Cerebral Palsy
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FOCUS QUESTIONS
1. In reaching a diagnosis, how would you differentiate among diplegic, hemiplegic, quadriplegic, and athetoid cerebral palsy?
2. What are the most common causes of each of the four types of cerebral palsy?
3. What risk factors are associated with cerebral palsy?
4. What disabilities are most likely to be associated with cerebral palsy, and how are they managed?

To be classified as cerebral palsy (CP), there must be difficulty in neuromotor control, a nonprogressive brain lesion, and an injury to the brain that occurred before it was fully mature. The term ‘cerebral palsy’ should be used only if a static encephalopathy exists. If there is any question that a progressive central nervous system disorder exists, the term ‘cerebral palsy’ should not be used diagnostically until the status of the lesion is clarified.

Although the primary abnormality must be a motor deficit, often there are many associated symptoms of cerebral dysfunction present.

Incidence and Prevalence
The prevalence of CP has changed very little over the past 40 years, in spite of many technological advances that have decreased mortality in compromised preterm and full-term infants. The prevalence rate has been estimated to be between 2 and 5 per 1000 live births. At 12 months of age, the prevalence rate was estimated to be 5.2 per 1000, but at 7 years of age, the rate was estimated to be 2 per 1000 live births. This indicates that many children who showed signs or experienced symptoms suggesting a motor disorder did not have CP on follow-up.

The past 3 decades have seen an increased survival rate of very small preterm infants, resulting in a change in the percentage rates of the different clinical types of motor disabilities among patients classified as having CP. Because preterm infants are susceptible to developing a spastic diplegia, this type of disability now is the most common. Of children whose CP is identified at 7 years of age, one third have a spastic diplegia.

There is a definite correlation between birthweight and/or gestational age and the incidence of CP.

Etiology
The insult to the brain may occur prenatally, perinatally, or postnatally. Until recently it was believed that hypoxic/ischemic incidents occurring perinatally caused the majority of CP cases. However, a collaborative perinatal project started in 1960 that followed 454,000 pregnant women and the outcome of their offspring revealed that adverse prenatal factors may be the primary culprits. An abnormal fetal brain or fetal environment may make the infant more vulnerable for being born prematurely or, if born full-term, may place him or her at risk for cardiorespiratory problems in the neonatal period.

In more than 50% of the patients who have CP, an etiology may not be evident, even after having obtained a comprehensive history for known high-risk factors and a genetic history; having performed a complete physical and neurologic examinations; and having evaluated metabolic, chromosomal, and neuroimaging studies. Although prematurity is the most common antecedent of CP, the majority of infants who develop this disorder have had full gestational terms. This paradox is explained by there being seven to ten times more full-term than preterm babies born.

The lower the birthweight, the higher the incidence of CP. However, even for very low-birthweight infants (<1500 g), there is only a 15% to 20% risk of CP. Preterm infants who develop intraventricular hemorrhage, especially with extension of the hemorrhage into the white matter, are at the greatest risk for developing CP.

However, the best predictor of CP is the presence of echodensities in the periventricular white matter that develop into cysts. If the cysts are more than 3 mm in diameter, it has been estimated that there is a 90% to 95% chance of developing a spastic diplegic CP.

The Apgar score, whether in preterm or full-term infants, has been a poor indicator of babies at risk for brain damage. For instance, with an Apgar score of 3 or less at 10 to 15 minutes, the risk of developing CP varies from 10% to 15%. On the other hand, the majority of full-term infants who develop CP have normal Apgar scores.

Table 1 lists the timing of pathologic periods in CP. It is evident that a relatively high percentage of infants who have CP had no identifiable risk factors and that postnatal causes are infrequent.

Certain types of insults have clinical pathologic correlations. For example, in preterm infants the germinal matrix is juxtaposed to the periventricular white matter. The germinal matrix of a preterm infant is susceptible to bleeding because it has a low resistance to hypoxia and/or ischemia. The immediate periventricular white matter contains the long-tract pyramidal fibers that descend to the spinal cord segments responsible for neuromotor control of the lower extremities. More peripheral are the pyramidal fibers destined for the cervical spinal cord segments. Consequently, the pathology in preterm infants usually is limited to the long tracts that supply the lower extremities, resulting in a spastic diplegia (eg, legs involved more than arms).

