Children with Disabilities

6th EDITION

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26 Cerebral Palsy

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Upon completion of this chapter, the reader will

- Understand the definition and causes of cerebral palsy
- Understand how cerebral palsy is diagnosed
- Know the various types of cerebral palsy and their characteristics
- Know the sensory, cognitive, and medical problems commonly associated with cerebral palsy
- Understand the range of therapeutic options available to help children with cerebral palsy
- Be knowledgeable about the medical and functional prognoses for cerebral palsy

JAMAL

Jamal is a 15-month-old boy who was seen by his primary pediatrician for a routine well-child checkup. Jamal's mother expressed concern that her son was not yet walking. The pediatrician had documented some modest delays in motor development at Jamal's 12-month office visit but had attributed these delays to the fact that Jamal was born 3 months prematurely. Jamal's mother had delivered him vaginally following spontaneous onset of labor at 27 weeks' gestation. This spontaneous labor was probably caused by **chorioamnionitis**, an infection of the membranes surrounding the fetus. Jamal's birth weight was 1 pound, 10 ounces. He required ventilator support for 5 days, and at 2 weeks of age, an ultrasound study of the brain demonstrated a possible abnormality of the white matter of the brain.

As of his 12-month well-child checkup, Jamal had been sitting on his own for about 2 months. The pediatrician had explained to Jamal's mother that based on age adjusted for prematurity, Jamal began sitting at an adjusted age of 7 months. This was only slightly delayed compared with the expected age of sitting independently. Jamal's leg muscles were also a little stiff, and he sat slouched forward, but his pediatrician knew that many premature infants have mild, temporary abnormalities of muscle tone (known as transient **dystonia** of prematurity) that resolve by 15–18 months of age.

At this 15-month visit, Jamal was sitting well but still sat with a slouch. He could crawl stiffly on all fours but was not yet pulling up to a standing position. Jamal showed a pronounced tendency to keep his legs stiffly extended with his toes pointed and feet crossed at the ankles (scissoring). His pediatrician was concerned that Jamal was showing signs of cerebral palsy.

WHAT IS CEREBRAL PALSY?

Cerebral palsy is a developmental disability. As is the case with other developmental disabilities, cerebral palsy is defined on the basis of specific functional characteristics rather than on the basis of the diverse causes of the condition. The hallmark of cerebral palsy is a significant impairment of functional mobility that is associated with signs of neurological dysfunction. As the term *cerebral* implies, the locus of the dysfunction is the brain. Cerebral palsy is further distinguished from other motor impairment syndromes by the recognition that signature features of the condition are known to be associated with disturbances of the immature,

or developing, brain. These disturbances most often occur during fetal development or in the perinatal period, but they may also occur during the first few years after birth. Whether the disturbances are a consequence of brain injury or relate to a genetically based problem with brain development, in all cases the presumption is that the processes resulting in the disturbance are time limited, or nonprogressive. Based on these observations, cerebral palsy may be defined as follows: Cerebral palsy is a disorder of movement and posture that is caused by a nonprogressive abnormality of the immature brain.

Modifications of this basic definition of cerebral palsy have recently been proposed that emphasize the complexity of the condition, including associated nonmotor impairments of sensation, cognition, communication, and behavior and associated medical conditions such as seizures (Bax et al., 2005). Although the neurological basis for cerebral palsy is considered nonprogressive, the functional consequences of the disorder can, in a sense, progress. For example, some children with more severe forms of cerebral palsy (e.g., spastic quadriplegia; see the Classifying the Subtypes of Cerebral Palsy section) are prone to develop orthopedic complications such as hip dislocation, scoliosis, and muscle contractures that may reduce their functional mobility over time (Liptak & Accardo, 2004).

WHAT CAUSES CEREBRAL PALSY?

Cerebral palsy is most often a consequence of brain injury, but in some cases may be due to genetically based problems with brain development (Nelson, 2002; O'Shea, 2002). Until the 1980s, it was thought that most cases of cerebral palsy resulted from "birth asphyxia" (hypoxic-ischemic encephalopathy; see Chapter 4), which has traditionally been defined as a disruption of blood flow (ischemia) and oxygen supply (hypoxia) to the brain as a consequence of problems encountered at the time of birth It is now clear, however, that true birth asphyxia is the cause of cerebral palsy in only a minority of cases (Hankins & Speer, 2003; Nelson & Grether, 1999; Pschirrer & Yeomans, 2000).

Prematurity-Related Cerebral Palsy

Jamal provides an example of cerebral palsy that is most likely related to complications of prematurity. Premature infants, especially those born prior to 28 weeks' gestations (or with a birth weight less than 1,500 grams), are recognized to be at increased risk for cerebral palsy. The overall prevalence of cerebral palsy is approximately 2.0–2.5 per 1,000 in the general population (Hagberg et al., 2001; Reddihough & Collins, 2003; Winter et al., 2002); premature infants represent 40%–50% of this group. Among infants born prior to 28 weeks, more than 12% will ultimately be diagnosed with cerebral palsy (Marlow et al., 2005; O'Shea, 2002; Vohr et al., 2005). The increased risk of cerebral palsy in the premature infants relates to a special vulnerability of the white matter of the brain (Volpe, 2003; see Chapter 9). The white matter contains neuronal processes (the "wiring" of the brain), some of which are involved in the control of movement and regulation of muscle tone (see discussion on corticospinal pathways in the Delayed Motor Development section). Disruption of these pathways can result in the signs and symptoms of cerebral palsy. (Recent research also suggests that disruption of brain white matter is associated with subtle but important associated disturbances in the neurons, or brain cells, that compose the "gray matter" of the brain. These disturbances may explain the learning and cognitive problems encountered in children with cerebral palsy [Volpe, 2005].) The two common causes of white matter injury in premature infants are periventricular leukomalacia (PVL) and intraventricular hemorrhage (IVH) (see Chapter 9). Immaturity of brain development predisposes premature infants to both of these conditions.

Cerebral Palsy in Full-Term Infants

A wide variety of prenatal, perinatal, and genetic factors have been highlighted as possible causes of cerebral palsy in full term-infants, including birth asphyxia, congenital brain malformations, coagulation abnormalities, complications related to multiple gestation and intrauterine infection/inflammation (Nelson, 2002). Compared with those born prematurely, children born at term who subsequently develop cerebral palsy are more often small for gestational age or have malformations inside and outside the central nervous system (CNS), suggesting a problem with early fetal development in some cases (Krageloh-Mann et al., 1995). In addition, if a full-term infant experiences severe birth asphyxia, he or she may develop cerebral palsy, especially the athetoid or dystonic types (see Classifying the Subtypes of Cerebral Palsy section), which reflects damage to deep brain structures (e.g., the basal ganglia). In the past, high bilirubin levels in the immediate postnatal period resulted in a condition called kernicterus, which also caused athetoid cerebral palsy. Although kernicterus is rare now in developed countries, more subtle bilirubin-induced neurologic dysfunction (BIND) continues to be a concern (Shapiro, 2005).

Infection and Cerebral Palsy

In both term and preterm infants, direct infection of the fetus by viruses (e.g., cytomegalovirus, rubella; see Chapter 6) and other infectious agents (e.g., toxoplasmosis, a parasitic infection) has long been a recognized cause of cerebral palsy. Although these types of infections are a fairly uncommon cause of cerebral palsy, there has been increasing recognition that chorioamnionitis, or maternal intrauterine infection, may play a key role in the genesis of cerebral palsy. Chorioamnionitis predisposes to premature delivery and may also have direct adverse effects on the fetal brain (Jacobsson, 2004; Yoon, Park, & Chaiworapongsa, 2003). Complex relationships likely exist between chorioamnionitis and other clinical entities that are thought to cause cerebral palsy. For example, placental infection may contribute to the development of hypoxic-ischemic encephalopathy/birth asphyxia. Infection during pregnancy may also promote blood coagulation, leading to stroke-like events in the fetus (Leviton & Dammann, 2004; Nelson & Lynch, 2004). These relationships are currently the subject of intense investigation (Wu, 2002).

HOW IS CEREBRAL PALSY DIAGNOSED?

The diagnosis of cerebral palsy is based on the recognition of significant delays in motor development associated with signs of CNS dysfunction. Although newborns may have known risk factors for cerebral palsy (e.g., PVL), cerebral palsy cannot be diagnosed at birth. Children with more severe forms of cerebral palsy are usually diagnosed by 1 year of age; children with milder forms of cerebral palsy may not be diagnosed until 2 years of age (Aneja, 2004; Palmer, 2004; Russman & Ashwal, 2004).

Delayed Motor Development

Cerebral palsy is not the most common cause of delayed motor development. Children with motor delays in association with cognitive and learning disabilities far outnumber children with motor delays as a consequence of cerebral palsy (see Chapters 17 and 25). Cerebral palsy-related motor delay is distinguished by severity (children with cerebral palsy tend to have more severe motor delays) and by the presence of associated signs of upper motor neuron dysfunction.

