

Cerebral Palsy (CP)

Competency:

Know clinical signs and usual timing of symptoms associated with the cerebral palsy syndromes
Know risk factors for developing cerebral palsy in very low birth weight infants
Know risk factors for developing cerebral palsy in term/near term infants
Know which history and exam findings warrant referral to a specialist and further investigation
Know common morbidity, habilitative interventions and supports for a child with cerebral palsy.

Case: In a new patient clinic, you see a 15 month old girl for a well-child exam. She has recently moved to IL from another state. Your history and exam are significant for delayed motor milestones, and increased tone in both lower extremities, and some scissoring of lower extremities upon vertical suspension. You suspect that child has diplegic CP, and start talking to child's mom about details of child's past history and development. Child's mother has several questions along these lines:

Questions:

1. What are the common spastic CP syndromes by limb distribution?
2. What are the characteristics of the extrapyramidal CP syndromes (choreoathetosis, dystonia, ataxia)?
3. What are the common causes in preterm infants?
4. What are the common causes in term/near term infants?
5. What elements of history and exam should make a physician suspect CP in an apparently healthy child?
6. What is the differential diagnosis of motor delay?
7. What evaluation should a child with suspected CP have?
8. What management options are available for children with CP?
9. What is the prognosis for function, i.e. sitting, walking, self-care, communication, and learning for children with CP?
10. What medical conditions are children with CP at risk of developing?

References:

1. Ashwal, S. et al. Practice Parameter: Diagnosis assessment of the child with cerebral palsy. *Neurology*, 62. March (2 of 2), 2004: 851-863.
2. Fenichel, G. *Clinical Pediatric Neurology: A Signs and Symptoms Approach*. 5th edition. WB Saunders Co., 2005
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5. Goldstein M The Treatment of Cerebral Palsy: what we know, what we don't know. *J Pediatr* 2004;145:S42-46
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9. Accardo, J et al. Neuroimaging in Cerebral Palsy (*J Pediatr* 2004;145:S19-S27)

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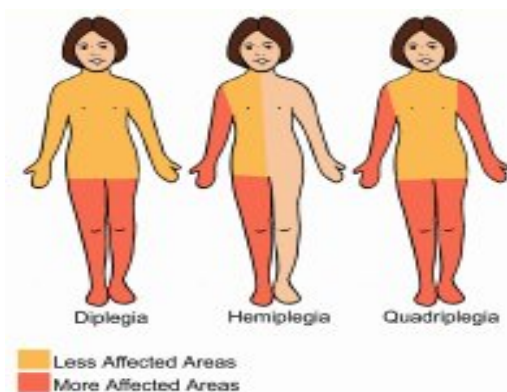
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Introduction

Cerebral palsy is a common problem and cause of disability. The worldwide incidence of CP is approximately 2-2.5 per 1,000 live births. Each year about 10,000 babies born in the United States develop Cerebral Palsy. This condition occurs more commonly in very preterm babies (10-20% ELBW, 5-10% VLBW, 3-5% LBW) especially those with IVH3-4, PVL, and ventriculomegaly. 95% of babies are born term (>37 weeks gestation), 4% are born moderate preterm at 32-36 weeks gestation, and 1% are born very preterm or <32 weeks gestation. Thus, the average child who develops CP was either term or a "big" preterm, i.e. >1500 g. Current evidence suggests that 70-80% of CP cases are due to prenatal factors, and that birth asphyxia accounts for <10% of Cerebral Palsy.

In Western Australia, which has one of the best CP registries, **Hemiplegia** was present in 36%, **Diplegia** in 32% and **Quadriplegia** in 16%. Ataxia was present in 7%, and extrapyramidal symptoms in 9.2%. Half of the individuals with CP use assistive devices such as braces, walker or wheelchairs, and almost 70% have other disabilities, primarily communicative, cognitive, learning, and attentional disorders. There are high rates of seizures, strabismus, hearing loss, and poor growth.



1. What is CP and what causes it?

CP is a disorder of aberrant control of movement and posture, appearing early in life secondary to a CNS lesion or dysfunction that is NOT a result of a progressive or degenerative brain disease. In other words, CP is caused by a *static* brain dysfunction (also called static encephalopathy).

It is believed that CP is due to a defect in a developing brain, a lesion that may have had its onset in prenatal, perinatal, or postnatal period. Thus, causes of CP may be manifold.

Here are some common etiologies manifesting as CP:

<u>Prenatal (~85% of total)</u>	<u>Perinatal</u>	<u>Postnatal</u>
extreme prematurity	IVH grade 3-4, PVL	infection
hyperarginemia		
genetic disease	infection	trauma
brain malformation	hypoxic-ischemic encephalopathy	
hydrocephalus	trauma	CO poisoning
meningitis	kernicterus	toxic ingestion
stroke		
toxemia		
maternal coagulopathy		
placental abruption		

4 types of cerebral palsy include:

Spastic

(70-80% of all) Defined as increased muscle tone of clasp knife quality, increased reflexes with tendency to clonus, and tendency for contractures;

Flexion contractures at elbow, difficulty with pronating or supinating forearms;

Difficulty with long sitting because of hamstring contractures, difficulty changing diapers because of decreased range in abduction of hips;

Scissoring in lower extremities, toe walking gait

Caution: idiopathic toe walking with ability to walk, climb and run is more often a marker for a developmental disability or autism, or congenital tight Achilles tendons.



