CEREBRAL PALSY

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DEFINITION

- Cerebral palsy describes a group of disorders attributed to non-progressive disturbances that occur in the developing fetal or infant brain and result in a cluster of disorders that impact development of movement and posture, causing activity limitation.
- It is non-contagious motor conditions that cause physical disability in human development

- In addition to primary impairments in gross & fine motor function, there may be associated problems with cognition, seizures, vision, swallowing, speech, bowel-bladder, & orthopedic deformities

HISTORY

- First identified by English surgeon **William Little** in 1860. (Little's disease)
  - Believed that **asphyxia during birth** is chief cause
- National Institute of Neurological Disorders & Stroke (NINDS) in 1980s suggested that only a small number of cases of CP are caused by lack of oxygen during birth

EPIDEMIOLOGY

- The incidence of CP is about 2 to 2.5 per 1000 live births
- The incidence is higher in males than in females
- Other associated problems include
  - Mental disadvantage (IQ < 50): 31%
  - Active seizures: 21%
  - Mental disadvantage (IQ < 50) and not walking: 20%
  - Blindness: 11%

CAUSES OF CP

- Prenatal (70%)
- Peri-natal (5-10%)
- Post natal (20%)

PRENATAL (BEFORE DELIVERY)

- Maternal infections E.g. rubella, herpes simplex
- Inflammation of placenta (chorion amnionitis)
- Intake of Oral Contraceptive drugs.
- Diabetes during pregnancy
- Genetic causes
- Exposure to radiations
- Maternal jaundice
- Seizure Disorder

PERI- NATAL (DURING DELIVERY)

- Prematurity- immature respiratory & cardiac function
- Asphyxia
- Maconeum aspiration
- Birth trauma
- Disproportion
- Breech delivery
- Rapid delivery
- Low birth weight

POST NATAL (AFTER DELIVERY)

- Brain damage secondary to cerebral hemorrhage, trauma, infection or anoxia
- Motor vehicle accidents
- Shaken baby syndrome
- Drowning
- Lead exposure
- Meningitis
- Encephalitis

CLASSIFICATION OF CP

- **Depending on the topographical distribution**
  - Monoplegic
  - Diplegic/ Paraplegic
  - Triplegic
  - Hemiplegic
  - Tetraplegic / Double hemiplegia

- **Monoplegia:** is one single limb being affected.
- **Diplegia:** LE affected, with little to no upper-body spasticity.
  - The most common form of spastic forms
  - Most people with spastic diplegia are fully ambulatory, but are "tight" & have scissors gait
  - Flexed knees & hips to varying degrees, & moderate to severe adduction are present
  - Often nearsighted & intelligence is unaffected
  - In 1/3rd of spastic diplegics, strabismus may be present.

- Hemiplegia:
  - The most ambulatory of all forms, although they generally have dynamic equinus on affected side
- Triplegia: three limbs affected usually both LL & one UL
- Quadriplegia: all four limbs more or less equally affected.
  - Least likely to be able to walk
  - Some children also have hemiparetic tremors (hemiballismus), & impairs normal movement

- **Depending on tone or movement patterns (physiologic)**
    - Spastic
    - Athetoid/ dyskinetic
    - Ataxic
    - Flaccid/ Hypotonic
    - Mixed

Spastic CP

- It is the most common type of CP, occurring in 70% to 80% of all cases.
- The cerebral cortex is affected
- Moreover, spastic CP accompanies any of the other types of CP in 30% of all cases
- It can be monoplegia, diplegia, triplegia, hemiplegia or quadriplegia.

Athetoid/ dyskinetic CP

- It is mixed muscle tone. Often show involuntary motions
- The damage occurs to extrapyramidal motor system & pyramidal tract
- It occurs in 10% to 20% of all cases
- In newborn infants, high bilirubin levels in blood, if left untreated, can lead to brain damage in certain areas (kernicterus).
- This may also lead to Athetoid CP.

ATAXIC CP

- It is caused by damage to cerebellum.
- They are least common types of CP, occurring only in 10% of all cases.
- Some of these individuals have hypotonia and tremors.

