Cerebral Palsy

_Cerebral:_ an adjective meaning of or pertaining to the brain

_Palsy:_ a noun meaning atonal muscular conditions characterized by _tremors_ of the body parts

Cerebral Palsy: What is it – really?

- **Static Encephalopathy**
  - There is a _lesion in the brain_ that results in a _non-progressive motor disability_.
  - The causative event must occur early in childhood, usually _before age 2_.
  - The spinal cord and muscles are structurally normal.

So what’s the goal?

Can you pick out the adult with CP?

The goal for these children, for their families, their medical care, their education, and society is for them to grow and develop to their _maximum capabilities_.

Disclosures

- I have no disclosures.
How is the child with CP different?

• In addition to typical childhood experiences, these kids go through (and have to deal with):
  – Multiple surgical procedures
  – Multiple hospitalizations
  – Physical therapy for years
  – Struggles for independence
    • From their bodies
    • From their parents

Family impact of a child with CP

• There are as many variations among families as there are manifestations of CP.
• “Families” can include single parents, teenage parents, caregiver-grandparents, and those with very little support.
• There is a high level of divorce amongst parents of kids with CP.

The whole picture

• The medical team must keep all of this in mind as they treat the whole child and their family.
• This person will eventually become an adult – how can you help them become the best adult they can be?

Isn’t CP going away?

• The incidence of cerebral palsy is increasing slightly.
  – It is between 2.4 and 2.7 per 1000 live births.
  – As technology has improved, there are fewer problems with deliveries, but some do still exist.
  – As technology has improved, the number of infants with very low birthweight that survive has increased.

A bit about cerebral palsy – from an orthopaedic surgeons perspective

Incidence
Classification

Classification

1. Geographic

2. Physiologic
Geographic Classification

Physiologic Classification

- **Spasticity**
  - damage to the motor cortex
  - disinhibition of pathologic reflex arcs leading to increased tone
  - velocity dependent
    - rapid stretch → high tone
    - slow stretch → less tone
- **Hypotonia**
  - abnormally decreased tone
  - more commonly seen in infancy, it can develop into spasticity
- **Dystonia**
  - increased tone
  - not velocity dependent
  - lead pipe: tone does not decrease with slow stretching

- **Athetosis**
  - abnormal writhing movements
  - damage to the basal ganglia
  - difficulty with speech, but can have normal intelligence
  - neonatal hemorrhages
- **Ataxic**
  - cerebellar lesion → disturbed balance, wide-based gait
  - Mixed movement disorder
    - Don’t miss this! The results of surgical treatment are unpredictable for all but purely spastic patients.
  - Go to video...

Orthopaedic Problems in CP

- **Spasticity**
  - Scoliosis
  - Contractures of joints
    - Planovalgus and cavusfeet, clubfeet, bunions
    - Knee flexion contractures
    - Adductor and ilipsoas contractures
  - Gait abnormality
  - Hip dislocation
  - Poor upper extremity function
  - Postural difficulty

That leaves us with... Spasticity!

- This is the underlying problem of almost all other orthopaedic manifestations.
- Remember these?
  - There is a lesion in the brain that results in a non-progressive motor disability
  - The spinal cord and muscles are structurally and biochemically normal

Spasticity

- Tight, stiff muscles or spasms that may make movement, posture, and balance difficult.
- Affects the ability to move limbs, or to move one side of the body.
- Sometimes spasticity is so severe that it gets in the way of daily activities, sleep patterns, and caregiving.
- Spasticity can be painful.
Evaluating Spasticity: Ashworth Scale

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increase in muscle tone</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in muscle tone, manifested by a catch and release, or by minimal resistance at the end range of motion when the affected part is moved in flexion or extension</td>
</tr>
<tr>
<td>1+</td>
<td>Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the range of motion</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in muscle tone through most of the range of motion, but the affected part is easily moved</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in muscle tone, passive movement is difficult</td>
</tr>
<tr>
<td>4</td>
<td>Affected part is rigid in flexion or extension</td>
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</tbody>
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How is spasticity treated?
- Rehabilitation Therapy
- Oral Medication
- Neurosurgeries
- Injection Therapy
- Orthopedic Surgery
- ITB Therapy (Baclofen Pump)

