Disclaimer:
Edited a book for primary care providers, mentioned in presentation.
Introduction

- CP is a common condition presenting in childhood that impacts individuals across the lifespan
- CP often more than just motor dysfunction
- Extremely heterogeneous
- CP impacts whole family
- Role of the PCP or general pediatrician is central
Topics for Review

- Definition and classification
- Early diagnosis and prognosis
- Nutrition/growth
- Therapy/fitness
- ICF: CP in Life
- Joints, bones, and surgical treatment
- Behavior/mental health
- Medical Comorbidities
- Equipment
- Dealing with school
- Family life/siblings
- Friendship
- Difficult decisions
- Coping
- Transition
- Guardianship
- Adulthood
How do we best help this child grow up well in his family and community?
Objectives

- Describe the clinical syndrome of cerebral palsy, its epidemiology, causation and classification
- Early identification, diagnosis, workup
- Primary management (PT, Ortho, Med) and the role of the pediatrician
- Review associated/secondary conditions
- Discuss resources to support children and youth with CP, and their families
How common is CP?

- Prevalence 2.7-3.6 per 1,000*
- 2013 Metanalysis of Prevalence Studies (International)
  - 2.11 per 1,000 live births pooled data

- BW < 1,000g: 56/1,000
- BW 1000-1499: 59/1,000
- BW 1500-2499: 10/1,000
- BW > 2500: 1.33/1,000

- GA < 28 weeks: 82/1,000
- GA 28-31 weeks: 43/1,000
- GA 32-36 weeks: 7/1,000
- GA > 36 weeks: 1.35/1,000

Prevalence of CP in US

- Overall Prevalence (3 sites):
  - 3.6/1,000 8-year old children
  - Spastic 2.8/1,000 (unilateral 0.8/1,000)
  - Dyskinetic/Ataxic 0.19/1,000
  - Mixed/Unspecified 0.6/1,000
  - Hypotonic 0.1/1,000
Early Diagnosis: Risk Factors & Tools

- Prematurity and/or Low Birthweight
- Asphyxia/Hypoxic-Ischemic Encephalopathy
- General Movements Assessment (GMA)
- Brain MRI
- Hammersmith Infant Neurological Exam (HINE)

Early Diagnosis: Presenting Signs

- Motor Delay
  - No sit by 9 mos, no walk by 15 mos
- Hypotonia or Hypertonia
- Asymmetry of tone or movement
- Early hand dominance
- Prolonged fisting (unilateral/bilateral)
- Atypical movement
  - “mermaid”, scissoring, commando crawl, toe walk/stand, arching, stiffness
Next Steps for Pediatrician

- Referral to medical specialist for diagnosis and differential consideration
  - Neurologist, Developmental Peds, PMR
- Consider referral to Peds Ortho
- Discuss possibility of CP with family
- Referral to PT and/or OT and Speech
- Consider evaluating hearing and vision
- Consider MRI and/or Genetics referral
New Idea...

- “Your child could have CP but we cannot tell for sure yet. Let’s wait and see...”

  versus

- “Your child is at high risk for CP. Let’s try to do something about it...”
Cerebral Palsy: Definition

A group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.

The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior; by epilepsy, and by secondary musculoskeletal problems.

*Rosenbaum, et al DMCN 49 (supp 109): 8-14, 2007*
Diagnosis of Cerebral Palsy

- Based on clinical history and examination
- Presence of motor delay and abnormalities of movement, reflexes and muscle tone
- No evidence of clinical deterioration
- Clinical picture fits definition
Medical Evaluation of Child with Clinical CP

All children with the diagnosis of CP

- Brain MRI
- Hearing evaluation
- Ophthalmologic evaluation
- Developmental screening

Children with characteristic risk factors or clinical presentations

- Genetic testing (phenotypic or MRI findings)
- Hematologic evaluation (history of stroke)
- Infectious work-up (evidence of perinatal infection: toxoplasmosis, CMV, HSV, etc.)
- Metabolic evaluation (uncertain diagnosis)
- EEG (clinical suspicion)

Proposed Components for the Classification of CP*

- Motor abnormalities
  - Motor typology
  - Functional status
- Associated impairments
- Anatomic & radiologic findings
  - Anatomic distribution and neuroimaging
- Causation and timing

*DMCN 47: 571-76, 2005
Classification of Motor Abnormalities

- Motor Typology
  - Spasticity
    - Bilateral vs Unilateral
  - Dyskinesia
    - Hyperkinetic (chorea)
    - Hypokinetiс (athetosis/dystonia)
  - Ataxia (titubation, dysmetria)
  - Mixed types (describe)
Functional Motor Abilities
GMFCS

- Level 1: Walks without restrictions
- Level 2: Walks without assistive devices; limitations outdoors/ in community
- Level 3: Walks with assistive devices; limitations walking outdoors/ in community
- Level 4: Self-mobility with limitations; transported or use power mobility in community
- Level 5: Self-mobility is severely limited even with assistive technology

GMFCS E & R Descriptors and Illustrations for Children between their 6th and 12th birthday

**GMFCS Level I**
Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.

**GMFCS Level II**
Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.

**GMFCS Level III**
Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.

**GMFCS Level IV**
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

**GMFCS Level V**
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

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GMFCS E & R Descriptors and Illustrations for Children between their 12th and 18th birthday

**GMFCS Level I**
Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping but speed, balance and coordination are limited.

