Cerebral Palsy

4.0 Contact Hours

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Cerebral Palsy

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Cerebral palsy is a non-progressive neuromuscular disorder. It is used to describe a group of motor syndromes resulting from disorders of early brain development. It is often associated with epilepsy and abnormalities of speech, vision and intellect. It is difficult to estimate the precise magnitude of the problem since milder cases are likely to be missed. Cerebral palsy is classified physiologically, topographically, etiologically and functionally.

Because brain development continues during the first two years of life, cerebral palsy can result from brain injury occurring during the prenatal, perinatal, or postnatal periods. The history, combined with a neurologic examination establishing that motor deficit is due to a cerebral abnormality, establishes the diagnosis of cerebral palsy. Because of the heterogeneous nature of cerebral palsy, it is difficult to make generalized statements regarding treatment, and it is best to have an individualized approach to each patient and his or her needs.

Upon completion of this course, the learner will be able to:

1. Define cerebral palsy
2. Discuss the epidemiology of cerebral palsy
3. Discuss the various classifications of cerebral palsy
4. Discuss the clinical manifestations of various types of cerebral palsy
5. Elaborate ON the various causes of cerebral palsy
6. Discuss the management of cerebral palsy

Definition

Cerebral palsy is defined as a non-progressive neuromuscular disorder of cerebral origin. It is 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.

Magnitude of the problem

Cerebral palsy is the most common cause of chronic motor disability that begins in childhood. It is difficult to estimate the precise magnitude of the problem since milder cases are likely to be missed. It is the most common childhood physical disability and affects 2 to 2.5 children per 1,000 born in the United States.

Types of cerebral palsy

Cerebral palsy is classified physiologically, topographically, etiologically and functionally.

Physiological classification

This is determined on the basis of the nature of the movement disorder, muscle tone, and anatomic distribution. A single patient may have more than one type. Spastic CP is the most common, accounting for about two thirds of cases.

1. Pyramidal (or spastic) CP**:** This type is characterized by neurologic signs of upper motor neuron damage with increased "clasp knife" muscle tone, increased deep

tendon reflexes, pathologic reflexes, and spastic weakness. Spastic CP is subclassified on the basis of distribution:

- Hemiparesis: Primarily unilateral involvement, with the arm usually moving more than the leg
- Quadriparesis: All limbs are involved, with the legs often more involved than the arms
- Diparesis: Legs are much more involved than the arms, which may show no or only minimal impairment

2. Extrapyramidal (nonspastic or dyskinetic) CP**:** This type is characterized by prominent involuntary movements or fluctuating muscle tone, with choreoathetosis as the most common subtype. Distribution is usually symmetric among the four limbs.
3. Hypotonic CP**:** This type manifests as generalized muscle hypotonia that persists with normal or increased deep tendon reflexes. Many patients with this condition develop cerebellar deficits of incoordination and ataxia, and about one third of patients have severe retardation.
4. Ataxic CP**:** Primarily cerebellar signs are seen.
5. Mixed types

<u>Topographical classification</u>

This is based on which limbs are involved.

- Monoplegia – Only one limb is involved.
- Paraplegia – Both the legs are involved
- Hemiplegia – Both upper and lower limbs are involved on one side
- Triplegia – Three limbs are involved
- Quadriplegia – all the four limbs are involved
- Diplegia – The lower limbs are more involved than the upper limbs.

Etiologic classification

This classification is based on the underlying cause of cerebral palsy-

- Prenatal causes – Infection, metabolic disorders, anoxia, toxins, genetic disorder and infarction
- Perinatal causes – Anoxia
- Postnatal causes – Toxins, trauma and infection

Functional classification

Functional classification is based on the degree of disability in the cerebral palsy infants –

Class I – No limitation of activity

Class II – Slight to moderate limitation

Class III- Moderate to great limitation

Class IV – No useful physical activity

Causes

Cerebral palsy is a static neurologic condition resulting from brain injury that occurs before cerebral development is complete. Because brain development continues during the first two years of life, cerebral palsy can result from brain injury occurring during the prenatal, perinatal, or postnatal periods.

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).

In contrast with popular perception, large clinical epidemiologic and longitudinal studies indicate that perinatal asphyxia is an important-but relatively minor-cause.

Estimates range from a low of 3% to a high of 21%. In most cases, the events leading to CP occur in the fetus before the onset of labor or in the newborn after delivery.

