. The reflected wave reaches the transducer and produces a two-dimensional (2D) image. The conversion of mechanical energy back to electrical energy is known as the *reverse piezoelectric effect*. The greater the number of waves reflected by the acoustic interface, the brighter the image appears on the screen, and the fewer the waves reflected by the acoustic interface, the darker the image appears on the screen.


**SCANNING BASICS**

As stated earlier, the images produced by the ultrasound machine provide a
limited field of view. Images seen on the screen produce a 2D “slice” through the structure being examined. Therefore it is imperative that the examiner follow a protocol to ensure that the entire area is interrogated, examining all tissues in orthogonal planes. Musculoskeletal tissues are best described by their echotexture, which represents the internal echo pattern specific to certain tissues. The relative brightness or darkness of a structure imaged with ultra-sound is referred to as echogenicity. An image is said to be hyperechoic if it is brighter compared with another image. If an image is darker than another image, it is said to be hypoechoic. If an image appears to be absent or devoid of any image, it is described as anechoic.

Ultrasound is also associated with various artifacts. Anisotropy, a common artifact, is created when an ultrasound beam does not hit a structure at 90 degrees. As stated earlier, the ultrasound wave will travel until it hits a structure, whereupon it will be reflected back to the transducer to produce an image. The greater the portion of the wave that is reflected or the greater the interface differential, the more echogenic will be the image produced. If the wave does not hit the structure perpendicularly, only a part of the wave will be reflected back to the transducer, changing a hyperechoic image to a hypoechoic image. This creates the artifact known as anisotropy. Anisotropy occurs more commonly in tendons. To avoid anisotropy, the examiner must be careful to ensure that the transducer is oriented at 90 degrees to the targeted tissue.

There is an inverse relationship between ultrasound frequency and penetration depth. Typically, linear array high-frequency transducers produce an image with better resolution at the cost of decreased depth penetration. However, evaluation of deeper tissues may necessitate the use of a low-frequency curvilinear transducer, which provides greater depth of penetration but at the cost of lower resolution. The depth of the ultrasound image can be adjusted using the settings on the ultrasound unit. By increasing the depth, structures that lie deeper can be brought into an appropriate field of view. Equivalent to adjusting the focus on a camera, the structure being examined should be placed within the focal zone in order to enhance tissue resolution. Gain relates to the overall brightness seen on the ultrasound screen and can be increased or decreased based on subjective appearance. Time gain compensation (TGC) is similar, although the gain at various tissue depths can be adjusted appropriately. Many ultrasound machines have predefined settings for specific body parts being examined.

Connolly D, Berman L, McNally E: The use of beam angulation to overcome anisotropy when viewing human tendon with high frequency linear array
NORMAL TISSUES

Distinguishing structures seen on ultrasound can be a difficult task for novice ultrasonographers; however, most musculoskeletal tissues have a characteristic echogenic pattern when viewed in orthogonal planes. As stated earlier, it is important to describe structures seen on ultrasound in terms of echogenicity, echotexture, susceptibility to anisotropy, compressibility, and presence or absence of blood flow. The normal appearance of several soft tissues is described below and summarized in Table 39–1.

<table>
<thead>
<tr>
<th>Structure</th>
<th>Echogenicity</th>
<th>Transverse Appearance</th>
<th>Longitudinal Appearance</th>
<th>Susceptibility to Anisotropy</th>
<th>Compressible</th>
<th>Doppler Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tendons</td>
<td>Hyperechoic</td>
<td>“Broom end”</td>
<td>Fibrillar</td>
<td>High</td>
<td>No</td>
<td>Negative</td>
</tr>
<tr>
<td>Ligaments</td>
<td>Hyperechoic</td>
<td>“Broom end”</td>
<td>Fibrillar</td>
<td>High</td>
<td>No</td>
<td>Negative</td>
</tr>
<tr>
<td>Nerves</td>
<td>Mixed</td>
<td>“Honeycomb”</td>
<td>Fascicular</td>
<td>Mild</td>
<td>No</td>
<td>Negative</td>
</tr>
<tr>
<td>Muscles</td>
<td>Mixed</td>
<td>“Starry night”</td>
<td>Feathery</td>
<td>Mild</td>
<td>Yes</td>
<td>Negative</td>
</tr>
<tr>
<td>Vessels</td>
<td>Anechoic</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>Yes</td>
<td>Positive</td>
</tr>
<tr>
<td>Bone</td>
<td>Hyperechoic</td>
<td>Linear, smooth</td>
<td>Linear, smooth</td>
<td>—</td>
<td>No</td>
<td>Negative</td>
</tr>
</tbody>
</table>

N/A, not applicable.

Tendons exhibit a fibrillar pattern when imaged longitudinally and a “broom end” pattern when imaged transversely. As these tissues are typically hyperechoic and highly susceptible to anisotropy, it is important to focus the ultra-sound waves perpendicularly when imaging tendons. Tendons are not compressible and do not normally have any blood flow seen on Doppler examination. Longitudinal and transverse views of various tendons are shown in Figures 39–1 through 39–18.

▲ Figure 39–1 Longitudinal view of the supraspinatus tendon shown underneath the deltoide muscle and the bursa.
Figure 39–2 Longitudinal view of the supraspinatus tendon with an anechoic area in the middle of the tendon representing a tear.

Figure 39–3 Transverse view of the supraspinatus tendon. (BICEPS TRANS,
transverse biceps tendon.)

▲ Figure 39–4 Transverse view of the supraspinatus tendon with a hypoechoic area representing a tear.
Figure 39–5 View of the infraspinatus tendon in the longitudinal view.
Figure 39–6 Longitudinal view of the subscapularis as it inserts onto the lesser tubercle (LT).

Figure 39–7 Transverse view of the biceps tendon between the greater and lesser tubercles.
Figure 39–8 Longitudinal view of the biceps tendon insertion and the brachial vessel running with the tendon.
Figure 39–9 Comparison of common wrist flexor origins. The tendon originating from the left medial epicondyle has a mixed echotexture compared with the tendons from the right medial epicondyle.

Figure 39–10 Longitudinal view of the common extensor tendons as they originate from the lateral epicondyle.
Figure 39–11 Longitudinal view of the distal quadriceps tendon as it inserts onto the patella (PAT). The suprapatellar recess is also labeled. (FP, fat pad.)

Figure 39–12 Transverse view of the second through fourth wrist extensor
tendon compartments. (ECRB, extensor carpi radialis brevis; ECRL, extensor carpi radialis longus; EDC/EIP, extensor digitorum communis/extensor indicis proprius; EPL, extensor pollicis longus.)

Figure 39–13 Longitudinal view of the proximal patella tendon.
Figure 39–14 Longitudinal view of the distal patella tendon with the infrapatellar bursa seen deep to the patella tendon.

Figure 39–15 Longitudinal view of the distal Achilles tendon (ACH) as it
inserts onto the calcaneus.

▲ Figure 39–16 Longitudinal view of a thickened Achilles tendon (ACH) with mixed echogenicity proximal to its insertion onto the calcaneus. This is representative of a tendinopathy.

▲ Figure 39–17 Transverse view of the distal Achilles tendon (ACH) as it inserts onto the calcaneus. The retrocalcaneal (RETROCALC) bursa is identified in the image.
Figure 39–18 Short axis comparison of left and right, midportion plantar fascia (PF). Compared with the asymptomatic left side, notice the large area of hypoechochogenicity of the symptomatic right PF. This represents a tear versus chronic changes. (asx, asymptomatic; SYX, symptomatic.)


Ligaments

Ligaments have a similar appearance to tendons. When imaged longitudinally, they have the appearance of a fibrillar pattern, and when imaged in the transverse plane, a “broom end” appearance. Compared with tendon fibers, the fibers in ligaments appear less compact. When scanning tissues, ligaments can be distinguished from tendons by the fact that ligaments will be seen originating
and terminating on bony structures. Ligaments are hyperechoic and are susceptible to anisotropy. Similar to tendons, these structures normally lack any blood flow and are not compressible. Longitudinal and transverse views are shown in Figures 39–19 through 39–23.

Figure 39–19 Ulnar collateral ligament (UCL) seen connecting the distal humerus to the ulna.
Figure 39–20 Longitudinal view of the medial collateral ligament (MCL).
Figure 39–21 Transverse view of the proximal medial collateral ligament (MCL).

Figure 39–22 Transverse view comparisons of a painful left medial collateral ligament (MCL) and the right asymptomatic ligament. The left ligament is thickened with mixed echogenicity, which is representative of a partial tear. (nl, normal; pain, painful.)
Figure 39–23 Longitudinal view of the proximal lateral collateral ligament (LCL).

Nerves

Nerves have a mixed hyperechoic and hypoechoic pattern that is attributed to the fascicles and their surrounding connective tissue. In the transverse plane, nerves have a distinguishable “honeycomb” pattern. In the longitudinal plane, they have a characteristic fascicular pattern. Nerves are mildly susceptible to anisotropy, but are not compressible. Blood flow is typically not visualized within nerves. Figures 39–24 through 39–27 show several examples.

Figure 39–24 Transverse view of the ulnar nerve seen in the ulnar groove.
Figure 39–25 Transverse view of the radial head where the supinator muscle, the posterior interosseus nerve (PIN), and the superficial radial nerve can be seen (SUP RAD).

Figure 39–26 Transverse view of the ulnar nerve as it passes through Guyon’s canal, which is composed of the pisiform and the hamate bone.
Figure 39–27 Transverse view of the median nerve (MN) as it passes through the carpal tunnel. (FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus.)

Muscles

Muscles have a mixed echogenicity appearance. In the transverse plane, muscles have a “starry night” appearance. In the longitudinal plane, they exhibit a “feathery” appearance. Muscle is slightly compressible and is mildly susceptible to anisotropy. There is an absence of blood flow within muscles as visualized on ultrasound, although vasculature may be identified running with nervous tissue within fascial planes. Figure 39–28 through 39–33 show several commonly imaged muscles in contrasting views.
Figure 39–28 Transverse view of the quadriceps (QUAD) muscles. (RF, rectus femoris; VI, vastus intermedius; VL, vastus lateralis; VM, vastus medialis.)
**Figure 39–29** Transverse view of the proximal rectus femoris (RF) and the vastus intermedius (VASTUS INT) with the femur seen deep to these muscles.
**Figure 39–30** Anechoic area (***) representing a tear through the rectus femoris (RF). (VASTUS INT, vastus intermedius.)

**Figure 39–31** Transverse view of the semimembranosus (SM) with an area of mixed echotexture, representing a partial tear.
Figure 39–32 Transverse view of the posterior medial knee with an anechoic
cystic structure, marked with calipers, just lateral to the semimembranosus (SM),
representing a Baker’s cyst.


Vessels

In general, blood vessels are anechoic tubular structures that lack echotexture when visualized under ultrasound. Under Doppler examination, blood flow is readily visualized. Arteries and veins can be distinguished based on imaging features. Arteries are mildly compressible and are often seen pulsating under ultrasound examination. Veins are more readily compressible and typically have reduced flow compared with arteries. Examples of both can be seen in Figure 39–34.
**Figure 39–34** Transverse view of the medial ankle. (A, artery; FDL, flexor digitorum longus; FHL, flexor halluces longus; MED MAL, medial malleolus; PT, posterior tibialis; TN, tibial nerve; V, vein.)

**Bone**

Bone is a well-defined, linear structure that is smooth in appearance and is typically hyperechoic compared with its surrounding structures. Because most of the ultrasound beam is reflected back to the transducer, structures deep to bone cannot be visualized. On the monitor this is depicted as an area devoid of shadows beyond the bony structure. This is referred to as *acoustic shadowing*. Figures 39–35 through 39–41 present several views.
Figure 39–35 Transverse view of the proximal patella.


Figure 39–36 Longitudinal view of the medial epicondyle with common origin of wrist flexor tendons.
Figure 39–37 View of the posterior glenohumeral joint using a 3–5 MHz curvilinear transducer. (HH, humeral head.)
Figure 39–38  Coronal view of the acromioclavicular (AC) joint, depicted with the joint capsule, clavicle, and acromion.
Figure 39–39 Longitudinal view of the anterior talofibular ligament (ATFL)
as it attaches between the talus and the lateral malleolus (LAT MAL).


Figure 39–40 Longitudinal view of the proximal plantar fascia as it originates from the calcaneus (CALC).
DIAGNOSTIC MUSCULOSKELETAL ULTRASOUND IMAGING PROTOCOLS

It bears repeating that the quality of the ultrasound examination is dependent upon the skill of the examiner as well as the ability to scan the entire structure. To produce a complete and consistent examination, it is important for the examiner to develop a structured protocol whenever performing a musculoskeletal ultrasound examination. The American Institute of Ultrasound in Medicine (AIUM) is a multidisciplinary association dedicated to advancing the safe and effective use of ultrasound in medicine. In association with the American College of Radiology (ACR), the AIUM has recommended protocols for performing a musculoskeletal examination. Additionally, the European Society of Musculoskeletal Radiology has developed protocols for examining different anatomic structures; these protocols can be referenced online.

▲ Figure 39–41 Transverse view of the proximal plantar fascia as it originates from the calcaneus (CALC).

Peripheral injections are among the most common procedures performed in an outpatient musculoskeletal medicine practice. Such injections serve a valuable role in the diagnosis and treatment of peripheral joint–mediated pain. Proper training, preparation, and technique is paramount to the performance of safe and effective peripheral joint and musculo-skeletal injections.

Peripheral soft tissue or intraarticular injections commonly involve the injection of a local anesthetic in conjunction with corticosteroid. The local anesthetic is used to minimize the post-traumatic pain associated with joint or tissue needle penetration. In addition, the use of local anesthetic can serve a valuable diagnostic role during the immediate postinjection period by evaluating the injected structure as a pain generator. The use of a pain diary in conjunction with an injection can be particularly valuable in evaluating the degree and duration of pain relief obtained from the injected anesthetic. Steroids act as potent antiinflammatories; they can serve a therapeutic role in diminishing joint or tissue inflammation and can provide pain relief of longer duration. While local anesthetics generally take effect on the order of seconds to minutes, steroid injections often take several days to reach maximum efficacy.

For many injections it has become increasingly evident that surface anatomy landmarks alone are often not reliable enough for accurate needle placement, particularly in patients with suboptimal anatomy or body habitus. The use of image guidance in the form of fluoroscopy or ultrasound has become essential for the safe and accurate performance of many injection procedures, and the discussion that follows includes recommendations and technical considerations.

The first part of this chapter reviews basic principles of peripheral joint injection procedures. The second half describes individual joint and soft tissue injection techniques. Techniques for nerve blocks, neurolysis, trigger point
injections, and spinal injections, as well as the diagnosis and pathogenesis of musculoskeletal conditions, are reviewed and discussed elsewhere in this text. The reader is referred to the index for these topics. In addition, a separate chapter is dedicated to the emerging role of ultrasound in musculoskeletal medicine (see Chapter 39).

**PRINCIPLES OF PERIPHERAL JOINT INJECTION**

**Medications**

Musculoskeletal injections most often involve the injection of two classes of medications: corticosteroids and local anesthetics. The local anesthetic results in immediate post-procedure pain relief, which may be useful for diagnostic purposes, and the steroid reduces inflammatory-mediated pain with longer duration of effect. Other commonly injected substances for musculoskeletal disorders include hyaluronic acid and, more recently, autologous platelet-rich plasma (PRP) or conditioned serum (ACS).

**A. Local Anesthetics**

Local anesthetics act by reversibly blocking the axon sodium channels, preventing sodium ion influx and action potential generation. The degree and duration of neural blockade is dependent on the volume, concentration, and proximity to the targeted nerve. The addition of a sympathomimetic such as epinephrine reverses the inherent vasodilatory effect of local anesthetics and in doing so decreases systemic absorption and secondarily increases concentrations, hastening onset, prolonging duration of action, and reducing toxicity. Local anesthetics typically are prepared with a pH of 5–6. The addition of sodium bicarbonate raises the pH and in doing so increases diffusion through the axon membrane and hastens onset. In the presence of inflammation, tissue pH is often lowered, thereby slowing local anesthetic onset of action. In addition, by buffering the injected solution, sodium bicarbonate reduces the burning pain associated with injection.

Local anesthetics, when administered using proper technique at reasonable doses and concentrations, are extremely safe medications that are widely used and well tolerated. Toxicity is generally associated with high volumes or concentrations or inadvertent intravascular injection. Central nervous system toxicity may cause confusion, convulsions, respiratory arrest, seizures, or death.
Other serious side effects include cardiodepression, anaphylaxis, and malignant hyperthermia. When injecting large volumes of local anesthetic, withdrawing the plunger after each milliliter to check for heme and injecting contrast under live fluoroscopy is recommended to reduce risk of intravascular injection.

The two groups of local anesthetics include the amino esters and the amino amides. The ester group has an ester linkage and is broken down rapidly by the plasma pseudo-cholinesterase. The ester anesthetics are less commonly used owing to their short half-lives, instability in solution, propensity to degrade at high temperatures, and higher rate of allergic reactions. The shorter half-lives of esters may lower risks of toxicity. The amide group has an amide linkage; these agents are hydrolyzed in the liver. Consequently, caution is advised in patients with impaired hepatic function due to decreased ability to metabolize these agents. Methyl-paraben, a preservative, is commonly used with local anesthetics and is a common allergen. Allergic reactions to amides are generally related to methyl-paraben preservatives. The use of preservative-free anesthetics is mandatory for injections in or near the epidural space or thecal sac.

Lidocaine is the most commonly used local anesthetic owing to its rapid onset, intermediate duration, and safety profile, which allows for a relatively high maximum dose in comparison to effective dose. Bupivacaine is another commonly used local anesthetic, particularly for neural blockade, because of its longer duration of action. However, bupivacaine is limited by its longer onset of action and greater cardiotoxicity. Caution is advised when performing intraarticular injection with bupivacaine, which has shown chondrotoxic effects in animal studies and human case reports. Table 40–1 reviews the pharmacokinetics associated with the most commonly injected local anesthetics.

Table 40–1 Pharmacokinetics of local anesthetics.
B. Corticosteroids

Corticosteroids all demonstrate some degree of glucocorticoid as well as mineralocorticoid activity. Antiinflammatory and immunosuppressive effects are mediated by the glucocorticoids whereas salt and water balance is affected by mineralocorticoids. The corticosteroids used in musculo-skeletal injections have primarily glucocorticoid effects. Corticosteroids are commonly mixed with local anesthetics for musculoskeletal injections. The use of local anesthetics in conjunction with the injected steroid serves dual roles, diluting the steroid to permit greater spread of injectate across inflamed structures, as well as aiding in diagnosis.

Table 40–2 describes the common indications for glucocorticosteroid injections. The degree and duration of pain relief depend on many factors; in general, greater improvement is seen in inflammatory diseases as compared with degenerative diseases. Short-acting corticosteroids are rarely used for intraarticular injection as they are rapidly metabolized by the hypervascularized synovium of inflamed joints. Typically, systemic absorption following injection of intermediate- to long-acting corticosteroids begins at 48 hours and continues for several weeks. The most commonly used corticosteroids include methylprednisolone, triamcino-lone, betamethasone, dexamethasone, and hydrocortisone. Glucocorticosteroids and their potencies are compared with hydrocortisone in Table 40–3. Dexamethasone and betamethasone demonstrate the greatest degree of glucocorticoid (antiinflammatory) activity, whereas hydrocortisone demonstrates the greatest degree of mineralocorticoid properties.
Table 40–2 Indications for glucocorticosteroid injections.

<table>
<thead>
<tr>
<th>Intraarticular</th>
<th>Nonarticular</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteoarthritis</td>
<td>Trigger finger</td>
</tr>
<tr>
<td>Rheumatoid arthritis (adult and juvenile)</td>
<td>Tenosynovitis</td>
</tr>
<tr>
<td>Crystalline arthropathies (gout and pseudogout)</td>
<td>Ganglion cyst</td>
</tr>
<tr>
<td>Systemic lupus erythematosus and mixed connective tissue disorder</td>
<td>Epicondylitis</td>
</tr>
<tr>
<td>Shoulder periarthritis (adhesive capsulitis)</td>
<td>Tendonitis</td>
</tr>
<tr>
<td>Seronegative spondyloarthropathies</td>
<td>Entrapment neuropathies</td>
</tr>
<tr>
<td>Axial spine pain</td>
<td>Bursitis</td>
</tr>
<tr>
<td></td>
<td>Plantar fasciitis</td>
</tr>
<tr>
<td></td>
<td>Neuromas</td>
</tr>
<tr>
<td></td>
<td>Radiculopathy</td>
</tr>
<tr>
<td></td>
<td>Trigger points</td>
</tr>
<tr>
<td></td>
<td>Sympathetic-mediated disorders</td>
</tr>
</tbody>
</table>

Table 40–3 Glucocorticosteroid pharmacokinetics.

<table>
<thead>
<tr>
<th>Glucocorticosteroid Type</th>
<th>Relative Potency</th>
<th>Duration of Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hydrocortisone</td>
<td>1</td>
<td>Short</td>
</tr>
<tr>
<td>Prednisolone</td>
<td>4</td>
<td>Intermediate</td>
</tr>
<tr>
<td>Methylprednisolone acetate</td>
<td>5</td>
<td>Intermediate</td>
</tr>
<tr>
<td>Triamcinolone</td>
<td>5</td>
<td>Intermediate</td>
</tr>
<tr>
<td>Betamethasone</td>
<td>25</td>
<td>Long</td>
</tr>
<tr>
<td>Dexamethasone</td>
<td>25</td>
<td>Long</td>
</tr>
</tbody>
</table>

Adverse effects of corticosteroids include elevation in blood glucose, predisposition to local infection, osteonecrosis, tendon rupture, and postinjection pain flare. Other side effects include psychosis, facial flushing, injection site hypopigmentation, subcutaneous fat atrophy, increased appetite, dyspepsia,
malaise, and insomnia. Serious side effects may include fluid retention leading to renal or congestive heart failure. The adverse effects of corticosteroids should be carefully considered in patients with uncontrolled blood glucose, active infection, ulcerative disease, uncontrolled hyper-tension, congestive heart failure, renal failure, or preexisting psychiatric illness or mood instability. For diabetic patients, a plan for blood glucose monitoring and glycemic coverage should be in place prior to injection.

C. Hyaluronic Acid

Another option for intraarticular injection involves the use of viscosupplementation for the treatment of osteoarthritis (OA)–related pain. Viscosupplementation involves the injection of hyaluronic acid, a glycosaminoglycan composed of repeating disaccharides of glucuronic acid and N-acetylglucosamine found naturally in synovial fluid. In the United States, the injection of hyaluronic acid is approved by the Food and Drug Administration (FDA) only for the treatment of pain associated with knee OA. Currently, there is limited evidence for the use of hyaluronic acid in the treatment of hip OA, and future indications are pending.

The concentration and molecular weight of hyaluronic acid are reduced in joints with OA. This reduction may be responsible for the decreased viscosity and elasticity noted in osteoarthritic joints. Although the mechanism of action of hyaluronic acid is not known completely, it is thought that these injections help in restoring the depleted hyaluronic acid in synovial fluid and within cartilage. Interestingly, although the half-life of hyaluronic acid is only 8.8 days, the onset of effect and duration of effect are actually much longer, with peak effects in pain and function scores occurring 5–13 weeks post-treatment. Therefore, there may be other mechanisms of action.

Hyaluronic acid may have additional antiinflammatory and antinociceptive effects. Several formulations are available, ranging from low molecular weight (Hyalgan) to high molecular weight (Synvisc). A Cochrane review on viscosupplementation that compared hyaluronic acid to steroid injection identified statistically significant differences in pain at 5–13 weeks postinjection. In addition, hyaluronic acid injection was found to have fewer systemic side effects with more prolonged effects than intraarticular corticosteroids. The safety of repeated exposure to hyaluronic acid injection has been well established, and repeat cycles of injection are recommended in patients who have previously responded favorably. The most common side effect is a local mild and self-limiting inflammatory response. Cross-linking of hyaluronic acid may also be
related to a severe inflammatory response (commonly referred as pseudosepsis) that occurs within 24–72 hours of the injection, and it is important to rule out septic or acute inflammatory arthropathy.

D. Sterile Dextrose
Sterile dextrose at a concentration of 12.5–25% is a commonly used prolotherapy agent. In vitro studies on human chondrocytes and fibroblasts exposed to dextrose solution showed an increase in growth factor production. Sterile dextrose injections are generally performed with a 4-week interval between treatments and are repeated until at least 80% pain relief or plateau in clinical improvement is achieved. Postinjection antiinflammatory medications are avoided to allow the inflammatory healing phase to proceed unimpeded. There are indications from limited weak clinical trials that sterile dextrose has beneficial effects in pain relief and functional outcomes, with a good safety profile, in patients with knee OA.

E. Autologous Platelet-Rich Plasma
Autologous PRP, a concentration of human platelets in a small volume of plasma, is a regenerative form of therapy that is thought to augment tissue healing through natural healing cascades via growth factors released from the platelets. PRP has been used in the treatment of muscle, tendon, and cartilage injury. The use of PRP has been found to enhance production of hyaluronic acid and angiogenic growth factors in in-vitro studies of human synovial cells. The administration of PRP is generally performed with at least a 2-month interval between treatments and is repeated until at least 80% pain relief or plateau in clinical improvement is achieved. As with sterile dextrose injection, postinjection antiinflammatory medications are often avoided to allow the inflammatory healing phase to proceed.

F. Autologous Conditioned Serum
ACS (Orthokine, Regenokine) is an immunomodulator that has been available for use in Germany since 2003 but is not yet approved by the FDA. It is produced by removing 60 mL of a patient’s own blood, incubating the sample at 37 °C for 24 hours, and then centrifuging it to extract what is thought to be a potent interleukin-1 receptor antagonist. This substance, termed ACS, is then injected into an affected area with the goal of reducing pain and inflammation. The treatment is repeated a total of six times over 3 weeks. Limited studies thus
far have shown ACS to be an effective and well-tolerated option in the treatment of early knee OA.

### Adverse Effects

Complications following intraarticular injections include systemic toxic reactions; epinephrine reactions; allergic reaction; vasovagal response; nerve injury; infection; postinjection acute synovitis; ligament, tendon, or cartilage injury; tissue atrophy; and bleeding. Although the incidence of such complications is very low, several factors may increase patients’ risk. Patients who are receiving anticoagulant or antiplatelet agents, and those with a history of coagulopathy, are at greatest risk of hemarthrosis. Patients with diabetes mellitus are at greater risk for infection, which can lead to septic arthritis. The incidence of infection following joint injection ranges from 1 in 3000 to 1 in 50,000. Complications related to intraarticular injection are described in Table 40–4. Other complications that are unique to specific procedures are discussed in the context of each injection technique later in the chapter.

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**Table 40–4** Adverse effects of intraarticular injection.
<table>
<thead>
<tr>
<th>Complications</th>
<th>Comments</th>
</tr>
</thead>
</table>
| Systemic toxic reaction               | From accidental IV injection of local anesthetic  
  CNS effects: facial numbness or tingling, headache, restlessness, vertigo, tinnitus, slurred speech, seizures,  
  CNS depression, and coma  
  CVS effects: cardiovascular depression, prolonged PR interval, prolonged QRS  |
| Epinephrine reaction                  | Adrenaline effects: tremors, tachycardia, hypertension, and apprehension  |
| Allergic reaction                     | Para-aminobenzoic acid (PABA), a metabolite of ester anesthetics, is more likely to cause hypersensitivity reactions than amide anesthetics  |
| Vasovagal reaction                    | Secondary to physiologic and psychological factors of the procedure itself, resulting in dizziness, sweating, pallor, bradycardia, hypotension, and syncope  |
| Systemic steroid effects              | Hyperglycemia; flushing, warmth, diaphoresis of face and torso; hypothalamic–pituitary axis suppression  |
| Nerve injury                          | From an unintentional intraneurial injection  |
| Infection                             | Incidence of 1:1000–1:25,000  |
| Postinjection acute synovitis, “flare” | Occurs in 1–6%, lasting up to 48 hours; higher frequency in long-acting glucocorticosteroid preparations; treated with NSAIDs or local ice application, or both  |
| Ligament, tendon, or cartilage injury | From local effects of glucocorticosteroids; weight-bearing joints should not be injected more frequently than every 3–4 months  |
| Tissue atrophy of surrounding soft tissue and skin depigmentation | From periarticular injection or leakage of corticosteroids from the joint capsule; occurs in 5% of patients; may occur 1–6 months later  |
| Avascular bone necrosis               | Likely from systemic steroid therapy and not local intraarticular injections  |
| Bleeding                              | From accidental injury to adjacent vascular structures  |

CNS, central nervous system; CVS, cardiovascular system; NSAIDs, nonsteroidal antiinflammatory drugs.

Arthrocentesis

Arthrocentesis can be performed for diagnostic and therapeutic purposes in patients with joint effusions. By decreasing mechanical pressure associated with joint effusions and in turn reducing inflammatory mediators, joint aspiration in isolation can be a therapeutic procedure. The aspirate can be sent for fluid
analysis and cytology to differentiate infectious, hemorrhagic, inflammatory, and noninflammatory forms of arthritis. Table 40–5 provides details on characteristics of the joint aspirate in each of these four conditions. The techniques for arthrocentesis are the same as the intraarticular injection techniques described next and should precede the injection of medication into a joint with significant effusion.

***Table 40–5 Characteristics of joint aspirate.***

<table>
<thead>
<tr>
<th>Procedure Basics</th>
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</thead>
<tbody>
<tr>
<td>A. Preinjection Care</td>
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</table>

Prior to every injection procedure, informed consent should be obtained; this includes the discussion of benefits, limitations, risks, and alternatives. The patient should then be positioned to provide optimum exposure to the area of interest, and surface anatomy landmarks should be palpated and marked. Both
Patient comfort and physician comfort should be considered as well for optimal positioning. Recumbent positioning is preferred over sitting or standing positioning in the event that a patient develops a vagal response or syncopal episode as a result of the injection. Clinicians should be aware that some exquisitely sensitive patients may develop vagal responses even during informed consent or during early preparations for the injection procedure.

After landmarks are appropriately palpated, the injection site should be marked using a skin marker or skin impression. Next, the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. Evidence suggests that a chlorhexidine preparation may be more effective than alcohol or povidone–iodine. In addition, when using povidone–iodine, it is essential to allow a drying time of at least 90–120 seconds to maximize its bacteriocidal effects. In patients with low pain thresholds, topical anesthetics or vapocoolant sprays may be used to minimize pain. Prior to injection, aspiration should be performed to reduce the risk of intravascular injection.