Another clinical pathologic correlation is seen with bilirubin encephalopathy. Bilirubin is especially toxic to the neurons of the basal ganglia. Hyperbilirubinemia existing in an immature nervous system, if in toxic amounts, results in athetoid CP. Also, the auditory nuclei in the brain stem are especially susceptible to the toxic effects of bilirubin. Therefore, a high percentage of children who have athetoid CP have a neurosensory hearing loss.

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Classification
Classification of the cerebral palsies still is based on the type of motor disorder, not on etiology or pathology. Although the available neuroimaging techniques may help in defining the anatomic deficit better, they still will not reveal any pathology in the majority of children who have CP. The clinical classification is based on the extremities involved, the type of tone abnormalities, and the characteristics of the involuntary movements. One problem in using the type of motor dysfunction for clinical classification is that the neurologic picture may change as the infant grows older. Therefore, the exact type of motor deficit may not be clarified until 2 to 3 years of age. An added difficulty is that 20% to 30% of infants who appear to fit the criteria for the diagnosis of CP at 1 year of age will not show any manifestations of a motor deficit by 7 years of age. Pediatricians must be cautious in prognosticating whether an infant will or will not have CP or in offering a judgment as to future cognitive abilities.

Clinical Characteristics
Table 2 offers a clinical classification of cerebral palsy. Spastic diplegia encompasses the largest number of CP patients. In the past decade, the number of patients who have spastic diplegia have, relatively speaking, increased in numbers, concomitant with the increased survival rates of preterm infants.

Because the insults to the brain that produce CP often result in diffuse pathology, the mixed types of CP are not uncommon. Those clinicians who appraise movement disorders carefully find that more than 10% are of the mixed variety. Prior to the use of immunoglobulins to prevent blood incompatibility from developing in pregnant women, bilirubin encephalopathy with resulting kernicterus and athetosis occurred at a higher percentage than shown in Table 2. Now the most common cause of the athehtoid-dystonic type of CP is hypoxic-ischemic injury to the basic ganglia.

Ataxic-spastic CP and spastic CP frequently have similar antecedents. Hydrocephalus often is present in ataxic-spastic disorders. Genetic factors have been implicated in the pure ataxic CP. Also, it is necessary to be aware that when ataxia is the primary motor disorder, one should not immediately conclude that the diagnosis is CP. Ataxia is a relatively uncommon type of CP, but it is not uncommon as a manifestation of a progressive neurometabolic encephalopathy. The child who has nonprogressive ataxic CP frequently is mentally retarded.

Many children who have insults to their motor systems at birth will show transient hypotonia that develops into increased tone of the spastic or rigid type. A few will remain hypotonic throughout life. These children who have “ataxic CP” have severe motor and intellectual disabilities.

Clinical Approach to Diagnosis
Any infant whose motor development has been delayed must be considered suspect for having CP. Diagnosis is very difficult before 6 months of age, primarily because abnormalities in tone or reflexes or involuntary movements rarely manifest during early infancy. The primary reason for this is that much of the movement observed in infants younger than a few months of age is of reflex origin and not under voluntary motor cortical control. It is only with maturation of the cortex that the clinical picture of CP emerges more clearly. There often is additional diagnostic uncertainty after an acute perinatal insult to the brain. Tone and reflex abnormalities may be noted immediately and falsely indicate permanent damage to the central nervous system motor system. However, the tone and reflex changes may prove to be transient phenomena; after 2 to 12 months, these signs of CP disappear.

Delay in achieving motor milestones at an appropriate age is common for children who are retarded but who do not have CP. Adding to the diagnostic dilemma is that many of these children present with hypotonia. One should be much more highly suspicious of the presence of CP if there is a motor delay but evidence of normal cognitive development. However, that is not to say that children who have CP cannot be retarded; more than 50% of them are.

