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PAEDIATRICS NEUROLOGY

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Cerebral palsy (CP)

Cerebral palsy (CP) is a term used to describe a group of permanent disorders of movement and posture causing activity limitation, that are attributed to non-progressive disturbances in the in the developing fetal or infant brain.. A *static encephalopathy*, a term previously used, is now inaccurate because some of the neurologic features of CP, such as movement disorders and orthopedic complications, including scoliosis and hip dislocation, can change or progress over time. CP is often associated with epilepsy and abnormalities of speech, vision, and intellect, sensation as well as secondary musculoskeletal problems. Many children and adults with CP function at a high educational and vocational level, without any sign of cognitive dysfunction.

EPIDEMIOLOGY AND ETIOLOGY

CP is the most common form of chronic motor disability with a prevalence of 3.6 per 1,000 children with a male : female ratio of 1.4 : 1. In 80% of cases, features were identified pointing to antenatal factors causing abnormal brain development. Fewer than 10% of children with CP had evidence of intrapartum asphyxia. The prevalence of CP is increased among preterm infants, primarily because of intracerebral hemorrhage and periventricular leukomalacia (PVL). Intrauterine exposure to maternal infection (chorioamnionitis, urinary tract infection) was associated with a significant increase in the risk of CP in normal birth weight infants.

Classification of Cerebral Palsy and Major Causes

Physiologic	Topographic	Etiologic	Function
Spastic Athetoid Rigid Ataxic Tremor Atonic Mixed	Monoplegia Paraplegia Hemiplegia Triplegia Quadriplegia Diplegia Double hemiplegia	**Prenatal:** (e.g. infection, met-metabolic, anoxia, toxic, genetic, infarction). **Perinatal:** (e.g. Asphyxia) **Postnatal:** (toxin, trauma, infection)	**Class 1:** no limitation of activity. **Class 2:** slight to moderate limitation **Class 3:** moderate to great limitation **Class 4:** no useful physical activity.

Spastic Hemiplegia: the patient has 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. Walking is usually delayed until 18–24 mo, and a circumductive gait is apparent. Examination of the extremities may show growth arrest, 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 tiptoe because of the increased tone. Ankle clonus and

Babinski sign may be present; the deep tendon reflexes are increased.

About one third of patients with spastic hemiplegia have a seizure disorder; ≈25% has cognitive abnormalities including mental retardation. A CT scan or MRI study may show an atrophic cerebral hemisphere with a dilated lateral ventricle contralateral to the side of the affected extremities. Primary causes are cerebral infarction due to intrauterine or perinatal thrombo-embolism, like presence of anticardiolipin antibodies and inherited clotting disorders, such as factor V Leiden mutation), & brain malformation.

Spastic Diplegia is bilateral spasticity of the legs. It is strongly associated with damage to the immature white matter in preterm infant and approximately 15% in full term infants. The 1st indication of spastic diplegia is often noted when an affected infant begins to crawl, he tends to drag the legs behind more, commando crawl rather than using 4 limbs in reciprocal fashion. 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 tiptoe. Severe spastic diplegia is characterized by disuse atrophy and impaired growth of the lower extremities.

The prognosis for normal intellectual development is excellent for these patients, and the likelihood of seizures is minimal. Such children often have learning disabilities and deficits in other abilities, such as vision, because of disruption of multiple white matter pathways that carry sensory as well as motor information. The most common neuropathologic finding is periventricular leukomalacia, particularly in the area where fibers innervating the legs course through the internal capsule.

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, 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 (choreoathetoid or **extrapyramidal** CP): 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. Generally, upper motor neuron signs are not present, seizures are uncommon, and intellect is preserved in many patients. This form of CP is the type most likely to be associated with birth asphyxia & kernicterus. In the European CP study, 76% of patients with this form of CP had lesions in the basal ganglia and thalamus.

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. An **MRI** of the brain is indicated to determine the location and extent of structural lesions or associated congenital malformations or if spinal cord pathology is suspected. Additional studies may include **tests of hearing and visual** function. **Genetic evaluation** should be considered in patients with congenital malformations (chromosomes) or evidence of metabolic disorders.

