Cerebral Palsy Case Review

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Clinical Scenario

• T.M. is a 26 month old boy who has motor delay. He has been in physical therapy for 3-4 months and is making progress, but is not walking independently yet. His pediatrician and physical therapist recommend he gets an evaluation for Cerebral Palsy (CP).
What is CP?

- A heterogeneous group of permanent, non-progressive clinical syndromes characterized by dysfunction of movement, muscle tone, and posture
- The result of abnormalities to the developing fetal or infant brain
- Can be accompanied with disturbances of sensation, perception, cognition, communication and behavior, epilepsy and secondary musculoskeletal problems
Who does CP affect?

• It is the most common cause of childhood disability- occurs in 2/1000 live births
• It has a much higher prevalence in preterm infants compared to term infants; infants born at less than 28 weeks are at most risk
• The prevalence is also higher for low birth weight infants, with those born weighing less than 1500 g at greatest risk
How is CP diagnosed?

• CP is a clinical diagnosis and is based on:
  – Clinical history AND
  – A combination of clinical findings: motor delay, persistence of primitive reflexes, abnormal postural reactions, and tone abnormalities
• Clinical signs evolve as the nervous system matures
• Diagnosis of CP or “high risk of CP” should be made as early as possible
  – Early standardized assessments and testing for CP should be conducted in at risk populations
  – Infant can receive early intervention and surveillance to optimize neuroplasticity and prevent complications
  – Parents can receive psychological support and resources
What are the risk factors and causes of CP: The 4 P’s

• Prenatal
• Perinatal
• Postnatal
• Prematurity
Prenatal Risk Factors/Causes

• Congenital brain malformations
• Intrauterine stroke
• Intrauterine infections
• Intrauterine growth restriction
• Multiple gestation
• Genetic susceptibility
• Maternal substance abuse
Perinatal Risk Factors/Causes

- Hypoxic ischemic encephalopathy
- Stroke
- CNS infections
- Kernicterus
- Antepartum hemorrhage
- Severe placental pathology
Postnatal Risk Factors/Causes

• Trauma (accidental or non-accidental)
• Anoxic/ischemic insult
• Stroke
• Central Nervous System infections
Prematurity Risk Factors/CAUSES

- 5-15% of surviving very LBW (<1.5 kg = 3.3 lbs) infants develop CP
- Periventricular leukomalacia (PVL)
- Severe intraventricular hemorrhage (IVH)
  - Post-hemorrhagic hydrocephalus
  - Periventricular hemorrhagic infarct
Birth History

• He was born to a 30 year old woman at 27 weeks gestation.
• Complicated by maternal insulin dependent diabetes and pre-eclampsia. At time of birth he was blue
• Birthweight 2 lbs 4oz
• His APGAR scores were 6 and 9 at 1 and 5 minutes.
• He required resuscitation and was intubated and placed on ventilator
• His NICU course was complicated by Grade II IVH, NEC, and stage II ROP and feeding difficulties
• He had a seizure at day 2 of life
• He was discharged on Day 72 of life.
Clinical Case: Developmental History

• **Motor development**
  – Held head up while prone at 4-5 months
  – Rolled over at 7-8 months
  – Sat independently at ~12 months
  – Crawled at ~14 months
  – Pulled to stand at ~16 months
  – Currently cruises on toes

• **Language development**
  – Stringing vowels together
  – Making consonant sounds, a few single words

• **Social development**
  – Smiles
  – Curious about his environment
Clinical Case

- Medications: none
- Allergies: NKDA
- Family History: no CP, seizures, or learning disabilities
- Social History: No daycare, gets physical and occupational therapy in the home 1 hr/wk with Babies Can’t Wait
## Exam findings: Reflexes

