The Child With Cerebral Palsy

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Cerebral Palsy is not a disease

- It’s a symptom complex or “group of disorders”
- Causing activity limitation
- Attributed to a non-progressive disturbance
- Occurred in the developing fetal or infant brain (in general the disturbance resulting in CP is presumed to occur before the affected function has developed.
- Often accompanied by disturbances in sensation, cognition, communication, perception, perception, orthopedic issues, and/or behavior, and/or by a seizure disorder.
- While the brain lesion is static, the course of the condition is not
The most common motor disability of childhood

- 3.6 per 1000 live births
- 10,000 new cases per year
- 80% antenatal factors
Most common motor disability of childhood

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Spina Bifida</td>
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<tr>
<td>Autism</td>
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<tr>
<td>Brain Injury</td>
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<tr>
<td>Cerebral palsy</td>
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KEY FEATURES OF THE EPIDEMIOLOGY OF CP

- Found in about 1 in 500 school children
- Not as closely associated with “birth asphyxia” as once thought
- Increasing evidence for a role of antepartum infection
- Transient hypothyroxinemia may harmful?
- Lots of room for more research!
Epidemiology

- 5.2 per 1000 neonatal survivors at 12 months of age
- By age seven 2.5 per 1000 live births (but 10 times more likely to be retarded)
  - Nelson & Ellenberg *Pediatrics* 69:5, 1982
- 8000 new cases per year
- The overall prevalence of CP in 1-year survivors increased from 1975-1991 by 18%
- Inflammatory cytokines released during the course of intrauterine infection play a central role in the genesis of preterm parturition, fetal PVL, and cerebral palsy
What causes CP?
We’re not always sure……

- The rate of CP does not exceed 2% following placenta previa, abruptio placenta, breech, cord prolapse, nuchal cord, mid or high forceps delivery
- 75% of children with CP have normal Apgar scores
- Most children with an Apgar score of 3 at 10 and 15 minutes do not have CP
MAYBE IT’S BIRTH COMPLICATIONS?
## DO COMMON LABOR COMPLICATIONS CAUSE CP?

<table>
<thead>
<tr>
<th>Complication</th>
<th>CP Risk</th>
</tr>
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<tbody>
<tr>
<td>No complication</td>
<td>0.3%</td>
</tr>
<tr>
<td>Nuchal cord (18%)</td>
<td>0.3%</td>
</tr>
<tr>
<td>2nd stage ≥ 1 hour (10%)</td>
<td>0.3%</td>
</tr>
<tr>
<td>Meconium (20%)</td>
<td>0.4%</td>
</tr>
<tr>
<td>Mid or high forceps (8%)</td>
<td>0.4%</td>
</tr>
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Nelson KB, Ellenberg JH: JAMA 1984;251:1843-8
**DO UNCOMMON LABOR COMPLICATIONS CAUSE CP?**

<table>
<thead>
<tr>
<th>Complication</th>
<th>CP Risk</th>
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<tr>
<td>CORD PROLAPSE</td>
<td>0.3%</td>
</tr>
<tr>
<td>PLACENTA PREVIA</td>
<td>0.6%</td>
</tr>
<tr>
<td>BREECH POSITION</td>
<td>1.0%</td>
</tr>
<tr>
<td>ABRUPTIO PLACENTAE</td>
<td>1.9%</td>
</tr>
</tbody>
</table>

Nelson KB, Ellenberg JH: *JAMA* 1984;251:1843-8
DO LABOR COMPLICATIONS MATTER AT ALL TO CP RISK?

- **NO LABOR COMPLICATIONS**
  - CP RISK: 0.3%

- **ANY LABOR COMPLICATIONS**
  - CP RISK: 0.3%
APGAR SCORES AND CEREBRAL PALSY

WHAT ABOUT BIRTH ASPHYXIA CAUSING HYPOXIC ISCHEMIC ENCEPHALOPATHY (HIE)?
Not so much.....

