

## The Cerebral Palsy Patient's Musculoskeletal Exam:

How to perform, What not to miss, and Who to refer out to and When

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RWJBarnabas Health

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### My Background

- Medical School NYU School of Medicine, New York, NY
- Residency NYU Langone Orthopedic Residency, New York, NY
- Fellowship Nemours Children's Health, Wilmington, DE



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### Areas of Interest

- Neuromuscular care
- Pediatric trauma/fracture care
- Hip dysplasia
- Lower extremity deformity
- Gait abnormalities
- Foot deformities
- Clubfoot
- Osteogenesis imperfecta
- Muscle diseases



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### Goals

1. Understand pathophysiology and classification of CP from an orthopedic perspective
2. Have confidence in performing a consistent, thorough musculoskeletal exam for a neuromuscular (NM) patient
3. Understand when to refer out to different subspecialties
4. The role orthopedics play in early intervention for CP

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### Cerebral Palsy definition

**Cerebral Palsy (CP)** is a lifelong neurodevelopmental condition characterized by limitations in activity due to impaired development of movement and posture, manifesting as spasticity, dystonia, choreoathetosis, and/or ataxia. It results from maldevelopment attributed to malformation or injury to the fetal or infant brain that is not degenerative, although the manifestations may change with age. The phenotype of CP is complex and heterogeneous, with each person experiencing a unique presentation. In addition to motor dysfunction, persons with CP frequently encounter primary and secondary impairments across various areas of development and functioning, which can significantly impact their participation in daily life.

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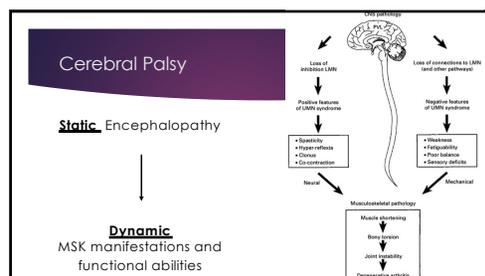
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### Cerebral Palsy – Epidemiology

- ▶ 2-4/1,000 live births
- ▶ No Nationwide surveillance in US
- ▶ Europe: 2/1,000
- ▶ Increased risk:
  - ▶ Multiple pregnancies
  - ▶ Low birthweight
  - ▶ Low gestational age

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### Cerebral Palsy – CP Like condition

- ▶ Classic CP was from birth anoxia
- ▶ "Modern" "typical" CP now from prematurity
  - ▶ Periventricular leukomalacia
- ▶ Other conditions give a musculoskeletal disability similar to CP
  - ▶ Chromosomal disorders
  - ▶ Perinatal infections
  - ▶ Perinatal strokes
  - ▶ Congenital Brain Malformations
- ▶ Certainly, may have different pathophysiology and different phenotypes
- ▶ "Lumpers or Splitters" - Ortho typically "Lumpers" due to similar motor impairments

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### Cerebral Palsy – Primary Impairments

- ▶ Abnormal muscle tone
- ▶ Loss of selective muscle control
- ▶ Impaired coordination and balance
- ▶ Weakness
- ▶ Loss of sensation

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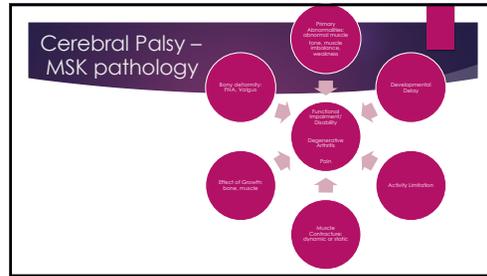
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- ### Cerebral Palsy – Orthopedic Care
- ▶ Hip dysplasia
  - ▶ Foot deformity
  - ▶ Spinal deformity
  - ▶ Upper extremity
  - ▶ Gait disturbances
  - ▶ specifically, crouch gait

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- ### Cerebral Palsy – Goals of Orthopedic Care
- ▶ Normalize, or decrease tone
  - ▶ Prevent muscle contracture:
    - ▶ ROM via therapy, bracing, lengthening, balancing
  - ▶ Correct bony deformity
  - ▶ Optimize function/Quality of Life

