Cerebral Palsy

Cerebral palsy (CP) is a diagnostic term used to describe a group of motor syndromes resulting from disorders of early brain development. CP is caused by a broad group of developmental, genetic, metabolic, ischemic, infectious, and other acquired etiologies that produce a common group of neurologic phenotypes. Although it has historically been considered a static encephalopathy, this term is now inaccurate because of the recognition that the neurologic features of CP often change or progress over time. In addition, although CP is often associated with epilepsy and abnormalities of speech, vision, and intellect, it is the selective vulnerability of the brain's motor systems that defines the disorder. Many children and adults with CP function at a high educational and vocational level, without any sign of the type of cognitive dysfunction that is generally implied by the term encephalopathy.

Epidemiology and Etiology.

CP is the most common and costly form of chronic motor disability that begins in childhood with a prevalence of 2/1000. The Collaborative Perinatal Project, in which approximately 45,000 children were regularly monitored from pregnancy to the age of 7 yr, found that most children with CP had been born at term with uncomplicated labors and deliveries. In 80% of cases, features were identified pointing to antenatal factors causing abnormal brain development. A substantial number of children with CP had congenital anomalies external to the central nervous system (CNS). Fewer than 10% of children with CP had evidence of intrapartum asphyxia. Intrauterine exposure to maternal infection (e.g., chorioamnionitis, inflammation of placental membranes, umbilical cord inflammation, foul-smelling amniotic fluid, maternal sepsis, temperature greater than 38°C during labor, and urinary tract infection) is associated with a significant increase in the risk of CP in normal birthweight infants. Another study found elevated levels of inflammatory cytokines in heelstick blood collected at birth from children who later were identified with CP. The prevalence of CP is increased among low birthweight infants, particularly those weighing less than 1,000 g at birth, primarily because of intracerebral hemorrhage and periventricular leukomalacia (PVL). Cramped synchronized general movements may be an early clinical finding in these infants. Although the incidence of intracerebral hemorrhage has declined significantly, PVL remains a major problem. PVL appears to reflect the enhanced vulnerability of immature oligodendroglia in premature infants to oxidative stress caused by ischemia or infectious/inflammatory insults. This information suggests that attempts to reduce the incidence of CP in term born infants should be directed toward increasing understanding of fetal developmental biology and that for premature infants strategies need to be developed to protect the vulnerable developing white matter.

Clinical Manifestations.

CP is generally divided into several major motor syndromes that differ according to the pattern of neurologic involvement, neuropathology, and etiology (Table 591-1). The physiologic classification identifies the major motor abnormality, whereas the topographic taxonomy indicates the involved extremities. CP is also commonly associated with a spectrum of developmental disabilities, including mental retardation, epilepsy, and visual, hearing, speech, cognitive, and behavioral abnormalities. The motor handicap may be the least of the child's problems.

Infants with spastic hemiplegia have decreased spontaneous movements on the affected side and show hand preference at a very early age. The arm is often more involved than the leg and difficulty in hand manipulation is obvious by 1 yr of age. Walking is usually delayed until 18-24 mo, and a circumductive gait is apparent. Examination of the extremities may show growth arrest, particularly in the hand and thumbnail, especially if the contralateral parietal lobe is abnormal, because extremity growth is influenced by this area of the brain. Spasticity is apparent in the affected extremities, particularly the ankle, causing an equinovarus deformity of the foot. An affected child often walks on tiptoes because of the increased tone, and the affected upper extremity assumes a dystonic posture when the child runs. Ankle clonus and a Babinski sign may be present, and weakness of the hand and foot dorsiflexors is evident. About one third of patients with spastic hemiplegia have a seizure disorder that usually develops during the first year or two, and approximately 25% have cognitive abnormalities including mental retardation. A CT scan or MRI study may show an atrophic cerebral hemisphere with a diluted lateral ventricle contralateral to the side of the affected extremities. An MRI is far more sensitive than CT for most lesions seen with CP, although a CT scan may be useful for detecting calcifications associated with congenital infections. Focal cerebral infarction secondary to intrauterine or perinatal thromboembolism related to thrombophilic disorders, especially anticardiolipin antibodies, is an important cause of hemiplegic CP. Family histories suggestive of thrombosis and inherited clotting disorders may be present and evaluation of the mother may provide information valuable for future pregnancies and other family members.
Table 591-1. Classification of Cerebral Palsy and Major Causes