When assessing a child who has a motor delay, it is necessary to decide where the pathology exists anatomically: centrally (eg, brain) or peripherally (eg, spinal cord, anterior horn cell, peripheral nerve, myoneural junction, or muscle). If the brain is the site of the pathology, the child is considered to have CP.

It also is important to determine if the motor delay is a manifestation of a progressive central nervous system disorder. Clues to a progressive disorder are: 1) regression; 2) normal development for a period of time, then a slowing; 3) consanguinity; 4) congenital skeletal anomalies such as pes cavus or scoliosis; and 5) neuromuscular stigmata. In certain progressive diseases, the evidence of regression may not occur until very late in the course; at the early onset of the disorder, only a delay in achievement of motor milestones.

### Table 1. Timing of Pathogenetic Periods in Cerebral Palsy*

<table>
<thead>
<tr>
<th>Type of Palsy</th>
<th>TERM BIRTHS N = 457 (%)</th>
<th>PRETERM BIRTHS N = 224 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Obvious prenatal cause or risk factor</td>
<td>24</td>
<td>6</td>
</tr>
<tr>
<td>Combined prenatal and perinatal factors</td>
<td>20</td>
<td>23</td>
</tr>
<tr>
<td>Purely perinatal factors</td>
<td>19</td>
<td>51</td>
</tr>
<tr>
<td>No identifiable risk factors</td>
<td>30</td>
<td>17</td>
</tr>
<tr>
<td>Obvious postnatal cause</td>
<td>8</td>
<td>2</td>
</tr>
</tbody>
</table>

*Adapted from Hagberg B and Hagberg G (210, pp 116–134)
(and not regression) will be the presenting symptoms.

Spastic paraparesis, if associated with a peripheral sensory disturbance, is not a manifestation of CP, but indicates a transverse myelitis, an intrathecal mass lesion, or a neurodegenerative spinal cord disorder.

Primary lower motor neuron lesions (anterior horn cell, peripheral nerve, myoneural junction, or muscle) are manifested by weakness, low tone, and decreased-to-absent reflexes. There is no involuntary movement.

**Early Signs**

It is difficult to diagnose CP before the infant is 6 months of age. However, a number of clues can raise the index of suspicion. These so-called high-risk findings can be assessed easily during a routine well-child examination. Many of the tests do not require a “cooperative” infant. Tone, primitive reflexes, and deep tendon reflexes are the mainstay of the examination.

**TONE CHANGES**

An infant who has CP may present with either low tone or increased tone of the trunk and/or appendicular muscles. The assessment of tone lacks precise parameters and often is based on clinical judgment. However, a few guidelines are available, although they lack specificity. Following are a few examples appropriate for clinical examination.

**Hypotonia.** The anterior scarf sign is elicited with the infant in the prone position. Both shoulders are held against the table, and with the head in the midline, the examiner pulls one arm across the infant’s chest toward the opposite ear. Increased range of motion of joints (hypotonia) is indicated if the elbow can be drawn past the chin. For the lower extremities, the abductor sign is elicited with the baby supine. The knees are extended and the hips abducted. Abduction greater than 160° suggests increased laxity of the hip joints.

**Hypertonia.** Assessment of hypertonia is difficult if a baby is crying, which is a common problem during routine examinations. However, this does not necessarily preclude a meaningful evaluation. To appreciate the value of this type of examination, it is helpful to know that when a newborn is held prone, a tonic labyrinth reflex results in increased flexor posturing of trunk and extremities. Placing the newborn baby supine results in a slight decrease in this flexor posturing toward more extension. As the newborn matures, the flexor posturing in the prone position diminishes and extensor tone tends to increase. The finding of premature development of extensor tone, or at the other extreme, a marked loss of flexor tone in the prone position should arouse suspicion of a motor problem.