In a large study of 49,000 infants, a low Apgar score correlated poorly with the development of CP. Of term infants with scores of 0-3 at 1 or 5 minutes, 95% did not develop CP. Of those with scores of 0-3 at 10 minutes, 84% did not develop CP. If the 10-minute Apgar score improved to 4 or more, the rate for CP was <1%. A low Apgar score (0-3) at 20 minutes, however, had an observed CP rate of nearly 60%. Conversely, nearly 75% of patients with CP had 5-minute Apgar scores of 7-10.

Prenatal causes

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. Neonatal risk factors for cerebral palsy include birth after fewer than 32 weeks' gestation, birth weight of less than 5 1b, 8 oz (2,500 g), intrauterine growth retardation, intracranial hemorrhage, and trauma.

Perinatal causes

Although cerebral palsy is rarely diagnosed in the fetus or newborn infant, the etiology is nearly always from a perinatal or neonatal cause. The contributions of various perinatal causes are –

- Prematurity and intrauterine growth rate restriction: 40% to 50%
- Birth asphyxia or birth trauma: 25% to 30%
- Neonatal stroke: 5% to 10%
- Toxoplasmosis, rubella, cytomegalovirus, herpes simplex, other infections, 5% to 10%

- Chromosomal abnormalities: 5% to 10%
- Inborn errors of metabolism: 5% to 10%
- Other known causes: 5% to 10% (neonatal sepsis/meningitis, kernicterus, hypoglycemia, environmental toxins, drug and alcohol exposure, maternal thyroid disease, postnatal infections and trauma, and others)
- Idiopathic: 5% to 10%

Postnatal causes

In about 10 to 20 percent of patients, cerebral palsy is acquired postnatally, mainly because of brain damage from bacterial meningitis, viral encephalitis, hyperbilirubinemia, motor vehicle collisions, falls, or child abuse.

Causes of cerebral palsy based on the type

1. Spastic cerebral palsy – Prematurity, ischemia, infection, endocrine causes
2. Spastic quadriplegia - Ischemia, infection
3. Hemiplegia - Thrombophilic disorders, Infection, Genetic/developmental, Periventricular hemorrhagic infarction
4. Extrapyramidal (athetoid, dyskinetic)- Asphyxia, Kernicterus

Clinical features

The clinical features vary depending on the type of cerebral palsy. The various types of CP and the clinical features are given below-

Spastic diplegia

Diplegia is the most common anatomical type of cerebral palsy, constituting approximately 50% of all cases. Patients with diplegia have motor abnormalities in all four extremities, with the lower extremities more affected than the upper. The close

proximity of the lower extremity tracts to the ventricles most likely explains the more frequent involvement of the lower extremities with periventricular lesions.

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.

This type of cerebral palsy is most common in premature infants; intelligence usually is normal. Most children with diplegia walk eventually, although walking is delayed usually until around age 4 years.

Spastic hemiplegia

In hemiplegia, one side of the body is involved, with the upper extremity usually more affected than the lower extremity. Patients with hemiplegia, approximately 30% of patients with cerebral palsy, typically have sensory changes in the affected extremities as well.

This type of CP occurs due to periventricular leukomalacia in neonates. 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 month, and a circumductive gait is apparent. Examination of the extremities may show growth arrest, particularly in the hand and thumbnail.

Severe sensory changes, especially in the upper extremity, are a predictor of poor functional outcome after reconstructive surgery. Hemiplegic patients also may have a leg-length discrepancy, with shortening on the affected side.

Spasticity is apparent in the affected extremities, particularly the ankle, causing an equinovarus deformity of the foot. An affected child often walks on tiptoe because of the increased tone, and the affected upper extremity assumes a dystonic posture when the child runs.

About one third of patients with spastic hemiplegia have a seizure disorder that usually develops in the 1st year. Approximately 25% have cognitive abnormalities including mental retardation.

Spastic quadriplegia

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. In quadriplegia, all four extremities are equally involved, and many patients have significant cognitive deficiencies that make care more difficult. Head and neck control are usually present, which helps with communication, education, and seating.

Athetoid CP

Athetoid CP is also called choreoathetoid or extrapyramidal CP. Athetoid cerebral palsy is less common than spastic cerebral palsy. This type is most likely to be associated with birth asphyxia**.** It is caused by an injury to the extrapyramidal tracts and is characterized by dyskinetic, purposeless movements that may be exacerbated by environmental stimulation. The clinical picture varies based on the level of excitement of the patient. In pure athetoid cerebral palsy, joint contractures are uncommon.

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. Seizures are uncommon, and intellect is preserved in many patients.

