

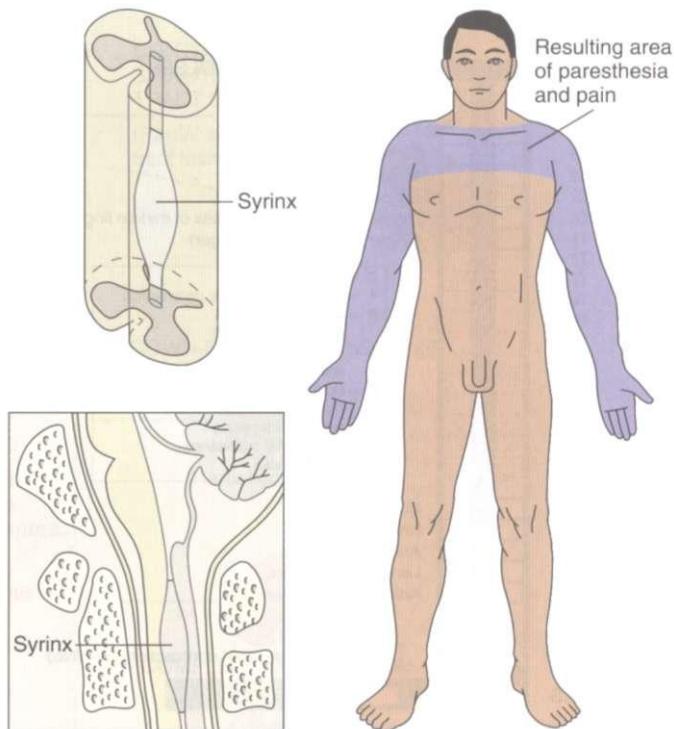
death occurring in the chronic phase (months to years). In the somatosensory system, shrinkage of the dorsal column nuclei and thalamus has been demonstrated in primates following upper limb deafferentation. Thus, both the somatosensory and motor systems are susceptible to atrophy after nervous system damage.<sup>42</sup>

**Neurapraxia.** The syndrome of neurapraxia is of special concern after athletic injury. Affected individuals experience dramatic, although transient, neurologic deficits, including tetraplegia. Transient tetraplegia most commonly occurs as a result of axial loading of the spine. In athletes with narrowing of the anteroposterior diameter of the spinal canal, both hyperextension and hyperflexion can lead to cord compression. This is referred to as the pincer mechanism. In the already stenotic canal, with hyperextension the cord is compressed between the posterior inferior margin of the superior vertebral body and the anterior superior aspect of the spinolaminar line of the subjacent vertebra. Conversely, in hyperflexion the cord is compressed between the anterior superior aspect of the spinolaminar line of the superior vertebra and the posterior superior margin of the inferior vertebra. In both cases, this sudden decrease in anteroposterior diameter of the spinal canal results in compression of the spinal cord. Many attempts have been made to quantitate the level of risk to these individuals from continued athletic participation; however, considerable controversy still exists.<sup>27</sup>

**Syringomyelia.** One type of pathologic condition that can appear over time in the spinal cord related to trauma is syringomyelia. It is a clinical syndrome that results from cystic cavitation and gliosis of the spinal cord. This is reported to occur in close to 2% of persons with paraplegia and in 0.2% of quadriplegic individuals. In the chronic spinal cord lesion, the cysts may continue to develop, become tubular in shape, like that of a syrinx, and extend over several spinal levels (Fig. 34-8). Post-traumatic syringomyelia can develop up to 30 years after the initial lesion, but most commonly occurs within 4 to 9 years after trauma. One mechanism of syrinx formation is an initial hematoma followed by resorption and formation of a cyst cavity.<sup>30</sup>

In some cases, there are multiple cavities. The cavity may occupy almost the entire cross-sectional area of the cord, compressing the posterior columns. As the cyst develops, usually below the level of the initial lesion, there can be significant pain as a result of the compromise of the central spinal cord structures, such as the substantia gelatinosa and the posterior root entry zone.<sup>73</sup> The spinothalamic tracts are involved, which can result in the sharp pain that is often the first presenting symptom. There can be lower motor neuron dysfunction, causing weakness, atrophy, and loss of reflex activity. Sensory loss is common, and the sympathetic nervous system can become involved, resulting in syndromes such as Horner's syndrome.<sup>48</sup>

The thoracic area of the spine is the most common site for the syrinx to develop, with descending and ascending fibers running in the walls of the cavitated lesions. The size and extent of the syrinx are represented by the symptoms, but because of location below the site of the lesion, changes are not easily recognized. The syringomyelia may



**Figure 34-8**

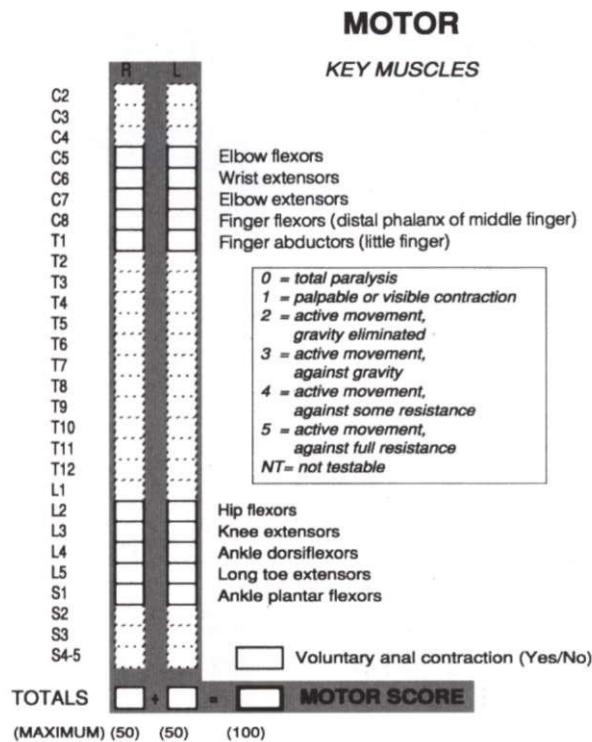
The syrinx formed in the late stages of spinal cord injury as a part of syringomyelia.

be responsible for the spasms, phantom sensations, reflex changes, and autonomic visceral phenomena that may occur. Scoliosis can result from the loss of input to the paraspinals.<sup>44</sup> Anything that blocks the free flow of cerebrospinal fluid (CSF) can keep this fluid from moving normally in and out of the head. Pressure can build up in the syrinx, causing expansion and possible rupture, damaging normal spinal cord tissue and injuring nerve cells. Many people with posttraumatic syringomyelia do not develop any symptoms until midlife or later.<sup>32</sup>

Syringomyelia can be a very disabling condition. Spasms, phantom sensations, and autonomic visceral (organ) changes can occur. Sexual dysfunction, muscle spasticity, and loss of bowel or bladder control can develop. The first symptom may be sharp pain. Muscle atrophy, stiffness, and weakness of the neck, back, shoulders, arms, or legs, along with loss of reflexes, are common. Symptoms may be distributed like a cape over the shoulders and back. Headaches and loss of sensation (pinprick and temperature) in the hands may be reported. The symptoms may only occur on one side of the body, depending on where the syrinx develops.

### Clinical Manifestations

**Level of Injury.** SCIs are named according to the level of neurologic impairment. Differences may exist in the motor versus sensory levels identified. The American Spinal Injury Association (ASIA) has created standards for assessment and classification that are widely used (Figs. 34-9 and 34-10). The sensory examination consists of testing 28 dermatomes on each side of the body using

**Figure 34-9**

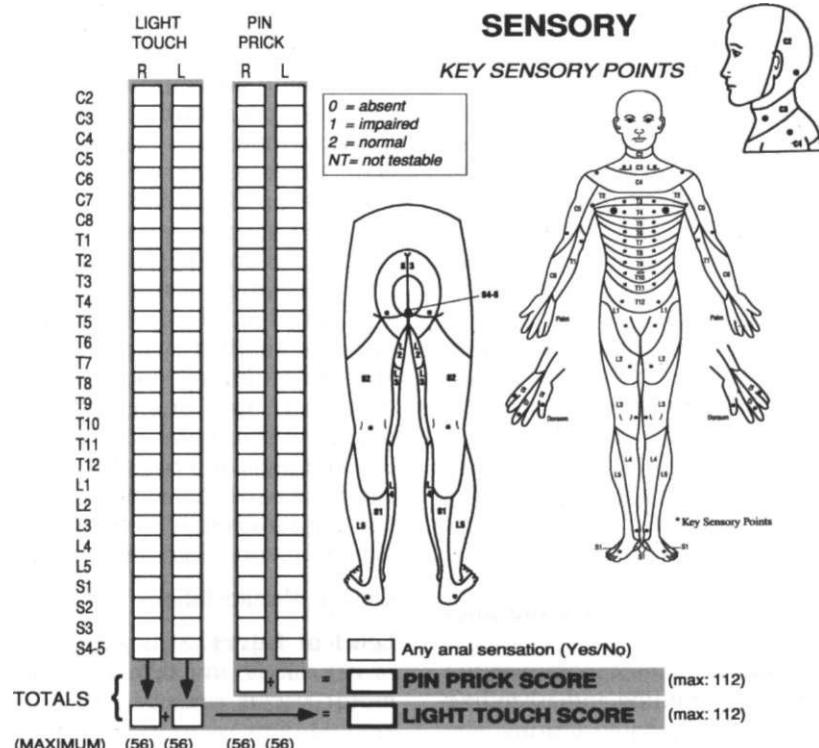
American Spinal Injury Association (ASIA) motor assessment form.  
(Courtesy American Spinal Injury Association International, Atlanta.)

pinprick and light touch, with the sensation in each then scored as follows: 0 = absent, 1 = impaired, 2 = normal. Sensation of the external anal sphincter is tested as yes or no. The level of inquiry reflects the most caudal level of the spinal cord that exhibits intact sensory and motor functioning.<sup>21,26</sup>

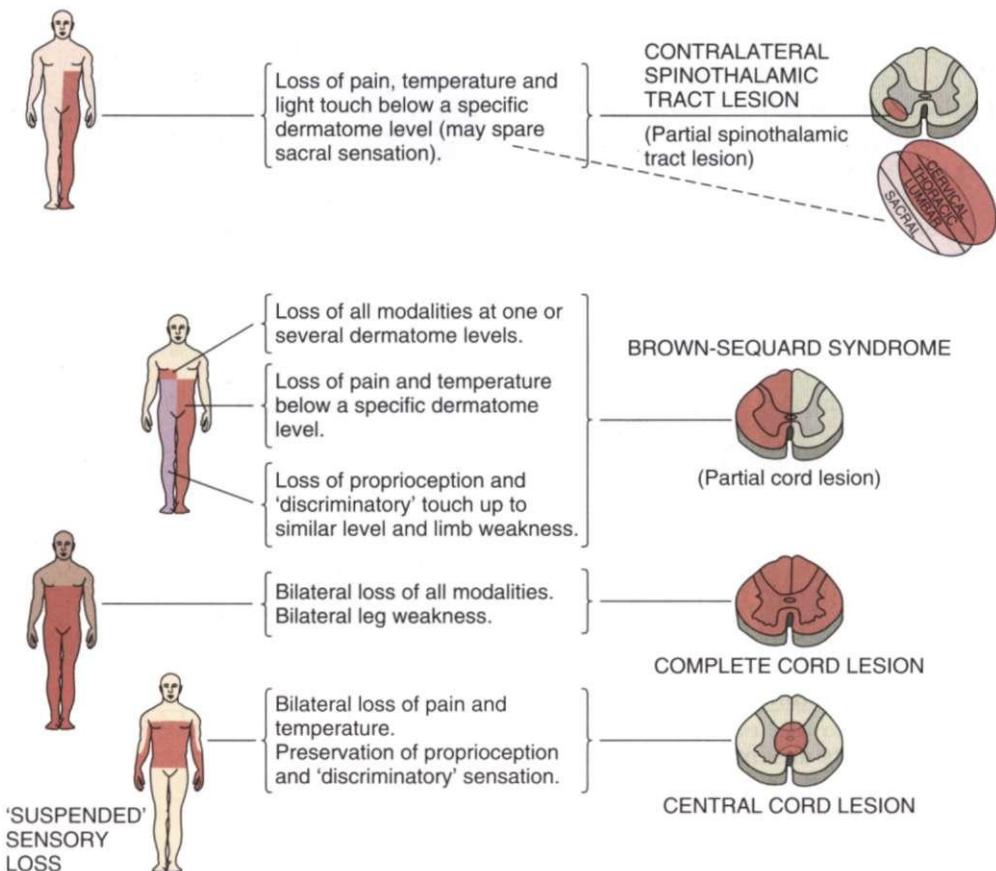
Identification of motor impairment is more problematic, given the dual innervation of many muscles. The strength of a given muscle is a reflection of the functioning of two or more cord segments. Loss of innervation from a spinal cord level results in weakness of the dual innervated muscle. To determine the level of innervation, the therapist looks for the muscle that has a two thirds muscle strength with a one third muscle strength in the next most rostral muscle. Volitional contraction of the anal sphincter is also noted. Asymmetric damage may result in different levels of neurologic impairment on the left versus the right side. The lesion can be then be reported as, for example, a right C5 and a left C6 lesion.

Lesions are reported as *complete* when there is complete loss of sensory and motor function below the level of the lesion. Complete lesions are a result of spinal cord transection, severe compression, or extensive vascular dysfunction.

*Incomplete* lesions are the partial loss of sensory and motor function below the level of the injury. Incomplete lesions often occur when there is contusion produced by bony fragments, soft tissue, or edema within the spinal canal. The resulting motor or sensory function is called

**Figure 34-10**

American Spinal Injury Association (ASIA) sensory assessment form. (Courtesy American Spinal Injury Association International, Atlanta.)

**Figure 34-11**

Spinal cord syndromes. Patterns of sensory loss and weakness. (From Lindsey KW, Bone I, Callander R: *Neurology and neurosurgery illustrated*, New York, 1986, Churchill Livingstone, p 188.)

**Box 34-1****AMERICAN SPINAL INJURY ASSOCIATION (ASIA) CLASSIFICATION OF SPINAL CORD INJURY-RELATED IMPAIRMENT**

- A = Complete: no sensory or motor function preserved in the segment S4-S5
- B = Incomplete: sensory but no motor function preserved below the neurologic level; sensory function extends through segment S4-S5
- C = Incomplete: motor function preserved below the neurologic level, and the majority of key muscles below the neurologic level have a muscle grade of less than 3
- D = Incomplete: motor function preserved below the neurologic level, and the majority of key muscles below the neurologic level have a muscle grade greater than or equal to 3
- E = Normal: sensory and motor functions normal

*sparing.* Box 34-1 describes the ASIA method for describing impairment related to SCI.

**Spinal Cord Injury Syndromes.** Within the category of incomplete spinal cord lesions are the recognizable syndromes that have been identified. Several syndromes are illustrated in Fig. 34-11.

*Brown-Sequard syndrome* is characterized by damage to one side of the spinal cord. The most common causes are

stab and gunshot wounds. Loss of the entire hemisection of the spinal cord is rare; the natural lesion is always irregular. There is weakness ipsilateral to the lesion. Lateral column damage results in abnormal reflexes, including a positive Babinski's sign, and clonus. Often there is ipsilateral spasticity in the muscles innervated below the lesion. As a result of dorsal column damage, there is loss of proprioception, kinesthesia, and vibratory sense. On the contralateral (opposite) side, there is pain and temperature loss starting a few levels below the lesion. The lateral spinothalamic tract ascends on the same side for several segments before crossing, giving rise to the discrepancy between the level and contralateral signs.

*Anterior cord syndrome* is frequently associated with flexion injuries and is often the result of loss of supply from the anterior spinal artery. Damage to the anterior and anterolateral aspect of the cord results in bilateral loss of motor function and pain and temperature sensation because of interruption of the anterior and lateral spinothalamic tracts and corticospinal tract.

*Central cord syndrome* is a result of damage to the central aspect of the spinal cord, often caused by hyperextension injuries in the cervical region. There is characteristically more severe neurologic involvement in the upper extremities than in the lower extremities. Peripherally located fibers are not affected, and therefore function is retained

in the thoracic, lumbar, and sacral regions, including the bowel, bladder, and genitalia.

*Posterior cord syndrome* is extremely rare, with preservation of motor function, pain, and light touch sensation. There is loss of proprioception below the level of the lesion, leading to a wide-based steppage gait.

*Conus medullaris syndrome* and *cauda equina syndrome* reflect damage at the base of the spinal cord and generally result in lower limb paralysis, reflexive bowel, or both.

The site of spinal cord damage determines the extent of the physical impairments. Injury of the cord in the cervical region creates tetraplegia, or paralysis of all four limbs. In addition to the limbs, the trunk and muscles of respiration are involved. Damage in the thoracic or lumbar region will result in paraplegia or paraparesis involving only the lower extremities and generally the lower trunk.

**Changes in Muscle Tone.** Paralysis of the voluntary musculature is the most obvious effect of SCI. Damage can involve the descending motor tracts, anterior horn cells, or spinal nerves, and it is often seen in combinations of these. Spinal shock is the loss of sensory, motor, and automatic control below the level of the lesion that occurs immediately after the trauma but resolves within a few weeks after injury.<sup>27</sup>

When the descending tracts are involved, immediate flaccidity is present and reflexes are absent. This is followed by autonomic symptoms, including sweating and reflex incontinence of bladder and rectum. Within weeks there is a gradual increase in the resting tone of the muscles innervated below the lesion, and reflexes reappear.

Spasticity is an inevitable consequence of spinal cord lesions. There is an essential or basic spasticity, which may be of some benefit to the individual when emptying the bladder or flexing the hip and knee. Excess spasticity is due to afferent stimuli. Spasticity can be made worse by the presence of constipation, infection, fracture, or a pressure sore below the level of the lesion, and it can be exacerbated by a sudden change in temperature or by physical or emotional stress. Typically, the flaccid condition lasts longer and spasticity comes later in a cervical injury compared with a thoracic injury.<sup>39,43</sup>

**Autonomic Nervous System Changes.** Autonomic dysreflexia (AD) can occur with a lesion above T5 and is the result of impaired function of the autonomic nervous system (ANS) caused by simultaneous sympathetic and parasympathetic activity. The ANS regulates body functions such as heart rate, blood pressure, and gland activity. Noxious stimuli, such as elevated blood pressure, overextended bladder or bowel, or other visceral stimuli, will typically elicit a sympathetic response, resulting in vasoconstriction and an increase in blood pressure. In the non-spinal cord injured individual, the descending sympathetic output compensates for this increase in blood pressure by causing vasodilation to bring blood pressure to a more normal level. Following SCI, sensory nerves below the level of the injury continue to transmit excitatory impulses, causing similar vasoconstriction and increased blood pressure. With the lack of sympathetic inhibitory output below the lesion, however, the blood pressure keeps rising unchecked. Secretions of neurotrans-

#### Box 34-2

#### SIGNS AND TRIGGERS OF AUTONOMIC DYSREFLEXIA

##### Signs

- Sudden and significant (>20 mm Hg) increase in both systolic and diastolic blood pressure above normal (Normal blood pressure when the lesion is above T6 is 90 to 110 mm Hg systolic and 50 to 60 mm Hg diastolic.)
- Onset of a sudden throbbing or pounding headache
- Sweating and flushing of the face, neck, or shoulders
- Goose bumps above the level of the lesion
- Blurred vision
- Visual field changes
- Nasal congestion
- Increased anxiety and apprehension without cause
- Changes in heart rhythm, such as arrhythmias, fibrillation, premature ventricular contractions

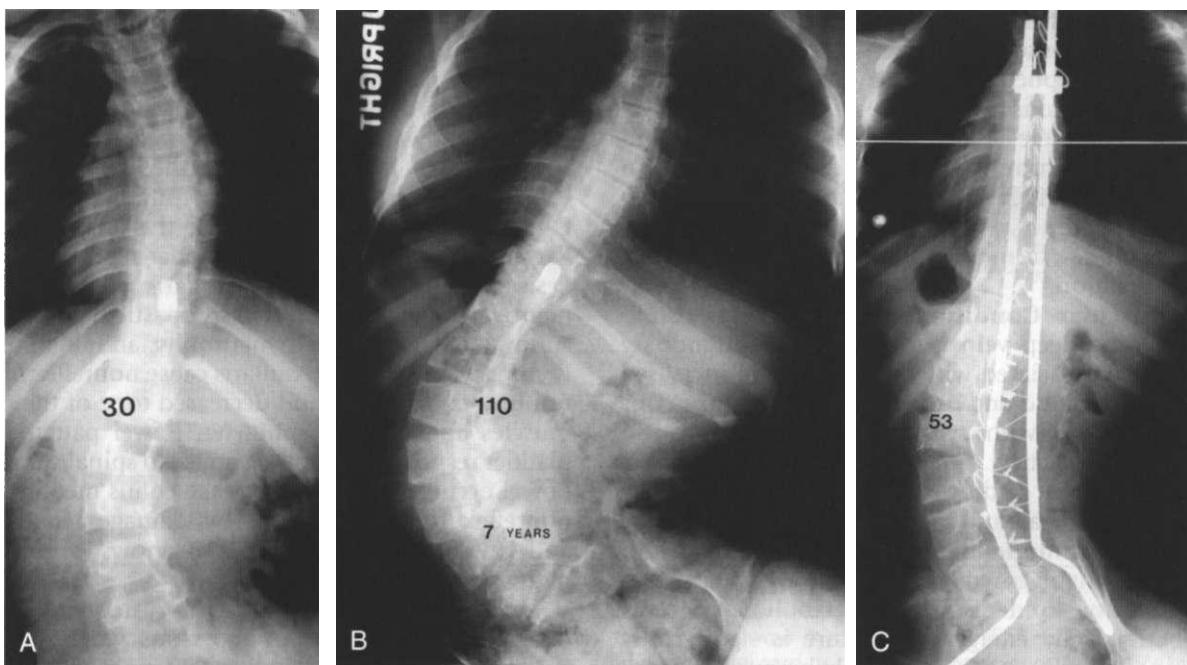
##### Triggers

- Full bladder
- Full or impacted bowel
- Scrotal compression
- Kidney stones
- Gastritis
- Contractions of labor and delivery
- Onset of menses
- Deep vein thrombosis
- Pulmonary embolus
- Pressure ulcers
- Insect bites
- Bruises caused by sharp objects
- Tight and constrictive clothing or apparatus
- Changes in temperature
- Pain or irritation below the level of the lesion

Adapted from Zabel RJ, Forest BH: Autonomic dysreflexia: an acute care emergency, *Acute Care Perspect* 7(3):9-14, 1999.

mitters, such as norepinephrine, epinephrine, and dopamine, support this sympathetic response. Control of the situation at the brainstem level leads to parasympathetic stimulation to slow the heart rate (bradycardia) through stimulation of the vagus nerve. This response is not strong enough to overcome the extreme vasoconstriction. The vasoconstriction continues above the level of the lesion and results in profuse sweating and skin flushing. A severe pounding headache follows, with sweating and chills without fever. The increase in blood pressure makes the person susceptible to subarachnoid hemorrhage, renal or retinal hemorrhage, and seizure or myocardial infarction. AD should be handled as a medical emergency.<sup>1,26</sup> See Box 34-2 for signs and triggers of AD.<sup>80</sup>

Loss of thermoregulation below the level of the spinal cord lesion is a result of the disruption of the autonomic pathways from the hypothalamus, resulting in subnormal body temperature in a normal ambient environment. Vasoconstriction, the ability to shiver, and the ability to sweat are lost. The body temperature then is greatly influenced by the external environment, and sensory feedback from the head and neck must be used to assist in regulating body temperature. The higher the lesion is, the more severe the problem becomes.



**Figure 34-12**

Progressive paralytic scoliosis after gunshot wound. **A**, Initial curve of 30 degrees. **B**, Seven years later, curve is 110 degrees. **C**, After fusion and segmental instrumentation, correction to 53 degrees. (From Canale ST, ed: *Campbell's operative orthopaedics*, ed 10, St Louis, 2003, Mosby.)

**Skeletal Changes.** Joint ankylosis caused by heterotopic ossification, or ectopic bone formation in the soft tissue such as the tendons and connective tissue, can limit range of motion, cause pain, and impair seating and posture. It often develops near the large joints, such as the anterior area of the hip, knee, shoulder, and elbow. It is always found below the level of the lesion, and it begins to develop within the first year after injury. The initial symptoms are soft tissue swelling, pain, redness, and increased temperature in the affected area. Changes in bony alignment can develop secondary to muscle imbalances caused by unopposed contractions. Scoliosis can develop over time due to lack of paraspinal support, as is evidenced in Fig. 34-12.

**Pain.** Individuals with SCI must deal with a number of secondary complications in addition to any disability caused by the injury itself. Pain, weakness, and fatigue appear to be most common and most closely linked to individual social and mental health functioning.<sup>41</sup> The number of reported pain sites increases with time, regardless of level or completeness of injury. While physical independence, mobility, and social integration remained relatively stable despite increasing numbers of pain sites, increases in depressive symptoms are associated with increased pain. Smokers with SCI report more pain sites than their nonsmoking counterparts.<sup>61</sup>

Pain caused by irritation of the nerve root is common, especially in cauda equina injury. Dysesthesia, impairment of sensation usually perceived as pain, can occur in areas with sensory loss and is often described as burning, pins and needles, or tingling. Disturbances of proprioception are related, and the person feels that a limb is in a different position than it is.

Musculoskeletal pain can result from faulty posture and overuse of limbs. Joint, ligament, and tendon deterioration is common, and secondary injury from muscle imbalances can come from functioning on the edge. Proper strength training can reduce these injuries and lead to more independence.

**Fatigue.** Complaints of fatigue, noted to be higher than in the general population, may be associated with changes in several systems. ANS changes with inadequate sweating and thermoregulation can cause activity intolerance. Psychologic well-being is associated with fatigue; depression and decreased community mobility can be predicted by increasing complaints of fatigue.

**Respiratory Complications.** Respiratory complications associated with spinal cord lesions can be life-threatening. Pulmonary complications are a common cause of death in both the acute and chronic phases. Spinal shock and poor management of edema can reduce vital capacity.

Lesions above C4 result in paralysis of muscles of inspiration and generally require artificial ventilation because of loss of the phrenic nerve innervation. Pulmonary complications with lesions at C5 through T12 arise as a result of loss of innervation of the muscles of expiration, the abdominal and intercostal muscles. The position of the diaphragm is compromised, and the abdominal musculature is unable to exert pressure during forced expiration. Paralysis of the external oblique muscles also inhibits the person's ability to cough and expel secretions.

An altered breathing pattern develops in conjunction with the loss of the diaphragmatic muscle, the intercostal muscles, and the accessory muscles of inspiration. The

upper chest wall flattens and the abdominal wall expands, leading to musculoskeletal changes in the trunk.

Aspiration and pneumonia occur frequently in individuals with SCI and is usually associated with high-level injuries and complete lesions and advanced age. Pneumonia is the most common cause of death, especially in the period immediately after the injury. With no other complications, proper rehabilitation, and stable respiration, the death rate from pneumonia matches that in the general population.<sup>60</sup>

**Cardiovascular Conditions.** Cardiovascular conditions, including deep vein thrombosis and pulmonary embolism, are associated with SCI because of increased coagulability of blood and decreased venous return. This may be associated with sympathetic dysfunction and unopposed vagal action. For long-term SCI, morbidity and mortality from cardiovascular causes now exceeds that caused by renal and pulmonary conditions, the primary causes of mortality in previous decades. Risk of cardiovascular involvement comes from a greater prevalence of obesity, lipid disorders, metabolic syndrome, and diabetes. Daily energy expenditure is significantly lower in individuals with SCI, not only because of a lack of motor function, but also because of a lack of accessibility and fewer opportunities to engage in physical activity. Autonomic dysfunction caused by SCI is also associated with several conditions that contribute to heightened cardiovascular risk, including abnormalities in blood pressure, heart rate variability, arrhythmias, and a blunted cardiovascular response to exercise that can limit the capacity to perform physical activity.<sup>56</sup>

**Metabolic Conditions.** Persons with SCI are prone to abnormal carbohydrate metabolism and are found to develop hyperinsulinemia and insulin resistance. During the acute phase of SCI, there is significant weight loss, especially with tetraplegia, associated with increased metabolic demands, muscle atrophy, and a negative nitrogen and calcium balance. Hypoproteinemia can be caused by pressure ulcers. Over time, there is usually an increase in body fat in proportion to lean tissue in the person with chronic SCI. A more sedentary lifestyle as a result of SCI may predispose a person to some of these conditions.