If the newborn baby is held in ventral suspension, the elbows, knees, and hips should be flexed and the neck slightly flexed. At about 6 weeks of age, this flexor posturing decreases but still is present; now the head is usually in line with the trunk. If the head is hyperextended when the infant is held in ventral suspension at this early age, this is considered to be an abnormality in extensor tone and may reflect an underlying motor disturbance. If the same baby is placed on the table prone and the buttocks are still riding high—that is, above the trunk—this would be due to increased flexion at the hips, which should have lessened by 6 weeks to 3 months of age. This is also an abnormal manifestation of tone.

When the baby is picked up from the supine position, the head should come up with the trunk at around 3 months of age. A persistent head lag can be due to increased extensor tone. In fact, if a baby seems to have good head control at 6 weeks of age, as indicated by the head being held above the trunk in ventral suspension, one can prove, by picking the baby up from the supine position, that this is not due to good voluntary control but is only a manifestation of increased reflexive extensor tone. If the baby has good voluntary head control, he or she should be able to bring the head up in line with the trunk. The head lagging behind is proof that the finding of good head control in ventral suspension was deceptive.

After ventral suspension is observed, the baby should be placed in vertical suspension. An infant who “scissors” (Fig. 1) after the age of 2 months is considered to have increased hip adductor tone, which is an early indicator that the infant eventually may develop more classic signs of CP.

**PRIMITIVE REFLEXES**

There is value in testing specific primitive reflexes during a routine physical examination. The asymmetric tonic neck reflex never should be obligatory. If the head is turned in one direction and the extremities move into the fencing position with

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**TABLE 2. Clinical Classification of Cerebral Palsy**

<table>
<thead>
<tr>
<th>TYPE</th>
<th>FREQUENCY %</th>
</tr>
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<tbody>
<tr>
<td>Spastic</td>
<td>70–80</td>
</tr>
<tr>
<td>Hemiparesis (monoparesis)</td>
<td></td>
</tr>
<tr>
<td>*Diplegia</td>
<td></td>
</tr>
<tr>
<td>Quadripleesis (double hemiplegia)</td>
<td></td>
</tr>
<tr>
<td>Dyskinetic</td>
<td>10–15</td>
</tr>
<tr>
<td>Athetoid</td>
<td></td>
</tr>
<tr>
<td>Dystonic</td>
<td></td>
</tr>
<tr>
<td>Chorea</td>
<td></td>
</tr>
<tr>
<td>Ballismus</td>
<td></td>
</tr>
<tr>
<td>Tremor</td>
<td></td>
</tr>
<tr>
<td>Rigid</td>
<td>5</td>
</tr>
<tr>
<td>Ataxic</td>
<td>1</td>
</tr>
<tr>
<td>Mixed</td>
<td>10–15</td>
</tr>
</tbody>
</table>

*Diplegia is becoming the most common type of spastic CP as more preterm infants survive.*
weight. This reflex usually disappears at about 4 months of age.

DEEP TENDON REFLEXES
To elicit the status of the deep tendon reflexes accurately, it is helpful to have the baby relaxed in the security of the mother’s lap. The primary area of concern is the presence of hyperreflexia, which indicates an upper motor neuron lesion. If the hyperreflexia is truly pathologic, there frequently is an increased sensory input zone for obtaining a primary motor response. For instance, if the examiner initially percusses over the deltoid and obtains a reflex contraction of the biceps, this would be considered a 4+ bicep jerk. Additionally, percussion over the shins or any place below the patellar tendon that results in knee extension is pathologic. Ankle clonus is not uncommon among normal infants up to a few months of age, but sustained ankle clonus is cause for concern.

The presence of decreased-to-absent deep tendon reflexes usually indicates a lower motor neuron disease.

Clinical Presentations of CP

SPASTIC HEMIPARESIS
A baby who eventually develops a spastic hemiparesis rarely shows any asymmetry of movement in the first few months of life. In fact, a Moro reflex will not be characterized by asymmetry of one upper extremity over the other. If the Moro response is characterized by this asymmetry, the examiner should look for a lower motor neuron lesion on the side of the extremity that responded poorly.