TREATMENT

A **multidisciplinary team** includes physicians, occupational and physical therapists, speech pathologists, social workers, educators, ophthalmologist, and developmental psychologists provide important contributions to the treatment of these children. Parents should be taught how to work with their child in daily activities such as feeding, carrying, dressing, bathing, and playing in ways that limit the effects of abnormal muscle tone. Children with **spastic diplegia** are treated initially with the assistance of adaptive equipment, such as walkers, 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 produces considerable improvement in selected patients with severe spastic diplegia. A **tight heel cord** may be treated surgically by tenotomy of the Achilles

tendon. **Quadriplegia** is managed with motorized wheelchairs, special feeding devices, & modified typewriters.
Lower urinary tract dysfunction should receive prompt assessment and treatment.
Several drugs have been used to treat spasticity, including oral dantrolene sodium, the benzodiazepines, and baclofen. Botulinum toxin injected into specific muscle groups for the management of spasticity shows a very positive response in many patients.

Mental Retardation (Intellectual Disability)

It is significantly sub-average general intellectual functioning with deficits in adaptive behavior and manifested during the developmental period. The onset is before age 18 years.
- Significantly sub-average intellectual functioning: an IQ score of ≈70.
- Concurrent deficits or impairments in present adaptive functioning (i.e., deficit in meeting the standards according the culture, e.g. communication, self-care, & home living).

Classification:
- Depending on I Q level:
Mild Mental Retardation: 52- ≈70.
Moderate Mental Retardation: 37- 51.
Severe Mental Retardation: 20- 36.
Profound Mental Retardation: below 20.
Mental retardation Severity Unspecified: when there is a strong presumption of mental retardation but the is untestable by standard tests

- Depending on levels of support required: intermittent, limited, extensive, or pervasive.

ETIOLOGY
2.5% of the population have mental retardation, and 85% of these individuals should fall into the range of mild mental retardation.
Mild MR is presumably a consequence of both genetic (children may inherit an intellectual impairment) and socioeconomic (poverty, undernutrition) factors. The specific causes of mild mental retardation are currently identifiable in <50% of affected individuals. The most common biologic causes of mild mental retardation include genetic syndromes with multiple minor congenital anomalies, fetal deprivation, prematurity, perinatal insults, intrauterine exposure to drugs of abuse, and sex chromosomal abnormalities.
While severe mental retardation is more frequently linked to biologic causes, a biologic cause (most commonly prenatal) can be identified in >75% of cases. Causes include chromosomal (Down syndrome) and other genetic syndromes (fragile X syndrome), abnormalities of brain development (lissencephaly), and inborn errors of metabolism/neurodegenerative disorders (mucopolysaccharidoses), Congenital infections, Perinatal causes (HIE, meningitis, IVH, PVL, fetal alcohol syndrome), Postnatal causes (Trauma, meningitis, hypothyroidism).

CLINICAL MANIFESTATIONS
Early diagnosis of mental retardation facilitates earlier intervention, decreasing parental anxiety, and greater acceptance of the child in the community. Most children with intellectual disability 1st come to the pediatrician's attention in infancy because of **dysmorphisms, associated problems, or failure to meet age-appropriate developmental milestones**.
Associated problems: e.g. seizures, cerebral palsy, autism.
Developmental delay:
- In early infancy, it includes a lack of visual or auditory responsiveness, unusual muscle tone (hypo- or hypertonia) or posture, and feeding difficulties.

- Between 6 and 18 mo of age, motor delay (lack of sitting, crawling, walking) is the most common complaint.
- Language delay and behavior problems are common concerns after 18 mo. **LABORATORY FINDINGS.**

The most commonly used medical diagnostic testing for children with mental retardation include **neuroimaging, metabolic, and chromosomal study, and EEG.** Decisions on diagnostic testing should be based on the medical/family history, physical examination, testing by other disciplines, and the family's wishes.

TREATMENT

Although mental retardation is not treatable, many **associated impairments** are amenable to intervention and, therefore, benefit from early identification.

The **behavioral/emotional** disorders are the primary cause for out-of-home placements, reduced employment prospects, and decreased opportunities for social integration. When intervention is needed, an environmental change, such as a more appropriate classroom setting, mental age here is used.

No agent has been found to improve intellectual function.

Education is the single most important discipline involved in the treatment of children with an intellectual disability. The educational program must be relevant to the child's needs and address the child's individual strengths and weaknesses.