Soon after SCI, bones start losing minerals and become less dense. This may be due to alteration of the ANS and circulatory system. Inactivity and lack of weight bearing also foster the development of osteoporosis. It is believed that individuals with SCI may have an earlier onset and a greater extent of osteoporosis and that it may affect different body regions. There is a greater risk of fracture with osteoporosis.

**Pressure Ulcers.** Pressure ulcers are a frequent complication of SCI. They arise primarily because of the pressure associated with lack of mobility and resulting pressure in the area of a bony prominence. Moisture, poor nutrition, complete lesions, acute illness, and cigarette smoking predispose the skin to breakdown. Persons who do not follow through on self-care requirements because of depression, lack of motivation, substance abuse, or alcoholism are also prone to develop more and deeper pressure ulcers. Initially, the sacrum, heel, and scapula are the most common sites of ulcer formation

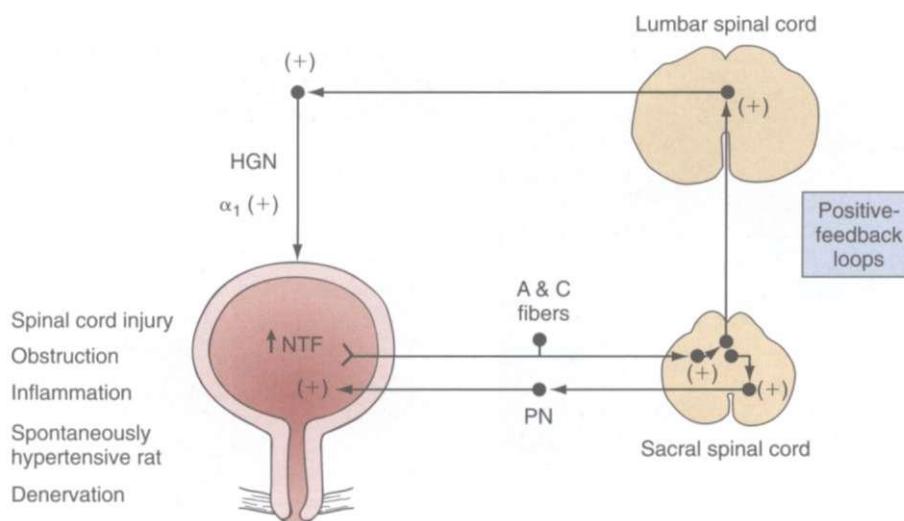
because of time spent in bed. As the individual begins to use a chair for mobility, the trochanter and ischium become common sites of pressure ulcers.<sup>75</sup>

**Bowel and Bladder Control.** Bowel and bladder control is always affected in the person with SCI. The spinal center for urination is the conus medullaris. Primary reflex control originates from the sacral segment. During the stage of spinal shock, the urinary bladder is flaccid. All muscle tone and bladder reflexes are absent. Lesions above the conus medullaris will cause a reflex neurogenic bladder, reflected by spasticity, voiding difficulties, detrusor muscle hypertrophy, and urethral reflux. Lesions at the conus medullaris cause nonreflex bladders, resulting in flaccidity and decreased tone of the perineal muscles and urethral sphincter. Bowel patterns mimic bladder responses in their response to spinal shock; reflex bowel occurs in lesions above the conus medullaris, and nonreflex bowel is caused by damage to the conus and cauda equina.<sup>47</sup> Fig. 34-13 shows bladder reflex pathways.

Urinary tract infection is the most frequent secondary medical complication seen in persons with SCI. This persists despite improved catheter materials and design and use of antibiotics. In concert with this is the increased concentration of calcium in the urinary system, which leads to formation of kidney stones. Calculi in the kidney are a complication found more frequently in individuals using an indwelling catheter.<sup>14</sup>

**Sexuality.** Sexual response is directly related to the level and completeness of injury. Sexual function relies on nervous pathways similar to those of the bladder and bowel and is altered as described earlier. There are two types of responses: reflexogenic, or a response to external stimulation seen in persons with upper motor neuron lesions, and psychogenic, a response that occurs through cognitive activity such as fantasy, associated with lower motor neuron lesions. Men with higher-level lesions can often achieve a reflexive erection but typically do not ejaculate. Those with lower lesions can more easily ejaculate, but achieving an erection is more difficult. With cauda equina lesions, erection and ejaculation are not usually possible. The primary reason for pursuing sexual activity is for intimacy needs, not fertility. Bladder and bowel concerns during sexual activity are not strong enough to deter the majority of the population from engaging in sexual activity. In addition, the occurrence of AD during typical bladder or bowel care is a significant variable predicting the occurrence and distress of AD during sexual activity.<sup>2</sup> Menses are typically interrupted for approximately 3 to 6 months; when restored, they can be another cause of AD. Fertility and pregnancy are uninterrupted, but the pregnancy must be observed closely, especially in the last trimester. Labor may begin without the woman's knowing it because of loss of sensation, and labor may initiate AD.<sup>64</sup>

**Sleep Disorders.** The prevalence of obstructive sleep apnea-hypopnea syndrome (OSAHS) is high after cervical cord injury. OSAHS is characterized by repeated oxygen desaturation. OSAHS should be suspected especially in individuals with daytime sleepiness, obesity, and frequent awakenings during sleep.<sup>49</sup> The changes in heart



**Figure 34-13**

Possible mechanisms underlying spasticity in bladder reflex pathways induced by various pathologic conditions. Bladders from rats with chronic spinal cord injury exhibit increased level of neurotrophic factors (NTFs), such as nerve growth factor. NTFs can increase the excitability of C-fiber bladder afferent neurons and alter reflex mechanisms in parasympathetic excitatory pathways in the pelvic nerve (PN) as well as in sympathetic pathways in the hypogastric nerve (HGN). These reflex circuits are organized in the spinal cord as positive-feedback loops that induce involuntary bladder activity. (From Wein AJ, Kavoussi LR, et al: *Campbell-Walsh urology*, ed 9, Philadelphia, 2007, Saunders.)

rhythm associated with OSAHS include sinus arrhythmia, severe bradycardia, and ventricular and supraventricular tachycardia. The risk of sudden death, particularly of cardiovascular cause, is well known.<sup>16</sup>

## MEDICAL MANAGEMENT

**DIAGNOSIS.** Delayed recognition of SCI is a significant problem in emergent care of traumatic injuries, occurring in more than 20% of cases.

Lateral film studies with plain radiographs are a rapid and effective way of evaluating cervical SCI, with the ability to detect approximately 85% of such injuries. When the open-mouth odontoid view and supine anteroposterior view are added, the accuracy rises to almost 100%. Any area that is inadequately demonstrated in the three-view spinal series is examined by computed tomography (CT). Flexion-extension studies are used primarily to evaluate instability caused by occult ligamentous injury and should not be done if there is any neurologic, bony, or soft tissue injury. CT demonstrates soft tissue structures and allows visualization of the bony limits of the spinal canal in the axial plane. CT is superior to other diagnostic procedures in demonstrating impingement on the neuronal canal.<sup>19</sup>

Fig. 34-14 shows a Jefferson fracture seen on x-ray and magnetic resonance imaging (MRI) scan. Fig. 34-15 compares fractures at time of injury and after stabilization. Myelography is indicated for optimal visualization of compression of the spinal cord after trauma. Myelography alone is rarely indicated, and it is used in conjunction with CT. In many cases, MRI has replaced myelography.

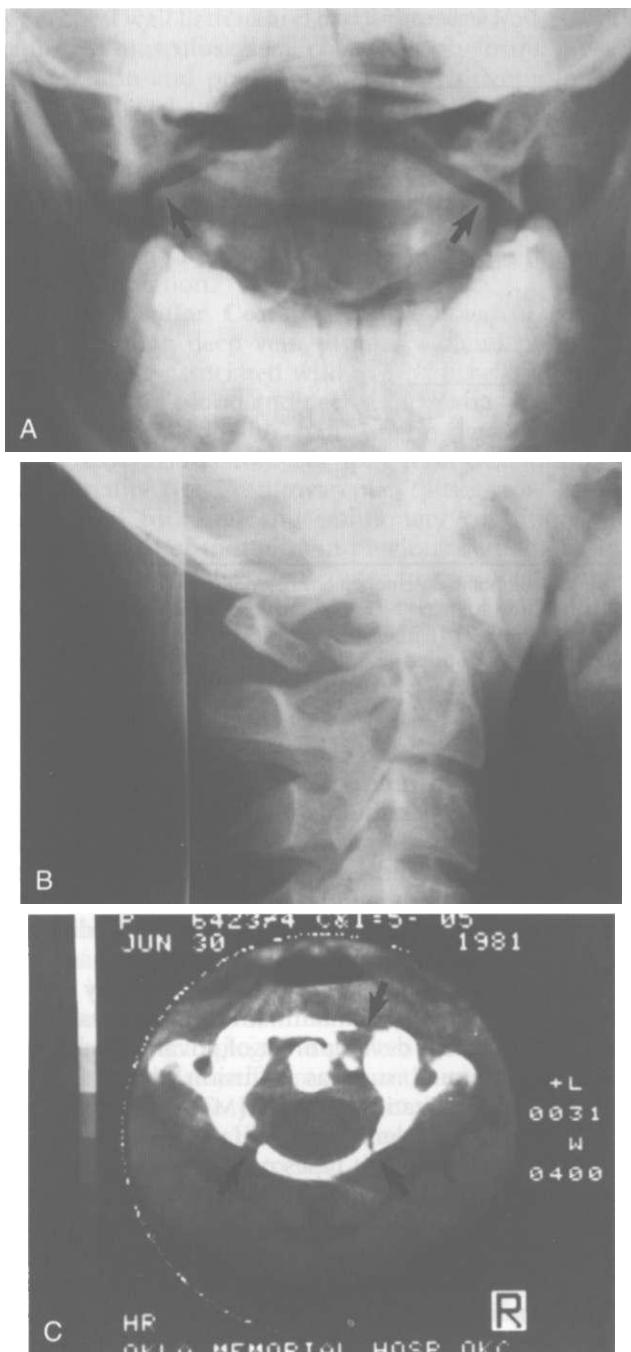
In acute SCI, MRI is sometimes problematic because of its limited use around ferromagnetic objects such as respirators, oxygen tanks, and traction devices. When

these obstacles do not exist, the extent of spinal cord damage and the possibility of disk herniation can be more readily assessed by MRI. The presence of intradural or extradural hematoma can often be demonstrated on MRI. MRI is useful in excluding spinal cord contusion or hemorrhage in persons with neurologic deficits and normal CT scans and plain films.<sup>21</sup>

MRI is insensitive to signals in the chronically injured cord because of lack of inflammation, hemorrhage, and swelling. The recent development of advanced structural imaging techniques, such as diffusion tensor imaging (DTI) and magnetization transfer (MT), is beginning to overcome these problems. In fact, lack of inflammation is advantageous for these types of images. DTI provides a unique image contrast called diffusion anisotropy. This reveals tissue organization at the microscopic level based on the average motion of water molecules.<sup>7</sup>

Advanced MRI techniques are better than conventional MRI in visualizing chronic SCI. The development of functional MRI (fMRI), currently useful in imaging the brain, and its application to the spinal cord should revolutionize the field of regeneration. To date, translation of brain fMRI techniques to the spinal cord has been hampered by the spinal cord's small size and mobility, and artifacts induced by pulse of CSF flow. Understanding recent advances in MRI technology and imaging is important to the discussion of regeneration. Transplanted cells can now be tracked after they are placed in living organisms using MRI. Neural stem cells are labeled with paramagnetic agents before they are transplanted, and MRI tracks their distribution and migration after they are placed in the damaged spinal cord.

In spinal trauma with severe neural malfunction, neuropsychologic studies help in determining which neural elements are involved, which spinal segment is responsi-



**Figure 34-14**

Fracture of C1 (Jefferson fracture). **A**, Anteroposterior view showing lateral displacement of the lateral mass and articulating facets of C1 on C2. **B**, Oblique view illustrating disruption of the posterior aspect of the ring of C1. **C**, Computed tomographic scan revealing the true extent of the injury. (From Green NE, Swirotkowski MF, eds: *Skeletal trauma in children*, ed 3, Philadelphia, 2003, Saunders. Courtesy of Dr. Teresa Stacy.)

ble for mechanical or other irritation, and whether the lesion is chronic, acute progressing, or resolving. Neurophysiologic studies allow intraoperative monitoring and include somatosensory evoked potentials (SEPs), motor evoked potentials (MEPs), neurography, F-wave and H-reflex electromyography (EMG), and sympathetic skin

#### Box 34-3

##### COMBINED EVALUATION OF HEAD AND NECK INJURIES

- Note exact time of injury. Management decisions are based on duration of symptoms.
- Assess loss of consciousness. Management of unresponsive athletes should follow the ABCs of trauma care (i.e., check airway, breathing, and circulation).
- Assess peripheral strength and sensation without moving the athlete's head or neck.
- Palpate the neck for asymmetric spasm or tenderness at the spine.
- Assess isometric neck strength without moving the athlete's head or neck.
- Assess active range of motion at the neck.
- Perform axial compression and Spurling test; if negative, athlete may be moved.
- Assess recent memory and postural instability.
- Inquire about symptoms such as headache, nausea, dizziness, or blurred vision.

From Whiteside JW: Management of head and neck injuries by the sideline physician, *Am Fam Physician* 74(8):1357-1362, 2006.

response (SSR). SEPs and MEPs are useful in the investigation of the CNS. EMG, neurography, and F-wave and H-reflex studies are used for evaluation of the peripheral component of spinal injury.<sup>79</sup>

A unique SCI syndrome, burning hands syndrome, was first described in sports injury. This syndrome appears to be a variation of central cord syndrome associated with severe burning paresthesias and dysesthesias in the hands and/or the feet. Other signs of neurologic dysfunction are minimal or absent. Over 50% of the time there is an underlying spinal fracture-dislocation. It is important to differentiate this syndrome from the much more common and usually innocuous "burning" or "stinging" of brachial plexus origin. Box 34-3 outlines the combined evaluation of head and neck injuries that should be performed beginning at the sideline.<sup>79</sup>

Diagnosis of syringomyelia may be delayed because the early symptoms are so similar to those of other, more common neurologic disorders. The accident that caused the initial damage may have been months to years previous to the onset of symptoms. MRI is the best imaging study for diagnosing this disorder in the beginning stages. MRI will show the syrinx number, size, and location, and any other abnormalities of the spine and spinal cord. Functional MRI allows the surgeon to see the spinal fluid pulsating within the syrinx.

**TREATMENT.** Interventions at several levels are required to improve mortality, morbidity, and quality of life. This begins with early surgical stabilization and with pharmacologic treatments aimed at blocking excitotoxicity and apoptosis. Prevention of the delayed wave of cell death that occurs in the weeks following injury is critical at the secondary level. Once injury is complete, the focus shifts to promoting regeneration. Pharmacologic treatments



**Figure 34-15**

A 21-year-old man involved in a motor vehicle accident sustained a burst fracture of L1 and L3. The patient had an incomplete spinal cord injury. **A**, A preoperative lateral view shows loss of height predominately at L1. **B**, A sagittal-cut magnetic resonance image shows compression at both L1 and L3. **C**, An axial-cut computed tomographic (CT) scan at L3 shows a retropulsed fragment filling half the canal. **D**, An axial CT scan at L1 shows a fracture of the lamina and retropulsion of a fragment into canal. **E**, This injury was stabilized with Isola instrumentation combining both pedicle screws and laminar hooks. Sagittal alignment was maintained. **F**, Postoperative anteroposterior radiograph showing a cross-connection added for additional stability. (From Browner BD, Jupiter JB, Levine AM, et al: *Skeletal trauma: basic science, management, and reconstruction*, ed 3, Philadelphia, 2003, Saunders.)

and transplantation are paired with the appropriate physical activity in order to optimize regeneration. It is now clear that spontaneous regeneration can be facilitated, and there is potential for optimizing regeneration even long after the injury and possibly extending throughout the remaining lifespan.

**Emergent Care.** The emergent phase of care is crucial for the person with traumatic SCI. It can make the difference between living the rest of life with a disability or recovering with only temporary neurologic deficits. An incomplete injury can be made worse by mishandling and can be made better by prompt attention to critical procedures. Box 34-4 describes guidelines for essential trauma care.<sup>11,30</sup>

Assessment of the likelihood of SCI includes understanding the mechanics of the trauma and obtaining vital signs to determine if the individual is in neurogenic shock. Movement of the distal components of the body

reflects the intactness of the spinal cord. In the case of a cervical injury, paradoxical respiration or abdominal breathing may be present, and immediate immobilization should be instituted. Use of a rigid collar and spinal board can help to prevent movement of the spinal column. Oxygen and medication should be given to control the hyperperfusion and swelling of the spinal cord. Transport should be swift, with care taken to avoid physical jarring caused by an uneven road surface and sudden stops.<sup>12-18</sup>

Monitoring in the critical care phase includes cardiac and neurologic status. Orthopedic management may begin at this phase and includes closed and open reduction of the vertebrae and decompression of the spinal cord. Insertion of a halo is common in treatment of cervical injuries, and a halo can be used without surgical intervention. In the more than 50% of individuals with SCI requiring surgery, fusion and internal fixation is the

**Box 34-4****ESSENTIAL TRAUMA CARE SERVICES ENDORSED BY IATSC AS THE "RIGHTS OF THE INJURED"**

- Obstructed airways are opened and maintained before hypoxia leads to death or permanent disability.
- Impaired breathing is supported until the injured person is able to breathe adequately without assistance.
- Pneumothorax and hemothorax are promptly recognized and relieved.
- Bleeding (external or internal) is promptly stopped.
- Shock is recognized and treated with intravenous fluid replacement before irreversible consequences occur.
- The consequences of traumatic brain injury are lessened by timely decompression of space-occupying lesions and by prevention of secondary brain injury.
- Intestinal and other abdominal injuries are promptly recognized and repaired.
- Potentially disabling extremity injuries are corrected.
- Potentially unstable spinal cord injuries are recognized and managed appropriately, including early immobilization.
- The consequences to the individual of injuries that result in physical impairment are minimized by appropriate rehabilitative services.
- Medications for these services and for the minimization of pain are readily available when needed.

From Mack C, Lormand JD, Goosen J, et al: *Guidelines for essential trauma care*, Geneva, 2004, World Health Organization; with permission.

IATSC, International Association for Trauma Surgery and Intensive Care.

most common procedure performed. The goal is to restore spinal alignment, establish spinal stability, and prevent further neurologic deterioration, enhancing recovery. Blood pressure dysregulation in persons with SCI may reflect increased vascular nitrous oxide. Treatment of hypotension using nitrous oxide inhibition has been shown to be effective in this population.<sup>71</sup>

Pharmacologic control of edema, blood flow, and secondary neurologic sequelae shows promise in current studies. Corticosteroids such as methylprednisolone have shown good results in large doses, but the side effects must be considered. MRI findings suggest that methylprednisolone therapy in the acute phase of SCI may decrease the extent of intramedullary spinal cord hemorrhage.<sup>50</sup> Although the concern about steroids continues, methylprednisolone should be administered to individuals with incomplete cervical SCI according to the Second National Acute Spinal Cord Injury Study protocol.<sup>69</sup>

Drugs that block opiate receptors appear to protect the spinal cord by decreasing endotoxic and hemorrhagic shock. Preservation of spinal cord function can be achieved by modulation of the neurotransmitters that are produced when there is injury to the nervous system. The dose-response curve remains narrow in most cases.<sup>76</sup>

When considering the appropriate type and dose of medication to be given to a person with an acute SCI, the degree of SCI must be assessed accurately. A moderate injury may require a different set of extracellular substances than that required in the secondary damage management for a severe injury.<sup>34</sup>

**Management of Complications.** Management of complications of SCI is critical. High cervical injuries require immediate placement of ventilation equipment and maintenance of pulmonary hygiene. Therapy consists of intermittent positive pressure breathing (IPPB), bronchodilators, and mucolytics. Prevention of pulmonary infection is critical in SCI.

In electrophrenic respiration the phrenic nerve is implanted with a single electrode, and the client carries a transmitter to activate the lung. This works well in persons with high cervical lesions and sparing of C3 to C5 anterior horn cells. This method of respiration more closely mimics normal physiology than does IPPB.<sup>12</sup>

Loss of ANS control affects the function of the cardiovascular system. Acute management of blood pressure is critical. The autonomic lesion predisposes persons with high spinal cord lesions to abnormal cardiovascular responses to vasoactive agents.

Treatment of spasticity includes use of muscle relaxants and spasmolytic agents.<sup>63</sup> Compared with oral baclofen, intrathecal baclofen infusion does not affect respiratory function and results in improved sleep continuity. Intrathecal baclofen infusion in therapeutic doses acts at the spinal level rather than at the supraspinal level.<sup>9</sup> Sustained-release fampridine is effective in individuals with chronic SCI.<sup>13</sup> One of the things that should be considered in the use of these medications is the fact that baclofen profoundly inhibits cell proliferation, survival, and differentiation, particularly myelination. Baclofen produces an irreversible loss of function. Baclofen is not an extremely effective oral agent, and there is now evidence that it can harm by inhibiting regeneration and recovery. Current research has shown that patterned activity, such as functional electrical stimulation (FES) bicycling (three times per week for 1 hour each time) may be a better way to control spasticity than medications.<sup>7</sup> Peripheral nerve blocks, such as botulinum toxin (Botox), provide a temporary reduction of spasticity. If there is long-term, severe spasticity, the contractile potential of the muscle can be modified by surgery. Therefore, spasticity-related interventions need to be aimed at what matters most to the individual. It is critical for clinicians to understand individuals' experiences to make accurate assessments, effectively evaluate treatment interventions, and select appropriate management strategies.<sup>52,53</sup>

**Pain Management.** Despite the fact that SCI causes loss of sensation, there is often significant pain that develops over time. Pain in SCI is classified in many different ways associated with intrinsic or neurogenic dysfunction, such as pain associated with syringomyelia or central cord pain, peripheral nerve pain, and musculoskeletal or mechanical pain. Psychogenic pain is also addressed in some classifications.

Management of neurogenic pain in SCI is by systemic or local drug therapy and by neuroaugmentative and neurodestructive intervention. The pharmacologic approach includes nonsteroidal analgesics, opioids, antidepressants, and anticonvulsants.<sup>38</sup> Pregabalin (Lyrica) is associated with relief of central neuropathic pain and with reduction in pain-related sleep interference and significant improvement in sleep problems. Action on

centrally located calcium channels may be important in the effectiveness of pregabalin in managing central neuropathic pain.

Neuroaugmentative procedures include transcutaneous electrical nerve stimulation (TENS), epidural spinal stimulation, and central thalamic stimulation. Neurodestructive procedures include both chemical and surgical destruction of nervous structures. Procedures may include deafferentation, interruption of ascending pain systems, or destruction of cells in the dorsal horn.

Diffuse, chronic, and dysesthetic pain following SCI has been described by several authors using different terms. Dysesthetic pain syndrome is difficult to treat. This pain is distinguished by its quality and is usually described using words such as *burning*, *stabbing*, *crushing*, *pressing*, or *pounding*, referred to as allesthesia and allodynia, and can be called the "central Tinel" sign. Light touch or tapping over areas rostral to the level of injury can be painful, and pain can be triggered by nonnoxious stimuli such as movement of the bed. It can also be triggered by noxious stimuli, such as smoking or gastrointestinal disturbances.<sup>74</sup>

Suburothelial injection of botulinum A toxin can effectively inhibit the occurrence of neurogenic detrusor overactivity, providing increased bladder capacity and improved incontinence grade in individuals with SCI. The therapeutic effect declines gradually after 3 months, and all symptoms return within 6 months.

Decompressive surgery is performed in individuals with spinal cord syringomyelia, depending on which area is affected. Surgery to create a pseudomeningomyelocele, an artificial CSF reservoir, performed to normalize the CSF flow, has been shown to be effective. By draining the cyst, it is possible to prevent the cyst from reexpanding. Draining the fluid can relieve pain, headache, and a sensation of tightness in the head or neck. In a dural graft procedure, the space around the spinal cord is enlarged to allow free flow of fluid and reduce pressure.

*Strategies for Spinal Cord Repair.* Nervous system repair is now feasible, and there is much research related to various strategies. A small degree of regeneration can result in recovery of function. A substantial loss of spinal cord tissue does not preclude function based on corticospinal tracts. There are two main regenerative approaches that are currently being studied: (1) optimizing spontaneous regeneration to restore function, and (2) transplanting stem cells. At this time there is no treatment that can clearly aid regeneration, but given the following information, primarily based on animal studies, it is likely that it will be developed in the near future.<sup>7</sup>

Embryonic stem cells are true stem cells that show unlimited capacity for self-renewal. In contrast, adult stem cells are progenitor cells or cells that are immature or undifferentiated. Their capacity for unlimited self-renewal and plasticity has not been as comprehensively demonstrated. Most progenitor cells are dormant or possess little activity in the tissue in which they reside. They exhibit slow growth, and their main role is to replace cells lost by normal attrition. Upon tissue damage or injury, progenitor cells can be activated by growth factors or cytokines, leading to increased cell division important

for the repair process. Progenitor cells participate in the normal maintenance of the CNS. These mechanisms include production and replacement of cells lost to normal aging and cell turnover.

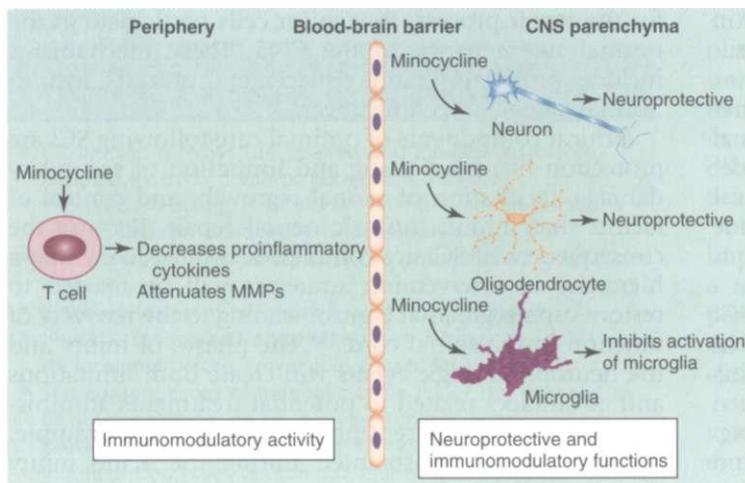
Critical components of optimal care following SCI are protection of neural tissue and limitation of secondary damage, facilitation of axonal regrowth, and control of factors that inhibit intrinsic neural repair. Because the consequences of SCI are complex, it is most likely that a hierarchy of intervention strategies will be needed to restore suprasegmental control leading to the recovery of function in the spinal cord.<sup>8,35</sup> The phases of injury and the neurophysiologic events will create both limitations and advantages related to potential treatments administered during the different phases of injury. For example, progenitor cells transplanted during the acute injury phase are vulnerable to the same set of cell death mechanisms predominant during the secondary phase of acute injury. Understanding the function of growth-inhibiting factors within the adult CNS may lead to control of the neural destruction after SCI.<sup>9</sup>

Improving regeneration of axons after SCI has been attempted by transplantation of various cell types. Success has been achieved in creating and sustaining function in both neurons and glial cells. Two CNS areas have shown the most response, the olfactory bulb and the hippocampal dentate gyrus. However, there has been some limited success in other area of the brain.<sup>51</sup>

Astrocytes can express molecules that are both growth permissive and growth inhibitory at the same time. Reactive astrocytes appear to create an inhibitory environment within the injured spinal cord and form an astrogliotic scar that acts as a physical and chemical barrier to axonal regeneration. Maintaining an environment to support the growth of axons may involve the selective removal of astrocytes from the site of injury.<sup>20,67</sup> In contrast, glial restricted precursors (GRP)-derived astrocytes (GDAs) may promote axonal regeneration via suppression of astrogliosis, realignment of host tissues, and delaying of expression of inhibitory proteoglycans. The glia, not the neurons, are the critical elements in preventing growth and in restoring it. Neurons retain the power to grow, and their sprouts only await the provision of a suitable glial pathway to be able to advance across the lesion.