Upper Motor Neuron Dysfunction in Cerebral Palsy

The upper motor neuron (UMN) system is not a discrete anatomical entity but refers collectively to the motor control systems based in the brain and spinal cord. The UMN system is distinguished from the lower motor neuron (LMN) system, which refers collectively to the peripheral nerves and the muscles that they innervate. The primary components of the UMN system are the **pyramidal tract** or **corticospinal pathways** and the **extrapyramidal system** (see Figure 26.1). These systems are differentially affected by disturbances to the developing brain depending on the timing and nature of these disturbances. Generically, UMN dysfunction is characterized by positive and negative signs. The positive signs refer to the presence of atypical neuromotor features, such as increased or decreased muscle tone, atypical reflex patterns, and involuntary movements. The negative signs refer to absent or deficient neuromotor functions, including poor motor control, poor balance, weakness, and easy fatigability. Virtually all children with cerebral palsy manifest the negative signs of UMN dysfunction to one degree or another. Significant problems with motor planning and motor control in particular are a hallmark of cerebral palsy, and tend to contribute most to deficits in functional mobility. The profile of positive signs tends to vary more from child to child and contributes to the classification of the subtypes of cerebral palsy (see Classifying the Subtypes of Cerebral Palsy).

Spasticity

Spasticity refers to a group of neuromotor signs that are seen in association with disturbances in the pyramidal component of the motor control system. The pyramidal system is composed of neurons that extend from the motor cortex to the brain stem and spinal cord. These corticospinal pathways directly control movement and also influence muscle tone and deep tendon reflexes (e.g., the familiar knee-jerk response) by inhibiting spinal cord mechanisms that control these processes. In the absence of normal corti-

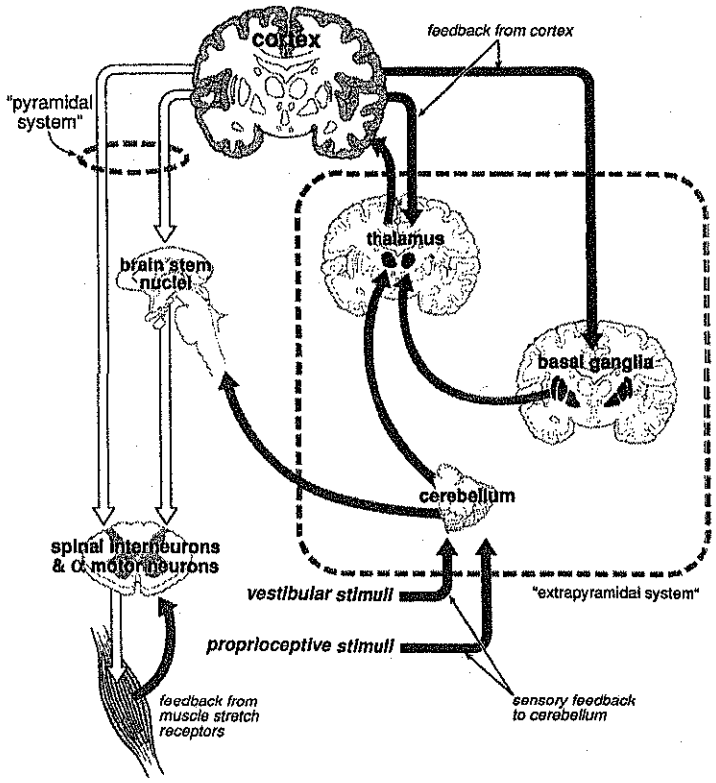

Figure 26.1. The motor control system. The upper motor neuron (UMN) system consists of the pyramidal and extrapyramidal systems. The pyramidal system connects the motor control center of the cortex to the brainstem and spinal cord and is responsible for the direct control of movement and muscle tone. The extrapyramidal system consists of deep brain structures (especially the basal ganglia and cerebellum) and works primarily by modifying and refining the output of the pyramidal system. The lower motor neuron (LMN) system consists of the muscles and the nerves that connect the muscles and the spinal cord, including the nerves that comprise the stretch reflex mechanism. (From Pellegrino, L., & Dormans, J.P. [1998]. Definitions, etiology, and epidemiology of cerebral palsy. In J.P. Dormans & L. Pellegrino [Eds.], *Caring for children with cerebral palsy: A team approach* [p. 10]. Baltimore: Paul H. Brookes Publishing Co.; reprinted by permission.)

cospinal inhibition, the spinal cord influences predominate, resulting in **spastic** hypertonicity (increased muscle tone) and exaggerated reflex responses, two of the hallmark features of spastic cerebral palsy. The clinical signs associated with spasticity are examples of release of inhibition phenomena, which characterize many forms of UMN disturbance.

Persistent Primitive Reflexes

Another example of release of inhibition phenomena relates to the abnormal persistence of primitive reflex patterns in some children with cerebral palsy. **Primitive reflexes** are called primitive because they are present in early life (in some cases, during intrauterine development) and because they are thought to be controlled by the primitive regions of the nervous system: the spinal cord, the labyrinths of the inner ear, lower brain areas, and the brain stem. Familiar examples of primitive reflexes include the suckling reflex and the hand-grasp reflex in the newborn. As the cortex matures, these reflexes are gradually suppressed and integrated into voluntary movement patterns. The process of integration is usually complete by 12 months of age. Because of a disturbance in normal maturation of corticospinal pathways in children with cerebral palsy, there is a tendency for these

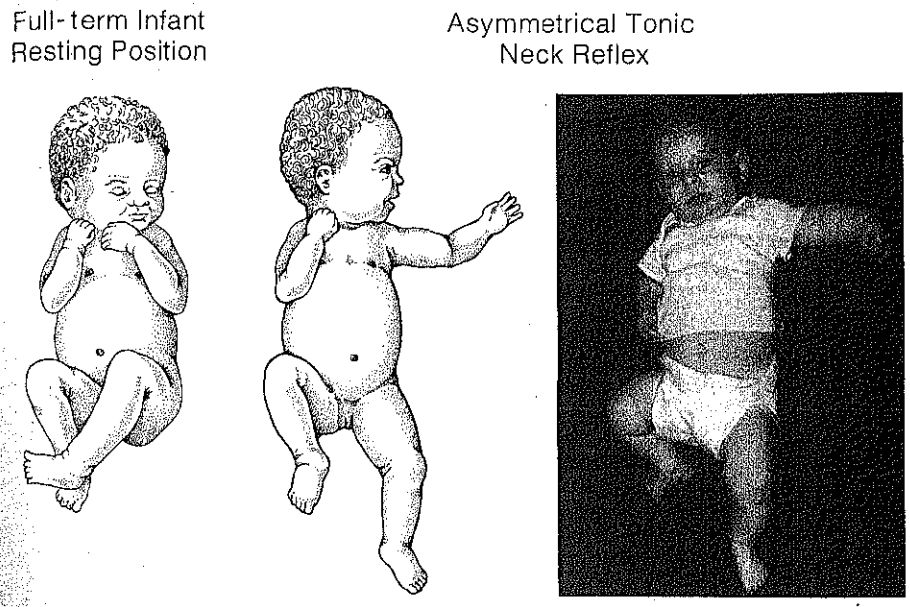

Figure 26.2. The asymmetric tone neck reflex. In the typical newborn infant, when the head is actively or passively turned to the side, the arm and leg on the same side will extend and the arm and leg on the opposite side will flex, resulting in a "fencing" posture. The opposite pattern occurs when the head is turn to the other side. In typically developing infants, the reflex fades (is integrated) by about 6 months of age and is never obligatory (the infant can break through the pattern with spontaneous movement, even in the newborn period). In children with cerebral palsy, the reflex tends to be more pronounced, persists beyond the expected age, and may be obligatory.

primitive reflex patterns to persist beyond early infancy. Among the primitive reflexes, the **asymmetric tone neck reflex** (Figure 26.2) and the **tonic labyrinthine response** (Figure 26.3) are particularly helpful in the diagnosis of cerebral palsy.

Involuntary Movements and Ataxia

Children who have cerebral palsy as a consequence of disturbances in the extrapyramidal system will sometimes exhibit atypical, involuntary movements, also know as **dyskinesias.** Rapid, random, jerky movements are known as **chorea**; slow, writhing movements are called **athetosis** (when seen together, these movements are called **choreoathetosis**). **Dystonia** refers to **rigid** posturing centered in the trunk and neck. **Ataxia** seen in association with cerebral palsy is characterized by abnormalities of voluntary movement involving balance and position of the trunk and limbs in space. For chil-

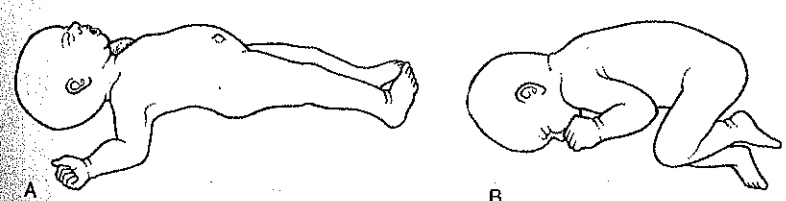

Figure 26.3. The tonic labyrinthine reflex. A) When the child is in the supine position with the head slightly extended, retraction of the shoulders and extension of the legs is observed. B) The opposite occurs when the infant is in the prone position with the head slightly flexed. In typically developing infants, the reflex pattern is barely evident in the newborn period; in children with cerebral palsy, the pattern may dominate posture and movement and may persist throughout life.

dren who can walk, this is noted most especially as a wide-based, unsteady gait. Difficulties with controlling the hand and arm during reaching (causing overshooting or past-pointing) and problems with the timing of motor movements are also seen.

