

# Cerebral Palsy

Cerebral palsy (CP) is defined as a nonprogressive disorder of posture and movement, often associated with epilepsy and abnormalities of speech, vision, and intellect resulting from a defect or lesion of the developing brain. CP is a common disorder, with an estimated prevalence of 2/1,000 population.

**EPIDEMIOLOGY AND ETIOLOGY.** The reported prevalence rate of CP is 4/1,000 live births. Birth asphyxia was an uncommon cause of CP; moreover, most high-risk pregnancies resulted in neurologically normal children. Although a cause for CP could not be identified in most cases, a substantial number of children with CP had congenital anomalies external to the central nervous system (CNS), which may have placed them at increased risk for developing asphyxia during the perinatal period. An Australian study comparing children with spastic CP with a group of matched controls had similar findings. Less than 10% of children with CP had evidence of intrapartum asphyxia. Although the increased survival of premature infants from improved perinatal care has resulted in more children with CP, the rate did not increase. These studies suggest that future developments aimed at enhancing perinatal care will have minimal impact on the incidence of CP and that research might be directed more profitably to the field of developmental biology in order to understand the pathogenesis of CP.

**CLINICAL MANIFESTATIONS.** CP may be classified by a description of the motor handicap in terms of physiologic, topographic, and etiologic categories and functional capacity. 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. The 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; the deep tendon reflexes are increased; 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 computed tomography (CT) scan or magnetic resonance imaging (MRI) may show an atrophic cerebral hemisphere with a dilated lateral ventricle contralateral to the side of the affected extremities. Intrauterine thromboembolism with focal cerebral infarction may be one etiology; CT or MRI at birth in infants with focal seizures often demonstrates the area of infarction.

Spastic diplegia refers to bilateral spasticity of the legs. The first indication of spastic diplegia is often noted when the 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-stance crawling movement. If the spasticity is severe, the application of a diaper is difficult owing to 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. This lesion is noted among premature infants.

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 owing to supranuclear bulbar palsies and often lead to aspiration pneumonia. At autopsy, the central white matter is disrupted by areas of necrotic degeneration that may coalesce into cystic cavities. 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 quadriplegia often have evidence of athetosis and may be classified as mixed CP.

Athetoid CP is relatively rare, especially since the advent of aggressive management of hyperbilirubinemia and the prevention of kernicterus. These infants are characteristically hypotonic and have poor head control and marked head lag. Feeding may be difficult, and tongue thrust and drooling may be prominent. The athetoid movements may not become evident until 1 yr of age and tend to coincide with hypermyelination of the basal ganglia, a phenomenon called status marmoratus. Speech is typically affected owing to involvement of the oropharyngeal muscles. Sentences are slurred, and voice modulation is impaired. Generally, upper motor neuron signs are not present, seizures are uncommon, and intellect is preserved in most patients.

**DIAGNOSIS.** A thorough history and physical examination should eliminate a progressive disorder of the CNS, including degenerative diseases, spinal cord tumor, or muscular dystrophy. Depending on the severity and the nature of the neurologic abnormalities, a baseline electroencephalogram (EEG) and CT scan may be indicated to determine the location and extent of structural lesions or associated congenital malformations. Additional studies may include tests of hearing and visual function. As CP is usually associated with a wide spectrum of developmental disorders, a multidisciplinary approach is most helpful in the assessment and management of such children.

**TREATMENT.** Parents should be taught how to handle their child in daily activities such as feeding, carrying, dressing, bathing, and playing in ways that will 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 will prevent the development of CP in the infant at risk or that it will correct the neurologic deficit, but there is ample evidence that therapy optimizes the development of the abnormal child. The child with spastic diplegia is treated initially with the assistance of adaptive equipment, such as walkers, poles, and standing frames. If the patient has marked spasticity of the lower extremities or if there is 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. The quadriplegic patient 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 the 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 utilized 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. Occasionally, patients with incapacitating athetosis will respond to levodopa, and children with dystonia may benefit from carbamazepine or trihexyphenidyl.