- Intrapartum asphyxia: a rare cause of CP
  - Blair & Stanley J Peds April 1988
- 1975-1980 All children with spastic CP born in Western Australia (n=183)
- Matched group of controls (n=549)
- Info on perinatal events was collected
- Birth asphyxia \textit{was} associated with CP (RR 2.84; 95\% confidence interval)
- BUT of all children with spastic CP intrapartum asphyxia was the possible cause of their brain damage in only 8\% (15 of 183).
HOW ABOUT PREMATURITY?
PREMATURITY IS THE MOST IMPORTANT KNOWN ANTECEDENT
40-fold higher risk in infants < 1,500 g
Possibly increasing in prevalence due to increased VLBW survival
IVH (grades III and IV) is an important cause
The brain lesion resulting after IVH is periventricular leukomalacia (PVL)
PVL is the most important identifiable risk factor in the development of CP
BUT MOST KIDS ARE BORN AT TERM WITHOUT COMPLICATIONS.
Risk of CP and gestational age
But there are some risk factors

- Low birth weight
- Prematurity (<1500 grams)
- Maternal MR
- Maternal sz
- Previous pregnancy loss
- Multiple pregnancies
- Motor deficit in an older sibling
- Male gender
- Infections (rubella, toxoplasmosis, CMV, etc)
- Intrauterine infection w/chorioamnionitis
- Jaundice (kernicterus)
- Asphyxia
- Maternal thyroid deficiency (*Bernal and Nunez 1995*).
- Non-vertex presentation
- Fetal inflammatory response has been found to be related to white matter injury and CP

(Bernal and Nunez 1995).
Diagnostic clues but certainties

- Failure to meet expected developmental milestones
- Failure to suppress obligatory primitive reflexes.
- Abnormal muscle tone: hypotonic / hypertonic. Often will have early hypotonia followed by hypertonia as the brain myelinates.
- Definite hand preference before age 1 year
- Asymmetric crawling or failure to crawl
- Growth disturbance especially failure to thrive
- Persistent primitive reflexes
Persistent primitive reflexes - ATNR
Persistent Primitive Reflexes - Moro
Differential

- Hereditary spastic paraplegia
- Rett syndrome
- Tethered spinal cord
- Genetic d/o (named and unnamed)
  - Brown Vialetto VanLaer
  - Aicardi
- Charcot Marie Tooth Disease
- Spinal Muscular Atrophy
- Spina Bifida
- Polio
- Myopathy
Mean Age for Diagnosis

- Quadriplegia - 5 months
- Diplegia - 12.5 months
- Hemiplegia - 21 months
Classification based on topography

- Monoplegia: one limb involved
- Diplegia: primarily legs are involved
- Hemiplegia: one side of body involved
- Quadriplegia: all four limbs and trunk involved
Classification based on tone disorder

- Spasticity: velocity-dependent increase in tone
- Athetosis: slow, writhing, involuntary movements especially in distal extremities
- Chorea: abrupt, irregular, jerky movements usually in the head, neck, and extremities
- Dystonia: slow, rhythmic movements with tone changes generally found in the trunk and extremities
- Ataxia: unsteadiness with uncoordinated movements; wide-based gait
Associated Deficits

- Mental retardation: 30-50%
- Seizures: half of all CP, but 70% of hemiplegics
- Oromotor: sucking, swallowing, excessive drooling, articulating, poor dentition, FTT
- Pulmonary - aspiration
- GI: reflux, constipation
- Ocular/Visual: strabismus, refractory errors
- Perceptual deficits
- behavioral disorders
- bladder/bowel control
Is this CP?
Is this CP?

Hypotonia (decreased muscle tone)
Is this CP?
Do Musculoskeletal Complications Help in Dx?

- Caused by imbalance between agonists and antagonists
- Muscles in a growing child tend to shorten if not fully stretched daily
- Progression may be slowed by consistent proper positioning
Do Contractures Help in Dx?
How about spasticity?
Is this CP?
Is this CP?
Diagnostic Testing

- Neuroimaging (MRI preferred) to determine the presence and nature of any brain abnormality (normal results do not exclude CP)
- Possible genetic evaluation to assure that the condition is not progressive
- Because of the high incidence of associated conditions screen for:
  - mental retardation
  - ophthalmologic disorders
  - hearing impairments
  - Speech and language disorders
What’s not CP?

- Anything degenerative
- If there are no tone abnormalities (idiopathic toe walking)
- If it’s a genetic syndrome (Aicardi syndrome, Palizaeus-Merzbacher)
- If it’s not caused by a brain lesion (spina bifida or spinal cord injury)
Children, on average, reach about 90% of their motor function (as measured by the GMFM-66) by around age 5 years or younger, depending on their GMFCS level.

- A: lift and maintain head in a vertical position with trunk support by a therapist while sitting;
- B: when in a sitting position on a mat, a child can maintain sitting unsupported by his/her arms for 3 seconds;
- C: ability to walk forward 10 steps unsupported;
- D: walking down 4 steps alternating feet with arms free.
THE END