Automatic Movement Reactions and the Development of Voluntary Control

As primitive reflexes diminish in intensity in the typically developing child, **postural reactions,** also known as **automatic movement reactions,** are developing (Figure 26.4). Some of the more important of these reactions include righting, equilibrium, and protective reactions, all of which enable the child to have more complex voluntary movement and better control of posture. These automatic movement responses serve a crucial supporting role in the development of specific motor milestones; delayed or absent development of these responses in children with cerebral palsy makes a significant contribution to their motor disability.

Walking and the Diagnosis of Cerebral Palsy

Many children with cerebral palsy first come to professional attention because of delayed walking. This developmental milestone has a powerful intrinsic meaning for parents and professionals alike. Most adults know that children begin walking at about 1 year of age, and there is an implicit understanding that a child's first steps mark the transition from infancy to toddlerhood. When a child does not make this transition at the expected time, it is more difficult to ignore than other delays in development. To walk, a child must be able to maintain an upright posture, move forward in a smoothly coordinated manner, and demonstrate protective responses for safety when falling. Even a child with the mildest form of cerebral palsy has difficulty attaining the continuous changes in muscle tone that are required for typical walking. The child's walk or gait is affected in many ways. Scissoring, the most common gait disturbance, occurs because of increased tone in the muscles that control adduction (movement toward the mid-line) and internal rotation of the hips. Toe walking results from an **equinus** position of the feet (Figure 26.5) and increased extensor tone in the legs. In children without cerebral palsy, a protective reaction called the parachute response develops by 10 months of age. This is manifest by forward extension of the arms when falling forward. Many children with cerebral palsy have delayed or absent development of this response, which makes walking inherently unsafe.

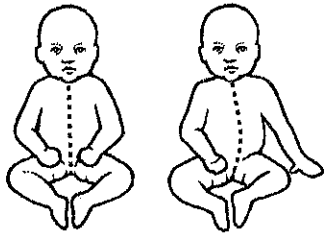

Figure 26.4. Automatic movement responses: the lateral prop reaction. At about 6 months of age, typically developing infants have already developed good postural control of the head and trunk (righting or equilibrium responses) and can stop themselves from falling forward when placed in sitting by extending their arms in front of them (forward prop response). By 6 months of age, most infants can also catch themselves when falling to the side by extending the arm on the same side (lateral prop response). This automatic movement reaction is critical for independent sitting and may be delayed or absent in children with cerebral palsy. (From Pellegrino, L., & Dormans, J.P. [1998]. Making the diagnosis of cerebral palsy. In J.P. Dormans & L. Pellegrino [Eds.], *Caring for children with cerebral palsy: A team approach* [p. 39]. Baltimore: Paul H. Brookes Publishing Co.; reprinted by permission.)

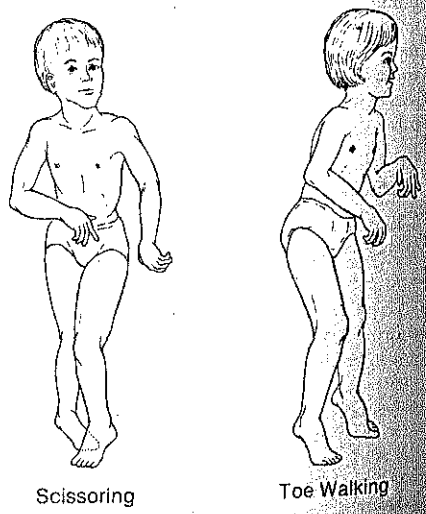

Figure 26.5. Scissoring results from increased tone in the muscles on the inner aspect of the thigh that tend to pull the legs together and turn the legs inward. Toe walking is due to tightness of the calf muscles and Achilles tendon and increased extensor tone in the legs.

Classifying the Subtypes of Cerebral Palsy

Cerebral palsy is often classified according to the type of motor impairment that predominates (Figure 26.6; Koman, Smith, & Shilt, 2004), with spastic cerebral palsy being the most common type. Spastic cerebral palsy is further categorized according to the distribution of limbs involved. In **spastic hemiplegia,** one side of the body is more affected than the other; usually, the arm is more affected than the leg. Because the motor neurons that control one side of the body are located in the opposite cerebral cortex, a right-side hemiplegia implies damage to or dysfunction of the left side of the brain, and vice versa. In **spastic diplegia,** the legs are more affected than the arms. This is the type of cerebral palsy most frequently associated with prematurity (and is the type that Jamal exhibits in this chapter's case study). In **spastic quadriplegia,** all four limbs and usually the trunk and muscles that control the mouth, tongue, and pharynx are affected. The severity of the motor impairment in spastic quadriplegia implies wider cerebral dysfunction and worse outcome than for the other forms of spastic cerebral palsy. Individuals with spastic quadriplegia often have intellectual disability, seizures, sensory impairments, and medical complications.

Dyskinetic cerebral palsy (also known as extrapyramidal cerebral palsy) is characterized by abnormalities in muscle tone that involve the whole body. Changing patterns of tone from hour to hour and day to day are common. These children may exhibit rigid muscle tone while awake and normal or decreased tone while asleep. Involuntary movements are often present, although they are sometimes difficult to detect, and are the hallmark of this type of cerebral palsy. The term **athetoid cerebral palsy** characterizes a form of dyskinetic cerebral palsy associated with choreoathetosis. Similarly, **dystonic cerebral palsy** is associated with prominent involuntary posturing and dystonia.

Ataxic cerebral palsy is characterized by abnormalities of voluntary movement involving balance and position of the trunk and limbs in space (ataxia). Ataxic cerebral palsy may be associated with increased or decreased muscle tone.

The term **mixed cerebral palsy** is used when more than one type of motor pattern is present and should be used only when one pattern does not clearly predominate over another. The term *total body cerebral palsy* is sometimes used to emphasize that certain types of cerebral palsy (dyskinetic, ataxic, mixed, and spastic quadriplegia) involve the entire musculoskeletal system to a greater or lesser degree; other forms

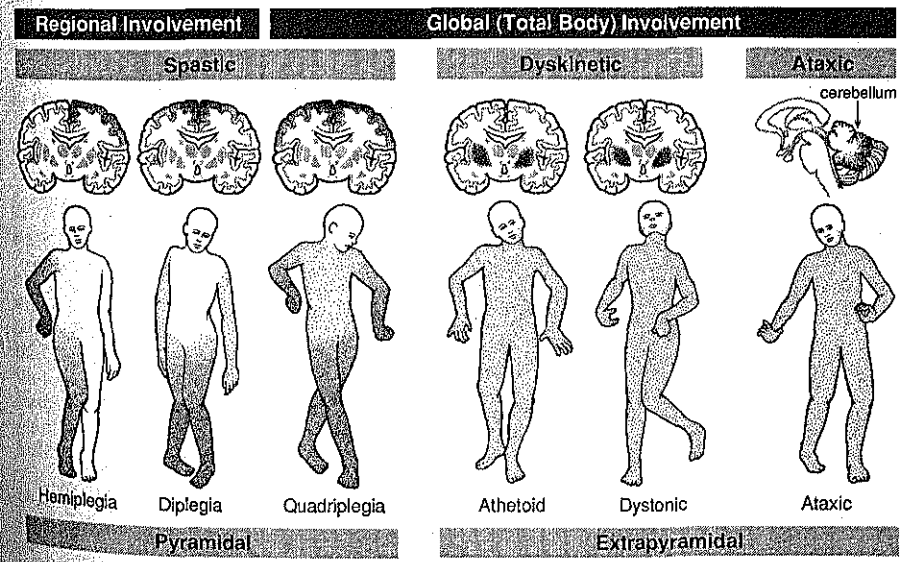

Figure 26.6. Different regions of the brain are affected in various forms of cerebral palsy. In this figure, the darker the shading, the more severe the involvement.

of spastic cerebral palsy (diplegia, hemiplegia) are localized to particular regions of the body.

Characterizing the Degree of Functional Disability Associated with Cerebral Palsy

Although there is some clinical utility in classifying cerebral palsy on the basis of neuromotor characteristics (e.g., spasticity, dyskinesias), there tends to be a great deal of functional variability within specific subtypes. For example, some children with spastic diplegia may be able to walk independently, whereas others depend on a wheelchair for mobility. A number of functional assessment systems have been developed to address this issue (Oeffinger et al., 2004; Palisano et al., 1997; see Table 26.1). These systems provide a means to classify individuals based on meaningful functional distinctions. Using this type of functional classification scheme is often helpful for planning therapeutic interventions and establishing goals for habilitation (see the Habilitation section) and is more predictive of long-term functional outcome in comparison with the prognostic value of the traditional, impairment-focused classification scheme (Rosenbaum et al., 2002).

