CEREBRAL PALSY: An Integrated Approach

Michael J. Ward, MD
Associate Professor, CHS Orthopedics and Rehabilitation Medicine
University of Wisconsin Medical School
November 5, 2016
WHAT IS CEREBRAL PALSY?
Modern consensus definition:

- Group of disorders of movement and posture
- Non-progressive etiology
- Damage to the fetal or infant brain
- Often accompanied by co-occurring problems with sensation, perception, communication, and/or behavior and/or seizure disorder

Bax 2005 DMCN
Diagnosis of Cerebral Palsy has 4 requirements:
1. Non-progressive impairment
2. Immature or developing
3. Brain (*cerebral*)
4. Abnormal motor development (*palsy*)
DIAGNOSIS:
Non-progressive

Excludes conditions which cause ongoing brain injury over time: neurodegenerative disorders

Also excludes conditions which resolve
DIAGNOSIS: Non-progressive

However, symptoms can transform through the life span even when the primary brain injury has not worsened or improved since birth

*CP is non-progressive, but not unchanging*
DIAGNOSIS: Immature or developing brain

When does development end?
- Embryonic formation of organs
- Birth
- 1 year: common end point for CP diagnosis
- 2-3 years: Brain myelination completed
- 7-9 years: Maturation of motor skills
- 16-18 years: Physical maturity
- Social maturity
DIAGNOSIS:
Immature or developing brain

Brain injury causing cerebral palsy usually occurs before birth or shortly after...

...and sometimes we do not know when it happens.
DIAGNOSIS: Immature brain

Presentation of symptoms in CP:

- Sometimes noted right after birth
- Typically by 6-12 months
- Mild cases may not be noticed until 12-18 months
DIAGNOSIS: Brain impairment

Excludes motor problems from diseases of:

EXAMPLES

<table>
<thead>
<tr>
<th>Spinal cord</th>
<th>Spina bifida</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscles</td>
<td>Muscular dystrophy</td>
</tr>
<tr>
<td>Nerves</td>
<td>Spinal muscular atrophy</td>
</tr>
</tbody>
</table>
DIAGNOSIS: Etiology

Includes many causes of early brain injuries:

- PVL: Brain damage with prematurity
- Birth hypoxia: Lack of oxygen to whole brain
- Brain malformation: Abnormal pattern of cell growth
- Prenatal stroke: Blood supply interruption
- Encephalitis: Brain infection, reaction to infection
- Hyperbilirubinemia: Jaundice
- Other
DIAGNOSIS: Etiology

Can be caused by a combination of factors

Occasionally the factors are never known
DIAGNOSIS: Etiology

Most common etiology:

Complex series of events in the brain set in motion after birth among newborns with prematurity and very low birth weight

Currently largest single etiology of cerebral palsy
DIAGNOSIS: Etiology

Prematurity and low birth weight associated with PERIVENTRICULAR LEUKOMALACIA:

Peri = around
Ventricular = deep brain fluid spaces
Leuko = white matter
Malacia = thinning
DIAGNOSIS:
MRI with Periventricular leukomalacia

Normal brain

PVL
Cerebral Palsy: Cranial imaging findings

- PVL
- Gray matter
- Basal ganglia
- Malformation
- Miscellaneous
- Normal

Bax JAMA 2006
DIAGNOSIS: Disturbance of motor development

CP is usually described by type of motor problem

Spastic types most common, and described by distribution

- Quadriplegic: both arms and both legs
- Hemiplegic: Arm and leg on both sides
- Diplegic: Both legs more impaired than both arms
DIAGNOSIS

Disturbance of motor development

CP is usually described by type of motor problem

Other types:

- Dyskinetic: abnormal involuntary movements
  - Dystonia
  - Athetosis
  - Chorea
- Ataxic: coordination issues

Many people have more than one movement challenge
DIAGNOSIS: Types by motor pattern

- Extrapyramidal
- Other
- Diplegic
- Quadriplegic
- Hemiplegic
DIAGNOSIS: Disturbance of motor development

There is *partial* correlation between etiology and type of motor problem:

**ETIOLOGY**

**MOTOR TYPE**
DIAGNOSIS: Disturbance of motor development

There is \textit{partial} correlation between etiology and type of motor problem:

\begin{tabular}{ll}
\textit{MRI abnormality} & \textit{Motor problem} \\
PVL & Diplegia \\
Birth Hypoxia & Quadriplegia and dystonia \\
Prenatal stroke & Hemiplegia
\end{tabular}
DIAGNOSIS:
MRI with Periventricular leukomalacia

Normal brain  PVL
MOTOR DELAYS: GMFCS
Gross Motor Classification System

Track curves of motor development in children with CP from early milestones to adult skills achievement.