It is usually at about 4 months of age that a mother notices her infant tending to maintain a fisted hand position on one side compared with the contralateral extremity. It may be difficult to discern an asymmetry of tone or reflexes in the upper extremities in the clinic. Tone and reflex changes rarely are evident in young infants and, even if present, having the baby relaxed enough for an accurate measurement frequently is impossible. Therefore, if the formal neurologic examination does not result in confirmatory “hard” signs of CP, the examiner should not rule out this possibility, especially if the history indicates early development of “handedness.” It is usually at approximately 6 months of age that tone and reflex asymmetry in the upper extremity evolves. The same baby may crawl late and, if so, will use a nonalternating pattern that involves pushing off with the arm and leg on the normal side and dragging the contralateral extremities. Even at 10 months of age, asymmetry in re-
flex and tone may not be evident in the lower extremities. However, these findings will become more apparent at approximately 12 to 15 months of age.

The first sign of spasticity in the upper extremities is tightness of the elbow flexors and wrist pronators. By comparing differences in the ease of supination and pronation at the wrist, one may be able to judge any inequality in resistance to passive movement. Spasticity of the upper extremity features resistance when attempting supination. In the lower extremities, the first muscle that develops spasticity is usually the gastrocnemius. The examiner will notice that it is more difficult to dorsiflex the ankle of the involved extremity compared with the normal one.

Another method for comparing tone is to hold the arms in a position so that the hands can be “flapped” up and down rapidly. One occasionally may be able to make a judgment of asymmetry in terms of the ease of movement. Flapping of the ankles can be assessed similarly.

The final extent of the spasticity and the motor disability may not become apparent until 2 to 3 years of age.

ATHETOID CP

Newborns who develop bilirubin encephalopathy will show signs of kernicterus at about 3 to 5 days of age. The infant will develop a high-pitched cry, have increased extensor tone manifested as opisthotonus, and have increased deep tendon reflexes. If the baby survives, the increased extensor tone will decrease gradually at about 2 weeks of age, and by 6 weeks of age, the tone may be variable or possibly normal. Even if the infant has normal tone, crying will result in an abnormal exaggeration of extensor tone. As the infant reaches 3 months of age, tone will have decreased further to the point where the baby appears “floppy.” Deep tendon reflexes most likely will be within normal limits at this stage. The primary neurologic finding beyond the decreased tone is an obligatory asymmetric tonic neck reflex.

This infant will be delayed in achieving motor milestones, although the motor delay will not be on the background of reflex changes or involuntary movements. For example, at 10 months of age, the infant will use a raking movement when reaching out for a pellet instead of the expected pincer grasp. This immature raking motion will be done without any involuntary movements. It is not until about 12 to 18 months of age that the infant will show athetoid-dystonic posturing on voluntary movement. It is around this period that the hypertonia gradually changes to increased tone of the rigid or cogwheel type. During these sequential changes, the deep tendon reflexes will continue to be normal. However, the asymmetric tonic neck reflex will persist. The hypertonus and the involuntary movements may increase in severity gradually over the next year.

These dramatic changes on the background of a static encephalopathy may cause the pediatrician to suspect a progressive central nervous system lesion. However, knowing about the presence of neonatal hyperbilirubinemia and the natural course of bilirubin encephalopathy will give the progressive changes in signs and symptoms a reasonable explanation.

Preterm infants are susceptible to developing a spastic diplegia, the most common type of disability in CP.

SPASTIC DIPLEGIA

Many preterm infants who have CP will develop spastic diplegia. It usually is manifested initially by delayed sitting, especially if the trunk is involved. Crawling, standing, and ambulation also will be delayed. In infancy, low tone of the lower extremities may be present. It is not until after 1 year of age that hypertonia (spasticity) will be evident. However, there are clues to indicate that the initial low tone does not suggest a lower motor neuron lesion, but reflects an upper motor neuron lesion. Increased deep tendon reflexes and occasionally unsustained bilateral clonus will be present. Also, one may be able to elicit abnormal postural reactions. For example, when the baby is picked up quickly and placed in the vertical position, he or she may exhibit a scissoring of the legs due to spasticity of the hip adductors. Also, when the infant is held vertically and the soles touch a hard surface, he or she will exhibit a positive supporting reaction with plantar flexion at the ankles and extensions at the knees and the hips. A positive supporting reaction usually disappears at about 4 to 6 months of age.