Table 26.1. Summary of Gross Motor Function Classification System (GMFCS)

	Age			
Level	< 2 years	2–4 years	4–6 years	6–12 years
I: Walks without restrictions	Sits well (hands free to play), crawls and pulls-to-stand; walks between 18 and 24 months without a device	Gets up and down from floor to standing without help; walking is preferred method of mobility	Walks indoors and outdoors; climbs stairs; starting to run/jump	Independent walking, running, jumping, but speed, balance, and coordination reduced
II: Walks without device; restricted community mobility	Sits but may need hands for balance; may creep or crawl; may pull-to-stand or cruise	Floor sits, but hard to keep both hands free; mobility by crawling, cruising, or walking with assistive device	Transfers with arm assist; walks without device at home, short distances outside; climbs stairs with railing; no running or jumping	Independent walking but limitations in challenging circumstances; minimal running, jumping
III: Walks with assistive device; limited community mobility	Sits with low back support; rolls and creeps	Floor sits, often W-sitting, needs help getting to sit; creeping and crawling primary means of mobility; limited assisted standing/walking	Sits in regular chair with pelvic support to allow free hands; walks with device on level surface; transported for long distances	Walks indoors or outdoors with assistive device; wheelchair mobility or transport for long distances
IV: Limited self-mobility; power mobility	Has head control, but needs trunk support for sitting; rolls to back, may roll to front	Needs hands to maintain sitting; adaptive equipment for sitting/standing; floor mobility only (rolling, creeping, or crawling without reciprocal leg movements)	Adaptive seating needed for maximum hand function; needs assistance for transfers; walks short distances with assistance; power mobility for long distances	Maintains function achieved by ages 4–6 or relies more on power wheelchair for self-mobility
V: Self-mobility severely limited even with assistive devices	Limited voluntary control of movement; head and trunk control minimal; needs help to roll	Limited control of movement and posture; all areas of motor function are limited; adaptive equipment does not fully compensate functional limitations for sitting and standing; no independent mobility (requires transport); some children achieve very limited power mobility with extensive adaptations		

Source: Palisano, Rosenbaum, Walters, et al. (1997).

Establishing the Etiology of Cerebral Palsy

Diagnosis of the etiology, or cause, of cerebral palsy occurs in parallel with the process that establishes the disability or functional diagnosis of cerebral palsy itself (Ashwal et al., 2004). Establishing the etiology of cerebral palsy may have important implications for treatment, prognosis, and recurrence risk. Information gleaned from the medical history and physical examination is often critical in establishing etiology (e.g., knowing that a child was born prematurely and has signs of spastic diplegia strongly suggests the possibility of cerebral white matter injury). With regard to specialized diagnostic testing, brain imaging is especially helpful (Accardo, Kammann, & Hoon, 2004; Ancel et al., 2006). Ultrasonography is used for fetal and neonatal screening and can distinguish large malformations of the brain and abnormalities related to brain hemorrhage or injury (i.e., IVH, PVL). Computed tomography (CT) and especially magnetic resonance imaging (MRI) provide more detailed resolution of anatomical structures than ultrasound and may help to define the cause of cerebral palsy. Newer techniques—such as positron emission tomography (PET), functional magnetic resonance imaging (fMRI), **single photon emission computed tomography (SPECT),** and diffusion tensor imaging (DTI)—complement CT and MRI (primarily in the research setting) by providing information about brain metabolic function, which in some cases is abnormal even when brain structure appears to be normal (Davidson, Thomas, & Casey, 2003; Mohan, Chugani, & Chugani, 1999; Watts et al., 2003).

WHAT OTHER IMPAIRMENTS ARE ASSOCIATED WITH CEREBRAL PALSY?

All children with cerebral palsy have problems with movement and posture. Many also have other impairments associated with damage to the CNS. The most common associated disabilities are intellectual disability, visual impairments, hearing impairments, speech-language disorders, seizures, feeding and growth abnormalities, and behavior/emotional disorders.

Assessment of intellectual functioning in children with cerebral palsy may be difficult because most tests of cognition require motor or verbal responses. Even taking this into account, approximately one half of children with cerebral palsy have intellectual disability, and many of those with average intelligence exhibit some degree of learning disability (Nordmark, Hagglund, & Lagergren, 2001). There is a general correlation between physical and cognitive disability: Children with more severe forms of cerebral palsy are a greater risk for more significant intellectual disability.

Visual impairments are also common and diverse in children with cerebral palsy (Guzzetta, Mercuri, & Cioni, 2001). The premature infant may have severe visual impairment caused by retinopathy of prematurity (see Chapter 11). Nystagmus, or involuntary oscillating eye movements, may be present in the child with ataxia. Children with hemiplegia may present with homonomous hemianopsia, a condition causing loss of one part of the visual field. Strabismus, or squint, is seen in many children with cerebral palsy. Children with cerebral palsy are also more prone to hyperopia (farsightedness) than children without cerebral palsy are (Sobrado et al., 1999).

Hearing, speech, and language impairments are also common, occurring in about 30% of children with cerebral palsy. Children with congenital rubella or other intrauterine viral infections often have high-frequency hearing loss (see Chapter 12). Dyskinetic cerebral palsy is associated with articulation problems, as choreoathetosis affects tongue and vocal cord movements. Expressive or receptive language disorders are commonly observed among children with cerebral palsy who do not have intellectual disability and may be a harbinger of a learning disability, such as specific reading disability (see Chapter 25).

Approximately 40% of children with cerebral palsy also develop seizures (Nordmark et al., 2001). Children with more severe cognitive and physical disability are more prone to seizures, as are children whose cerebral palsy is a consequence of brain malformation, infection, or gray matter injury. Partial epilepsy (see Chapter 29) is the most common form of seizure activity in all children with cerebral palsy, and is especially common in children with hemiplegia who have seizures (Carlsson, Hagberg, & Olsson, 2003).

Feeding and growth difficulties are often present in children with cerebral palsy (Samson-Fang et al., 2002) and may be secondary to a variety of problems, including hypotonia, weak suck, poor coordination of the swallowing mechanism, tonic bite reflex, hyperactive gag reflex, and exaggerated tongue thrust. These problems may lead to poor nutrition and may require the

use of alternative feeding methods, such as tube feeding (see Chapter 31). Medical problems related to poor gastrointestinal motility (including gastroesophageal reflux and constipation) tend to add to these difficulties. Poor nutrition and lack of weight-bearing activities also leads to **osteopenia**, or weak bones related to reduced bone mineral density, making children with cerebral palsy more prone to fractures. Bisphosphonates (drugs used to treat osteoporosis in older adults) have been shown to be effective for osteopenia is cerebral palsy (Henderson et al., 2002; Henderson et al., 2005).

Poor health in children with cerebral palsy significantly contributes to the societal and functional disadvantages they experience as a consequence of their disability (Liptak & Accardo, 2004; Samson-Fang et al., 2002). A comprehensive health plan implemented in the context of a well-defined "medical home" is a critical component to assuring that the health needs of the children are adequately addressed (Cooley & American Academy of Pediatrics Committee on Children with Disabilities, 2004).

WHAT CAN BE DONE TO HELP CHILDREN WITH CEREBRAL PALSY?

Habilitation

Cerebral palsy is a lifelong disability that has different functional implications at different stages of the life cycle. For families and professionals involved in the care of children with cerebral palsy, the ultimate goal of any treatment or intervention is to maximize functioning while minimizing any disability-related disadvantages. This is accomplished by recognizing the specific abilities and needs of the individual child as they occur within the context of his or her family and community. Habilitation is an intervention strategy that is family focused and community based. Ideally, it is conceived and implemented as a comprehensive program designed to facilitate adaptation to and participation in an increasing number and variety of societal environments, including home, school, clinic, child care, neighborhood, and day treatment programs (Pellegrino, 1995). The ultimate goal of intervention is to enhance participation in these environments and to afford access to new environments in a manner that is mutually satisfying for the individual and the community.

Early Intervention, School, and Therapy

For most children with cerebral palsy, the process of habilitation begins at home under the auspices of state-administered early intervention programs. These programs emphasize involvement of parents so that they can learn effective methods of working with their child (Guralnick, 1998). Programs are individualized according to the specific needs of the child and the family and while emphasizing home-based services may also provide consultative and center-based interventions (see Chapter 33).

For many children with cerebral palsy, entry into preschool represents the first major step into the wider community. Difficulties are often encountered in accommodating the physical, nutritional, and medical needs of these children at this stage. For school-age children, concerns regarding motor function and medical needs continue, but increased attention is focused on concerns related to learning disabilities, attention and behavior problems, intellectual disability, and sensory impairments. For many children, these comorbid conditions, rather than their motor disability, put them at greatest disadvantage relative to their peers.

Children with cerebral palsy have traditionally been segregated into classrooms with designations such as "multiply disabled" and "orthopedically impaired," sometimes without proper regard for their cognitive skills. The trend now, however, is toward inclusion in general education classrooms, which under ideal circumstances can accommodate the needs of children with a range of abilities. Inclusive environments require significant collaboration between the general and special education models and work best when a team of educators and paraprofessionals is associated with each classroom (see Chapter 34).