Inflammatory reactions in the CNS have a dual nature; they may be neuroprotective as well as neurotoxic. Studies illustrate that the nervous and immune systems have overlapping rules of organization and intercellular communication. As a result, both systems express a host of common cytokines and neurotrophic factors that regulate cell survival and function. These shared mediators enable the two systems to engage in cross-talk and may provide a molecular explanation for neuroprotective effects of inflammation, which have been observed in animal models of CNS inflammation and trauma. T cells and natural killer (NK) cells in the spinal cord of rats can reduce the extent of neuron injury after ventral root avulsion. Hippocampal neurogenesis can be restored in mice with SCI by the transfer of CNS-reactive T cells.

Remyelination of bare axons after nervous system injury can be promoted by injecting endogenous stem

**Figure 34-16**

Peripheral and central functions of minocycline. Minocycline has immunomodulatory activity in the periphery and both immunomodulatory and neuroprotective capacity within the central nervous system. [From *The Lancet Neurology* Volume 3, Issue 12, 2004, Fig. 3, Elsevier.]

cells that can be mobilized to become oligodendrocytes by exogenously delivering growth factors such as NT-3 or BDNF.<sup>37</sup> Schwann cells have been used to facilitate a permissive environment for the injured spinal cord to regenerate. Previous experiments have shown compressive mechanical stress to be important in stimulating the regenerative behavior of Schwann cells. Transplantation of highly permissive Schwann cell-enriched peripheral nerve grafts may enhance regeneration in SCI.<sup>32</sup> Fig. 34-16 demonstrates the neuroprotective processes that may be integral within the immunomodulation.

Peripheral nerves may be able to provide an axonal bridge across the longer areas of spinal cord damage by activating nerve impulses carried from the brain, through intercostal nerve axons grown from implanted nerve, into the isolated distal end of the transected spinal cord, bridging the transection and connecting with neurons in the grey matter of the isolated distal segment of the spinal cord. It is possible that inferior-to-superior nerve bridging can produce return of function, just as superior-to-inferior nerve bridging does. This is related to the concept that axons have a "relentless compulsion" to grow until they participate in return of function. Regeneration-associated genes (RAGs) are associated with developing neurons and as a response to CNS injury, are considered an important element for biochemical therapies driving regeneration of the axon.<sup>26</sup> Electrical stimulation may provide an avenue for accelerated axonal outgrowth from the proximal nerve stump. (See Special Implications for the Therapist: Traumatic Spinal Cord Injury at the end of this chapter.)

A newly discovered cell line, hNT2.17, which expresses an exclusively neuronal phenotype, is a human-derived cell line that may be used to treat neuropathic pain and tactile and thermal allodynia, once it is further tested for safety and approved by the Food and Drug Administration. A dose of these human cells could be delivered with a spinal tap and affect the intrathecal spinal environment for sensory system modulation.<sup>24</sup>

**Stem Cell Transplantation.** Transplantation of stem cells is being studied extensively in relation to the treatment of SCI. Processes include producing regenerative

growth factors, expressing substances capable of breaking down scar tissue and modulating the immune system's response to injury. It appears that there will be potential for reprogramming the host microenvironment; for example, embryonic stem cell transplantation reduces macrophage influx by more than 50%.

An immature CNS does not produce the same inhibitory effect on axon growth as the mature CNS does. After spinal cord lesions and transplantation of spinal cord tissue, there is extensive growth of descending axons into the transplants. A transplant of fetal spinal cord tissue may serve as a bridge to permit the regrowth of axons from spinal and supraspinal levels across the site of SCI.<sup>3</sup> Transplants combined with neurotrophin treatment appear to have an additive effect compared with each intervention individually. Transplants and exogenous application of neurotrophic factors up-regulate cellular programs associated with regrowth and increase the extent of axonal regrowth with a favorable environment.<sup>3</sup>

Human umbilical cord blood stem cells (hUCB) hold great promise for therapeutic repair after SCI. Human embryonic stem cells (hESCs) may offer a renewable source of a wide range of cell types for use in research and cell-based therapies to treat disease. Cografted neural stem cells and NT-3 gene-modified Schwann cells promote the recovery of transected SCI and are a potential therapy for SCI. The grafted cells could enhance the survival of injured neurons in the inner pyramidal layer of sensorimotor cortex and could decrease the latency and increase the amplitude of cortical somatosensory and cortical motor evoked potentials, promoting limited structural and functional recovery.<sup>18,33</sup> The CNS offers the benefit of transplantation into an immunologically privileged site.

**PROGNOSIS.** The prognosis for recovery and repair depends on the phase of the injury and the age of the individual, with the best potential related to a younger age. The chronic consequences of SCI are related to maintenance as well as plasticity and repair. More than 90% of persons admitted to an acute care hospital for treat-

merit of SCI are ultimately discharged home. Morbidity and mortality during the first 4 weeks following SCI are most often related to paralysis of the respiratory muscles. The presence of either proteinuria with protein of 500 mg/day or greater is associated independently with increased mortality in the chronic SCI population.<sup>32</sup>

Long-term urinary tract infection continues to be a cause of death, but control of sepsis has improved markedly since 1970. This is primarily because of improvement in bladder training, antibiotic treatment, control of fluid intake, and surgery for obstruction of the lower urinary tract. Another common cause of death is respiratory disease, and this is the leading cause of death among high cervical injury clients. Pneumonia continues at a rate higher than in the general population, with pulmonary edema associated with injuries above T6. Heart disease is common, including myocardial infarction, cardiac arrest, myocarditis, and pulmonary embolism. However, the mortality rate is not much higher than that in the general population and is improving with increased pharmacologic control and improved medical knowledge of the cardiovascular changes accompanying SCI.

Most motor recovery occurs during the first 6 months, and strength can continue to increase with appropriate facilitation. The muscles graded 1 to 3 in the zone of partial preservation have potential to recover motor function. Overall, more than one half of the SCI population will have return of some neurologic function. Compression fractures have the most favorable prognosis for return of function, with crush fractures having the least chance for return of function.<sup>44</sup> Skeletal complications are related to the deformity and degenerative changes associated with nonuse of extremities. These can lead to pain and further neurologic compromise.

Prognosis related to mobility is a concern to most persons, especially those with thoracic-level injury. Preservation of axonal integrity and regrowth of neural tissue will have a significant effect on the recovery of mobility after SCI. Turning this nervous system recovery into improved functional status is part of current and ongoing research in the rehabilitation field.

People with SCI experience significant problems in a number of areas of life, resulting in ongoing stress related to pain, lack of income and money problems, spasticity, stress and worries, and difficulty in their sex lives. These problems do not appear to be highly correlated with aging, suggesting that they will not necessarily become more problematic, nor are they likely to self-remediate.<sup>45</sup>

**6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure**

**7A: Primary Prevention/Risk Reduction for Integumentary Disorders**

Rehabilitation to enhance the function and lifestyle of SCI clients has traditionally been aimed at helping clients achieve the ability to perform most daily tasks. The goal is to reach the point where the disability is no longer the major focus of clients' lives. Included are psychosocial adjustment, physical skills, health maintenance, and vocational adjustments. One goal of rehabilitation is to involve clients in all aspects of rehabilitation, provide the information and environment to foster independence, and have clients work as partners with the clinicians. Rehabilitation involves setting outcome goals related to changes in future lifestyle. Clients must sort through what they must give up, what they will have to do differently, and what they can still enjoy.

Among young adults, 40% to 50% of people with traumatic SCI are unmarried at the time of the incident. Available caregiver support is more limited, and external support systems will need to be established. For older individuals sustaining SCI, there are more preexisting medical conditions, more secondary medical complications, and more need for long-term care.

It is critical throughout the rehabilitation process that the therapist be prepared for the systemic and neurologic changes that could be life-threatening, such as orthostatic hypotension and AD.

Guidelines for management of these conditions as defined by the Consortium for Spinal Cord Medicine begin with monitoring blood pressure and pulse rate. Identification and elimination of the causal factor and notification of the occurrence to other team members are critical.<sup>46</sup> Refer to Box 34-2 for symptoms and triggers of AD.

Physical therapists are concerned with mobilization of the client, beginning in intensive care. Emphasis here is on respiratory management. Postural drainage, Trendelenburg's and reverse Trendelenburg's positioning, and assisted coughing must be initiated. Monitoring of atrial blood gas level and oxygen saturation at rest and during activity is critical. Box 34-5 outlines the considerations that should be included in determining the most appropriate parameters regarding mobility in clients at this level. An active program to increase strength and excursion of the diaphragm and other innervated accessory muscles of respiration, such as the sternocleidomastoid and trapezius muscles, is critical for pulmonary health. Included in the early program are range of motion, the beginning of a strength and endurance program, and education of both the client and family about the process of therapy. The goal of early intervention is to prevent the secondary sequelae, such as pressure sores or contractures, that would interfere with the rehabilitation process and to begin to prepare the musculoskeletal system for a different method of mobility.<sup>48</sup>

*Continued.*

## SPECIAL IMPLICATIONS FOR THE THERAPIST 34-1

### Traumatic Spinal Cord Injury

#### PREFERRED PRACTICE PATTERNS

- 4A: Primary Prevention/Risk Reduction for Skeletal Demoralization
- 5H: Impaired Motor Function, Peripheral Nerve Integrity, and Sensory Integrity Associated with Nonprogressive Disorders of the Spinal Cord

Orthotic management of the unstable spinal column is often necessary, and the therapist should be familiar with the types of orthotic devices used. Fig. 34-17 illustrates several orthoses used with different levels of spinal cord lesions.<sup>59,60</sup>

As Box 34-6 demonstrates, the level of the lesion will determine the degree to which independence can be expected in certain activities. The development of wheelchair skills includes transfers, propelling the wheelchair, electric wheelchair propulsion, management of the wheelchair components, and, most important, weight shifts in the chair to prevent skin breakdown. Adaptations within the environment are necessary for the client with weak upper extremities or poor hand control.

When possible, the client should be taught to maintain range of motion of the extremities independently. Adequate hamstring length is critical for performance

of transfers and independent dressing. In determining the training program for an SCI client, there must first be an assessment of physical capabilities and skills. The therapeutic program must work toward developing the necessary range of motion, strength, and skills. Skills needed for mobility without equipment may be different from skills needed for mobility with equipment. In some cases, transfers into and out of bed can only be done with special equipment, such as a transfer board or loop ladder.<sup>46,65</sup>

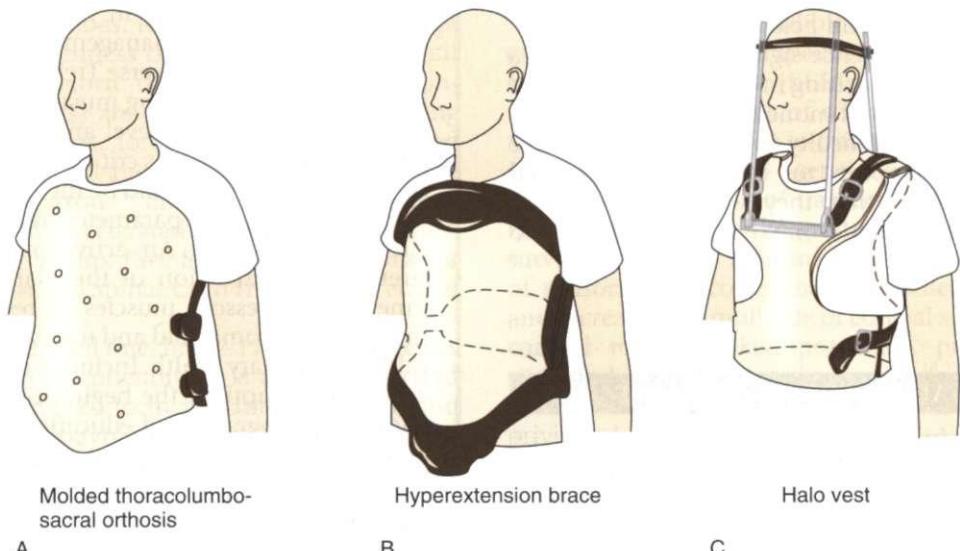
While working with clients on seated stability and functional movement, clinicians should be encouraged to incorporate bilateral reach tasks, because bilateral reach has the strongest relationship to performance of activities of daily living (ADLs).<sup>66</sup> Seventy-five percent of individuals with SCI sustain at least one fall per year. Even though most injuries are minor, there is potential for fracture and reduced community mobility. Factors perceived to contribute to falls most often were decreased strength in the trunk and lower extremities, loss of balance, and hazards in the environment.<sup>10</sup>

Rehabilitation traditionally focused on social reintegration is now giving way to inclusion of restoration of function by means of regeneration.<sup>67</sup> An approach known as activity-based rehabilitation, or ABR, is a noninvasive approach to enhance recovery and optimize spontaneous regeneration by inducing regular activity in the injured nervous system. The activity-based hypothesis implies that nervous system injury produces a complex secondary phase of progressive deterioration in the integrity of neural circuitry. It is believed that the cellular events that occur during development are similar to the processes that promote regeneration. ABR is based on the theory that patterned neural activity assists the system in optimizing cellular regeneration.

#### Box 34-5

##### **SAFETY PARAMETERS TO BE ADDRESSED IN FUTURE MOBILITY STUDIES**

- What are the ventilator settings at which mobilization should be withheld?
- Fever is known to increase oxygen consumption—should mobilization be withheld in febrile patients?
- Is there a dose of norepinephrine that predicts harm if mobilization occurs?
- How soon after respiratory failure or shock should mobilization be implemented?
- How should the appropriate mode and intensity of mobilization be selected?
- What is appropriate action if a decrease in oxygenation or blood pressure occurs?



**Figure 34-17**

**A**, Molded thoracolumbosacral orthosis, designed to control extension and rotary movements. **B**, Hyperextension brace, which restricts flexion in the thoracic area. **C**, Halo vest, which restricts upper thoracic and cervical motion.

**Box 34-6****DISABILITIES ASSOCIATED WITH LEVEL OF INJURY****C1-C5 Quadriplegia****Dressing**

Dependent in all dressing activities

**Bathing**

Dependent in all bathing activities

**Communication**

Independent with assistive devices for verbal communication (C1-C3)

Independent verbal communication (C4-C5)

Assistive devices necessary for keyboarding, writing, page turning, and use of telephone

**C6-C8 Quadriplegia****Dressing**

Independent with assistive device in bed (C7) or wheelchair (C8)

Minimal assistance with lower body dressing in bed

Moderate assistance undressing lower body in bed

Able to dress and undress in wheelchair with assistive devices (C8)

**Bathing**

Minimal assistance for upper body bathing and drying

Moderate assistance for lower body drying (C6)

Independent with assistive devices (C7-C8)

Assistive devices (tub chair necessary for tub or shower)

**Communication**

Independent in verbal communication

Assistive devices necessary for keyboarding, writing, and use of telephone

Setup required (C6)

**T1 and Below Paraplegia****Dressing**

Independent with use of assistive device

**Bathing**

Independent with use of assistive device (tub bench or cushion on bottom of tub)

**Communication**

Independent

Modified from Altrice MB et al: Traumatic spinal cord injury. In Umphred DA, ed: *Neurological rehabilitation*, St Louis, 1995, Mosby-Year Book, pp 502-506.

ABR therapy includes the use of FES to drive mobility in the extremities. FES can be used for muscle strengthening during the recovery period. Muscles are strengthened by cyclic, isotonic FES activation increasing muscle force, bulk, and fatigue resistance. FES may also increase sensory awareness and muscles for augmentation of voluntary control. Supported standing and limited distances of ambulation can be part of a program to improve respiratory control. Loading of the joints can be achieved with the use of stimulation in multiple sites to achieve co-contraction necessary for standing. Another important effect of ABR is the reduction of complications. The severe complications that accompany immobility include infections, skin breakdown, spasticity, pathologic fractures, deep vein thrombosis, pain, and AD.

Electrical stimulation reverses the natural recruitment order of muscle fibers, resulting in excessive fatigue compared to a natural contraction. Attempts to minimize fatigue related to FES have led to use of units that are fired by feedback on voluntary muscle activity or control stimulation through weight shifting. The FES is intended to enhance the contraction as the extremity moves through the cycle. Stimulation of the common peroneal nerve can facilitate the flexor swing, and stimulation of the upper portion of the hamstring and gluteus can facilitate the stance phase. Stimulation of the lower portion of the hamstring and quadriceps can support correction of knee hyperextension (*genu recurvatum*).<sup>53</sup> Neuroprostheses via surgically implanted FES device are rehabilitative tools with the potential to increase independence. The muscles stimulated include the vastus lateralis, gluteus maximus, semimembranosus, and erector spinae. The implanted standing neuroprosthesis appears to be a clinically acceptable and effective means of providing the ability to exercise, stand, and transfer to selected individuals with paraplegia or low quadriplegia through the coordinated activation of the paralyzed lower limb musculature. FES is not intended to create mobility when there is no potential for walking but is a tool to augment natural recovery that would lead to walking with crutches rather than wheelchair mobility. FES is used to augment walking.

Computer-controlled surface stimulation of three muscle groups in each leg (quadriceps, hamstrings, and gluteal muscles) allows paralyzed individuals to rotate the wheels of a bicycle under their own muscle power despite lack of volitional control of muscles. FES ergometry is the only method by which paralyzed individuals are able to obtain the benefits of exercise. An hour of FES ergometry is the equivalent of 6000 steps. Such patterned stimulation activates the lumbar gait central program generator, sending a normal pattern of neural activity up the cord. In addition to providing the benefits of physical reconditioning, FES ergometry is done to replicate normal levels of patterned neural activity in the spinal cord below the lesion level in an effort to optimize spontaneous regeneration. Home-based, self-administered neuromuscular electrical stimulation resistance exercise therapy consisting of 80 contractions per week improves arterial health after SCI, which may reduce the risk of future cardiovascular disease.<sup>55</sup>

Evidence now indicates hope for more sparing of descending tracts at the level of the lesion. Even minimal sparing of white matter has a profound impact on recovery of function.<sup>4</sup> The potential for training motor responses associated with reflexes below the level of the lesion has been identified. Walking is typically a priority concern for many clients with SCI, especially those with paraplegia. Interventions such as body weight-supported treadmill training have shown potential for improved locomotion.<sup>6</sup> Walking and balancing abilities are related, and balance reactions must be integrated into overground carryover. Partial body support treadmill training and

*Continued.*

supported overground walking are used with dorsiflexion assisted through FES and noxious stimulation driving flexor responses.<sup>28,54</sup> Studies of these techniques are finding that the context specificity of training affects the outcomes, and future studies are directed toward discovering the most efficient and effective interventions to produce walking and the health benefits that are associated.

Deficits in upper extremity function in individuals with tetraplegia are primarily due to the loss of motor pathways. Detrimental cortical reorganization, however, may create further loss of function. An intensive training intervention may induce both functional and neurophysiologic changes by driving cortical reorganization.<sup>55</sup> FES-assisted hand movement can enable clients with tetraplegia to perform most of their simple ADLs.

Exercise is critical to the individual with SCI to maintain cardiac fitness levels. There are numerous reports of strategies for exercises.<sup>56</sup> Circuit resistance exercise (CRT) improves muscle strength, endurance, and anaerobic power in individuals with paraplegia while significantly reducing their shoulder pain.<sup>57</sup> Skeletal muscle atrophy is associated with accumulation of greater intramuscular fat in thigh muscle groups in SCI and continues to increase over time in incomplete SCI.<sup>58</sup> There is a link between adiposity (accumulation of fat in adipose tissue) and defining characteristics of metabolic syndrome. Adiposity is related to dyslipidemia, vascular inflammation, hypertension, and insulin resistance.<sup>59</sup> There is persistent adaptive capability within chronically paralyzed muscles. Preventing musculoskeletal adaptations after SCI may be more effective than reversing changes in the chronic condition.<sup>60</sup>

Being independent of personal assistance in ambulation is related to lower levels of pain interference with life activity and work performance and less use of prescription medication to treat pain. Pain interference appears to be more problematic for those who are partially ambulatory than for individuals who are independent in ambulation.<sup>61</sup>

Age, employment status, motor level and completeness of injury, and ambulatory mode (use of hand-propelled or motorized wheelchair, use of crutches or canes, or walking independently) are independently associated with health-related quality-of-life scores (HRQoL). Chronic cough, chronic phlegm, persistent wheeze, dyspnea with ADLs, and lower forced expiratory volume and forced vital capacity are each associated with a lower HRQoL.<sup>15,40</sup>

The importance of specific staff qualities, the need for a vision of future life possibilities, the importance of peers, the necessity for relevant program content, and the importance of the ability to reconnect the past to the future has been documented by individuals with SCI. If rehabilitation services are to be evidence based, relevant, and effective in meeting the needs of people with SCI, they must be informed by the perspectives of people with SCI. The most important dimension of rehabilitation for people with SCI is the caliber and vision of the rehabilitation staff.

## References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. The reader can view the reference source and access it on-line whenever possible. There are a total of 80 cited references and other general references for this chapter.

# CHAPTER 35

## Cerebral Palsy

ALLAN GLANZMAN

### CEREBRAL PALSY

#### Overview

Cerebral palsy (CP) is a nonprogressive lesion of the brain occurring prior to 2 years of age resulting in a disorder of posture and voluntary movement. CP may be accompanied by impairment of speech, vision, hearing, and perceptual function. Common comorbidities include visual and hearing deficits, seizure disorders, hydrocephalus, microcephaly, scoliosis, hip dislocation, and mental retardation.

#### Classification

CP is often classified by the type of muscle tone, distribution of limb involvement, or functional skills. The types of muscle tone include hypotonia (low tone); hypertonia (high tone, spasticity); ataxia; and choreoathetosis or dystonia.

*Choreoathetosis* is characterized by involuntary distal writhing movements (athetosis) and poorly graded proximal voluntary movement (chorea). *Spasticity* is graded most commonly by using the modified Ashworth scale and is characterized by a velocity-dependent resistance to passive stretch.<sup>10</sup> *Dystonia* is characterized by sustained muscle contraction resulting in sustained end-range posture. *Ataxia* is characterized by diametric movement patterns. The patterns of motor involvement and distribution are described in Table 35-1.

Spastic CP, particularly quadriplegia and spastic diplegia, accounts for the majority of cases. Hemiplegia, ataxia, dystonia, and choreoathetoid CP affect a relatively smaller number of children. New cases of choreoathetoid CP have become rare in the United States and Canada as a result of improved prenatal care in the prevention of Rh incompatibility and hyperbilirubinemia. The disorder remains a problem in developing countries.

Functional skills can also be used to classify individuals with CP. The Gross Motor Function Classification System (GMFCS) provides a five-level system to classify motor involvement of children with CP on the basis of their functional status and their need for assistive technology and wheeled mobility (Box 35-1). The GMFCS provides a means of grading age-related developmental skill.<sup>31,32</sup>

Level 1 includes children with neuromotor impairments whose functional limitations are less than what is typically associated with CP. It also includes children who have traditionally been diagnosed as having "minimal brain dysfunction" or "cerebral palsy of minimal severity." The distinctions between levels I and II therefore are not as pronounced as the distinctions between the other levels, particularly for infants less than 2 years of age.<sup>31,32</sup>

The descriptions of the five levels are broad and not intended to describe all aspects of the function of individual children. The focus is on determining which level best represents the child's present abilities and limitations in motor function. Emphasis is on the child's usual performance in home, school, and community settings (not best performance).<sup>32</sup>

The levels are described on a time line including before second birthday, between second and fourth birthdays, between fourth and sixth birthdays, and between sixth and twelfth birthdays. Distinctions between adjacent levels are outlined in Box 35-1.

#### Incidence and Etiologic and Risk Factors

The reported incidence of CP ranges from 1.5 to 4.7 cases per 1000 births in the United States.<sup>28</sup> Advances in perinatal care have improved the chances for survival of infants of extremely low birth weight and immature gestational age.<sup>22</sup>

Despite the increased use of fertility drugs, survival of infants in multiple births, and survival of extremely-low-birth-weight infants, the incidence (number of cases occurring over a certain period) of neurodisabilities, including CP, has remained constant among surviving premature infants.<sup>26,33</sup> In fact, incidence may have begun to show some recent decline,<sup>21</sup> although the prevalence (overall number of cases present at a specified time) has increased because of the improved survival rates.

The cause of CP may be unknown and is often multifactorial. In children of normal birth weight who have disabilities associated with CP, 80% of the disabilities are a result of factors occurring before birth and 20% are attributed to factors occurring around the birth or in the immediate postbirth period (first 4 weeks of life).

**Table 35-1** Classification of Cerebral Palsy

Type	Distribution or Description
<b>Spastic</b>	
Monoplegia	Only one limb affected
Diplegia	Involves trunk and lower extremities; upper extremities to a lesser degree
Hemiplegia	Primarily one total side involved; upper extremity usually more than lower extremity
Quadriplegia (tetraplegia)	Involvement of all four limbs, head, and trunk
<b>Ataxia</b>	Irregularity of muscular action manifested by dysmetria; may be pure or combined with other forms
<b>Dyskinesia (choreoathetosis)</b>	Impairment of the power of voluntary movement; poor control of proximal movement (chorea) alternating with repetitive, involuntary, slow, writhing movements (athetosis); movements increase with emotional stress and around adolescence; often associated with rigidity or spastic quadriplegia or diplegia
<b>Hypotonia</b>	Abnormally reduced tension or muscle tone; accompanied by variable degrees of weakness

**Table 35-2** Risk Factors for Cerebral Palsy

Prenatal	Perinatal	Postnatal
Maternal infection <ul style="list-style-type: none"> <li>• Rubella</li> <li>• Cytomegalovirus infection</li> <li>• Herpes simplex</li> <li>• Toxoplasmosis</li> </ul>	Prematurity <ul style="list-style-type: none"> <li>• Mechanical birth trauma</li> <li>• Breech delivery</li> <li>• Forceps delivery</li> <li>• Twin or multiple births</li> </ul>	Neonatal infection (meningitis, encephalitis) <ul style="list-style-type: none"> <li>Environmental toxins</li> <li>Trauma</li> <li>Kernicterus</li> <li>Brain tumor</li> <li>Anoxia (e.g., near drowning, assault)</li> <li>Cerebrovascular accident</li> <li>Neonatal hypoglycemia</li> <li>Acidosis</li> </ul>
Maternal diabetes	Prolapsed umbilical cord or umbilical cord flow abnormalities	
Rh incompatibility	Low birth weight (<1750 g)	
Toxemia (undiagnosed or untreated)	Small for gestational age (SGA)	
Maternal malnutrition	Low Apgar scores (<4 at 5 min)	
Maternal thyroid disorders	Placenta previa (intrauterine bleeding)	
Maternal seizures	Abruption placentae	
Maternal irradiation		
Abnormal placental attachment		
Congenital anomalies of the brain		
Coagulation factor abnormalities		
Factor V Leiden mutation		
Antiphospholipid antibodies		

In children of low birth weight who develop disabilities associated with CP (approximately 0.7 per 1000 live births) uncertainty remains as to when the brain damage occurred (i.e., during embryonic development or during or after birth). Any prenatal, perinatal, or postnatal condition that results in cerebral anoxia, hemorrhage, or damage to the brain can cause CP (Table 35-2). CP is the second most common neurologic impairment in childhood (mental retardation is the first).<sup>19</sup>

### Pathogenesis

No consistent or uniform pathology is associated with CP. Several types of neuropathic lesions have been identified on the basis of autopsy: (1) hemorrhage below the lining of the ventricles (subependymal or interventricular), (2) hypoxia causing encephalopathy, and (3) malformations of the central nervous system (CNS).<sup>34</sup>

Until recently, interventricular hemorrhage (IVH) was the most common form of brain injury in the premature infant (Fig. 35-1). In recent years the incidence of IVH has declined from an incidence of 49% in very-low-birth-weight infants to 20% in the same population.<sup>35\*</sup> As a

result, periventricular white matter injury has become the more common cause of long-lasting brain injury in this population.

