CEREBRAL PALSY

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CEREBRAL PALSY

Cerebral palsy (CP) can be defined as a central motor dysfunction affecting muscle tone, posture, and movement that is attributed to nonprogressive disturbances in the developing fetal or infant brain.

Despite being described as a nonprogressive disorder, the clinical expression of brain injury or insult changes over time. Therefore, the condition should be viewed as a dynamic disorder that evolves because of factors such as growth, nervous system maturation, and aging.

EPIDEMIOLOGY AND ETIOLOGY

Cerebral palsy is the most common neuromotor disorder in childhood, with an overall incidence of 2.6- 2.9 cases per 1,000 live births in the United States. Within developed countries, studies estimate the prevalence of CP as nearly 1- 4 per 1,000 live births. In developing countries, available estimates of prevalence are similar.

CP is caused by a broad group of developmental, genetic, metabolic, ischemic, infectious, and other etiologies that produce a common group of neurologic phenotypes. Thus, CP should be based on phenotype rather than etiology.

The prevalence of CP is higher for children born preterm or at a low birthweight, though this is influenced by sex, ethnicity, and socioeconomic status. There is a 30–40% greater prevalence in males. Prevalence is higher in low- and middle-income versus high-income communities.

In addition to prematurity and birthweight, numerous other prenatal and perinatal risk factors have been reported. These risk factors include antenatal infection (chorioamnionitis, urinary tract infection), multiple pregnancy, and neonatal infection. Infertility treatments are also associated with a higher rate of CP, probably because these treatments are often associated with multiple pregnancies. CP is most often multifactorial, and multiple risk factors coexist. The cerebral disruption associated with CP can occur prenatally, perinatally, or postnatally in the first 2 years of life, given that brain development is ongoing during this critical period.

Congenital CP (due to cerebral injury/maldevelopment before birth) accounts for 85–90% of total cases, whereas acquired CP (due to cerebral injury after 1 month of life) is responsible for the remaining cases.

The major lesions that contribute to CP in preterm infants are intracerebral hemorrhage and periventricular leukomalacia (PVL). Although the incidence of intracerebral hemorrhage has declined significantly, PVL remains a major problem. White matter abnormalities (loss of volume of periventricular white matter, extent of cystic changes, ventricular dilation, thinning of the corpus callosum) present on MRI at 40 weeks of gestational age in former preterm infants are a predictor of later CP.

In term births, causes historically have primarily been thought to be by events during labor and delivery, causing hypoxia. The mechanisms are predominantly the result of cerebral ischemia and excitotoxicity. The cause can be obvious (i.e., placental abruption, meconium aspiration), though at other times the etiology can be difficult to pinpoint.

Risk factors can include eclampsia, hypercoagulability, and placental pathology. For some, no predisposing clinical factors are identified.

Hypoxic-ischemic encephalopathy (HIE) may be decreasing as an apparent cause of CP in developed countries. Therapeutic hypothermia may reduce the risk of CP in term patients with HIE.

There are several causes of acquired CP. The most common cause in this category is perinatal stroke, which can be ischemic, hemorrhagic, or thromboembolic. The second most common cause is meningitis or encephalitis during infancy. Kernicterus is a rare cause of CP in developed countries, though cases (particularly in very preterm infants) persist.

Cryptogenic CP traditionally refers to an individual in whom no clear perinatal etiology has been identified and accounts for ~30% of cases. Chromosomal disorders have been identified in ~30% of patients with CP.

CLINICAL MANIFESTATIONS

There are several classification systems to describe CP. One such classification system starts by determining the type of motor involvement: spastic or extrapyramidal. Spastic CP can then further truncated topographically, whereas extrapyramidal is further categorized based on the type of involuntary movement seen.

In extrapyramidal CP, the brain injury or insult spares the pyramidal tracts that cause spasticity, resulting in disorders of movement, coordination, and balance. Clinically, these patients exhibit dystonia and/or choreoathetosis (collectively referred to as dyskinetic) or ataxia associated with lesions in the cerebellum or its connections.