For children with cerebral palsy, therapy may come in many different forms. Most children receive traditional forms of speech, occupational, and physical therapy. The most common method of motor therapy for the young child with cerebral palsy is **neurodevelopmental therapy (NDT)**, an approach employed by both occupational and physical therapists that is designed to provide the child with sensorimotor experiences that enhance the development of more typical movement patterns (see Chapter 37; Campbell, 2000). An individualized program of positioning, therapeutic handling, and play is developed for the child. Program goals include the normalization of tone and the improved control of movement during functional activities. Less traditional therapies, including **hippotherapy** (therapeutic horseback riding) and aquatic therapy (a variation of physical therapy performed in the water) are commonly employed therapeutic options that have mainly anecdotal support for efficacy (Meregillano, 2004). Most therapists utilize an eclectic mix of techniques in the pursuit of improved function and mobility based on the specific set of goals established for a specific child.

Recently, a promising technique known as constraint-induced therapy, or forced-use therapy, has been introduced to help children with hemiplegia cerebral palsy (Taub et al., 2004; Willis et al., 2002). The technique involves "constraining" the more functional arm or hand to force use of the less functional upper extremity. Randomized control trial suggests that the technique may be of significant benefit over traditional therapy alone (Taub et al., 2004).

Physical exercise is important to strengthen muscles and bones, enhance motor skills, and prevent contractures. In addition, the social and recreational aspects of organized physical activities can be highly beneficial (see Chapter 38). Many popular activities, including swimming, dancing, and horseback riding, can be modified so that children with cerebral palsy can participate. In addition, the Special Olympics has enabled thousands of children and young adults with intellectual disabilities to take part in various sporting events. The rewards of engaging in competitive sports are invaluable for enhancing self-esteem and providing a sense of belonging to a peer group. Parents and professionals should encourage all children to participate in whatever physical activities their interests, motivation, and capabilities allow (see Chapter 38).

Bracing, Splinting, and Positioning

Therapists make frequent use of braces and splints (referred to collectively as **orthotic** devices) and positioning devices as aids in the pursuit of functional goals for children with cerebral palsy. These devices are used to maintain adequate range of motion, prevent contractures at specific joints, provide stability, and control involuntary movements that interfere with function. One of the most commonly prescribed orthotics is a short leg brace, known as an ankle-foot **orthosis** (AFO). The AFO stabilizes the position of the foot and provides a consistent stretch to the Achilles tendon (see Chapter 37). A variety of splints may be used to improve hand function. For example, the resting hand splint is commonly used to hold the thumb in an abducted (away from the mid-line) position and the wrist in a neutral or slightly extended position. This helps the child keep his or her hand open, and tends to prevent the development of hand deformities (Figure 37.1). Another type of brace called a body splint is made of a flexible, porous material and controls abnormal tone and involuntary movements by stabilizing the trunk and limbs. Most pediatric braces and splints are custom-made from plastics that are molded directly on the child, so they must be monitored closely and modified as the child grows or changes abilities.

Positioning devices are used to promote skeletal alignment, to compensate for abnormal postures, or to prepare the child for independent mobility (Figure 26.7). Proper positioning geared to the age and functional status of the child is often a key intervention in addressing the tone and movement abnormalities associated with cerebral palsy. For children who must sit for extended periods or who use a wheelchair for mobility, a carefully designed seating system becomes an all-important component of their habilitation. Careful attention to functional seating may also have long-term benefits in the prevention of contractures and joint deformities related to spasticity (Myhr et al., 1995).

Adaptive Equipment

A wide variety of devices is available to aid mobility. For children who are ambulatory, crutches, walkers, and canes may help in the attainment of walking or in improvement of the quality and range of ambulation. The forearm, or Lofstrand, crutch is used in preference to the familiar under-the-arm crutch. A posterior walker (the child is positioned in front of the walker, rather than behind) with wheels is used in preference to a standard forward-position walker without wheels. Canes are used less commonly.

For children with limited walking skills, wheelchairs are essential for maximizing mobility and function. A wheelchair with a solid seat and back is usually recommended. Some children, however, have difficulty using this type of chair unless modifications are made. The addition of head and trunk supports or a tray may be needed for the child who lacks postural control

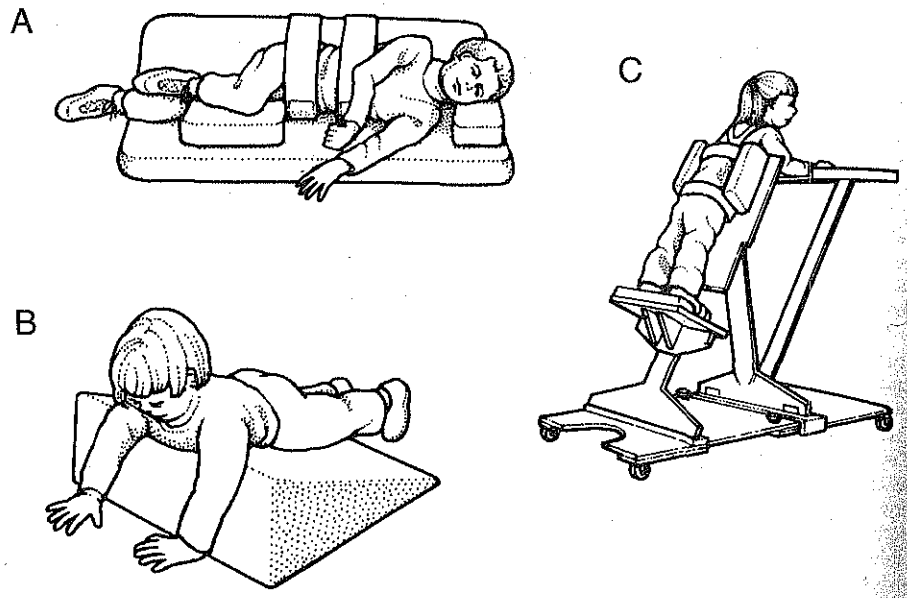

Figure 26.7. a) Child in sidelyer, b) child on prone wedge, and c) child in prone standing device.

due to low tone. The child with limited head control or with feeding difficulties may benefit from a high-backed chair that can be tilted back 10–15 degrees (Figure 26.8a). This helps to maintain the child's body and head in proper alignment.

Special seating cushions or custom-molded inserts that conform to the contours of the body can offer necessary support for the child with orthopedic deformities such as scoliosis. Motorized (power) wheelchairs can enhance the independence of children who are able to use them. Although these usually have an easily manipulated joystick for controlling both speed and direction (Figure 26.8b), other types of switches are available for children who cannot control their hand movements.

Special supportive strollers are an alternative to wheelchairs for mobility within the community or for the young child whose potential for ambulation has yet to be determined. These are lightweight and collapsible yet support the back and keep the hips properly aligned (Figure 26.8c).

Car seats are essential to the safety of all children who ride in automobiles. Several manufacturers offer adapted car seats that meet federal safety guidelines as well as provide proper support for the child with cerebral palsy. Often these models include a base that allows the seat to be used as a stroller or a positioning chair outside of the car. Car beds and special straps are also available for children who have more severe disabilities or who require these special adaptations temporarily (e.g., following surgery).

Assistive Technology

Assistive technology devices are often an important part of the habilitation plan for children with cerebral palsy (see Chapter 36). The technology involved may be as simple as Velcro or as complex as a computer chip. Although it is often true that the simplest intervention is the best, it cannot be denied that the computer has become the hero of assistive technology. Computers can be used to control the environment, provide a lifeline with the outside world, enable a person to work at home, facilitate artificial speech and sight, and provide entertainment. The real potential of this technology to improve the quality of life for children with disabilities is just beginning to be realized. Enthusiasm for its use, however, is tempered by

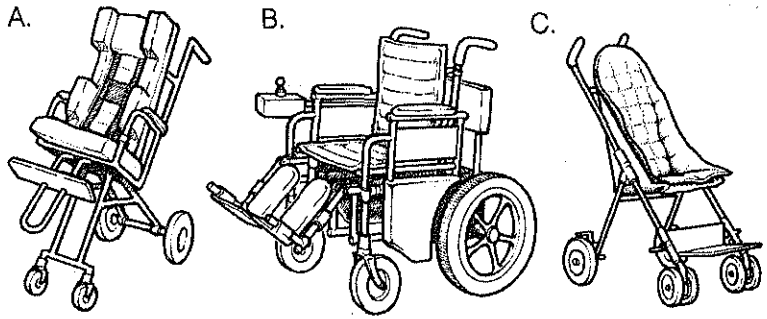

Figure 26.8. Wheelchairs. a) High-backed, tilting chair with lateral inserts and head supports. b) Motorized wheelchair with joystick control. c) Supportive collapsible stroller.

problems with access related to the cost and availability of durable hardware and well-designed software (see Chapter 36).