<table>
<thead>
<tr>
<th>Reflex</th>
<th>Age of Appearance</th>
<th>Age of Disappearance</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primitive Reflexes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moro (Startle)</td>
<td>34-36 weeks</td>
<td>5-6 months</td>
</tr>
<tr>
<td>Tonic Neck Response</td>
<td>38-40 weeks</td>
<td>2-3 months</td>
</tr>
<tr>
<td>Galant (Trunk incurvation)</td>
<td>38-40 weeks</td>
<td>1-2 months</td>
</tr>
<tr>
<td>Palmar Grasp</td>
<td>38-40 weeks</td>
<td>5-6 months</td>
</tr>
<tr>
<td>Plantar Grasp</td>
<td>38-40 weeks</td>
<td>9-10 months</td>
</tr>
<tr>
<td>Rooting</td>
<td>38-40 weeks</td>
<td>2-3 months</td>
</tr>
<tr>
<td><strong>Postural Reflexes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive Support Reflex</td>
<td>3-4 months</td>
<td></td>
</tr>
<tr>
<td>Landau</td>
<td>4-5 months</td>
<td></td>
</tr>
<tr>
<td>Lateral Propping</td>
<td>5-7 months</td>
<td></td>
</tr>
<tr>
<td>Parachute</td>
<td>8-9 months</td>
<td></td>
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</tbody>
</table>
Physical Exam

- Weight 10 kg Height 105cm  BMI. 13.6  Head circumference 25\textsuperscript{th} percentile
- HR 80. RR 20 T. 97.8
- GEN: NAD
- HEENT: mild drooling, Mild esotropia on his left eye, TM WNL, throat clear, nares patent
- CV: RRR w/o murmur
- RESP: Mild rhonchi but clear breath sounds
- Abd: mildly distended, palpable stool, non tender
- Ext: Mild Plantar flexion contractures of 5 degrees in right Achilles
Physical Exam

- Neurologic Exam:
  - Mental Status: Awake and alert. Regards the examiner and interested in his environment, says hi and no
  - CNs: PERRL, EOMs full, symmetric facial movements, tongue midline
  - Motor: Mild spasticity at the biceps, moderate spasticity at the hip adductors, hamstrings, and gastrocs. Moves all extremities against gravity, has better controlled movements of arms than legs. Walks with walker and assist from mother with crouch
  - Sensation: Intact to light touch
  - Coordination: No ataxia or dysmetria
  - Deep Tendon Reflexes: 2 in the upper extremities, 4 in lower extremities with cross-adduction at the patella and a few beats of clonus at the ankles
What tests do we do?

- Neuroimaging
  - Brain MRI should be performed
    - Abnormalities on MRI are noted in 90% of children with CP
    - 99% of preterm infants; 92% of term infants
- If brain MRI is normal
  - Spine MRI
  - Blood work looking for genetic and/or metabolic problems
  - Lumbar puncture
# What are the types of CP?

<table>
<thead>
<tr>
<th>Classification</th>
<th>Percentage affected</th>
<th>Common Etiologies</th>
<th>Clinical Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spastic Diplegia</td>
<td>13-25%</td>
<td>Periventricular leukomalacia</td>
<td>Both legs &gt; arms</td>
</tr>
<tr>
<td>Spastic Hemiplegia</td>
<td>21-40%</td>
<td>Neonatal stroke, Cortical malformations</td>
<td>One side of the body, Arm &gt; leg, Sensory deficits</td>
</tr>
<tr>
<td>Spastic Quadriplegia</td>
<td>20-43%</td>
<td>CNS infection, Cerebral dysgenesis, Perinatal/postnatal events</td>
<td>All limbs, Arms ≥ legs</td>
</tr>
<tr>
<td>Dyskinetic</td>
<td>12-14%</td>
<td>Severe perinatal asphyxia</td>
<td>Involuntary movements, Exacerbated by stress, excitement, or fever</td>
</tr>
<tr>
<td>Ataxic</td>
<td>4-13%</td>
<td>Early prenatal events, Genetic</td>
<td>Uncoordinated, unsteady movements, Slow, jerky, and/or explosive speech</td>
</tr>
</tbody>
</table>
What are next steps?