**GMFCS Level II**
Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.

**GMFCS Level III**
Youth are capable of walking using a hand-held mobility device. Youth may climb stairs holding onto a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility.

**GMFCS Level IV**
Youth use wheeled mobility in most settings. Physical assistance of 1-2 people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.

**GMFCS Level V**
Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.
Prevalence by GMFCS*

- Level 1 32%
- Level 2 27%
- Level 3 12%
- Level 4 14%
- Level 5 15%

* Australian Cerebral Palsy Registry
What does this mean for average practice of 2000 children?

- 7 children with cerebral palsy
  - 4 children GMFCS 1 or 2
    - Probably all spastic
    - 2 hemiplegia and 2 diplegia
  - 1 child GMFCS 3 (probably spastic)
  - 1 child GMFCS 4 (spastic or mixed tone)
  - 1 child GMFCS 5 (spastic or mixed tone)
- Rarely, 1 child with ataxia/dyskinesia

MACS

- I: Handles objects easily and successfully
- II: Handles most objects but with reduced quality and/or speed (self-modification)
- III: Handles objects with difficulty; needs help to prepare and/or modify activities
- IV: Handles a limited selection of easily-managed objects in adapted situations
- V: Does not handle objects; severely limited

* Eliasson, et al, DMCN: 48:549-54
CFCS

- I: Effective sender and/or receiver with familiar and unfamiliar partners
- II: Effective but slower paced sender/receiver with all partners
- III: Effective sender/receiver with familiar partners only
- IV: Sometimes effective with familiar
- V: Seldom effective even with familiar

EDACCS

I: Eats and drinks safely
II: Eats and drinks safely but some limitations to efficiency
III: Some limitations to safety; may be limitations to efficiency
IV: Eats and drinks with significant limitations to safety
V: Cannot eat or drink safely

* DMCN: online, December 2013
Associated Impairments & Secondary Conditions

- Impact on the health and quality of life of child and family
- Add family stress
- Potentially preventable, manageable, or remediable
Associated Impairments & Secondary Conditions

- Cognitive impairments
- Sensory impairments
- Seizures
- Strabismus
- Behavioral disorders
- Mental health
- Malnutrition
- Orthopedic problems
- Reflux/constipation
- Chronic/acute aspiration
- Chronic pain
- Constipation
- Drooling
Radiographic/Anatomic Findings

- Diffuse vs Focal
- Types
  - Maldevelopment 10%
  - Deep gray matter
  - White matter
  - Vascular
- Normal 15%

- Unilateral
  - Right vs Left
- Bilateral
  - Symmetric
  - Asymmetric

Himmelmann, et al, DMCN, 59:57-64, 2017
Causation and Timing

Causal Pathways/Risk Factors

- Medical history
- Clinical syndrome
- Neuroimaging
  - Calcifications
  - Distribution
  - Timing
- Genetics (10-30%?)
- Hematology
- Infection
  - CMV, toxo, bacteria
  - Hypoxic ischemic
- Birth asphyxia
- Kernicterus
- Trauma
- Hemorrhage
- Prematurity
## Causation and Timing

<table>
<thead>
<tr>
<th>Timing</th>
<th>Type of Mechanism</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-natal</td>
<td>Intrauterine pathological processes</td>
<td>Placental vascular disease</td>
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<tr>
<td></td>
<td></td>
<td>Intrauterine growth retardation</td>
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<tr>
<td></td>
<td></td>
<td>Infection with fetal inflammatory response</td>
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<tr>
<td></td>
<td></td>
<td>Congenital/genetic anomalies</td>
</tr>
<tr>
<td>Peri-natal</td>
<td>Peri-partum events</td>
<td>Birth asphyxia</td>
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<tr>
<td></td>
<td></td>
<td>Chorioamnionitis</td>
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<tr>
<td></td>
<td></td>
<td>Placental abruption</td>
</tr>
<tr>
<td>Post-natal</td>
<td>Neonatal complications</td>
<td>Intraventricular hemorrhage</td>
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<tr>
<td></td>
<td></td>
<td>Sepsis/meningitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Periventricular leukomalacia</td>
</tr>
<tr>
<td></td>
<td>Late complications</td>
<td>Hypoxic-ischemic brain injury</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Non-accidental trauma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Meningitis/encephalitis</td>
</tr>
</tbody>
</table>

Adapted from Figure 1, (Stavsky, Mor et al. Frontiers in Pediatrics 5: 21, 2017)
Genetics and the cause of CP?