Diagnosis of Cerebral palsy

Diagnostic criteria for Cerebral palsy

Cerebral palsy can be diagnosed using Levine (POSTER) criteria. POSTER stands for –

P - **P**osturing/abnormal movements

O - **O**ropharyngeal problems (e.g., tongue thrusts, swallowing abnormalities)

S - **S**trabismus

T - **T**one (hyper- or hypotonia)

E - **E**volutional maldevelopment (primitive reflexes persist or protective equilibrium reflexes fail to develop

R - **R**eflexes (increased deep tendon reflexes/persistent Babinski's reflex)

Abnormalities in four of these six categories strongly point to the diagnosis of cerebral palsy.

In a child with cerebral palsy motor deficit is always present. The usual presenting complaint is that child is not reaching motor milestones at the appropriate age. Medical history establishes that the child is not losing function. This history, combined with a neurologic examination establishing that motor deficit is due to a cerebral abnormality, establishes the diagnosis of cerebral palsy. Serial examinations may be necessary if the history is unreliable. Other causes of neonatal hypotonia include muscular dystrophies, spinal muscular atrophy, Down's syndrome, spinal cord injuries.

Laboratory tests are not necessary to establish the diagnosis. Workup is helpful for assessment of recurrence risk, implementation of prevention programs, and medico legal purposes. Metabolic and genetic testing should be considered if on follow-up the child has

- Evidence of deterioration or episodes of metabolic decompensation
- No etiology determined by neuroimaging
- Family history of childhood neurologic disorder associated with cerebral palsy
- Developmental malformation on neuroimaging

An EEG should be obtained when a child with cerebral palsy has a history suggestive of seizures. Children with cerebral palsy should be screened for ophthalmologic and hearing impairments, speech and language disorders. Nutrition, growth, and swallowing function should be monitored. Neuroimaging is recommended if the etiology has not been established previously; for example, by perinatal imaging. MRI, when available, is preferred to CT scanning because of higher yield in suggesting an etiology, and timing of the insult leading to cerebral palsy.

<u>Early identification of infants at risk for developmental disabilities</u>

Early recognition of infants at risk for development disability is important. The early identification of a developmental delay is a challenge to all physicians, however. It is difficult to differentiate between infants who are lagging behind in skill acquisition but will achieve the usual developmental milestones and infants who are truly deviating from

the expected pattern. Identifying children in the first year of life provides the opportunity for early referral for interventional services and diagnosis.

Risk category for developmental delay by medical diagnosis -

High risk

- Birth weight less than 1250 g
- 30 weeks gestation or less
- Intraventricular hemorrhage/periventricular leukomalacia
- Severe perinatal asphyxia
- Severe neurologic problems
- Bronchopulmonary dysplasia that requires home oxygen
- Complex congenital/cyanotic heart disease
- Abnormal neurologic examination at discharge
- Significant feeding problems/requirement of gavage feeding
- Intracranial pathology: congenital or acquired
- Extracorporeal membrane oxygenation
- Diaphragmatic hernia
- Persistent pulmonary hypertension of the newborn/required inhaled nitric oxide /oscillatory ventilator
- Significant circulatory failure
- Congenital viral infection (HIV, TORCH)
- Prolonged or persistent hypoglycemia
- Multiple/major congenital anomalies and genetic disorders

Moderate risk

- Birth weight between 1250 and 1500 g
- Prolonged ventilation and high-frequency ventilation
- Surgical: cloacal anomalies/gastroschisis/omphalocele
- Tracheostomy
- Metabolic disorders

Early Markers of Cerebral Palsy

The high risk newborn babies should be followed up for early identification of neuromotor disability so that appropriate stimulation therapy can be initiated to enhance neuromotor development. The following clinical markers should be looked for to make an early diagnosis of cerebral palsy –

1. Episodes of inconsolable crying, chewing movements, lip smacking, excessive sensitivity to light and noise, spontaneous Moro response
2. Persistent neck tonic posturing beyond 4 weeks
3. Clenched fists with thumb adducted and flexed across the palm beyond 8 weeks
4. Paucity or absence of fidgety limb movements during 6-12 weeks
5. Abnormalities of tone(usually hypertonia but occasionally hypotonia) as assessed by scarf sign and various angles
6. Persistence of automatic reflexes beyond 4-5 months (Moro reflex, grasp reflex, asymmetric tonic neck reflex)
7. Persistent asymmetry of posture, tone, movements and reflexes
8. Slow head growth

The following table gives ages at appearance and disappearance of common developmental reflexes. Absence of parachute response and Landau reflex and persistence of other automatic reflexes beyond the ages mentioned in the table are indicative of cerebral palsy.