Periventricular lesions can be either cystic, as in periventricular leukomalacia (PVL), or more diffuse and result in abnormal myelination (Fig. 35-2). Diffuse periventricular myelination abnormalities can be found in up to 65% of premature infants when they reach full-term (9 months from conception). The incidence of PVL has declined somewhat, and PVL is seen in only 5% of this population. During the preterm period infants are at heightened risk for ischemia in the periventricular areas. The heightened risk is the result of passive-pressure circulation in the premature infant. The autoregulation of CNS blood flow normally present in full-term infants is absent, and the CNS blood pressure is more dependent on peripheral pressure. The premature infant between 23 and 32 weeks' gestation is at the highest risk of periventricular injury. As the periventricular white matter begins to myelinate, the risk of hypoxic injury declines.

Hypoxic injury can also occur in the full-term infant; however, this represents only a small portion of infants with CP. It often occurs in the presence of bradycardia,

**Box 35-1****GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM FOR CEREBRAL PALSY (GMFCS)**

The GMFCS is based on self-initiated movement with emphasis on sitting (truncal control) and walking. The focus is on determining which level best represents the child's present abilities and limitations in motor function. User instructions and more information are available on-line at <http://www.canchild.ca/Portals/0/outcomes/pdf/GMFCS.pdf>.

**Before Second Birthday**

- Level I** Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand, and take steps holding onto furniture. Infants walk between 18 months and 2 years of age without the need for any mobility device.
- Level II** Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomachs or crawl on hands and knees. Infants may pull to stand and take steps holding onto furniture.
- Level III** Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.
- Level IV** Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone. Physical impairments limit voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting positions. Infants require adult assistance to roll.
- Level V**

**Between Second and Fourth Birthdays**

- Level I** Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.
- Level II** Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture, and walk using an assistive mobility device as preferred methods of mobility.
- Level III** Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on their stomachs or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using an assistive mobility device and adult assistance for steering and turning.
- Level IV** Children floor sit when placed but are unable to maintain alignment and balance without use of their hands for support. Children frequently require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on stomach, or crawling on hands and knees without reciprocal leg movement.
- Level V** Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for though the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

**Between Fourth and Sixth Birthdays**

- Level I** Children get into and out of, and sit in, a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors, and climb stairs. Emerging ability to run and jump.
- Level II** Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for any assistive mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.
- Level III** Children sit on a regular chair but may require pelvic or trunk support to maximize hand function. Children move in and out of chair sitting using a stable surface to push or pull up on with their arms. Children walk with assistive an mobility device on level surfaces and climb stairs with assistance from an adult. Children frequently are transported when traveling for long distances or outdoors on uneven terrain.
- Level IV** Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a power wheelchair.
- Level V** Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

**Between Sixth and Twelfth Birthdays**

- Level I** Children walk indoors and outdoors, and climb stairs without limitations. Children perform gross motor skills including running and jumping but speed, balance, and coordination are reduced.
- Level II** Children walk indoors and outdoors, and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines, and walking in crowds or confined spaces. Children have at best only minimal ability to perform gross motor skills such as running and jumping.

**Box 35-1****GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM FOR CEREBRAL PALSY (GMFCS)—cont'd**

<b>Level III</b>	Children walk indoors and outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Depending on upper limb function, children propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain.
<b>Level IV</b>	Children may maintain levels of function achieved before age 6 or rely more on wheeled mobility at home, school, and in the community. Children may achieve self-mobility using a power wheelchair.
<b>Level V</b>	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

***Distinctions Between Levels I and II***

Compared with children at level I, children at level II have limitations in the case of performing movement transitions, walking outdoors and in the community, the need for assistive mobility devices when beginning to walk, quality of movement, and the ability to perform gross motor skills such as running or jumping.

***Distinctions Between Levels II and III***

Differences are seen in the degree of achievement of functional mobility. Children at level III need assistive mobility devices and frequently orthoses to walk, while children at level II do not require assistive mobility devices after age 4.

***Distinctions Between Levels III and IV***

Differences in sitting ability and mobility exist, even allowing for extensive use of technology. Children at level III sit independently, have independent floor mobility, and walk with assistive mobility devices. Children at level IV function in sitting (usually supported) but independent mobility is very limited. Children at level IV are more likely to be transported or use power mobility.

***Distinctions Between Levels IV and V***

Children at level V lack independence even in basic antigravity postural control. Self-mobility is achieved only if the child can learn how to operate an electronically powered wheelchair.

At the time this chapter went to press, the final phase in content validation for a 13- to 18-year age band of the GMFCS was in process. The 13- to 18-year age band will incorporate the International Classification of Functioning, Disability, and Health concepts of environmental and personal factors. Readers are encouraged to access the CanChild website for availability ([www.canchild.ca/](http://www.canchild.ca/)).

intrauterine growth retardation, and preeclampsia and may also be facilitated by the presence of infection.<sup>44</sup>

Hypoxic-ischemic injury can be the result of three possible underlying causes: (1) decreased perfusion resulting from systemic hypotension and poor autoregulation of cerebral blood flow; (2) emboli, which block distal perfusion, and thrombosis; or (3) clot formation from polycythemia or a hypercoagulable state.<sup>55</sup> Hypoxic-ischemic injury is known to disrupt the normal metabolic processes, starving the cells of oxygen because of poor perfusion and poor oxygen delivery to the cells, resulting in a reliance on the cells' limited ability to maintain homeostasis through anaerobic energy metabolism.

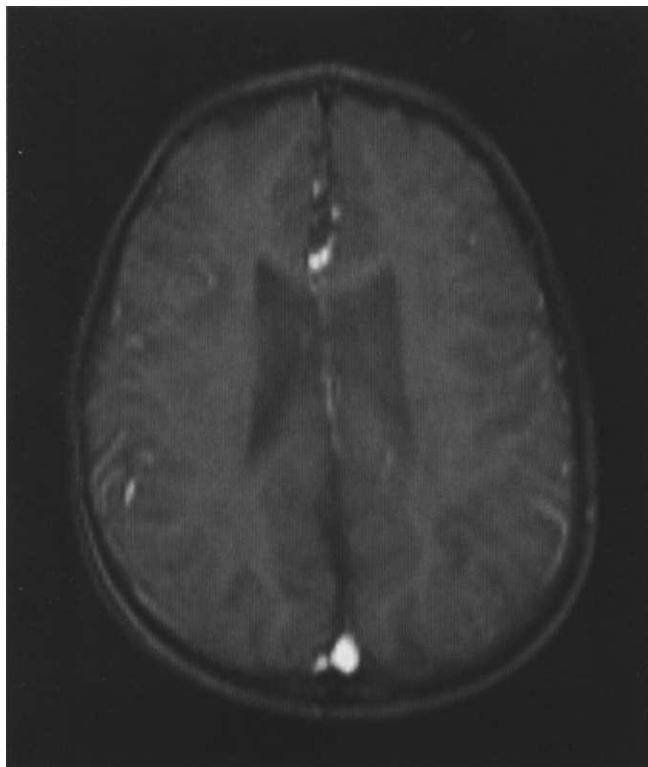
Eventually, insufficient energy is all that is available for powering the sodium-potassium pump in the cell membrane, and the ionic gradients across the cell membranes break down. The resulting influx of calcium begins a cascade that, along with the osmotic pressure gradient which has developed, ends in cell death. The second phase of cell damage occurs with reperfusion when vaso-dilatation allows increased blood flow and oxygen free radicals (see Fig. 6-2) are released that trigger programmed cell death or apoptosis. The severity and topography of the damage depends on the gestational age at the time of the injury and the degree of injury sustained.<sup>7</sup>

The primary hypoxic-ischemic lesion found in the premature infant is PVL (bilateral necrosis of the white

matter of the brain adjacent to the lateral ventricles), present in 42% of term and 87% of preterm infants with CP.<sup>25,52</sup> A portion of premature infants demonstrate impaired autoregulation of cerebral circulation and also demonstrate a passive-pressure cerebral circulation (i.e., blood pressure in the CNS is not able to remain constant with fluctuations in the peripheral circulation, placing them at risk of cerebral injury when fluctuations in peripheral pressure occur). The periventricular arterial border zones are at particular risk for hypoxic-ischemic injury in these children.<sup>52</sup>

Focal injury to the brain can also result from hemorrhage and ischemia, with the resulting collection of blood creating injury from direct mechanical pressure on the tissue and secondary ischemia. In the premature infant, hemorrhage of the germinal matrix (the cells from which the nervous system arises; in the adult, these cells, called ependymal cells, lie adjacent to the ventricular system) into the lateral ventricle (see Fig. 35-1) is a common cause of CP and can result in venous infarction of the periventricular area with a resulting cystic lesion in that portion of the brain.<sup>52</sup>

Hypoxic-ischemic injury in the mature neonate most commonly results from either selective neuronal cell damage or parasagittal brain damage. These hypoxic-ischemic insults affect the border zones of the major cerebral arteries, either in the cerebral cortex, cerebellum,



**Figure 35-1**

Magnetic resonance image of an interventricular hemorrhage with expansion of the lateral ventricles. This child was born at 26 weeks' gestation and diagnosed with diplegic cerebral palsy. (Courtesy Allan Glanzman, Children's Seashore House of the Children's Hospital of Philadelphia, PA.)

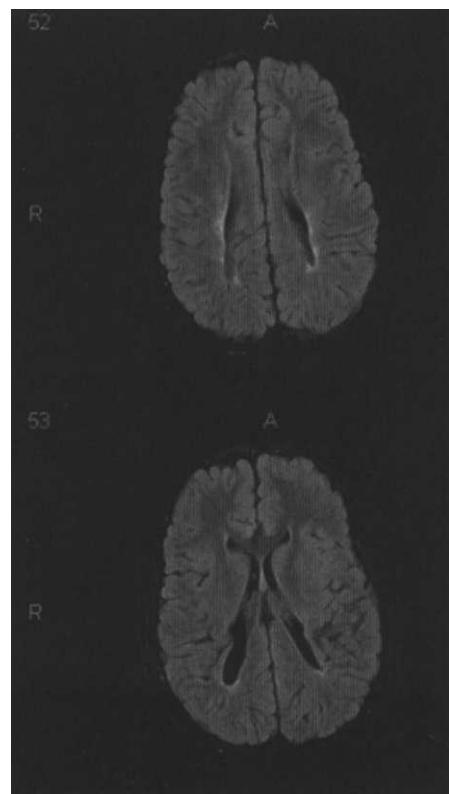
or the parietal or occipital regions. Focal or multifocal brain damage can result from either arterial embolism or venous thrombosis and is more common in the more mature neonate. The incidence of this mechanism of injury increases with gestational age greater than 28 weeks and typically presents as a unilateral injury.<sup>7</sup>

Children who develop CP fail to demonstrate normal CNS maturation after a CNS injury. Persistence of immature layers of the primary motor cortex is often present, and many of the other layers demonstrate abnormalities, particularly those with projections to the pyramidal tract.<sup>2</sup>

### Clinical Manifestations

Although the neurologic manifestations of CP are non-progressive, the motor impairments change with growth and maturation and may become more apparent as the affected child grows. Clinical manifestations of motor impairments associated with CP may include alterations of muscle tone, delayed postural reactions, persistence of primitive reflexes (Fig. 35-3), delayed motor development, and abnormal motor performance (e.g., delay in movement onset, poor timing of force generation, poor force production, inability to maintain antigravity postural control, decreased speed of movement, and increased co-contraction).<sup>14</sup>

Persistence of primitive reflexes and impaired motor function can affect the head, neck, trunk, and extremities



**Figure 35-2**

Magnetic resonance image of a periventricular leukomalacia with cystic formation extending into the parenchyma in a child with quadriplegic cerebral palsy. Top and bottom are serial sections in the same brain. In this child, the ventricles are a normal size. The abnormal finding is in the bottom slice where the cystic changes (black) extend into the brain tissue. (Courtesy Allan Glanzman, Children's Seashore House of the Children's Hospital of Philadelphia, PA.)

and impair sucking and swallowing, resulting in feeding difficulties, a major focus of occupational and speech therapists (Fig. 35-4).

Associated disabilities may include cognitive impairments (e.g., mental retardation, learning disabilities, seizure disorders); sensory impairments (in vision, hearing); and constipation or bowel and bladder incontinence with their associated problems (e.g., poor hygiene, skin problems).<sup>29,34</sup>

Microcephalus and hydrocephalus are also common findings, with the latter being the result of increased intracranial pressure. Behavioral signs of increased intracranial pressure accompanying hydrocephalus may include extreme irritability, vomiting, and eventually delay in reaching developmental milestones, resulting from pressure-induced damage as discussed earlier.

Musculoskeletal problems of altered muscle tone, muscle weakness, and joint restrictions are common and can result in functional and orthopedic impairments. For example, the abnormal pull of the spastic iliopsoas and adductor muscles are the initiating deforming force in hip dislocations (Fig. 35-5).

When spasticity and contracture of the iliopsoas occur, the medial joint capsule is compressed and the femoral head is pushed laterally. As lateral drift of the femoral head occurs, the iliopsoas insertion on the lesser trochan-



**Figure 35-3**

Asymmetric tonic neck reflex. Four-year-old with quadriplegic cerebral palsy demonstrating the asymmetric tonic neck reflex. This primitive reflex contributes to an obligatory change in body posture resulting from a change in head position. With head turning to one side, the arm and leg on the same side extend while the arm and leg on the opposite side flex. This posture resembles a fencing position. (Courtesy Allan Glanzman, Children's Seashore House of the Children's Hospital of Philadelphia, PA.)

ter becomes the center of rotation. Acetabular development ceases when the femoral head is completely displaced laterally, and further hip flexion pushes the head posteriorly to complete the dislocation (Fig. 35-6).<sup>9</sup>

Joint restrictions associated with CP are a result of a decrease in the number of sarcomeres<sup>44</sup> per muscle fiber. Muscles also demonstrate an increased variation in fiber size and type<sup>37</sup> with both hypertrophy and atrophy present, possibly representing an ongoing dynamic process. Increases in fat and fibrous tissue and a decrease in blood flow have been identified.<sup>36</sup> In this process, bone grows faster than muscle, resulting in a disadvantageous length-tension relationship of the muscle and an increased risk of subsequent contracture.<sup>43</sup> A characteristic decrease in muscle mass also results in decreased muscle power and endurance.

Changes in muscle tone affect a person's ability to control movement, resulting in poor selective control of muscles, poor regulation of activity and muscle groups, decreased ability to learn unique movements, inappropriate sequencing of movements, and delayed anticipatory postural response (Table 35-3). Most often, the timing and sequence of muscle activity are also affected.

A significant number of children with a diagnosis of spastic CP present with low muscle tone early in the first year of life and later develop spasticity. They often have



**Figure 35-4**

Symmetric tonic neck reflex. The same 4-year old with quadriplegic cerebral palsy as in Fig. 35-3 demonstrates another primitive reflex known as the symmetric tonic neck reflex (STNR). When the head and neck are extended, the arms extend; flexion usually predominates in the lower extremities. Flexion of the head and neck causes flexion in the upper extremities and extension in the lower extremities (not shown). In the normal infant the asymmetric tonic neck reflex and STNR are typically integrated by 6 to 8 months. Integration of the STNR allows voluntary flexion of both arms and legs needed to sit comfortably. Prior to 6 to 8 months, these reflexes can be observed in developing infants but when present are not obligatory (i.e., the person can voluntarily move out of the position). (Courtesy Allan Glanzman, Children's Seashore House of the Children's Hospital of Philadelphia, PA.)

insufficient flexor skills to position themselves against gravity for activities such as lifting the head, reaching, and kicking. The child will attempt to develop alternative strategies to complete the tasks. If control is not available, these strategies result in postures that allow completion of a particular sequence but do not allow for subsequent movement and transitions.

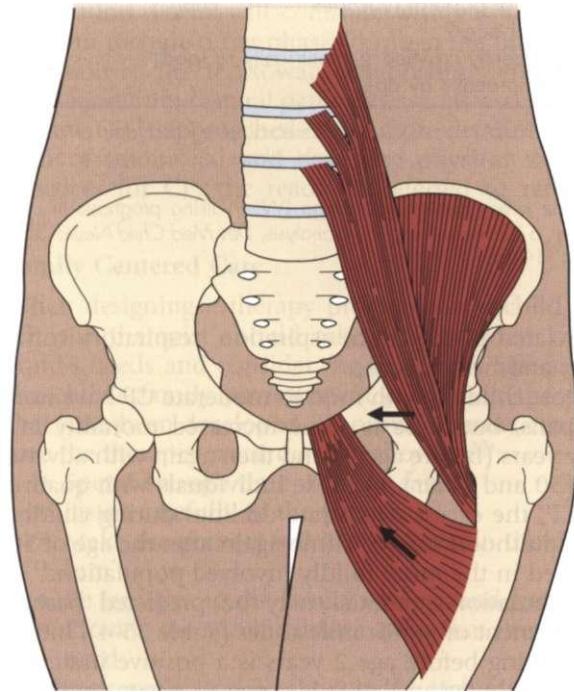
Examples of such situations are a wide-based sitting posture that allows the child to maintain sitting but decreases the ability to turn and rotate in and out of the position. Pulling to stand with the arms only (without using the lower extremities) is another example of an alternative strategy used by children with CP. If practiced and repeated over time, these abnormal movements become habitual and are difficult to change.

Each type of CP is characterized by its own clinical picture based on the presence and extent of these clinical manifestations. The progression of motor development associated with each type of CP is beyond the scope of this book. The reader is referred to other texts for a more detailed discussion.<sup>15-45</sup>

## MEDICAL MANAGEMENT

**DIAGNOSIS.** Observation, a good history, and a neurologic examination will provide the physician with the information necessary to make an accurate and early diagnosis. The diagnostic studies performed depend on clinical findings. For example, electroencephalography (EEG) is indicated when seizures are present or suspected; hip radiographic films are indicated to rule out hip dislocations and should be followed over time, particularly in the presence of spasticity.

Blood or urine screening tests may be used to rule out certain metabolic diseases, and a thorough workup should be undertaken if a history reveals a progressive course of positive family history. A computed tomo-



**Figure 35-5**

Spastic iliopsoas and adductor muscles are the initiating deforming force in acquired spastic hip dislocation.

graphic (CT) or magnetic resonance imaging (MRI) scan can provide information on the location of the insult.

**TREATMENT.** Comprehensive and cooperative planning with an interdisciplinary team including physicians, therapists, nurses, special educators, psychologists, social workers, nutritionists, and family members is essential.

Some of the most common medical management strategies include pharmacologic intervention, neurosurgical intervention, and orthopedic surgery. Skeletal muscle relaxants (e.g., baclofen, diazepam, dantrolene,



**Figure 35-6**

Anteroposterior radiograph of a young child with spastic quadriplegia and subsequent hip dysplasia with subluxation on the left. Note that a line drawn vertically down from the outermost edge of the acetabulum would bisect the head of the femur. Failure of the acetabulum to deepen with weight bearing resulting in hip dysplasia and subluxation occur as a result of the inability to weight bear and abnormal muscular forces pulling on the bone. The standard measurement for hip dislocation is a migration percentage. This is done by drawing Hilgenreiner's line, which provides a horizontal reference to the pelvis and then drawing Perkin's line perpendicular to Hilgenreiner's line from the outermost edge of the acetabulum. (Courtesy Allan Glanzman, Children's Seashore House of the Children's Hospital of Philadelphia, PA.)

**Table 35-3** Effects of Changes in Muscle Tone

### Effects of Moderate Hypertonia

Decreased movement: gross motor delays  
Tightness/contractures of hip and knee flexors, lower extremity adductors, gastrocnemius, and soleus (heel cords)  
Weak trunk; unstable kyphotic sitting  
Flexed, adducted, pronated upper extremity (severe cases)  
Significant upper extremity compensations (pulling up with arms, using arms for sitting balance)  
Standing and ambulation usually require assistance (splinting, walking aids)  
Usually at least one orthopedic (surgical) intervention required by age 5, often secondary to hip subluxation  
Adaptive equipment required by age 5 (e.g., stander, wheelchair or stroller, adaptive chair, walker)

### Effects of Moderate Hypotonia

Decreased movement: gross motor delays  
Possible lower extremity external rotation tightness  
Truncal kyphosis  
Compensatory widened lower extremity base of support in prone position, creeping, sitting, standing, and walking  
Delayed ambulation  
Unstable (pronated) feet  
Future difficulties with hopping, jumping, skipping, and higher-level balance skills

botulinum toxin) can be used to assist in controlling increased spasticity and can be administered orally (baclofen, dantrolene, and diazepam), intrathecally (baclofen), or directly to muscles through injection at the motor point (botulinum toxin).

Intrathecal administration of baclofen (through the sheath of the spinal cord into the subarachnoid space) uses an implantable intrathecal infusion pump to deliver medication to the spinal cord without the associated CNS sedation found with oral administration. After the pump is implanted, the dosage can be titrated to the optimal level for each person. Any attempts to control excess muscle tone (pharmaceutically or otherwise) should always be paired with functional goals to take advantage of the modulated tone.<sup>1,3</sup>

Motor point blocks can also be used to control spasticity and can be paired with serial casting to increase muscle length. Muscles such as the gastrocnemius, hip adductors, or hamstrings are injected with a botulinum toxin (or phenol) to create a temporary denervation and to decrease tone and increase movement.<sup>11,47</sup>

The type A botulinum toxin (Botox) is injected directly into the muscle at the motor point and is used to blockade the neuromuscular junction by acting presynaptically to reduce the release of acetylcholine. Muscle weakness and decrease in muscle spasm occur in 3 to 7 days and gradually reappear in 4 to 6 months.

Successful use of botulinum toxin type A in the upper extremity and at lower doses has been reported.<sup>19,20,42</sup> The effects of these injections will wear off anywhere from several weeks to several months later.

Dorsal rhizotomy (surgically identifying the posterior roots of the spinal cord and selectively resecting some of them) to reduce spasticity has been used over the past decade. This is usually performed at the L2 to L5 spinal levels for clients with spastic diplegia or mildly increased tone who are independent ambulators but who have abnormalities of posture and gait.<sup>51</sup> A rhizotomy may also be used effectively for clients with severe positioning difficulties such as severe quadriplegia. This procedure may reduce muscle tone enough to facilitate personal hygiene and provide improved sitting and comfort.

Orthopedic surgery may include muscle lengthening or releases (e.g., adductors, iliopsoas, hamstrings) to address contracture, muscle transfers (e.g., rectus femoris or tibialis posterior) to increase control or decrease excessive muscle pull, or bone procedures (e.g., femoral derotational osteotomy [see Fig. 23-12, B]; acetabular augmentation; triple arthrodesis; spinal fusions) to correct bony deformity, hip dislocation, or scoliosis.

Orthotic intervention may be used to maintain flexibility, support or stabilize a joint, or improve alignment. The ultimate goal is to delay the development of fixed contractures and improve function.

**PROGNOSIS.** Little information is available about the causes of death of people with disabilities resulting from CP. No national registry of names of people with CP is available, and cause of death is not usually listed on a death certificate even if the CP was a meaningful contributing factor to the cause of death. Currently available data indicate that common causes of death in this population

**Table 35-4** Predictors of Ambulation for Cerebral Palsy

Predictors	Ambulation Potential
<b>By Diagnosis</b>	
Monoplegia	100%
Hemiplegia	100%*
Ataxia	100%
Diplegia	60%*-90%
Spastic quadriplegia	0%-70%
<b>By Motor Function</b>	
Sits independently by age 2	Good†
Sits independently by age 3 to 4	50% community ambulation
Presence of primitive reflexes beyond age 2	Poor
Absence of postural reactions beyond age 2	Poor
Independently crawled symmetrically or reciprocally by age 2.5	Good†

\*From Pallas Alonso CR et al: Cerebral palsy and age of sitting and walking in very low birth weight infants, *An Esp Pediatr* 53:48-52, 2000.

†da Paz Júnior, Burnett SM, Braga LW: Walking prognosis in cerebral palsy: a 22-year retrospective analysis, *Dev Med Child Neurol* 36:130-134, 1994.

are related to infection, aspiration, respiratory compromise, and heart disease.<sup>48</sup>

Most children with mild to moderate CP have normal lifespans, but there is some increased mortality in the early years (before age 4) and then again with advancing age (50 and older). In those individuals with quadriplegic CP, the excess death rate declines during childhood and adulthood only to climb again after the age of 50, as is noted in the more mildly involved population.<sup>41</sup>

Ambulation potential may be predicted based on achievement of motor milestones (Table 35-4). Independent sitting before age 2 years is a positive indicator of future ambulation.<sup>53</sup> If it is going to occur, ambulation usually takes place by 8 years of age.

## SPECIAL IMPLICATIONS FOR THE THERAPIST 35-1

### Cerebral Palsy

#### PREFERRED PRACTICE PATTERNS

**4A:** Primary Prevention/Risk Reduction for Skeletal Demineralization

**4I:** Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Bony or Soft Tissue Surgical Procedures

**5C:** Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Congenital Origin or Acquired in Infancy or Childhood

**6E:** Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure

**7A:** Primary Prevention/Risk Reduction for Integumentary Disorders

In addition to the treatment options discussed in the previous section, physical therapists are exploring a more focused and proactive approach of activity-based intervention through intense activity training protocols, lifestyle modifications, and mobility-enhancing devices. Increased motor activity has been shown to lead to better physical and mental health and improve various aspects of cognitive performance.<sup>163</sup>

Activity-based programs for individuals with CP focus on maximizing physical function while preventing secondary musculoskeletal impairments; foster cognitive, social, and emotional development; and potentially promote or enhance neural recovery.<sup>164</sup>

With new research information about the role of neural recovery in damaged nervous systems, therapists can expect to see continued changes in philosophy and intervention approaches with this unique population. Focus will continue with early intervention but include other phases through the lifespan. As attention is directed toward establishing, enhancing, and maintaining neural pathways, we may see changes in how CP is approached. For a more detailed discussion of traditional and emerging physical therapist practices for CP, the reader is referred to reference 16a.

### Family Centered Care

When designing a therapy program for a child with CP, the therapist should take a broad view of the child's needs and consider the interactive effects that the child's family environment creates on the goals that have been developed. To provide family-centered care the therapist must do the following<sup>455</sup>:

- Spend enough time with the family
- Listen carefully to the parents
- Make the parents feel like partners in the child's care
- Be sensitive to the family's values and customs
- Provide the specific information that the parents need

The strengths and weaknesses of each family need to be assessed and considered when designing a given child's program, and the therapist needs to consider what the impact of carrying out the program will be on the family and, as a result, on the child.