• Screen for associated medical problems (seizures, nutritional deficiencies, vision, hearing, and muscle/bone abnormalities, etc)
• Set goals with the patient and primary caregivers
• Interventions focused on maximizing quality of life by reducing the burden of disability
• Multidisciplinary team approach (neurology, physiatry, orthopedics, therapy services, etc)
YOU'RE MY WHAT, SIR?

PHYSIATRIST. I'LL BE COORDINATING YOUR CARE AND REHABILITATION.

LIKE YOU, MOST PEOPLE CAN'T EVEN PRONOUNCE "PHYSIATRIST." IT DOESN'T TRIP OFF THE TONGUE AS EASILY AS, SAY, THE WORD "SURGEON..."
GMFCS

GMFCS Level I

GMFCS Level II

GMFCS Level III

GMFCS Level IV

GMFCS Level V
Equipment

• He has a pair of hinged AFO's
• A convaid Stroller for long distances
Patient A

- He is in the 5% for height and weight
- Drooling
- Palpable Stool
Growth and Nutrition

- Growth failure related to inadequate energy intake
- Children with CP have a higher energy cost of walking than the typically developing children
- High prevalence of children with advanced (7%) or delayed (10%) skeletal age
- Parents overestimate nutritional intake
Growth and Nutrition

- Higher Fat deposition in Muscles
- Impaired Control of Oromotor musculature
- Impaired ability to handle food and drink
- Difficulty transitioning to more caloric dense food
- Prolonged meal times
Gastroenterology

- Delayed Gastric Emptying
- GER → chronic anemia/ dental erosions
- Constipation
- Hyperosmolarity
- Compound?
- Gravity
Delayed Milestones

• He was a premature baby with CP and ROP
• Late talker, delayed speech
Vision Impairments

• Common complication
• Impairments vary and be multiple
  • Acuity
  • Field loss
  • Processing (cortical visual impairment)
  • Eye-hand coordination
Hearing Impairment

- Sensorineural loss more common
- Early assessment and intervention critical
- Follow-up evaluations important in congenital CMV infection because late onset hearing loss common
- Will impact speech-language development
Respiratory Issues

• Upper Airway Obstruction
• Parenchymal lung Disease
  – Aspiration, secretion, GER
• Restrictive Lung Disease
  – Dystonia, scoliosis, obesity, frailty
Oromotor Impairments

- Skin problems
- Cosmetic

- Treatment options:
  - Robinul
  - Scopolamine
  - Atrovent neb
  - Artane
  - Atropine ophthalmic gtts orally
  - Botulinum Toxin injections
  - Excise, ligate or reroute salivary glands
Learning Issues

- 25-50% of children with CP will have some learning issues.
- Assessment can be very challenging due to visual, hearing, and motor impairments.
- Specialized IEP with family input important.
- Neuropsychology evaluation.
- Hand Eye Coordination may decrease performance IQ scores.
- Children in school before 2 yo → inc verbal IQ not performance.
Motor Delays

Delayed Motor Skills
Sat unsupported at 2 years old
Just started walking
Functional Prognosis

<table>
<thead>
<tr>
<th></th>
<th>Good prognosis for independent walking</th>
<th>Poor prognosis for independent walking</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head control</td>
<td>by 9 months</td>
<td>none by 20 months</td>
</tr>
<tr>
<td>Sitting</td>
<td>by 24 months</td>
<td>none by 48 months</td>
</tr>
<tr>
<td>Floor mobility</td>
<td>by 30 months</td>
<td>none by 48 months</td>
</tr>
</tbody>
</table>

Ambulation:
- 75% of all with spastic CP
- 85% with diplegia
- 70% with quadriplegia
- Most with hemiplegia
- >3 abnormal reflexes at 18 months
Spasticity

- Toe walking
- Foot Inversion
- Crouched Gait
- Oral Meds- Baclofen
- Phenol: Hamstrings/Adductors
- Botulinum Toxin:
  - Gastrocnemius/Soleus
  - Posterior Tibialis
- Serial casting on Left
Equipment