Babies who have a spastic diplegia frequently will not use an alternating pattern with their lower extremities when crawling, but will exhibit an “army” crawl using their upper extremities.

ATAXIC CP

Babies who have ataxic CP present with low tone of trunk and extremities, usually with normal deep tendon reflexes. As they mature, they tend to support their weight with their knees extended so as to lock them in position, and they use a very wide base for support. It is rare to see any overt signs of ataxia, such as an intention tremor or head titubation in infancy. A complicating factor may be a spastic component to the condition. In such cases, the infant usually will be hypotonic and not spastic, but hyper-

SPASTIC QUADRIPE SIS

Infants who have this type of CP also pass through a stage of hypertonia prior to the appearance of spasticity and increased muscle tone. Early manifestations of spasticity are similar to those described in other spastic clinical types and affect both lower and upper extremities.

Management

Informing the family about a suspected or established diagnosis of CP should be compassionate and factual, but should acknowledge the limitations of early prognostication. Enrollment in an early intervention program will provide family support for coping, parental education for handling a child at home, and treatment
to promote motor and other developmental skills of the child. PL99-457 is a federal law that mandates such programs in all states.

The long-range management of children who have CP addresses both motor dysfunction and associated nonmotor deficits. Equally important are measures to assist adjustment of the family because the home milieu will affect the child’s emotional and personality development.

**MOTOR DYSFUNCTION**

In general terms, the aim of treating the physical disability is to improve function and to prevent secondary complications of the musculoskeletal system. The pediatrician must establish realistic expectations for the child’s motor function and ensure that progressive contractures and deformities do not interfere with optimal outcome.

**When assessing a child who has a motor delay, it is necessary to determine where the pathology exists anatomically. . . If the brain is the site of the pathology, the child is considered to have CP.**

Physical therapy is a treatment used frequently to promote gross motor skills. Methods proposed for CP are numerous, and new ones continue to emerge. Most techniques focus on specific aspects of abnormal movement control as a key to successful remediation. Studies on the efficacy or comparative effectiveness of different therapeutic methods are scarce or nonexistent.

Occupational therapy concentrates on eye-hand coordination and upper extremity motor control to attain the highest level of independence in self-care and other activities of daily life that involve hand function. Pseudobulbar palsy in bilateral impairment of the motor system leads to oral motor dysfunction manifested as early sucking, swallowing, and chewing difficulties. In the hands of trained occupational or speech therapists, proper positioning and feeding methods help to alleviate these problems. Parental involvement and training in special feeding techniques should be emphasized. Early feeding difficulties often precede impaired speech production.

Assistive devices and adapted functional techniques offer compensatory means for independence in many gross and fine motor activities that otherwise are not feasible to achieve. Ambulation with crutches or wheelchair and use of adapted feeding utensils or writing implements are only some examples. A properly constructed wheelchair insert can modify abnormal tone and posture, improve sitting balance, and thereby allow better arm use. Rehabilitation engineering revolutionized the application of electronic and computer technology for disabled persons. Motorized wheelchairs, nonverbal communication, and environmental control devices have opened up new possibilities for children who have severe physical handicaps but adequate cognitive function.

Biofeedback with electromyography and auditory signal monitoring has been used to modify the activity of selected muscle groups. Improved gait has been reported in children who have hemiparesis, but long-term follow-up indicates that the initial good response was lost by 6 to 12 months after discontinuation of treatment. Biofeedback training of the orbicularis oris also has been recommended to decrease drooling. Experience with long-range effectiveness is limited.

Bracing usually is prescribed to correct abnormal posture of the ankle and foot, such as toe walking, in ambulatory children. Braces extending above the knee or hip largely have been abandoned as a walking aid for children who have CP. An inhibitory walking cast that maintains the foot in good anatomic alignment stretches a tight heel cord and may decrease the tendency to toe walk, at least temporarily. Positioning braces and applying splints at rest can help to prevent or delay contractures of the Achilles tendon, hamstring, or hip adductors.