If the cost (emotional, social, financial) is too great, the family may choose to abandon the intervention. As a result, the child may lose ground in terms of altered musculoskeletal alignment and decreased function, and the family must bear the emotional impact.

If the therapist is able to match the program with the family's cultural expectations, ability to participate, and emotional and financial resources, then a partnership with the family can develop that will most benefit the child in the long run. Expecting from family members only what they can succeed at and providing support and education where it is needed to help the family grow and care for the child with special needs will create the best therapeutic environment to allow the child to thrive.

There is often a fine line between balancing the natural history of the condition with the family's com-

mitment and understanding in maximizing the child's quality of life. In addition, families make choices in terms of providing for the child with CP. Often these choices must take into consideration other family members, expectations of themselves, expectations of the child, and, as mentioned, cultural and ethnic beliefs that may not match up with the health care professional's defined goals and plans for intervention.

### Early Intervention

A general review of intervention studies shows that children benefit from early intervention compared with those children not involved in specific programmed activities. Programming focused on cognitive outcomes has relatively stronger support<sup>5</sup> than programming aimed at solely motor outcomes.<sup>23</sup> The potential for improvement is better for children less than 9 months old but no greater than 2 years of age at a minimum frequency of intervention of two times per week.<sup>8</sup>

Early and accurate identification of CP provides the most likely opportunity for facilitation of optimal motor development.<sup>40</sup> Many motor milestone checklists are available from which a comparison with the normal can be made.<sup>24</sup> In fact, the gross motor function of children with CP and outcomes of intervention have often been evaluated using measures on children without motor impairment.

A more meaningful approach would be to make management decisions and evaluate intervention outcomes based on expectations for children with CP of the same age and gross motor function.<sup>31</sup> This type of evaluation can be made by using assessment tools specifically designed to evaluate the child with CP (e.g., Gross Motor Function Assessment<sup>38</sup>; Gross Motor Function Classification System<sup>31,32</sup>). An assessment of management practices with guidelines for the management of clients with CP is available,<sup>15</sup> as is a model for the acquisition of basic motor abilities and intervention implications.<sup>6</sup>

A significant number of children with a diagnosis of spastic CP present with low muscle tone early in the first year of life and later develop spasticity. They often have insufficient flexor skills to position themselves against gravity for activities such as lifting the head, reaching, or kicking. The child will attempt to develop alternative strategies to complete the tasks. If control is not available, these strategies result in postures that allow completion of a particular sequence but do not allow for subsequent movement and transitions.

Examples of such situations are a wide-based sitting posture that allows the child to maintain sitting but decreases the ability to turn and rotate in and out of the position. Pulling to stand with the arms only (without using the lower extremities) is another example of an alternative strategy used by children with CP. If practiced and repeated over time, these abnormal movements become habitual and are difficult to change.

*Continued.*

### Postoperative Concerns

After orthopedic surgery, the therapist can assist in reducing muscle spasms that increase postoperative pain by moving and turning the child carefully and slowly; however, adequate postoperative pain management should include medication (e.g., codeine and diazepam [Valium]) prescribed by the surgeon.

In the case of postoperative casting, the therapist can instruct the family to wash and dry the skin at the edge of the cast frequently, inspecting often for signs of skin breakdown. Repositioning and ventilation under the cast with a cool-air blow dryer can also assist in preventing skin breakdown. A flashlight can be used daily to inspect beneath the cast.

Surgical procedures (orthopedic or neurosurgical) may expose areas of underlying muscle weakness and instability. It is critical that an intensive therapy intervention program begin after surgery to assist with strengthening and improving functional performance.

### Assistive Technology

Properly prescribed assistive technology is vital in allowing the child with CP the least restrictive access to both the physical and social environment and is a critical part of the overall management of the child with CP. Assistive technology includes any device used to increase, maintain, or improve the functional ability of a person with a disability (Fig. 35-7).

This equipment can be either low tech (standers, positioning equipment, communication boards, or wheelchairs) or high tech (switch toys, power wheelchairs, or computer-based communication systems), as long as it is provided with a functional goal (Fig. 35-8).

Quality of life has become a new focus in the management of all clients seeking health care services. Mobility impairment limits can negatively effect overall development, including social, cognitive, emotional, and physical development. An increased emphasis on powered mobility to increase voluntary activity, function, and independence has contributed to improved quality of life for many individuals with CP (Fig. 35-9).

For children who are dependent for mobility, power mobility can be an option and can be successful in children as young as 2 years of age with corresponding cognitive skills.<sup>12,13,46</sup> These systems can be controlled with a standard joystick or adapted for control with a variety of switch systems.

These systems allow control with the head (either by a switch array or by proportional control), control through the use of individual switches, or control by a single switch through a scanning program (Fig. 35-10). When computer access is educationally appropriate, the same wheelchair-based control system can be adapted through an infrared link and mouse emulator to operate the computer. The mouse emulator is an electronic link that allows use of the joystick to control the mouse, usually by infrared beam.



**Figure 35-7**

Tub lift. A battery-powered tub lift has been very successful with this child, who has spastic quadriplegic cerebral palsy. With help, he transfers from the toilet next to the tub to the tub seat. With assistance, he swings his legs into the tub, and then can independently operate the unit to lower (and later elevate) himself. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

Use of mobility and speech-generating (communication) devices is encouraged earlier and with children at all levels of motor disability, including those with severe involvement. Differences in clinical practice and debate continue over providing an external means of mobility in favor of promoting more voluntary activity. More definitive research guidance is needed in these areas.

Proper positioning is critical to the child with CP, both from a functional perspective and to help prevent the soft tissue limitations that can develop over time. Appropriate positioning has been shown to encourage smoother and faster reach, decrease extensor tone, increase vital capacity, and improve performance on cognitive testing.<sup>30</sup>

In addition to proper wheelchair position, time out of the chair to counteract the flexed posture of the body is necessary. A standing program can be initiated between 12 and 18 months of age in the child who is not pulling to stand independently to maintain flexibility and provide the normal weight-bearing forces across the hip joint.

Standing helps orient children to the upright position, assists with visual perception, and can aid in digestion and elimination. Therapists hear anecdotal



**Figure 35-8**

Mulholland Walkabout. This 3-year-old girl with spastic quadriplegia can propel this wheeled upright walker through space to explore her environment and play where she wants to. Although she may not become a functional ambulator, the use of this equipment is developmentally appropriate. She could be a candidate for independent wheelchair mobility, but her parents have deferred this decision for now. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)



**Figure 35-9**

New power wheelchair. This 4-year-old child with cerebral palsy receives a new power wheelchair with a seat elevator to enable him to reach age-appropriate items on countertops and tabletops. Trunk supports and footplates help with alignment. The joystick on the left allows him to navigate independently. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)



**Figure 35-10**

This young lady with spastic quadriplegic cerebral palsy uses a DynaVox speech-generating device with a Tash Microlite switch on the left. By moving her head, she is able to hit the switch with her cheek to stop the electronic scan where she wants it to create a message. Stealth neck rest provides suboccipital head support. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

reports of decreased constipation and urinary tract infections in response to standing (Fig. 35-11). Standing also provides relief from pressure and can be used for prolonged muscle stretching, especially in the older or larger child.

Positioning for feeding for the child with CP is often critical for his or her ultimate success and safety at this task. The child's head and neck posture, pelvic posture, and inclination with respect to gravity are important considerations. A team approach using the skills of the physical and occupational therapist in conjunction with those of the speech therapist is essential in designing a program to optimize the child's oral motor skills (Fig. 35-12).

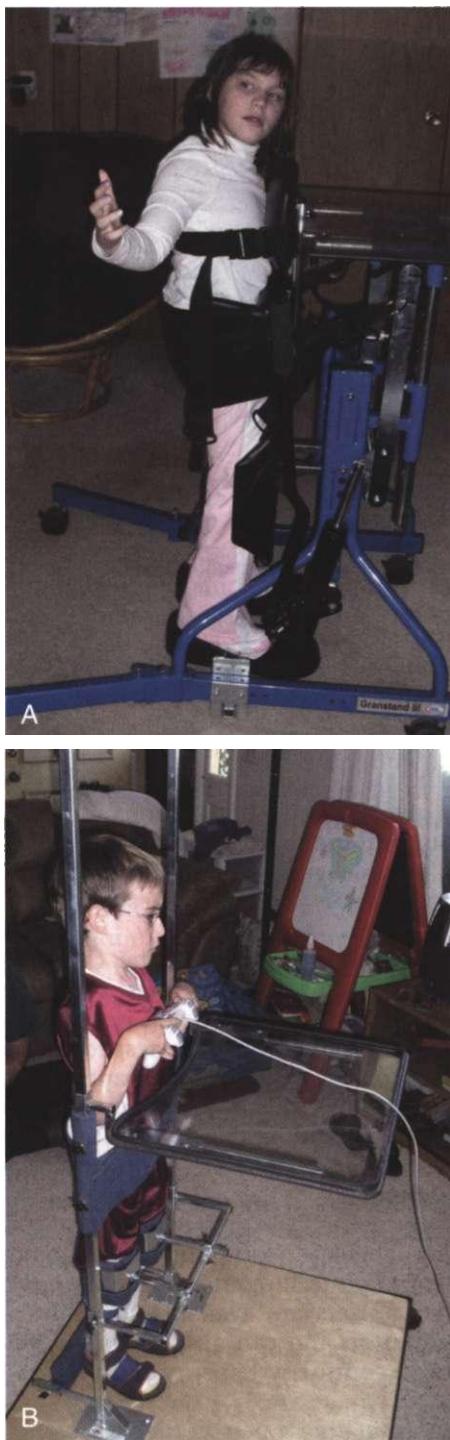
For children with expressive communication deficits, sign language, communication boards, and a variety of high-tech communication systems with voice synthesizers are available to augment spoken communication and can also be linked with wheelchair-based control systems in the power wheelchair user.

When evaluating a person for assistive technology, consideration should be given both to the individual's unique abilities and challenges and to the environment in which the equipment will be used. The products should provide the person the greatest degree of functional independence in all the environmental situations encountered. The barriers in each environment may vary, and thus the solutions by necessity may be different in different environments.

#### Manual Passive Range-of-Motion Exercise

It is generally accepted that manual straight-plane passive range-of-motion (ROM) exercise for children

*Continued.*



**Figure 35-11**

Standing frame. Many different types and styles of standers are available with a variety of adaptive features. **A**, Young girl with spastic hemiplegia from a birth injury/infection drives her power chair up to the stander. With assistance, she is able to get a seat sling under her buttocks to lift her up to standing. The sling is significant because it allows the parent to avoid lifting her into the stander. Shoe holders guide the placement of her feet. **B**, This standing frame offers an additional fun feature: the ability to operate a PlayStation. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)



**Figure 35-12**

Young boy with schizencephaly and cerebral palsy with severe involvement has a planar seating system with postural components. A tilt-in-space feature and deep ischial ledge formed in the seat keep his pelvis aligned and hips back. Hip flexion is combined with the medial thigh support between his legs to keep his knees apart and allow him to relax. Shoulder pad retractors are a feature added when it was discovered that downward pressure and anterior support at the shoulders improved head control for this child. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

with a chronic neurologic disorder such as CP is not, by itself, the best way to meet the physiologic requirements necessary to stretch a muscle. However, passive trunk rotation has been found to be useful in assisting with general flexibility and modulation of increased tone for persons with spastic quadriplegia.

Instead, splinting or positioning that offers a low-load prolonged stretch for longer than 30 minutes or that is used throughout the day is recommended. For example, splints such as lower extremity ankle-foot orthoses (AFOs) to maintain ankle ROM or supported standing to control lower extremity flexion contractures and assist with hip development and stability may be implemented. However, manual passive ROM exercise is not without its applications and is best combined with a well-thought-out positioning and splinting program.

Other interventions used by therapists to improve ROM and facilitate motor development or improve function include relaxation techniques such as neutral warmth or acupressure points; serial or tone casts (often in conjunction with botulinum toxin A injections); therapy ball activities; aquatic programs; and manual therapy techniques.

**Table 35-5** General Foot and Ankle Splinting Guidelines

Splints	Status	Application
Solid AFO neutral to +3 degrees DF	Nonambulators, beginning standers	<ol style="list-style-type: none"> <li>1. Less than 3 degrees of DF</li> <li>2. <i>Genu recurvatum associated with decreased ankle DF or weakness</i></li> <li>3. Need for medial-lateral stability</li> <li>4. Nighttime/positional stretching</li> </ol>
AFO with 90 degrees posterior stop and free DF (hinged AFO)	Clients with some, but limited, functional mobility	Applications 1-4 above, but need more passive DF during movement, such as ambulation, squatting, steps, and sit to stand
Floor reaction AFO (set dorsiflexion depending on weight line in standing)	Crouch gait Full passive knee extension in standing	For clients with decreased ability to maintain knee extension during standing and ambulation
SMO	Standers/ambulators with pronation at the ankles	<ol style="list-style-type: none"> <li>1. Need medial-lateral ankle stability</li> <li>2. Would like opportunity to use active plantar flexion</li> <li>3. Decreased DF not a problem during gait</li> </ol>

AFO, Anklefoot orthosis; DF, dorsiflexion; SMO, supramalleolar orthosis.

## Orthoses

AFOs are probably the most commonly used orthoses for children with CP. A rigid polypropylene AFO is used to provide medial-lateral stability to the foot and ankle while at the same time assisting with foot clearance during gait. The AFO can be set at +3 degrees of dorsiflexion to facilitate the increased ankle dorsiflexion necessary for the swing phase of gait or to decrease genu recurvatum (hyperextension at the knee) through ground reaction forces.

Hinged AFOs may be recommended once a child is moving in the upright position, especially when the child is beginning to walk, squat, or move up and down stairs, both to allow active ankle motion and to allow normal tibial progression during the stance phase of gait. A more flexible plastic such as copolymer or a thinner polypropylene may be used in the lighter child to provide more dynamic use of the foot musculature. In this case the term *dynamic ankle-foot orthotic* (DAFO) is used.

In some cases dorsiflexion assist hinges may be used, either with a plantar flexion stop or in the more mild cases with free plantar flexion. Care must be taken to choose the correct degree of hinge strength (or an adjustable hinge) so as not to create a crouched posture.

A supramalleolar orthosis (SMO) provides medial-lateral stability for the foot and ankle while allowing free plantar flexion and dorsiflexion. It is always helpful to have whatever plantar flexion is available, since this motion helps decelerate the limb during middle and late stances and facilitates the initial progression of the limb during late stance and early swing.

The SMO can be used when decreased active ankle dorsiflexion and excessive genu recurvatum are not problems. Extending the SMO proximally to the malleoli provides important support, whereas support distal to the malleoli usually shifts the deformity in a proximal direction. General guidelines and recommendations for foot and ankle splinting can be found in Table 35-5 (Fig. 35-13).

**Figure 35-13**

Orthoses. Ankle-foot orthoses as seen from behind this client in a standing frame are used for foot alignment and inhibition of tone associated with spastic quadriplegia. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

## Adolescents with Cerebral Palsy<sup>31a</sup>

Therapists are encouraged to include older children and teens in problem solving to help them become more self-sufficient, assuming more responsibility during this developmental phase despite their limitations in physical capability. Providing adolescents the opportunity to participate in planning and decision making is important for transition planning.

This may include decisions about assistive technology, environmental modifications, health and fitness,

*Continued.*



**Figure 35-14**

Adult with cerebral palsy. This 33-year-old man with athetoid cerebral palsy uses a speech-generating (communication) device and power chair; the joystick to operate the chair is under the client's right hand. The communication device can be folded and moved out to the side to allow for transfers. The chair has power tilt for independent position changes and pressure relief. Ankle huggers wrapped around the ankles keep his legs from flailing. A neoprene chest harness was added later to help provide external stability and increase control of his movements (not shown). This individual has clearly communicated how much he likes having the chest support, saying he feels much more in control of his body with it on. Straps and supports are fashioned with buckles, because Velcro is not strong enough to hold this client. The additional supports help reduce athetoid movements and improve function. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

and prevention of secondary musculoskeletal impairments. Likewise, the therapist can work closely with those individuals interested in participating in recreation and sports activities.

Client-centered assessment of strengths and needs identifying self-care, productivity, and leisure activities is possible and has been reported with this population.<sup>31a</sup>

### Adults with Cerebral Palsy

Therapists must also recognize and address the ongoing and unique needs of adults with CP (Fig. 35-14). With improved understanding of CP and its associated long-term complications and with improved health care, increased longevity has brought a new area of concern for children with CP living into adulthood: effects of the aging process. Decline of already impaired muscle strength and elasticity and bone density can lead to loss of ambulatory status.<sup>10a</sup>

Group homes, independent living centers, and sheltered workshops are now making it possible for many



**Figure 35-15**

Adult with cerebral palsy. Adults with moderate to severe effects of cerebral palsy can face some difficult physical challenges. This 21-year-old woman has a power chair with seat elevator, power tilt in space, and power elevating leg rests she can operate herself. Each leg rest can be raised or lowered separately to her comfort. This client changed her lower extremity position frequently to manage pain related to spasticity and immobility. A spring upper extremity assist on the left helps keep her hand on a modified joystick to allow her to independently control her chair. (Courtesy Tamara Kittelson-Aldred, Access Therapy Services, Missoula, MT. Used with permission.)

nonambulatory adults with disabilities to function independently or semiindependently. Regular daily living assistance is required by adults with spastic quadriplegia, especially in the area of lifts and transfers.

Degenerative arthritis, severe joint contractures, and other orthopedic deformities present the most common and challenging problems in this population (Fig. 35-15). Moderate to intense pain is a significant problem for the majority of adults with CP, accompanied by depressive symptoms interfering with activities. Further research is needed to determine the functional impact of this problem and appropriate interventions.<sup>18,39</sup>

Management strategies for older children and adults are different by virtue of their ability to participate and understand the aims of therapy. Therapy to maintain functional skills through the adolescent growth spurt, when weight gain, weakness, and atrophy often result in a decline in function, is essential. Aerobic training may prevent deterioration in body composition and muscle strength.<sup>50</sup>

Strengthening has become an integral part of therapy programs for individuals with CP and is especially helpful in this population. Measuring isokinetic strength is considered reliable in this population and should be used in rehabilitation protocols.<sup>4</sup> Isokinetic strengthening three times per week for 8 weeks can

improve muscle strength and gross motor skills<sup>27-50</sup> and increase cadence<sup>17</sup> for those people who remain ambulatory into adulthood. A traditional upper extremity strengthening program of 6 to 10 repetitions three times per week for 8 weeks has been useful in improving speed and endurance in independent wheelchair propulsion.

## References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. The reader can view the reference source and access it on-line whenever possible. There are a total of 54 cited references and other general references for this chapter.

# CHAPTER 36

## Epilepsy

KENDA S. FULLER

### EPILEPSY

#### Overview and Definition

*Epilepsy* comes from the Greek word meaning possession. The Greek people believed that seizures were caused by demons. Stigma and prejudice surrounding epilepsy continue, and people living with epilepsy often are reluctant to admit it or to seek treatment.<sup>2</sup>

A fundamental difference exists between seizure and epilepsy. Epilepsy is defined as a chronic disorder of various causes characterized by recurrent seizures—sudden, usually unprovoked attacks of subjective experiential phenomena, altered awareness, involuntary movements, or convulsions. Although a diagnosis of epilepsy requires the presence of seizures, not all seizures imply epilepsy. Epilepsy in children is usually considered constitutional, whereas in the older person, it is probably related to a provoking cause that develops over time.<sup>29</sup>

A seizure is a finite event; it has a beginning and an end. Seizures are a result of paroxysmal excessive discharge of cerebral neurons resulting in transient impairment or loss of consciousness. Seizures can be induced in any normal human brain by a variety of different electrical or chemical stimuli. The cerebral cortex in particular contains within its anatomic and physiologic structure a mechanism that is inherently unstable and capable of producing a seizure. Seizures are a relatively common symptom of brain dysfunction, and they may occur in many acute medical or neurologic illnesses in which brain function is temporarily disrupted. These seizures are most often self-limited and do not persist after the underlying disorder has resolved. Seizures also can occur as a reaction of the brain to physiologic stress, sleep deprivation, fever, and alcohol or sedative drug withdrawal. Isolated seizures also occur sometimes for no discernible reason as unprovoked events in presumably healthy people. None of these kinds of seizures represents epilepsy.<sup>6</sup>

#### Incidence

Epilepsy affects about 45 million people worldwide. Incidence is highest among young children and the elderly, and men are affected slightly more often than women (1.5:1). In the United States figures range from 31 to 57 per 100,000 population.

The incidence in the older population, once thought to be low, has increased to approximately 77 per 100,000. In individuals over age 80, this incidence rises to 159 per 100,000. Epilepsy is the third most common serious neurologic disease of old age after dementia and stroke.<sup>26,30</sup>

Although seizure activity in children is rare, at less than 1%, seizures are the most common symptoms requiring medical attention in the infant. It has been estimated that 75% of cases of epilepsy have their onset before 20 years of age.

#### Etiologic and Risk Factors

In many individuals, several predisposing factors coexist, and the development of epilepsy reflects the interaction of acquired brain pathology and genetic predisposition. Although genetic diseases account for only about 1% of cases, heritable factors are important in a much higher percentage, especially in children. Forms that are demonstrably more heritable than others are termed *idiopathic* or *primary epilepsies*. Common features include a variable family history, generalized spike-wave abnormality on electroencephalogram (EEG), and onset in childhood or adolescence.

Although mutations in single genes account for a few rare epileptic syndromes, in most cases of epilepsy, data are most consistent with complex, polygenic influences.<sup>19</sup> One gene for juvenile myoclonic epilepsy has been located on chromosome 6. Mutations of the EFHC1 gene may underlie different syndromes. The function of EFHC1 is unclear; roles in apoptosis and ciliary function have been proposed.<sup>3</sup> This genetic predisposition to seizure activity may explain why one individual with brain damage develops epilepsy while another with similar damage does not develop seizure activity.<sup>22</sup>

**Adult-Onset Seizure Disorder.** The causes of symptomatic epilepsy are multiple, and the symptoms may be transient, resolving after treatment of the primary disorder. Box 36-1 describes some of the causes of symptomatic epilepsy. Epilepsy can be caused by virtually any major category of serious disease or human disorder. It can result from congenital malformations, infections, tumors, vascular disease, degenerative diseases, or injury. In people over age 50, cerebrovascular disease is the most common cause of seizures, which often accompany or follow a stroke, and increases the risk of seizure by

**Box 36-1****POTENTIAL CAUSES OF ACUTE SYMPTOMATIC SEIZURES****Medical Conditions****Metabolic Derangements**

- Hyponatremia (<120 mEq/L)—especially acute
- Hypernatremia (>150–155 mEq/L)—especially acute
- Hypoglycemia (<40 mg/dL)
- Hyperglycemia (>400 mg/dL)
- Hyperosmolality (>320 mOsm/L)
- Hypocalcemia (<7 mg/dL)
- Respiratory alkalosis—acute

**Drug-Induced Seizures**

- Isoniazid, penicillins
- Theophylline, aminophylline
- Lidocaine
- Meperidine
- Ketamine, halothane, enflurane, methohexitol
- Amitriptyline, maprotiline, imipramine, doxepin, fluoxetine
- Haloperidol, trifluoperazine, chlorpromazine
- Ephedrine, phenylpropanolamine, terbutaline
- Methotrexate, BCNU (carmustine), asparaginase
- Cyclosporine
- Cocaine (crack), phencyclidine, amphetamines
- Alcohol (withdrawal)

**Illnesses**

- Eclampsia
- Hypertensive encephalopathy
- Liver failure
- Polyarteritis nodosa
- Porphyria
- Renal failure
- Sickle cell disease
- Syphilis
- Systemic lupus erythematosus
- Thrombotic thrombocytopenic purpura
- Whipple's disease

**Neurologic Conditions**

- Angitis of the nervous system
- Meningitis
- Encephalitis
- Acute head trauma (impact seizures)
- Stroke
- Brain abscess
- Brain tumor

Adapted from Pedley TA: The epilepsies. In Goldman L: *Cecil textbook of medicine*, ed 22, Philadelphia, 2004, Saunders, Chap 434, Table 434-1.

17%.<sup>15–28</sup> It is believed that perhaps even in the absence of stroke, cerebrovascular disease may predispose an individual to seizure activity. Seizures may be the first symptom of an intracranial mass, and it is suggested that 10% to 15% of older adult-onset epilepsy is a result of neoplasm. When the seizure is because of a permanent lesion or scar, the seizure activity may persist. Head trauma is the most common preventable cause of epilepsy. More than 3000 cases of epilepsy are added every year in the U.S. population because of head injury.<sup>27</sup> Subdural hematoma can cause seizure activity and is

common in older adults, often after a fall that may have seemed trivial.<sup>25</sup> Pneumonia, which carries the possibility of hypoxia, especially in the aged, can cause seizures that can become recurrent.<sup>27</sup> Alcohol abuse often leads to uncontrolled seizure activity.

Hormonal changes with increase in the levels of estrogen that occur during ovulation and menstruation can be a trigger for seizure in some women. Fertility appears to be reduced by seizure activity when it is triggered by hormonal changes. Disruptions of reproductive function in women include anovulatory cycles that may increase the risk for infertility, migraine, emotional disorders, and reproductive cancers. Moreover, both epilepsy itself and use of medications have been implicated as causal or contributory factors that can alter reproductive hormone levels and promote the development of reproductive endocrine disorders, especially polycystic ovarian syndrome. Menopause also tends to occur earlier in women with epilepsy, and this early menopause is associated with a history of high seizure frequency.<sup>9</sup>

**Infantile Seizures.** The age-dependent appearance of spontaneous seizures in the primary epilepsies appears to depend on a critical period in cerebral maturation when the genetically determined defect is expressed clinically as a manifest change in behavior. Changes known as microdysgenesis occur in the brains of these infants. These changes include an increase in cell density, abnormal arrangements of cortical neurons, and an increase in white matter neurons.<sup>7</sup>

Seizures may occur in the neonatal period, within the first 24 to 72 hours, and they are usually of focal cerebral origin. The seizure type relates to the level of development of the brain. The seizures tend to be unilateral because of the immaturity of the forebrain and corpus callosum, limiting movement from one hemisphere to the other. Seizure activity represents the relative development of the limbic system, diencephalon, and brainstem. Hypoxic-ischemic brain insult is the most common cause of neonatal convulsions and is because of compromise of oxygen to the brain during or before delivery. Seizures resulting from cerebral contusion are often a result of prolonged or traumatic labor.

The other important cause of seizures arising in the early neonatal period is hypoglycemia, most often seen in babies who are small for gestational age. The neurologic symptoms of hypoglycemia consist of irritability, drowsiness, hypotonia, and apnea. Approximately half of the babies with hypoglycemia develop neurologic symptoms, and about one fourth of these go on to develop seizures.

Hypocalcemia (serum level below 7 mg/100 mL) may occur in the first 2 to 3 days of life either in low-birth-weight infants or in association with the complications of birth asphyxia. It may then contribute to seizures but is rarely the primary cause. Hypercalcemia occurring at age 6 to 8 days is usually the primary cause of the clinical features of neonatal tetany, including jitteriness and jaw, knee, and ankle clonus. Hypocalcemia is rare in infants who are breastfed or fed formula created to simulate human milk. Hypernatremia, resulting from alteration in sodium level, may also lead to neonatal seizures. Hypernatremia is associated with dehydrating illnesses.