Hinged AFO’s
Convaid Stroller
Orthotics

- Provide support
- Limit motion
- Improve function
- Delay or prevent deformity
- AFO’s, SMAFO’s, UCB’s
Orthotics

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Wheelchairs

- Rigid vs folding
- Manual vs power
- Upright vs Tilt-in-space vs reclining
- Sling back vs linear seating
Positioning

- Well fitting wheelchair/seating system
- Adequate support at head, trunk, and feet
- Anti-thrust cushion to provide flexor pattern at hips
Usually the best special seat is one that provides the least amount of support needed to help the child do the most for herself.

For a child whose body stiffens like this, don't do this, when all that is needed is this.
Community Activities

CATALYST SPORTS
BECOME INSPIRED

BlazeSports
America

RHYTHM WORKS
Integrative Dance
Community Activities

I fell down on your sidewalk and bumped my knee.

Fortunately, we have a world famous orthopedic surgeon right here.
ORTHOPEDICS

• Persistent muscle imbalance can cause muscle/tendon contractures --> bone/joint deformities

• Abnormal forces across joints --> prolonged abnormal posture, increased energy expenditure

• Impairment of function and quality of life
Common Conditions

• Neuromuscular hip dysplasia
• Scoliosis
• Joint contractures
• Limb length inequality
• Planovalgus foot
• Cavovarus foot
• Equinus contracture
The Spastic Hip

• overactive hip flexors and adductors
• scissoring, intoeing, symptomatic subluxation, frank dislocation
• relationship to spinal deformity controversial but present in some cases
• can make walking in the diplegic and sitting in the quadriplegic difficult
Question?

Doc, Do we really need that xray?
Scrutton et al, DMCN 2001

- migration % (femoral head containment in acetabulum) is best radiographic guide
- all children with bilateral CP should have pelvic X-Ray at 30 months of age
- clinical examination is an unreliable method of screening hips for risk of subluxation
# Neuromuscular Hip Dysplasia

- **Hip dislocation related to GMFCS**
  - 0% GMFCS I
  - 15% GMFCS II
  - 41% GMFCS III
  - 69% GMFCS IV
  - 90% GMFCS V

- **Consequences**
  - Pain (~50%)
  - Early OA
  - Difficulty with positioning
  - Difficulty with hygiene
  - Skin breakdown over bony prominences
Natural History of the Hip in C.P.

- initially hips are radiographically normal
- lateral subluxation, secondary to adductor spasticity and excessive anteversion & valgus, leads to acetabular dysplasia then to dislocation

17 Months
Why Treat?

• how many of these subluxed/dislocated hips will cause problems?
  – ~50%, and probably more if parental denial removed from the equation.
  – difficulty sitting, diapering, perineal care, pain with motion, decubiti

• if patient able to perceive pain and communicate, you’ll know it hurts

• more treatment options early on than later
3 Phases of Treatment

- Prevention
- Reconstruction
- Palliative
Treatment of the Hip

• first: prevention-- proper positioning, standing balance, ? night time bracing/splinting

• close observation with radiographs looking for early subluxation, dysplasia, dislocation
Prevention

• **Indications**
  – Under age 8 years
  – Hip abduction < 30 deg
  – MI 25-60%

• **Spasticity Management**

• **Soft tissue lengthening**
  – **Adductor** release
  – **Hip flexor** release
    – Ambulatory: psoas only
  – **Hamstring** release
    – If popliteal angle greater than 45 deg

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**TABLE 1: Indications for Soft-Tissue Releases in Patients with Cerebral Palsy**

| Patients ≤7 yr of age | Migration percentage of ≥25% and abduction of ≤30°
| Indication for a repeat soft-tissue release | Migration percentage of 25%-50% and abduction of 31°-45°, if migration percentage increases 10% per year
| Patients ≥8 yr of age | Migration percentage of ≥50% and abduction of ≤45°
| If migration percentage ≥40%, proceed to osseous hip reconstruction |
Treatment of the Dislocated Hip