Universal precautions and sterile technique should be used and maintained throughout the procedure. The choice of sterile versus nonsterile gloves remains physician dependent for peripheral joint and soft tissue injections. If the region to be injected is to be palpated following skin preparation, then sterile gloves should always be used. If nonsterile gloves are to be used, full sterile precautions must be maintained at the site of injection. Patients should always be queried regarding latex allergies if latex gloves or dressings are to be used.

The choice of needle length and gauge is dependent on the type of injection performed, potential need for joint aspiration, and patient’s body habitus. The choice of needle, syringe, and injectate are dependent on the structure being injected and are discussed later in the chapter.

**B. Postinjection Care**

After the injection, a sterile adhesive bandage should be applied and the patient monitored for any immediate postinjection reaction or bleeding. If local anesthetic is being injected and the injection is serving a diagnostic role, a pain diary should be provided to the patient to more objectively evaluate numerical pain scores during the immediate postinjection period. Patients should be monitored for 15–20 minutes following injection and, if treated in an outpatient setting, should be advised ahead of time to have another person come with them in the event that significant paresthesia, proprioceptive loss, or focal weakness develop that would impair their ability to drive themselves home.
Patients should be advised of the possibility of short-term symptom aggravation, consisting of postinjection acute synovitis within the first 24–48 hours, which usually resolves spontaneously. To minimize postinjection inflammation, patients are advised to apply ice to the site for 15–20 minutes two to three times a day for the first 24–48 hours and use nonsteroidal antiinflammatory drugs as needed for postinjection discomfort. Patients should be instructed that a baseline level of pain may return for a period of time as the local anesthetic wears off, before the steroid starts to act. They should further be instructed to limit usage of the joint, at least for the first 24 hours, particularly for the weight-bearing joints. Activity restriction has been shown to prevent injury and prolong the effect of the injected steroid. In addition, patients should be informed of potential “red flags” and given instructions to call the treating physician if any of the following are noted: temperature exceeding 100.3°F, erythema, drainage, or effusion.

Injection frequency should be determined based on degree and duration of response to previous injections, the nature of disease process, medical comorbidities, symptom severity, and clinical judgment. Injection of large joints should not be performed more than three to four times per year and not more than 10 times cumulatively. Injections of small joints should not be performed more than two to three times per year and not more than four times cumulatively.


Neidel J, Boehnke M, Kuster RM: The efficacy and safety of intraarticular corticosteroid therapy for coxitis in juvenile rheumatoid arthritis. Arthritis...
PERIPHERAL JOINT INJECTION

Temporomandibular Joint Injection

A. Indications
Common indications for temporomandibular joint (TMJ) injection include TMJ syndrome or arthritis. Patients usually present with pain and stiffness localized to the TMJ. The pain may also radiate to nearby facial structures, and commonly patients experience headaches in association. TMJ imaging should be obtained prior to injection.

**B. Relevant Contraindications**

There are no specific contraindications.

**C. Equipment and Medication**

*Equipment:* 27-gauge, 1-inch needle; 3-mL syringe for medication.

*Image guidance:* computed tomography (CT), fluoroscopy, or ultrasound guidance is optional.

*Medication:* 40-mg of methylprednisolone and 0.5 mL of 2% lidocaine.

**D. Patient Preparation**

The patient is placed in a supine or lateral recumbent position. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry.

**E. Technique and Relevant Anatomy**

The joint is localized below the zygomatic arch, 1–2 cm anterior to the tragus. The landmark is the inferolateral margin of the mandibular condyle. The joint is easily localized by opening and closing the mouth. The needle is inserted perpendicularly, with the tip angled slightly superiorly into the joint. Studies have demonstrated greater efficacy with ultrasound-guided approaches in children with juvenile idiopathic arthritis.

**F. Potential Complications**

The temporal artery lies posterior to the joint and should be avoided. The facial nerve may also be inadvertently blocked, causing facial muscle weakness.

UPPER LIMB INJECTION

Shoulder Joint Injections

Shoulder injections are among the most common injection procedures performed in a musculoskeletal medicine practice. Shoulder pain is often poorly localized, and physical examination findings can overlap widely among disorders. Therefore, these injections are often equally valuable for both diagnostic and therapeutic roles.

Sternoclavicular Joint Injection

A. Indications

Pain secondary to arthritis is the most common indication for intraarticular injection of the sternoclavicular (SC) joint. Other indications include post-traumatic pain associated with subluxation. OA of the SC joint is a relatively uncommon pathologic finding. The SC joint acts as a fulcrum for the shoulder and serves as an attachment site for the axial to appendicular skeleton. Patients may experience pain localized to the sternum, and pain may be increased with shoulder range of motion. The medial clavicle, manubrium of the sternum, and first rib articulate to form the SC joint, which is a true synovial joint. The joint is stabilized by the anterior and posterior SC ligaments. The SC joint may be subluxed, elevating the proximal clavicle in relation to the sternum and serving as a pain generator. Imaging should always be obtained prior to injection, and CT is the preferred imaging modality for the SC joint.

B. Relevant Contraindications

Acute trauma or dislocation is a contraindication to injection.

C. Equipment and Medication

Equipment: 25-gauge, 1.5-inch needle; 3-mL syringe for medication.
**Image guidance:** CT, fluoroscopy, or ultrasound guidance is optional.

**Medication:** 40 mg of methylprednisolone acetate or equivalent and 1 mL of 1% preservative-free lidocaine.

**D. Patient Preparation**

The patient is placed in a supine position. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. The use of a vapocoolant spray is optional.

**E. Technique and Relevant Anatomy**

The needle is inserted at the indentation of the SC joint and advanced medially at a 45-degree angle until the needle sinks into the joint, taking care not to advance too deeply.

**F. Potential Complications**

The brachiocephalic vessels are located posterior to the joint and are at risk of injury if the needle is advanced too deeply. The lungs are located posterior and lateral to the joint, and pneumothorax is an unlikely but potential risk if the needle is advanced in a direction that is too far lateral and too deep.

**Glenohumeral Joint Injection**

**A. Indications**

The most common indications for intraarticular injections of the glenohumeral (GH) joint include adhesive capsulitis, primary or secondary (post-traumatic) OA, and rheumatoid arthritis after more conservative treatment approaches have failed. GH injections can also serve a diagnostic role in evaluating intraarticular pathology as the pain generator.

Patients typically present with decreased range of motion, pain, and weakness associated with pain. There may be palpable crepitus with shoulder motion. Shoulder imaging may provide diagnostic information and is recommended prior to injection. High volumes of injectate, up to 100 mL, can be used for distention arthrography for the treatment of adhesive capsulitis using the anterior approach under fluoroscopic guidance.
B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication

*Equipment:* 22-gauge, 3.5-inch needle; 5-mL syringe for contrast with extension tubing; 10-mL syringe for medication.

*Image guidance:* fluoroscopic or sonographic guidance is recommended.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 9 mL of 1% preservative-free lidocaine. *Contrast:* up to 5 mL of iohexol.

D. Patient Preparation

The GH intraarticular injection can be performed using an anterior or a posterior approach. The anterior approach is recommended when using fluoroscopic guidance. The patient should be supine with the shoulder externally rotated to open up the joint space. Next, the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexi-dine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy

The fluoroscopic beam is generally oriented in an anterior–posterior plane. Based on the amount of shoulder external rotation, several degrees of oblique rotation may be required to maximize the joint space. The target is the junction of the middle and lower third of the GH joint. The needle should be advanced to the humeral head and then gently walked off medially into the joint. The glenoid labrum should be avoided; hence the recommendation for image guidance when utilizing the anterior approach. Intraarticular injection is confirmed with injection of enough contrast (approximately 5 mL) to confirm proper placement arthrographically and rule out intravascular injection (*Figure 40–1*). Following contrast administration, the steroid solution is administered, the needle removed, and an adhesive dressing applied.
F. Potential Complications
Complications include iatrogenic injury to the glenoid labrum, and bleeding or hematoma from unintentional injury to the axillary neurovascular bundle if the needle is placed in too medial a location.

Subacromial Shoulder Injection

A. Indications
The subacromial shoulder injection is principally indicated for treatment of subacromial bursitis, rotator cuff impingement, or tendinosis. These three
conditions typically coexist and can be difficult to differentiate individually. This approach also serves an important diagnostic role in assessing the subacromial joint space and identifying shoulder impingement as the pain generator. Subacromial shoulder injections best target the supraspinatus tendon, which passes closest through the subacromial space in comparison with the other rotator cuff tendons.

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication

*Equipment:* 22-gauge, 1.5-inch straight or 3.5-inch spinal needle (based on patient body habitus); 10-mL syringe for medication.

*Image guidance:* sonographic guidance is preferred over fluoroscopic guidance to allow visualization of soft tissue structures and is considered optional.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 5 mL of 1% preservative-free lidocaine.

D. Patient Preparation
The posterior approach is the most commonly used approach for subacromial injection, but lateral or anterior approaches can be used as well. The posterior approach has been found to have better accuracy for intraarticular needle placement as compared with the other approaches. The landmarks are more easily palpated and the joint space is wider posteriorly than anteriorly. Ultrasound-guided GH intraarticular injections have outcomes that are superior to blind injections. With the posterior approach the patient is generally in a seated position with the shoulder internally rotated, allowing the arm to hang. This positioning allows gravity to displace the humeral head from the acromion, increasing the subacromial space. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy
The posterolateral corner of the scapula is generally easily palpable and should be identified. The needle is inserted 1 cm or 1 fingerbreadth inferior to the
posterolateral angle of the acromion while directing the tip anteromedially toward the coracoid process (Figure 40–2). The needle should be aspirated prior to injection to reduce the risk of intravascular injection and to draw off any fluid from the subacromial bursa. If resistance is encountered with medication injection, the needle bevel can be rotated or the needle can be withdrawn slightly to permit a low-pressure injection.

▲ Figure 40–2 Subacromial shoulder injection using a posterior approach with needle tip insertion slightly inferior to the acromion.

F. Potential Complications
Complications include iatrogenic injury to the rotator cuff or labrum with improper technique.

▶ Acromioclavicular Joint Injection

A. Indications
Intraarticular injection of the acromioclavicular (AC) joint is indicated for treatment of pain secondary to arthritis as well as pain with grade 1 AC ligament sprain after a trial of more conservative approaches has failed. Patients
commonly present with localized pain and tenderness over the AC joint, and pain with ranges of motion, particularly crossed arm adduction. Pathologic AC joint findings may be seen on radio-graphs, CT scan, and magnetic resonance imaging (MRI).

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication

*Equipment*: 25-gauge, 1-inch needle; 3-mL syringe for medication; 3-mL syringe for contrast if fluoroscopic guidance used.

*Image guidance*: sonographic or fluoroscopic guidance is optional.

*Medication*: 1 mL of solution containing 20 mg of methylprednisolone acetate or equivalent and 1% preservative-free lidocaine.

*Contrast*: 0.5 mL of iohexol.

D. Patient Preparation
The superior approach is used to inject the AC joint. The joint has a small capacity of approximately 1 mL. The patient should be in a seated position and can hold a weight to further separate the AC joint. If fluoroscopy is used, the patient is seated with the C-arm rotated to provide an anteroposterior view to maximize the visualized joint space and assess needle depth. The accuracy of image-guided AC intraarticular injections is superior to that of blind injections. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy
The AC joint lies at the lateral margin of the clavicle. It is generally easily palpable as a distinct sulcus formed by the articulation of the clavicle with the acromion. The needle is inserted inferiorly into the sulcus formed by the lateral end of the clavicle and the medial edge of the acromion using intermittent fluoroscopy to assess needle depth. Approximately 0.5 mL of iohexol can be injected to confirm the proper location and rule out intravascular placement. The volume of contrast should be minimized because of the small joint capacity. The medication is then slowly injected. The small joint capacity may necessitate a
higher pressure injection. If significant resistance is encountered with the injection, the needle bevel should be rotated. If resistance is encountered midway through medication injection, the joint may have reached capacity.

F. Potential Complications
Pneumothorax represents a potential complication with misplaced injection.

#### Ulnohumeral Joint Injection

A. Indications
Intraarticular injection of the elbow is indicated for treating pain in rheumatoid or other inflammatory arthritis affecting the elbow after conservative therapies have failed. Patients generally present with localized pain and tenderness over the elbow and pain with range of motion.

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication

*Equipment:* 22-gauge, 1.5-inch needle; 3-mL syringe for medication.

*Image guidance:* sonographic or fluoroscopic guidance is optional.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 1% preservative-free lidocaine.

D. Patient Preparation
The patient is positioned with the elbow flexed at 50–90 degrees. The elbow is placed on a flat surface with the forearm pronated. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy
The intraarticular injection is performed between the posterior olecranon process and the lateral epicondyle of the humerus when using the posterior approach. Alternatively, using the posterolateral approach the needle is inserted medi-ally
toward the radial head in the center of triangle formed by the lateral epicondyle, the radial head, and the tip of the olecranon process. Aspiration should be performed prior to injection to rule out intravascular injection. If resistance is encountered either the joint capacity has been reached or the bevel is obstructed by joint margins. In such cases, the needle should be rotated or pulled back slightly and injection should be reattempted.

F. Potential Complications
If the injection is performed too far medially, there is a risk for intraneural injury to the radial nerve.

Lateral Epicondylitis Injection

A. Indications
Lateral epicondylitis may represent the most common elbow pathology and most common elbow injection in a musculo-skeletal practice. Lateral epicondylitis describes a syndrome of pain emanating from the common extensor tendon insert on the lateral epicondyle. Despite its name, lateral epicondylitis is rarely a true inflammatory condition and, as such, is generally a tendinopathy rather than tendinitis. The pathophysiologic process involves degenerative changes within the tendon, with fibroblastic and vascular hyperplasia. As a result, steroid injections for lateral epicondylitis are generally more effective early in the course of symptoms when a true inflammatory response exists. Other treatments, such autologous blood and PRP injections, have demonstrated efficacy in small studies.

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication
Equipment: 25-gauge, 1.5-inch needle; 3-mL syringe for medication.
Image guidance: sonographic guidance is optional.
Medication: 40 mg of methylprednisolone acetate or equivalent and 1 mL of 1% preservative-free lidocaine.
D. Patient Preparation

The patient is positioned with the elbow flexed at 50–90 degrees. The elbow is placed on a flat surface with the forearm pronated. The lateral epicondyle as well as common extensor tendon should be marked in advance and are generally easily palpable, particularly if the patient performs active wrist extension to identify the mobile tendon. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy

Once the lateral epicondyle has been demarcated and the region prepared in a sterile fashion, the needle is directed toward the lateral epicondyle at the site of attachment of the common extensor tendon. Aspiration should be performed prior to injection to rule out vascular penetration. If resistance is encountered the needle may lie within the belly of the tendon and requires repositioning.

F. Potential Complications

If the injection is performed too far medially, there is a risk of intraneural injury to the radial nerve. In addition, there is risk of tendon weakening or rupture, particularly if steroid is injected within the body of the tendon.

Medial Epicondylitis Injection

A. Indications

Medial epicondylitis, also known as “golfer’s elbow,” is a result of pathologic changes to the musculotendinous origin at the medial epicondyle. Medial epicondylitis is much less common than lateral epicondylitis. However, the same pathophysiologic process applies and is likely related to repetitive overuse of the flexor–pronator musculature, with resultant microtearing. The pronator teres and flexor carpi radialis are the two most commonly involved tendons.

B. Relevant Contraindications

There are no specific contraindications.
C. Equipment and Medication

*Equipment:* 25-gauge, 1.5-inch needle; 3-mL syringe for medication.

*Image guidance:* sonographic guidance is optional.

*Medication:* 40 mg of methylprednisolone acetate or equivalent and 1 mL of 1% preservative-free lidocaine.

D. Patient Preparation

The elbow is placed on a flat surface with the forearm fully supinated. The medial epicondyle as well as flexor–pronator tendons should be marked in advance and are generally easily palpated, particularly if the patient performs active wrist flexion to identify the mobile tendon. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy

Once the medial epicondyle has been demarcated, the needle is directed toward the medial epicondyle adjacent to the insertion of the flexor tendon. Aspiration should be performed prior to injection to rule out intravascular placement. If resistance is encountered, the needle may lie within the tendon and requires repositioning. Care must be taken to avoid injection directly into the tendon, which can irreversibly weaken it.

F. Potential Complications

If the injection is performed too far posteriorly, there is a risk for intraneural injury to the ulnar nerve. In addition there is additional risk of tendon weakening or rupture, particularly if steroid is injected within the body of the tendon.

Olecranon Bursa Injection

A. Indications

Prior to injecting for olecranon bursitis it is mandatory to rule out an infectious etiology. Nonseptic olecranon bursitis is often related to overuse but can be seen in conjunction with rheumatoid arthritis and crystalloid arthritis. Patients typically present with a swollen, enlarged, and sometimes painful olecranon
bursa sac. The role of injection is often to both drain the bursa sac and inject steroids to reduce any inflammatory-mediated pain. Studies have demonstrated a reduced risk of bursa fluid reaccumulation in conjunction with intrabursal methylprednisolone injection.

**B. Relevant Contraindications**

There are no specific contraindications.

**C. Equipment and Medication**

*Equipment:* 25-gauge, 1.5-inch needle if injecting, only; 18-or 20-gauge, 1.5-inch needle if aspirating; 10-mL syringe for aspiration; 3-mL syringe for medication.

*Image guidance:* sonographic guidance is optional.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 1% preservative-free lidocaine.

**D. Patient Preparation**

The injection can be performed with the patient seated, supine, or prone. The elbow is flexed to 90 degrees, and the olecranon bursa is generally easily palpated and demarcated when an effusion is present. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

**E. Technique and Relevant Anatomy**

Once the olecranon bursa has been demarcated, the needle is directed in a lateral to medial direction into the bursa. Using this approach reduces the risk of ulnar nerve injury associated with injection. If a large-gauge needle is to be used to attempt aspiration, the skin should be anesthetized with 1% lidocaine prior to injection. Aspiration should be performed prior to injection to rule out intravascular placement and draw off any bursa fluid. When aspirating, a “zig-zag” approach is recommended to reduce the risk of tract formation.

**F. Potential Complications**

If the injection is performed too far posteromedially, there is a risk of intraneural injury to the ulnar nerve.
First Carpometacarpal Joint Injection

A. Indications
Injection of the first carpometacarpal (CMC) joint is indicated for treating pain in OA and inflammatory arthritis after more conservative treatment approaches have failed. The first CMC joint, also known as the trapeziometacarpal joint, is the most common location of OA in the hand, frequently associated with overuse stress. Patients usually present with localized pain or tenderness at the first CMC joint and pain with activity. There is reduced grip and pinch strength secondary to pain.

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication
*Equipment:* 25-gauge, 1.5-inch needle; 3-mL syringe for medication.
*Image guidance:* fluoroscopic or sonographic guidance is optional.
*Medication:* 20 mg of methylprednisolone acetate or equivalent and 0.5 mL of 1% preservative-free lidocaine.

D. Patient Preparation
The injection is generally performed with the patient seated and the wrist pronated. The articulation between the trapezium and first metacarpal in the “snuff box” is palpated and marked. The radial artery should be palpated as well to avoid arterial puncture. To avoid puncture the injection should be performed on the ulnar side of the extensor pollicis brevis tendon. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

E. Technique and Relevant Anatomy
Once the injection site is determined and the skin prepared, the needle is directed into the joint. In arthritic joints, osteophytes are commonly encountered, making entry more difficult. Thumb traction can be applied to increase the joint space. Aspiration should be performed prior to injection to rule out vascular penetration.
and draw off any bursal fluid prior to injecting medication. Injection volume is limited due to the small capacity of the joint. If performed under fluoroscopic guidance, the volume of contrast injected should be minimized as well.

**F. Potential Complications**

Unintentional injuries to the radial artery, radial sensory nerves, extensor pollicis longus and brevis, and abductor pollicis longus are some of the potential risks of this procedure.

**De Quervain’s Tenosynovitis Injection**

**A. Indications**

De Quervain’s stenosing tenosynovitis involves the first dorsal compartment of the wrist and is commonly seen with overuse stress. Involvement of the abductor pollicis longus or extensor pollicis brevis tendons, which are responsible for radial abduction of the thumb, may occur. Patients generally complain of pain or swelling along these tendons with pain-inhibited grip strength and tenderness overlying the tendon at the radial styloid. Finkelstein’s test, which produces pain when thumb flexion is combined with ulnar deviation of the wrist, is diagnostic.

**B. Relevant Contraindications**

There are no specific contraindications.

**C. Equipment and Medication**

*Equipment:* 25-gauge, 1.5-inch needle; 3-mL syringe for medication.

*Image guidance:* sonographic guidance is optional.

*Medication:* 20 mg of methylprednisolone acetate or equivalent and 0.5 mL of 1% preservative-free lidocaine.

**D. Patient Preparation**

The injection is generally performed with the patient seated and the radial side of the wrist up. The abductor pollicis longus and extensor pollicis brevis tendons are identified and marked by passively flexing and extending the thumb. The radial artery should be palpated as well to avoid arterial puncture. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine,
chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

**E. Technique and Relevant Anatomy**

The entry site should overlie the tendon at the base of the first metacarpal. The needle is directed at a shallow angle distal to proximal along the tendon sheath toward the radial styloid. Aspiration should be performed prior to injection to reduce the risk of intravascular injection. If resistance is encountered, this may indicate needle placement within the tendon. The needle should be directed more superficially, taking care to avoid injecting within the body of the tendon.

**F. Potential Complications**

Steroid injection directly into the tendon can increase the risk of rupture.

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**Carpal Tunnel Injection**

**A. Indication**

Carpal tunnel syndrome is the most common entrapment neuropathy and results from compression of the median nerve within the carpal tunnel. It generally affects the dominant hand prior to the nondominant hand but can be present bilaterally. Carpal tunnel syndrome may be related to overuse and is commonly seen in association with OA, rheumatoid arthritis, diabetes mellitus, hypothyroidism, and pregnancy. Patients generally complain of pain, numbness, and tingling affecting the volar surface of the first three digits along with the radial half of the volar surface of the ring finger. Decreased strength or atrophy of the abductor pollicis brevis may indicate more advanced disease or axon loss. Carpal tunnel injection is indicated for the treatment of mild to moderate carpal tunnel syndrome. The injection deposits medication in the ulnar bursa proximal to the carpal tunnel.

**B. Relevant Contraindications**

There are no specific contraindications.

**C. Equipment and Medication**

*Equipment*: 27-gauge, 1-inch needle; 3-mL syringe for medication.
**Image guidance:** sonographic guidance is optional.

**Medication:** 40 mg of methylprednisolone acetate or equivalent and 0.5 mL of 1% preservative-free lidocaine.

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**D. Patient Preparation**

The injection is generally performed with the patient seated and the wrist supinated. The median nerve generally lies between the palmaris longus tendon and the flexor carpi radialis tendon. The entry site should be marked at a site proximal to the distal wrist crease and ulnar to the palmaris longus tendon. In cases where the palmaris longus tendon is absent the entry site is marked at the midpoint between the ulnar and radial styloid process. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray is optional.

**E. Technique and Relevant Anatomy**

Once the injection site is marked, the needle is advanced proximal to distal at a shallow angle of approximately 30 degrees directed toward the ring finger. The needle is aspirated to avoid intravascular injection. The solution is slowly injected. If the patient develops exacerbation of pain or median paresthesias, this may indicate nerve pressurization or intraneural placement; in such cases, the needle should be redirected or the injection discontinued.

**F. Potential Complications**

Complications include intraneural injection of the median or ulnar nerve.

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LOWER LIMB INJECTION

Hip Joint Injection

A. Indications

Pain arising from the hip joint usually is perceived in the groin or proximal anteromedial thigh region. Patients often present with pain, stiffness, and decreased range of motion, particularly internal rotation of the hip. Several pain generators can produce groin or anterior thigh pain; for this reason, intraarticular injection of the hip can serve an important diagnostic role. In addition, when a corticosteroid is included the injection can serve a therapeutic role, particularly for the treatment of pain from hip OA. Although viscosupplementation is not FDA approved for intraarticular hip injection, several studies support this
emerging indication. Hip radio-graphs should always be obtained prior to injection.

**B. Relevant Contraindications**

There are no specific contraindications.

**C. Equipment and Medication**

*Equipment:* 22-gauge, 3.5-inch spinal needle; 10-mL syringe for medication; 5-mL syringe with extension tubing for contrast.

*Image guidance:* fluoroscopic guidance is recommended.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 5 mL of 1% preservative-free lidocaine.

*Contrast:* approximately 2 mL of iohexol.

**D. Patient Preparation**

The patient is positioned supine on the fluoroscopy table with the hip in extension and external rotation. When intraarticular hip injection is performed using surface anatomy landmarks alone, without image guidance, Leopold and colleagues report an accuracy of only 60% using an anterior approach. Consequently, fluoroscopic guidance is highly recommended to minimize extraarticular injections and to minimize risk. The anterior superior iliac spine as well as the femoral artery are palpated and marked prior to injection. The needle entry site is located 2 cm below the anterior superior iliac spine and 3 cm lateral to the palpated femoral pulse at the level of the superior edge of the greater trochanter. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional.

**E. Technique and Relevant Anatomy**

Once the patient is positioned, fluoroscopy is used to verify proper injection site at the superior–lateral aspect of the femoral neck. If paresthesias are encountered, the needle should be withdrawn and redirected. The onset of paresthesias may indicate that the needle is passing near or contacting the femoral or lateral femoral cutaneous nerve. The needle should pass through the hip capsule until periosteum is contacted. The stylette should be removed and extension
tubing attached. Prior to contrast injection, the joint should be aspirated to draw off any fluid and rule out intravascular placement. Next, approximately 2 mL of contrast should be injected under live fluoroscopy for arthrographic verification (Figure 40–3). Following injection of medication, the needle should be withdrawn, the region cleansed, and an adhesive bandage applied.

\[\text{Figure 40–3} \text{ Intraarticular hip injection under fluoroscopic guidance. Contrast pattern demonstrating proper intraarticular placement.}\]

\textbf{F. Potential Complications}

Without fluoroscopic guidance there is increased risk of injuring the femoral nerve, femoral artery, or lateral femoral cutaneous nerve. Avascular necrosis of the hip has been reported after multiple intraarticular hip injections.

\textbf{Ischial Bursa Injection}
A. Indications
Injection of the ischial bursa is indicated for patients with ischial or ischiogluteal bursitis. Patients report pain, particularly when sitting on hard surfaces, and examination reveals focal tenderness overlying the ischium. Patients with hamstring insertional tendinosis can present with pain in a similar location but typically also have pain with resisted hamstring contraction at the ischium. Ischial bursitis can coexist with hamstring tendinosis, and an ischial bursa injection can serve therapeutic roles for both conditions. The injection can also serve a diagnostic role.

B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication
Equipment: 25-gauge, 3.5-inch spinal needle; 10-mL syringe for medication; 5-mL syringe with extension tubing for contrast.

Image guidance: fluoroscopic guidance is recommended.

Medication: 80 mg of methylprednisolone acetate or equivalent and 5 mL of 1% preservative-free lidocaine.

Contrast: approximately 1 mL of iohexol.

D. Patient Preparation
The patient is positioned prone on the fluoroscopy table. The point of maximal tenderness at the ischial tuberosity is demarcated. The ischial tuberosity should be easily identified with fluoroscopy. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional.

E. Technique and Relevant Anatomy
Following skin preparation and local anesthesia, a 25-gauge, 3.5-inch spinal needle is directed toward the inferior margin of the ischial tuberosity. If the patient develops pares-thesias, this may indicate that the needle is passing too close to the sciatic nerve and the needle should be withdrawn and redirected medially. The needle is advanced until periosteum is contacted. The stylette is removed and extension tubing attached. Prior to contrast injection, the joint
should be aspirated to draw off any fluid and rule out intravascular placement. Next, approximately 1 mL of contrast should be injected under live fluoroscopy for verification of bursography (Figure 40–4). If resistance is encountered, the bevel may be occluded by bone or tendon; in such cases, the needle should be withdrawn slightly and the bevel rotated. Following medication injection, the needle should be withdrawn, the region cleansed, and an adhesive bandage applied.

![Figure 40–4 Ischial bursa injection under fluoroscopic guidance.](image)

**F. Potential Complications**

Sciatic nerve injury can occur if the needle is directed too far laterally.

**Trochanteric Bursa Injection**

**A. Indications**

The greater trochanter serves as an insertion site for the hip girdle musculature. Greater trochanteric bursitis is a common source of pain involving the lateral thigh. Patients commonly describe lateral thigh pain as hip pain. However, it is important to recognize that an intraarticular source of hip pain generally is
located in the groin and proximal thigh, whereas lateral thigh pain is often related to a trochanteric bursitis or insertional tendinosis. A greater trochanteric bursa injection may be therapeutic for the treatment of either condition. The lateral thigh is a common pain referral site and, as such, greater trochanteric bursa injection can also serve an important diagnostic role.

**B. Relevant Contraindications**
There are no specific contraindications.

**C. Equipment and Medication**

*Equipment:* 25-gauge, 3.5-inch spinal needle; 10-mL syringe for medication; 5-mL syringe with extension tubing for contrast if fluoroscopy is used.

*Image guidance:* fluoroscopic guidance is optional.

*Medication:* 80 mg of methylprednisolone and 5 mL of 1% preservative-free lidocaine.

*Contrast:* approximately 1 mL of iohexol.

**D. Patient Preparation**

If performed with image guidance the patient can be placed in a prone, supine, or lateral recumbent position with the affected side up. The choice of a supine or prone approach should reflect the location of the fluoroscopy equipment, so that the affected side is opposite the equipment, facilitating physician access to the injection site. Although fluoroscopy is considered optional for trochanteric bursa injections, it is highly recommended for patients in whom the greater trochanter is not easily palpated. The point of maximal tenderness at the greater trochanter is demarcated. The skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional.

**E. Technique and Relevant Anatomy**

Following skin preparation and local anesthesia, a 25-gauge, 3.5-inch spinal needle is directed toward the greater trochanter until periosteum is contacted. The stylette is removed and extension tubing attached. Prior to contrast injection, the joint should be aspirated to rule out intravascular placement. Next, approximately 1 mL of contrast is injected under live fluoroscopy for
verification of bursography (Figure 40–5). If resistance is encountered the bevel may be occluded by bone or tendon; in such cases, the needle should be withdrawn slightly and the bevel rotated. Following medication injection, the needle should be withdrawn, the region cleansed, and an adhesive bandage applied.