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### Cerebral Palsy – Classification

- ▶ Depends on location and extent of injury
- ▶ Spastic, hypotonic, or dystonic
- ▶ Hemiplegia, diplegia, superimposed, quadriplegia, or triplegia

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### Cerebral Palsy – Classification GMFCS

- ▶ Valid: Based on GMFM
- ▶ Reliable
- ▶ Stable (Relatively)
- ▶ Prognostic: Predicts Natural History
- ▶ Goal Setting
- ▶ Monitoring but not outcome measure



The image shows six illustrations of children performing different activities, each labeled with a GMFCS level: GMFCS Level I (walking), GMFCS Level II (walking with a cane), GMFCS Level III (walking with a walker), GMFCS Level IV (using a wheelchair with manual propulsion), GMFCS Level V (using a wheelchair with manual propulsion and a communication device), and GMFCS Level VI (using a wheelchair with manual propulsion and a communication device).

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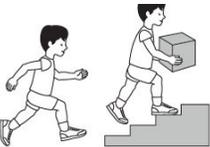
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### GMFCS I

- ▶ typically developed, except balance and coordination limited



The image shows a child walking up a set of stairs, carrying a box. Below the illustration is the label "GMFCS Level I".

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**GMFCS 2**

► Walk, may use cane/crutch occasionally; minimal jumping/running



**GMFCS Level II**

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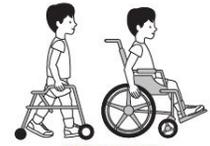
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**GMFCS 3**

► Crutch/walker indoors; self-propelled chair; can do long distances



**GMFCS Level III**

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**GMFCS 4**

► Need assistance, independent use of power chair, assistance with walker



**GMFCS Level IV**

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### GMFCS 5

- ▶ Dependent on aide in all settings, manual wheelchair only; difficulty with head/trunk postures

GMFCS Level V

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### Early Diagnosis

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### Early Diagnosis

- ▶ Important to diagnose early as:
  - ▶ Maximize the neuroplasticity to maximize the child's movement and cognitive outcomes
  - ▶ Early, regular monitoring and treatment for the known musculoskeletal complications of cerebral palsy can prevent the onset of hip dislocation, scoliosis and contracture
  - ▶ Parents experience more depression and stress when they are disoriented with the diagnostic process; families prefer early diagnosis, followed by early intervention and parent-to-parent support.
  - ▶ The lack of intense early intervention may restrict the infant's motor and cognitive gains\*\*

\*\*Bickel et al. 2003  
\*\*\*Nurjan et al. 2016

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### Early Diagnosis

- ▶ An international clinical practice guideline (detailed on the AACPD website) shows that using 3 tests together in combination, enables early diagnosis of cerebral palsy at 12 weeks of age with over 95% accuracy.
- ▶ The 3 tests are:
  - ▶ A brain scan (MRI) showing damage to the movement areas of the brain
  - ▶ A movement test where the child's movement is scored to be of low quality from video footage (General Movements Assessment)
  - ▶ A scored neurological test showing either asymmetries between the left and right or atypical postures (Hammersmith Infant Neurological Examination).

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### Early Diagnosis

- ▶ Prechtl's General Movements Assessment (GMA)
  - ▶ Birth to 20 weeks
  - ▶ Video assessment of "writhing" (6-9 weeks), "fidgeting" (12-20 weeks)
  - ▶ How the CNS is developing

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### Early Diagnosis

- ▶ Hammersmith Infant Neurological Exam (HINE)
  - ▶ 3 to 24 months
  - ▶ 26 assessment items including cranial nerve function, movements, reflexes, protective reactions and behavior and age dependent items reflecting gross and fine motor function

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## Early Intervention

- ▶ Randomized control trials have indicated that:
  - ▶ Infants with hemiplegic CP who receive early Constraint Induced Movement Therapy (CIMT) have better hand function than controls short-term and probably substantially better hand function long-term.\*
  - ▶ Infants with any type and topography of cerebral palsy, who receive "GAME" (Goals - Activity - Motor Enrichment, which is an early, intensive, enriched, task-specific, training based interventions at home), have better motor and cognitive skills at 1-year, than those who received usual care\*\*
  - ▶ Improvements are even better when training occurs at home) because children learn best in supported natural settings, where training is personalized to their enjoyment - translating to more intense, specific and relevant practice.\*\*\*