Reflex	Age of appearance	Age of disappearance
Rooting	Birth	3 months
Moro	Birth	5-6 months
Palmar grasp	Birth	6 months
Tonic neck	Birth	9-10 months
Adductor spread of knee jerk	Birth	7-8 months
Landau	10 months	24 months
Parachute	8-9 months	Persists in normal children

A child's development should follow a predictable pattern. Reviewing a child's history of acquisition of developmental milestones offers a way to detect deviations from normal. A developmental history is usually organized by domains of development. Areas to be included are gross motor skills, fine motor skills, social interaction, language, and self-help. By eliciting a developmental history at each visit, the pediatrician has an opportunity to create a supportive relationship with the parents and educate the family on normal development of their child.

Treatment

Because of the heterogeneous nature of cerebral palsy, it is difficult to make generalized statements regarding treatment, and it is best to have an individualized approach to each patient and his or her needs. In some centers, a multidisciplinary team approach—including physical, occupational, and speech therapy; orthotics, nutrition; social work, orthopedics and general pediatrics—has been successful. Four basic treatment principles exist.

- The first is that although the central nervous system injury, by definition, is nonprogressive, the deformities caused by abnormal muscle forces and contractures are progressive.
- The second, which can be a source of frustration, is that the treatments currently available correct the secondary deformities only and not the primary problem, which is the brain injury.
- The third is that the deformities typically become worse during times of rapid growth. For some patients, it may be beneficial to delay surgery until after a significant growth spurt to decrease the risk of recurrence.
- The fourth is that operative or nonoperative treatment should be done to minimize the impact it has on the patient's socialization and education.

It is important to be aware of these timing issues when considering any form of treatment in this patient population. It also is important to recognize that for most patients a combined approach using nonoperative and operative methods is more beneficial rather than one form of treatment alone.

Nonoperative Treatment

Nonoperative modalities, such as medication, splinting and bracing, and physical therapy, commonly are used as primary treatment or in conjunction with other forms of treatment such as surgery. A wide variety of medications have been used to treat cerebral palsy. The three most common agents are diazepam and baclofen, which act centrally, and dantrolene, which acts at the level of skeletal muscle. Because these drugs increase inhibitory neurotransmitter activity, common systemic side effects include sedation, balance difficulties, and cognitive dysfunction, which can have a dramatic detrimental effect on ambulation, education, and communication.

Botulinum toxin type A (Botox) has been used to weaken muscles selectively in patients with cerebral palsy. It also is safer than these other agents because it binds selectively to the neuromuscular junction and not to other surrounding tissues. This effect begins approximately 24 hours after injection and lasts 2 to 6 months. The maximal safe dose of BTX-A based on primate data is 36 to 50 U/kg body weight; however, most studies report doses of less than 20 U/kg body weight. BTX-A has been shown to be effective when used in conjunction with other modalities, such as physical therapy or serial casting. The most common side effects are local pain and irritation from the injection.

Physical therapy is an essential component in the treatment of patients with cerebral palsy. Physical therapy typically is used as a primary treatment modality and in conjunction with other modalities, such as casting, bracing, BTX-A, and surgery.

Bracing, as with physical therapy and medication, typically is used in conjunction with other modalities. Bracing in patients with cerebral palsy most commonly is used to prevent or slow progression of deformity. The most commonly used braces for the treatment of cerebral palsy include ankle-foot orthoses, hip abduction braces, hand and wrist splints, and spinal braces or jackets.

Operative Treatment

Operative treatment typically is indicated when contractures or deformities decrease function, cause pain, or interfere with activities of daily living. It is the only effective treatment when significant fixed contractures exist. Surgical procedures should be

scheduled to minimize the number of hospitalizations and interference with school and social activities. “Birthday surgery” or multiple procedures performed at different times, should be avoided whenever possible.

Operative treatment of deformities related to cerebral palsy can be divided into several groups, including procedures to

(1) Correct static or dynamic deformity

(2) Balance muscle power across a joint

(3) Reduce spasticity (neurectomy)

(4) Stabilize uncontrollable joints. Often, procedures can be combined

Neurosurgical Intervention

Selective dorsal root rhizotomy is a technique to reduce spasticity and balance muscle tone in carefully selected patients. In patients with cerebral palsy, the normal central nervous system inhibitory control of the gamma efferent system is deficient, leading to the exaggerated stretch reflex response. The goal of selective dorsal rhizotomy is to identify the rootlets carrying excessive stimulatory information and section them to reduce the stimulatory input from the dorsal sensory fibers

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