Athetoid (10-20% of all) Defined as dominated by athetoid involuntary movements of hand, feet, arms, muscles of the face or tongue. Movements are usually writhing in nature, exacerbated by stress, disappear in sleep.

Hypotonic/Ataxic (5-10% of all) Least common type of CP, manifested by poor coordination, unsteady gait, difficulty with rapid or precise movements.



The 4th type is **mixed CP**: the most common form includes spasticity and dystonic/athetoid movements.

2. What elements of history and physical exam make a physician suspect CP in an apparently healthy child?

The diagnosis of CP always involves **gross motor delay** with an abnormal neurological exam, and the usual presenting complaint for which medical evaluation is sought is that the child is not reaching motor milestones at the appropriate chronological age. (e.g. no rolling over, not sitting, not crawling, not walking) It is critical to establish that the child is not continuing to lose function, assuring that the patient **does not have a progressive disease** (e.g. hydrocephalus, leukodystrophy, stroke). Thus, a history of losing motor or any other developmental milestones is *inconsistent with CP*, and further investigation is warranted to establish treatable causes of CP, especially hydrocephalus and posterior fossa processes.

Of note, serial examinations may be necessary to assure the diagnosis of CP, especially when history is not reliable. Another important but often overlooked symptom of CP is **abnormally early hand dominance** (within the 1st year of life). Most children with CP come to pediatrician's attention before 2 years of age.

Common physical findings in affected limbs include:

- Fisted hands, Flexed arms, Difficulty with pronation or supination
- Scissoring of lower extremities, Crouched gait, Equinovarus feet (club foot)
- Spasticity : clasp knife increased tone, esp. upon fast movement of extremity
- Abnormally brisk reflexes, possibly clonus, usually at the ankle
- Upgoing toes (positive Babinski sign)
- Dislocated/subluxed hip joints from unopposed increased tone in hip adductor
(This is perhaps the most important deformity for the general pediatrician to detect!)

3. What evaluation should a child with suspected CP have?

- Detailed history, including birth, family, developmental history, and physical exam
- Referral to a developmental specialist or child neurologist for confirmation of diagnosis
- Referral to early intervention. Remember that all children with cerebral palsy learn.
- Support for the family
- Be cautious in attributing causality to hypoxic ischemic encephalopathy

- **Neuroimaging?**

CT scan (77% abnormal yield) –useful if abnormal, as it often reveals a potentially treatable lesion (e.g. Hydrocephalus, AVM)

MRI (89% abnormal yield)—higher abnormal yield than CT, useful to pinpoint timing of brain injury (pre/peri/postnatal), often suggests etiology.

- **Metabolic and genetic testing?**

Not routinely recommended since metabolic or genetic causes for CP are rare (0-4% of all cases). However, if history or neuroimaging findings are atypical and do not point to a specific structural abnormality, metabolic and genetic testing should be considered. (Hyperarginemia, glutaric aciduria, and homocystinuria are among common treatable metabolic causes of cerebral palsy).

All children with ataxia benefit from a MRI with visualization of posterior fossa and cervical spinal cord.

- **Coagulation studies?**

Children with **hemiplegic CP** often have a **stroke episode** as an etiology. In children, the most common causes of stroke are: hypercoagulable state, congenital heart disease, and infection. Current evidence suggests that for children with suspected *hemiplegic CP*, coagulations studies are useful in determining the cause of CP.

4. What management options are available for children with CP?

- Monitoring of growth, nutrition, vision, hearing, sensory skills
- Physical and occupational therapy to promote position for function, use of equipment, and developmental manipulative skills
- Botulinum toxin injections to reduce spasticity
- Phenol injections to reduce spasticity
- Baclofen infusion to reduce spasticity in dystonic and quadriplegic cerebral palsy
- Braces and/or tendon release surgery to increase function of lower limbs
- Selective posterior rhizotomy in select cases of diplegic CP

5. What is the prognosis for function, i.e. sitting and walking for children with CP?

NB! Prognosis for function in the future is very dependent on age and ability level of the patient at the time of diagnosis.

If not sitting at all by age 4, 99% will not stand/walk?

If not having head control by age 1, very unlikely by stand/walk?

If sitting by age 2, 100% likely to stand/walk?

6. What are the medical conditions that children with CP are at risk for developing?

- Mental retardation, esp. in children with quadriplegia(52%)
- Epilepsy (45%)
- Speech and Language disorders (38%)
- Vision defects (28%)
- Hearing defects (12 %)
- Oral-motor dysfunction, dysphagia, and GERD requiring surgical correction

Children with hemiplegia and diplegia have as their major challenge communicative, learning and attentional disorders. Failure to address these issues is a major barrier to chances for independent living as a young adult. It is in this area that pediatricians can make a difference.

References:

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