▲ Figure 40–8 Greater trochanteric bursa injection under fluoroscopic guidance. Contrast pattern outlining the greater trochanteric bursa.

F. Potential Complications
There are no specific complications.

Knee Joint Injection
A. Indications
The knee is the most commonly injected joint in the body. Joint effusions are easily palpable in the knee. Intraarticular injection of the knee is most commonly performed for the treatment of pain secondary to OA. Patients generally present with pain and tenderness at the knee, which is worse with activity. There may be weakness in the quadriceps muscles from disuse. The appearance of the knee
may be of genu varum in medial compartmental arthritis and genu valgum in lateral compartmental arthritis. Flexion contracture can be seen, as well, in patients with advanced disease. Radiographs should be obtained prior to injection. The knee joint is the only site that has been approved by the FDA for injection of viscosupplementation.

B. Relevant Contraindications

There are no specific contraindications.

C. Equipment and Medication

*Equipment:* 22-gauge, 1.5-inch needle, if injecting without aspiration; 18-gauge, 1.5-inch needle, if aspirating effusion or injecting viscosupplementation; 10-mL syringe for medication.

*Image guidance:* fluoroscopic or sonographic guidance is optional.

*Medication:* 80 mg of methylprednisolone acetate or equivalent and 5 ml of 1% preservative-free lidocaine.

D. Patient Preparation

The knee joint can be accessed using suprapatellar or infrapatellar techniques. Each technique can be performed using either a medial or a lateral approach, based on the location of greatest degeneration. The suprapatellar injection should be performed with the patient supine and the knee in slight flexion. The infrapatellar approach can be performed with the patient supine and the knee fully flexed, or with the patient seated and the knee flexed to 90 degrees. Once the target site has been identified, the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional. Imaging guidance for intraarticular knee injections has been found to have superior accuracy compared with anatomic guidance.

E. Technique and Relevant Anatomy

Following skin preparation and local anesthesia, the needle can be directed into the joint. If aspiration of an effusion or injection of viscosupplementation is to be performed, an 18- or 20-gauge needle should be used with a suprapatellar approach. If only steroids will be injected, a 22-gauge needle can be used to
minimize tissue trauma and associated pain.

For the suprapatellar approach, the superior lateral or medial corner of the patella should be palpated and displaced slightly to avoid needle contact with the patella upon entry. For the lateral suprapatellar approach, the needle is inserted at an angle of 45 degrees and directed medially 1 cm superior and lateral to the patella. With the medial suprapatellar approach, the needle is inserted laterally 1 cm superior and medial to the patella and directed laterally at an angle of 45 degrees.

Although the infrapatellar approach may allow easier access to the joint in patients with advanced OA, there is a higher risk of meniscal and articular cartilage injury. When using an infrapatellar approach, the inferior medial or inferior lateral corner of the patella should be palpated. The needle is placed within the triangle formed by the tibial plateau, patellar tendon, and femoral condyle and directed toward the intercondylar notch.

After the needle is inserted into the joint, a 10-mL syringe can be used to aspirate any joint effusion and rule out intravascular injection. If there is any concern for infection, inflammatory arthritis, or crystalline arthritis, the fluid should be sent for analysis. The medication can then be injected and should flow without significant resistance. If resistance is encountered, the needle should be withdrawn slightly or the bevel rotated 180 degrees. The needle is then removed, the site cleansed, and an adhesive bandage applied.

F. Potential Complications
Septic arthritis is a rare but serious complication.

▶ Pes Anserine Injection

A. Indications
The pes anserine bursa is located at the proximal medial edge of the tibia and is a common source of medial knee pain. The bursa underlies the conjoined tendon formed by the sartorius, gracilis, and semitendinosus muscles. Pes anserine bursitis is a common cause of medial knee pain, can be seen with overuse or direct trauma, and is particularly common in association with knee OA, particularly in patients with genu valgus, which places additional stresses on the medial knee insertions.
B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication
*Equipment:* 25-gauge, 1.5-inch needle; 10-mL syringe for medication.
*Image guidance:* sonographic guidance is optional.
*Medication:* 40 mg of methylprednisolone acetate or equivalent and 2 mL of 1% preservative-free lidocaine.

D. Patient Preparation
The patient should be supine with the knee slightly flexed. The entry site is identified at the tendinous insertion of the conjoined tendon at the proximal medial tibia. The point of maximal tenderness is demarcated. After the target site has been identified the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional.

E. Technique and Relevant Anatomy
Following skin preparation and local anesthesia, the needle can be directed toward the pes anserine bursa. Once periosteum is contacted, the syringe should be aspirated to draw off any bursal fluid and rule out intravascular placement. The medication can then be injected. The needle is then withdrawn, the region cleansed, and an adhesive bandage applied.

F. Potential Complications
There are no specific complications.

Tibiotalar Joint Injection

A. Indications
The common indication for intraarticular injection of the tibiotalar joint is pain secondary to OA. The patient presents with pain and tenderness at the ankle, and pain that is worse with activity, especially with ankle dorsiflexion. Radiographs should be obtained prior to injection.
B. Relevant Contraindications
There are no specific contraindications.

C. Equipment and Medication

*Equipment:* 25-gauge, 1.5-inch needle; 5-mL syringe for medication; 5-mL syringe with extension tubing for contrast.

*Image guidance:* fluoroscopic or sonographic image guidance is recommended.

*Medication:* 40 mg of methylprednisolone acetate or equivalent and 4 mL of 1% preservative-free lidocaine.

*Contrast:* approximately 1–2 mL of iohexol.

D. Patient Preparation
The patient is positioned supine on the fluoroscopy table with the ankle in 45 degrees of plantar flexion. The talus is palpated with foot in neutral position to serve as a landmark. The anterior medial approach is the preferred approach as it avoids the neurovascular structures. For the anterior medial approach, a soft spot is identified and marked lateral to the medial malleolus and medial to the anterior tibial tendon and extensor hallucis tendon. After the target site has been identified, the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional. Fluoroscopic or ultrasound guidance has shown superior accuracy of injection.

E. Technique and Relevant Anatomy
The needle is inserted into the tibial joint surface at a 45-degree angle slightly laterally, superiorly, and posteriorly (Figure 40–6). After the needle is inserted into the joint, aspiration is performed to remove any joint effusion and rule out intravascular injection. Next, 1–2 mL of contrast is injected under live fluoroscopy, again to confirm proper articular placement and rule out intravascular injection. Following demonstration of proper arthrography, medication is injected, the needle removed, and an adhesive bandage applied.
F. Potential Complications
There are no specific complications.

Subtalar Joint Injection

A. Indications
Intraarticular injection of the subtalar joint is performed in patients who have pain associated with arthritis. Patients generally complain of pain within the heel that is worse with ambulation and stairs. There may be increased pain with calcaneal adduction. Radiographs should be obtained prior to injection.

B. Relevant Contraindications
There are no specific contraindications.
C. Equipment and Medication

Equipment: 25-gauge, 1.5-inch needle; 5-mL syringe for medication; 5-mL syringe with extension tubing for contrast.

Image guidance: fluoroscopic guidance is recommended.

Medication: 40-mg methylprednisolone acetate or equivalent and 2 mL of 1% preservative-free lidocaine.

Contrast: approximately 1–2 mL of iohexol.

D. Patient Preparation

The subtalar joint can be approached either medially or laterally. In the medial approach, the patient lies supine with the leg in abduction and external rotation. The medial malleolus and the sustentaculum tali (a bony protrusion that is 1 inch below the medial malleolus) are identified. The subtalar joint is slightly posterior and superior to the sustentaculum tali, where the needle is inserted perpendicularly. In the lateral approach, the patient lies prone with the foot in the neutral position. After the target site has been identified, the skin should be thoroughly cleansed with antiseptic agents (povidone–iodine, chlorhexidine, or alcohol) and allowed to dry. A fenestrated sterile drape can then be applied. The use of a vapocoolant spray or local skin anesthesia, or both, is optional. The use of ultrasound guidance provides superior accuracy in needle placement.

E. Technique and Relevant Anatomy

The needle is inserted perpendicular and just inferior to the lateral malleolus and at the midpoint between the lateral malleolus and the Achilles tendon. After the needle is inserted, the joint should be aspirated to remove any effusion and rule out intravascular injection. Next, 1–2 mL of contrast should be injected under live fluoroscopy to confirm intraarticular placement and rule out intravascular injection. Following demonstration of proper arthrography, medication is injected, the needle removed, and an adhesive bandage applied.

F. Potential Complications

There are no specific complications.


Wheelchairs & Assistive Devices

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WHEELCHAIRS

Canes, walkers, crutches, manual wheelchairs, power wheelchairs, and scooters are examples of mobility assistive equipment (MAE). Power wheelchairs and scooters are referred to as power mobility devices (PMDs). The first section of this chapter reviews the basic components of manual wheelchairs, power wheelchairs, and scooters. Included in this discussion are criteria used when prescribing a mobility device and documentation requirements. Also discussed are the types of wheelchairs used by patients who have had a stroke, spinal cord injury (SCI), or amputation; those who are obese; as well as pediatric patients with cerebral palsy.

In order to prescribe the most appropriate MAE, a physician needs to know the key elements of the patient’s medical history, past medical history, social history, current functional status, and recent changes in the medical condition. Equally vital components of the decision-making process are the cognitive status of the patient and his or her ability to use the equipment. The amount of assistance needed for transfers, activities of daily living (ADLs), and mobility within the home are extremely important, as well as whether the patient has assistance from a home health aide or family member. The physician needs to fully examine the patient and assess the range of motion and strength in all joints. Joint contractures, spasticity, amputations, joint abnormalities, and weakness affect the patient’s ability to safely transfer, ambulate, and perform ADLs.

An important factor considered by the U.S. Centers for Medicare and Medicaid Services when approving a patient for a PMD is the inability to
perform mobility-related activities of daily living (MRADLs), such as toileting, feeding, dressing, grooming, and bathing in the home using a cane, walker, or manual wheelchair. Documentation of functional mobility should include the following information:

- Mobility limitations and how they interfere with the performance of ADLs.
- Ability to use a cane or walker independently to accomplish all MRADLs in the home in a safe and timely fashion.
- Ability to use a manual wheelchair to meet mobility needs in the home.
- Changes in the patient’s condition or functional limitations that now necessitate a PMD.
- Physical and mental abilities that may prevent safe transfer into, and operation of, a PMD.
- Whether the patient is willing and motivated to use a PMD in the home.
- Whether the patient’s typical environment supports the use of a wheelchair or PMD.

**CLINICAL EVALUATION**

Before prescribing an MAE or a PMD, a physician must perform a face-to-face examination with the patient, and the date of the examination must be documented. The face-to-face examination should include the patient’s past and present medical history, social history, functional mobility and ADL status, review of systems, and a physical examination. It should also include the weight and height of the patient and whether the patient has the ability to safely propel a manual wheelchair or safely drive a PMD. The physical examination should include documentation of cardiopulmonary, neurologic, and musculoskeletal abnormalities. Range of motion in all limbs and the presence of contractures, swollen joints, or amputations should be described.

**Patient History**

The impact of the medical history on the patient’s functional status should be highlighted. The social history should document whether the patient has a wheelchair-accessible home, especially if the patient is applying for a PMD. The number of steps required to enter the home and whether the patient has a wheelchair ramp are extremely important pieces of information.
Physical Examination

The patient’s strength and endurance should be assessed and his or her ability to perform ADLs should be documented. Head and trunk control and the ability to sit upright without support should be assessed. A gait analysis should be performed, and the ability to safely stand and transfer should be evaluated and documented. The patient’s cognitive status and ability to safely maneuver a manual wheelchair or PMD should be noted. Patients with severe cardiac or pulmonary disease who experience dyspnea on exertion or at rest may have normal muscle strength; however, they may be unable to propel a manual wheelchair secondary to shortness of breath. These patients may also be unable to safely perform MRADLs in a reasonable amount of time and may benefit from a PMD; this should be so noted in the assessment. Multiple studies have noted that propelling a manual wheelchair increases the energy expenditure in users. Some multiple sclerosis patients with increased tone and decreased coordination may be unable to safely ambulate, push a manual wheelchair, or perform MRADLs and may require a PMD to help maintain their independence. Ataxia, coordination issues, and proprioception problems should be assessed and the effect on mobility and ADLs documented. Documentation of past or present sacral decubitus ulcers and skin integrity should be noted as this information will help to determine the appropriate wheelchair cushion.

Insurance Authorization

In the United States, wheelchair home accessibility requirements vary with each insurance company. Some insurance companies do not require a wheelchair ramp. Others require a wheelchair ramp if there are steps to enter the home or apartment and will automatically deny authorization for a PMD if the patient does not have a wheelchair-accessible home. Still others require a photograph of the ramp at the patient’s place of residence and a home functional evaluation performed by a physical therapist to document the functional need for a PMD. Medicare requires adequate access between rooms, adequate maneuvering space, and surfaces appropriate to operation of a PMD. At present installation of a wheelchair ramp is not covered by most major insurance companies and is a direct cost to the patient. Patients with limited income may apply for a wheelchair ramp though federal, state, and local government resources, such as local agencies on aging (eg, Philadelphia Corporation for Aging), Maximus, or Liberty Resources.
A letter of medical necessity must be written or a functional mobility examination form completed based on the face-to-face examination. The specific documentation requirements vary with each insurance company. Some insurance companies specify a time frame for the completion of required documentation after the face-to-face examination. Medicare requires a seven-element prescription, including the beneficiary’s name, description of item that is ordered, date of completion of the face-to-face examination, pertinent diagnoses or conditions that relate to the need for the PMD, length of need, physician’s signature, and date of physician’s signature.

Medicare requires all documentation to be completed and submitted within 45 days after the face-to-face examination. Some managed care insurance companies, upon review of the initial letter of medical necessity, may request additional documentation, which must be submitted within a certain time frame (usually 15 days). In most cases if the time limit for completion of documentation has expired the patient must be reexamined and the process begun anew.


WHEELCHAIR PRESCRIPTION

Six key measurements are needed to properly fit a patient for a manual or power
wheelchair: seat width, seat depth, leg length, seat height, arm height, and back height. All measurements should be obtained with the patient in the seated position. Weight and height should also be recorded.

▶ Seat Width

To determine seat width, the widest area across the thighs or hips is measured, in inches, and 2 inches (5 cm) is then added to the number obtained. The adjusted measurement provides a 1-inch (2.5-cm) space on each side of the seat, between the hips and the armrest skirt guard, that should be sufficient to allow room for a prosthesis, brace, or coat. The examiner should be able to run both hands at the same time vertically along the side of the wheelchair seat and touch the hips and the armrest skirt guard simultaneously. The standard wheelchair seat width is 18 inches (46 cm).

A patient will have difficulty propelling a wheelchair if the seat is too wide. A wide wheelchair may prevent the patient from being able to reach the hand rims on a manual wheelchair, causing him or her to lean toward one side of the wheelchair while propelling and resulting in unequal weight distribution. Patients with poor trunk control need lateral support to maintain a proper upright posture. Insufficient lateral support from a wide wheelchair and truncal instability may lead to the development of scoliosis or contractures due to muscle imbalance. A small, narrow wheelchair seat will be uncomfortable and too tight. A narrow wheelchair seat may cause pressure along the hips and thighs from the armrest guard, which may lead, initially, to skin irritation and, with continued pressure, to a pressure ulcer. Transfers may also be difficult because it will be harder for the patient to stand up if the wheelchair seat is too tight. A small wheelchair may prohibit the use of a prosthesis, brace, or coat.

▶ Seat Depth

The seat depth is measured from the posterior buttock to the popliteal fossa, and 1–2 inches (2.5–5 cm) is then subtracted from the number obtained. The examiner should be able to insert 2–3 fingerbreadths horizontally between the edge of the seat and the popliteal fossa. The wheelchair seat should not cause pressure in the popliteal fossa. A standard wheelchair seat depth is 16, 18, or 20 inches (41, 46, or 51 cm) in adults. Longer seat depths require custom orders. If the seat is too long, the edge of the seat will press into the popliteal fossa,
causing irritation of the skin and impeding circulation. A long seat may also prevent the patient from sliding back fully into the seat. This will prevent the patient from sitting properly against the wheelchair and will result in a slumped posture. A short wheelchair seat will not provide enough support to the thigh, which will cause more pressure at the ischial tuberosity, increasing the risk of decubitus ulcer. The unusual weight distribution caused by a short wheelchair seat also makes the patient prone to falling out of the wheelchair, especially on uneven surfaces.

**Leg Length**

The leg length is measured from the knee bend at 90 degrees to the bottom of the heel. A long leg length will result in the footrest not providing support to the leg; additionally, the footrest may bump curbs and other obstacles. The bottom of the footrest is usually 2 inches (5 cm) from the floor. A short leg length will cause extreme flexion of the hip and raise the knee, causing increased pressure on the ischial tuberosity. This positioning will make the patient prone to decubitus ulcers and will lead to hip and knee flexion contractures. The thigh should be parallel to the seat cushion. A raised knee will also impede the patient’s ability to pull the wheelchair underneath a table.

**Seat Height**

The seat height is the height of the seat platform with respect to the floor. Wheelchair seats are not always level and some patients prefer to have their seat slanted toward the back to provide stability. The seat height is measured from the knee bend at 90 degrees to the bottom of the heel; 2 inches (5 cm) is then added to allow for clearance of the footrest. Next, the thickness of the wheelchair cushion when compressed is subtracted from the previously calculated number. Factors to consider include the height of the wheelchair cushion and the compressibility of the cushion. Foam wheelchair cushions vary in stiffness and density; some foam cushions will compress to half their normal height, whereas others (eg, antidecubitus cushions with a contoured nondeforming foam base) are not compressible.

A seat height that is too low will cause the patient’s feet to drag and raise the thigh and knee, causing increased pressure on the ischial tuberosity. This type of positioning makes the patient prone to decubitus ulcers and contractures at the
hip and knee. Additionally, when the seat is too low patients with poor trunk control may exhibit a slumped posture, which could lead to scoliosis. A low seat height will also impair transfers to different surfaces and make reaching for objects more difficult.

Conversely, a seat height that is too high will prevent the feet from touching the floor; consequently, some patients will not be able to use their feet to help propel the wheelchair. Patients may also have difficulty reaching the hand rims for propulsion, and transfers may be impaired. A too-high seat may impair the ability to perform ADLs, reach for objects, and pull the wheelchair underneath a table.

Arm Height

The arm height is measured from the seat to the bottom of the elbow flexed at 90 degrees, and 1 inch (2.5 cm) is then added to the number obtained. The appropriate arm height will provide proper positioning of the upper extremities and give support to the arms and shoulders. If the patient is sitting on a cushion, the arm height should be measured from the top of the cushion. A high arm height will cause the shoulders to be pushed upward, resulting in pressure and pain in the glenohumeral joint. The patient will not be able to comfortably rest the upper extremities on the armrest and the upper extremities will not have support and stability. If the arm height is too low, no support will be provided for the upper extremities. Unsupported upper extremities tend to pull the body forward, causing a slouched posture. The patient may also lean forward, which will result in poor posture, leading to kyphosis or scoliosis and compromised respirations in patients with poor trunk control. Without armrest support for the upper extremities, shoulder subluxation may worsen in stroke patients.

Back Height

The back height determination is dependent on the patient’s level of functional mobility and the type of wheelchair prescribed. The patient’s balance, coordination, arm strength, the amount of trunk support needed, and the upper extremity wheelchair propulsion ability are important factors to be considered in the back height determination. The backrest should provide support and help to maintain proper posture. For patients with good trunk control and posture, a wide variety of wheelchair back heights is available, ranging from just below the
pelvis, as in sports wheelchairs, to the midback height used on a standard wheelchair.

For a manual self-propelled wheelchair, the back height is measured from the seat to the axillary floor with the shoulder flexed at 90 degrees; 4 inches (10 cm) is then subtracted from the number obtained. The top edge of the back upholstery should be slightly below the inferior angle of the scapula. At this height the scapula and shoulder will be free for easier propulsion of the wheelchair. For patients with poor trunk control who require more back support and do not self-propel or those who plan to use a power wheelchair, the back height is measured from the seat to the shoulder. If the wheelchair is a recliner or has a tilt mechanism, a headrest is needed for head support and control. Wheelchair users with kyphosis or scoliosis will need a higher back height to provide lateral support and truncal stability to help prevent progression of the disease.

When measuring the wheelchair back height, it is important to consider the wheelchair cushion thickness because the cushion will affect the seat height and ultimately the back height. A low back height will not provide enough support for the upper trunk. In patients with poor trunk control and muscle weakness, this will lead to poor posture and the development of scoliosis or kyphosis. The patient can easily lean backward over the top of the upholstery, and this weight shift may cause the chair to tip over backward. A high back height will interfere with propelling the wheelchair, because the high back will limit the movement of the upper arm and shoulder and impair scapular mobility. The scapula may also rub against the back of the wheelchair causing irritation.

**MANUAL WHEELCHAIR TYPES & COMPONENTS**

There are several manufacturers of manual wheelchairs (ie, Pride, Invacare, Drive, Everest and Jennings, and Quickie), with inventory ranging from high-end models to low-end basic wheelchairs. Differences between models reflect the materials used in fabrication, style, design, and the various features available. However, the basic components of manual wheelchairs are similar across all brands. These components include the frame, seat, back, tires, wheels, hand rims, casters, fork and stem, axle plate, axles, headrest, armrest, leg rest, footrest, seat cushions, back cushions, and accessories (Figure 41–1). As previously noted, the standard wheelchair seat width is 18 inches (46 cm), and the standard seat depth is 16, 18, or 20 inches (41, 46, or 51 cm). A seat depth greater than 20 inches (51 cm) is a custom order.
Figure 41–1 Standard manual wheelchair. (Reproduced with permission from Sunrise Medical (US) LLC.)

Seat Types

- **Sling seat**—This is the standard option on most wheelchairs and is made of nylon or vinyl. Prolonged sitting in a wheelchair will cause the sling seat to sag.

- **Solid seat insert**—The insert is made of wood and is used to prevent sagging of the sling seat. It provides a good base of support for a wheelchair cushion. The solid seat insert may attach to the wheelchair with clips or be a separate item placed underneath a cushion.

- **Folding solid seat**—This option allows the seat to fold to the side when the wheelchair is folded, permitting easier transport.

  Stroke patients with limited mobility would benefit from a solid seat insert
and seat cushion to prevent poor posture while sitting in the wheelchair for long periods of time.

### Wheels & Tires

The components of the wheel include the type of tire, push rim, tire rim, magnesium wheel or wire spoke wheel, and the hub. A standard self-propelled adult manual wheelchair has two large wheels in the rear and two smaller wheels called casters in the front. Rear wheels are usually 24 inches (61 cm) in diameter and casters are 5 or 8 inches (13 or 20 cm) in diameter, depending on the model. Several types of tires are available for wheelchairs, depending on the activity performed.

#### A. Tires

Numerous types of tires are available in different widths and tread thicknesses to accommodate functional mobility needs. Thin, smooth tires have less resistance, are easier to propel, and are well suited for sports activities. These tires are designed for speed, maneuverability, and endurance. Thin tires with a small amount of tread are suitable for indoor use. Wide tires with thick tread are more suitable for outdoor use and provide better traction on uneven rough terrain.

#### B. Casters

Casters are the small wheels in the front of the wheelchair used for steering and maneuvering and are made of aluminum or cast plastic. Caster sizes vary from 2 inches (5 cm) to 8 inches (20 cm) in diameter. Caster tire choices include solid rubber (polyurethane), spoke, pneumatic, and semi-pneumatic. Pneumatic casters provide a more comfortable ride; however, maintenance is required. A standard manual hemi-wheelchair usually has 8-inch (20-cm) solid rubber casters because this size is more suitable for different environments and terrains. A solid rubber caster is maintenance free. Small casters are used on basketball and tennis wheelchairs and are more likely to get caught in elevator doorways and sidewalk cracks.

### Fork & Stem Assemblies

The fork and stem assembly attaches the casters (small front wheels) to the wheelchair. The lengths of the fork and stem determine the seat-to-floor height.
Some wheelchairs have adjustable forks with different positions for wheel attachment; other, standard manual wheelchairs have fixed nonadjustable forks. The stem is available in different lengths and is specific to each type of caster. The fork and stem assemblies adjust the seat tilt and determine the height from seat to floor.

**Axle Plates & Axles**

Rear wheels are attached to the wheelchair via the axle plate. Different types of axle plates are available; some are adjustable, allowing the rear wheel to move forward or backward, or the wheel to be raised or lowered, which will affect the height of the wheelchair seat.

A standard axle plate has limited adjustability, depending on the model of the wheelchair. The rear wheels may have a few inches of adjustability either forward or backward, and the seat height may have two variable positions. In a stroke patient, the wheel is placed in a higher axle position to decrease the seat-to-floor height.

**Leg Rests**

The front rigging of the wheelchair consists of the leg rest and footrest. The leg rests are used to provide support to the lower extremities, and the footrest provides support to the feet.

**Anti-Tippers**

Anti-tippers are devices that attach to manual wheelchair frames to keep patients from tipping the wheelchair over backward or forward (Figure 41–2). Anti-tippers may be fixed or removable, and can be attached to the rear of the wheelchair or to the front rigging.
Wheelchairs are prone to tip backward when patients attempt to stand up from the wheelchair or to propel it up an incline, ramp, or hill. The combination of propelling a manual wheelchair up a hill while simultaneously pushing on the hand rims transfers the center of gravity behind the rear wheels, which can cause the wheelchair to tip backward. Anti-tippers can also be attached to the front of the wheelchair to prevent the wheelchair from tipping forward. Forward tipping is less common but may occur when a patient tries to pick an item up off of the floor by bending forward. Forward tipping can also occur with any activity that causes the center of gravity and weight to shift forward over the casters.

A variety of anti-tipper styles are available to accommodate different wheelchairs. Anti-tippers are optional equipment on most manual wheelchairs.
except tilt-in-space and recliner wheelchairs. The disadvantage of anti-tippers is that they may interfere with climbing over curbs or other small obstacles. Rear anti-tippers may need to be manually turned upward when tilting the wheelchair backward to traverse curbs. When propelling the wheelchair on uneven terrain, the anti-tippers may contact the ground, preventing the user from making further progress.

## Wheel Locks

Wheel locks keep the wheelchair stationary, preventing it from moving when the patient transfers in or out of the wheelchair, and from rolling inadvertently when stopped on an inclined surface. The type of wheel lock prescribed for a patient is determined by upper extremity strength, hand grip, and functional dexterity.

## Hand Rims

Hand rims are located on the outside of the wheel and are used to propel the wheelchair. Several types are available with each wheelchair model. Upper extremity strength, hand grip, amputations, contractures, spasticity, and presence or absence of dysesthesia are some of the factors to be considered when selecting the appropriate hand rim.

## Armrests

Armrests provide support for proper positioning of the upper extremities. Several options are available. The type of armrest chosen will depend on the patient’s mobility and his or her ability to perform ADLs. Examples follow:

- **Adjustable height, full-length, removable armrest**—This type of armrest extends from the back to the front of the wheelchair, is removable, and is height-adjustable for proper positioning of the upper extremities. The length of this armrest will prevent a stroke patient from moving close to a table; however, some patients may require the longer length for stability and support during transfers.

- **Adjustable height, desk-length, removable armrest**—This armrest extends from the back of the wheelchair forward to a point about three-quarters the length of the seat. The armrest is removable and has adjustable heights. Desk-length
armrests allow the wheelchair to be positioned closer to a table or desk without removing the armrest.

• **Flip-back, non–adjustable height armrest**—This type of armrest flips backward to allow for easier transfers; however, the height is not adjustable. The armrest remains attached to the wheelchair. The flip-back feature is beneficial because some patients have difficulty retrieving wheelchair armrests from the floor once they have been removed from the wheelchair. The patient may not require an adjustable-height armrest if the arm is properly supported.

• **Flip-back, adjustable-height armrest**—This armrest flips backward for increased clearance and easier transfers, and the height is adjustable. Most patients require adjustable-height armrests for proper positioning of the upper extremities.

• **Space saver armrest (wraparound armrest)**—The armrest is curved inward and decreases the overall width of the wheelchair without limiting the seat width.

• **Elevating arm rests**—This style of armrest assists with edema control and proper positioning of the upper extremity. Several different styles are available.

• **Arm trough**—An arm trough can aid hemiplegic patients with upper extremity positioning and prevent the arm from hanging at the side of the wheelchair, which could exacerbate shoulder subluxation.

▶ **Lap Trays**

Trays are available for hemi-wheelchairs that can be used for meals or performing ADLs. Trays provide increased work surface for activities.

▶ **Seatbelt**

Most injuries among electric wheelchair users occur as a result of tipping and falling accidents. Many of these accidents occur when ascending curb cuts, ascending a 45-degree curb, or descending a 5-degree ramp. Research studies have found that leg rests and seatbelts provide restraint and reduce the risk of being ejected from the wheelchair, and increase safety when traversing common obstacles. Seatbelts provide stability and safety to the patient, and the type of seatbelt prescribed is based on patient preference and upper extremity strength.
and functional ability. Wheelchair seatbelt types include auto, airplane, and Velcro.


Wheelchair Seat Cushions

More than 40 different types of cushions are available for wheelchairs. Wheelchair cushions are designed for comfort; some also improve pressure distribution and prevent pressure ulcers, or improve pelvic stability. The type of cushion prescribed for a wheelchair user should be based on the patient’s individual needs. Some patients require only a basic cushion for comfort, because they are able to weight shift, transfer, and ambulate. Nonambulatory wheelchair-bound patients who are unable to transfer and stand independently require a more complex seat cushion that will prevent pressure ulcers and provide truncal and pelvic stability.

Basic foam cushions are used mainly for comfort. Skin-protective cushions may be made of foam, air, or gel and are recommended for patients with current skin breakdown or those at high risk for breakdown. Positioning cushions are mainly designed for patients who need support to maintain a proper sitting posture. Combination skin-protection and positioning cushions are designed for patients who have skin breakdown issues and need support to maintain proper positioning in a wheelchair.

The basic foam cushion for a wheelchair is a 2-inch (5-cm) polyfoam cushion with a cover. One example, the Jay basic cushion, is described as a soft molded foam cushion with mild contour to provide posture stability. This cushion has a nonskid bottom with a Velcro attachment and a moisture-resistant cover. This type of cushion is ideal for patients with good functional ability, who can weight shift and have a lower risk for skin breakdown.

