Cerebral palsy (CP)

It is a diagnostic term used to describe a group of motor syndromes. A static encephalopathy, a term previously used, is now inaccurate because of the recognition that the neurologic features of CP often change or progress over time. Although CP is often associated with epilepsy and abnormalities of speech, vision, and intellect, it is the selective defect of the brain’s motor systems that defines the disorder.

**Epidemiology and etiology.**

CP is the most common form of chronic motor disability with a prevalence of 2/1000.

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 low birth weight infants, particularly those weighing less than 1,000 g at birth, primarily because of intracerebral hemorrhage and periventricular leukomalacia (PVL).

<table>
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<th>Classification of Cerebral Palsy and Major Causes</th>
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<td><strong>Physiologic</strong></td>
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<td>Athetoid</td>
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<td>Ataxic</td>
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<td>Tremor</td>
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**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. Walking is usually delayed until 18–24 mo, and a circumductive gait is apparent. Examination of the extremities may show growth arrest. 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 a Babinski sign may be present, the deep tendon reflexes are increased. About one third of patients with spastic hemiplegia have a seizure disorder; ≈25% have 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. The
neuropathology is stroke. The causes are: Thrombophilic disorders, infection, periventricular hemorrhagic infarction, genetic.

**Spastic Diplegia** is bilateral spasticity of the legs. The 1st indication of spastic diplegia is often noted when an affected infant begins to crawl (tends to drag the legs behind more, commando crawl). 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. The most common neuropathologic finding is periventricular leukomalacia, particularly in the area where fibers innervating the legs course through the internal capsule. The probable causes of PVL are: prematurity, ischemia, infection, endocrine/metabolic.

**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 quadripareisis often have evidence of athetosis and may be classified as having mixed CP. The probable causes are: ischemia, infection, endocrine/metabolic, genetic.

**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. 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 the type most likely to be associated with birth asphyxia & kernicterus.

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 scan of the brain is indicated to determine the location and extent of structural lesions or associated congenital malformations; an MRI scan of the spinal cord is indicated 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 (chromosomes) or evidence of metabolic disorders.

TREATMENT.
A multidisciplinary team include 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. Intrathecal baclofen has been used successfully in selected children with severe spasticity. Botulinum toxin injected into specific muscle groups for the management of spasticity shows a very positive response in many patients. Patients with rigidity, dystonia, and spastic quadriplegics sometimes respond to levodopa, and children with dystonia may benefit from carbamazepine or trihexyphenidyl.
Mental Retardation (Intellectual Disability)

It is significantly sub-average general intellectual functioning, existing concurrently 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 or below on an individually administered IQ test.
- Concurrent deficits or impairments in present adaptive functioning (i.e., deficit in meeting the standards expected for his or her age by his or her cultural group) in at least two of the following areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety.

Classification:
- Depending on IQ level:
  Mild Mental Retardation: 52- ≈70.
  Moderate Mental Retardation: 36- 52.
  Severe Mental Retardation: 20- 36.
  Profound Mental Retardation: below 20.
- Depending on levels of support required (adaptive function): intermittent, limited, extensive, or pervasive.

ETIOLOGY.
2.5% of the population should 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, easing of parental anxiety, and greater acceptance of the child in the community. Most children with intellectual disability first come to the pediatrician's attention in infancy because of dysmorphisms, associated dysfunctions, or failure to meet age-appropriate developmental milestones. Associated dysfunctions: are neurologic disorders (seizures, cerebral palsy, autism) that are seen more frequently in conjunction with mental retardation than in the general population.

Developmental delay: In early infancy, failure to meet age-appropriate expectations may include 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. Earlier identification of atypical development is likely to occur with more severe impairments.

LABORATORY FINDINGS.
The most commonly used medical diagnostic testing for children with mental retardation include neuroimaging; metabolic, and chromosomal study; and electroencephalography (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. Aggression, self-injury, oppositional-defiant behavior, and mental illness (mood and anxiety disorders) occur with greater frequency in this population than among children with typical intelligence. These behavioral/emotional disorders are the primary cause for out-of-home placements, reduced employment prospects, and decreased opportunities for social integration. In assessing the behavior, one must also consider whether it is inappropriate for the child's mental age, rather than the chronological age. When intervention is needed, an environmental change, such as a more appropriate classroom setting, No agent has been found to improve intellectual function. Medication may be helpful in treating associated behavioral and psychiatric disorders or associated dysfunctions.