<table>
<thead>
<tr>
<th>Motor Syndrome</th>
<th>Neuropathology</th>
<th>Major Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spastic Diplegia</td>
<td>Periventricular Leukomalacia (periventricular leukomalacic [PVL])</td>
<td>Prematurity Infarction Infection Endocrine/metabolic (e.g., thyroid)</td>
</tr>
<tr>
<td>Spastic quadriplegia</td>
<td>PVL Multicystic encephalomalacia Malformations</td>
<td>Ischemia Infection Endocrine/metabolic Genetic/developmental</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>Stoke: in utero or neonatal</td>
<td>Thrombophilic disorders Infection Genetic/developmental Periventricular hemorrhagic infarction</td>
</tr>
<tr>
<td>Extrapyramidal (athetoid, dyskinetic)</td>
<td>Basal ganglia Pathology: putamen, globus pallidus thalamus</td>
<td>Asphyxia Kernicterus Mitochondrial Genetic/metabolic</td>
</tr>
</tbody>
</table>

Spastic diplegia is bilateral spasticity of the legs greater than in the arms. The first indication of spastic diplegia is often noted when an affected infant begins to crawl. The child uses the arms in a normal reciprocal fashion but tends to drag the legs behind more as a rudder (commando crawl) rather than using the normal four-limbed crawling movement. If the spasticity is severe, application of a diaper is difficult because of the excessive adduction of the hips. Examination of the child reveals spasticity in the legs with brisk reflexes, ankle clonus, and a bilateral Babinski sign. When the child is suspended by the axillae, a scissoring posture of the lower extremities is maintained. Walking is significantly delayed, the feet are held in a position of equinovarus, and the child walks on tiptoes. Severe spastic diplegia is characterized by disuse atrophy and impaired growth of the lower extremities and by disproportionate growth with normal development of the upper torso. The prognosis for normal intellectual development is excellent for these patients, and the likelihood of seizures is minimal. The most common neuropathologic finding is periventricular leukomalacia, particularly in the area where fibers innervating the legs course through the internal capsule. MRI is very useful for evaluating the severity of white matter injury and for excluding other brain lesions.

Spastic quadriplegia is the most severe form of CP because of marked motor impairment of all extremities and the high association with mental retardation and seizures. Swallowing difficulties are common as a result of supranuclear bulbar palsies, often leading to aspiration pneumonia. The most common lesions seen on pathologic examination or on MRI scanning are severe PVL and multicystic cortical encephalomalacia. Neurologic examination shows increased tone and spasticity in all extremities, decreased spontaneous movements, brisk reflexes, and plantar extensor responses. Flexion contractures of the knees and elbows are often present by late childhood. Associated developmental disabilities, including speech and visual abnormalities, are particularly prevalent in this group of children. Children with spastic quadriparesis often have evidence of athetosis and may be classified as having mixed CP.

Athetoid CP, also called choreoathetoid or extrapyramidal CP, is less common than spastic cerebral palsy. Affected infants are characteristically hypotonic with poor head control and marked head lag and develop increased variable tone with rigidity and dystonia over several years. Feeding may be difficult, and tongue thrust and drooling may be prominent. Speech is typically affected because the oropharyngeal muscles are involved. Speech may be absent or sentences are slurred, and voice modulation is impaired. Generally, upper motor neuron signs are not present, seizures are uncommon, and intellect is preserved in many patients. This form of CP is also referred to as dyskinetic CP in Europe and is the type most likely to be associated with birth asphyxia. Extrapyramidal CP secondary to acute intrapartum near-total asphyxia is associated with bilaterally symmetric lesions in the posterior putamen and ventrolateral thalamus. These lesions appear to be the correlate of the neuropathologic lesion called status marmoratus in the basal ganglia. Athetoid CP can also be
caused by kernicterus secondary to high levels of bilirubin, and in this case the MRI scan shows lesions in the globus pallidus bilaterally. Extrapyramidal CP can also be associated with lesions in the basal ganglia and thalamus caused by metabolic genetic disorders such as mitochondrial disorders and glutaric aciduria. MRI scanning and possibly metabolic testing are important in the evaluation of children with extrapyramidal CP to make a correct diagnosis of etiology.