Spastic CP accounts for 80% of cases, whereas extrapyramidal makes up 20% of cases (15% dyskinetic and 5% ataxic).

- **Spastic hemiplegia** has decreased spontaneous movements on the affected side and shows hand (handedness) preference at a very early age. The arm is often more involved than the leg, and difficulty in hand manipulation is evident by 1 year of age. Walking is usually delayed until 18- 24 months, and a circumductive gait is apparent.
  Examination of the extremities may show growth arrest leading to shortened limbs and decreased muscle bulk on the affected side.
  An affected child often walks on tiptoe (toe-walking) because of the increased tone in the antigravity gastrocnemius muscles and tight, contracted Achilles tendon; the affected upper extremity assumes a flexed posture when the child runs.
  Ankle clonus and a Babinski sign may be present, the deep tendon reflexes are increased.

- **Spastic monoplegia** is when only one limb is affected and may not be as obvious as other types of CP. Depending on which limb is affected, the child's motor disability ranges from challenges with either fine or gross motor skills. A monoplegia that affects the arm may result in challenges with bimanual tasks, whereas when the legs are involved, toe walking may be seen.

- **Spastic diplegia** is bilateral spasticity of the legs that is greater than in the arms. Spastic diplegia is strongly associated with injury to the immature white matter during the vulnerable period between 20 and 34 weeks of gestation, hence seen in those born prematurely. The first clinical indication of spastic diplegia is often noted when an affected infant begins to crawl. The child uses the arms in a

normal 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 symmetric spasticity in the legs with brisk reflexes, ankle clonus, and a bilateral Babinski sign.
When the child is suspended by the axillae, an extended scissoring posture of the lower extremities is maintained. Walking can be 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, impaired growth of the lower extremities, and disproportionate growth with normal development of the upper torso.

- **Spastic quadriplegia** is the most severe form of CP because of marked motor impairment of all extremities and the high association with other comorbidities, including intellectual disabilities, seizure disorders, communication, and visual impairment, and feeding difficulties. Swallowing difficulties are common as a result of supranuclear bulbar palsies, often leading to aspiration pneumonia and growth failure.
  Neurologic examination shows increased tone and spasticity in all extremities, decreased spontaneous movements, brisk reflexes, and plantar extensor responses. Flexion contractures of the knees, elbows, and wrists are often present by late childhood. Children with spastic quadriparesis can also have extrapyramidal findings given the diffuse involvement of the brain injury.

Extrapyramidal CP can be divided into the two main types of involuntary movement seen: ataxia and dyskinesias. In this type of CP, injury is typically to the subcortical areas, which are centers for coordination in movement and balance. Injury may not produce weakness, but rather the inability to voluntarily control movements. This type is less common than spastic CP and makes up approximately 15–20% of patients with CP.

- **Ataxic CP** is the rarest form, whose clinical picture is variable, ranging from hypotonia to mild spasticity in addition to incoordination, depending on the other systems involved. Walking gait is often very wide and sometimes irregular. Control of eye movements and depth perception can be impaired. Often, fine motor skills requiring coordination of the eyes and hands, such as writing, are difficult.
- **Dyskinetic CP** is further divided into two groups: athetoid and dystonic. Athetoid CP includes cases with involuntary movement, especially in the arms, legs, and hands. With the current management of Rh and ABO incompatibility, the incidence of CP characterized by athetosis has markedly decreased. Athetoid CP is often caused by kernicterus secondary to high levels of bilirubin, and in this case, the MRI scan shows lesions in the globus pallidus bilaterally, or it may be normal. Cases are still seen as a result of HIE as well. The affected infant is initially hypotonic, but a tendency toward arching and opisthotonos (dystonia) is noted, and obligatory tonic neck reflexes are present.
- **Dystonic CP** : encompasses cases that affect the trunk muscles more than the limbs and result in a fixed, twisted posture. Toward the end of the first year, repetitive, involuntary movements become more consistent with the full-blown picture of dystonic CP. Dystonia in CP presents as hypertonia, involuntary postures and movements, or a combination.