Managing Spasticity and Dystonia

The motor impairment associated with cerebral palsy is multifaceted, and the specific profile of skills and deficits varies greatly from child to child. The core impairment in all children with cerebral palsy, however, is a significant deficit in motor planning and motor control. Although this deficit is the primary target of therapy, it tends to be refractory to medical interventions. By contrast, specific manifestations of the UMN syndrome associated with cerebral palsy, namely spasticity and dystonia, are amenable to a variety of physical, pharmacologic, and surgical techniques. Spasticity and dystonia therefore represent tempting targets for intervention. A potential pitfall in targeting these symptoms is lack of sufficient care in establishing the connection between the impairments (spasticity or dystonia) and the disability (functional deficits). For some children, it is possible to significantly reduce spasticity or dystonia without improving (and in some cases even worsening) functional outcome. Consultation with experienced professionals who are intimately familiar with a child's particular pattern of skills and impairments is critical to the proper selection specific interventions.

The primary goals in treating spasticity and dystonia are to improve function, to prevent or postpone the musculoskeletal complications attendant to these conditions, and to ease the care of the child with significant muscle tightness. Different treatment options operate at different levels of the neuromotor apparatus (Figure 26.9); treatments may be used singly, sequentially, or simultaneously depending on the specific clinical circumstance. Because dystonia is caused by disturbances in the extrapyramidal component of the motor control system, pharmacological interventions for this condition must operate at the level of the CNS (brain and spinal cord). By contrast, the mechanisms that generate muscle spasticity operate from the level of the CNS down to the level of the muscle itself; a wider variety of therapeutic techniques are therefore available for targeting and modulating the effects of spasticity.

Casting Tone-reducing, or inhibitive, casts are used in some centers as an adjunct to more traditional methods of managing spasticity (Law et al., 1991). The casts are made for arms or legs and can be designed either for immobilization or to be used during weight-bearing activities. Benefits of inhibitive casting include improved gait and weight bearing, increased range of motion, and improved functional hand use. Casts position the limbs so that spastic muscles are in lengthened positions, being gently stretched. Serial application of casts (serial casting) can allow the therapist to increase range of motion gradually when contractures are present. After maximal range and position have been achieved, a cast is worn intermittently to maintain the improvement. Casting is now most often used in conjunction with other therapeutic modalities, especially following use of botulinum toxin (Glanzman et al., 2004; Kay et al., 2004; Wasiak, Hoare, & Wallen, 2004).

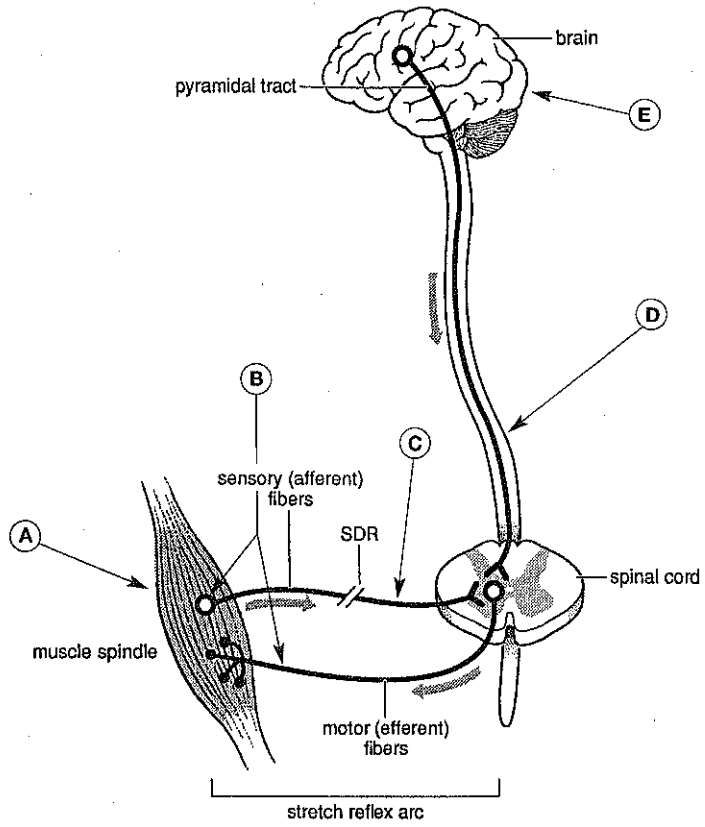

Figure 26.9. Levels of intervention for spasticity and dystonia. a) Inhibitive casting, physical therapy, exercise, and medications such as dantrolene directly affect tone at the muscle level. b) Nerve blocks, motor point blocks, and botulinum toxin work at the level of muscle and nerve entry into muscle. c) Selective dorsal rhizotomy reduces spasticity by interrupting the sensory component of the stretch reflex arc. d) Medications such as baclofen reduce spasticity at the level of the spinal cord. e) Medications for spasticity such as diazepam and medications for dystonia work at the level of the brain. (From Pellegrino, L., & Dormans, J.P. [1998]. Definitions, etiology, and epidemiology of cerebral palsy. In J.P. Dormans & L. Pellegrino [Eds.], *Caring for children with cerebral palsy: A team approach* [p. 46]. Baltimore: Paul H. Brookes Publishing Co.; adapted by permission.)

Nerve Blocks, Motor Point Blocks, and Botulinum Toxin Several injectable agents are available that can be used to target spasticity in particular muscle groups. Local anesthetic agents injected into the nerves that supply spastic muscles produce a temporary, reversible conduction block and are used for diagnostic purposes. Longer-lasting effects are achieved by injecting chemical agents, such as diluted alcohol or phenol, which denature muscle and nerve protein at the point of injection. Direct injections of denaturing agents into motor nerves, called **nerve blocks**, are sometimes used but carry the risk of sensory loss due to damaged sensory nerve fibers that are bundled together with motor fibers. A **motor point block** effectively interrupts the nerve supply at the entry site to a spastic muscle without compromising sensation. The main side effect of the procedure is localized pain that may persist for a few days after the injection. Inhibition of spasticity lasts for 4–6 months, and the procedure can be repeated after the initial effect has worn off. This temporary reduction of spasticity allows for more effective application of physical therapy to improve range of motion and function and may make it possible to postpone orthopedic surgery.

Injectable botulinum toxin (Botox) has been introduced as an alternative to motor point blocks and has largely supplanted alcohol and phenol in many clinical applications (Jefferson, 2004; Mooney, Koman, & Smith, 2003; Morton, Hankinson, & Nicholson, 2004; Pidcock, 2004). Botulinum toxin is produced by the bacterium that causes **botulism** and is among the most potent neurotoxins known. It works by blocking the nerve–muscle junction. When the toxin is absorbed into the general circulation (as with botulism), death may result from paralysis of respiratory muscles. Small quantities, however, can be safely injected directly into spastic muscles without significant spread of the toxin into the bloodstream. This results in weakening of the muscle and reduction of spasticity for 3–6 months (the antispastic effects of the injections dissipate over time). Although botulinum toxin is used mainly to treat spasticity in muscles of the limbs and trunk, novel uses, included injection of the salivary glands to reduce drooling (a common problem in many children with cerebral palsy) are now being reported (Jongerius et al., 2004). A large number of studies have demonstrated the efficacy and safety of botulinum toxin as a therapeutic modality in cerebral palsy (Jefferson, 2004; Mooney et al., 2003). Although clarification is still needed regarding the definition of clinical indications and outcomes, the use of injectable botulinum toxin has become a mainstay in the management of spasticity in cerebral palsy and has also found applications for specific types of dystonia (Gordon, 1999).

Oral and Intrathecal Medications A variety of orally administered medications have been used to improve muscle tone in children with spasticity and rigidity (Krach, 2001). Although no drug has proved helpful for treating choreoathetosis, several drugs used in **Parkinson's disease**, including carbidopa-levodopa (Sinemet) and trihexyphenidyl (Artane), have been helpful for some children with dystonic cerebral palsy. The medications most commonly used to control spasticity and rigidity are diazepam (Valium), baclofen, and dantrolene (Dantrium). Diazepam and its derivative compounds, lorazepam (Ativan) and clonazepam (Klonopin), affect brain control of muscle tone, beginning within half an hour after ingestion and lasting about 4 hours. Withdrawal of these drugs should be gradual, as physical dependency can develop. Side effects include drowsiness and excessive drooling, which may interfere with feeding and speech.

Baclofen has been most commonly used to treat adults with multiple sclerosis and traumatic damage to the spinal cord. Drowsiness, nausea, headache, and low blood pressure are the most common side effects of the oral form of the medication in children with cerebral palsy. About 10% of children treated with baclofen experience side effects unpleasant enough to necessitate discontinuation of the medication. Care must be taken when stopping the medication to gradually taper it, as rapid withdrawal can lead to severe side effects, including hallucinations.

Dantrolene works on muscle cells directly, as a calcium channel blocker, to inhibit their contraction. It is usually given two to three times daily. Side effects include drowsiness, muscle weakness, and increased drooling. A rare side effect of this drug is severe liver damage, so liver function tests should be performed periodically.

Although a variety of additional medications are becoming available for the treatment of spasticity in children with cerebral palsy, most cause problematic side effects similar to those described for diazepam, baclofen, and dantrolene, and none are clearly superior to these medications (Krach, 2001).