### Pathogenesis

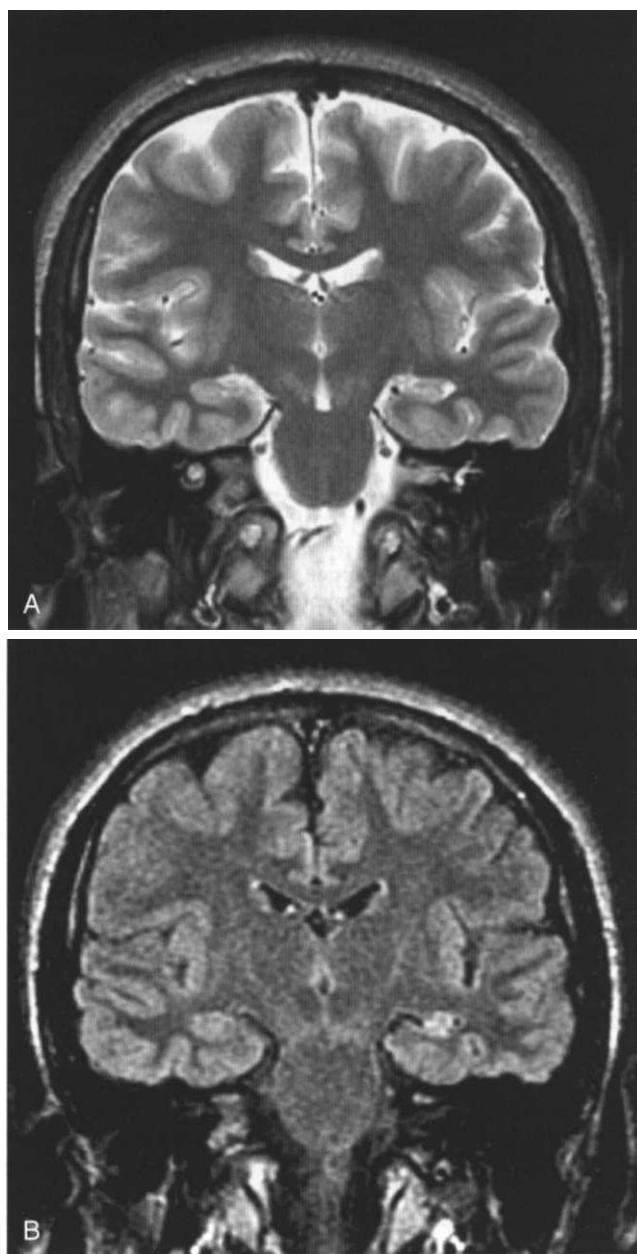
All seizure activity is a result of chaotic electrical discharge in the central nervous system (CNS). Epileptic seizures result from the sudden, excessive electrical discharges of large aggregates of neurons. Because there are no consistent, demonstrable pathologic changes in the brains of individuals with idiopathic generalized epilepsy, susceptibility to these seizures most likely results from inherited biochemical, membrane, or neurotransmitter defects that result in abnormal excitability within the involved circuits.

The main inhibitory neurotransmitter of the CNS is  $\gamma$ -aminobutyric acid (GABA). When stimulated, GABA receptors modulate chloride ion flux, inhibiting membrane depolarization. GABA antagonists or functional depletion of GABA increases membrane depolarization and may result in seizures. GABA agonists (direct or indirect) therefore play a vital role in seizure termination. Loss of GABA-mediated inhibition results in seizures.<sup>31</sup>

Activation of the N-methyl-D-aspartate (NMDA) type of glutamate receptors potentiates cellular excitability and leads to sustained neuronal depolarization and calcium influx. Extracellular potassium and intracellular calcium concentrations increase and contribute to the overall excitability of the epileptic neuronal aggregate. During the seizure itself, neurons are tonically depolarized and fire continuously in a sustained, high-frequency discharge (tonic phase). The seizure ends as phasic repolarizations interrupt the continuous firing pattern (clonic phase) and gradually restore membrane potentials to normal or to a temporary hyperpolarized state (postictal depression). Prolonged NMDA receptor activation and excessive accumulation of intracellular calcium also result in neuronal toxicity and may lead to cell death. In some areas of the cortex such as the hippocampus, subsets of neurons that normally fire in bursts may serve as pacemaker cells for other groups of neurons during epileptogenic activities. In the focal epilepsies, abnormal neuronal behavior originates in and may remain confined to a restricted area of the cortex.<sup>19</sup>

The thalamus plays a role in generating generalized seizures and the generalized spike-wave EEG patterns that accompany them. The substantia nigra also is crucial to the expression of generalized convulsions, especially the tonic phase; GABAergic inhibitory transmission in the substantia nigra plays a regulatory role in the propagation of primary and secondarily generalized seizure discharges. Generalized seizures and the rhythmic spike-wave discharges are dependent on ionic conductance of the neurons in the thalamic nucleus reticularis, allowing them to function as pacemaker control cells.

About 70% to 80% of complex partial seizures arise from the temporal lobe, and more than 65% of these originate in mesial temporal lobe structures, especially the hippocampus, amygdala, and parahippocampal gyrus. Remaining cases of complex partial seizures arise mainly from the frontal lobe, with smaller percentages originating in the parietal and occipital lobes. Hippocampal or mesial temporal sclerosis, or scarring, is characterized by variable degrees of pyramidal cell loss and gliosis in the hippocampal subfields and dentate gyrus. This



**Figure 36-1**

Temporal mesial sclerosis—coronal magnetic resonance image (MRI). The coronal projection is essential to reveal hippocampal abnormalities. Fluid-attenuated inversion recovery (FLAIR) MRI is superior to T2 weighting to show signal abnormalities, because the saturation nullifies the signal from the cerebrospinal fluid. **A**, A T2-weighted scan shows volume reduction of the left hippocampus. **B**, A FLAIR sequence shows the abnormal high signal (arrow), not seen on the T2 scan. [From Adam A, Dixon AK, Grainger RG, et al, eds: *Grainger and Allison's diagnostic radiology: a textbook of medical imaging*, ed 4, Philadelphia, 2001, Churchill Livingstone.]

condition represents the most common pathologic substrate of partial epilepsy in adolescents and adults.<sup>11</sup> Fig. 36-1 shows mesial sclerosis identified through coronal magnetic resonance imaging (MRI).

The hippocampus plays a critical role in both seizure activity and mood disorders. This suggests that pathology in this area of the brain might provide a link between

epilepsy and depression. Remodeling of the hippocampal spine synapses may play a significant role in the neurobiology of depression and the effects of antidepressant therapy. Because the effects of estrogens on hippocampus parallel those of antidepressants, loss of estrogen appears to be a critical contributor to the etiology of depressive disorders. The increased incidence of depression observed in women with epilepsy might therefore reflect a hormonal deficiency state, although it is probably not the only factor that contributes to depression.<sup>8</sup>

Shifts in levels of brain-derived neurotrophic factor (BDNF) in the brain may represent another common link between the distinctive patterns of epilepsy and depression seen in women. Seizure incidence varies across the reproductive cycle, peaking in the periovulatory period. Dramatic fluctuations in estrogen levels in women may explain their greater vulnerability to depression, even as estrogen-related surges in BDNF expression may lead to an increased propensity for seizures.

Another potential explanation of the facilitation of seizures is *kindling*. Kindling refers to the processes that mediate long-lasting changes in brain function in response to repeated, gradually augmented stimulation of the brain resulting in epileptiform activity. Kindling also refers to sensitization of neuronal tissue by the addition of a drug or electrical stimulus that renders it susceptible to subsequent seizure activity. This may explain why exposures to localized, repetitive, low-intensity electrical stimuli induce increasingly pathologic responses. This is a possible mechanism that exists between the occurrence of a brain insult and the later onset of epilepsy. The kindling model is currently used in the study of partial epilepsy, particularly that involving the mesial temporal structures.<sup>9</sup>

Chronic focal epileptogenic lesions can cause distant areas to become capable of generating abnormal electrical discharges and seizures. This focus continues to function independently even after ablation of the primary lesion. This has important implications for the surgical treatment of epilepsy.

### Clinical Manifestations

**Signs and Symptoms.** In most individuals, seizures occur unpredictably at any time and without any relationship to posture or ongoing activities.<sup>10,14</sup> In some individuals, seizures are provoked by specific stimuli such as flashing lights or a flickering television. See Box 36-2 for the typical triggers to seizure. The presence of focal signs after the seizure suggests that the seizure may have a focal origin. Prodromal symptoms (premonitory symptoms that indicate an impending seizure) may include headache, mood alterations, lethargy, and myoclonic jerking.

In the tonic phase of a tonic-clonic seizure, the body becomes rigid and the person is at risk of falling. A cry may be uttered, and the person may become cyanotic. In general the jaw is fixed and the hands are clenched. This phase usually lasts for 30 to 60 seconds. The clonic phase begins with rhythmic, jerky contractions and relaxation of all body muscles, especially in the extremities. Fig. 36-2 demonstrates the tonic and clonic phases of seizure. Biting of the tongue, lips, or inside of the mouth may

### Box 36-2

#### EVENTS THAT MAY TRIGGER SEIZURE

- Stress
- Poor nutrition
- Missed medication
- Skipping meals
- Flickering lights
- Illness
- Fever and allergies
- Lack of sleep
- Emotions such as anger, worry, fear
- Heat and humidity



A Tonic phase



B Clonic phase

**Figure 36-2**

Tonic and clonic phases of seizure activity. (From Black JM: Medical-surgical nursing: clinical management for positive outcomes, ed 7, Philadelphia, 2004, Saunders.)

occur. Saliva is blown from the mouth, with a froth appearing on the lips.

Minor motor seizures are reflected by involuntary jerking of the major muscles (myoclonus), momentary loss of muscle movement (akinesia), or total loss of muscle tone, which causes the person to fall to the floor (ataxia).

Nonconvulsive status epilepticus is difficult to define, but it may be described as a syndrome in which the most fundamental feature is a change in the individual's behavior. A degree of clouding of mental processes occurs, ranging from drowsiness and confusion to disorientation and dysphagia.

### MEDICAL MANAGEMENT

**DIAGNOSIS.** The history obtained from the client and the observations of bystanders are of importance in establishing a diagnosis and classifying the seizure disorder correctly. To determine seizure type, a series of questions must be answered regarding the location of the seizure activity, the level of consciousness, the level of generalized motor activity, and the preceding level of seizure

activity. The state of the client after the episode, including the level of confusion, sleepiness, or headache, must be determined. These data may be gathered from the paramedic or emergency personnel in the case of an individual who has been found on the floor.

It is important to recognize events that may mimic a seizure but are not related to the diagnosis of epilepsy. Early morning confusion and headache could be related to hypoglycemia in individuals on hypoglycemic medications. If the event is a transient ischemic attack, a loss of consciousness usually does not occur, and the neurologic insult is in the form of weakness or numbness. With a seizure, however, a twitching and tingling is evident. Behavioral disturbances as seen in dementia usually have a predictable pattern, such as occurring at a certain time of day.<sup>12</sup> Syncope, or a transient loss of consciousness, is related to an acute change in cerebral perfusion. The related transient cerebral anoxia may itself cause seizure activity. Recurrent cardiac arrhythmia can be confused with recurrent seizures.<sup>17</sup>

On the other hand, epileptic events may not be recognized. Because the temporal lobe is the terminus of the vestibular pathways, the symptoms of activity may be nonspecific dizziness, or episodic vertigo. Fluctuations in mental status may be attributed to other causes or regarded as a dementing process.

**Electroencephalography.** The EEG has a central role in the diagnosis of epilepsy. The EEG records the integrated electrical activity generated by synaptic potentials in neurons in the superficial layers of a localized area of cortex. In the epileptic focus, neurons in a small area of the cortex are activated for a brief period in a synchronized pattern and then inhibited. Interictal (between-seizure) activity on the EEG provides strong presumptive evidence that the event was a seizure. The best way to diagnose the presence of seizures and to classify the seizure is to observe, simultaneously, the seizure and the associated EEG recording. A normal reading does not rule out the diagnosis, and in the older individual, it may be more difficult to distinguish normal from abnormal.<sup>30</sup>

Prolonged EEG monitoring with simultaneous closed-circuit video recording is reserved for complicated cases of protracted and unresponsive seizures. It provides an invaluable method for recording ictal seizure events that are rarely captured during routine EEG studies. This technique is extremely helpful in the classification of seizures because it can accurately determine the location and frequency of seizure discharges while recording alterations in the level of consciousness and the presence of clinical signs.<sup>14</sup>

Metabolic studies are ideally performed at the time of the seizure occurrence, when values are most likely to be abnormal. Lumbar puncture should be considered for children with repeated seizures and other evidence of neurodevelopmental disability. It may be useful for detecting low cerebrospinal fluid glucose in glucose transporter disorder; alterations in amino acids, neurotransmitters, or cofactors in metabolic disorders; or evidence of chronic infection.

Aside from glucose determination, laboratory testing such as serum electrolyte levels and toxicology screening should be ordered based on individual clinical circum-

stances, such as evidence of dehydration. An EEG is not warranted after a simple febrile seizure but may be useful for evaluating individuals with an atypical feature or with other risk factors for later epilepsy. Similarly, neuroimaging is also not useful for children with simple febrile convulsions but may be considered for children with atypical features, including focal neurologic signs or pre-existing neurologic deficits.<sup>14</sup>

Specialized neuropsychologic testing is often needed in evaluating clients with seizures. These tests not only help determine general intelligence and state of brain functioning but also often help to localize lesions.

A radiographic examination of the brain is indicated to rule out mass effect or vascular disease. MRI is the method of choice, since the lesions that cause epilepsy are often subtle.

**TREATMENT.** Control of seizures is paramount in the treatment of epilepsy, but it is only part of the treatment. Support and education provided by health care professionals is critical to amelioration of the behavioral, social, and economic consequences of uncontrolled seizures. Reassurance should be given that for most individuals, epilepsy does not indicate serious brain damage. The risk-to-benefit ratio for antiepileptic treatments is one of the biggest challenges in seizure management. Because anti-epileptic drugs have various adverse effects, which may interfere with normal developmental processes and affect cognitive functions, physicians must weigh the adverse effects of more aggressive treatments against the benefits of complete seizure control.<sup>4</sup>

Antiepileptic drugs carry risks of side effects that are particularly important in children. The decision as to whether or not to treat children and adolescents who have experienced a first unprovoked seizure must be based on a risk-benefit assessment that weighs the risk of having another seizure against the risks of long-term anti-epileptic drug therapy. There appears to be no benefit of treatment with regard to the prognosis for long-term seizure remission.<sup>13</sup> Suppressing interictal discharges can improve behavior in children with epilepsy and behavioral problems, particularly partial epilepsy. Focal discharges may be involved in the underlying mechanisms of behavioral problems in children with epilepsy.<sup>23</sup>

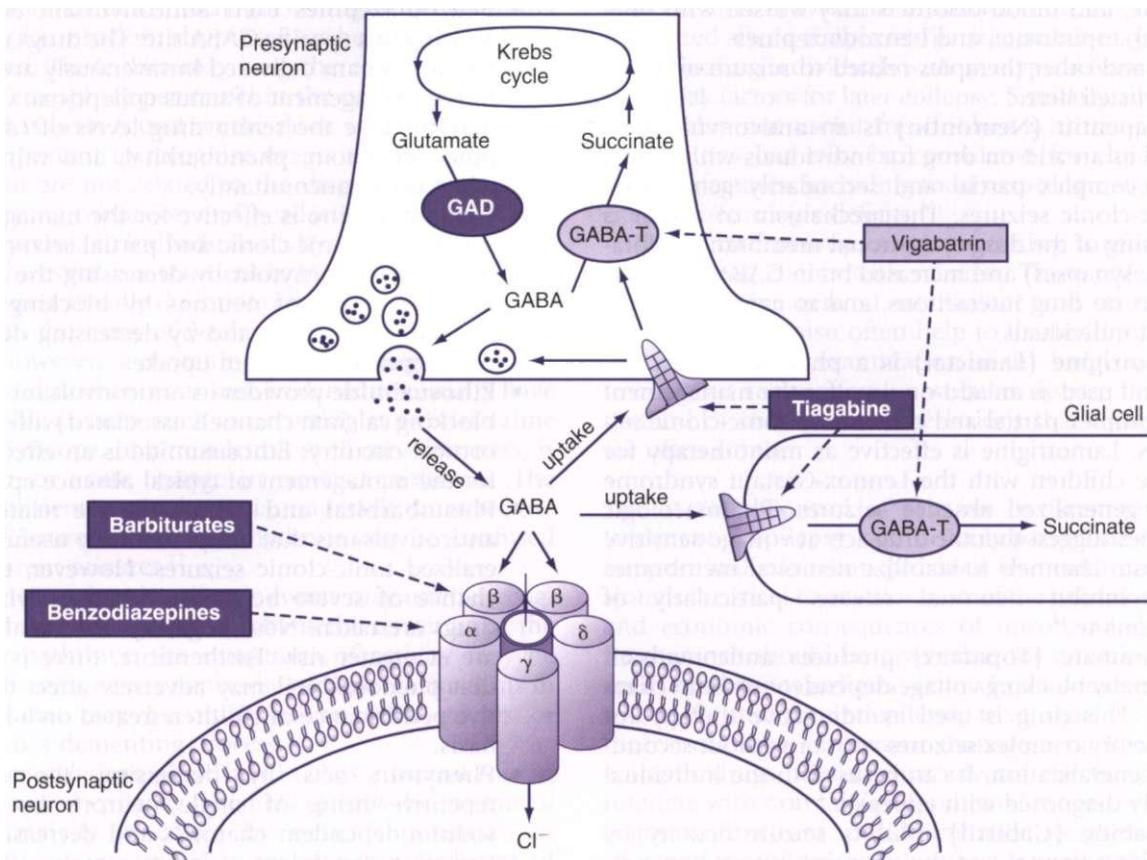
Antiepileptic medications are used according to the type of seizure. In most people with seizures of a single type, satisfactory control can be achieved with a single anticonvulsant drug. Monitoring of plasma drug levels has increased the ability to maintain the drug at the maximal tolerated dose. Generally, drug testing should be performed during the course of treatment and again when good seizure control has been established. Subsequent tests may be performed with changes in control or if side effects occur. Dose adjustments are carefully monitored, and noncompliance can be identified. It sometimes takes a trial of several different medications at different doses to find the best fit. In people who do not comply with the drug regimen, no drug may be better than an inconsistent dosage.<sup>1,3</sup>

Older individuals more often have difficulty with the neurologic side effects of dizziness, imbalance, drowsiness, and tremors. Cognitive deficits may worsen with

topiramate, and mood disorders may worsen with phenobarbital, topiramate, and benzodiazepines.

Drugs and other therapies related to seizure management are listed here:

- **Gabapentin (Neurontin)** is an anticonvulsant used as an add-on drug for individuals with refractory complex partial and secondarily generalized tonic-clonic seizures. The mechanism of action is binding of the drug to neuronal membranes (glutamate synapses) and increased brain GABA turnover. It has no drug interactions, and so can be used for most individuals.
- **Lamotrigine (Lamictal)** is a phenyltriazine compound used as an add-on drug for the management of complex partial and generalized tonic-clonic seizures. Lamotrigine is effective as monotherapy for some children with the Lennox-Gastaut syndrome and generalized absence seizures. Pharmacologic studies suggest that the drug acts at voltage-sensitive sodium channels to stabilize neuronal membranes and inhibit neuronal release, particularly of glutamate.<sup>14</sup>
- **Topiramate (Topamax)** produces anticonvulsant action by blocking voltage-dependent sodium channels. This drug is used as adjunctive therapy for refractory complex seizures with or without secondary generalization. It can be used in the individual newly diagnosed with epilepsy.<sup>14</sup>
- **Tiagabine (Gabitril)** inhibits seizure activity by blocking reuptake of the neuroinhibitory transmitter GABA into neuronal and glial cells and is effective in the management of complex partial seizures as an add-on drug.
- Levetiracetam (Keppra) acts by an unknown mechanism and is indicated for use as an adjunctive treatment for partial seizures.
- **Retigabine** is structurally and functionally unrelated to any antiepileptic drugs in use today and was discovered through screening. The drug acts on the potassium channels and through an ancillary mechanism of potentiating GABA-evoked currents. Since the discovery that mutations in the genes that encode KCNQ2 and KCNQ3 are associated with benign familial neonatal convulsions, more attention has been paid to retigabine. The drug is particularly effective against kindled seizures. Retigabine is also effective against neuropathic pain in animals.<sup>21</sup>
- Divalproex **sodium** (Depakote) and **sodium valproate** (Valproate) are from the chemical compound valproic acid and are broad-spectrum anticonvulsants. They act by blocking voltage-dependent sodium channels and increase calcium-dependent potassium conductance. These drugs are useful for the management of many seizure types, including generalized tonic-clonic, absence, atypical absence, and myoclonic seizures. They rarely induce behavioral changes but are associated with mild gastrointestinal disturbances, alopecia, tremor, and hyperphagia. Two rare but serious side effects of valproate are a Reye-like syndrome and irreversible hepatotoxicity.<sup>14</sup>
- Benzodiazepines exert anticonvulsant activity by binding to a specific GABA site. The drugs diazepam and lorazepam delivered intravenously are used for initial management of status epilepticus. Clobazam may increase the serum drug levels of carbamazepine, phenytoin, phenobarbital, and valproic acid when used concomitantly.
- Carbamazepine is effective for the management of generalized tonic-clonic and partial seizures. It acts similarly to phenytoin by decreasing the sustained repetitive firing of neurons by blocking sodium-dependent channels and by decreasing depolarization-dependent calcium uptake.
- Ethosuximide provides its anticonvulsant action by blocking calcium channels associated with thalamocortical circuitry. Ethosuximide is an effective drug for the management of typical absence epilepsy.
- Phenobarbital and primidone are relatively safe anticonvulsants that are particularly useful for generalized tonic-clonic seizures. However, there is a chance of severe behavioral changes when these drugs are taken. Neurologically abnormal children are at greater risk. Furthermore, there is evidence that phenobarbital may adversely affect the cognitive performance of children treated on a long-term basis.
- Phenytoin acts by decreasing the sustained repetitive firing of single neurons by blocking sodium-dependent channels and decreasing depolarization-dependent calcium uptake. Phenytoin is used for primary and secondary generalized tonic-clonic seizures, partial seizures, and status epilepticus.<sup>14</sup>
- Vigabatrin acts by binding to the degradative enzyme GABA transaminase receptor, causing an increase in GABA levels and inhibition of neurotransmission. It is effective in infantile spasms, particularly in children with tuberous sclerosis. Fig. 36-3 diagrams the pharmacologic effects of some antiepileptic drugs at the GABA class A ( $\text{GABA}_A$ ) receptor.
- Adrenocorticotropic hormone (**ACTH**) is the preferred drug for the management of infantile spasms, although the dose and duration of therapy are not uniform. Prednisone is equally effective. ACTH and prednisone are equally effective for the treatment of cryptogenic and symptomatic seizures, and control can be expected in approximately 70% of individuals. There is no relationship between the ease or degree of seizure control and ultimate neurologic and cognitive outcome. The response to medication is usually apparent within a few weeks of therapy, but one third of individuals who respond suffer relapse when the ACTH or prednisone is discontinued.
- Ketogenic diet is a valuable therapeutic approach for epilepsy, used mostly with children. It is a diet high in fats, with enough protein for normal growth and energy and a low threshold for carbohydrates. Although the mechanism by which the diet protects against seizures is unknown, there is evidence that it causes effects on intermediary metabolism that influence the dynamics of the major inhibitory and excitatory neurotransmitter



**Figure 36-3**

Pharmacologic effects of antiepileptic drugs at the  $\gamma$ -aminobutyric acid class A ( $GABA_A$ ) receptor. Barbiturates bind to a  $\beta$ -subunit of the  $GABA_A$  receptor to potentiate the action of the endogenous agonist GABA and prolong the opening time of the chloride ion channel. Benzodiazepines bind to an  $\alpha$ -subunit of  $GABA_A$  to potentiate the action of GABA and increase the frequency of opening of the chloride ion channel. Vigabatrin irreversibly binds to GABA transaminase (GABA-T) to inhibit degradation of the inhibitory neurotransmitter GABA. Tiagabine blocks the uptake of synaptically released GABA into both presynaptic neurons and glial cells, allowing GABA to remain at the site of action for longer periods. GAD, Glutamic acid decarboxylase. (From Leach JP, Brodie MJ: *Lancet* 351:203, 1998.)

systems in brain. During consumption of the ketogenic diet, marked alterations in brain energy metabolism occur, with ketone bodies partly replacing glucose as fuel. Whether these metabolic changes contribute to acute seizure protection is unclear; however, the ketone body acetone has anticonvulsant activity and could play a role in the seizure protection afforded by the diet. In addition to acute seizure protection, the ketogenic diet provides protection against the development of spontaneous recurrent seizures in models of chronic epilepsy. The use of valproic acid is contraindicated in association with the ketogenic diet, because the risk of hepatotoxicity is enhanced.<sup>10</sup>

Anticonvulsive drugs are safe, but side effects do occur, especially at the start of drug therapy. Side effects of the medication may be ataxia, dysarthria, dizziness and blurring, or double vision. Fatigue is a common complaint.<sup>2</sup> When phenytoin is taken, osteomalacia may occur as a result of increased metabolism of vitamin D. Hyponatremia can occur at low doses of carbamazepine and should be of concern when combined with a diuretic or in individuals with cardiac failure.<sup>24</sup> Newer medications, such as lamotrigine, gabapentin, vigabatrin, and topiramate, are shown to be effective as add-on drugs demonstrating low

interaction with other drugs.<sup>2</sup> These drugs currently are more expensive, and studies have not shown them to be more effective when used as monotherapy compared with the current standard regimen. Allergic reactions are often in the form of a rash, and in these cases a change to another medication should be made.

When drug therapy does not control the seizures or drugs become toxic at effective dosages, then surgical treatment is indicated. Lobectomies, cortical resections, and sectioning of the corpus callosum are the types of surgeries most often performed. Hemispherectomies, the removal of one of the hemispheres, can be effective for the severe, uncontrollable seizures usually found in children.

Vagal nerve stimulation can be provided through an implantable pulse generator. By stimulating the left vagal nucleus, an inhibitory projection influences the entire cerebral cortex. A 50% reduction in seizure with vagal nerve stimulation has been reported. Recent studies indicate that it may be a safe adjunctive therapy for individuals with seizure disorders refractory to other therapies.

Among the complications associated with treatment with antiepileptic drugs is the reduction of efficacy of oral contraceptives, most of which involve hormonal intervention in the reproductive cycle to prevent ovulation

and to inhibit fertilization and implantation of the ovum. A variety of pituitary hormone abnormalities in women with epilepsy are known to be caused by antiepileptic drugs, including elevated prolactin levels, disrupted luteinizing hormone and follicle-stimulating hormone levels, and abnormal concentrations of sex hormones. Seizure control must be the primary goal of therapy, with birth control a secondary concern. Understanding the continuum of estrogenic modulation of hippocampal BDNF levels may be relevant to the understanding, treatment, and even prevention of these disorders in women, possibly by the use of novel neurotrophin- and estrogen-based therapies for their management.<sup>6</sup>

Many people with epilepsy experience a higher frequency of seizures with large doses of caffeine. Amphetamines and other stimulants should be avoided, and some asthma drugs can increase the incidence of seizure activity.

Some evidence shows that biofeedback and conditioning techniques are effective in helping control epilepsy in some people.<sup>7</sup>

**PROGNOSIS.** Persons with epilepsy have increased mortality rates compared with the general population. Much of this increased risk occurs in individuals with symptomatic epilepsy in whom mortality relates to the underlying condition.