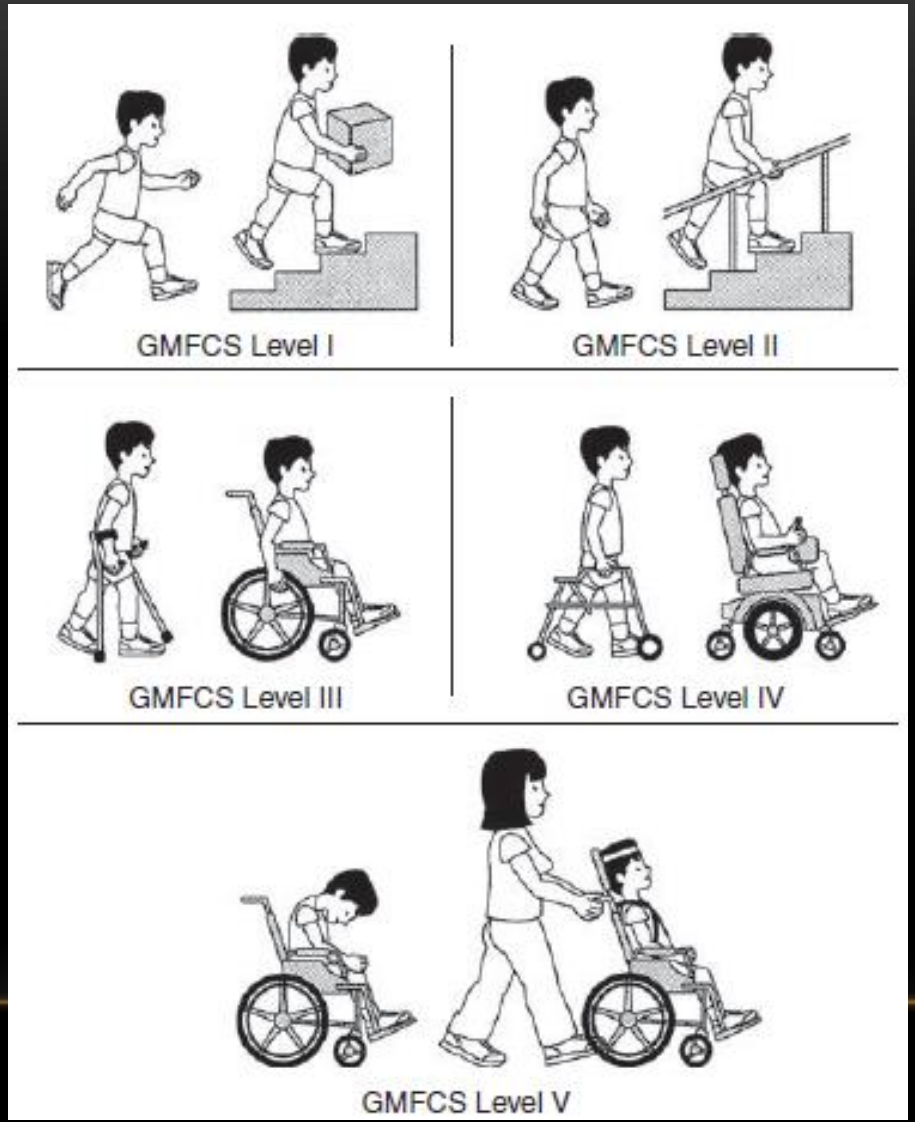
HYPOTONIC CP

- Hypotonic CP have musculature that is limp, and can move only a little or not at all (Floppy child)
- The location of damage is wide spread in the CNS

MIXED CP

- Signs & symptoms of spastic CP is seen with any other type of CP
- Most commonly mixed with Athetoid

- **Depending on functional level (Gross Motor Function Classification System)**
    - It classifies according to age categorized activity level


GMFCS Level I
GMFCS Level II
GMFCS Level III
GMFCS Level IV
GMFCS Level V

PATHOPHYSIOLOGY

- Periventricular leukomalacia (PVL) is the most common finding in CP
- Corticospinal tract fibers to LL are medial to those of UL in periventricular white matter.
- Thus children with PVL typically have spastic diplegia (common type of CP)

- Bilirubin encephalopathy in basal ganglia is seen in athetoid CP following a diagnosis of kernicterus
- Focal cortical infarcts involving both grey & white matter are found in patients with hemiparesis, & are typically related to MCA strokes
- Brain malformations can be found on neuroimaging in approximately 10% of children

CLINICAL FEATURES OF CP

SIGNS & SYMPTOMS (SPASTIC)

- Hypertonia
- Exaggerated reflexes & +ve babinsky
- Clonus
- Poor voluntary movement
- Scissoring gait
- Low intelligence & loss of memory
- Epilepsy
- Loss of Synergistic pattern
- Contracture, deformity & wasting
  - Adduction & IR of shoulder
  - Flexion of elbow & pronation of forearm
  - Wrist flexion & thumb inside hand
  - Flexion & adduction of hip
  - Knee flexion
  - PF of ankle

EXTRAPYRAMIDAL CP

- May affect limb, face, tongue & speech
- Postural instability
- Decreased movement in prone position
- Fluctuation of tone from high to low
- Reflexes are usually normal & muscles are able to contract
- Decreased stability
- Emotional liability
- Sucking & feeding problems
- Delayed head & trunk control
- May be either quadriplegic or rarely hemiplegic
- Subtypes - dystonic, athetoid, choroid, hemiballismic, rigid

ATAXIC / HYPOTONIC

- Inco-ordination
- Intension tremor
- Hypotonia
- Nystagmus
- Diminished reflexes
- Speech, visual, hearing & perceptual problems
- Joint hypermobility
- Dysmetria
- Incontinence
- Postural instability
- Gait disturbances
- Imbalance & lack of trunk control

RISK FACTORS

RISK BABIES

- Biological risk
- Established risk
- Environmental & social risk

BIOLOGICAL RISK

- Birth weight of 1500g or less
- Asphyxia with apgar score <3 in 1 min after birth or <6 in 5 min after birth
- Gestational age of 32 weeks or less
- Ventilator requirement for 36 hours or more
- Intracranial hemorrhage
- Recurrent neonatal seizures (3 or more)
- Feeding dysfunction
- Meningitis