Table 638.1 Classification of Cerebral Palsy and Major Causes

MOTOR SYNDROME (APPROX % OF CP)	NEUROPATHOLOGY/MRI	MAJOR CAUSES
Spastic diplegia (35%)	Periventricula leukomalacia Periventricular cysts or scars in white matter, enlargement of ventricles, squared- off posterior ventricles	Prematurity Ischemia Infection Endocrine/metabolic(e.g., thyroid)
Spastic quadriplegia (20%)	Periventricular leukomalacia Multicystic encephalomalacia Cortical malformations	Ischemia, infection Endocrine/metabolic, genetic/developmental
Hemiplegia (25%)	Stroke: in utero or neonatal Focal infarct or cortical, subcortical damage Cortical malformations	Thrombophilic disorders Infection Genetic/developmental Periventricular hemorrhagic infarction
Extrapyramidal (athetoid, dyskinetic) (15%)	Asphyxia: symmetric scars in putamen and thalamus Kernicterus: scars in globus pallidus,hippocampus Mitochondrial: scarring of globus pallidus,caudate, putamen, brainstem No lesions: ? dopa- responsive dystonia	Hypoxia Kernicterus Mitochondrial Genetic/metabolic

comorbidities

Nearly all individuals with CP will have one or more medical, neurologic, or psychiatric comorbidities. Neuropsychiatric comorbidities that occur at higher rates with CP include intellectual disability, anxiety, depression, and attention-deficit/hyperactivity disorder (ADHD) compared with the general population. Other associated comorbidities are common and include chronic pain, sleep difficulties, urinary dysfunction, decreased bone health, sialorrhea, respiratory disorders, feeding and growth challenges, gastrointestinal disorders, including constipation and dysmotility, speech/communication difficulties, visual impairment, and hearing loss.

DIAGNOSIS

i. CP is a clinical diagnosis that considers elements from the history, physical examination, and neuroimaging. The history should focus on whether there was perinatal or postnatal injury to the brain, what factors led to this injury, and contributory pregnancy factors. Another important factor is the developmental trajectory. It is important to quantify rates of development in the various domains and whether that trajectory has been one of continued acquisition of skills, plateauing, or regression.

❖ No loss of acquired milestones (regression)—helps preclude a progressive disorder of the central nervous system (CNS), including degenerative diseases, metabolic disorders

ii. The examinations are necessary to characterize abnormalities of tone, reflexes, movements, posture, and balance that may be consistent with the diagnosis of CP.

iii. An MRI scan of the brain is indicated to determine the location and extent of structural lesions or associated congenital malformation.

iv. Additional studies may include tests of hearing and visual function.

v. Genetic evaluation should be considered in patients with congenital malformations, evidence of metabolic disorders (e.g., amino acids, organic acids, MR spectroscopy), clinical features atypical of CP, or when there are no perinatal risk factors, especially in term infants.

TREATMENT

The treatment strategy for CP is best developed in a multidisciplinary team (physiatry, orthopedics, neurology/neurodevelopmentalists, neurosurgery) as well as physical and occupational therapy, speech pathology, social work, primary care physicians, developmental pediatricians, and educators maximize the chances of success.

➢ Rehabilitative strategies include orthotics, casting, and physiotherapy.

➢ Pharmacotherapy :

- Baclofen is routinely favored; other antispasticity medications such as tizanidine, dantrolene, and benzodiazepines are also available.
- Second-line medications such as clonidine or gabapentin.
- Medications used to treat dystonia include enteral baclofen, benzodiazepines, trihexyphenidyl, clonidine, dantrolene, levodopamine, and gabapentin, but practice varies widely.
- Tetrabenazine can be useful for hyperkinetic movement disorders, including athetosis or chorea.

➢ The management of focal/segmental spasticity or dystonia includes :Targeted injections botulinum toxin A

➢ Neurosurgical options include intrathecal baclofen (ITB), selective dorsal rhizotomy (SDR), and deep brain stimulation (DBS).