A. Skin-Protective Cushions

Gel-filled cushions range from basic cushions to custom-modified seat cushions for patients at high risk for skin breakdown. One example, the Jay 2 custom cushion, has a firm contoured foam base with more than 2 inches (5 cm) of gel
in the gel pack, allowing it to conform to boney prominences. An accessories pack is available, which includes adductor wedges, hip guides, abductor pads, and pelvic obliquity pads. Additional gel packs can be added to the cushion to individualize it to patient needs. Some other examples of skin-protective cushions are the Invacare Matrix, a molded foam cushion with a dual layer of fluid sacs (gel), and the Matrix airflow cushion, which consists of a foam base and air flotation cells on top of the base. Many other options are also available. One example, the Pride Synergy cushion, has a molded foam base with a cutout over the sacral area and is available with the choice of either twin cell–fluid insets or viscoelastic foam inserts. Another protective cushion, the Supracor Stimulite, consists of flexible honeycomb cells, which flatten when compressed to relieve pressure. The cells are perforated to allow airflow and assist with stimulating capillary blood flow.

B. Positioning Cushions

Patients who need moderate assistance in positioning can benefit from an air cushion with multiple air cells. This type of cushion can be used with or without a contoured silicone foam base that, additionally, provides lateral and preischial contoured support. An example of this type of cushion is the Pride Synergy Spectrum Air. The Jay 2 Deep Contour is an ultra-lightweight cushion containing 3 inches (7.6 cm) of gel arranged in a tripad design covering the coccyx and both buttocks. A 2-inch (5-cm) layer of soft foam lies below the gel pad, and the base is made of precontoured foam, which provides pelvic stability and helps with positioning. A third example of a positioning cushion is the Invacare Matrix Stabilite Contour, which provides pelvic stability and leg support. The cushion is fitted with a “ThinAir” bladder to assist with pressure redistribution.

C. Combination Cushions

Many type of cushions in this group provide dual benefits by relieving pressure and maintaining the desired position of patients in a wheelchair, thus preventing deformities. An example is the Jay 2 Positioning and Pride Synergy Solution cushion, which has a contoured foam base with twin cell gel inserts in the ischial area. A modification, the Pride Synergy Solution 1 cushion, consists of a high-density contoured foam base with thin gel overlay. Finally, the Invacare Matrix Flotech cushion is made of a molded foam cushion and a dual layer of fluid sacs (gel).

Many studies have evaluated the effectiveness of different wheelchair
cushions on the prevention of sacral decubitus ulcers. One study in elderly nursing home patients found that skin-protective cushions (air, viscous fluid, and foam or gel and foam) lowered pressure ulcer incidence as compared with segmented foam cushions and should be used to help prevent skin breakdown. The inferior surface of the ischial tuberosity receives the greatest subcutaneous stress, and protection of this body part helps prevent pressure ulcers. Another research study noted that foam seat cushions up to 3.2-inch (8-cm) thick were effective in reducing subcutaneous stress to the area of the ischial tuberosity; however, increasing the thickness of the foam cushion beyond 8 cm was ineffective in reducing subcutaneous stress to the ischial tuberosity. Finally, foam inserts (viscoelastic polyurethane foam) such as those used in custom-molded wheelchair seats were found to produce lower peak interface pressures and better pressure distribution than gel inserts.


D. Backrest Cushions

Several types of backrests are available for wheelchairs and scooters. The standard backrest on a manual wheelchair is a sling back. A sling backrest is usually made of nylon or vinyl upholstery and is lightweight and foldable. This type of backrest provides no lateral support to the trunk to assist with prevention or management of trunk alignment problems. Clinical research has shown that a sling backrest may affect sitting posture and produce kyphosis and posterior tilting of the pelvis, leading to deformities of the spine and pelvis. The sling backrest is prescribed for patients with good trunk control who have the ability
to sit upright without support but is not appropriate for patients with poor trunk control.

Patients with poor trunk control may require a more rigid backrest with lateral trunk stability to help maintain proper wheelchair positioning. Rigid backrests range from basic low-cost styles that provide minimal support to semi-custom styles that provide posterior lateral pelvic stability and trunk support. Some styles have lumbar support, lateral support wedges, and posterior lateral support.

Semi-custom rigid backrest cushions are available that provide lateral support, posterior support, thoracic pads, lumbar pads, or sacral pads to accommodate different types of trunk or spinal abnormalities (Figure 41–3). Patients who sit in a wheelchair for long periods of time may also benefit from a more supportive backrest because prolonged wheelchair sitting with a sling backrest will cause the backrest to sag, especially if patients are obese.

![Figure 41–3 Jay 2 backrest with lateral support. (Reproduced with permission from Sunrise Medical (US) LLC.)](image)

Adjustable-tension backrests consist of three to eight horizontally positioned adjustable straps for the thoracic, lumbar, and sacral regions (Figure 41–4).
Tension is adjusted to the patient’s preference. This backrest provides more support than a standard sling backrest with limited lateral trunk support.

▲ **Figure 41–4** Jay precision-adjustable tension backrest. (Reproduced with permission from Sunrise Medical (US) LLC.)


**MANAGEMENT CONSIDERATIONS**

In addition to the basic standard wheelchair, several specialized wheelchairs are available, ranging from hemi-wheelchairs to heavy duty, and from sport to special-use categories, to suit the needs of specific patient populations. The prescription of a wheelchair for a patient involves the careful evaluation of the person who will use it to ensure the best match of a device with the individual’s functional abilities and planned activities. A brief survey of the main types of wheelchairs, and patients for whom they might be most beneficial, follows.

1. **Hemi-wheelchairs & Lightweight Wheelchair Options for Stroke Patients**
**A. Hemi-wheelchairs**

Stroke patients with hemiplegia or hemiparesis can benefit from the use of a hemi-wheelchair. Manual wheelchair propulsion requires the use of both upper extremities or of one arm and one leg for wheelchair maneuverability. The wheel axle on a hemi-wheelchair is set lower to the ground, which decreases the seat-to-floor height and allows the patient’s foot to touch the ground. A hemiplegic patient usually propels a manual wheelchair with one arm and one foot on the ground. The foot helps steer the direction of the wheelchair. A hemi-wheelchair can be constructed of standard aluminum or lightweight aluminum.

**B. Lightweight Wheelchairs**

Stroke patients who are unable to push a standard hemi-wheelchair might be able to push a lightweight wheelchair. A lightweight wheelchair is lighter and easier to propel than a standard wheelchair. A lightweight wheelchair is made of aluminum, and the weight of this chair varies between 24 and 36 lb (11 and 16 kg), depending on the model, manufacturer, and optional equipment. The weight capacity is up to 250 lb (113 kg). Most lightweight wheelchairs have nylon upholstery to decrease the weight of the wheelchair. Some models are customizable regarding seat width, seat depth, casters, and wheel options to fit a variety of patients with different heights, body sizes, and disabilities. Ideally, patients with significant weakness should have a wheelchair evaluation to determine whether they are will be able to push a lightweight wheelchair or an ultra-lightweight wheelchair.

**C. Ultra-Lightweight Wheelchairs**

An ultra-lightweight wheelchair (high-strength lightweight) is the lightest of all manual wheelchairs and is manufactured by most wheelchair companies. The wheelchair is made of aluminum or titanium, a higher quality of aluminum. Titanium wheelchairs are more costly and often not covered by insurance companies. Ultra-lightweight wheelchairs have a high degree of adjustability and are customizable. This type of wheelchair can be easily adjusted to fit a stroke patient who requires a lower floor-to-seat height. The weight of the wheelchair varies from 14 to 26 lb (6 to 11 kg), and the weight capacity is up to 265 lb (120 kg) in some models. The frame choice for ultra-lightweight wheelchairs includes nonfolding and folding models. Nonfolding ultra-lightweight wheelchairs are popular with wheelchair users who want increased response and maneuverability and are willing to sacrifice the ease of portability for decreased weight.
Rigid ultra-lightweight wheelchairs are lighter in weight, without the folding mechanism and additional hardware. Folding models are easier to transport in an automobile but are heavier because of the folding mechanism and hardware. Some models are appropriate for patients with severe weakness who are unable to propel a lightweight wheelchair. Others are designed for very active patients. Wheelchair users report that ultra-lightweight wheelchairs are more comfortable and have better ergonomics than lightweight wheelchairs. Medicare reimburses the cost of this wheelchair only for individuals who engage in frequent activities that cannot be performed with a standard wheelchair or a lightweight wheelchair, or patients who require a seat width, seat depth, or seat height that cannot be accommodated in a standard or a lightweight wheelchair. The home environment is also important, because increased surface resistance, such as high carpet pile, decreases wheelchair velocity. The weight of the wheelchair also decreases self-selected wheelchair velocity and increases the resistive forces.

D. One-Arm Drive Wheelchairs

Another wheelchair option for a stroke patient is a one-arm drive manual wheelchair. The one-arm drive wheelchair is designed for patients who have no functional use of the lower extremities and only one functional upper limb. A one-arm drive wheelchair may be extremely useful for a stroke patient with hemiplegia or a patient with bilateral transtibial amputations and unilateral upper extremity hemiplegia. There are two types of one-arm drive wheelchairs; one has a lever drive and the other, a double hand rim. The lever drive one-arm wheelchair has a lever mounted to the front casters. The direction of the wheelchair and all aspects of wheelchair manipulation are controlled by the single lever. Pumping the lever forward moves the wheelchair forward, and pumping it backward moves the chair backward.

The double hand rim, one-arm drive wheelchair has two hand rims on the same side of the wheelchair. The large hand rim controls the wheel on the same side, and the small hand rim controls the opposite wheel. The patient can propel the manual wheelchair and control all aspects of wheelchair manipulation with both hand rims on the same side. The scissor- or bar-type conversion mechanism used to attach the inner hand rim to the opposite side of the wheelchair makes this type of wheelchair heavier than a standard wheelchair. Folding a double hand rim, one-arm drive wheelchair is also slightly more difficult because of the conversion mechanism.
Hand Rims

A wide variety of hand rims is available to suit patient preference and needs, including the following options:

- **Chrome**—The standard option on stock manual wheelchairs.
- **Aluminum anodized**—Cold to the touch in the winter and hot in the summer; consequently, patients usually have to wear wheelchair gloves when temperatures are extreme.
- **Ultra-lite**—Of lighter weight; used on lightweight wheelchairs.
- **Plastic coated**—The plastic coating it is not temperature sensitive and provides increased grip.
- **Projections**—Hand rims are available with various protrusions designed to improve wheelchair propulsion (Figure 41–5). The projections come in different sizes and shapes (eg, knobs, bumps, or nubs). Hand rims with projections are beneficial for patients with poor hand grip who are unable to grasp a regular-sized rim. The projections are spaced evenly around the handrim in a vertical, a horizontal, or an oblique direction, and allow patients with severe hand weakness to propel the wheelchair using their palms.

![Projection Handrims](image)

▲ Figure 41–5 Hand rims with projections for patients with hand weakness. (Reproduced with permission from Sunrise Medical (US) LLC.)

- **Painted**—This choice is mainly cosmetic.
• **Natural fit**—An extra wide hand rim, which is easier to grip, and provides a nonslip surface.

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### Fork & Stem Assemblies

#### A. Standard Fork

The standard fork on most wheelchair models is usually fixed and nonadjustable. This means that the seat-to-floor height cannot be adjusted to suit a particular user’s needs.

#### B. Multi-Position Fork

In contrast, a multi-position fork has several positions for caster attachment, thereby allowing the seat-to-floor height to be individualized. This option, in a hemi-wheelchair, would be beneficial for a stroke patient.

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### Wheels

Most standard hemi-wheelchairs have magnesium (“mag”) wheels. These wheels are maintenance free; made of molded composite material (plastic, nylon, or aluminum); and usually have fewer than 10 spokes. When first introduced, the mag wheel was generally heavier in weight than the standard wire spoke wheel; however, recent innovations have produced a mag wheel that is comparable in weight to the standard wheel.

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### Tires

Standard 1-inch (2.5-cm) solid rubber or plastic (polyurethane) is the most commonly ordered tire for a hemi-wheelchair because this tire requires no maintenance and is well suited for indoor use. A solid rubber tire is heavier than a pneumatic tire, and one tire may weigh 1–2 lb (0.45–0.9 kg) more than a pneumatic tire.

Treaded pneumatic is made of rubber, has an inner tube filled with air, and provides a more cushioned ride; however, this type of tire requires maintenance. The air inner tube can be easily punctured with a nail, resulting in a flat tire. An optional zero-pressure inner tube can be inserted into the pneumatic tire to prevent the tire from going completely flat after being punctured with a nail. The
tire is more suitable outside in soft, sandy, or rough terrain. This type of tire is more difficult for a stroke patient to manage.

**Leg Rest**

Swing-away, removable, nonelevating leg rests allow for easier transfers. In the nonelevating leg rest, there is no calf pad for leg support. This type of leg rest may not be appropriate for a stroke patient with hemiplegia of the lower extremity because there is no support for the hemiplegic leg. And, because it does not allow for elevation of the leg, this leg rest would not be appropriate for a patient with lower extremity edema.

A swing-away, removable, elevating leg rests is more appropriate for a stroke patient with hemiplegia. This type of leg rest allows for easier transfers, assists with edema control of the lower extremities, and includes a calf pad to provide support to the leg.

**Footrest**

Footrests provide support to the feet. Different types are available, depending on the model of the wheelchair, and most have a nonslip surface. Some footrests have adjustable height, depth, and angles to accommodate deformities, contractures, or spasticity. Models may flip up on one side to allow for easier transfers. Others fold back under the seat or are detachable. Footrests may be made of aluminum, rubber, foam, or a composite material. The width and depth of the footrest are variable and depend on the width of the wheelchair frame and the amount of support needed for the foot. Custom footrests are available for patients with contractures or severe deformities of the feet.

Most hemi-wheelchairs have two individual footrests that fold and flip up on both sides and swing away to allow for easier transfers. These footrests are generally made of lightweight composite material. Aluminum footrests are stronger and heavier, and usually are found on heavy-duty wheelchairs or older model wheelchairs. For patients who do not wear shoes, a foam footrest is more appropriate than either aluminum or composite.

**Wheel Locks**

Several types of wheel locks are available for a hemi-wheelchair. With the push-
to-lock option, pushing the brake forward, away from the wheelchair, locks the brake. The disadvantage is that the brake may interfere with swinging the leg rest out. The pull-to-lock mechanism involves pulling the brake toward the wheelchair to lock the brake. A third option is the extension handle, essentially a long handle added to the short brake to extend the length of the brake handle, making it easier to lock or unlock the wheelchair. This is very helpful for the stroke patient who has hemiplegia and uses one arm to lock and unlock both brakes.

## Accessories

Available accessories for a hemi-wheelchair include a crutch holder; which allows the user to transport crutches while propelling the wheelchair; an oxygen tank holder, which is mounted to the rear of the wheelchair or scooter; and side guards (plastic or fabric), which prevent clothing from becoming caught in the wheel.

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### 2. Lightweight & Sports Wheelchairs for Patients with Low Spinal Cord Injuries

The standard of care for a manual wheelchair in a patient with a low-level SCI is a lightweight customizable wheelchair. However, one study found that socioeconomically disadvantaged patients were less likely to receive customizable wheelchairs. Young, active paraplegic patients with low SCIs and good trunk stability often prefer a sports-type wheelchair for mobility because of its sporty styling and ease of maneuverability ([Figure 41-6](#)).
A sports-type manual wheelchair is constructed of lightweight aluminum aircraft alloy (titanium), and the style of the wheelchair is based on the patient’s functional status and the sport to be played. Sports wheelchairs are available for basketball, tennis, and bike racing. The T-frame design, including wheels, weighs approximately 9–16 lb (4–7 kg). A sports wheelchair has effortless turning, increased speed, and less rolling resistance than a standard wheelchair. The rear wheels have cambers, which can be adjusted to tilt the wheels up to 16 degrees. The tilted wheels increase the base width of the wheelchair, allowing for faster speed and faster turns. Sports-type manual wheelchairs appeal to younger, school-aged patients who may be involved in school activities or recreational sports programs. Basic sports-type manual wheelchair models are usually approved by insurance companies; however, it is very difficult to gain approval for a high-end sports wheelchair, and other funding sources may need to be investigated.
Sports wheelchairs used for basketball or tennis have two large rear wheels (22–27 inches [56–68 cm]) and one to two small front wheels (3 inches [7.6 cm]), depending on the model and manufacturer (Figure 41–7). The small front wheels improve turning and speed, and decrease resistance. Racing wheelchairs have two large wheels (28 inches [71 cm]) in the rear and one smaller front wheel (20 inches [51 cm]).

![Image of a sports tennis wheelchair. (Reproduced with permission from Sunrise Medical (US) LLC.)](image)

**Tires**

Several types of sports wheelchair tires are available for specific activities. Thin, smooth tires have less resistance, require less energy for propulsion, and are well suited for sports activities. These tires are designed for speed, maneuverability, and endurance.

Slick pneumatic tires are designed for high speeds. A Kevlar tire, made of the same material used for bulletproof vests, is high performance and ideal for
sports. Clincher is a high-performance tire designed for basketball, tennis, and cycling. It is constructed with inserts that prevent the pneumatic tire from going completely flat after the tire has been punctured.

Fork and stem options for sports wheelchairs include a multi-position fork that can be placed in several positions for caster attachment (see earlier discussion) and quick-release caster and fork assemblies, which allow casters to be easily removed for transport. (Without this option, a wrench or pliers would be needed to remove the casters.)

### Axle Plates & Axles

Rear wheels are attached to the wheelchair by means of the axle plate. Some patients with low-level SCIs prefer the rear wheels to be angled outward, increasing the wheel camber. The wheel camber refers to the spacing between the top points of the wheel and the bottom points. Increasing the wheel camber increases the wheelbase, and thus the width of the wheelchair. It also shifts the seat lower and moves the center of gravity backward. As a result of the wider wheelbase there is improved lateral stability and turning speed, which is important in activities such as wheelchair basketball. Increasing the wheel camber results in increased rolling resistance, a decrease in wheelchair velocity, and an increase in power output. Studies have found that anterior axle positions decrease forces needed to propel the wheelchair, and posterior axle positions increase forces.

Propelling a wheelchair on high-pile carpet usually requires more force; however, the amount of force needed is decreased when the rear wheel is moved forward into an anterior axle position. Median nerve injuries have been correlated with increased propulsion frequency and a higher rate of push rim force, because increased propulsion frequency decreases median sensory nerve amplitude. Unfortunately, moving the rear axle forward also decreases stability, makings it easier to flip the wheelchair over backward. Conversely, while moving the rear wheel backward increases stability, it also increases the propulsion frequency, lowering the median sensory nerve amplitude. Both the weight of a manual wheelchair and the axle position will have an impact on the amount of force needed to propel the wheelchair; decreasing the wheelchair weight will reduce the peak force needed to propel the wheelchair regardless of axle position.

Wheelchair axles for SCI patients fall into three general groups. A quick-release axle allows easy removal of rear wheels for transport in the trunk of an
automobile. A curved axle plate has multiple adjustments for fine tuning the seat height, seat angle, and rear wheel camber, thereby allowing tires to be tilted in sports wheelchairs. Finally, an offset axle plate allows the center of gravity to be adjusted by moving the rear wheel axle either forward or backward from its original position.

Armrests

Sports wheelchairs generally do not have armrests. Although armrests provide support to the upper extremities, they increase the weight of the wheelchair, decrease freedom of motion, and reduce maneuverability. Armrests also interfere with wheelchair propulsion over long distances.

Leg Rests

A swing-away removable, nonelevating leg rest is preferable for patients in sports wheelchairs (see earlier discussion). Active patients who are paraplegics will require leg straps to secure the legs to the leg rest.

Footrest

Foam-padded footrests made of molded plastic or chrome covered in foam are frequently used in sports wheelchairs because of the reduced weight.

Wheel Locks

Active wheelchair users who wish to propel their wheelchairs quickly prefer to have the wheel lock underneath the seat to prevent hand injuries associated with quick maneuvering. In a standard wheelchair, the push–pull lock is located in front of each rear wheel. Fast, quick maneuvering, such as is needed in basketball, tennis, or racing, can cause hand injuries when using the standard push–pull lock. Among available alternative wheel lock styles is a compact version with a small scissor-type brake that is located under the seat and is usually made of plastic. The scissor-type lock does not interfere with the leg rests and allows for easier transfers. This brake is more expensive and is usually made of metal.
Accessories

Sports wheelchairs have spoke guards that cover the spokes on the wheels to prevent objects or fingers from getting caught in the wire spoke wheels.

Skin-Protective Seat Cushions

Seat cushions are important elements of sports wheelchairs, helping to prevent and minimize skin breakdown from the increased shearing force effect associated with increased mobility and pressure. Several options are available. Roho is an air-filled cushion made of neoprene rubber. This cushion requires maintenance and periodic inflation with an air pump. The Roho High-Profile (single compartment) cushion has 4-inch (5-cm) interconnected air cells, which are filled with air (Figure 41–8) by means of an inflation pump.

Figure 41–8 Roho high-profile seat cushion. (Reproduced with permission from The ROHO Group, Inc.)

3. Wheelchairs for Patients with High Spinal Cord Injuries

Patients with high-level SCIs (C5 or above), who have tetraplegia and poor trunk and head control, may benefit from a recliner wheelchair. These patients require attendant care for ADLs and are nonambulatory. A recliner wheelchair is usually prescribed for a disabled individual who is unable to sit upright for extended periods of time. The patient may also have difficulty changing positions and weight shifting. This type of wheelchair is also beneficial for patients with hip abnormalities, hip contractures, or inability to flex 90 degrees at the hip. Patients wearing a body brace with a fixed hip joint may also find this type of wheelchair useful. Patients with unstable blood pressure (eg, orthostatic hypotension) may benefit from a reclining wheelchair. The back of the wheelchair can recline either fully or partially (semi-reclining to approximately 45 degrees); however, a home health aide or family member will have to manually lower the back of the wheelchair. A reclining wheelchair is difficult to self-propel because of the placement of the wheels in a more posterior position.

The tilt-in-space wheelchair is another manual wheelchair option for a patient with a high-level SCI who is unable to transfer or ambulate (Figure 41–9). This type of wheelchair is manufactured by several companies and is available in both manual and powered models. A tilt-in-space wheelchair is frequently used for less active patients who sit in a wheelchair for long periods of time and require caregiver assistance. It is useful for patients who are prone to pressure ulcers, and those who are unable to weight shift to alleviate sacral pressure. This type of wheelchair is beneficial in patients who have poor head and trunk control, associated with risk of aspiration, and improves respiration and positioning for digestion. A standard tilt-in-space wheelchair provides pressure relief and weight shifting by tilting the entire frame backward; however, the angle of the seat and back do not change. The tilt range is −5 degrees to 50 degrees in most models; however, some tilt-in-space wheelchairs recline up to 120 degrees.
Figure 41–9 Motorized tilt-in-space wheelchair. (Reproduced with permission from Sunrise Medical (US) LLC.)

Tilting and reclining increase the seating surface area across the buttocks and back, which helps to prevent pressure ulcers. A tilt-in-space wheelchair enhances skin perfusion over the ischial tuberosities at 15, 25, and 35 degrees of tilt when combined with 120 degrees of recline. At 35 degrees of tilt, skin perfusion is also improved when combined with 100 degrees of recline as compared with an upright position. A tilt-in-space wheelchair allows patients with limited hip and knee movement secondary to contractures to be easily repositioned with minimal shearing forces. Adjusting the degree of the tilt during the day improves sitting tolerance and decreases abnormal muscles tone or reflex responses in severely disabled children. The wheelchair reduces attendant care needed for repositioning and reduces the number of transfers in and out of the wheelchair during the day. Manually operated models require attendant care while other models are electric and can be operated by the wheelchair user independently. However, the tilt-in-space wheelchair is larger, heavier, and more difficult for caregivers to maneuver. Tilt-in-space wheelchairs are available in adult and pediatric sizes. Pediatric models have adjustable seat depths to allow for a
growing child.

**Headrest**

Headrests provide support to the head and neck musculature, which is extremely important in patients with poor head and neck control. Headrests improve posture, which in turn improves breathing, especially in patients with neuromuscular diseases. They also help to prevent head and neck deformities in patients prone to contractures. Headrest cushions and pads are designed to provide support to the occipital or suboccipital region, or both. Lateral support and anterior support are also available. Various headrest pads are available with different degrees of head control.

Some headrests provide minimal support and control while others are extremely supportive and provide anterior, posterior, and lateral support (Figure 41–10). The headrests are designed to easily swing away to allow for easier transfers. Static anterior head supports, such as forehead straps, should be used when patients have very poor head control. The straps keep the head from falling forward. Foam and gel inserts are available for patients who are prone to skin breakdown. Some manufacturers (eg, Invacare and Permobil) produce headrests designed specifically for their own wheelchairs. Others (eg, Whitmyer, Freedom Designs, Otto Bock Healthcare, and Stealth Products) manufacture universal headrests that can be adapted to fit a variety of wheelchair models. These headrests are made of contoured foam and are available in several styles. The pads vary in height from 2.5 inches (6 cm) for pediatric models to 12 inches (30 cm) for adults. Different types of mounting systems are available, which can be fitted to a variety of wheelchairs and head supports.
Leg Rests

Options for a recliner wheelchair include telescoping, elevating, and removable leg rests. The leg rest extends and elevates to provide support to the legs. Tilt-in-space options include a nonelevating, removable leg rest, which is often used to accommodate lower extremity contractures. Elevating removable legrests allow for easier transfers and assist with edema control.

Skin Protective Seat Cushions

The same cushions described as appropriate for lightweight and sports wheelchairs are appropriate for patients with high-level SCIs (see earlier discussion). The wheelchair frame can be fitted with custom seat and back cushions to accommodate patient needs.
4. Wheelchairs for Amputee Patients

An amputee manual wheelchair is extremely important for patients with a bilateral below-the-knee (transtibial) amputation or bilateral above-the-knee (transfemoral) amputation. Bilateral amputation of the lower limbs shifts the body weight distribution posteriorly over the rear wheels, making a wheelchair more likely to tip over backward. To prevent backward tipping, the wheel axle in an amputee wheelchair is moved further back. Patients with one lower extremity amputation (transtibial or transfemoral) may also benefit from this type of wheelchair as it minimizes the possibility of tilting backward when traversing curbs or wheelchair ramps. Amputee wheelchairs provide limb support to a patient with a transtibial amputation, helping to keep the knee of the amputated side straight to prevent a knee flexion contracture.

5. Standard, Heavy-Duty, & Bariatric Wheelchairs for Overweight or Obese Patients

Obese patients weighing less than 250 lb (113 kg) can generally use a standard
manual wheelchair for mobility. The standard wheelchair is made of aluminum, weighs less than 36 lb (16 kg), and has a maximum weight capacity of 250 lb (113 kg). Although the standard seat width is 18 inches (46 cm), an extra-wide (20-inch [51-cm]) seat is also available. Nevertheless, excess weight in such patients increases the stress on all wheelchair components. Over time, prolonged sitting can cause the sling seat and back to sag, which provides less support to the patient, resulting in poor posture or kyphoscoliosis. Additionally, patients with severe disability who are unable to weight shift or transfer independently are prone to sacral decubiti and contractures. For these reasons, obese patients, chronic wheelchair users, and patients with severe disabilities or those who sit in a wheelchair for prolonged periods of time can benefit from a solid seat insert, seat cushion, and a back cushion to prevent upholstery sagging and potential complications.

Obese patients (> 250 lb [113 kg]) require a heavy-duty wheelchair, which is a wheelchair that is reinforced with a double axial under the seat. The weight of the wheelchair is 36–42 lb (16–19 kg) and the weight capacity for this wheelchair is 250–450 lb (113–204 kg), depending on the model. A wheelchair width greater than 22 inches (56 cm) is considered a heavy-duty wheelchair. The wheelchair seat width may be 22, 24, or 26 inches (56, 61, or 66 cm). A heavy-duty wheelchair is usually too wide to fit through bathroom doorways in older homes. Heavy-duty wheelchairs up to 26 inches (66 cm) wide can be folded easily for transport.

Patients who are morbidly obese will require a bariatric manual wheelchair. The weight capacity for such wheelchairs is usually in excess of 450 lb (204 kg), and the chairs themselves may weight more than 42 lb (19 kg), depending on the model. Some models can accommodate up to 1000 lb (454 kg). The seat of a bariatric wheelchair is usually wider than 26 inches (66 cm). Most models are reinforced with double axles and are not foldable. However, the backrest and wheels on some models can be removed, to allow for easier transport. An extra-wide doorway in a handicap-accessible living environment is usually required to accommodate this type of wheelchair.

6. Wheelchairs for Pediatric Patients with Cerebral Palsy

Pediatric patients are measured for wheelchairs in the same manner as adults. The standard pediatric wheelchair seat width is 12–18 inches (30–46 cm). Most pediatric wheelchairs have adjustable frames, which allow for changes in seat
depth and seat width to accommodate a growing child (Figure 41–11). Children with trunk weakness or instability may require a five-point harness to maintain an upright posture. Patients with trunk weakness will also require lateral thoracic support for trunk stability. These pads prevent side leaning, which can lead to scoliosis in some patients.

*Figure 41–11* Pediatric wheelchair that allows for growth of the patient. (Reproduced with permission from Sunrise Medical (US) LLC.)

Patients with spastic cerebral palsy, SCI, or other neurologic illness that result in lower extremity spasticity and increased adductor tone may benefit from an adductor pommel. The pommel is a cushion that is used to slightly abduct the thighs to assist with abduction and prevent an adductor contracture. It
is attached to the wheelchair or wheelchair cushion and is easily removed to allow for easier transfers. Hip guides are another option to help maintain proper alignment of the hips and pelvis.

Depending on the child’s age and disability, he or she may be able to self-propel; however, pediatric wheelchairs are designed to accommodate pushing by a caregiver. Most push handles have adjustable heights to accommodate the caregiver. Some pediatric wheelchair models also have an adjustable center of gravity and rear wheel axle adjustability, which can convert the wheelchair from everyday use to a sports wheelchair. Some pediatric wheelchairs are foldable; others have a rigid frame with easy-release rear wheels to allow for easier transport. Nonambulatory patients with severe disabilities may benefit from a tilt-in-space wheelchair to help alleviate pressure and prevent decubiti.

