Neurological disorders -IV

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CEREBRAL PALSY (CP)

Definition

- A group of motor syndromes resulting from disorders of early brain development.

- Cerebral palsy (CP) is a non-progressive, non-fatal, non-curable disorder of the CNS resulting from malfunction of the brain centers and pathways .

- Characterized by motor disability and/or other manifestations of organic brain damage

  Seizures,

  Mental retardation,

  Sensory and learning defects

  Behavior and emotional disturbances.

- The term static encephalopathy is not entirely accurate as the neurological features of cerebral palsy often change or progress overtime.

Etiology

Prenatal causes

A-Genetic (Hereditary)

B-Acquired (Environmental)

Intrauterine infections as toxoplasmosis, rubella, cytomegalovirus, herpes, mumps, syphilis.

Fetal anoxia as placental separation

Maternal Trauma

Toxemia of pregnancy

Maternal malnutrition and vitamin deficiency

Maternal pelvic irradiation

Natal and perinatal causes

- Anoxia: Hypoxic ischemic encephalopathy
- Trauma: birth injuries and intracranial hemorrhage
- Prematurity because of frequent association with anoxia, respiratory problems and intracranial hemorrhage

Postnatal causes

- Infections as meningitis and encephalitis
- Trauma: skull fracture and hemorrhage
- Kernicterus
- Cerebrovascular accidents: thrombosis and embolism

Clinical Classification

Spastic Cerebral Palsy

Clasp-Knife hypertonia

Markedly exaggerated deep tendon reflexes with sustained clonus

Positive Babiniski sign.

Persistence of primitive reflexes as Moro reflex

Abnormal tone leads to contractures most commonly at hips, knees and ankles

It may be

Hemiplegic: hemiparesis affects one side of the body and arms more than legs.

Quadriplegic: quadriparesis affects both sides of the body and arms as much as or more than legs.

Spastic diplegia: both legs are spastic and arms are less affected or not affected at all.

Dystonic (Athetoid) cerebral palsy (10%)

- Irregular and involuntary movements of some or all muscle groups
- Athetosis is the commonest form with slow twisting movement.
- Other movements as chorea and Dystonia may occur

Ataxic cerebral palsy (5%)

- Disturbance in balance and equilibrium
- Hypotonia, weakness, uncoordinated movement and intention tremors.

Cerebral infantile hypotonia (5%)

- Presents like floppy infant
- Hypotonia of the trunk and head is marked
- Tendon reflexes are preserved and brisk

Mixed cerebral palsy (10%)

- spasticity and ataxia may coexist

Functional Classification

Mild

- the child is able to ambulate, uses his arms and speak. He does not need special care.

Moderate

- the involvement affects the child but not sufficient to disable him completely. Such patients need special care

Severe

- the involvement affects the child entirely so that he is bed ridden or restricted to a wheel chair.

Clinical picture to detect early CP

Infants who show

- poor suckling,
- increased or decreased tone,
- abnormal reflexes,
- irritability, convulsions or drowsiness in the neonatal period

are at risk of having cerebral palsy in the future.

- Usually cerebral palsy is not diagnosed until several months have passed
- when it becomes obvious that motor development is abnormal or delayed .
- An infant with spastic hemiplegia will be noted to have developed hand preferences before one year that is much earlier than normal.

- **The persistence of the primitive reflexes** beyond the time they usually disappear is suspicious.

- **the characteristic involuntary movements of Dystonic cerebral palsy**

1- are not usually obvious until near the end of the first year of life or even latter,

2- affected children are usually very floppy and show delayed motor development in infancy.

children with ataxic cerebral palsy are

- hypotonic
- show delayed motor development
- subsequently show intention tremor .

Treatment

This is best done by the cooperation of

- pediatrician,
- physiotherapist, orthopedist,
- surgeon,
- psychiatrist
- the family.

- **Pediatrician:** to take care of the child throughout the disease, to supervise nutritional status, prescribe muscle relaxants and other necessary medications.

a-Oral dantrolene sodium

b-Benzodaizepines.

c-Oral & Intrathecal Baclofen.

- Physiotherapist: to do physiotherapy to prevent muscle contractures and assist the child for better control of muscles.

- Family: the parents are taught to perform exercise at home to prevent contractures.

- **Orthopedist and surgeon: to deal with contractures and provide stability or for cosmetic reasons.**

a-Hip adductor tenotomy.

b-Psoas muscle transfer and release.

c-Achilles tenotomy.

d-Rhizotomy.

e-Botolinum Toxin (Botox) injection into spastic muscles

FLOPPY INFANT

Definition

- Floppy infant is that who present with severe persistent hypotonia since birth or early infancy.

Etiology

- Central causes
  - Atonic cerebral palsy
  - Chromosomal diseases; e.g. Down syndrome
  - Cerebral anomalies
  - Other causes as metabolic and degenerative diseases

Neuromuscular causes

- Werding-Hoffman disease
- Congenital muscle dystrophy
- Congenital myopathies
- Benign congenital hypotonia
- Other causes as metabolic diseases and congenital myasthenia gravis.

Clinical picture

Hypotonia

Hypotonia of limbs

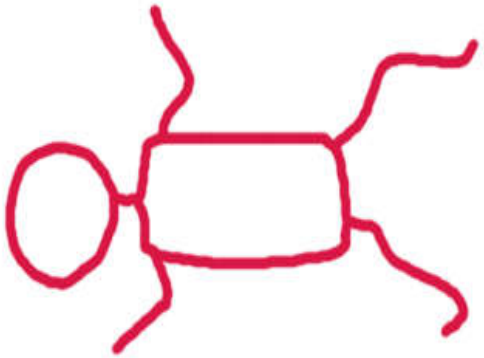
limbs are abducted and slightly flexed resembling frog legs position.

Hypotonia of trunk

curved trunk when suspended in prone position over examiner's palm.

Hypotonia of neck

the head lags backward when the infant is pulled up from his hands while in supine position.



In central causes:

mental retardation is common. Convulsions may occur. Tendon reflexes are exaggerated.

In peripheral causes:

Mentality is usually normal. Tendon reflexes are weak or absent.

Features of the cause

Abnormal features in chromosomal disorders

Persistent primitive neonatal reflexes in cerebral palsy.

Tongue fasciculations in Werding-Hoffman disease

Thin muscles bulk in congenital myopathy and muscle dystrophy.

Investigations

- **Radiological**

  CT and MRI: may show

  1-brain atrophy,

  2-anomalies or

  3-degenerative disease

- **Laboratory**
  - **Chromosomal karyotype**
  - **Metabolic screen**
  - **Creatinine phosphokinase level: elevated in muscle dystrophy.**

- Others
  - EMG
  - Nerve conduction velocity
  - Muscle biopsy

Treatment

- Treatment is directed towards treatable causes.


Thank you