Death from asphyxia is the greatest concern in instances when the individual has a seizure during eating or when breathing passages are compromised during the seizure or in the postepileptic phase. Drowning during a seizure has been documented and remains a serious consequence of swimming or bathing alone. There is twentyfold increased risk of sudden unexplained death, presumably secondary to cardiac arrhythmia, pulmonary edema, or myocardial infarction. Sudden unexplained death is the most common cause of seizure-related mortality in persons with severe, chronic epilepsy.<sup>8</sup> With status epilepticus, 3% of children and 10% of adults die during an attack.<sup>22</sup>

Duration of epilepsy is a composite factor that probably reflects the influence of several factors acting in combination. The longer an individual suffers from intractable epilepsy, the higher the possibility of exposure to factors that result in seizure-induced neuronal damage, the more prolonged the exposure to antiepilepsy drugs, and the greater the risk of seizure-related accidents, including closed head injuries.<sup>18</sup>

The correlation between depression and epilepsy is strong, and attempted suicide is higher than the norm in those with epilepsy. People with epilepsy are more likely to be hospitalized for depression. Young people are at particular risk for development of depression and other psychiatric disorders and are less likely to be treated. Despite evidence that two thirds of children with epilepsy have a diagnosable psychiatric disorder, only about one third receive psychiatric treatment.<sup>9</sup>

In people with epilepsy of no known cause and for whom a diagnosis is made before age 10, a 75% remission rate (defined as five seizure-free years) is seen. A child with epilepsy who has been free of seizures for more than 4 years while taking antiepileptic drugs has about a

70% chance of remaining in permanent remission when the drugs are withdrawn.

Chronic epilepsy is more likely when associated neurologic impairment is present at birth, when the seizures begin before 2 years of age, and when complex partial seizures are predominant.<sup>16</sup>

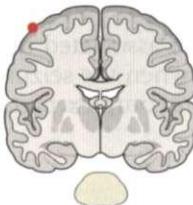
## Classification of Seizures

An essential distinction in the modern system of classification is between partial and generalized seizures. Some classifications include a third group, partial seizures progressing to generalized seizures. Fig. 36-4 shows the seizure types, the typical area of the brain in which the seizure begins, and where it may travel. The following seizure types have been classified by the International League Against Epilepsy (Box 36-3). The initial events of a seizure, described either by the individual or by an observer, are usually the most reliable indication to determine whether a seizure begins focally.<sup>9</sup> Fig. 36-5 charts the proportion of seizure types as a function of age.

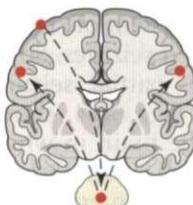
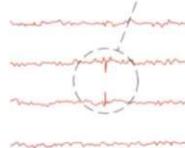
### Partial Seizures

Partial seizures have clinical or EEG evidence of a local onset. The abnormal discharge usually arises in a portion of one hemisphere and may spread to other parts of the brain during a seizure. The syndrome is characterized by the locus of onset of the attacks. Consciousness is not depressed, and individuals can interact normally with their environment except for limitations imposed by the seizure on specific localized brain functions. The seizures are identified as temporal, frontal, parietal, or occipital. Sensory symptoms such as localized paresthesias, numbness, vertigo, auditory hallucinations, and unformed visual hallucinations occur with seizures beginning in the corresponding primary sensory areas. Psychoillusions arise from ictal discharges in limbic and association cortex and include dysmnesic symptoms, such as feelings of familiarity (*deja vu*) and unfamiliarity (*jamais vu*); dreamy states, such as feelings of unreality and depersonalization; time distortion; emotional symptoms, such as fear or depression; visual illusions, such as multiple images (*polyopsia*) or distortions of size (*micropsia* and *macropsia*); and hallucinatory phenomena, such as unpleasant smells, stereotyped visions, or familiar voices. Autonomic symptoms reflect ictal involvement of limbic structures that lie in the mesial temporal or frontal lobe and project to the hypothalamus and brainstem.<sup>19</sup> This group encompasses the most frequent and severe epilepsy problems of adults. Partial seizures can be subdivided into three groups: simple partial seizures, complex partial seizures, and partial seizures becoming secondarily generalized.

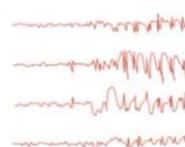
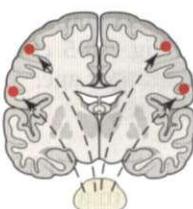
Many symptoms or phenomena can be the expressions of simple partial seizures. Subjective sensory and psychoillusions collectively are termed *auras* and affect about 60% of individuals with focal epilepsy. Examples of autonomic phenomena include an epigastric rising sensation (especially common with seizures beginning in the mesial temporal lobe), nausea, lightheadedness, pallor or flushing, pupillary dilation, piloerection, salivation, and urinary incontinence.

**CORTICAL ORIGIN****Partial (focal) seizures**

- Simple partial seizure:
  - Motor
  - Sensory
- Complex partial seizure, Syn. Psychomotor temporal lobe epilepsy

**Focal EEG abnormality****Partial seizures progressing to generalized tonic/clonic (grand mal) epilepsy**

- Simple or → progressing
- Complex partial

**Focal → generalized EEG abnormality****SUBCORTICAL ORIGIN****Generalized seizures**

- Absences (petit mal)
- Tonic seizures
- Tonic/clonic seizures (grand mal)
- Akinetic seizures
- Infantile spasms

**Generalized EEG abnormality****Box 36-3****INTERNATIONAL LEAGUE AGAINST EPILEPSY CLASSIFICATION OF EPILEPTIC SEIZURES AND SEIZURES AND SYNDROMES****Classification of Seizures****Partial Seizures (Focal Origin)**

- Simple partial seizures (consciousness not impaired)
  - With motor signs (including jacksonian, versive, and postural)
  - With sensory symptoms (including visual, somatosensory, auditory, olfactory)
  - With psychic symptoms (including dysphasia, hallucination, and affective changes)
  - With autonomic symptoms
- Complex partial seizures (consciousness is impaired)
  - Simple partial onset followed by impaired consciousness
  - With impairment of consciousness at onset
  - With automatisms
- Partial seizures evolving to secondarily generalized seizures

**Generalized Seizures (Nonfocal Origin)**

- Absence seizures
- Myoclonic seizures; myoclonic jerks (single or multiple)
- Tonic-clonic seizures
- Tonic seizures
- Atonic seizures

**Unclassified Epileptic Seizures****Classification of Epileptic Syndromes****Idiopathic Epilepsy Syndromes (Focal or Generalized)**

- Benign neonatal convulsions
- Benign partial epilepsy of childhood

- Childhood absence epilepsy
- Juvenile myoclonic epilepsy
- Idiopathic epilepsy, otherwise unspecified

**Cryptogenic or Symptomatic Epilepsy Syndromes (Focal or Generalized)**

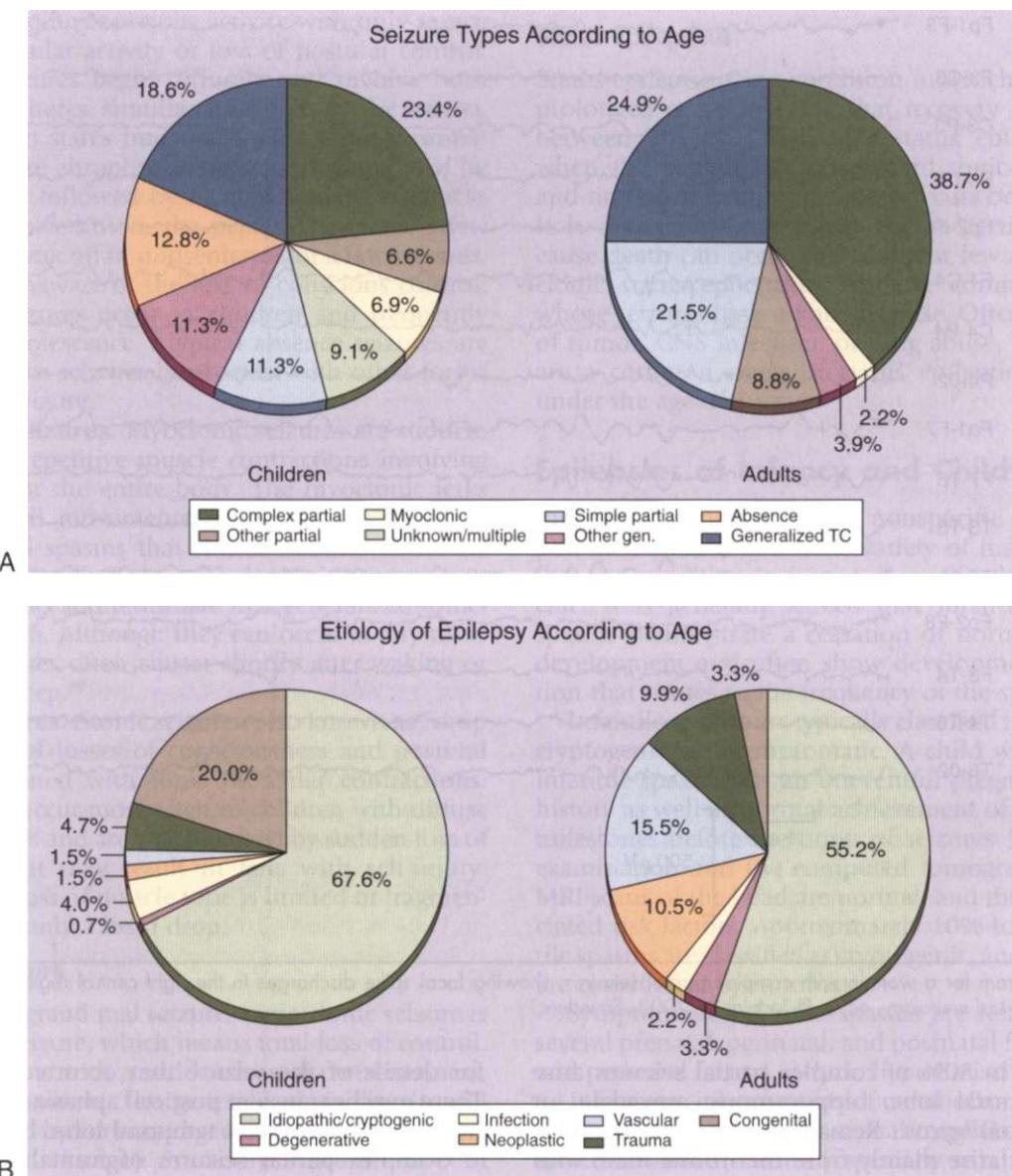
- West's syndrome (infantile spasms)
- Lennox-Gastaut syndrome
- Epilepsia partialis continua
- Temporal lobe epilepsy
- Frontal lobe epilepsy
- Posttraumatic epilepsy
- Other symptomatic epilepsies, otherwise unspecified

**Other Epilepsy Syndromes of Uncertain or Mixed Classification**

- Neonatal seizures
- Febrile seizures
- Reflex epilepsy
- Adult nonconvulsive status epilepticus
- Other unspecified

**Figure 36-4**

The nature of the attack is used to classify seizure types. (From Lindsay KW, Bone I, Callander R: *Neurology and neurosurgery illustrated*, New York, 1986, Churchill Livingstone.)

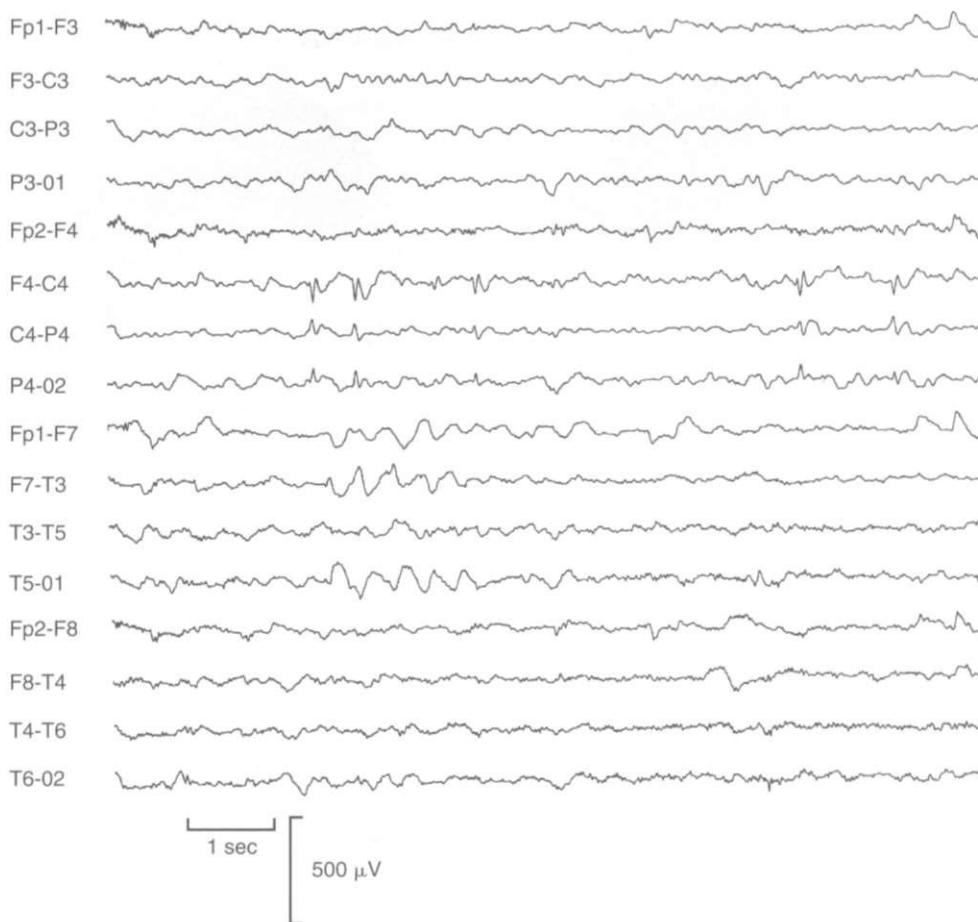
**Figure 36-5**

**A**, Proportion of seizure types as a function of age in newly diagnosed cases of epilepsy in Rochester, Minnesota, 1935 to 1984. **B**, Etiology of epilepsy in all newly diagnosed cases in Rochester, Minnesota, 1935 to 1984. TC, Tonic-clonic. (Modified from Hauser WA, Annegers JF, Kurland LT: Incidence of epilepsy and unprovoked seizures in Rochester, Minnesota: 1935-1984, *Epilepsia* 34:453-468, 1993.)

**Simple Partial Seizures.** Simple partial seizures (formerly known as focal seizures) are associated with preservation of consciousness and unilateral hemispheric involvement. Simple partial seizures result when the ictal discharge occurs in, and remains limited to, a circumscribed area of cortex. This site often is termed the epileptogenic focus. They may be manifested by focal motor symptoms (jerking) or somatosensory symptoms (paresthesias or tingling) that spread to different parts of the body. Because of their large cortical representation, muscles of the face and hand often are involved. Ictal discharges often involve supplementary or other secondary motor areas of the frontal lobe and produce contralateral flexion and elevation of the arm, contralateral turning of the head and eyes, and tonic extension of the ipsilateral arm (the so-called fencer's posture). Light

flashes, buzzing, or abnormal sensations of taste and smell represent involvement of the areas of the brain that control visual, auditory, or olfactory and gustatory responses. Sometimes autonomic symptoms create nausea, pallor, flushing, or pupillary dilation. Cognitive and affective changes can occur. Psychotic responses to seizure activity include illusions or hallucinations, and a sudden sense of fear is common. Focal weakness may follow a simple partial motor seizure; numbness, a sensory seizure; and blindness or amblyopia, an occipital lobe seizure. These reversible neurologic deficits collectively are called Todd's paralysis and rarely last for more than 48 hours.

**Complex Partial Seizures.** Complex partial seizures are associated with alteration or loss of consciousness and bilateral hemispheric involvement.



**Figure 36-6**

Electroencephalogram for a woman with complex partial seizures, showing focal spike discharges in the right central region. (From Goetz CG, ed: *Textbook of clinical neurology*, ed 2, Philadelphia, 2003, Saunders.)

About 70% to 80% of complex partial seizures arise from the temporal lobe, hippocampus, amygdala, or parahippocampal gyrus. Remaining cases of complex partial seizures arise mainly from the frontal lobe, with smaller percentages originating in the parietal and occipital lobes. Many complex partial seizures evolve from simple partial seizures; consciousness becomes impaired as the seizure progresses. Complex partial seizures preceded by an olfactory aura are called uncinate fits because of their origin in or near the uncus of the medial temporal lobe. Uncinate fits may have a higher association with brain tumors than other types of complex partial seizures. In temporal lobe seizures, loss of consciousness results when the ictal discharge spreads bilaterally to involve areas of the hippocampus and amygdala; the parahippocampal gyri; and, to some extent, the entorhinal cortex and frontal regions.<sup>19</sup>

The person appears dazed and confused with random walking, mumbling, head turning, or pulling at clothing. Automatic behaviors, without conscious control, may manifest but cannot be recalled by the person. There may be clumsy perseveration of ongoing motor tasks, such as eating, drawing, walking, or washing dishes. Complex partial seizures usually last 45 to 90 seconds and are followed by confusion and disorientation lasting several more minutes. Characteristically, individuals are amnesic

for details of the seizure that occurred after the aura. There may be transient postictal aphasia when the seizure involves the dominant temporal lobe.

Complex partial seizures of frontal lobe origin are atypical and often differ dramatically from seizures originating in the temporal lobe. Although there are many variations, frontal lobe complex partial seizures tend to begin and end abruptly; have few, if any, postictal symptoms; and involve often bizarre motor manifestations, such as asynchronous thrashing or flailing of arms and legs, pelvic thrusting, pedaling leg movements, and loud vocalizations that may appear as psychiatric disorders.<sup>19</sup> Fig. 36-6 presents the EEG of a woman with complex partial seizures, showing focal spike discharges in the right central region.

**Partial Seizure Evolving to Secondarily Generalized Seizures.** A partial seizure secondarily generalized is a generalized tonic-clonic seizure that develops from either a simple partial seizure or a complex partial seizure and has convulsive manifestations.

#### Generalized Seizures

In a generalized seizure, localized onset is not evident and the brain shows diffuse EEG abnormalities.

**Absence Seizures.** Generalized absence seizures, formerly called petit mal seizures, consist of the sudden

cessation of ongoing conscious activity with only minor convulsive muscular activity or loss of postural control. Generalized seizures begin diffusely and involve both cerebral hemispheres simultaneously from the outset. The person often stares into space. Onset and termination of attacks are abrupt. Absences are not preceded by an aura and are followed by normal activity. If attacks occur during conversation, the person may miss a few words or may break off in midsentence for a few seconds. The person is unaware of the loss of conscious control. Often, these seizures occur in children and frequently disappear by adolescence. Atypical absence seizures are similar to absence seizures but coexist with other forms of generalized seizure.

**Myoclonic Seizures.** Myoclonic seizures are sudden, brief, single or repetitive muscle contractions involving one body part or the entire body. The myoclonic jerks range from small movements of the face or hands to massive bilateral spasms that simultaneously affect the head, limbs, and trunk. Repeated myoclonic seizures may seem to crescendo and terminate in a generalized tonic-clonic convulsion. Although they can occur at any time, myoclonic seizures often cluster shortly after waking or while falling asleep.<sup>19</sup>

**Atonic Seizures.** Atonic seizures, also known as "drop attacks," are brief losses of consciousness and postural tone not associated with tonic muscular contractions. Atonic seizures occur most often in children with diffuse encephalopathies and are characterized by sudden loss of muscle tone that may result in falls with self-injury. Sometimes the loss of muscle tone is limited or fragmentary, producing only a head drop.

#### Tonic-Clonic Seizure

Formerly called grand mal seizure, tonic-clonic seizure is the archetypal seizure, which means total loss of control. The seizure begins with a sudden loss of consciousness, and falls are common. The generalized rigidity (tonic) phase is followed by very rapid generalized jerking movements (the clonic stage). In adults, incontinence of bowel and bladder occur. Generalized tonic-clonic seizures result in many striking but transient physiologic changes, including hypoxia, lactic acidosis, elevated plasma catecholamine levels, and increased concentrations of serum creatine kinase, prolactin, corticotropin, Cortisol, (3-endorphin, and growth hormone. In the tonic phase respiration can cease briefly. Recovery may be swift after a short seizure, but a prolonged seizure may induce a deep sleep. Altered speech and transient paralysis or ataxia may follow, as well as headache, disorientation, or muscle soreness. Another seizure may follow without recovery of consciousness or, after recovery of consciousness, the person may experience seizure again.<sup>22</sup> As recovery progresses, many individuals complain of headache, muscle soreness, mental dulling, lack of energy, or mood changes lasting 24 hours. Complications include oral trauma, vertebral compression fractures, shoulder dislocation, aspiration pneumonia, and sudden death, which may be related to acute pulmonary edema, cardiac arrhythmia, or suffocation. Although this is the type of epilepsy that most people associate with the disorder, it is less common than partial seizures.

#### Status Epilepticus

Status epilepticus is a condition in which seizures are so prolonged or so repeated that recovery does not occur between attacks. Convulsive status epilepticus occurs when the person has generalized tonic-clonic seizures and no return to consciousness occurs between seizures. It is a medical emergency. The molecular events that cause death can occur with the first few seizures. Tonic-clonic status epilepticus is more common in people whose seizures have a known cause. Often it is the result of tumor, CNS infection, or drug abuse. Febrile seizures are a common cause of status epilepticus in children under the age of 3 years.

#### Epilepsies of Infancy and Childhood

Infantile spasms represent a nonspecific reaction on the part of the brain to a wide variety of insults. It is likely that the condition is more age specific than disease specific. It is generally agreed that infants who develop spasms demonstrate a cessation of normal psychologic development and often show developmental deterioration that relates to the frequency of the spasms.

Infantile spasms are typically classified into two groups: cryptogenic and symptomatic. A child with cryptogenic infantile spasms has an uneventful pregnancy and birth history as well as normal achievement of developmental milestones before the onset of seizures. The neurologic examination and the computed tomographic (CT) and MRI scans of the head are normal, and there are no associated risk factors. Approximately 10% to 20% of infantile spasms are classified as cryptogenic, and the remainder are classified as symptomatic.

Symptomatic infantile spasms are related directly to several prenatal, perinatal, and postnatal factors. Prenatal and perinatal factors include hypoxia-ischemia, congenital infections, inborn errors of metabolism, tuberous sclerosis, and prematurity. Postnatal conditions include CNS infections, head trauma (especially subdural hematoma and intraventricular hemorrhage), and hypoxic-ischemic encephalopathy. The fact that infantile spasms and immunizations often occur simultaneously around 6 months of age is a coincidence of timing rather than a cause-and-effect relationship with any immunization antigen.

Infants with cryptogenic infantile spasms have a good prognosis, whereas those with the symptomatic type have an 80% to 90% risk of mental retardation. The underlying CNS disorder has a major role in the neurologic outcome. Several theories have been advanced with regard to the pathogenesis of infantile spasms, including dysfunction of the monoaminergic neurotransmitter system in the brainstem, derangement of neuronal structures in the brainstem, and an abnormality of the immune system. Another possibility is that specified stresses or injury to an infant during a critical period of neurodevelopment causes corticotropin-releasing hormone (CRH) overproduction, resulting in neuronal hyperexcitability and seizures. The number of CRH receptors reaches a maximum in the infant brain, followed by spontaneous reduction with age, which perhaps accounts for the eventual resolution of infantile spasms even without therapy.

Exogenous ACTH and glucocorticoids suppress CRH synthesis, which may account for their effectiveness in treating infantile spasms.

### **Severe Myoclonic Epilepsy of Infancy**

Severe myoclonic epilepsy of infancy is a syndrome of early normal development followed by treatment-resistant seizures of various types and by psychomotor retardation, usually beginning around 5 to 6 months of age. These attacks are often long and may include status epilepticus. Mental retardation is noted in all cases.

### **Benign Myoclonic Epilepsy of Infancy**

Benign myoclonus begins during infancy and consists of clusters of myoclonic movements confined to the neck, trunk, and extremities. The myoclonic activity may be confused with infantile spasms; however, the EEG is normal in individuals with benign myoclonus. The prognosis is good, with normal development and the cessation of myoclonus by 2 years of age. An anticonvulsant is not indicated.

### **Lennox-Gastaut Syndrome**

Lennox-Gastaut syndrome usually begins between 1 and 6 years of age. The most common seizures are atonic-akinetic, resulting in loss of postural tone. Violent falls occur suddenly followed by immediate recovery and resumption of activity, with the attack lasting less than 1 second. Injuries to the head and face are common. Tonic attacks consist of sudden flexion of the head and trunk. Clusters of attacks are common, followed by automatic behavior. Consciousness is usually clouded rather than completely lost. Neurologic abnormalities such as spasticity are common in severely affected patients.<sup>16</sup>

### **Acquired Epileptic Aphasia (Landau-Kleffner Syndrome)**

Acquired epileptic aphasia (Landau-Kleffner syndrome) is characterized by an acquired aphasia in the absence of other neurologic abnormalities. Often epileptic seizures and psychomotor disturbances develop at the same time or shortly afterward. This disorder often begins between ages 3 and 9. The age of onset is critical for the long-term loss of language. It is postulated that recently acquired skills are particularly vulnerable to any disturbance of brain function. If the formation of intracerebral connections is prevented by abnormal neuronal firings of epilepsy during optimal periods of language learning in children, then such connections may never become functional and the ability to develop language is lost.<sup>11</sup>

### **Benign Childhood Epilepsy with Centrotemporal Spikes**

Benign childhood epilepsy with centrotemporal spikes typically occurs between the ages of 3 and 13 and is characterized by brief, simple partial hemifacial motor seizures. Childhood epilepsy with occipital paroxysms is a syndrome similar to benign childhood epilepsy with centrotemporal spikes, but it includes visual symptoms at onset, and some children have associated migraine headache.

### **Childhood Absence Epilepsy**

Childhood absence epilepsy begins between ages 4 and 10 years. The attacks may occur many times a day. Absence attacks often can be precipitated by hyperventilation. The attacks were previously described as petit mal and are characterized by a blank stare with unresponsiveness, rhythmic blinking, and, sometimes, a few small clonic jerks of arms or hands. Behavior and awareness return immediately to normal. There is no postictal period and usually no recollection that a seizure has occurred. The attacks last for between 10 and 45 seconds. Longer absence attacks are accompanied by automatisms, usually of a perseverative type, in about 70% of cases. Absence seizures commonly coexist with generalized tonic-clonic or myoclonic seizures. Untreated, absence seizures can occur hundreds of times each day, a condition referred to as pyknolespy. Fig. 36-7 shows the EEG of a child with absence epilepsy.

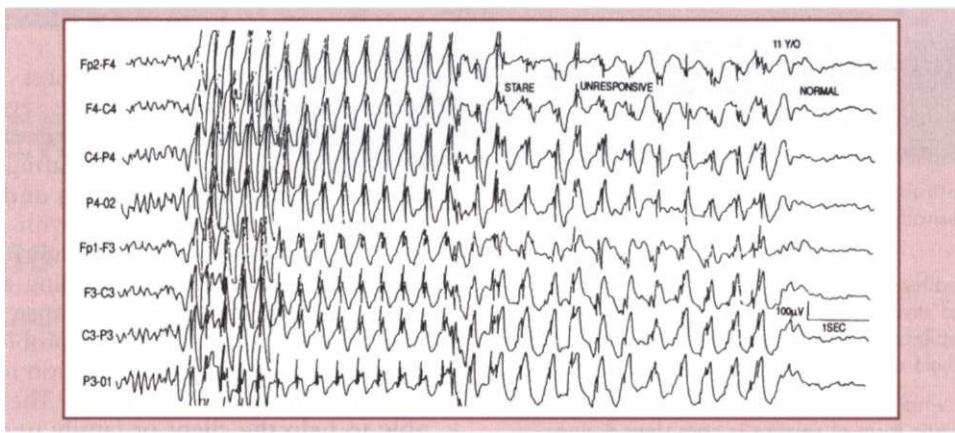
Lapses of awareness that have a more gradual onset, do not resolve as abruptly, and are accompanied by autonomic features or loss of muscle tone are called atypical absence seizures. These events occur most often in children with mental retardation, and they do not respond as well to antiepileptic drug treatment. Typical and atypical absence seizures also must be distinguished from complex partial seizures manifested only by brief lapses of consciousness, because cause, treatment, and prognosis differ among these three seizure types.<sup>19</sup>

### **Juvenile Myoclonic Epilepsy (Janz Syndrome)**

Juvenile myoclonic epilepsy (Janz syndrome) has its onset in early adolescence, between ages 8 and 20 years, in otherwise healthy individuals with normal intelligence and a family history of similar seizures. Consciousness is unimpaired, and mental retardation is not evident. It begins with early morning jerks of the head, neck, and upper limbs, making hair combing and toothbrushing difficult. As the myoclonus tends to abate later in the morning, most individuals do not seek medical advice at this stage and some deny the episodes. A few years later, early morning generalized tonic-clonic seizures develop. A gene locus has been identified on chromosome band 6p21.