Footrest

Cerebral palsy patients with lower extremity contractures at the hip, knee, or ankle may not be able to use standard footrests and may require adjustable footrests to accommodate flexion or extension contractures. Some patients have a combination of spasticity and weakness and have difficulty keeping their feet on standard footrests.

Proper choice of footrests is important to prevent deformity and injury. Several types of footrest are suitable for pediatric wheelchairs. The standard footrest, consisting of two individual footrests, is prescribed for patients with normal range of motion and no contractures at the knee and ankle. The patient should have enough strength and ability to keep the foot on the footrest. Toe straps are available to assist with foot positioning. Angle-adjustable footrests are designed for patients with contractures of the knee and ankle. The angle of the footrest is modifiable to accommodate flexion or extension contractures. Angle-adjustable styles are available as either two individual footrests or a single one-piece footrest. The one-piece footrest is designed for more disabled wheelchair users who have difficulty keeping their feet on individual footrests. The feet can be secured in place with a foot strap if needed. Some models flip up to allow for easier transfers while other styles are detachable with and without tools. Custom-padded footboards are also available.

Gel and foam footrest covers can be used to provide comfort, support, and protection to the feet in patients who have contractures or deformities. Gel footrest covers help alleviate pressure on the feet and prevent pressure sores. The foam footrest is made of compressed foam and is beneficial for patients who do
not wear shoes.

Accessories

Accessory components for a pediatric wheelchair are similar to those previously described for an adult wheelchair.

7. Other Types of Wheelchairs

Traveler’s Wheelchair

A traveler’s wheelchair is a custodial-type wheelchair with four small wheels (Figure 41–12). The patient is unable to propel this type of wheelchair and has to be pushed by an attendant. This wheelchair is frequently used in hospitals and in some airports for transport and is available in adult and pediatric sizes. It is not as heavy as a standard wheelchair and is easily transported in the trunk of an automobile. This type of wheelchair is useful for patients who need wheelchair assistance primarily when ambulating longer distances. It is not as supportive and comfortable as a standard wheelchair. A traveler’s wheelchair has fixed full-length armrests that are bolted to the wheelchair and are not removable or adjustable. The armrest has a fixed height and extends from the back to the front of the wheelchair. The fixed armrests make transfers more difficult. A threaded axle is used on traveler’s wheelchair; the rear wheel is bolted in place and has no adjustability.
Motorized Wheelchair

Motorized wheelchairs are prescribed for patients who are unable to propel a manual wheelchair or drive a motorized scooter. To qualify for a motorized wheelchair, patients are sometimes required by their insurance companies to have a wheelchair-accessible environment. The patient’s ability to safely maneuver a motorized wheelchair should also be evaluated; some patients may need physical therapy training to learn to use a motorized wheelchair safely.
Many insurance companies also require a home physical therapy evaluation. Research shows that stroke patients with and without visuospatial neglect can learn to maneuver a motorized wheelchair safely with appropriate training. Motorized wheelchairs are available in standard and custom models; all are equipped with a rechargeable battery.

A. Controls

Normal wheelchair direction and speed is controlled through a joystick. Some severely disabled wheelchair users may not have functional use of their hands and may require alternative wheelchair control mechanisms using another part of the body. Alternative wheelchair controls include chin control, sip-and-puff, head control, speech control, tongue control, mouthpiece, or a custom-designed device.

1. Chin control—A chin-mounted joystick is used to control the direction of the wheelchair. The chin sits in a cup-shaped joystick, and various neck movements control the speed and direction of the motorized wheelchair. Good head control is required for this device.

2. Sip-and-puff control—Patients with high-level SCIs or severely disabled patients may be able to use this option (Figure 41–13). The user controls the movement of the motorized wheelchair by sipping (inhaling) or puffing (exhaling) into a pneumatic tube. Sipping causes a negative pressure on the tube, and puffing results in a positive pressure. Sharp sips and puffs change the direction and speed of the wheelchair, and low-level sips and puffs control steering.
3. Head control—This option relies on devices mounted in the headrest that are activated with head movement to control wheelchair maneuvering. This control mechanism requires good head control.

4. Speech control—Speech control systems use words to control the direction of the wheelchair. Some systems are available for navigation and can be linked to computer systems for communication. This system is frequently used by patients
with high-level SCIs (C1–C4).

5. **Tongue control**—This option utilizes a tongue touch keypad built into a dental mouthpiece that is activated by a touch of the tongue. Touching the front pad increases wheelchair speed, and touching the rear pad slows the wheelchair.

**B. Seats**

Two seat options are available for motorized wheelchairs: captain and “rehab” seating.

1. **Captain seating**—This option consists of a standard seat with contoured foam cushions covered in upholstery. This type of seating does not provide control to the trunk, pelvis, or head and is not appropriate for patients with poor head control or inability to sit upright without support.

2. **“Rehab” seating**—This option is designed for individuals who are more severely disabled, and those who may not be able to sit upright independently. The backrests and seat cushions are the same cushions used in manual wheelchairs. Some patients may require a special backrest and seat cushion to accommodate trunk and pelvic weakness, decubitus ulcers, contractures, musculoskeletal deformities, or neurologic abnormalities. A tilt-in-space feature is available on some models for patients who are unable to weight shift.

**C. Drive Mode**

Motorized wheelchairs are available with front-wheel drive, mid-wheel drive, or rear-wheel drive. Front-wheel drive wheelchairs navigate tight corners well and are good on uneven terrain and hills but have a slow driving speed (5–5½ mph [8–9 kph]). Mid-wheel drive wheelchairs are the most maneuverable in small spaces and have the smallest turning radius. They are also more stable when traversing common obstacles. Rear-wheel drive wheelchairs drive a straighter course at high speeds and have the fastest speed. However, they also have the largest turning radius and are more difficult to use in small spaces.


Standing Wheelchairs

Standing wheelchairs raise a person from a seated position to a standing position. The standing wheelchair supports the legs and trunk and allows the patient to stand and talk at eye level or reach for items in a high cabinet. A standing wheelchair can be manual or motorized; similarly, the lifting mechanism can be either manual or powered. The benefits of standing include improved circulation, improved urinary and bowel function, and increased bone density. Despite documentation in the literature of these benefits, approval for this type of wheelchair is extremely difficult to obtain from insurance companies. Medicare may cover a portion of the cost, and other funding sources should be investigated.

MOTORIZED SCOOTERS

The same measurements used to size a patient for a wheelchair are used for a scooter; however, differences in the design of scooters can affect specific sizing decisions. The seat width of a scooter is more accommodating than that of a wheelchair because the armrest width is adjustable. Scooter armrests do not have side guards to limit and confine the body. Therefore, the armrests can be moved out to enlarge the overall width, for example, providing a 1-inch (2.5-cm) space on each side of the scooter seat to accommodate a coat. This armrest adjustment allows for proper positioning of the upper extremities without prescribing a wider scooter. Patients considered for a scooter should have good lower extremity strength and the ability to keep their feet on the scooter platform. Because a scooter does not have elevating leg rests, which are useful in treating patients with significant edema of the lower extremities, it may be less appropriate than a wheelchair for such patients. Use of both upper extremities is required to drive a motorized scooter; consequently, the patient should have good arm strength, good intrinsic strength, and the ability to grip a handlebar.

Motorized scooters are prescribed for patients who need assistance in mobility and are unable to propel a manual wheelchair. The patient must have
the ability to drive a motorized scooter, which requires two arms for steering. Because the seat on a motorized scooter has limited support, the patient must have good head and trunk control, the ability to sit upright, and the ability to transfer independently. Additionally, the scooter provides no support for the low extremities, so the patient must have the ability to control movement in the legs. Patients with knee extension contractures and those who are unable to flex the knee may not be appropriate candidates for a motorized scooter.

The turning radius for a motor scooter is much larger than that for a motorized wheelchair, which makes using a scooter more difficult in a small apartment. A scooter is more compatible for outdoor use. A major complaint from patients is difficulty maneuvering a larger scooter on a city bus. The weight and height of a patient are important factors to be considered in prescribing a scooter. There are three categories of motorized scooters: portable compact, three-wheeled, and four-wheeled.

**Portable Compact Scooter**

This is a small, lightweight scooter, which can be easily broken down into smaller parts that will fit into the trunk or back seat of an automobile. It has the best turning radius of all scooters and is easy to maneuver in small, crowded areas. The weight capacity is 250 lb (113 kg). This scooter is small and compact and may not accommodate obese patients or those taller than 5 feet, 7 inches (170 cm).

**Three-Wheeled Scooter**

A three-wheeled scooter has two wheels in the back and one articulating wheel in the front (Figure 41–14). It is heavier and more durable than a portable scooter, with more legroom, which may better accommodate a taller patient. The ride is more comfortable, and the turning radius is comparable to that of a portable scooter. This scooter is well suited for indoor use; when used outdoors it functions best on smooth, paved surfaces.
A four-wheeled scooter has two wheels in back and two wheels in front, which provides more stability than a three-wheeled model. It is bigger, heavier, faster, more reliable, and provides a more comfortable ride than other models, and is stable outdoors on smooth or uneven terrain. However, the turning radius and maneuverability of the four-wheeled scooter is not as good as that of either a portable or a three-wheeled scooter.

**ASSISTIVE DEVICES**

Assistive devices, such as canes, walkers, and crutches, can be used to aid in balance and stability, pain relief, fatigue, and offloading of extremities during weight bearing. When selecting an appropriate assistive device for a patient, it is important to clarify the reason for using the device. It is also vital to know of any specific weight-bearing restrictions, musculoskeletal precautions, or

▲ Figure 41–14 Three-wheeled scooter. (Reproduced with permission from Pride Mobility Corp.)
environmental barriers.

**DETERMINATION OF WEIGHT-BEARING STATUS**

Four categories of weight bearing are usually differentiated when describing functional mobility or restrictions in patients: (1) weight bearing as tolerated, (2) partial weight bearing, (3) toe-touch weight bearing, and (4) non–weight bearing. Descriptions and concerns related to each are summarized below.

▶ **“Weight Bearing As Tolerated” (WBAT)**

When patients are approved for “WBAT,” there are no specific weight-bearing restrictions. The patient is permitted to fully bear weight through the identified extremities. The patient may be unable to bear weight on an extremity due to pain, decreased range of motion, or weakness, but should be encouraged to bear as much weight as can be tolerated to as mimic normal gait as closely as possible.

▶ **“Partial Weight Bearing” (PWB)**

The patient who is restricted to “PWB” may put up to 50% of full body weight through the involved extremity, or about half as much as he or she would place on the extremity during normal weight bearing. The designation of PWB may be further clarified by identifying an area of the body that can bear the weight, for example, “PWB to left lower extremity, with heel weight bearing only.” An assistive device should be used to help offload the affected extremity.

▶ **“Toe-Touch Weight Bearing” (TTWB)**

When a patient has a restriction of “TTWB,” the affected extremity can touch down on the floor for balance purposes only but should not be used to bear any weight. An examiner should be able to place a hand under the involved extremity to ensure that no actual weight is being placed on it. Patients who have been given TTWB restrictions are often instructed to visualize having an egg underneath the involved toes. The weight transmitted though this extremity should be slight enough that the egg would not be cracked or broken.
**“Non–Weight Bearing” (NWB)**

The patient who is “NWB” cannot use the designated extremity at all for weight bearing, and the extremity must be kept off of the ground at all times. An assistive device can be utilized to assist functional mobility.

**CANES**

The primary purpose of canes is to widen the patient’s base of support, which can improve balance and redistribute weight to decrease pain or help with ambulation. Canes should not be used to offload an extremity in a patient with weight-bearing restrictions (ie, NWB, PWB, or TTWB). Canes rely on single upper extremity involvement to aid balance and share in weight distribution.

If the cane is being used solely to create a wider base of support in a patient with balance problems, it can be used in either hand. Factors influencing which hand should hold the cane include patient comfort and preference, the desire to use the nondominant hand to keep the dominant hand free, or to use the unaffected hand if there is an existing upper extremity limitation or restriction. Depending on the cause of the instability, the cane may provide better balance in one hand versus the other. A simple gait trial using each hand can help determine the benefits of using one hand instead of the other.

If the cane is being used to redistribute weight in a patient with pain or weakness, it is typically used on the side opposite to the affected side. Contralateral usage provides the opportunity to shift weight from the involved leg to the uninvolved leg and upper extremity. Different types of canes vary in the amount of support they provide. The discussion that follows describes the most common types, along with advantages and disadvantages of each.

1. **Single-Point Cane**

Single-point canes (SBCs) allow for one extra point of contact with the floor combined with weight bearing through one upper extremity (Figure 41–15). SPCs vary in make and shape. Typically they are constructed of either wood or aluminum and have a rubber tip, and a handle with a grip. The wooden cane is not adjustable and must be cut to size. Aluminum SPCs are adjustable to size. The handle may be U-shaped or have an anteriorly offset handle for greater stability.
Advantages & Disadvantages

SPCs are lightweight, easily adjustable, inexpensive, and easy for patients to use on stairs. However, they provide the least amount of stability compared with other canes.

Fitting

The cane should be adjusted for size with the patient standing up straight and the arms relaxed at the sides. The handle of the cane should be at the height of the patient’s wrist. This allows for a slight bend in the elbow when gripping the handle, which helps to absorb shock and maintain a straight posture through different phases of gait and on varying surfaces. An alternative method aligns the handle with the patient’s greater trochanter, but this method can be slightly less accurate depending on body type.
Management Considerations

A. Gait Pattern
First, the cane and the opposite or contralateral leg are moved in conjunction. Next, the ipsilateral leg steps past. The contralateral leg is usually the affected side, so by moving this leg and the cane at the same time, the patient’s weight remains centered over the unaffected leg.

B. Use on Stairs
The main goal of using any assistive device on the stairs is to prevent a fall down the stairs. Thus, the assistive device should always remain even with or lower than the patient’s body. When ascending stairs, a banister or railing should be grasped when available. This may involve moving the cane to the affected side. First, the patient should step up with the unaffected leg. Next, the patient should advance the cane and the affected leg to the same step. This pattern should be continued up the stairs.

When descending, the cane should be moved first, followed by the affected side, and then the unaffected side.

2. Quad Cane
The distinguishing characteristic of a quad cane is the use of four rubber-tipped points (feet) of floor contact instead of one (Figure 41–16). This provides a wider base of support for the cane and can increase overall stability. The feet of the cane closest to the user are positioned flush to decrease the chance of tripping. The two feet on the other side are usually splayed out to increase the base of support. The handle can typically be adjusted 180 degrees to accommodate left-handed or right-handed users. The size of the bases varies, causing the base of support to vary. Quad canes with small bases are referred to as narrow based, and those with large bases are called wide based.
Figure 41–16 A, B: Four-point stability of the quad cane.
Advantages & Disadvantages

Three design features of quad canes that provide advantages to patients with mobility issues are their adjustable height, variable bases, and a greater degree of support. Quad canes are slightly heavier than SPCs. Although they can provide greater stability at a slower gait cadence, quad canes may rock from back to front when the patient uses a quick cadence or when all four feet are not placed evenly on the floor. Depending on the size of the base, they can be awkward to use on stairs and even create a tripping hazard.

Management Considerations

A. Fitting
The height of a quad cane is determined in the same way as an SPC. An important consideration is making sure that the feet with the flatter edge are toward the patient, with the splayed feet away from the patient.

B. Use on Stairs
When using a quad cane on the stairs, it is important to ensure that all four feet of the cane are in full contact with the stair. This may be more difficult with wide-based quad canes and may require angling the cane or rotating it 90 degrees. Special consideration should be given to patients with cognitive, visual, or sensory deficits, or those with significant balance deficits.

3. Hemi-Walker
Despite inclusion of the term walker in its name, the hemi-walker is yet another variation of a four-legged unilateral assistive device. It has an inverted U-shaped handle, a crossbar for support, and four widely spaced legs. As with a quad cane, the legs closest to the patient are flush, while the two outside legs extend laterally to give an extremely large base of support. The legs are rubber tipped. In some models, the supporting crossbar has a handle grip, which the user can push on when transferring from sitting to standing. Hemi-walkers are adjustable to accommodate varying patient heights. The outer legs are usually hinged, enabling them to fold against the inner legs for transport and storage.
Advantages & Disadvantages

Hemi-walkers are very sturdy compared with other canes and have the largest base of support of all unilateral devices. This larger base of support can assist in exaggerated weight shifting toward the hemi-walker to aid ambulation. Models with a high and low handgrip can be used effectively for transfers. Those that fold can be transported and stored easily.

Because of their bulk, hemi-walkers are heavier than other canes. This results in a slower gait pattern and increased fatigue. They are also more expensive than other canes. The wide base of support on one side of the body can pose a tripping hazard to the patient and others if not properly placed. A hemi-walker should not be used on stairs because it can be a tripping hazard and cause anterior–posterior loss of balance. Consequently, a helper would be needed to bring the hemi-walker up or down the steps and to provide assistance, as necessary, to a patient who was using a railing to climb up or down.

Management Considerations

A. Fitting

As with other canes, a hemi-walker can be used for weight distribution and stability. The four legs can be independently adjusted to accommodate different patient heights. As with other devices, the upper hand grip should be even with the patient’s wrist while he or she stands straight with the arms at the sides. When folded, the outer legs will be longer than the inner legs because of the splayed leg design.

B. Use on Stairs

As noted, a hemi-walker is not designed to be used on stairs.

CRUTCHES

Crutches rely on bilateral upper extremity weight bearing to provide lower extremity offloading (Figure 41–17). They are especially useful if a patient has lower extremity weight-bearing restrictions. Crutches can also be used for balance, to redistribute weight associated with pain, and to increase endurance. As with canes, different choices of crutches are available to fit individual patient needs and preferences.
1. Axillary Crutches

Axillary crutches are typically made of wood or aluminum, and are adjustable for height. The top of the crutch is padded and fits under a patient’s axilla to stabilize the device. Although padded, the axillary portion of the crutch is not meant to be a weight-bearing surface. The padding exists to prevent rubbing or irritation of the crutches on the torso. The axilla contains sensitive structures such as nerves and blood vessels that can become stretched and damaged if exposed to repetitive weight bearing.

Axillary crutches also have padded handgrips. These grips are designed to support all upper extremity weight bearing. The handle grip heights can be adjusted to accommodate different arm lengths. Like an SPC, an axillary crutch has a single rubber tip that contacts the ground.

▶ Advantages & Disadvantages
Axillary crutches are lightweight, inexpensive, and adjustable, can be used on stairs, and can provide full offloading of a lower extremity. These crutches are also compact enough to be used in most home environments, including narrow halls and pathways.

On the other hand, because crutch walking involves simultaneously using two separate assistive devices, patients must have a requisite degree of balance, strength, and coordination. Furthermore, crutches by their nature reduce a patient’s ability to use his or her hands in functional tasks. And, as mentioned, because the crutches fit under the axilla and contact the torso, patients with a painful torso may not tolerate their use.

Management Considerations

A. Fitting

The height of crutches is adjustable, usually about 8 inches (20 cm) from the smallest to tallest height, using either compressible locking pins or a combination of screw, washer, and wing nut. For most crutches the patient heights that can be accommodated are listed, either on the packaging or on the adjustable portion of the crutch. Before fitting crutches, it is important to know the height of the patient. If he or she is near the upper or lower height limit of the crutches, it may be helpful to have the next size available to test for fit.

The height of the axillary pad on the crutch is the determining factor when fitting the height of the crutch. With the patient standing as tall as possible, arms relaxed at the side, the axillary pad should reach a point 2 inches (5 cm) below the axilla. An easy way to measure this allowance is by inserting 2 fingerbreadths between the axilla and the pad of the crutch. Crutches that are too high can compress nerves and blood supply in the axilla, while those that are too low can result in instability as the crutch pad slides away from the axilla.

The handgrips can also be adjusted to accommodate different arm lengths. Usually they are adjusted by screw, washer, and wing nut. As with canes, the hand grips should be even with the patient’s wrist when the patient stands straight with the arms relaxed at the side. This permits a slight bend of the elbow, which assists with gait cycles and uneven terrain.

B. Use on Stairs

1. Ascending stairs—Ascending stairs with axillary crutches involves upper
extremity strength and balance. The patient should start as close to the stairs as possible. The uninvolved extremity should be advanced to the next step while the crutches and involved extremity remain behind. If the involved extremity is not permitted to bear weight (ie, NWB restriction), the patient will need to hop to the next step while pressing down through the handgrips of each crutch. Next, the crutches and involved extremity follow to the same step, using the sequence of unaffected (“good”) leg, then crutches, then affected (“bad”) leg. This pattern continues up the stairs. The crutches should never be higher on the steps than the patient’s body as this can cause the patient to fall backward down the steps. They should always remain lower than or even with the body.

The presence of a sturdy handrail or banister helps ensure stability when ascending steps. In this scenario, both crutches still need to travel up the steps so that the patient can ambulate once at the top. To accomplish this, both crutches can be held together under the arm opposite the railing. As with two-crutch stair climbing, the patient’s unaffected leg should advance first, followed by the crutches and the affected side. Once at the top of the stairs, the crutches can be replaced under each arm. An alternative is to have someone help by bringing the second crutch to the top of the stairs.

2. Descending stairs—Descending the steps involves balance. The crutches should go first, followed by the affected side, and then the unaffected leg (ie, crutches, “bad” leg, “good” leg). The patient’s head and shoulders should lean slightly forward during this process to avoid posterior loss of balance. If a handrail is present, the patient can transfer both crutches under the arm opposite the railing.

2. Lofstrand Crutches

An alternative to axillary crutches is Lofstrand or forearm crutches. These crutches stop at the forearm instead of extending to the axilla. They have C-shaped cuffs or braces that encircle the forearm for stability. They also have handgrips, and one point of contact with the ground. The height of the crutch as well as the cuff portion can be adjusted for proper fit.

Advantages & Disadvantages

Forearm crutches allow the patient to release the handgrips without having to release the crutches. This allows for usage of the hands and can help conserve
energy. Lofstrand crutches can also be used effectively on stairs, and can be stored and transported more easily than axillary crutches owing to their shorter height.

Because Lofstrand crutches do not extend past the elbow, they introduce another degree of freedom compared with axillary crutches. This means that they provide less stability than axillary crutches. They may also be more expensive than axillary crutches.

Management Considerations

A. Fitting

The height of the hand grip of the Lofstrand crutch is adjusted in the same way as a cane or axillary crutch. An additional adjustment can be made to the forearm cuff so that the cuff is as proximal as possible, about an inch from the elbow.

B. Use on Stairs

Lofstrand crutches can be used to ascend and descend stairs following the pattern previously described for axillary crutches, but more easily. Rather than having to place two axillary crutches on one side in order to hold onto a railing, the patient with Lofstrand crutches can grasp the railing while the crutch of the grasping hand is still attached to the arm.

3. Platform Crutches

Platform crutches provide an alternative when weight bearing through the hand, wrist, and distal forearm are prohibited, but offloading of a lower extremity is required (Figure 41–18). The examiner should first ensure that the patient is permitted to bear weight through the proximal forearm.
Figure 41–18 Right-sided platform crutch used to offload the distal forearm and wrist.

The platform attachment is a padded trough that supports the patient’s medial to proximal forearm. The trough has a Velcro strap to secure the forearm and a vertical handgrip that the patient can use for extra support. The attachment can be adjusted to fit most axillary crutches. Adjustment in height can be made to both the crutch and the platform attachment.

Advantages & Disadvantages

Platform crutches can make ambulation a reality when a patient has both upper and lower extremity weight-bearing restrictions. The platform crutch can be used in conjunction with a normal axillary crutch on the uninvolved side to offload a lower extremity. If a patient has bilateral wrist and hand involvement, two platform crutches can be used. Alternately, when patients can bear weight through their forearm on one side, but cannot bear any weight on the other upper extremity, a single platform crutch can be used to help the patient fully or partially offload a lower extremity.

Use of platform crutches involves a greater degree of balance than axillary crutches. More standing time and adjustment prior to ambulating may be needed to ensure proper placement of the crutch under the axilla and the platform under the forearm, with the strap attached. This extra stance time and setup time make sit-to-stand transfers more precarious and time consuming. Platform crutches can also be difficult to use on stairs. Grasping of a railing becomes more difficult,
and assistance from another person may be needed to bring one of the crutches up the steps.

**Management Considerations**

**A. Fitting**

The height of the crutch to which the platform is attached is fitted in the same way as an axillary crutch, by allowing a 2-fingerbreadth space between the top of the crutch and the patient’s axilla (refer to earlier discussion). However, instead of adjusting the grip height, the platform height is adjusted so that the patient can stand upright, and not have to bend over to lean on the upper extremity platform.

**B. Use on Stairs**

Platform crutches can be used to navigate stairs following the pattern previously described for axillary crutches. However, owing to the bulk of the platform, and the inability to use the hand and wrist on the affected side, transferring two crutches under one arm to grasp the railing on the other side becomes almost impossible. Thus, when using platform crutches, the patient either must use just the crutches and no railing, or have a helper bring the regular axillary crutch up the stairs. Another option is stair bumping, but again this can prove hazardous when trying to transition from standing to sitting onto the steps, and it is difficult to bring both crutches up the steps without help. Patients using this option will likely need assistance.

**WALKERS**

Walkers encompass a group of assistive devices that provide four points of contact with the ground, effectively providing a stable frame. Walkers are typically used to increase balance, to offload a lower extremity, or to conserve energy in patients with endurance limitations.

**1. Standard Walker**

A standard walker has four legs with rubber tips that contact the ground. Each leg is adjustable for height. The walker forms a three-sided frame that the patient can walk within to provide forward and lateral support. Walkers are usually
constructed of a lightweight metal such as aluminum. The walker is advanced by lifting from the lateral hand supports, placing it forward, and then stepping toward into the frame. Standard walkers may have fixed frames or the ability to fold for easier portability and storage.

Advantages & Disadvantages

Standard walkers provide significant stability. The frame is rigid and can be used to hang other equipment, such as oxygen tanks, chest tubes, and so on. For home use, baskets or bags can be hung on the walker to store household items. Folding walkers can be stored easily and are more portable than rigid walkers.

Standard walkers require upper body strength, balance, and endurance to advance the walker after each step. Also, advancing the walker involves stopping and starting, making a person’s gait less efficient. Walkers are wide, and may be difficult to maneuver in tight spaces. Four legs provide extra tripping hazards when transitioning from smooth surfaces to raised surfaces such as carpets or rugs. Walkers cost more than other assistive devices, and even when foldable can be awkward to transport. Folding walkers can be used on stairs, but only with extreme caution.

Management Considerations

A. Fitting

Each of the four legs can be adjusted to obtain the right height via compressible locking pins. The holes on each leg for the adjusting pin are usually numbered to ensure that the legs are the same height. As with other assistive devices, the handgrips should be measured to be as high as the patient’s wrists when in a neutral standing position.

Because the walker is meant to fit around the user in front and laterally, the patient’s body habitus must be taken into consideration. A standard walker should comfortably contain the patient’s body. If girth, edema, or external equipment makes it impossible for the patient to fit within the walker during gait, a wider walker (bariatric) should be used. Most aluminum walkers are suitable for patients weighing 250–300 lb (113–136 kg); a bariatric walker can be used for patients who exceed this weight limit.
B. Use on Stairs

Walkers are challenging to use in environments that include steps. Many people who need walkers position one upstairs and one downstairs to eliminate the need to maneuver the walker up and down the steps. The patient can then use a railing to get up the steps, or use an alternative strategy, such as stair bumping.

If a railing is present and the walker folds, it can be used with extreme caution on a stairway. The walker should be folded and then lifted to the first step. The patient then presses through the railing on one side and the walker with the other hand in order to step up to the first step. Care must be taken to avoid tripping over the legs of the walker. Once safely on the same step as the walker, the patient can then raise the walker to the next step and repeat. When descending, the walker should come first, followed by the patient to the same step. This should only be attempted after thorough training and only by patients with good balance and upper body strength.

2. Rolling Walker

An alternative to a standard walker with four legs is a rolling walker with wheels on both front legs (Figure 41–19). The wheels allow the walker to be pushed forward when advancing instead of having to lift it. The back legs are identical to those of a standard walker and do not have wheels. This design prevents the walker from rolling freely and uncontrollably for the user and maintains stability. Gliders can be ordered and used on the back legs to allow the walker to roll over uneven surfaces such as carpets more easily.
Advantages & Disadvantages

Because the wheels decrease the need to start and stop, and lift the walker to advance it, rolling walkers require less energy and can be more effective for patients with endurance limitations. Rolling walkers can promote a more fluid, natural gait pattern in patients who must rely on a walker. However, the wheels make a rolling walker even more precarious to use on stairs.

Management Considerations

Management issues are similar to those previously discussed for a standard walker, but as noted use of a rolling walker on stairs is not advisable.
3. Rollator Walker

A third type of walker, known as a rollator, provides a bench inside the walker frame for the user to sit on when he or she becomes fatigued, and hand brakes that can be set prior to transitioning between sitting and standing. Rollators often have wheels for all four legs, and added stability is provided by having hand brakes. Some models are designed with one front wheel instead of two, similar to a tricycle.

Advantages & Disadvantages

Rollators provide the added safety of having a seat available whenever needed, helping to conserve energy in patients who are easily fatigued. They are bigger than, heavier than, and not as easy to transport as a rolling walker. They are also more expensive than a rolling walker. Rollators cannot be used on stairs.

Management Considerations

Management concerns are similar to those applicable to a standard walker, but as noted a rollator should not be used on stairs.

4. Platform Rolling Walker

Walkers require the usage of both hands to steer and maneuver and to offload a lower extremity. If a person has impairment to one distal forearm or hand, a platform attachment can be applied to the affected side, similar to the padded platform with a Velcro strap used on a platform crutch (Figure 41–20).
Advantages & Disadvantages

Platform rolling walkers allow usage of a walker even if a hand or distal forearm is impaired. As with a platform crutch, increased standing time and adjustment of straps occur with each usage. The platform also makes the walker almost impossible to use on stairs, and more awkward to transport.

Management Considerations

Management issues are similar to those previously discussed for platform crutches.


Medical Rehabilitation

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Primary care is coordinated, comprehensive, and personal care, available on both a first-contact and a continuous basis. It can be defined by several tasks: (1) medical diagnosis and treatment; (2) psychological diagnosis and treatment; (3) personal support of patients of all backgrounds; (4) communication of information about diagnosis, treatment, prevention, and prognosis; (5) maintenance of patients with chronic illness; and (6) prevention of disability and disease through detection, education, behavioral change, and preventive treatment.