Diagnosis.
A thorough history and physical examination should preclude a progressive disorder of the CNS, including degenerative diseases, metabolic disorders, spinal cord tumor, or muscular dystrophy. The possibility of anomalies at the base of the skull or other disorders affecting the cervical spinal cord needs to be considered in patients with little involvement of the arms or cranial nerves. An MRI scan of the brain is generally indicated to determine the location and extent of structural lesions or associated congenital malformations, and an MRI scan of the spinal cord is worthwhile if there is any question about spinal cord pathology. Additional studies may include tests of hearing and visual function. Genetic evaluation should be considered in patients with congenital malformations or evidence of metabolic disorders. Because CP is usually associated with a wide spectrum of developmental disorders, a multidisciplinary approach is most helpful in the assessment and treatment of such children.

Treatment.
A team of physicians from various specialties, as well as occupational and physical therapists, speech pathologists, social workers, educators, and developmental psychologists provide important contributions to the treatment of these children. Parents should be taught how to handle their child in daily activities such as feeding, carrying, dressing, bathing, and playing in ways that limit the effects of abnormal muscle tone. They also need to be instructed in the supervision of a series of exercises designed to prevent the development of contractures, especially a tight Achilles tendon. There is no proof that physical or occupational therapy prevents development of CP in infants at risk or that it corrects the neurologic deficit, but ample evidence shows that therapy optimizes the development of an abnormal child.

Children with spastic diplegia are treated initially with the assistance of adaptive equipment, such as walkers, poles, and standing frames. If a patient has marked spasticity of the lower extremities or evidence of hip dislocation, consideration should be given to performing surgical soft tissue procedures that reduce muscle spasm around the hip girdle, including an adductor tenotomy or psoas transfer and release. A rhizotomy procedure in which the roots of the spinal nerves are divided has produced considerable improvement in selected patients with severe spastic diplegia. A tight heel cord in a child with spastic hemiplegia may be treated surgically by tenotomy of the Achilles tendon. Quadriplegia is managed with motorized wheelchairs, special feeding devices, modified typewriters, and customized seating arrangements.

Communication skills may be enhanced by the use of Bliss symbols, talking typewriters, and specially adapted computers including artificial intelligence computers to augment motor and language function. Significant behavior problems may substantially interfere with the development of a child with CP; their early identification and management are important, and the assistance of a psychologist or psychiatrist may be necessary. Learning and attention deficit disorders and mental retardation are assessed and managed by a psychologist and educator. Strabismus, nystagmus, and optic atrophy are common in children with CP; thus, an ophthalmologist should be included in the initial assessment. Lower urinary tract dysfunction should receive prompt assessment and treatment.

Several drugs have been used to treat spasticity, including dantrolene sodium, the benzodiazepines, and baclofen. These medications are generally ineffective but should be considered if severe spasticity is not controlled by other measures. Intrathecal baclofen has been used successfully in selected children with severe spasticity. This experimental therapy requires a team approach and constant follow-up for complications of the infusion pumping mechanism and infection. Botulinum toxin is undergoing study for the management of spasticity in specific muscle groups, and the preliminary findings show a positive response in those patients studied. Patients with rigidity, dystonia, and spastic quadriplegia sometimes respond to levodopa, and children with dystonia may benefit from carbamazepine or trihexyphenidyl. Hyperbaric oxygen does not improve the condition of children with CP.