Predicts general trends at 5 functional levels
MOTOR DELAYS: GMFCS

Level I: Walks without limitations
Level II: Walks with limitations
Level III: Ambulation with device only
Level IV: Limited mobility, power wheelchair
Level V: Dependent manual wheelchair
This graph shows the observed and predicted GMFM-66 scores for children in GMFCS Levels I through V. The curved solid lines indicate average performance. The horizontal dotted lines on the right of the figures indicate the band expected to encompass 50% of children’s limits of development. The solid vertical lines indicate the average age (in years by which children are expected to reach 90% of their motor development potential). The dotted vertical lines indicate the bands expected to encompass 50% of age-50 values around the average. The absence of 50% bands in level IV and level V indicates low variation in age-50 values.

MOTOR DELAYS: REHABILITATION INTERVENTIONS

Physical therapy
Orthopedic surgery
Spasticity reduction
Casting/splinting
Bracing
Mobility aids

*Help but do not change the GMFCS level (usually)*
DIAGNOSIS:
Disturbance of motor development

Required for diagnosis

*CP is not an exclusively motor condition*
Modern consensus definition:

- Group of disorders of movement and posture
- Non-progressive etiology
- Damage to the fetal or infant brain

- Often accompanied by co-occurring problems with sensation, perception, communication, and/or behavior and/or seizure disorder

Bax 2005 DMCN
CEREBRAL PALSY
Associated concerns

Cognitive
– Cognitive impairment 40-60%
– Learning disabilities common
– Attention deficit disorder
– Other behavioral challenges
– Language disorders
CEREBRAL PALSY
Associated concerns

Sensory abnormalities:
- Hearing loss 7-12%
- Abnormal control of eye motions 20-60%
- Visual impairment overall 80%
- Visuoperceptual abnormality also frequent
- Tactile impairment 50-75%
- Balance system impairment
- Sensory integration challenges
CEREBRAL PALSY
Associated medical concerns

Seizures 30-50%
CEREBRAL PALSY
Associated medical concerns

Autonomic nervous system also affected:

- Abnormal digestive motility
- Temperature instability and cold or hot limbs
- Sweating changes
- Bladder dysfunction
- Breathing irregularities
- Sleep disorders
CEREBRAL PALSY
Associated concerns

Secondary problems: Gastrointestinal
- Swallowing difficulties
- Malnutrition
- Growth delays
- Gastric reflux
- Constipation
- Drooling
- Dental changes
CEREBRAL PALSY
Associated concerns

Many orthopedic complications:
- Abnormal hip growth
- Osteoporosis and fractures (even in children)
- Scoliosis
- Joint limitations
- Musculoskeletal pain
Combining all of this provides a more complete description of CP:

<table>
<thead>
<tr>
<th>Type</th>
<th>Spastic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distribution</td>
<td>Quadriplegic</td>
</tr>
<tr>
<td>Etiology</td>
<td>VLBW and prematurity</td>
</tr>
<tr>
<td>MRI Imaging</td>
<td>Periventricular leukomalacia</td>
</tr>
<tr>
<td>Functioning</td>
<td>GMFCS V</td>
</tr>
<tr>
<td>Associated</td>
<td>Cognitive, visual, orthopedic, etc.</td>
</tr>
</tbody>
</table>
DIAGNOSIS:
MRI with Periventricular leukomalacia

Normal brain

PVL
Cerebral Palsy: Goals of an Integrated Approach

Understand the whole person
Address all types of questions
Evaluate unique medical issues with CP