The Basics of Cerebral Palsy

General Presentation

Cerebral Palsy (CP) is a term used to describe a heterogenous group of motor impairment syndromes caused by a **non-progressive** lesion of the developing brain. It is synonymous with the term “static encephalopathy”. However, although the disorder itself is non-progressive, the lesions themselves and their clinical expression may change over time as the brain matures.

The incidence of CP is 2.0 to 2.5 per 1000 live births in developed countries and is higher in premature infants. In general, males are affected slightly more than females, in a 1.2:1 ratio.

The cause is unknown in the majority of patients and the lesion itself can arise prenatally, perinatally or postnatally. In a minority of cases, single or multiple risk factors can be identified. The presentation of CP can vary widely, from mild impairments of advanced gross motor function to a severe limitation of self-mobility even with supportive aids.

There are multiple classification systems:

1) Anatomic:

- *Diplegia* = lower limbs affected more than upper limbs
- *Hemiplegia* = one side of the body affected more than the other
- *Quadriplegia* = all limbs are similarly affected
- *Paraplegia* = only lower limbs affected

2) Physiologic:

- *Spasticity* (75%) = upper motor neuron lesion leading to velocity dependent increased resistance in response to passive stretch. This results in increased muscle tone, hyperreflexia, extensor plantar responses and clonus.
- *Ataxia* (15%) = cerebellar lesion resulting in uncoordinated movements. This often presents as hypotonia in the first year of life with delayed milestones.
- *Dyskinesia* (including choreoathetosis) (5%) = Basal ganglia lesion which results in abnormalities of voluntary movement. It is a collective term for athetosis, chorea, & dystonia.
    - *Athestosis* refers to slow, writhing involuntary movements of the distal muscles
    - *Chorea* refers to rapid, irregular, involuntary movements of individual or small groups of muscles. This affects term infants and in most cases results from severe perinatal asphyxia.
    - *Dystonia* refers to repetitive, patterned, twisting and sustained movements of the trunks or limbs that may be either slow or rapid
- *Pure hypertonia without spasticity* (1%)

3) **Functional:**
    - Mild, moderate, and severe limitations

Usually you will see CP described as a combination of these classifications. For example, severe spastic quadriplegia meaning a severe form of spasticity in all four limbs and so on. CP is characterized by abnormalities of movement and posture. Here we will focus on the type and distribution of the motor abnormalities remembering that there is often substantial overlap between clinical syndromes.

Table 1:

Spastic	Ataxic	Dyskinetic
Diplegia	Simple ataxia	Mainly dystonic
Hemiplegia	Ataxia diplegia	Mainly athetoid
Quadraplegia		

CP is associated with many disorders of cerebral function. Associated neurological deficits can include seizures (35-50%), mental retardation (65%), visual deficits (50%), psychiatric disorders and speech impairment.

Other associated disorders include growth failure, orthopaedic disorders such as hip subluxation, dislocation and dysplasia, contractures, osteopenia and dysfunctional voiding.

Questions to ask

A complete history is key to diagnosing and managing children with CP. The following are key elements of the history not to miss:

History of Presenting Illness:

CP commonly presents with delayed milestones or other concerns with development, abnormal movement, abnormal tone ("floppy" or stiff & difficult to dress or put into car seat), asymmetric movements (e.g., asymmetric crawling)

Review of Systems:

Hearing loss, "lazy eye", abnormal oculomotor or oromotor patterns, constipation, dysphagia, GERD, dental caries, recurrent chest infections (2^o to aspiration), pressure ulcers, seizures

Past Medical History:

Pregnancy History: Exposures to drugs or toxins, acute maternal illnesses, maternal thyroid disorder, gestational diabetes, prenatal ultrasounds, extent of prenatal care and any other complications.

Birth History: Gestational age, multiple births (e.g. twins) delivery type, presentation, birth measurements, APGAR scores, birth complications (e.g. resuscitation needed, time in hospital)

Neonatal History: infections, jaundice, investigations required & results if known

Growth & Development:

You must complete a very thorough review of gross motor, fine motor, speech & language, and social milestones specific for age. Particularly note any regressions as this is a very worrisome sign.

Family History:

Ask about any family history of seizures, mental retardation, or other neurological disorders.