- Genetics studies have shown 7-15% CNV in CP (*similar to autism*): deletions or duplications
- Chromosome 22, 1q21, Xp21.2 (DMD), 2.25, DIP2C as examples
- Many of the genes have been found in ASD and other ND disorders
- Similar findings in unpublished data on 100 kids with hemiplegia

## Summary of Classification

<table>
<thead>
<tr>
<th>Primary Component</th>
<th>Potential Sub-Component</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Motor Abnormalities</strong></td>
<td>Motor Typology</td>
<td>Spasticity &amp; dystonia</td>
</tr>
<tr>
<td></td>
<td>Functional Status</td>
<td>GMFCS IV, MACS III</td>
</tr>
<tr>
<td><strong>Associated impairments</strong></td>
<td>Neurological comorbidities</td>
<td>Seizures, ID**, CVI***, dysphagia</td>
</tr>
<tr>
<td></td>
<td>Medical/Orthopedic secondary conditions</td>
<td>Contractures at elbows, knees, hips;</td>
</tr>
<tr>
<td></td>
<td></td>
<td>constipation; malnutrition; drooling</td>
</tr>
<tr>
<td>Anatomic &amp; Radiologic Findings</td>
<td>Anatomic distribution of spasticity</td>
<td>Bilateral, asymmetric (R&gt;L)</td>
</tr>
<tr>
<td></td>
<td>Neuroimaging findings/classification</td>
<td>Malformation: schizencephaly</td>
</tr>
<tr>
<td>Causation and Timing</td>
<td>Genetic (&quot;intrinsic&quot;) factors</td>
<td>Gene deletion syndrome</td>
</tr>
<tr>
<td></td>
<td>Medical (&quot;extrinsic&quot;) factors</td>
<td>Cocaine exposure in utero</td>
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<td></td>
<td>Timing of injury or perturbation</td>
<td>Early second trimester; born at term</td>
</tr>
</tbody>
</table>
Why is Classification Important?

Classification aids

- Conceptualization
- Prognosis
- Communication
- Research
- Treatment
- Matching right treatment with right child
Cerebral palsy is never the same disease twice

-Michael Shevell
Options for Treatment

- Physical/Occupational/speech therapy (including early intensive)
- Bracing/adaptive equipment
- Orthopedic surgery
- Neurotoxins and oral medications
- Selective dorsal rhizotomy
- Intrathecal baclofen pump
- Alternative therapies
Interventions for children with CP: state of the evidence

- 166 articles
- 64 interventions
- 131 outcomes
- 16% "green light"
- 6% "red light"
- "worth it" line
New Ideas for Treatment

- Prevention and mitigation of severity
  - Magnesium sulfate, EPO, cooling
  - Stem cells (?)
  - Early rehabilitation “bundles”
- Intensive early therapies (i.e. CIMT)
- “Maintenance therapy”
- Therapy intensives (goal directed)
- Focus on function and participation
ICF Framework

Health Condition (CP)

Body Structures and Functions

Activities

Participation

Environmental Context and Personal Factors
ICF Framework

Health Condition (CP)

PVL, spasticity, Contracture, deformity

Poor gait, self care

Popular, outgoing Cheerleader

Family support, school principal, Wheelchair, SEML
Outcome measures

- Development of many valid and reliable measures that quantitate status and change
- Broadened scope of “outcomes”
- Informed by ICF
- Development of specific measures in each domain of the ICF
- Construct of Quality of Life
F Words of Childhood Disability

- Function – focusing on the “can do”
- Family – context for everything
- Fitness – need to be “fitter”
- Fun – life is about “doin’ stuff”
- Friends – quality not quantity
- Future – all children are “becoming”

What comes next?

- Role of genetics
- Stem cells
- Early diagnosis
- Intensive early intervention
- Comparative work
- Melding research and practice (CPRN)

https://cprn.org/
Transition to Adulthood

- Important process for all children with disabilities or chronic illnesses
- Identify primary care doctor
  - Geriatric expertise helpful
- PM&R for musculoskeletal consult
- PT +/- Physical trainer
- Psychiatry or Neurologist
- Consider attorney for POA
Summary

- Cerebral palsy (CP) is a complex, heterogeneous syndrome
- Definition and classification of CP are important for clinical care and research
- Treatments are available with variation in evidence base
- Resources available for families
- Pediatrician and primary provider play central role in growing up well
What is most important?

- Listen to parents and children
- Care for the entire family (siblings)
- Coordinate and interpret specialty care
- Identify strengths of the child and foster maximum independence
- Family, friends and community
- Inspire and challenge children
- Facilitate transition to adult care
Tools for Families (and others)
Useful Links

- http://yourcpf.org/
- https://abc.go.com/shows/speechless
- https://www.aacpdm.org/
- http://cpdailyliving.com/
- https://cpnowfoundation.org/
Thank you!

“Yes, you are on the frontlines of your patients’ medical care. But more importantly, to the parents of a child with complex cerebral palsy, you are on the frontlines of hope.”

Carol Shrader (mother of triplets, two of whom have CP)