Antispasticity medications affect muscle tone through different mechanisms of action. Diazepam is a centrally acting muscle relaxant with sedative side effects that limit its use as an antispasticity agent. Dantrolene sodium reduces spastic hypertonicity by decreasing the contractile properties of skeletal muscle fibers. Liver function needs to be monitored, although hepatotoxicity has not been reported in children. Baclofen is a gamma-aminobutyric acid derivative that is thought to inhibit neurotransmission. This drug is more effective for spasticity of spinal origin, but it may be used for adjunctive treatment in CP that is accompanied by severe spasticity.

Topical injection of phenol, alcohol solution, or botulinum toxin into the motor points or motor nerves of a spastic muscle creates a temporary neuromyelosis and consequent tone reduction lasting 4 to 6 months. The procedure usually is a temporizing measure and most often is applied to alleviate spasticity of the ankle plantar flexors and hip adductors.

Surgical therapy—orthopedic surgery—often is recommended to improve both function and appearance and to prevent or correct deformities. For example, lengthening of a tight or contracted heel cord corrects toe walking and enhances the efficiency of gait. Other frequently performed soft-tissue procedures include release of spastic hip and knee flexors and tenotomy of the hip adductors. In severely affected children, the primary reason for surgery is to prevent progressive skeletal deformities that inevitably follow spastic muscle imbalance. Soft-tissue procedures alone are not successful when an underlying bone deformity already is present. Osteotomy or other bone procedures need to be considered in such cases. Acquired hip dislocation resulting from spasticity of the hip adductors and flexors is a serious complication in bilateral lower extremity involvement that is especially frequent in the nonambulatory group of patients. In spastic quadriparesis, scoliosis can develop in the presence or absence of hip dislocation. After postoperative immobilization, intensive physical therapy is required to regain strength and function. Long-range maintenance of surgical correction is difficult in severely affected children whose active function is limited.

Selective posterior rhizotomy is the
current neurosurgical approach to alleviate spasticity. The procedure consists of L2 to L4 laminectomy to localize the posterior roots in the cauda equina. Electrical stimulation applied to the individual rootlets is monitored by electromyography. Rootlets that produce clonic or sustain stimulus response, especially with spread to other muscle groups, are severed because they are thought to be responsible for spastic disinhibition. The procedure is recommended mostly for those children who have spastic diplegia but who can walk and have adequate strength, motor control, balance, and reasonable intelligence. Daily physical therapy is recommended for 6 to 12 months after surgery. Postoperative complications are rare and include transient paresthesias and loss of bladder control. Although the procedure reduces spastic hypertonicity, whether it significantly influences the natural course of motor disability remains a matter of debate.

ASSOCIATED DYSFUNCTIONS

Audiometric testing is part of the diagnostic evaluation. High-frequency hearing loss is typical of athetoid CP caused by bilirubin encephalopathy and is present in 70% of cases. The frequency of sensorineural hearing loss is approximately 30% in other clinical types. Early intervention to reduce hearing loss is necessary to avoid interference with speech development.

Speech and language development should be monitored because children who have CP have an increased incidence of dysarthria, verball dyspraxia, or other central language disorders. Inadequate language development also may reflect a cognitive deficit. Speech impairment is a sign of pseudobulbar palsy associated with bilateral spasticity or atresia. Speech production defects range from a mild articulation disorder to severe dysarthria with complete lack of intelligible verbal communication. Milder forms of articulation defects can benefit from speech therapy. Augmentative, nonverbal communication training for both spoken and written language is the method of choice when there is a profound speech production deficit. Many children who have severe motor and speech disability were thought to be retarded in the past but they proved to have good educational potential by using this method of communication.