Intrathecal baclofen therapy is a newer therapeutic modality that allows for the direct delivery of antispasticity medication (baclofen) into the spinal fluid (intrathecal) space, where it can inhibit motor nerve conduction at the level of the spinal cord (Disabato & Ritchie, 2003; Fitzgerald, Tsegaye, & Vloeberghs, 2004; Tilton, 2004). A disk-shaped pump is placed beneath the skin of the abdomen, and a catheter is tunneled below the skin around to the back, where it is inserted through the lumbar spine into the intrathecal space. The intrathecal medication most often used is baclofen, which is stored in a reservoir in the disk that can be refilled with a needle inserted into the reservoir through the skin. The medication is delivered at a continuous rate that is computer controlled and adjustable. Because the drug is delivered directly to its site of action (the cerebrospinal fluid), much lower dosage may be used to achieve benefit, with a reduced risk of side effects. Improvements in lower extremity, upper extremity, and even oral-motor function have been observed. The main benefit of the method is dramatic reduction in spasticity and adjustable dosing (Fitzgerald et al., 2004). The main dis-

advantages are fairly common although usually manageable side effects, including hypotonia (low muscle tone), increased seizures in individuals with known epilepsy, sleepiness, and nausea/vomiting (Gilmartin et al., 2000). Complications related to mechanical failures and infection and the need for intensive and reliable medical follow-up are also significant concerns (Murphy, Irwin, & Hoff, 2002).

Selective Dorsal Rhizotomy Selective posterior rhizotomy is a neurosurgical procedure that reduces spasticity by interrupting the sensory, or afferent, component of the deep tendon (or stretch) reflex. This reflex mechanism is exaggerated in children with spastic forms of cerebral palsy. The surgery reduces spasticity permanently in the legs but not in the arms, so its use is confined mainly to children with spastic diplegia. Uncertainties exist regarding long-term functional outcomes in children who undergo this procedure (Koman et al., 2004).

Another neurosurgical procedure currently under investigation, deep brain stimulation (DBS), has been proposed as a method to reduce choreoathetosis and dystonia associated with some forms of extrapyramidal cerebral palsy. Initial results are promising, but DBS is still in its infancy, and information is lacking regarding its use in children (Krauss et al., 2003).

Managing the Musculoskeletal Complications of Cerebral Palsy

Because of the abnormal or asymmetrical distribution of muscle tone, children who have cerebral palsy are susceptible to the development of joint deformities. The most common of these result from permanent shortening or contracture of one or more groups of muscles around a joint, which limits joint mobility. Orthopedic surgery is done to increase the range of motion by lengthening a tendon, by cutting through muscle or tendon (release), or by moving the point of attachment of a tendon on bone. For example, a partial release or transfer of the hip adductor muscles may improve the child's ability to sit and walk and may lessen the chances of a hip dislocation (Hagglund et al., 2005; Stott, Piedrahita, & American Academy for Cerebral Palsy and Developmental Medicine, 2004). A partial hamstring release, involving the lengthening or transfer of muscles around the knee, also may facilitate sitting and walking. A lengthening of the Achilles tendon at the ankle improves walking (Figure 26.10).

More complicated orthopedic procedures may be required for correction of a dislocated hip. If this is diagnosed when there is a partial dislocation (called **subluxation**), release of the hip adductor muscles alone can be effective (Figure 26.11). If the head of the femur is dislocated more than one third to one half of the way out of a hip joint socket, a more complex procedure, a varus derotational osteotomy, may be necessary. In this operation, the angle of the femur (the thigh bone) is changed surgically to place the head of the femur back into the hip socket (Figure 26.12). In some cases, the hip socket also must be reshaped to ensure that the hip joint remains functional. Sometimes muscle releases or lengthening are performed at the same time as these bony procedures.

For ambulatory children with cerebral palsy, deciding which type of surgery is most likely to improve function is a complex issue. Computerized gait analysis conducted prior to surgical intervention has become increasingly common as an aid in the decision-making process. Precise measurements obtained through motion analysis, force plates, and electromyography offer detailed information relating to specific abnormalities at each lower extremity joint as well as the muscle activity that controls motion through all phases of the gait (Cook et al., 2003). Such precise definition is not possible through clinical observation alone. Preoperative gait analysis helps to determine exactly which procedures are likely to be successful. Postoperative analysis can provide an objective measure of outcome.

In addition to treating contractures and dislocations, orthopedic surgeons also are involved in the care of scoliosis, a complication of both spastic and nonspastic forms of cerebral palsy. If untreated, a spinal curvature can interfere with sitting, walking, and self-care skills. If severe enough, it also can affect lung capacity and respiratory efforts. Treatment of significant scoliosis ranges from a molded plastic jacket or a chair insert to invasive surgery to straighten the spine as much as possible. This surgery involves using rods and wires to hold the spine in

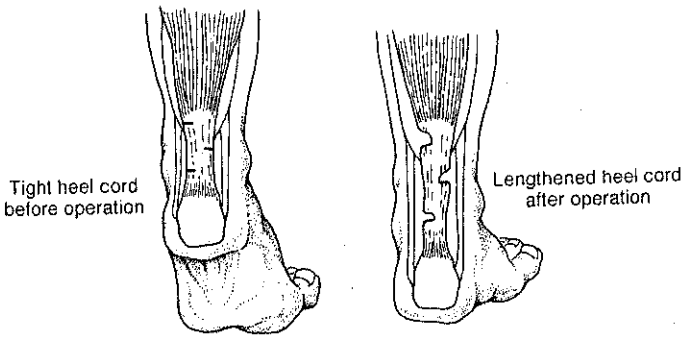

Figure 26.10. Achilles tendon lengthening operation. When the heel cord is tight, the child walks on his or her toes. Surgery lengthens the heel cord and permits a more flat-footed gait.

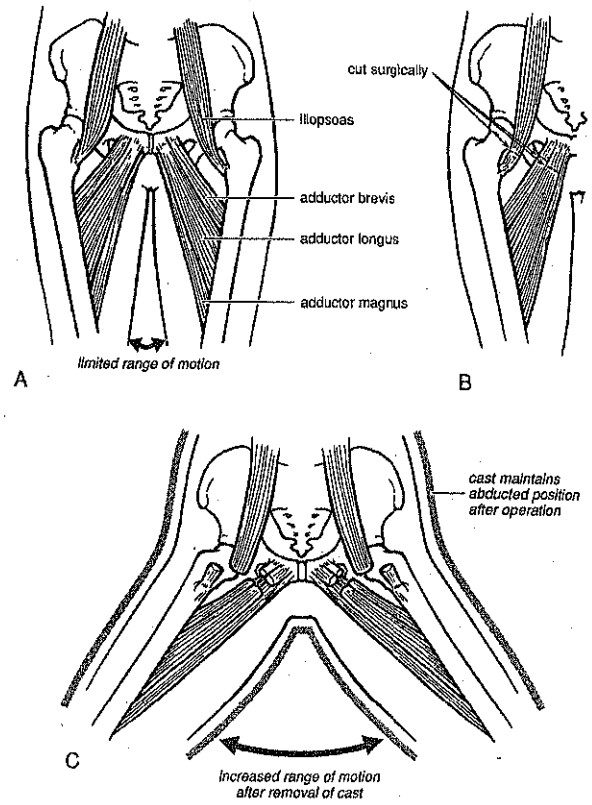

Figure 26.11. Adductor tenotomy. This operation is done to improve scissoring (Figure 26.5) and to prevent hip dislocation caused by contractures of the adductor muscles in the thigh (A). In this procedure, the iliopsoas, adductor brevis, and adductor longus muscles are cut, leaving the adductor magnus intact (B). The child is then placed in a cast for 6–8 weeks to maintain a more open (abducted) position (C). The muscles eventually grow together in a lengthened position, allowing improved sitting and/or walking.

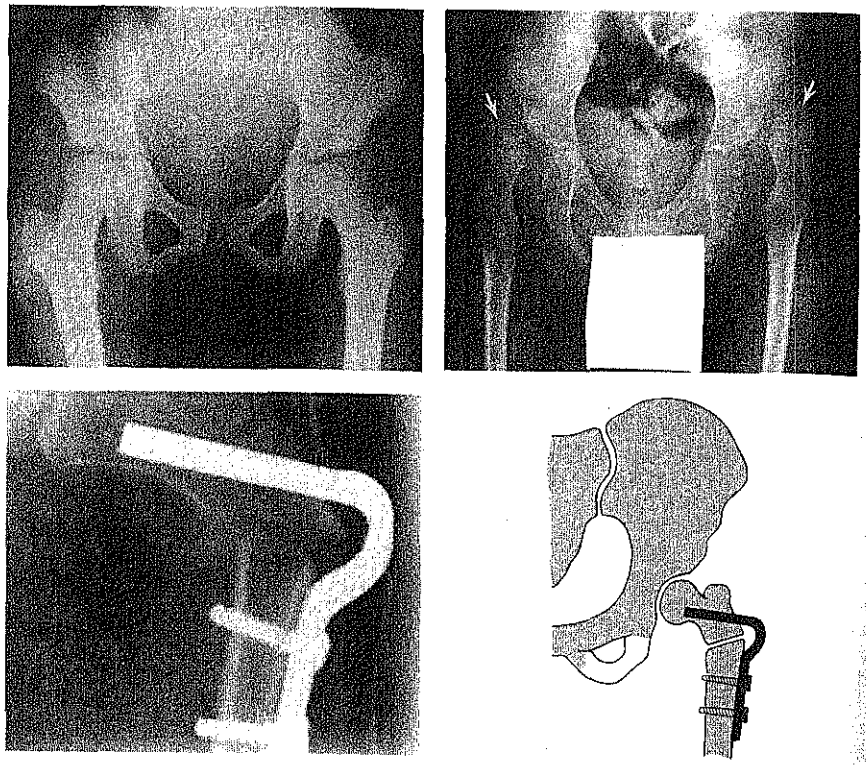

Figure 26.12. Dislocation of the hip. The upper X rays (frontal view) show a normal hip (left X ray) and a hip dislocated on both sides (right X ray). The arrows indicate the points of dislocation. The lower pictures show the results of a varus derotational osteotomy to correct the left-hip dislocation. The femur has been cut and realigned so that it now fits into the hip socket. Pins, which are later removed, hold the bone in place until it heals.

an improved alignment while bone graft material fuses the spine in position (Figure 26.13).