Specific known risk factors that should be considered:

Prenatal	Perinatal	Postnatal
Maternal Factors: - thyroid disorder - mental retardation - seizure disorder - iodine deficiency - previous pregnancy loss - toxin exposure **Pregnancy Factors:** - congenital malformations - infections - polyhydramnios - treatment of mother with thyroid hormone, estrogen, or progesterone - severe maternal proteinuria or high blood pressure - third trimester bleeding - multiple gestations	- prematurity (< 32 wks) - birth weight <2500g - nonvertex and face presentations - chorioamnionitis - birth asphyxia - meconium aspiration - pneumothorax - Rh incompatibility - Placental abruption	- infections (e.g., meningitis or encephalitis) - parenchymal or intraparenchymal hemorrhage - jaundice causing kernicterus - prolonged ventilation - hypoxic ischemic encephalopathy (HIE)

Differential Diagnosis

The diagnosis of cerebral palsy can be challenging because although CP is defined as a static condition, clinical findings change as the nervous system matures. The diagnosis involves a combination of findings including various neurological signs, motor delay, abnormal postures and persistence of primitive reflexes. Remember, CP is a diagnosis of exclusion. Your differential should broadly include:

- Neurological disorders
    - Hereditary neurodegenerative diseases – usually presents as milestone regression
    - Rett syndrome – a neurodevelopmental disorder that occurs primarily in females in which development is initially normal for at least 6 months. This is followed by progressive loss of language, purposeful hand movements, deceleration of head growth and development of stereotypic hand movements (wringing/washing). There is severe regression of motor milestones.
    - Tethered Spinal Cord – presents as lower limb spasticity with deficits to bowel and bladder function.
    - Disorders of muscle weakness
    - Disorders of ataxia – such as ataxia telangiectasia and Nieman-Pick disease
    - Bulbar and oromotor dysfunction
- Metabolic disorders
    - Glutaric aciduria type 1 – may present with dystonia and choreoathetosis
    - Urea cycle disorders – may present with diplegia and/or quadraplegia
- Endocrine disorders
    - Thyroid Dysfunction
- Malignancy
    - Intracranial neoplasms (may also present with focal neurological deficits)

Physical Exam:

Physical exam should always begin with general appearance, vital signs and growth parameters. A very thorough neurological examination is required to aid with the diagnosis of CP.

General appearance: dysmorphic features, FTT, abnormal postures, asymmetry of movement

Head & Neck: cataracts or retinopathy (2º to congenital infections), microcephaly, visual field deficits, strabismus, EOMs, hearing impairment, dental caries

Respiratory: respiratory distress, signs of (children with CP are often prone to recurrent respiratory infections)

CVS: congenital heart defects (2º to congenital infections)

Abdo: stool masses

MSK: limb leg discrepancy (2º to hemiplegia), early hand preference (before 2 yrs), involuntary movements (athetosis, chorea, dystonia), gait abnormalities, joint contractures, asymmetry of voluntary limb movements, scoliosis, limitations in range of motion

Neurological: Complete a full neurological exam:

- Cranial nerves – pay attention to oculomotor and oromotor functions

- Sensory – difficult in a newborn bit try to assess tracking and hearing
- Motor – muscle bulk, tone, power
    - Tone may be increased with clasp knife response or decreased with extreme hypotonia
    - Assess for fisting (hands tightly clenched in a fisting with the thumb tucked inside the fist) – normally infants should not be fisted all the time
- Reflexes – deep tendon and primitive reflexes
    - Pay particular attention to persistence of primitive reflexes (Moro, rooting, etc) and hyperreflexia, extensor plantar response beyond the appropriate age
- Cerebellar - assess for any ataxia, involuntary movements
- Gait and Posture – assess for abnormal movements

Derm: assess for presence of any neurocutaneous stigmata and pressure ulcers

Investigations:

- Labs:
    - Basic metabolic work-up to rule out an underlying metabolic disorder including glucose, ammonia, lactate, blood gas, plasma amino acids and urine organic acids
    - TSH to rule out thyroid abnormality
    - Chromosomal analysis if genetic syndrome suspected

- Imaging studies:
    - EEG if you want to rule out seizures
    - MRI is recommended to identify structural lesions and contribute to defining a cause of lesion, best after 2-3 weeks of age
    - Head CT can identify some malformations, hemorrhrage, or periventricular leukomalacia. Recommended if MRI unavailable.
    - Spinal MRI may be indicated if tethered spinal cord needs to be ruled out.

Conclusion:

In summary, cerebral palsy is a diverse group of syndromes characterized by motor delay, postural disorders, and neurologic dysfunction that is non-progressive though the clinical picture can change as the brain matures. There are a variety of associated signs and symptoms and diagnosis requires a thorough history and physical examination. There are a variety of underlying etiologies and a wide range in severity making CP difficult to diagnose.

Children with cerebral palsy require a complex health care team including the family, paediatrician, family physician, orthopaedic surgeon, neurologist, physiotherapists, occupational therapist and often respite care for severe cases. Close monitoring and follow-up with these children with give them the best quality of life they can achieve.

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