This chapter addresses the common clinical problems encountered by physiatrists in office practice of adult patients with disability. In this setting, the physician’s responsibilities and tasks extend beyond the narrow technological confines of medical diagnosis and treatment. As the number of patients requiring rehabilitation increases, the medical problems of those with physical disabilities become more complex, and the availability of primary care physicians who understand the needs of chronically ill patients become relatively less available, it becomes more important for physiatrists to understand the tasks that comprise the clinical work of physicians providing primary care for patients with such problems.
Historically, there has been little coordination across the multiple settings, providers, and treatments encompassed in the care of chronically ill patients. The treatments for chronic diseases are often complicated, making it difficult for patients to comply with treatment protocols. Effective medical care usually requires longer visits to the physician’s office than is common in acute care. Moreover, in treating chronic illnesses, effectiveness of the same intervention, whether medical or behavioral, may differ depending on when in the course of the illness the intervention is suggested. Fragmentation of care is a risk for patients with chronic diseases, because frequently multiple chronic diseases coexist. Physiatrists, because of their broad scope of practice, are ideally positioned to assume the role of gatekeeper in the overall medical management of patients with chronic illness and disability.

**PREVENTIVE SCREENING**

Preventive screenings are an important part of health promotion efforts. Many preventive screenings have been recognized as a cost-effective way to identify and treat potential health problems before they develop or worsen. However, it can be challenging to keep up with the latest scientific thinking regarding screenings. Age- and gender-specific preventive screening recommendations exist for dozens of health concerns, but the recommendations may vary from organization to organization and are frequently changed as new information becomes available.

Physicians must acknowledge their primary role in prevention as that of educators. Accurate information regarding risk factors is most likely to reinforce health-enhancing behavior and alter self-destructive behavior. The physician must appreciate the potential for behavior modification and familiarize himself or herself with local resources. Routine screening for specific diseases, the maintenance activity most closely identified with the physician, should be performed selectively. The limits of screening tests as well as their potential health benefits should be clearly understood by every physician, especially those who are managing patients with chronic disabling conditions.

Screening tests are performed to identify asymptomatic disease. The alternative is to wait until patients present with symptoms and then make the diagnosis. The practical objective of screening is prevention of morbidity and mortality—not simply early diagnosis. There is little benefit to the patient, and perhaps considerable harm, in advancing the time of diagnosis of a disease for which earlier treatment does not influence outcome.
Several official entities have recommended or published screening protocols and guidelines aimed at improving early detection and hence prevention of common diseases (Table 42–1). Among the most commonly performed screening measures in the general population are those developed to identify risks for hypertension, heart disease, breast cancer, prostate cancer, and colon cancer.

Table 42–1 Routine health screening and examination guidelines.
<table>
<thead>
<tr>
<th>Screening Measure</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Obesity/Weight control</td>
<td>Check weight annually if body mass index (BMI) is within normal range.</td>
</tr>
<tr>
<td></td>
<td>Check more frequently if patient is participating in a weight-loss management program.</td>
</tr>
<tr>
<td>Blood pressure</td>
<td>Check annually with each medical history review and physical examination if blood pressure is &lt; 120/80 mm Hg.</td>
</tr>
<tr>
<td></td>
<td>Check more frequently if blood pressure is higher.</td>
</tr>
<tr>
<td>Breast disease</td>
<td>Monthly self-breast examinations.</td>
</tr>
<tr>
<td></td>
<td>Annual mammogram and clinical breast examination for all women aged 40–69 years or younger if high risk.</td>
</tr>
<tr>
<td></td>
<td>After age 70 years, examinations and screenings at provider or patient discretion</td>
</tr>
<tr>
<td>Colon cancer</td>
<td>Colonoscopy at age 50 years and then every 10 years.</td>
</tr>
<tr>
<td></td>
<td>Or, sigmoidoscopy or barium enema every 3–5 years plus annual stool test for blood.</td>
</tr>
<tr>
<td></td>
<td>Check more frequently if patient is at high risk.</td>
</tr>
<tr>
<td>Prostate cancer</td>
<td>Digital rectal examination annually for all men aged 50 years and older.</td>
</tr>
<tr>
<td></td>
<td>Prostate-specific antigen (PSA) test is inconclusive.</td>
</tr>
<tr>
<td>Vision/Glaucoma</td>
<td>Every 2–4 years until age 65, then every 1–2 years.</td>
</tr>
<tr>
<td></td>
<td>Annually if at high risk (diabetes).</td>
</tr>
<tr>
<td>Audiology/Hearing</td>
<td>As needed.</td>
</tr>
<tr>
<td>Dental examination</td>
<td>Annually.</td>
</tr>
<tr>
<td>Osteoporosis/Bone density</td>
<td>Dual-energy X-ray absorptiometry (DEXA) scan for postmenopausal women aged 65 years and older and for men aged 70 years and older, or younger if at risk.</td>
</tr>
<tr>
<td>Mental health</td>
<td>Discuss mood and memory concerns at each visit.</td>
</tr>
<tr>
<td>Smoking cessation</td>
<td>At each visit.</td>
</tr>
</tbody>
</table>

*The U.S. Preventive Services Task Force (USPSTF) recommends against PSA-based screening for prostate cancer (grade D recommendation). This recommendation applies to men in the general U.S. population, regardless of age.*

**HEALTH MAINTENANCE IMMUNIZATIONS**

Immunizations are an effective and important means of controlling many communicable diseases through primary prevention. Their underuse stems, in large part, from a failure of public education and access to health care delivery. In general, live attenuated vaccines provide more complete and longer lasting
immunity than inactivated agents. However, because live vaccines can produce serious disseminated disease in the immunosuppressed host, these preparations should be avoided in patients who are immunologically deficient.

### Vaccination Recommendations

Patients who are chronically ill should be evaluated to determine their immunization status, and whether they would benefit from additional immunizations, including those described below. As noted, vaccines that contain a live virus are generally contraindicated in immunocompromised individuals. Rarely, individuals with allergies can have reactions to components of individual vaccines. Specific cautions and exclusions relating to routinely administered vaccines can be found on the Center for Disease Control and Prevention’s vaccine and immunization web pages (see [www.cdc.gov/vaccines/vpd-vac/should-not-vacc.htm#mmr](http://www.cdc.gov/vaccines/vpd-vac/should-not-vacc.htm#mmr)).

**A. Streptococcus pneumoniae Vaccine**

The vaccine is effective against 23 serotypes of *S pneumonia*, and is recommended for individuals with asplenia, sickle cell disease, or any debilitating disease, as well as those older than 65 years of age. The dose should be repeated in 5–6 years.

**B. Influenza Vaccine**

The influenza vaccine given seasonally is an inactivated vaccine containing two type A and one (or two) type B strains of the influenza virus that have been identified as most likely to cause illness in a given year. The vaccine is generally effective within 2 weeks and must be readministered each year. The Advisory Committee on Immunization Practices (ACIP) of the Centers for Disease Control and Prevention (CDC) currently recommends universal vaccination of all individuals older than 6 months of age. High-risk groups (ie, those with chronic illness or immunocompromising conditions, pregnant women, health care professionals, and those in contact with high-risk persons), and older adults should receive priority vaccination. Trivalent inactivated vaccine is contraindicated in persons with severe egg allergy, and it should be used with caution in patients with Guillain-Barré syndrome.

**C. Measles, Mumps, and Rubella (MMR)**
Children in the United States usually receive the live-virus vaccine against measles in combination with mumps and rubella (German measles) as part of the standard childhood immunizations. MMRV is a combination vaccine that adds protection against varicella. Because this is a live-virus vaccine, it should not be administered to immunocompromised patients. In addition, some patients with allergies may have reactions to components of the vaccine.

D. Measles Vaccine

The live-virus measles vaccine (Attenuvax) is recommended for all susceptible people older than 12 months of age (ie, anyone who does not have documented immunity against measles). Although usually given as part of the MMR vaccination, measles vaccine may be administered separately to individuals who are at risk for contracting infection because of incomplete or missed childhood vaccinations. Persons born before 1956 are considered immune. Individuals born after 1956 should have received two doses of live measles vaccine, given not less than 1 month apart. However, some adults born after 1956 may have received only the first dose as a child. These individuals should be given a second dose of the vaccine to ensure complete protection against the virus. The same cautions listed earlier for MMR administration apply to measles vaccination.

E. Varicella Vaccine

Varicella vaccine (Varivax) is recommended for all older individuals who are not known to be immune to the varicella virus. A past history of chicken pox is sufficient for assuming immunity. If an adult patient is unsure whether he or she had a prior infection, immune status should be evaluated before administering the vaccine as up to 91% of patients so tested are immune.

F. Hepatitis A Vaccine

Unless otherwise contraindicated, inactivated hepatitis A vaccine (Havrix, VAQTA) is recommended in persons 2 years of age or older who are at increased risk of infection with hepatitis A virus, including those with chronic liver disease and travelers outside the United States (except for northern and western Europe, New Zealand, Australia, Canada and Japan).

G. Hepatitis B Vaccine

Hepatitis B vaccine is recommended for all individuals who are or may be at
increased risk for infection with hepatitis B virus, including all adolescents. Booster doses can be given every 7 years but are not currently recommended. If an individual’s antibody level is greater than 10 IU/mL, a booster dose is not needed. If a patient has been exposed to hepatitis B and has had the complete vaccination series but the antibody level is unknown, hepatitis B immune globulin (HBIG) should be administered along with a booster dose.

**H. Tetanus and Diphtheria (Td)**

Many children and young adults in the United States have been vaccinated against tetanus and diphtheria as part of the standard childhood vaccinations. Tetanus booster is recommended once every 10 years after the primary series. The booster may be given at 5 years for “dirty” wound management.

**I. Meningococcal Vaccine**

Adults 55 years of age and younger who require meningococcal vaccination should receive the quadrivalent meningococcal polysaccharide vaccine. Individuals without a functioning spleen and those with persistent complement deficiencies should routinely receive a two-dose series with 2 months between doses, then booster doses every 5 years. Persons infected with HIV do not require booster doses after the initial two-dose series. Other adults who are at increased risk of infection (eg, college students living in dormitories, laboratory personnel at risk of exposure) should receive a single dose of vaccine; revaccination is not required unless the first dose was given before 16 years of age.

**J. Herpes Zoster Vaccine**

A single dose of the herpes zoster vaccine (Zostavax) is recommended for adults aged 60 years and older, regardless of their history. Vaccine effectiveness decreases with age; therefore, patients should be immunized as soon as possible after age 60. The vaccine contains a live virus and thus is contraindicated in persons with immunocompromising conditions.

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Centers for Disease Control and Prevention: 2009 Adult vaccination coverage.
SMOKING CESSATION

Tobacco use is implicated in one in five deaths in the United States per year. Smoking is considered the most important of the modifiable risk factors for ischemic heart disease and stroke. There are both pharmacologic and nonpharmacologic options to aid in smoking cessation. Nonpharmacologic options include receiving brief advice from a health care provider and participating in behavioral therapy. Options such as acupuncture and hypnotherapy, while anecdotal evidence might prove otherwise, have not been shown to be statistically more effective than placebo. Pharmacologic options, however, are well studied and have been shown to be quite effective. These include bupropion, nortriptyline, varenicline, and nicotine replacement therapy (NRT). NRT has a mild side-effect profile and has proven effective in clinical trials. If patients are unable to adhere to NRT, a trial of bupropion, nortriptyline, or varenicline may be considered next. There are no recommendations at this time as to which is the superior. The physician must consider pricing, patient preference, availability, and potential side effects when prescribing these agents.

Centers for Disease Control and Prevention: Health Effects of Cigarette
CARDIOVASCULAR DISORDERS

HYPERTENSION

ESSENTIALS OF DIAGNOSIS

► Hypertension in adults is defined as a systolic blood pressure of at least 140 mm Hg, or a diastolic blood pressure of at least 90 mm Hg, or both.
► Screening is the primary method of detection.
► Nonmodifiable risk factors include older age, male gender, African-American race, and a family history of hypertension.
► Modifiable risk factors include obesity, sedentary lifestyle, excessive alcohol consumption, tobacco use, increased sodium intake, decreased potassium intake, vitamin D deficiency, and stress.
► Untreated hypertension may lead to cardiovascular disease, peripheral vascular disease, renal disease, and other end-organ damage.

General Considerations

Hypertension in adults is defined as a systolic blood pressure of at least 140 mm Hg, or a diastolic blood pressure of at least 90 mm Hg, or both (Table 42–2). Screening is the primary method of detecting hypertension. Essential hypertension, also known as idiopathic or primary hypertension, accounts for approximately 90% of cases, has no identifiable cause, and is often asymptomatic. It is an important risk factor for subsequent cardiovascular disease, peripheral vascular disease, and stroke. Secondary hypertension has an identifiable and often treatable or reversible cause; examples include obstructive...
sleep apnea and use of certain medications (eg, birth control pills and decongestants).

**Table 42–2 Classification and management of blood pressure in adults.**

<table>
<thead>
<tr>
<th>Blood Pressure Classification</th>
<th>Systolic Blood Pressure (mm Hg)</th>
<th>Diastolic Blood Pressure (mm Hg)</th>
<th>Lifestyle Modification</th>
<th>Initial Drug Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt; 120 and &lt; 80</td>
<td>Encourage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prehypertension</td>
<td>120–139 or 80–89</td>
<td>Yes</td>
<td>No antihypertensive drug indicated</td>
<td></td>
</tr>
<tr>
<td>Stage 1 hypertension</td>
<td>140–159 or 90–99</td>
<td>Yes</td>
<td>Thiazide-type diuretics for most patients. May consider ACEI, ARB, BB, CCB, or combination.</td>
<td>Drug(s) for compelling indications. Other antihypertensive drugs (diuretics, ACEI, ARB, BB, CCB) as needed.</td>
</tr>
<tr>
<td>Stage 2 hypertension</td>
<td>&gt; 160 or &gt; 100</td>
<td>Yes</td>
<td>Two-drug combination for most patients (usually thiazide-type diuretic and ACEI, or ARB or BB or CCB).</td>
<td>Drug(s) for compelling indications. Other antihypertensive drugs (diuretics, ACEI, ARB, BB, CCB) as needed.</td>
</tr>
</tbody>
</table>

ACEI, angiotensin-converting enzyme inhibitors; ARB, angiotensin II receptor blocker; BB, β blocker; CCB, calcium channel blocker.

*Refer to Table 42–3.


Nonmodifiable risk factors include increasing age, male gender, African-American race, and a family history of hypertension. Modifiable risk factors include obesity, sedentary lifestyle, excessive alcohol consumption, tobacco use, increased sodium intake, decreased potassium intake, vitamin D deficiency, and stress.

Hypertension causes increased systemic vascular resistance (afterload), which leads to concentric left ventricular hypertrophy and decreased left ventricular function. Thus, the chamber often dilates, leading to heart failure.
Hypertension also accelerates atherosclerosis, leading to a higher incidence of coronary artery disease (CAD).

Clinical Findings

A. Symptoms and Signs
Most people with hypertension have no signs or symptoms, even when blood pressure readings reach dangerously high levels. A few individuals with early-stage hypertension may have dull headaches, dizziness, or nosebleeds but these findings typically do not occur until blood pressure elevation has reached a severe stage.

B. Diagnostic Testing
Hypertension is identified by means of blood pressure readings using an appropriately sized cuff. When warranted, an electrocardiogram can help identify cardiac abnormalities associated with hypertension (eg, left ventricular hypertrophy). A basic metabolic panel can provide evidence of primary and secondary contributors to hypertension, including low potassium level and renal dysfunction (blood urea nitrogen and creatinine).

Complications
Most complications of hypertension result from end-organ damage. Hypertension is a major risk factor for CAD, which can result in angina and myocardial infarction. Congestive heart failure with left ventricular hypertrophy is another common result of chronic uncontrolled blood pressure elevation. In addition, hypertension predisposes patients to peripheral vascular disease and is associated with an increased risk of aortic dissection.

Uncontrolled hypertension can lead to intracerebral hemorrhage, transient ischemic attacks, and other stroke subtypes, as well. Severely elevated pressures can cause hypertensive encephalopathy. Kidney disease with eventual renal failure can occur in patients with chronic hypertension as a result of decreased glomerular filtration rate and tubular dysfunction.

Treatment
A. Primary Prevention

Treatment of hypertension is often initially addressed with lifestyle modification techniques, and this alone may be adequate for some patients. Patients should be advised to limit sodium intake and alcohol consumption, lose weight, exercise regularly, stop unnecessary medications that may contribute to hypertension, stop smoking or other use of tobacco products, and engage in stress management practices. Additional risk factors, if identified, should be addressed (eg, low potassium level, contributing to renal problems).

Use of tobacco and alcohol should be questioned and addressed by all health care providers, and the benefits and methods of smoking cessation discussed at every visit. Alcohol consumption should be limited to the equivalent of 30 mL (in men) or 15 mL (in women) of ethanol per day. Some evidence suggests that the cardiovascular benefits of alcohol may be greater for red wine than for other alcoholic drinks. Overweight or obese people need to be advised to lose weight. Consumption of fruits and vegetables is usually sufficient for adequate potassium intake; however, some patients with low potassium levels may benefit from supplementation.

Ambulatory blood pressure self-monitoring should be recommended for patients with prehypertension or hypertension as part of their regular monitoring.

B. Pharmacotherapy

Pharmacologic intervention includes the use of various antihypertensive agents (refer to Tables 42–2 and 42–3). Thiazide diuretics are considered first-line agents in African-American patients as this population often has a type of hypertension that is more “salt-sensitive.” Monitoring for hypokalemia is important while patients are taking these drugs. If patients also have diabetes, however, an angiotensin-converting enzyme (ACE) inhibitor is the initial choice. Angiotensin II receptor blockers (ARBs) also inhibit the renin–angiotensin–aldosterone system. & blockers decrease heart rate, cardiac output, and renin release. Calcium channel blockers cause vasodilation of arteriolar vasculature. Vasodilators such as minoxidil and hydralazine are used often in conjunction with & blockers.

Table 42–3 Drug therapy for compelling indications of hypertension in adults.
HYPERLIPIDEMIA

ESSENTIALS OF DIAGNOSIS

- Excess serum cholesterol, especially low-density lipoprotein cholesterol (LDL-C) level > 160 mg/dL, or excess triglycerides (> 250 mg/dL).

ACEI, angiotensin-converting enzyme inhibitors; ARB, angiotensin II receptor blocker; CCB, calcium channel blocker.

Patients are usually asymptomatic; however, rarely symptoms of lipid deposition or pancreatitis may be noted.

LDL-C is considered to be the major contributor to CAD risk; high-density lipoprotein cholesterol (HDL-C) is cardioprotective.

General Considerations

Hyperlipidemia is a group of disorders characterized by an excess of serum cholesterol, especially excess LDL-C, or an excess of triglycerides, or both. On its own it is often asymptomatic. Hyperlipidemia is often genetically determined, but it can be caused or amplified by abnormal diet, drugs, and certain disease conditions. Hyperlipidemia is one of the most important and modifiable risk factors for CAD as it causes accelerated atherosclerosis.

Clinical Findings

A. Symptoms and Signs

Hyperlipidemia is usually asymptomatic. In rare cases, patients may experience recurrent abdominal pain as a result of pancreatitis secondary to very high triglyceride levels of greater than 1000 mg/dL. Similarly, although there are often no signs associated with hyperlipidemia, at extremely high levels, as occurs in familial hypercholesterolemia, lipid deposition may produce masses that are observable on physical examination. Patients may present with xanthomas, which appear as hard yellowish masses on the extensor tendons of the hands, the Achilles tendons, plantar tendons, and at the insertion of the patella tendon. Xanthelasmas can also be seen and are described as yellowish plaques on eyelids, but these are not specific for hypercholesterolemia or hyperlipidemia.

B. Laboratory Findings

Cholesterol and triglyceride levels are routinely evaluated in primary care visits. The determination of what constitutes “ideal,” “low,” and “high” cholesterol continues to be refined as epidemiologic studies provide evidence about long-term consequences relating to these values.
1. **Total cholesterol**—Ideal total cholesterol is currently equated with a level less than 200 mg/dL, but some evidence suggests that levels between 60 and 200 mg/dL may be associated with an increased risk of CAD. A borderline level is considered to be 200–240 mg/dL, and high total cholesterol is deemed to be any level greater than 240 mg/dL. High total cholesterol is associated with a severe increased risk of CAD and requires medical intervention.

2. **LDL-C**—LDL is considered to be the major contributor to CAD risk because it is the most atherogenic of all lipoproteins. It is a calculated measurement derived from the total cholesterol value. Ideal LDL is currently considered to be a level less than 130 mg/dL; borderline is 130–160 mg/dL; and high is greater than 160 mg/dL.

3. **Triglycerides**—An ideal triglyceride level is considered to be less than 125 mg/dL; borderline is 125–250 mg/dL; and high is greater than 250 mg/dL.

4. **HDL-C**—HDL has a protective effect as it removes excess cholesterol from arterial walls. For every 10 mg/dL increase in HDL levels, the CAD risk is decreased by 50%. A low HDL level of less than 35 mg/dL is a major independent risk factor for CAD, whereas high HDL of greater than 60 mg/dL counteracts risk to some degree (see later discussion).

# Treatment

Treatment goals are based on absolute serum levels of lipids in combination with risk stratification of patients. Evidence shows that effective therapy to lower serum LDL-C is associated with dramatic benefits in terms of short-term morbidity and mortality in patients with CAD, and long-term morbidity and mortality even in low-risk patients. The short-term goal is to reduce LDL levels, while the long-term goal of treatment is to reduce the risk of atherosclerosis and CAD.

## A. Primary Prevention

1. **Identification of risk factors**—The patient’s risk for CAD events is determined, based on the Framingham score for 10-year risk of developing myocardial infarction or death, to aid in planning specific treatment interventions. In tabulating this score, major risk factors are giving one point each; these include hypertension (blood pressure > 140/90 mm Hg, or treatment
for hypertension), cigarette smoking (any within the past month, HDL-C level < 40 mg/dL), male gender and age greater than 45 years, female gender and age greater than 55 years, and family history of premature CAD (clinical disease or sudden death in a first-degree male relative before age 55, or first-degree female relative before age 65). If HDL-C level is 60 mg/dL or greater, one point is subtracted from the total.

Updated recommendations from the National Cholesterol Education Program Adult Treatment Panel (NCEP-ATP) III define target levels for treatment based on the Framingham score and include optional, more aggressive, lower target LDL-C goals in patients at higher risk. The target LDL-C levels defined by NCEP are listed in Table 42–4.

Table 42–4 Target goals for treatment of elevated low-density lipoprotein cholesterol (LDL-C).\(^a\)
2. **Lifestyle modification**—Many of the measures previously described for prevention of hypertension and hyperlipidemia are included in the treatment recommendations for lifestyle modifications to improve LDL and HDL levels. In particular, exercise is beneficial in increasing HDL-C level.

### B. Pharmacotherapy

HMG-CoA reductase inhibitors (statins) are the medication of choice for patients at high risk of CAD. These agents have been shown to reduce mortality from cardiovascular events and significantly reduce total mortality by lowering LDL-C. They are also indicated as secondary stroke prophylaxis. Statins are considered to be more effective than other medications in reducing LDL-C;
however, they are less effective than fibrates for reducing triglycerides and raising HDL-C. Liver function tests should be monitored at first monthly for 3 months, and then every 6 months, in all patients taking these drugs.

Niacin is a second-line agent in the treatment of hyperlipidemia. It lowers triglyceride and LDL levels and increases HDL levels but should be avoided in diabetic patients, in whom it may worsen glycemic control. At levels prescribed for antilipemic treatment, niacin is often poorly tolerated. The most common side effect, cutaneous flushing, can be reduced by taking aspirin 30 minutes prior.

Bile acid–binding resins lower LDL but increase triglyceride level. They are often effective when used in combination with statins or niacin to treat severe disease in high-risk patients. However, these agents are often poorly tolerated because of side effects, including gastrointestinal discomfort.

Fibrates are used when all the other pharmacologic options fail. They lower very low-density lipoprotein (VLDL) and triglyceride levels while increasing HDL. Side effects include gastrointestinal distress and, less commonly, gynecomastia, gallstones, weight gain, and myopathies.


CORONARY ARTERY DISEASE

ESSENTIALS OF DIAGNOSIS

➤ May present as stable angina pectoris, unstable angina, myocardial infarction, or sudden cardiac death, or be completely asymptomatic.

➤ Stable angina produces chest pain or a substernal pressure sensation (heaviness, pressure, or squeezing), usually lasts 1–5 minutes, is brought on
by exertion or emotion and relieved with rest or nitroglycerin.

- Classic symptoms are crushing substernal chest pain, radiating to the left neck, jaw, or arm, and a sensation similar to gastroesophageal reflux.
- Atypical symptoms are common in women and in the elderly.

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**General Considerations**

Coronary artery disease (CAD) is the clinical manifestation of the pathologic features seen in atherosclerosis of the coronary arteries. It is the most common type of heart disease and is the leading cause of death for both men and women in the United States. Every year, more than 400,000 Americans die from CAD.

Nonmodifiable risk factors include age, gender, and family history of CAD. In premenopausal women, estrogen has cardioprotective properties. Modifiable factors account for more than 90% of the population-attributable risk of a first myocardial infarction and include smoking, dyslipidemia, hypertension, diabetes mellitus, abdominal obesity, psychosocial factors, regular alcohol consumption, and lack of regular physical activity. Controlling risk factors often can lessen genetic influences and help prevent CAD, even in older adults.

**Clinical Findings**

**A. Symptoms and Signs**

CAD can present as stable angina pectoris, unstable angina, myocardial infarction, or sudden cardiac death, yet for some individuals can be completely asymptomatic. Stable angina appears as chest pain or a sensation of substernal pressure (often described as heaviness, pressure, or squeezing) that usually lasts for 1–5 minutes, is brought on by exertion or emotion, and is relieved with rest or nitroglycerin. The classic picture is crushing substernal chest pain, radiating to the left neck, jaw, or arm, and a sensation similar to gastroesophageal reflux. Up to one third of patients are asymptomatic or have other symptoms, such as dyspnea, syncope, diaphoresis, weakness, nausea, and vomiting. This atypical presentation is more common in patients who are diabetic, female, elderly, or postoperative.
B. Diagnostic Tests

Diagnosis of CAD relies on findings obtained primarily from electrocardiographic studies. One or more of the following diagnostic studies may be ordered: electrocardiogram, exercise stress test, echocardiography, Holter monitoring, and cardiac catheterization. In patients with unstable angina, the diagnostic workup needs to exclude a myocardial infarction; these patients should be stabilized with medical management prior to undergoing stress testing or catheterized initially. (Additional information about cardiac evaluation of at-risk patients appears in Chapter 23.)

Treatment

A. Primary Prevention

Patients should be educated about risk factors for CAD and actions they can take to decrease their risk of disease. Smoking cessation is paramount; patients often require assistance in identifying options and methods that will increase their chances of quitting successfully. Blood pressure control is likewise crucial, and measures aimed at reaching a goal of less than 130/60 mm Hg may include use of pharmacologic agents, if necessary (see Hypertension, earlier). Hyperlipidemia should be addressed with lifestyle modifications, including regular exercise and decreased intake of saturated fat and cholesterol. Medications such as HMG-CoA reductase inhibitors (statins) may be prescribed to help to reduce LDL level (see Hyperlipidemia, earlier).

B. Inpatient Care

Treatment involves risk-factor modification for all patients with CAD, as previously discussed. Medical therapy for patients with stable angina includes aspirin, as it decreases morbidity and reduces the risk of a myocardial infarction. β-Blockers decrease cardiac workload and have been shown to reduce the frequency of coronary events. Nitrates reduce preload through generalized vasodilation and also offer the benefit of symptom relief. Revascularization may be preferred in high-risk patients.

Patients with unstable angina should be admitted to the hospital for continuous cardiac monitoring. Aggressive medical management is warranted, as well as oxygen therapy and pain relief with nitrates and morphine. (See Chapter 23 for further details.)
C. Exercise Therapy

The exercise prescription for a patient with CAD needs to address type, intensity, duration, and frequency of exercise. There should be a particular emphasis on aerobic, isotonic exercises that involve larger muscle groups. Some patients with CAD, including those with congestive heart failure, severe valvular disease, and uncontrolled arrhythmias, should avoid resistive and isometric exercises. It is common to use a target heart rate to assess exercise intensity. For detailed discussion of exercise in a post–myocardial infarction patient, refer to Chapter 23 on cardiac rehabilitation.


PULMONARY DISORDERS

SLEEP APNEA

ESSENTIALS OF DIAGNOSIS

- Cessation of airflow for > 10 seconds during sleep.
- Oxygen saturation usually decreases by > 4% during the apneic episodes.
- Diagnosis is confirmed by polysomnography.

General Considerations

Sleep apnea is defined as a cessation of airflow for greater than 10 seconds during sleep. It becomes clinically significant at 10–15 episodes per hour, and in
severe cases patients may have more than 40 episodes per hour. Oxygen saturation usually decreases by more than 4% during the apneic episodes. Two classes of sleep apnea are differentiated: central and obstructive.

Diagnosis is confirmed by polysomnography (sleep study). Presence or absence of inspiratory effort during the apneic episode differentiates between obstructive and central apnea. Oxygen desaturation of less than 85% or a change of greater than 4% are significant. The frequency of hypoxic apneic episodes determines the severity of the disease.

Obstructive Sleep Apnea

Obstructive sleep apnea (OSA) occurs despite continuing ventilatory effort. The obstructive episode is usually followed by a loud snore. Patients have daytime hypersomnolence, snoring, and may have headaches, recent weight gain, and hypertension. OSA is frequently associated with an abnormal airway, myxedema, and obesity.

Treatment of persistent and significant OSA involves either nasal continuous positive airway pressure (nCPAP) or bilevel positive airway pressure (BiPAP). With nCPAP, air at constant pressure (5–15 mm H₂O) is supplied by means of a well-sealed nose mask. This acts like a “splint” to the pharynx to keep it open at night and is a very effective measure to prevent nocturnal hypoxemia. BiPAP is similar but can be used with a nasal or full-face mask and allows independent adjustment for inspiratory and expiratory pressures. This improves comfort and compliance.