Uncontrolled drooling is another manifestation of pseudobulbar palsy and represents a socially embarrassing handicap. Some children improve spontaneously by 5 to 10 years of age. For persistent drooling, anticholinergic drugs, biofeedback, or behavioral intervention may be tried. Transposition of the parotid and/or submandibular ducts, extirpation of the submandibular gland, or resection of the chorda tympani are surgical procedures for redirecting or decreasing salivation.

There is no consensus as to the optimal surgical approach for controlling drooling. Most surgeons report a 60% to 80% success rate with any of the techniques used. Depending on the surgical approach, complications include xerostomia, thick secretions that impair the ability to swallow food, an increase in dental care, and pain and swelling of the parotid gland. Rerouting the parotid duct to the posterior pharynx may increase the pulmonary complications of posterior drooling. Treatment should be based on assessment of each child’s drooling problem.

Assessment of vision is part of routine evaluation. Refractive errors and retinopathy of prematurity are associated visual complications in CP. The incidence of strabismus is approximately 75%, with the highest frequency seen in the spastic clinical types of CP. Timely referral to an ophthalmologist and early treatment will prevent amblyopia.

Approximately 50% of the children who have CP develop some form of seizure disorder. The highest incidence is in those who have spastic hemiparesis and quadriplegia. Atypical psychomotor seizures may be difficult to recognize. The choice of anticonvulsant medications depends on the type of seizure disorder. Side effects on behavior and alertness also should be considered.

Assessment of intelligence in those who have CP requires the skills of a psychologist experienced with physical disabilities. Mental retardation occurs in about 50% of those affected, and children of normal intelligence may have a learning disability. Planning for appropriate educational placement should start before the child reaches school age. Intellectual and learning abilities are the deciding factors for selecting the most suitable school. Environmental adaptations can accommodate physical disability and are mandated in the United States by PL94-142, the Education of All Handicapped Children Act. Children who require special education should be mainstreamed for selected activities.

The energy expenditure for walking and other physical activities is several times above that of normal in motor disabilities. This should be considered in dietary guidance of active children who have CP. On the other hand, a low level of physical activity in severely handicapped children may lead to obesity. The increased weight may increase the energy expenditure of a limited ambulator to such a degree that a wheelchair existence becomes necessary. This is not an unusual scenario among adolescents.

Participation in “adaptive” sports should be encouraged. Swimming and horseback riding seem to be the most popular for children who have CP, but virtually all sports can be adapted to a motor disability, especially with wheelchair use.

Psychosocial Guidance

Anticipatory guidance of the family and, at the appropriate age, of the child is perhaps the most complicated aspect of management. Preoccupation with treatment of the motor disability should not distract attention from the need to foster an independent, resilient personality, with determination and drive to succeed. Children who have CP should have a variety of social and recreational opportunities similar to those of nondisabled youngsters. Acquisition of good social skills during childhood is essential for attaining maximal functional and vocational potential in adult life.

Outcome

In predicting long-range outcome, one must consider multiple variables. Physical and intellectual abilities are the primary determinants of func-
tional potential, but the achievement of these expectations also depends on psychosocial adjustment.

Children who have hemiparesis walk by 1.5 to 3 years of age; 80% to 90% of children who have diplegia, 70% of those who have dyskinesia, and 50% who have quadriplegia attain some mode of ambulation. In these clinical types the ability to maintain independent sitting by 1.5 to 2 years offers a good prognosis for community ambulation. Children who learn to sit between 2 and 4 years usually become household ambulators, and some may walk short distances outdoors with assistive devices. Walking is not expected of children who cannot sit by 4 years of age. Despite the unilateral hand impairment, hemiparesis does not exclude independence in daily activities. Motor control of the upper extremities is adequate to perform activities of daily living in diplegia and usually in cases of mild ambulatory quadriplegia. Some children who have quadriplegia and rely on wheelchair mobility may achieve partial independence in self-care. About 25% of this group requires help for all activities. Athetoid and ataxic movement disorders tend to compromise fine hand dexterity to a greater extent than ambulation. Mental retardation delays but does not exclude walking if it is permitted by the motor disability. Achievements in daily living skills are consistent with intellectual competence, regardless of the degree of physical disability.

SUGGESTED READING