OUTCOME

Life Expectancy

Although most children with cerebral palsy will live to adulthood, their projected life expectancy is somewhat less than that of the general population (Hemming et al., 2005; Katz, 2003). Outcome varies for each type of cerebral palsy. A child with mild left hemiplegia probably will have a typical life span, whereas a child with spastic quadriplegia may not live beyond age 40 (Strauss & Shavelle, 2001). Children with very severe impairments, measured in terms of functional characteristics, have the poorest outcome. For example, children who cannot lift their heads and are fed via gastrostomy tube may not survive to adulthood (Strauss, Shavelle, & Anderson, 1998). Excess mortality for people with cerebral palsy may also be due to factors not directly connected with the cerebral palsy itself. Rates of mortality due to breast cancer, brain tumors, circulatory and digestive diseases, and accidents are higher for people with cerebral palsy than for the general population, suggesting that inadequate medical surveillance and psychosocial issues may play roles in excess mortality (Cooley & American Academy of Pediatrics, 2004; Strauss, Cable, & Shavelle, 1999).

Walking

When a child is first diagnosed with cerebral palsy, one of the first questions that arises in the minds of parents is whether their child will

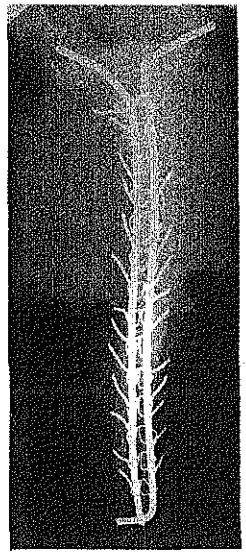

Figure 26.13. Treatment of scoliosis may require spinal fusion. This X ray shows improved scoliosis following a Luque procedure. During this surgery, the position of the spine is improved using metal hooks, rods, and wires while bone graft material fuses the spine in position.

eventually be able to walk. In addressing this question, it is important to recognize that "walking" can refer to several levels of ability. A child may be able to walk independently or may need crutches or a walker. A child may be able to walk long distances (community ambulation), short distances only (household ambulation), or only in the context of therapy (exercise ambulation). In general, children with better motor skills at a younger age (e.g., being able to sit and pull-to-stand before 2 years of age) have a better prognosis for walking than those with less well-developed skills. Recent research has provided greater specificity to this observation. For example, the previously described Gross Motor Function Classification System (GMFCS) can be used to estimate prognosis for walking (Rosenbaum et al., 2002; Wood & Rosenbaum, 2000). Children at any given level within the classification scheme tend to stay at that same level. In general, children at GMFCS Level of I or II will have a good prognosis for some degree of independent walking, children at Levels III and IV with have a variable prognosis for walking with some form of assistance, and children at Level V have a poor prognosis for any type of walking. Precise probability curves for ambulation have been published that allow even more exact predictions of ambulatory potential based on motor functioning at 2½ years of age (Wu et al., 2004). For example, using these curves it can be predicted that a child who is able to roll, sit, and pull-to-stand at 2½ years of age has a greater than 70% probability of being able to engage in some form of ambulation at age 7 years. By contrast, a child who can roll but cannot sit or pull-to-stand at 2½ years has approximately a 25% chance of walking and would most likely do this with the help of an **assistive device.**

Societal Independence

The ability to participate independently and effectively in a variety of societal settings is a complex function of a child's profile of abilities and disabilities but is also affected by environmental factors (e.g., family, neighborhood, economic) and by a child's health status (Liptak & Accardo, 2004). Motor skills and mobility may not be the primary determinant of societal independence. When asked, parents often identify communication and socialization as the functional areas of greatest concern to them. A child's ability to successfully participate in society is probably more strongly related to cognitive and interpersonal strengths than to physical ability.

Although about half of the individuals who have cerebral palsy have average intelligence, most of these individuals still have difficulty leading completely typical lives (Murphy, Molnar, & Lankasky, 1995). Studies suggest that employability is not related solely to the degree of disability but to a variety of other factors including family support, quality of educational programs, and the availability of community-based training and technical support (Russman & Gage, 1989). In a study of young adults with cerebral palsy (van der Dussen et al., 2001), 75% were fully independent with activities of daily living, 90% moved independently indoors, and 70% moved independently outdoors. The study also found that 77.5% of these individuals had adequate communication for telephone conversation, 30% lived with their parents, 12.5% lived with a partner, and 32.5% lived alone. In addition, 53% had some form of secondary education, but only 36.3% had paid employment. It is hoped that these figures will improve as a result of federal mandates (e.g., the Americans with Disabilities Act [ADA] of 1990, PL 101-336), which define the rights of people with disabilities and are making inroads into societal

perceptions of disability. Once society recognizes that functional outcomes are related as much to societal conditions as they are to the characteristics of a particular child with a disability, society's perception of outcome will undergo a major shift. Ultimately, strengthening supports to families (Raina et al., 2005) improving schools, increasing opportunities for employment, and changing attitudes about disabilities in society at large may do as much for children with cerebral palsy as traditional therapy and medical interventions.

SUMMARY

Cerebral palsy is a developmental disability that results from damage to or dysfunction of the developing brain. The impairments associated with cerebral palsy are nonprogressive but permanent. Varying degrees of ability related to functional mobility, daily living skills, and communication/socialization skills result from these impairments. Habilitation is an interdisciplinary strategy that seeks to maximize function and minimize the disadvantage a person experiences as a consequence of disability or societal circumstances. Efforts founded on the principles articulated in the ADA will create new opportunities for greater participation and enhanced quality of life for people with cerebral palsy.

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27 Movement Disorders

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Upon completion of this chapter, the reader should be able to

- Define and recognize different types of movement disorders that occur in children
- Understand treatment approaches to and outcomes of several common movement disorders in children
- Recognize certain normal behaviors that are frequently mistaken for movement disorders

Movement disorders are common in childhood, with certain types occurring in more than 10% of school-age children (Kurlan et al., 2001). They may occur as a primary disorder or secondary to other disorders of the nervous system such as cerebral palsy or traumatic brain injury. Movement disorders may be mistaken for other episodic disorders that are common in childhood, such as seizures. Furthermore, movements that are within the range of typical childhood behaviors may be misdiagnosed as abnormal, leading to unnecessary testing.

REGGIE

At 7 years of age, Reggie's parents began to notice that he would intermittently tilt his head to the left or right with a sudden, quick motion. Within a few months, they observed the gradual onset of episodic movements, including eye blinking, shoulder shrugging, and twitching at the corner of his mouth. The movements gradually subsided, but Reggie began to frequently clear his throat for no apparent reason. He was seen by his pediatrician, who examined his oropharynx and sent a rapid test for streptococcal infection. His physical examination was normal, and the strep test was negative. The movements and sounds were increased by stress, anxiety, and fatigue, and they seemed better when he was concentrating on other activities. When asked to suppress the throat clearing or the movements, Reggie could do so only briefly. Reggie's movements were noticed by his teachers and classmates at school, causing hurtful comments from peers and his withdrawal from social activities. More than 1 year after the movements began, Reggie was referred to a child neurologist, who observed Reggie's **tics.** The neurologic examination was otherwise normal. Reggie was diagnosed with Tourette syndrome and started on therapy with clonidine. After a number of dose adjustments, his tics improved. At 9 years of age, Reggie now has many friends in his peer group and is doing well in school.

DEFINITION AND CLASSIFICATION OF MOVEMENT DISORDERS

Movement disorders consist of either 1) a loss or poverty of movement (akinesia) or slowness (bradykinesia) of movement that is not associated with weakness or paralysis, or 2) an excess of abnormal involuntary movements (dyskinesia). Various systems exist for classifying movement disorders. In one approach, disorders are categorized on the basis of whether they are **paroxysmal** (occurring in episodes of sudden onset); transient developmental phenomena (resolving in early childhood); secondary to a noninherited, static injury; or a manifestation of a hereditary or metabolic disorder. Using this system, there is the potential for overlap among categories. For example, tics may occur as a paroxysmal disorder, may be a transient developmental phenomenon, or may be due to a