ESTABLISHED RISK

- Hydrocephalus
- Microcephaly
- Chromosomal abnormalities
- Musculoskeletal abnormalities (CDH, AMC, limb deficiencies)
- Multiple births more than twins
- Brachial plexus injuries
- Myelodysplasia
- Congenital myopathies
- Inborn errors of metabolism
- HIV infection

ENVIRONMENTAL / SOCIAL RISK

- Parental age less than 17
- Poor quality infant parent attachment
- Maternal drug or alcohol abuse
- Behavioral state abnormalities (lethargy, irritability)

SCREENING OF CP

PRENATAL SCREENING

- Routine check up for mother during pregnancy is beneficial for the mother & the foetus
- Health education
  - Diet advice (avoid tobacco & alcohol)
  - Exercise on prescription
  - Sleep & working habits

- **Clinical examination**
  - Breast condition
  - Height of uterus
  - Position of foetus
  - Samples of blood & urine

- **For special test condition sought are**
  - Phenylketonuria (Phenylalanine)
  - Glycosuria (Excess sugar)
  - Albuminuria
  - Rh incompatibility (+ & -)
  - Congenital syphillis
  - Rubella, AIDS
  - Neural tube defect

- **Special test for screening**

- **USG from 8-12 weeks**
  - For the assessment of the gestational period
  - Congenital abnormalities in various organ defect
- **Amniocentesis** from 16-18 weeks of pregnancy
  - To find chromosomal defect, if the test is positive terminate pregnancy
- **Chorionic villus sampling technique** in 8-11 weeks of pregnancy
  - Sample of tissues from placenta to test for chromosomal abnormality,

POST NATAL SCREENING

- Starts in the immediate neonatal period & during the first two years
  - Assessment of Developmental milestone
  - Any biochemical defects
  - Hearing & visual problems
  - Behavioral changes

- **Clinical methods (at birth)**
  - APGAR score
  - Examination of weight, height, head circumference
  - Gestational age
  - Musculoskeletal defects

- **Chemical methods**
  - Blood sample from heel prick at 2-5 days of age & repeated where necessary
    - Respiratory conditions, cardiac pathology, haemoglobinopathies can be detected
    - Neuromuscular pathologies (cpk level)
    - Metabolic disorders
    - Gene abnormalities

- **Electronic scanning**
  - USG, CT Scan, MRI
    - To find out AVM, hemorrhage, cyst, leucodystrophies (Degeneration of white mtter) etc.

DIAGNOSIS

- The diagnosis of CP depends on patient's history & on the basis of significant delay in gross & fine motor function, with abnormalities in tone, posture, & movement on neurological examination.
- Once diagnosed with CP, further diagnostic tests are optional.

- MRI is preferred over CT due to diagnostic yield & safety.
- The CT or MRI also reveals treatable conditions, such as hydrocephalus, AVM, subdural hematomas etc.
- Diagnosis, classification, & treatment are often based on abnormalities in tone
- Apgar scores have sometimes been used as one factor to predict whether or not an individual will develop CP

DIAGNOSTIC TOOLS

- **Movement Assessment of Infants (MAI):** able to predict CP at 4 months (identifies motor delay)
- **Alberta Infant Motor Scale (AIMS)** is able to predict CP at 6 months (Identifies motor delays & measures changes in motor performance over time)
- **Bayley scale** is able to predict CP at 1 year (Identifies devt delay in gross & fine motor, & cognitive domains)

MANAGEMENT

- Medical
- Surgical
- Rehabilitative

MEDICAL MANAGEMENT

- Oral medications such as baclofen, diazepam, and trihexyphenidyl as well as therapeutic botulinum toxin (Botox)
- Children with dystonic CP have dopa-responsive dystonia, with improved motor function using levodopa
- Children with basal ganglia/thalamic injury from perinatal asphyxia may develop improved expressive speech & hand use with trihexyphenidyl

SURGICAL MANAGEMENT

- Dorsal rhizotomy reduces spasticity
- Joint & Tendon release most often performed on hips, knees, & ankles.
- The insertion of a baclofen pump usually during young adolescence.
  - usually placed in left abdomen - a pump that is connected to spinal cord,
  - sends bits of Baclofen to relax muscle
- Bony correction E.g. femur (termed femoral anteversion or antetorsion) & tibia (tibial torsion).

PROGNOSIS

- CP is not a progressive but the symptoms can become more severe over time
- Prognosis depends on intensity of therapy during early childhood
- Tend to develop arthritis at a younger age than normal because of pressure placed on joints by excessively toned & stiff muscles

- Intellectual level among people with CP varies from genius to intellectually impaired
- The ability to live independently with CP varies widely depending on severity of each case.
  - Some individuals with CP are dependent for all ADL.
  - Some can lead semi-independent lives, needing support only for certain activities.
  - Still others can live in complete independence.