Mild to moderate OSA can be successfully treated with weight loss; avoidance of alcohol, sedatives, and hypnotics; and not sleeping in the supine position. Nasal and intraoral patency devices may also be helpful. For moderate OSA, treatments include uvulopatopharyngoplasty, nCPAP, BiPAP, or a combination of these measures.

Central Sleep Apnea (CSA)

CSA occurs in less than 5% of sleep apnea patients. Although usually observed in neurologically impaired patients (eg, those with Cheyne–Stokes breathing), it frequently is seen in healthy patients at high altitudes for the first time and in patients suffering from congestive heart failure. Treatment mainly involves avoidance of central nervous system depressants such as alcohol, sedatives, and
hypnotics. Weight loss and avoidance of sleep deprivation are also helpful.


CHRONIC OBSTRUCTIVE PULMONARY DISEASE

ESSENTIALS OF DIAGNOSIS

► A group of diseases that includes emphysema, chronic bronchitis, and peripheral airway disease.

► Primary cause is long-term exposure to inhaled lung irritants, especially cigarette smoke.

► Lack of abnormal lung fibrosis is a distinguishing feature (in contrast to interstitial airway disease).

► Chest radiographs are preferable to spirometry for diagnosis of exacerbations.

General Considerations

Chronic obstructive pulmonary disease (COPD) is a progressive disease that usually involves components of emphysema and chronic bronchitis, often with predominance of one or the other condition. Peripheral airway disease, previously considered a distinct entity, is now thought to represent an early form of emphysema or early chronic bronchitis, and hence is encompassed within this group as well. In COPD, there is no abnormal fibrosis in the lung, a finding that distinguishes COPD from interstitial lung disease. COPD is usually treated with
bronchodilators and anticholinergics.

**Clinical Findings**

Emphysema is defined histologically by enlargement of the airspace distal to the terminal bronchioles, with destruction of the alveolar septa. There are two major types: centroacinar, usually seen in cigarette smokers; and panacinar, classically seen in patients with α₁-antitrypsin deficiency. Altered lung mechanics in patients with emphysema result in air trapping, decreased elastic recoil, increased compliance, and increased total lung capacity.

Chronic bronchitis, in contrast, has a clinical definition. It is defined as excess bronchial mucous secretion for at least 3 consecutive months for at least 2 years. Chronic bronchitis occurs in approximately 20% of adult men, and one cause is prolonged irritation by cigarette smoke.

Peripheral airway disease results from increased tortuosity, inflammation, and fibrosis of the small airways of the lung. Rather than a distinct entity, it is now thought to represent an early stage of emphysema or bronchitis.

The best prognostic indicator in COPD is the forced expiratory volume in 1 second (FEV₁). In both smokers and nonsmokers, FEV₁ decreases about 15–30 mL per year due to the normal aging process. An FEV that is less than 80% of predicted equates to compromised airway function indicative of COPD.

**Treatment**

For all patients with acute exacerbation of COPD, the American College of Physicians and American College of Chest Physicians (ACCP) recommend the following actions:

- Obtain a chest radiograph, because up to 23% of images show new infiltrates that change the chosen therapy.
- Do not use spirometry to diagnose or assess the severity of an exacerbation.
- Because inhaled anticholinergic drugs (eg, ipratropium [Atrovent]) are equally effective and more benign than inhaled β₂ agonists, start treatment with an inhaled anticholinergic drug and move to an inhaled β₂ agonist only after the maximum dose of the anticholinergic drug is achieved.
A. Oxygen Therapy

The focus of oxygen therapy for COPD patients in respiratory distress is to give enough oxygen to achieve 92% oxygen saturation (Sao₂), or as close to this level as possible. This is a required end point in initial management. Failure to treat hypoxia leads to further end-organ damage, worsening of pulmonary vasoconstriction, and sometimes death.

1. Continuous oxygen use—Criteria for starting continuous oxygen are resting partial arterial oxygen tension (Pao₂) less than 55 mm Hg, or Sao₂ less than or equal to 88%, or Pao₂ less than 59 mm Hg; or Sao₂ greater than 89% with evidence of cor pulmonale, dependent edema suggesting congestive heart failure, or erythrocytosis (hematocrit > 55%). Continuous oxygen use, if needed according to the abovementioned criteria, increases life span. It is the only treatment modality that decreases morbidity and mortality.

2. Intermittent oxygen use—Some patients have similar findings of hypoxia and desaturation during low-level exercise or sleep and can benefit from supplemental oxygen during these activities. However, long-term studies have not been done to evaluate outcomes in this patient group.

B. Smoking Cessation

The single most important intervention in smokers with COPD is to encourage smoking cessation. Simply telling the patient to quit succeeds 5% of the time. Behavioral approaches, ranging from clinician counseling to intensive group programs, may improve cessation rates. Pharmacologic therapy includes nicotine replacement, bupropion, and varenicline. Combining pharmacotherapy with behavioral approaches has also been recommended.

C. Pulmonary Rehabilitation

For a detailed discussion of this topic, please refer to Chapter 24.

ASTHMA

ESSENTIALS OF DIAGNOSIS

- Reversible inflammatory response of the airway with a multifactorial etiology.
- Diagnosis must demonstrate reversible bronchospasm and elicit a history compatible with asthma.

General Considerations

Asthma is a reversible, inflammatory condition of the airways with a multifactorial etiology. The inflammatory response has an acute phase and a late phase. In some patients with asthma the disease response is mediated by immunoglobulin E. Most asthmatics, whatever the etiology, develop a nonspecific airway hyperresponsivity, as shown by exacerbations that occur with exposure to cold, dust, and viral infections.

Clinical Findings

A. Symptoms and Signs
Early in an asthmatic attack, bronchospasm is the major finding, but later on, airway inflammation, airway edema, and increased airway secretions with possible mucous plugging may dominate, especially in status asthmaticus. Asthmatics usually present with some combination of dyspnea, cough, and wheezing, but on initial presentation patients may have complaints only of a chronic cough. The cause of asthma is often not discovered, especially in adult-onset disease, but occupational causes should be considered. Acetylsalicylic acid (ASA)–sensitive asthmatics may also be sensitive to other nonsteroidal antiinflammatory drugs (NSAIDs). Triad asthma is a clinical syndrome characterized by ASA sensitivity, asthma, and nasal polyposis; most patients, however, usually have just two of the three findings.

B. Diagnostic Tests

Diagnosis of asthma requires demonstration of reversible bronchospasm as well as a history compatible with asthma. If these are not present, a challenge test (ie, methacholine test) may be performed to induce bronchospasm. Response to a bronchodilator is defined as an increase in FEV₁ or forced vital capacity by 12% and 200 mL. Exercise-induced asthma is diagnosed by a decrease in FEV₁ of 15% or more after a thermal challenge (either exercise or hyperventilation).

Treatment

The most effective treatment for asthma is removing the known causative agents. This includes smoking cessation and minimizing exposure to secondhand smoke. Pharmacologic treatment includes antiinflammatory agents (corticosteroids and nedocromil), direct bronchodilators (β₂ agonists, anticholinergics, and methylxanthines), mast cell stabilizers (cromolyn), and leukotriene inhibitors.


DERMATOLOGIC DISORDERS

DERMATITIS

ESSENTIALS OF DIAGNOSIS

- Inflammation of the skin, usually from an allergic reaction to an allergen or prolonged exposure to an irritation.
- May be secondary to stress or a neurologic disorder.

General Considerations

Dermatitis is an inflammation of the skin that usually occurs in response to prolonged exposure to an irritant or an allergen. It can occur in insensate or immobile patients who are unable to detect or move away from an irritating stimulus. The condition is classified into varying forms, including contact, atopic, herpetiformis, seborrheic, nummular, stasis, perioral, and infective. Manifestations may range from skin rashes to bumpy rashes or blisters.

Contact dermatitis results from exposure to a topical irritant (eg, chemicals found in detergents, soaps, and other products). Seborrheic dermatitis is usually caused by physical stress and neurologic conditions, such as Parkinson’s disease. Varicose veins, chronic conditions, or infections that affect blood flow in the legs, and limited mobility associated with long-distance travel are potential causes of stasis dermatitis. The cause of atopic dermatitis is unknown, but the disease seems to result from a combination of genetic and environmental factors. Studies have found worsening of atopic dermatitis after long-term use of topical steroid cream (often used to treat the condition). In such cases, total cessation of the topical cream can lead to cure, although there is a period of severe rebound
between cessation of the topical agent and resolution of symptoms.

**Treatment**

Treatment creams that contain corticosteroids, use of wet compresses, and avoidance of identified allergens and irritants are components of most treatment plans. For some types of dermatitis, nonsteroidal medications may help relieve signs and symptoms. And for all types of dermatitis, occasional use of over-the-counter antihistamines can reduce itching.

Usatine RP, Riojas M: Diagnosis and management of contact dermatitis. Am Fam Physician 2010;82:249–255.

**INTERTRIGO**

**ESSENTIALS OF DIAGNOSIS**

- Bacterial, fungal, or viral infection of the skin.
- Develops at a site of broken skin as a result of inflammation, most often in body folds.
- Common in obese, diabetic, or immobilized patients.

**General Considerations**

Intertrigo refers to a bacterial, fungal, or viral infection that develops at a site of broken skin as a result of inflammation and the chafing of warm, moist skin. It is commonly seen in overweight individuals, because of an increase in the number of skinfolds, and in patients with diabetes mellitus. It is also common in patients who are restricted to bed rest, those who use diapers regularly, and those who have artificial limbs, all of which can lead to moisture-trapping against the skin. A common form is candidal intertrigo. Lesions appear red and raw, and may itch
and ooze.

**Treatment**

Infections can be treated with a topical or oral medications. The most common treatment is zinc oxide. For persistent intertrigo an antifungal cream is beneficial. Keeping the area dry, weight loss if the individual is overweight, weight shifting, good body and artificial limb hygiene, and use of absorbent fabrics or powders over the area will help prevent future occurrences, although relapses are common.

**CALLUSES & CORNS**

**ESSENTIALS OF DIAGNOSIS**

- Cutaneous responses that develop from inappropriate skin friction.
- Calluses are the skin’s response to friction, pressure, or irritation.
- Corns are seen in high numbers of patients with diabetes.

**General Considerations**

Calluses are toughened areas of skin that become thickened and hard in response to repeated friction, pressure, or other irritation. They are commonly found on the palms of the hands or soles of the feet. Although not harmful, calluses can lead to skin ulceration or infection. Callus formation is noted in many patients with diabetes mellitus and, together with absent foot pulses and formation of hammertoe, may be early signs an increased risk for foot ulcers.

Corns form when the pressure point against the skin traces an elliptical or semielliptical path during the rubbing motion, the center of which is at the point of pressure, gradually widening. They may be dry, waxy, or translucent in appearance and are commonly found over the metatarsal arch on the outside of the fifth toe. If there is constant stimulation of the tissue producing the corn, the
skin may continue to grow as a corn even after surgical removal of an existing corn.

The stiffness of a callus or corn, along with the shear movement and pressure that caused it, may damage the capillaries at the site, causing bleeding within the callous or corn.

**Treatment**

Abnormal anatomy of the feet or an abnormal gait pattern can lead to corn or callus formation, as can bony prominences in the feet and underlying disease (eg, diabetes). Footwear that is too tight or exerts friction at specific points can cause skin thickening that leads to corns and calluses. In such instances, protective pads or skin dressings may be used. Abnormalities in gait or movement that result in increased pressure to specific areas can also cause corns or calluses. This patient population should be evaluated for orthotics. Proper shoes should have a wide and deep toe box, an extra-wide sole, and a low heel counter. Patients who use manual wheelchairs may develop palm calluses, but gloves can be worn to decrease friction and protect skin integrity. Patients who use hand-held assistive devices may benefit from padding their devices to limit the risk of developing calluses. Because diabetes affects the capillaries, thickening of the skin with callus increases the difficulty of supplying nutrients to the skin. Diabetic patients and others with poor circulation or sensation should check their skin often for signs of rubbing and irritation.


**CELLULITIS**

**ESSENTIALS OF DIAGNOSIS**

- An infection of the skin and subcutaneous tissue.
General Considerations

Staphylococcal and streptococcal bacteria are the most common causes of cellulitis. Risk factors include skin ulceration, history of peripheral vascular disease, skin or surgical wounds, and immunosuppressant therapy. Symptomatology includes fever, a sudden and fast-growing rash, and tenderness. The skin at the site is warm, red, irritated, and painful, and may appear tight, glossy, and “stretched.” Lymphadenopathy present in the surrounding area or drainage may be visible on inspection.

Treatment

Antibiotics are the primary treatment, although irrigation and drainage may be warranted. Patients at high risk of developing cellulitis include those with skin trauma, diabetes mellitus, circulatory problems, liver disease, and skin disorders, as well as postsurgical, insensate, or immobile individuals. It is important to monitor the margins of the infected area to ensure adequate treatment.

Fungal Infections

ESSENTIALS OF DIAGNOSIS

- May be superficial, cutaneous, subcutaneous, or systemic.
- High-risk groups include immunosuppressed patients, diabetics, and the very young and elderly.
Fungal infections occur most often in people who are receiving antibiotics, have diabetes, are immunosuppressed, or are very young or elderly. Rehabilitation patients are at risk of developing fungal infections if their skin is not adequately cleaned or if unsanitary therapeutic equipment is shared between patients. In all cases, it is important to maintain contact precautions in the at-risk population.

**Clinical Findings**

Mycoses are classified according to the tissue levels initially colonized: superficial, cutaneous, subcutaneous, or systemic.

Superficial mycoses such as tinea versicolor produce spots that are either lighter than the skin or a reddish-brown. Factors that can cause the fungus to become more visible include high humidity, as well as immune or hormone abnormalities.

Cutaneous mycoses extend into the epidermis. These infections are restricted to the keratinized layers of the skin, hair, and nails. The resulting diseases are often called ringworm or tinea. Athlete’s foot is a common fungal disease that lives on dead skin and can cause skin inflammation and softening of nails.

Subcutaneous mycoses involve the dermis, subcutaneous tissues, muscle, and fascia. These infections are chronic and can be initiated by piercing trauma to the skin, which allows the fungi to enter. Such infections are difficult to treat and may require surgical management.

**Treatment**

Depending on the nature of the infection, a topical or systemic antifungal agent may be used, such as fluconazole, or, for systemic involvement, amphotericin B.

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**VIRAL SKIN DISORDERS (HERPESVIRUS)**
ESSENTIALS OF DIAGNOSIS

- Herpesviruses cause several types of skin infections.
- Cutaneous lesions of cold sores (HSV-1) or genital herpes (HSV-2) appear as watery blisters, which heal with scabbing.
- After an initial infection, the virus can remain dormant for long periods before being reactivated.
- Acute herpes zoster infection is characterized by pain, rash, and cutaneous sensory changes, but may also cause muscular weakness or neuropathy.

General Considerations

Herpes simplex viruses—type 1 (HSV-1), which produces most cold sores, and type 2 (HSV-2), which produces most genital herpes—are widespread and contagious pathogens. After an initial infection, they can remain dormant for long periods. They are transmitted through contact with an infectious area of the skin during periods of reactivation, when the infected person is producing and shedding the virus.

Clinical Findings

Symptoms of herpes simplex virus infection include watery blisters on the skin or mucous membranes that heal with a characteristic scab. However, mild or atypical symptoms may also occur. Acute herpes zoster is characterized by pain, rash, and cutaneous sensory changes. Some patients develop muscular weakness 2–3 weeks after the appearance of the rash. Herpesvirus infection can cause neuropathy. Although motor recovery is slow it is typically near-complete. Electrodiagnostic testing shows axon degeneration in a myotomal distribution.

Treatment

Antiviral medications such as acyclovir and famciclovir can reduce the
frequency, duration, and severity of outbreaks caused by herpesvirus infections. Topical acyclovir is ineffective in the treatment of herpes zoster skin manifestations; however, the oral form (IC Acyclovir) helps relieve symptoms. Oral famciclovir and valacyclovir are also effective. Immunocompromised patients should be treated with intravenous acyclovir.

SKIN TRAUMA

General Considerations

Skin trauma can occur in the form of cuts, burns, breakdown, or other injury. Chronically ill patients who are unable to weight shift frequently develop skin breakdown as a result of shearing forces at pressure points. Burns can be of various origins—thermal, electrical, chemical, and mechanical—and can cause varying degrees of damage to the body, and are described in detail in Chapter 25.

In a first-degree burn, damage is limited to the epidermis; the skin becomes moist and red and is painful for a short period of time. A second-degree, or partial-thickness, burn affects the epidermis and dermis, and may cause scarring. A third degree, or full-thickness, burn affects all the tissue layers and requires skin grafting to restore the integrity of the skin. Third-degree burns are initially not as painful as other burns, because nerve endings are also destroyed.

Treatment

There is some evidence that vitamin C and zinc supplementation aid in wound healing. In patients with skin breakdown or pressure sores, prealbumin levels should be monitored at least daily, and weight should be shifted and wounds monitored regularly. Bony prominences should be monitored for breakdown, especially in patients who are thin and frail.

Orthoses (eg, Multi Podus boots), splints, weight shifting, and resting limbs on pillows can help relieve focal pressure to decrease the risk of skin trauma. In patients with clenched fists, nail length should be monitored to prevent palmar breakdown.

Detailed discussion of burn management appears elsewhere (see Chapter 25). Scar maturation can require up to 1½ years, and measures to minimize scarring are ongoing. Application of pressure (25 mm Hg, 23 hours a day) is needed to prevent contracture of a scar. Patients should be positioned in extension and
abduction while maintaining range of motion. Patients who are not able to comply with positioning may require splinting. Owing to increased metabolic needs, burn patients may require additional pain medication to obtain sufficient pain relief. Nutritional intake should be evaluated carefully and changes implemented, as necessary, in response to positive nitrogen balance, increased metabolic rate, and possible loss of fat cells. Osteophytosis is often found at the olecranon or coracoid process. During skin healing the skin pulls a joint into hyperextension, which can cause joint subluxation.


ENDOCRINE & METABOLIC DISORDERS

DIABETES MELLITUS

ESSENTIALS OF DIAGNOSIS

- Elevated fasting blood glucose level ≥ 126 mg/dL, random glucose level ≥ 200 mg/dL along with hallmark symptoms (polyuria and polydipsia), or 2-hour glucose level ≥ 200 mg/dL after a 75-g glucose load.
- Slightly elevated blood glucose levels are permissible in diabetic patients who are undergoing acute rehabilitation as this is offset by an increase in the baseline metabolic rate.

General Considerations

Classification of diabetes mellitus has continued to evolve, from a focus on treatment to one that reflects the mechanism of dysfunction. Currently four classes are differentiated: (1) Type 1 diabetes, which is associated with &-cell
destruction, usually immune mediated. Most patients with this type of diabetes have an absolute deficiency of insulin and are prone to ketosis. (2) Type 2 diabetes, in which patients are insulin-resistant, eventually developing an insulin secretion problem. (3) Diabetes that results from endocrinopathies, pancreatic exocrine defects, infectious diseases, and genetic defects. (4) Gestational diabetes, affecting women during pregnancy. As with other forms of diabetes, prevalence of gestational diabetes has been increasing over the past 20 years.

Clinical Findings

Elevated glucose levels obtained and reconfirmed through any of the following studies are diagnostic: an elevated fasting blood glucose level of 126 mg/dL or greater, random glucose level of 200 mg/dL or greater along with hallmark symptoms (polyuria and polydipsia), or 2-hour glucose level of 200 mg/dL or higher after a 75-g glucose load.

Treatment

Guidelines for exercise prescription in most patients with type 1 or 2 diabetes include daily exercise for 20–30 minutes. For patients with non–insulin-dependent diabetes mellitus, caloric expenditure should be maximized if obese. Disease-specific precautions include good foot hygiene and care when prescribing & blockers, as these agents can interfere with the patient’s ability to identify hypoglycemic symptoms. Thiazide-like diuretics and & blockers are more likely to provoke hyperglycemia when compared with drugs that block the renin–angiotensin system. Calcium channel blockers may predispose patients to excessive heating and can cause problems in those with diabetic peripheral neuropathy. Neuropathy, arteriosclerosis, and microvascular disease all increase the risk of chronic wounds, which can result in amputation. This patient population is also at risk for retinopathy, which places them at greater risk for falls.

Patients with insulin-dependent diabetes mellitus should have good blood glucose control prior to starting an exercise program. Exercise is contraindicated if fasting serum glucose is greater than 400 mg. Exercise-induced hypoglycemia is the most common problem in exercising patients with diabetes; this can last for up to 6 hours after exercise.

Hyperthyroidism is a condition of the thyroid gland in which too much thyroid hormone is produced. Symptoms include difficulty with concentration, fatigue, heat intolerance, low blood glucose, low blood pressure, muscle aches, and increased appetite. Blood tests showing low thyroid-stimulating hormone (TSH) and high triiodothyronine (T3) and free thyroxine (T4) levels are diagnostic.

Treatment includes antithyroid medications, radioactive iodine, or surgery to remove the gland. Thyroid storm is a life-threatening emergency that can occur in patients with untreated or undertreated hyperthyroidism. Symptoms are severe and include agitation, confusion, tachycardia, and sweating. Immediate action, including administration of antithyroid medication, is required.

Unexplained proximal muscle weakness can be a sign of thyroid dysfunction and should trigger evaluation of the TSH level. There is an increased risk of osteoporosis in patients with hyperthyroidism, and therefore an increased risk of fracture.
Hypothyroidism is a condition of the thyroid gland in which not enough thyroid hormone is produced. Symptoms include sensitivity to cold, constipation, depression, fatigue, weakness and weight gain, slow speech, hoarseness, and musculoskeletal complaints, including muscle weakness. Musculoskeletal complaints include myalgia, weakness, stiffness, cramps, early fatigability, and muscular hypertrophy. Unexplained proximal muscle weakness should prompt evaluation of the TSH level, as this can be a sign of hypothyroidism or, conversely, hyperthyroidism (see earlier discussion). Hoffmann’s syndrome is a type of hypothyroidism that initially causes proximal weakness and hypertrophy of muscles, with later neurologic manifestations of hypothyroidism.

Blood tests showing low TSH, T₃, and free T₄ levels confirm the diagnosis. Treatment consists of thyroid replacement therapy. Myxedema coma is a rare complication of hypothyroidism in which symptoms of altered mental state and hypothermia develop rapidly in response to a stressor, often in elderly patients. This is a life-threatening medical emergency and requires administration of oxygen, thyroid hormones, and other agents to stabilize physiologic function.
Primary headaches include migraine, tension, and cluster headaches. Occipital neuralgia is a paroxysmal shooting pain in the dermatomes of the nervus occipitalis major or nervus occipitalis minor, or both. The pain originates in the suboccipital region and radiates over the vertex. This diagnosis can be confirmed with a diagnostic block. Cervicogenic headache is a unilateral head pain that is triggered by specific pressures or positioning. Treatment options include physical or manual therapy, preventive medicines, anesthetic blocks, injection procedures, and surgery.

A migraine is a severe headache that occurs with nausea, vomiting, or
phonophobia and photophobia. This headache starts as a throbbing pain and can last up to 48 hours. Other warning signs include yawning, difficulty concentrating, nausea, and trouble finding the right words. An aura may or may not be present. More common in women, migraines affect blood flow in the brain. Anxiety, odors, caffeine withdrawal, hormone levels, or lack of sleep may trigger migraine attacks.

Tension headaches occur when the muscles that cover the skull are stressed or spasm, causing pain. Common sites are the trapezius, levator scapulae, scalene, and temporal muscles. Triggers include physical or emotional stress. Tension headaches are the most common type of headache and can be treated with various modalities, including stretching, analgesics, trigger point injections, and botulinum toxin.

### Secondary Headaches

Secondary headaches are caused by a structural problem in the head or neck, including masses, bleeding, meningitis, or encephalitis. Secondary headaches resulting from head and neck trauma can cause bleeding between layers of the brain or nonbleeding injuries, such as concussions or whiplash. Such headaches may be a result of stroke, transient ischemic attack, arteriovenous malformation, aneurysm, or temporal arteritis. In cranial neuralgia, headaches occur because nerves in the head and neck become inflamed. Changes in the body environment, including hypertension, dehydration, hypothyroidism, renal disease, ear–nose–throat problems, or psychiatric disorders can also cause headaches. To diagnose secondary headaches, blood tests, head imaging, or lumbar puncture may be warranted. Attention should be paid to any patient who says that he or she is experiencing “the worst headache of my life,” as this can be a clue to a subarachnoid hemorrhage.


Tremor is an involuntary, often rhythmic oscillation that affects a patient’s movements.

Physical findings and electrodiagnostic testing are used to differentiate the underlying cause.

## General Considerations

Tremor is a component of several disorders that are prevalent among patients who are seen in both outpatient and inpatient rehabilitation settings. These involuntary, often rhythmic oscillations range from subtle to severe in their effect on patients’ movements but, when significant, can impair a patient’s ability to engage in activities of daily living and self-care.

Many different types of tremors are differentiated, including essential, resting, dystonic, psychogenic, and intention. Most tremors are caused by a problem in the brain, including multiple sclerosis, traumatic brain injury, and neurodegenerative diseases; these types of tremors, and their management, are described in earlier chapters. Amphetamines, corticosteroids, psychiatric medications, alcohol abuse or withdrawal, mercury poisoning, an overactive thyroid, or liver failure can also cause tremors. Over 50% of essential tremors are hereditary. An essential tremor can be exacerbated by stress, fever, physical exhaustion, or hypoglycemia. An example of a resting tremor is the pill-rolling tremor seen in patients with Parkinson’s disease. This tremor starts on one side of the body and progresses to the other side.

Dystonic tremors, seen in patients with dystonia, occur irregularly and are relieved by rest. Touching the affected muscle may reduce the severity. Psychogenic tremors are sudden in onset and remission, and the tremor disappears with distraction. Orthostatic tremors occur in legs and trunk immediately after standing. Intention tremors occur at the end of purposeful movement from point to point and are the result of cerebellar damage. This type of tremor is minimally treatable with medications or surgery, but wrist weights can be helpful.
Physical findings and electrodiagnostic testing are helpful in identifying tremors and differentiating their underlying causes. Electromyographic testing confirms the neurologic activity involved in tremor initiation, showing simultaneous bursts produced from simultaneous contractures of agonist and antagonist.

**Treatment**

The treatment of tremors includes β-blockers, primidone, anticholinergic drugs, deep brain stimulation to the thalamus, and surgical thalamotomy. Alcohol can decrease tremors, but rebound tremors may manifest. For details about parkinsonian tremor, and its management, see Chapter 19.

Safety precautions should be implemented for people with tremors. For instance, they should not use glass cups, and their cups should be half filled to avoid injury or spillage. If lower limb tremors are present, assistive devices should be used to prevent the risk of falls. Blood glucose levels should always be checked in a patient with a new tremor as hypoglycemia may be causative.


**SEIZURES**

**ESSENTIALS OF DIAGNOSIS**

- Common sequela after stroke or head trauma.
- Post-stroke seizures are associated with older age, confusion, and large parietal or temporal hemorrhages.
- Risk factors for post-traumatic seizures include cortical lesions, bilateral parietal contusion, dural penetration, intracranial operation, and multiple subcortical contusions.
General Considerations

As discussed in Chapter 36, seizures are a common sequela of stroke or head trauma. They are also associated with older age, confusion, and large parietal or temporal hemorrhages. Most seizures are tonic–clonic. Recurrent seizures are a potentially life-threatening complication of stroke as they can cause elevated intracranial pressure. If a patient experiences new seizures, benzodiazepines are the first-line agent. If there is no response to benzodiazepines, anticonvulsants should be used. An early seizure is one that occurs 1–2 weeks after a stroke; after that period it is regarded as late. Stroke patients requiring inpatient rehabilitation have a higher probability of developing seizure than those in the general stroke population.

Post-traumatic seizures are classified as partial (simple partial if consciousness is preserved; complex partial if consciousness is impaired) or general (grand mal or tonic–clonic). Most post-traumatic seizures are simple partial seizures. An immediate seizure is one that occurs in the first 24 hours postinjury. An early post-traumatic seizure occurs within the first week; one that occurs beyond 1 week is deemed late. The incidence of post-traumatic seizure varies depending on the severity of injury, time since injury, and risk factors related to duration of loss of consciousness (LOC). Patients with LOC lasting 30 minutes to 24 hours or skull fracture have a greater risk of post-traumatic seizure. Most of these seizures occur 1–3 months after injury. Risk factors include cortical lesions, bilateral parietal contusion, dural penetration, intracranial operation, and multiple subcortical contusions. There is a greater risk of development of post-traumatic seizures in the first 2 years postinjury. Seizure prophylaxis is recommended for 10 days if the patient has not had a seizure. Therapeutic anticonvulsant medication is used in patients who experience a late seizures.

Treatment

Although all anticonvulsants can cause sedation and cognitive deficits, carbamazepine and valproic acid are preferred in patients with traumatic brain injury as these agents have a better side effect profile. Long-term phenytoin use is associated with adverse cognitive effects.

General seizure care protocols are applicable to most patients in rehabilitation settings. A patient who is having a seizure should be monitored carefully for aspiration or increased temperature. Signs of neurologic
complications or Todd’s paralysis should be evaluated. Additional complications include rhabdomyolysis, lactic acidosis, neurogenic pulmonary edema, and hypoglycemia. Readers are referred to other sources, and to Chapter 36, for additional information.


**PSYCHIATRIC DISORDERS**

**DEPRESSION**

**ESSENTIALS OF DIAGNOSIS**

- Depression maybe a manifestation of systemic disorder.
- Post-stroke depression may be caused by catecholamine depletion.

**General Considerations**

Depression is a state of low mood and aversion to activity that can affect a person’s thoughts, behavior, feelings, and physical health. Symptoms include feeling sad, anxious, helpless, worthless, guilty, irritable, or restless. People with clinical depression may experience anhedonia, eating disorders, and problems in concentrating, remembering details, or making decisions; and may contemplate or attempt suicide. Insomnia, excessive sleeping, fatigue, loss of energy, pain, or digestive problems that are resistant to treatment may be present.

The incidence of major depression in the elderly varies from 16% to 30%. Depression in the elderly is often linked to progressive health problems and isolation, particularly after the loss of a spouse. Depressed mood is a normal reaction to certain life events, such as a recent amputation or complete spinal cord injury. Patients with depression that has an identifiable precipitator may
benefit from meeting with a psychologist, neuropsychologist, or religious leader, who can assist them in accessing the resources or treatment they may need.