### **Febrile Convulsions**

Febrile convulsions, the most common seizure disorder during childhood, generally have an excellent prognosis but may also signify a serious underlying acute infectious disease such as sepsis or bacterial meningitis. They are rare before 6 months and after age 5 and are a result of fever. The typical febrile convolution is brief, generalized, and tonic-clonic in sequence, and the body temperature is high. The seizures occur more often when the child is asleep. The convolution is often the first indication that the child is ill, because 90% of all seizures occur in the first 24 hours of fever. The rise in temperature, which may be a result of increased oxygen demands on cerebral oxidative mechanisms, may be the most important factor. Severe febrile seizures are most often unilateral, and a possibility exists for permanent brain damage, with the development of epilepsy, if the seizure lasts more than 30 minutes.<sup>16</sup>



**Figure 36-7**

Childhood absence epilepsy. Electroencephalogram shows the typical pattern of generalized 3-Hz spike-wave complexes associated with a clinical absence seizure. (From Goldman L: *Cecil textbook of medicine*, ed 22, Philadelphia, 2004, Saunders.)

Although most affected children have no long-term consequences, febrile seizures increase the risk of future epilepsy. This risk is low for most children, but increases when there is a family history of afebrile seizures or if there were neurologic abnormalities before the first febrile seizure. Febrile seizures neither are associated with nor do they cause mental retardation, below-average IQ, poor school performance, or behavior problems. Prophylactic treatment generally is not indicated because of the benign prognosis. Routine treatment of a normal infant with simple febrile convulsions includes a careful search for the cause of the fever; active measures to control the fever, including the use of antipyretics; and reassurance of the parents. Prolonged anticonvulsant prophylaxis for preventing recurrent febrile convulsions is controversial and no longer recommended. Antiepileptics such as phenytoin and carbamazepine have no effect on febrile seizures. Phenobarbital can be effective in preventing recurrent febrile seizures but may also decrease cognitive function in treated children compared with untreated children. Sodium valproate is also effective in the management of febrile seizures, but the potential risks of the drug do not justify its use in a disorder with an excellent prognosis regardless of treatment. The incidence of fatal valproate-induced hepatotoxicity is highest in children younger than 2 years of age. Oral diazepam is an effective and safe method of reducing the risk of recurrence of febrile seizures.<sup>14</sup>

### Posttraumatic Epilepsy

After penetrating wounds and other severe head injuries, about one third of individuals have seizures within 1 year. Although most individuals experience seizures within 1 to 2 years of injury, new-onset seizures still may appear 5 or more years later. Two thirds of individuals with posttraumatic epilepsy have partial or secondarily generalized seizures. Mild head injuries (e.g., uncomplicated brief loss of consciousness, no skull fracture, absence of focal neurologic signs, no contusion or hematoma) do not increase the risk of seizures to a clinically significant degree.

Impact seizures (a generalized convolution occurring at the time of, or immediately after, the injury) and early

seizures (seizures occurring within the first 1 to 2 weeks) represent acute reactions of the brain to the trauma. Seizures beginning after 10 to 14 days reflect an increased risk of posttraumatic epilepsy development.

Early seizures should be treated with phenytoin. To minimize complications from seizures occurring during acute management, phenytoin also should be given prophylactically for 1 to 2 weeks to individuals who have had severe head injuries. In the absence of overt attacks, phenytoin use should be discontinued after 2 weeks, because no data indicate that antiepileptic drugs prevent the development of later epilepsy.<sup>15</sup>

### Epilepsia Partialis Continua

Epilepsia partialis continua is characterized by continuous focal seizures that can involve part or all of one side of the body. In adults, epilepsia partialis continua occurs with severe strokes, primary or metastatic brain tumors, metabolic encephalopathies, encephalitis, and subacute or rare chronic inflammatory diseases of the brain. Antiepileptic drugs are usually ineffective, as are corticosteroids and antiviral agents. Seizures remit spontaneously in some cases. Rasmussen encephalitis is one cause of epilepsia partialis continua. The onset is usually before age 10. Sequelae include hemiplegia, hemianopia, and aphasia. The disease is progressive and potentially lethal but more often becomes self-limited with significant neurologic deficits. The disease may be due to autoantibodies that bind to and stimulate the glutamate receptors. Studies have identified cytomegalovirus in several surgical specimens of individuals with Rasmussen encephalitis.<sup>16</sup>

### SPECIAL IMPLICATIONS FOR THE THERAPIST

36-1

#### Epilepsy

##### PREFERRED PRACTICE PATTERNS

**5A:** Primary Prevention/Risk Reduction for Loss of Balance and Falling

**5B:** Impaired Neuromotor Development

**5C:** Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System

*Continued.*

**Box 36-4****COMMON MISCONCEPTIONS ABOUT EPILEPSY**

**Myth:** You can swallow your tongue during a seizure.

**Fact:** It's physically impossible to swallow your tongue.

**Myth:** You should restrain someone during a seizure.

**Fact:** Do not use restraint; the seizure will run its course and stop.

**Myth:** People with epilepsy should not be in jobs of responsibility and stress.

**Fact:** People with epilepsy hold many types of jobs; they often do not inform others of the disorder.

**Myth:** You can't tell what a person may do during a seizure.

**Fact:** The characteristic form of seizure is consistent during each episode. Behavior may be inappropriate for the time and place but will most likely not cause harm.

**Myth:** You can't die from epilepsy.

**Fact:** Status epilepticus can cause death. It should be treated as a medical emergency.

#### *System—Congenital Origin or Acquired in Infancy or Childhood*

#### **5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood**

#### **5E: Impaired Motor Function and Sensory Integrity Associated with Progressive Disorders of the Central Nervous System**

Understanding the facts about epilepsy is important for therapists who may encounter an individual with epilepsy or if a seizure occurs in the work environment. Box 36-4 gives some basic information regarding seizure.

The client who is experiencing a seizure normally only needs protection from injury in the environment. The therapist should make sure that no objects in the immediate area can be knocked onto the person and that the individual with seizure activity is lying on a surface that will prevent a fall. Rolling the person onto his or her side may help to keep the airway clear. Observation of physical manifestations, respiratory status, focal or general status, and duration of the seizure are important to the ongoing medical management. When the seizure appears to be generalized, observation of frothing at the mouth, deviation of the eyes, and incontinence will add information for the health practitioners attempting to control the client's seizures medically.

If the individual develops status epilepticus, emergency measures must be taken. Irreversible brain damage can result from hypoxia, and therefore an airway must be established, possibly through endotracheal intubation. Medication to suppress the CNS given at this point usually includes diazepam or lorazepam, and this is usually effective in controlling the seizure. If the seizure continues, phenobarbital may be administered, and if that is not successful, general

anesthesia may be given, and the person may then require ventilatory assistance.<sup>1,27</sup>

The psychologic consequences are of concern. Seizure activity can often cause severe loss of confidence and restriction of lifestyle that can lead to depression. For the therapist treating a client with epilepsy, it is important to have an understanding of the triggering activities associated with seizure. Knowing the type and frequency of seizures helps the therapist make recommendations regarding activities that can be engaged in safely. If compliance with the medication regimen appears to be a problem, the therapist should give the family information regarding the need to maintain consistent dosages. The therapist may be able to help the client or family understand the relationship between epilepsy and depression, and assist in the appropriate referral.

Evaluation of the home, work, and school environments should be performed so the therapist can make specific recommendations. The client sometimes needs to be encouraged to become part of the group activity. Too often, the client has been discouraged from engaging in sports or leisure activities even when the seizures are controlled. When a potential safety hazard exists, the therapist is well suited to recommend adaptations to equipment or the environment to maintain safety. Swimming can even be enjoyed as long as it is supervised directly.

Seizures often occur after activity, and so safety measures should be considered even after the activity is finished. Loss of fluids from sweating during exercise can affect the serum blood levels of medication and increase the metabolism of liver enzymes. Decisions about whether to engage in vigorous activity should be made on the basis of whether seizures are controlled by medication and only after close monitoring of blood levels of medication after exercise.

Side effects of medication can slow cognitive function or alter reaction time. Movement disorders, including nystagmus, ataxia, and dysarthria, may be related to medication. Lethargy, nausea, irritability, and skin rashes may be the result of intolerance to medication. When these symptoms are noted during intervention, the proper health care worker should be notified. The client and family should have a clear understanding of symptoms related to toxicity or non-therapeutic doses of medication.

Restriction of activity may be important in the first 2 to 3 months after the first seizure, after treatment is initiated and until it can be determined that further seizures are unlikely. When antiepileptic drugs are discontinued, activity should be limited initially. In the case of children for whom the epileptic syndrome progresses, limitations may change over time.

## References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. The reader can view the reference source and access it on-line whenever possible. There are a total of 31 cited references and other general references for this chapter.

# CHAPTER 37

## Headache

KENDA S. FULLER

### OVERVIEW

Headache is a common complaint in the history of clients receiving therapy. It has been determined in one study that only 4% of the total population has never experienced a headache.<sup>79</sup> Headache can be the major complaint that brings the client to therapy but is often a comorbidity that can limit participation in therapy. Individuals with chronic headaches report lifestyle changes to varying degrees. The ability to perform daily tasks is limited by feelings of fatigue, the need to stop activity in order to limit external impact such as light and sound, and the inability to concentrate on task at hand. Engaging in leisure activity is also decreased during headache periods.<sup>14</sup> Days of lost work have been carefully studied and compared to that in individuals who do not have chronic headaches and have been noted to be on average almost 6 days/yr for an individual with significant headache such as migraine. Tools designed to measure the impact of headache are the Migraine Disability Assessment (MIDAS) and the Headache Impact Test (HIT). The HIT is an Internet-based scale and can be found at [www.headachetest.com](http://www.headachetest.com). A shorter paper version is also available. These tools help to define aspects of the headache to increase communication to practitioners, describe severity, and record change over time as an outcome measure.<sup>9</sup>

Disability related to headache has been studied using the SF-36 Health Status Profile, with lower scores representing more disability than in the normal population. Scores are also lower than the population with osteoarthritis. Scores are particularly low in the physical and emotional areas, with low vitality scores, and follow the pattern of the individuals with depression. Depression occurs in 30% of individuals with headache and is strongly associated with being on disability or welfare, unemployment, and age under 50 years.<sup>45</sup>

The International Headache Society (IHS) classification system is the standard system used for both clinical and research purposes. Box 37-1 shows the major categories as defined by the IHS. According to this classification, primary chronic headaches are episodic in nature with headache-free intervals. Primary headaches are not caused by other diseases. Examples of primary headaches are migraine headaches, tension-type headaches, and cluster headaches. Fig. 37-1 illustrates the pain patterns associ-

ated with typical complaints of headache and head pain.<sup>15</sup>

The IHS classification is based on a descriptive system that categorizes primary headaches based on their symptom profiles. Alternatives for classification have been proposed that would consider the primary headaches as a continuum. For example, tension-type headaches can also have some of the symptoms ascribed to migraine when the headache is severe, and these headaches are often described as borderline migraine. Individuals with chronic migraine often also report headaches that are tension-type, and these have been called interval headaches. These headaches are not as severe in nature as the migraine. On the other hand, excessive muscle contraction does not play a critical role in tension-type headache, whereas muscle tenderness and pain are prominent in migraine. If the continuum concept is valid, the tension-type headache may be at one end of the distribution of severity and migraine headache may be at the other end. Many people have described "mixed" headache disorders in which tension headaches, posttraumatic headaches, or secondary headaches appear to trigger migraine headaches.<sup>74</sup>

Secondary headaches are caused by associated disease and are categorized by etiology. Secondary headaches have diverse causes, ranging from serious and life-threatening conditions such as brain tumors, strokes, meningitis, and subarachnoid hemorrhages to less serious but common conditions such as withdrawal from caffeine and discontinuation of analgesics. Secondary headaches can be associated with systemic illness, fever, and increased blood pressure. Although headache with hypertension is rare except with grossly high blood pressure, episodic hypertension can give rise to throbbing headache in the occipital region. Sinusitis and other general medical disorders can have headache as a symptom. Headaches caused by illness resolve when the underlying cause is successfully treated.<sup>2</sup>

Posttraumatic headache, another common but less specific headache, begins after flexion-extension injuries and is associated with brain injury. This headache may be localized to the site of injury, or it may be generalized with varying intensity. Common complaints in addition to head pain are irritability, lack of concentration, and memory loss. These symptoms are consistent with both chronic headache and mild or moderate head injury.

**Box 37-1****IHS CLASSIFICATIONS OF HEADACHES**

This classification is hierarchical and you must decide how detailed you want to make your diagnosis. This can range from the first-digit level to the fourth. First get a rough idea about the group to which the patient belongs. Is it, for example, 1. Migraine, or 2. Tension-Type Headache, or 3. Cluster Headache and Other Trigeminal Autonomic Cephalgias? Then obtain information allowing a more detailed diagnosis. The desired detail depends on the purpose. In general practice, only the first- or second-digit diagnoses are usually applied, while in specialist practice and headache centers a diagnosis at the third- or fourth-digit levels is appropriate.

**Part I: The Primary Headaches**

1. Migraine
  - 1.1. Migraine without aura
  - 1.2. Migraine with aura
  - 1.3. Childhood periodic syndromes that are commonly precursors of migraine
  - 1.4. Retinal migraine
  - 1.5. Complications of migraine
  - 1.6. Probable migraine
2. Tension-type headache
  - 2.1. Infrequent episodic tension-type headache
  - 2.2. Frequent episodic tension-type headache
  - 2.3. Chronic tension-type headache
  - 2.4. Probable tension-type headache
3. Cluster headache and other trigeminal autonomic cephalgias
  - 3.1. Cluster headache
  - 3.2. Paroxysmal hemicrania
  - 3.3. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
  - 3.4. Probable trigeminal autonomic cephalgia

**Part II: The Secondary Headaches**

5. Headache attributed to head and/or neck trauma
  - 5.1. Acute posttraumatic headache
  - 5.2. Chronic posttraumatic headache
  - 5.3. Acute headache attributed to whiplash injury [S13.4]
  - 5.4. Chronic headache attributed to whiplash injury [S13.4]
  - 5.5. Headache attributed to traumatic intracranial hematoma
  - 5.6. Headache attributed to other head and/or neck trauma [S06]
  - 5.7. Postcraniotomy headache
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to nonvascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homeostasis
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures
12. Headache attributed to psychiatric disorder

**Part III: Cranial Neuralgias, Central and Primary Facial Pain, and Other Headaches**

13. Cranial neuralgias and central causes of facial pain
14. Other headache, cranial neuralgia, central or primary facial pain

When dizziness is associated, it can further restrict activity. Posttraumatic headache is difficult to treat because of its multifactorial cause of onset and the mechanical damage associated with the trauma.

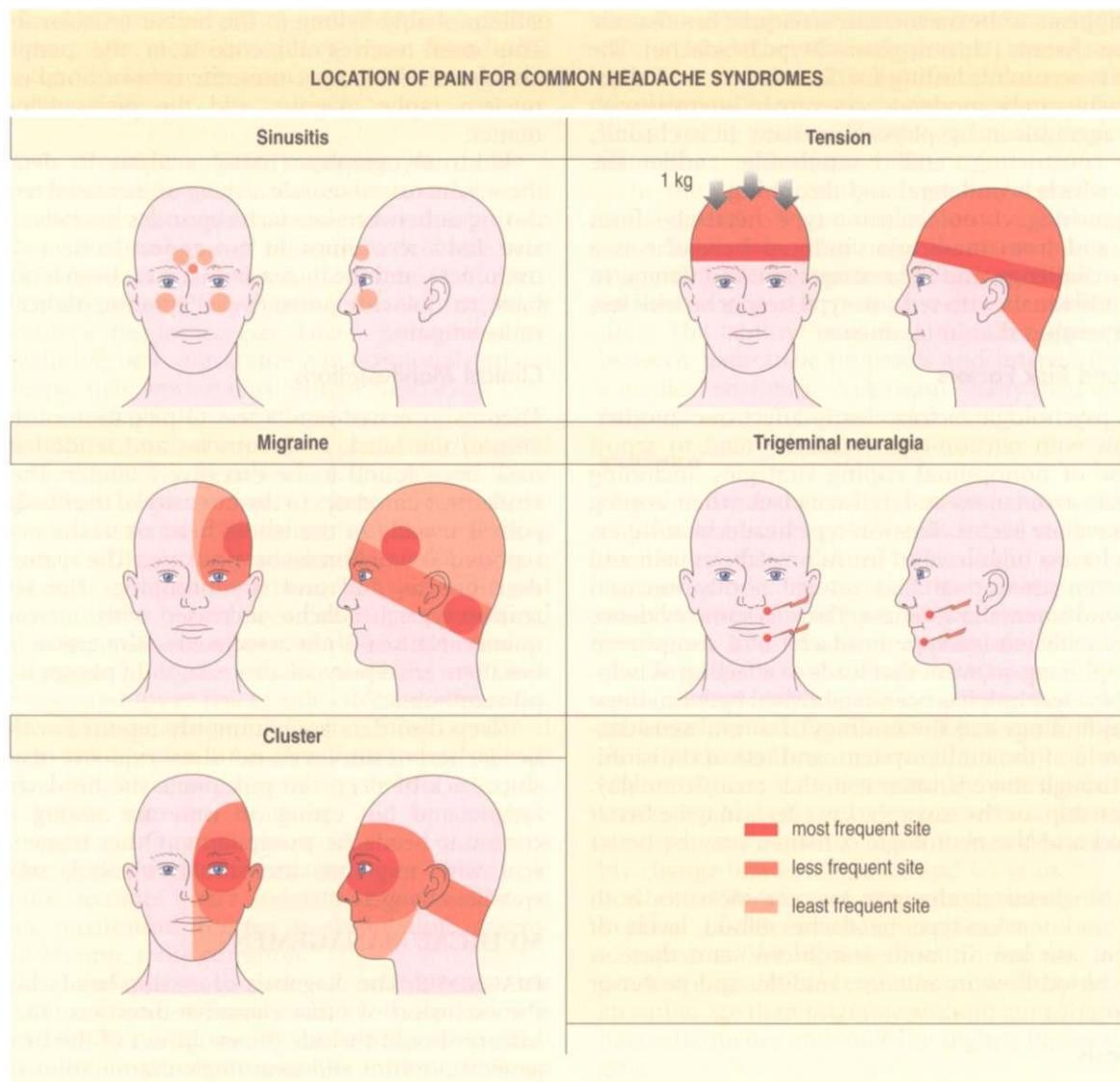
A sudden onset of severe headache is usually related to an intracranial disorder such as subarachnoid hemorrhage, brain tumor, or meningitis (see Chapter 29). These headaches will most often be accompanied by other neurologic signs, such as weakness, visual disturbances, and possibly altered mental status or coma. A structural, or space-occupying, lesion is suspected in headaches that disrupt sleep, are triggered by exertion, or cause excessive drowsiness. Headaches secondary to brain tumors, endocrinopathies, and other medical problems are only a very small percentage of all headaches.<sup>44</sup> Even though pathologic conditions account for only a few cases of headache pain, it is critical for the therapist to understand what mechanisms may be responsible and when the client may need medical or emergent care. Table 37-1 lists some of the causes of headache that may require immediate medical attention.

Headache can be difficult to evaluate; however, the intensity, quality, and site of pain may provide clues. Because the diagnosis of headache is essentially a clinical diagnosis, the physical therapist is often involved in the diagnosis. Awareness of the variety of manifestations can direct the intervention both medically and physically. Current understanding of the pathology and physiology of the headache puts it into the realm of the neurologic evaluation. The therapist needs to understand the cause, precipitating factors, and typical course of chronic headache pain. Understanding headache is a challenging task given the large number of headache syndromes and the possibility of a continuum within one diagnosis.<sup>31,68</sup>

**PRIMARY HEADACHES****Tension-Type Headache****Overview, Definition, and Incidence**

Tension-type headaches are the most common type of primary headache; as many as 90% of adults have had or will have tension headaches, representing 25 million people in the United States. Tension-type headaches occur primarily as a response to stress. Tension headaches are more common among women than men, especially with advancing age.<sup>8</sup>

The IHS divides tension-type headaches into subcategories that are believed to be varieties of the same disorder. Infrequent episodic headaches are those that occur less than 12 days/yr; frequent headaches occur from 12 to 180 days/yr. Chronic headaches are those reported more than 180 days/yr. The headaches are further defined by whether or not they are associated with pericranial pain. Prevalence appears to peak in 30- to 40-year-olds in both men and women, increasing with level of education in both genders.<sup>66</sup> The prevalence is between 16% and 39% in most studies. The prevalence of infrequent episodic headaches among 40-year-olds in the general population is 53%.<sup>62</sup>

**Figure 37-1**

Typical headache patterns. Primary headaches include tension-type, migraine, and cluster. Sinus headache, related to infection, is a common secondary headache. Trigeminal neuralgia is an example of cranial neuralgia. (See Chapter 40). (From Weinstein JM: Headache and facial pain. In Yanoff M, Duker JS, Augsburger JJ, et al, eds: *Ophthalmology*, ed 2, St Louis, 2004, Mosby.)

**Table 37-1** Pathologic Conditions Causing Headache

Pathologic Condition	Signs and Symptoms
Subdural hematoma	Mild to severe, intermittent headache; neurologic symptoms include fluctuating consciousness
Subarachnoid hemorrhage	Sudden onset, severe and constant headache; elevated blood pressure; can cause change in consciousness
Increased cranial pressure	Mild to severe headache; neurologic symptoms include hemiparesis, visual changes, and brainstem symptoms, such as vomiting, altered consciousness
Meningitis, viral and bacterial	Severe headache with radiation down neck; acute illness and fever; positive Kernig's sign
Brain abscess	Mild to severe headache; local or distant infection; fever may not be present; neurologic signs consistent with local site of infection
Central nervous system	Localized headache and focal neurologic symptoms; cranial nerve symptoms often seen
Central nervous system neoplasm	Localized headache and focal neurologic symptoms; cranial nerve symptoms often seen
Toxicity	Generalized headache, pulsating; other signs of toxicity may be present
Sinusitis	Frontal or dull headache, usually worse in morning; increased pain in cold damp air; nasal discharge
Otitis media, mastoiditis	Feeling of fullness in ear, stabbing pains in head, vertigo, and tinnitus

There appears to be an increase in headaches that are regarded as chronic (chronic tension-type headache). The headache is recurrent, lasting for 30 minutes to 7 days. The headache can be moderate to severe in intensity with minimal aggravation by physical activity. It is chronic, bilateral, constricting, and nonpulsatile, unlike the migraine, which is unilateral and throbbing.

Distinguishing chronic tension-type headache from migraine and from medication-induced headache is a diagnostic challenge, and the management is different. In general, individuals with tension-type headache seek less medical attention than migraineurs.<sup>72</sup>

### Etiologic and Risk Factors

Pain and psychologic factors clearly affect one another. Individuals with tension-type headache tend to report greater use of nonoptimal coping strategies, including withdrawal, avoidance, and self-criticism when coping with negative life events. Tension-type headache sufferers appear to have a high level of fearfulness about pain and may have an attentional bias toward bodily function versus environmental awareness. There is some evidence in females with tension-type headache of a component of catastrophizing an event that leads to a feeling of helplessness. No clear link has been established between these behavioral findings and the findings of central sensitization, the role of the limbic system, and reticular disinhibition, although there is interest in that area. Some day, the relationship, or the cause versus effect, may be better understood and the neurologic construct may be better defined.

Some biochemical changes are common to both migraine and tension-type headache. Blood levels of magnesium are low in both conditions, and there is increased blood flow in anterior, middle, and posterior cerebral arteries.

### Pathogenesis

The pain input to the brain may be increased because of activation or sensitization of peripheral sensory afferent neurons, possibly by endogenous substances such as serotonin and bradykinin. Increased hardness of muscle tissue in the upper neck is found in the absence of muscle firing.

Sensitization of second-order neurons and neurons at the level of the trigeminal nucleus alters nonpainful input so that it is perceived as noxious at the level of the cortex. Stimuli to skin, tendons, and muscle cause pain in the area around the head and, at the same time, in areas distant from the head. This may be consistent with referred hyperalgesia, relating to the convergence of multiple peripheral sensory afferents onto sensitized spinal cord neurons, which project to central structures that have another level of sensitization. Serotonin levels are likely to play an important role, as the headache pain can be modified by administration of drugs that work at the level of the serotonin receptor.<sup>3</sup>

Lack of normal inhibitory control in the central nervous system may also play a role in tension-type headache. Pain that can normally be down-regulated by interneurons may lose that function so that there is a loss of inhibitory control, or disinhibition. The neurons respon-

sible probably belong to the bulbar reticular formation. This area receives afferents from the periphery but also from limbic structures, the orbitofrontal cortex, the nucleus raphe magnus, and the periaqueductal grey matter.

Electromyographic (EMG) analysis to determine if there is increased muscle activity or increased resting tone during or between headache episodes has been inconclusive. EMG recordings do not appear to have diagnostic usefulness, and the increases that have been reported may have to do with protective adaptation rather than the cause of pain.

### Clinical Manifestations

There is increased tenderness to palpation of the tissues around the head. Both muscles and tendon insertions have been found to be excessively tender. The level of tenderness correlates to the intensity of the headache. The pain is usually in the whole head or in the neck and is reported as tightness or pressure. The pain is often described as dull and nonthrobbing. The severity of tension-type headache increases with increasing frequency. Nausea is not associated with tension headache, but there are reports of anorexia, mild photophobia, and phonophobia.<sup>56</sup>

Sleep disorders are commonly reported with tension headache, but studies do not show evidence of a relationship. Lack of sleep can precipitate the headache. Stress, fatigue and not eating on time are among the most common headache precipitants. Other triggers, consistent with migraine, are smoke, alcohol, smells, and weather changes.

### MEDICAL MANAGEMENT

**DIAGNOSIS.** The diagnosis of tension headache requires the exclusion of other causative disorders. The medical history should include the evolution of the headache. A general physical and neurologic examination should be performed to rule out a disease process, including palpation of the pericranial muscles to identify tenderness and trigger points. Referred pain patterns should also be recorded. The temporal, lateral pterygoid, masseter, sternocleidomastoid, and trapezius muscles should be looked at specifically. It should be noted whether the palpation is done during the headache or nonheadache phase, as there can be up to a 25% increase in pain perception during headache.

Except for their frequency and intensity, chronic tension-type headaches are similar to frequent episodic tension-type headaches. Chronic tension-type headaches occur most often in women over 50 years of age and have usually evolved from episodic tension headaches. Chronic tension-type headache may be linked with medication overuse (see later), and the diagnosis should be made only after there have been 15 days free of medication. It is important to recognize that individuals with confirmed migraine are prone to increased incidence of chronic tension-type headaches between migraine attacks. The individual will usually describe a difference between the headache types.

Tension-type headaches, because they occur with greater frequency and duration, and moderate intensity,