Cerebral Palsy (CP)

Learning objectives:
1. Define CP.
2. Discuss the etiologies of CP.
3. Discuss the topographic classifications of CP and the specific features of each type.
4. Know the common associated problems of CP.
5. Describe the important strategies of treatment of CP.

Definition:
Is a nonprogressive disorder of posture or movement caused by a lesion of the developing brain. Cerebral palsy is generally considered a static encephalopathy (ie, nonprogressive). However, the clinical presentation of CP changes as children mature.

Prevalence:
CP is the most common chronic motor disability in children with a prevalence of 2/1000 live births.
The prevalence of CP has increased among small premature infants as more have survived.

Etiology:
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.
1. Congenital CP (due to cerebral injury/maldevelopment before or during birth) accounts for 85–90% of total cases.
2. Acquired CP (due to cerebral injury after 1 month of life) is responsible for the remaining cases. The most common cause in this category is perinatal stroke, the second most common cause is meningitis or encephalitis during infancy.
3. Cryptogenic CP: when no clear perinatal etiology has been identified.

There is a significant association of CP with prematurity and low birth weight infants, particularly those weighing less than 1,000 g at birth, primarly because of intraventricular hemorrhage and periventricular leukomalacia (PVL).
PVL reflects the enhanced vulnerability of immature oligodendroglia in premature infants to oxidative stress caused by ischemia or infectious/inflammatory insults.

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Examples of common etiologies of CP:
1. Developmental anomalies of the brain.
2. Congenital infection.
3. Perinatal trauma (intracerebral hemorrhage, cerebral infarction, or both).
4. Hypoxic ischemic encephalopathy (HIE).
5. Hypoglycemia.
6. Hyperbilirubinemia.
7. Thromboembolic.
8. Prematurity.

Comorbidities
About 70% of cases have additional comorbidities like seizures, intellectual disabilities, speech and language impairment, vision deficits, and hearing loss….

Classification of cerebral palsy
Aims of classification:
1. Understanding the cause.
2. Monitoring comorbidities.
3. Treatment options.
4. Prognosis.
Classified according to the type of motor involvement into *spastic* (80% of cases) or *extrapyramidal* (20% of cases).
Spastic CP then further subdivided topographically into: spastic hemiplegia, spastic monoplegia, spastic diplegia, and spastic quadriplegia.
Extrapyramidal CP can be divided into the two main types of involuntary movement seen: ataxia and dyskinesias.

A. Spastic CP:

1. **Spastic hemiplegia:** one side of the body involved, with upper extremity spasticity more than lower extremity spasticity. Usually due to stroke (in utero or neonatal).
Clinical manifestations:
- decreased spontaneous movements on the affected side.
- hand preference at a very early age.
- delayed walking and a circumductive gait.
- examination of the involved extremity show growth arrest , spasticity , equinous deformity of the foot , increased deep tendon reflexes , ankle clonus and a Babinski sign .
- Seizure disorder that usually develops in the 1st 2 yr.

- About 25% have cognitive abnormalities including mental retardation.

- CT or MRI study of the brain may show atrophic cerebral hemisphere with a dilated lateral ventricle contralateral to the side of the affected extremity.

2. **Spastic diplegia:** is bilateral spasticity of the legs greater than the arms. The most common neuropathologic finding is periventricular leukomalacia (PVL). Infants with PVL are at higher risk of cerebral palsy because of injury to the corticospinal tracts that descend through the periventricular white matter. PVL is characterized by focal necrotic lesions in the periventricular white matter and/or more diffuse white matter damage.
Clinical manifestations:
-the 1st indication of spastic diplegia is often noted when an affected infant begins to crawl in which the child uses the arms in a normal reciprocal fashion but tends to drag the legs behind rather than using the normal four-limbed crawling movement .
- In severe cases, application of diaper is difficult because of the excessive adduction of the hips.
- The child may be unable to sit.
- Examination of the child reveals: spasticity in the legs with brisk reflexes, ankle clonus, and bilateral Babinski sign.
- When the child is suspended by the axillae, a scissoring posture of the lower extremities is maintained because of strong adductor muscles contractions.
- walking is delayed, the feet are held in a position of equinovarus, and the child walks on tiptoe.
- Disuse atrophy and impaired growth of the lower extremity in severe cases.
- Subluxation or dislocation of the hips is relatively common in children with severe spasticity.
- Intellectual development is excellent in these patients and the likelihood of seizures is minimal.

3. **Spastic quadriplegia:** is the most severe form of CP because of marked impairment of all extremities and the high association with mental retardation and seizures.
The most common lesions seen on pathological examination are severe PVL and multicystic cortical encephalomalacia.
Clinical manifestations:
- Profound developmental delay.
-neurological examination shows increased tone and spasticity in all extremities with decreased spontaneous movements and brisk reflexes.
- Flexion contractures of the knees and elbows.
- swallowing difficulty is common due to supranuclear bulbar palsies which may lead to aspiration pneumonia.
- Associated developmental disabilities, including speech and visual abnormalities is highly prevalent in this group of children.

B. Extrapyramidal CP:
Can be divided into the two main types of involuntary movement seen: ataxia and dyskinesias. It is less common than spastic CP and makes up about 15-20% of patients with CP.
1. **Ataxic CP:**

Characterized by cerebellar signs and abnormalities of voluntary movements.

2. **Dyskinetic CP**: is further divided into two groups: athetoid and dystonic.
Athetoid CP: involuntary movements affecting mainly the arms, legs, and hands often caused by kernicterus and HIE.
Dystonic CP: affect the trunk muscles more than the limbs and results in a fixed, twisted posture
The neuropathological lesions seen in this type of CP is basal ganglia lesions (mainly bilateral globus pallidus).

Clinical manifestations:
- Affected infants are characteristically hypotonic.
- Rigidity and dystonia develops over several years.
- Feeding difficulty, tongue thrust and drooling may be prominent.
- Speech problems e.g. slurred speech.
- Upper motor neuron signs are not present.
- Seizures are uncommon; intellect is preserved in many patients.

5. **Mixed CP:**
Two or more types codominant, most often spastic and athetoid.

Management
Includes investigations for the causative factors and screening for the associated comorbidities.
Treatment needs a team of physicians from various specialties as well as physical therapists, speech therapists, social workers, and developmental psychologists.
Management of the child with cerebral palsy should involve:
1. Management of the commonly associated disabilities and health problems.
2. Management of the consequences of the motor disorders:

- Spasticity management (antispasticity medications): several drugs have been used to treat spasticity, including: oral dantrolene sodium, benzodiazepines, tizanidine and baclofen.
Second- line medications such as clonidine or gabapentin may provide dual benefit for both tone management and other neurologic associations, including sleep disruption, pain, and irritability.
Intrathecal baclofen (ITB) has been successfully used in selected children with severe spasticity whose spasticity is not adequately treated with enteral baclofen or who are experiencing side effects such as sedation.

Botulinum toxin A injected into specific muscle groups may be used for localized spasticity.
Rhizotomy procedures (Surgical division of dorsal spinal roots) may be used to relieve spasticity in spastic diplegia.

- Medications for dystonia: e.g. trihexyphenidyl, levodopamine.
- Deep brain stimulation (DBS): could help patients with medically refractory dystonia. It involves the introduction of stimulating electrodes in areas of the brain such as the globus pallidus and the subthalamic nucleus, which are connected to an external pulse generators.

- Orthopedic problems:
E.g. adductor tenotomy to reduce muscle spasm around the hip, tenotomy of the Achilles tendon to treat a tight heel cord...

3. Physical and occupational therapy:
Is essential to enable children to achieve their optimal physical potential and independence.