Several psychiatric syndromes feature depressed mood as a main symptom. These include major depressive disorder, bipolar disorder, borderline personality disorder, and adjustment disorder. Depressed mood can also be associated with chronic disease states and exacerbations of systemic diseases, such as celiac disease, systemic lupus erythematosus, and others. It is also linked to anemia, chronic fatigue syndrome, fructose malabsorption, and lactose intolerance.

Post-stroke depression is reported in about 40% of stroke patients and their caregivers and is most prevalent at 6 months to 2 years. In patients, post-stroke depression may be related to catecholamine depletion or to the psychological response to personal loss. It is important to address this component in the patient’s care, as ongoing depression is associated with poorer functional outcomes after stroke.

Standardized questionnaires such as the Beck Depression Inventory and Hamilton Rating Scale for Depression may be used to evaluate patients with depressive symptoms.

**Treatment**

A range of treatment options is available for managing depression in various patient populations and settings. These include psychotherapy, pharmacotherapy (antidepressants), electroconvulsive therapy, music therapy, art therapy, group therapy, animal-assisted therapy, and light therapy. For patients with prolonged depression, trazodone and selective serotonin reuptake inhibitors or tricyclic antidepressants with the lowest anticholinergic effects may be helpful.

Patients should be encouraged to participate in a consistent physical exercise program to the extent possible, as this has been consistently linked with improved mood scores. Maintaining a gratitude journal has also shown benefit.

A maladaptive pattern of use of a substance that occurs on a continuum from mild to severe.

Patients may have underlying mental health issues, manifest co-addictions to multiple substances, or have problems with impulse control.

Tolerance occurs when more of the substance is needed in order to produce desired effects.

Substance abuse is present when an individual continues to use alcohol or drugs despite problems related to the substance. Patients with a substance use disorder demonstrate a maladaptive pattern of substance use that leads to significant impairment or distress. Tolerance occurs when the central nervous system requires more of the substance to produce the desired effect. When this point is reached, stopping the substance will cause withdrawal.

The high rates of suicide in alcoholics and drug abusers have been linked to physiologic distortion of brain chemistry and social isolation. Additionally, amphetamine, hallucinogen, and cocaine abuse have all been shown to induce psychiatric disorders. Withdrawal symptoms may continue for months after an individual stops taking benzodiazepines. Cannabis may trigger panic attacks and cause a dysthymic state. Severe anxiety and depression are commonly induced by sustained alcohol abuse.

Suspicion of substance abuse in a patient should trigger a thorough evaluation to identify possible coexisting psychological or physical disorders. Patients with a substance use disorder frequently have underlying mental health issues, manifest co-addictions to multiple substances, or have problems with impulse control. Treatment is multifaceted and usually includes cognitive–behavioral, family, or group therapy. Replacement therapies, such as buprenorphine and methadone, and antagonist medication are also used in the treatment of some patients with alcohol or opiate addiction.

FATIGUE

ESSENTIALS OF DIAGNOSIS

- Defined by exhaustion or lack of energy exacerbated by activity.
- Can be associated with muscle weakness, electrolyte abnormalities, and a vast array of systemic illnesses.

General Considerations

Fatigue is classified in one of three ways: (1) secondary to a medical condition, (2) physiologic (caused by an imbalance in energy expenditure; relieved with rest), or (3) chronic (lasting more than 6 months; not relieved with rest). When evaluating a patient with fatigue, it is necessary to differentiate this symptom from tiredness. Tiredness is characterized by the ability to be temporarily aroused by activity, whereas symptoms in persons with fatigue will worsen with activity. In the rehabilitation population, physiologic fatigue is commonly caused by a combination of overexertion and deconditioning.

Notable red flags associated with fatigue include unintentional weight loss, night sweats, lymphadenopathy, melena or hematochezia, hemoptysis, focal neurologic findings, sleep apnea, new cardiopulmonary symptoms, and inflammatory arthritis or vasculitis. Should symptoms persist for more than 1 month or if any red flags are present, further workup is warranted. In general, fatigue tends to be transient; less than 50% of patients actually receive a diagnosis confirming the etiology of their fatigue. However, the physician must be mindful that fatigue can persist, especially in the patients with brain injury, which can hinder activities of daily living and patients’ progress toward greater functional mobility.

Treatment
The treatment of fatigue is dependent on the patient’s primary diagnosis. Although fatigue is the primary complaint for an average of 5–7% of primary care visits, about 75% of these cases are single episodes not requiring specific followup. In other words, even without treatment, the great majority of patients improve with time. There are, of course, certain conditions in which fatigue is a concerning symptom, necessitating followup or treatment (particularly when associated with increasing intensity of fatigue, or other red flag symptoms noted earlier). The American Academy of Family Physicians recommends that exercise be prescribed for all patients with fatigue regardless of the etiology, as there is no evidence to suggest that exercise might cause clinical decline.

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**WEIGHT LOSS OR GAIN**

### General Considerations

In the evaluation of weight loss or gain in the primary care population, the physician must first consider whether the change is intentional. Intentional weight loss or gain may be associated with psychological conditions (anorexia nervosa, bulimia nervosa, binge eating) or psychosocial factors. Unintentional weight changes, however, often require further evaluation. In patients with physical disabilities, exercise can often be a challenge; weight gain is often an unfortunate consequence.

Significant weight loss or gain can lead to hormonal imbalances resulting in bone demineralization and osteoporosis, amenorrhea in women, muscle wasting, cardiovascular dysfunction, and death. Considerable weight gain and an associated sedentary lifestyle significantly increase the risk of developing osteoporosis.
Treatment

Significant and intentional weight loss or gain often requires psychological evaluation. For those with unintentional weight changes, however, a definitive diagnosis is needed prior to treatment initiation. Weight gain as a result of a medication adverse side effect may prompt a physician to look for medication alternatives. In the case of unintentional weight loss, several medications have been cited as potentially beneficial although not studied in all populations with weight loss. These include mirtazapine, dronabinol, megestrol, and oxandrolone.


INSOMNIA

ESSENTIALS OF DIAGNOSIS

- Difficulty initiating or maintaining sleep, or both, or nonrestorative sleep.
- Difficulty sleeping while in the hospital does not equate with insomnia.
- Occurs as a side effect of some drugs (eg, opioids decrease rapid eye movement sleep).

General Considerations
Insomnia is defined as difficulty initiating or maintaining sleep, or both, or nonrestorative sleep, associated with impairments of daytime functioning or marked distress for more than 1 month. It is categorized as transient, acute, or chronic.

Insomnia may have a multitude of causes. Among these are circadian rhythm disturbance (eg, shift work or jet lag), psychoactive drug use, pain, stress, anxiety, fluoroquinolone toxicity, restless legs syndrome, hormone shifts, dementia, neurologic disorders, brain lesions, or a history of traumatic brain injury. Sleep-onset insomnia can be indicative of anxiety disorders or the delayed sleep phase disorder. Nocturnal awakenings can be indicative of pain disorders or illnesses such as clinical depression.

The ability to sleep for long periods decreases with age. Medical conditions such as hyperthyroidism or polyuria may cause insomnia. Prescription sleep aids can produce rebound insomnia. Poor sleep can lead to irritability and decreased concentration and memory.

**Treatment**

Nonpharmacologic treatment strategies are superior to hypnotic medication for insomnia. The strategies include cognitive–behavioral therapy, attention to sleep hygiene, stimulus control, sleep-restriction therapy, patient education, and relaxation therapy. Reducing the temperature of blood flowing to the brain slows the brain’s metabolic rate, resulting in improvements in duration as well as quality of sleep. Electroencephalographic sleep studies can aid in diagnosis of underlying medical conditions.

Medications that have been used in the treatment of insomnia include benzodiazepines, opioids, diphenhydramine, melatonin and cyproheptadine. Many of these agents have side effects that can limit their utility in rehabilitation patients, particularly those who are elderly.

Benzodiazepines have a side effect profile that includes daytime fatigue, cognitive impairments, falls, and fractures—side effects to which elderly people are more sensitive. Some benzodiazepines have demonstrated effectiveness in sleep maintenance in the short term but in the longer term are associated with tolerance and dependence. Opioid medications can fragment sleep and decrease rapid eye movement sleep.

Although diphenhydramine is widely used as a nonprescription sleep aid, it has several drawbacks when used long term. Efficacy decreases over time, it is associated with next-day sedation, and anticholinergic side effects may appear...
with continued use. It can also induce dependence and rebound effects upon cessation. Diphenhydramine is not appropriate for use in geriatric patients.

Melatonin treats insomnia without altering the sleep pattern, and it does not impair performance related skills. Cyproheptadine enhances sleep quality, quantity, and appetite.


GASTROESOPHAGEAL REFLUX DISEASE

ESSENTIALS OF DIAGNOSIS

► Stomach contents leak from the stomach back into the esophagus, causing symptoms of heartburn, hiccups, hoarseness, or difficulty swallowing.
► Diagnosis is confirmed by esophagogastroduodenoscopy, barium swallow, esophageal pH monitoring, or esophageal manometry.

General Considerations

In gastroesophageal reflux disease (GERD), the stomach contents leak from the stomach back into the esophagus. Risk factors include alcohol use, hiatal hernia, obesity, pregnancy, scleroderma, and smoking; the condition may also be iatrogenic. Patients may complain of heartburn, hiccups, hoarseness, or difficulty swallowing. Among rehabilitation patients, aspirin use may sometimes trigger the development of GERD. Such patients are often newly started on aspirin therapy for stroke, coronary artery disease, or deep vein thrombosis prophylaxis. Aspirin-induced erosion of the gastric lining can cause symptoms of GERD.
Diagnosis can be confirmed using esophagastroduodenoscopy, barium swallow, esophageal pH monitoring, or esophageal manometry.

## Treatment

Treatment includes diet modification, proton pump inhibitors, or H$_2$ blockers. Cautious use of H$_2$ blockers is advised as these agents can cause confusion in geriatric populations. Fundoplication surgery is a treatment option in severe cases.


## PELVIC PAIN

### ESSENTIALS OF DIAGNOSIS

- Causes include dysmenorrhea, endometriosis, pelvic inflammatory disease (PID), ovarian cyst, or torsion.
- Patients with spinal cord injury may be unable to detect pelvic pain and should therefore be monitored routinely.

Female pelvic pain may be caused by visceral pain from a distended bladder or bowel. It may also be due to uterine pain sensitivity, or pelvic girdle pain. During pregnancy there is a loosening of the pelvic ligaments. After pregnancy, women may complain of pelvic pain, resulting from a misalignment of the pelvis.

Secondary dysmenorrhea is diagnosed when symptoms are attributable to an underlying disease, disorder, or structural abnormality either within or outside the uterus. Primary dysmenorrhea is diagnosed when these are not detected. Treatment includes NSAIDs or oral contraceptives.

Endometriosis occurs when cells that make up the lining of the uterus are
present outside the uterine cavity, most commonly on the ovaries. It can be associated with dyspareunia, dysuria, and constipation. Laparoscopy is the gold standard for diagnosis. Treatment options include pain medication, hormones, or a variety of surgical procedures.

PID may cause pelvic pain. The most common causes are infections with *Neisseria gonorrhea* or *Chlamydia trachomatis*; however, normal vaginal flora can be involved. Symptoms in PID range from subclinical to severe. Empiric antibiotic treatment is initiated to avoid serious complications. Acute PID is highly unlikely when recent intercourse has not taken place or an intrauterine device (IUD) is not being used. Pelvic and vaginal ultrasounds are helpful in the differential diagnosis of ectopic pregnancy of over 6 weeks. Although the PID infection may be cured, PID can cause scarring, chronic pelvic pain, and infertility.

Patients with a spinal cord injury may not be able to report pelvic pain, but if they demonstrate signs of autonomic dysreflexia and hypertension a pelvic examination should be performed.


**SEXUAL DYSFUNCTION**

**ESSENTIALS OF DIAGNOSIS**

- Sexual dysfunction is the inability to perform sexual activity, stemming from problems of desire, arousal, or orgasm.
- The cause may be physical, psychological, or pharmacologic in origin.
- Erectile dysfunction is the inability to develop or maintain an erection of the
General Considerations

Sexual dysfunction is defined as difficulty during normal sexual activity, including desire, arousal, or orgasm. Sexual dysfunction disorders are classified into sexual desire, arousal, orgasm, and pain disorders. These disorders have a wide range of causes. A decreased libido can result from low levels of sex hormone, aging, fatigue, pregnancy, medications, or psychiatric conditions. Antihypertensives, antipsychotics, antidepressants, sedatives, narcotics, antacids, nicotine, antihistamines, or alcohol can cause sexual dysfunction.

Erectile dysfunction is the inability to develop or maintain an erection of the penis. It may occur as a consequence of vascular disorders, diabetes mellitus, peripheral vascular disease, nerve injury, hypertension, or substance abuse. Hormone deficiencies, radiation therapy, chemotherapy, mumps, central nervous system tumors, and endocrine disorders may also cause erectile dysfunction. Sickle cell patients may experience priapism.

Orgasm disorders are persistent delays or absence of orgasm following a normal sexual excitement phase. The disorder can have physical, psychological, or pharmacologic origins. It is unclear what causes vaginismus, but it is thought that past sexual trauma is involved. Vulvodynia seems to be related to problems with the skin in the vulvar and vaginal areas.

Treatment

Treatment for male sexual dysfunction includes psychotherapy and lifestyle modifications. Medication options include sildenafil, tadalafil, vardenafil, or intracavernous pharmacotherapy. Treatment for female sexual dysfunction includes psychotherapy, pain relievers, desensitizing agents, lubricants, and hormone therapy. Women with arousal and orgasm disorders may benefit from use of a small vacuum device to increase blood flow to the clitoris and external genitalia.

Among post-stroke patients there is no significant change in sexual interest, but there is a decline in sexual behavior. The decrease in behaviors is attributed to medications such as antidepressants, antipsychotics, anticholinergics, opioids, and γ-aminobutyric acid agonists. Psychological concerns include decreased
sensation and issues relating to the custodial role of the patient’s partner or spouse. Treatment includes evaluation of an organic cause and supportive counseling.

Sexual dysfunction is common in patients with spinal cord injuries. Refer to Chapter 12 for more information. Patients with traumatic brain injury generally have a decrease in sexual function and satisfaction, regardless of where the injury occurs.


HIV INFECTION & AIDS

ESSENTIALS OF DIAGNOSIS

► Blood-borne virus (lentivirus, member of the retrovirus family) that causes acquired immunodeficiency syndrome (AIDS).

► Transmitted typically via unprotected sex, the sharing of intravenous needles, or from mother to child during birth or breastfeeding.

► Signs and symptoms vary depending on clinical stage (acute infection, clinical latency, or AIDS).

General Considerations

Human immunodeficiency virus (HIV) was first identified in the early 1980s when an unusual increase in several rare infections (especially Kaposi’s sarcoma and Pneumocystis carinii jiroveci pneumonia [PCP]), now known to be opportunistic in the setting of HIV, was reported in young homosexual men. Soon after, these infections were also noted in the intravenous drug–using population. Two types of HIV infect humans: HIV-1 and HIV-2. HIV-1 is the more virulent and transmissible, thus it is the cause of the majority of known
infections in the world. The CDC estimates there are 1.2 million people living with HIV in the United States. One in five of these patients are unaware of their status. For this reason, it is important to take a thorough history, specifically inquiring into risk-taking behaviors.

► Clinical Findings

During the acute phase of HIV infection (usually 2–6 weeks after infection), also called acute retroviral syndrome or primary HIV infection, patients may present with flulike symptoms (malaise, sore throat, fevers) or they may be entirely asymptomatic. During this stage, a large amount of virus is produced and the CD4 count can acutely drop. An HIV test is likely to be positive during the acute phase. Enzyme-linked immunosorbent assay (ELISA) is used for screening. If negative, but clinical suspicion remains high, retesting in 1 week is recommended. A positive ELISA should be confirmed with Western blot assay. Several other methods of detection are available, and the reader is referred to other sources for information.

► Complications

The most notable complications in patients with HIV infection are coinfection with hepatitis B or C as well as the many opportunistic infections associated with the virus. The latter include candidiasis, herpes simplex virus infection, PCP, Kaposi’s sarcoma, cytomegalovirus retinitis, and \textit{Mycobacterium avium intracellularare} infection. Additionally, the virus causing HIV as well as the medications used in its treatment may cause myopathy, neuropathy, or both.

► Treatment

Although there is no cure for HIV, the treatments available can lower the viral load to a level that is undetectable. The available medications include nonnucleoside reverse transcriptase inhibitors (NNRTIs), nucleoside reverse transcriptase inhibitors (NRTIs), protease inhibitors, entry or fusion inhibitors, and integrase inhibitors. Often at least three medications from two different classes are used in combination to prevent the virus from becoming immune to the treatment.
Prognosis

Many factors play in role in determining the prognosis of a patient with HIV. These include patient age, presence of coinfections or opportunistic infections, CD4 count, viral load, whether the patient is receiving treatment, and whether he or she is compliant with the treatment regimen. Overall, with the advent of antiretroviral therapy and earlier detection, patients are living longer with HIV.


CLINICAL CONCERNS IN GERIATRIC PATIENTS

OSTEOPOROSIS

ESSENTIALS OF DIAGNOSIS

► Primary risk factors include older age, northern European ancestry, positive family history, thin build, tobacco and alcohol use, and a patient who is a postmenopausal female.

► Laboratory studies may identify hormonal or metabolic conditions associated with secondary causes (eg, Cushing syndrome, hyperthyroidism, hyperparathyroidism, multiple myeloma, and hypogonadism).
Bone mineral density evaluation using dual-energy X-ray absorptiometry (DEXA) is preferred for diagnosis.

A bone mineral density T-score of −2.5 or lower indicates osteoporosis.

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**General Considerations**

Osteoporosis is common in the geriatric population and is generally suspected by clinical presentation. It can be divided into primary and secondary causes. Primary causes reflect predisposing risk factors associated with low bone density, such as older age, northern European ancestry, positive family history, thin build, tobacco and alcohol use, and a patient who is a postmenopausal female. Secondary causes include use of corticosteroids and conditions such as Cushing syndrome, hyperthyroidism, hyperparathyroidism, multiple myeloma, and hypogonadism.

**Clinical Findings**

The most common radiographic findings are multiple vertebral compression fractures. The diagnosis of osteoporosis is made by determining the patient’s bone mineral density (BMD). The following tests can all be used to evaluate bone density: quantitative computed tomography (CT), dual-photon absorptiometry, and dual energy X-ray absorptiometry (DEXA). DEXA is the preferred method—the newest, quickest, least expensive, and most accurate option. Quantitative CT scanning requires a large dose of radiation and is more expensive; and both CT scanning and dual-photon absorptiometry can be time consuming.

The BMD test reports two separate scores, referred to as the T-score and the Z-score, and compares these results with those of normal young healthy bone (T-score) or an age- and sex-matched control (Z-score). A T-score of −2.5 or lower indicates osteoporosis, and one of −1 to 2.5 suggests osteopenia. The Z-score is not used for treatment of osteoporosis but rather to determine whether the patient has accelerated bone loss. It is recommended that all women aged 65 years or older be screened using DEXA scans, and that selective screening be performed for women aged 60–64 years who have predisposing risk factors.
Treatment

Treatment and prevention of osteoporosis in premenopausal women centers on administration of elemental calcium, at a dosage of 1 g/day. For postmenopausal women, it includes administration of both elemental calcium, at 1.5 g/day, and bisphosphonates. The use of hormonal replacement therapy is controversial. Weight-bearing exercises or activities and adequate vitamin D (800 IU/day, needed for adequate calcium absorption) are required for both premenopausal and postmenopausal women.

It should be noted that few randomized controlled trials investigating antiosteoporotic agents with fracture end points have included participants over the age of 80 years. With regard to bisphosphonates, only pivotal trials of risedronate and zoledronic acid showing significant fracture reductions have included participants above this age group. Zoledronic acid has also been shown to reduce the rate of new clinical fractures after repair of a low-trauma hip fracture. More recently, denosumab has been associated with a significant reduction in the risk of new radiographic vertebral, hip, and nonvertebral fractures in women up to the age of 89 years with osteoporosis. Strontium ranelate and teriparatide have shown fracture reductions in populations that have included subjects over the age of 80 years. There has been evidence to show that a combination of calcium and vitamin D reduces nonvertebral fractures in older populations. The role of vitamin D alone is less clear, although there is the suggestion that it may be effective at higher doses.


Meunier PJ, Roux C, Seeman E, et al: The effects of strontium ranelate on the

DEMENTIA

ESSENTIALS OF DIAGNOSIS

- First symptom is forgetfulness.
- Differential diagnosis includes depression, cerebral hemorrhage, medication side effects, acute confusion, and infection.

General Considerations

Dementia is found in 1.5% of people aged 65–70 years and 25% of people aged 85 years and older. Dementia should be differentiated from depression, forgetfulness, cerebral hemorrhage, medication side effects, acute confusion, or infection. Most types are nonreversible. Although Alzheimer’s disease (discussed below) is the leading cause of dementia among adults of all ages, Lewy body disease is the leading cause of dementia in elderly adults. Other causes of dementia include brain injury; tumors; chronic alcohol abuse; changes in blood glucose, sodium, or calcium; low vitamin B\textsubscript{12}; and normal pressure hydrocephalus. Dementia may also be idiopathic.

Clinical Findings

Dementia encompasses deficits in language, memory, perception, emotional behavior, and cognitive skills. It first appears as forgetfulness. As the disorder progresses, patients’ walking speed decreases, and they have increased rigidity and difficulty with activities of daily living. The prevalence of falls and decreased mobility is higher in patients with dementia compared with healthy adults.

Lewy body disease is an umbrella term encompassing two related conditions:
the dementia associated with Parkinson’s disease and Lewy body dementia. Although the initial symptoms vary, later manifestations of these diseases can be difficult to distinguish. In both conditions, patients are hypersensitive to drugs that affect dopamine and cholinergic receptors.

**Treatment**

Intensive exercise and long-term physical therapy have been repeatedly shown to increase mobility and function. Benefits reported include reduction in falls, improved strength, and improved cognition. Caregiver strain can be improved by teaching family members a strength-based approach for selecting, developing, and implementing care goals, and teaching them how to use cognitive rehabilitation skills for addressing care needs.

Treatment of patients with mild to moderate dementia using cholinesterase inhibitors and memantine has resulted in statistically significant but clinically marginal improvement in measures of cognition and global assessment of dementia. There are questionable findings in severe dementia. Nutritional status should be monitored as patients with dementia have an increased risk of poor oral intake. They may also require assistance with personal hygiene and following or remembering commands. Repetition or writing in a journal are adaptive mechanisms that can be used in therapies. Patients usually have problems with poor sleep; however, treatment with commonly prescribed agents is problematic. Diphenhydramine can increase confusion, and temazepam can cause hypotension, tachycardia, and gait ataxia; zolpidem can increase confusion and falls.

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**ALZHEIMER’S DISEASE**

**ESSENTIALS OF DIAGNOSIS**

- Most common form of dementia.
- Usually slowly progressive, but the early-onset form may advance rapidly.
- Patients have difficulty naming items, acquiring new information, processing visuospatial information, performing executive functions, and carrying out activities of daily living.
- Later symptoms include loss of social inhibition, disturbed sleep cycle, and shuffling gait.

**General Consideration**

Alzheimer’s disease has an estimated prevalence of 1% among people aged 60–64 years and 40% among those older than 85. It is the most common form of dementia and generally progresses slowly. Initial symptoms may be subtle, but as the disease advances widespread impact on cognitive function and daily activities becomes increasingly apparent. Senile plaques and neurofibrillary degeneration are characteristically noted in the cortex and gray matter of patients with Alzheimer’s disease.

**Clinical Findings**

Patients have difficulty naming items, acquiring new information, processing visuospatial information, performing executive functions, and carrying out activities of daily living. As the disease progresses, there is a loss of social
inhibition and a disturbed sleep cycle. The confluence of impairments can have a negative impact on patient self-confidence and well-being. A shuffling gait with rigidity may also develop. Early-onset Alzheimer’s disease, before age 60, progresses more quickly. In all cases, the clinician should ensure that neurologic changes are not due to a brain tumor or stroke.

**Treatment**

Treatment goals include managing cognitive–behavioral and sleep problems. The home should be set up to maximize independence. Both pharmacotherapy and psychosocial treatment methods are used. Offering the appropriate support can have a positive effect on patients’ and caregivers’ ability to cope with the disease-related impairments in psychological functioning and on behavior.

**A. Pharmacotherapy**

Medications such as cholinesterase inhibitors (for mild disease) and *N*-methyl-<sub>d</sub>-aspartate (NMDA) inhibitors (for more severe disease) have shown efficacy in patients with Alzheimer’s disease. These agents are used to help slow down the progression of the disease; however, the side effect profile includes stomach distress, diarrhea, vomiting, muscle cramps, fatigue, agitation, and anxiety. Anticholinergic effects are common, and patients who take acetylcholine esterase inhibitors need to be monitored for bradycardia.

**B. Psychosocial Therapies**

Psychosocial methods focus on stimulating the individual’s functioning and preventing further disability through cognitive stimulation and exercise or by the application of compensatory aids. Methods now aim to accommodate the variation in individual needs, desires, and cognitive capacities of people with dementia.

Cognitive training in the early stages of Alzheimer’s disease improves cognition, decision making, and ability to carry out activities of daily living. The interventions should be structured, targeted, and combined with cognition-enhancing medication for maximum benefit. The goals of cognitive rehabilitation need to change as the disease progresses. Cognitive rehabilitation can address a patient’s memory problems by working to improve the current memory skills or by teaching use of compensatory aids.
C. Exercise

Increased physical activity is thought to help prevent Alzheimer’s disease. Regular physical exercise aims to stimulate the individual as an information-processing system and to positively influence the neurophysiologic or pathophysiologic processes underlying the (disturbed) system.


SEQUELAE OF ORGAN TRANSPLANTATION

General Considerations

As organ transplantation gains more success and is more widely practiced, the need for post-transplant rehabilitation will also be greater. Patients undergoing transplantation often endure long hospitalization courses, much of which time is spent confined to bed. This in combination with the use of immunosuppressive agents makes patients vulnerable to deconditioning and to many myopathic and neuropathic processes.

1. Tremors, Myopathy, & Neuropathy

ESSENTIALS OF DIAGNOSIS

- Immunosuppressant agents have neurotoxic properties; mild symptoms include tremors and neuralgia.
- In severe cases patients can have a spectrum of symptoms, including significant weakness, pain, seizures, and encephalopathy.
General Considerations

Post-transplantation neurologic complications are reported in 10–59% of transplant recipients. More specifically, 10–28% of patients using cyclosporine will develop some form of neurotoxicity. In fact, of the immunosuppressant agents used, calcineurin inhibitors are the main generators of neurotoxicity. Others include OKT3 and corticosteroids. Patients undergoing liver transplantation are more likely than any other transplant recipient to experience neurologic complications. Other causes of neurologic complications in the transplant recipient include metabolic abnormalities and infections, specifically opportunistic infections.

Clinical Findings

Clinical findings vary based on the type of neurologic insult experienced by the patient. The most commonly noted finding is tremor, experienced by over 20% of kidney transplant recipients. This tends to abate over time. Cases have been reported in which patients developed symmetric polyneuropathy as a result of axonal degeneration. Additionally, acute encephalopathy, and even cerebellar disorders of ataxia, tremor, hyporeflexia, and weakness, have been noted. Such sequelae, however, are severe and fortunately rare events.

Complications

Often, patients are left with few if any repercussions of neurotoxic events. However, certain phenomena can be debilitating, leaving patients with residual weakness and sensory deficits that can hinder their functional mobility and activities of daily living.

Treatment

The fine tremor associated most often with calcineurin inhibitor use is typically responsive to & blockers. When clinical signs of neurotoxicity are noted, the dosages of immunosuppressive agents may be decreased or one agent may be
exchanged for another in an attempt to quell the symptoms without allowing rejection to take place. Patients with presumed critical illness myopathy or polyneuropathy are treated supportively. Corticosteroids are weaned as soon as feasible, and early mobility with physical and occupational therapies is encouraged.

#### Prognosis

Commonly symptoms diminish or even resolve with decreased immunosuppressive dosages or cessation of the causative agent. For those with myopathies or polyneuropathies, recovery can take weeks to months.

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2. Electrolyte Imbalance

#### General Considerations

Certain electrolyte abnormalities are to be expected after organ transplantation. Hypokalemia, for example, can occur in the setting of any major surgery where fluid shifts occur and large doses of diuretics are used. Hyperkalemia, on the other hand, usually occurs as a result of renal tubular acidosis. This is typically caused by the commonly prescribed calcineurin inhibitors (cyclosporine, tacrolimus) and is often controllable by diet alone.

Hyperglycemia is another anticipated adverse reaction to the calcineurin inhibitors as well as to corticosteroids. This often resolves with cessation or reduction of the causative agent.

Hypomagnesemia is often indicative of a poor nutritional state. Many
transplant recipients have a low serum magnesium level preoperatively, which is then only exacerbated by surgery and often-lengthy hospitalization. Routine monitoring and supplementation are recommended.

Bone disease develops in a large proportion of solid organ transplant recipients, and its consequences can be devastating. Many patients have bone disease prior to the transplantation procedure as a direct result of organ failure. For these patients, the transplantation may actually impart some beneficial effects. For many more, the immunosuppressive agents used to prevent rejection of the new organ ultimately lead to further damage of an already weakened skeleton.

### Treatment

It is recommended that patients undergo a pretransplantation osteoporosis evaluation, including bone densitometry of spine and hip, and thoracic and lumbar spine radiographs. Postoperatively, weight-bearing and strengthening activities should be encouraged. Corticosteroids should be weaned as able. All patients should receive calcium (1000–1500 mg/day) and vitamin D (400–800 IU/day) supplementation, regardless of pretransplantation bone mineral density.


### 3. Inadequate Nutrition

It is recommended that patients undergo a pretransplantation nutrition assessment to address current nutritional deficits, post-transplantation concerns, as well as any organ failure-related symptoms affecting the patient’s current nutritional state. Malnutrition in both underweight and obese transplant recipients can affect wound healing, graft survival, and overall morbidity and mortality. Rejection, infection, hyperglycemia, and poor wound healing all have nutritional implications for the recipient. Lastly, it is important to be mindful that certain dietary products can affect the metabolism of immunosuppressive agents.
and that the medications themselves, by their nature, suppress natural immune responses.

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This chapter was completed prior to the release of the revised manual, DSM 5.