• what about RX of the symptomatic frank dislocation?
  – 1st: avoid it happening by early intervention!!
  – 2nd: if not longstanding, try hip reconstruction with femoral/pelvic osteotomy, soft tissue releases
  – last: no great option, but femoral head resection probably gives best result (70%)
  – ??Other options: valgus osteotomy, THR, hip fusion. Patient selection CRUCIAL.
Reconstruction

• Indications
  – Recommended above age 4 years
  – For MI > 60% and dislocated hips
  – Must not have degenerative changes

• Soft tissue lengthening

• Femoral osteotomy
  – Varus derotational
  – +/- shortening

• +/- Pelvic osteotomy
  – Lateral Coverage acetabuloplasty
  – PAO if triradiate closed
Hip Reconstruction
Salvage

• **Indications**
  – Painful dislocated hip with advanced degenerative changes
  – Skeletal maturity

• **Conservative Tx:**
  – Pain Management
  – Phenol/ Steroids

• **Femoral**
  – Castle
  – McHale
  – Fusion
  – THA
Femoral Head Resection Interposition Arthroplasty
RESULTS

• Early soft tissue releases may prevent progressive subluxation in 60% of patients.
• Bone +/- soft tissue procedures can achieve 70-80% success if done prior to fixed dislocations
• Address tone to improve results
Scoliosis and Hip Dysplasia

• Incidence GMFCS IV/V  75%
• Pelvic obliquity
• Severity of Spinal deformity
• Skeletal Maturity
 Neuromuscular Scoliosis

• Correlates with level of function

• Goals differ from the more common idiopathic population

• Must weight risk/benefits based upon comorbidities, pulmonary function, nutritional status
Clinical Signs of Neuromuscular Scoliosis

- Change in posture/seating
- Pelvic obliquity
- C-shaped or ‘sweeping curvature’ of spine
Neuromuscular Scoliosis

- Non ambulatory patients
- Triad
  - Scoliosis
  - Pelvic obliquity
  - Hip dislocation
Relationship of Deformities

- 1st and 2nd decade of life
- Orthotic
- Wheelchair Modifications
- Late surgical management when severe and rigid deformities.
Non Operative Management of Neuromuscular Scoliosis

• Team Approach
• Address Function
  – Tolerate (spastic vs flaccid)
  – difficult
• Address spinal deformity
  – Partially correct
• Address pelvic obliquity
  – Improve truncal balance
• Unlikely to prevent curve progression
Treatment

• Non operative Rx-
• Observation- mild curves only
  – TLSO
    • GI issues and Pulmonary problems
  – Seating support
    • Lateral supports and modular seating

Operative Rx- documented curve progression and deterioration in function
Benefits of Bracing

• Stable sitting
• Easier use of upper limbs
• Distract of the abdomen from collapsing trunk
• Increased diaphragm excursion
12 yo female GMFCS 5

- Seizure disorder
- Feeds by mouth/ No GT
- Difficulty with sitting, pressure points over the ribs.
- Have attempted multiple seating adjustments
- Difficulty with soft brace due to constipation
- Limited but pain free hip motion
- No problems with perineal care
Hips or Spine?

Severe Spinal Curve

Significant pelvic Obliquity

Tackle the Spine
Hip or Spine?

Address the hips first
Goals of surgery

- Level pelvis
- Head over shoulders
- Shoulders over pelvis
- NOT complete correction of curve
- Pain Free Hips
- Improvement of ADLs
- Improve Quality of Life
POST OP PSF T2-Pelvis
Fast Forward 2 years post op

- Complains bilateral hip pain
- Painful ROM
- No problems with perineal care
Take Home

Scoliosis

Pelvic Obliquity

Hip Dislocation
Your greatness is not what you have, but what you give.