Rehabilitation Therapy
- Physical or occupational therapy can include:
  - Stretching of muscles
  - Gait training
  - Splinting (disuse therapy)
  - Work on functional tasks
  - Provide some limited bracing

Oral Medication
- GABA receptors are the main inhibitors of the motor control system in the brain and spinal cord
- Baclofen – an analog of GABA that binds to the receptors but does not activate GABA
  - Works in the CSF (BBB)
  - Hard to build a “tolerance”
- Diazepam

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Oral Medication

- Chlorazepate
- Clonazepam
- Ketazolam
- Trazodone
- Dantrolene
- Tizanidine
- Clonidine
- Cannabis
- Cyclobenzaprine

Neurosurgery

- Dorsal Rhizotomy
  - Cut the dorsal sensory nerve rootlets
  - Contain afferent sensory nerves from the muscle spindles
  - Can be very effective if the right nerve roots are sectioned
  - Can be very bad if the laminectomy produces hyperlordosis

Injection Therapy

- Botox
  - Competitive inhibitor of presynaptic cholinergic receptor with a finite lifetime
  - Binds irreversibly
  - Used to maintain joint motion during rapid growth when surgery is inappropriate
  - Works in an area of 3cm from injection site, lasts 3-4 months
- Alcohol or phenol denervation

Orthopaedic Surgery

- Contractures
  - Due to a muscle that has not grown sufficiently to its anatomically required length
  - Does it actually treat spasticity or just the secondary effects?
  - Actually, hyperreflexia is dependent on a certain amount of tension, which is reduced when the muscle is lengthened
  - This can help with gait, but beware the overlengthening
- Bone surgery
  - Derotational osteotomies
  - Extension or flexion osteotomies
  - Guided growth
  - Bone shortening
  - Varus or valgus osteotomies

Intrathecal Baclofen

- Baclofen can also be given intathecally
- Already across the BBB
- 30mg TID (90mg) oral = 300mcg/day IT
- Anterior pump → posterior catheter
- Test dose
- Less pain

Intrathecal Baclofen

- Who is a candidate?
  - Those with spasticity of cerebral origin (at least 1 year post TBI)
  - Children who have had ineffective or intolerable side effects of oral baclofen
Benefits of ITB Therapy

- Reduced spasticity\(^1,2\) and spasms\(^3,8,9\)
- Increased independence allowing people to better perform activities of daily living including feeding or dressing themselves, sitting more comfortably, or transferring more easily\(^10-13\)

Contraindications

ITB Therapy and implantation of the SynchroMed programmable pump are contraindicated:

- In the presence of infection or spinal anomalies
- When the pump cannot be implanted 2.5 cm or less from the surface of the skin
- In patients whose body size is not sufficient to accept the pump bulk and weight
- In patients with hypersensitivity to baclofen

Safety and effectiveness in pediatric patients below the age of 4 have not been established.

Patient Selection

- Appropriate patient selection is essential to therapeutic success
- Consider ITB Therapy for patients who:
  - Have severe spasticity of cerebral or spinal origin\(^1\)
  - Demonstrate positive response to a single bolus dose of Lioresal\(^\text{tm}\)
    Intrathecal (baclofen injection) during the screening test\(^1\)
  - Are refractory to oral baclofen or experienced intolerable side effects at effective doses\(^1\) (spasticity of spinal origin)
  - Have experienced traumatic brain injury at least 1 year prior to being considered for long-term ITB Therapy\(^2\)
  - Have sufficient body mass to support the bulk and weight of the pump\(^2\)

\(^1\) Lioresal Intrathecal package insert
\(^2\) Medtronic information for prescribers for SynchroMed pump