\*Blanton et al. 2019  
 \*\*Auzan et al. 2016  
 \*\*\*Novak et al. 2006; Raitani et al. 2012

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## Upper and Lower Extremity Evaluation

**Upper Extremities:**

Bilateral upper extremities have grossly symmetrical alignment

No obvious swelling or pain with palpation

Elbow flexion contracture  right  left  degrees right

Finger flexion contracture  right  left  degrees

A flexion in palm deformity  PRESENT/ABSENT  on the left hand, and  PRESENT/ABSENT  on the right hand

Swan neck deformity  right  left

Plan in the upper extremities during ROM testing was absent

**Brace evaluation:** Brace  (eg braces) fit w/ without signs of skin irritation or breakdown

**Distal:**

Normal heel to toe progression  YES/NO (default YES)

Upper extremity posturing  YES/NO

Angulae gain  YES/NO

Transferability  YES/NO

Barthel

in  hands and  others

**Neurologic:**

Muscle tone  normal in all extremities.

Spasticity muscle test

Sensations  YES/NO  YES/NO  YES/NO  YES/NO

Strength  NORMAL/ABNORMAL

Grip

Ballistic

**Lower Extremities:**

	Right	Left
Hips- Flex	<input type="checkbox"/> 120	<input type="checkbox"/> 120
Hips-Extend	<input type="checkbox"/> 30	<input type="checkbox"/> 30
Hips-ADD (Hip-knee flex)	<input type="checkbox"/> 45	<input type="checkbox"/> 45
Hips- ABD (Hip-knee extension)	<input type="checkbox"/> 45	<input type="checkbox"/> 45
Hips- Internal	<input type="checkbox"/> 75	<input type="checkbox"/> 75
Hips- External	<input type="checkbox"/> 45	<input type="checkbox"/> 45
Hips- Internal (prone)	<input type="checkbox"/> 75	<input type="checkbox"/> 75
Hips- External (prone)	<input type="checkbox"/> 45	<input type="checkbox"/> 45
Plantar angle	<input type="checkbox"/> 40	<input type="checkbox"/> 40
Distal leg/proximal angle	<input type="checkbox"/> 40	<input type="checkbox"/> 40
Knee extension	<input type="checkbox"/> 0	<input type="checkbox"/> 0
Ankle dorsiflex- Extension	<input type="checkbox"/> 15	<input type="checkbox"/> 15
Ankle plantarflex- Flexion	<input type="checkbox"/> 15	<input type="checkbox"/> 15
Heel to toe	<input type="checkbox"/> 100	<input type="checkbox"/> 100
Thigh Foot Angle	<input type="checkbox"/> 5	<input type="checkbox"/> 5
Barthel ADL	<input type="checkbox"/>	<input type="checkbox"/>

Condition of heel: left  right

Condition of ball: left  right

Condition of ball: left  right

Condition of ball: left  right

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## Upper Extremity Evaluation

- ▶ Elbow flexion contractures
- ▶ Wrist flexion contractures
- ▶ Finger flexion contractures
- ▶ Thumb in palm
- ▶ Swan neck



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### Upper Extremity Evaluation

- Coordination
- Fine motor

Table 8.2. Upper extremity functional patterns.	
Type 0	No active function in the entire upper extremity
Type 1	Proximal function, none to minimal distal function (uses hand as a paperweight/posting device)
Type 2	Mass grasp, poor active control and strength, poor fine motor control
Type 3	Fair active grasp/release (able to place object with fair accuracy), poor thumb opposition
Type 4	Good active grasp/release, fair thumb opposition (key pinch only)
Type 5	Normal to near-normal function, good thumb opposition, able to perform sophisticated fine motor tasks (e.g., buttoning clothes)

Each type is further subdivided into A, no contractures; B, dynamic contractures only; and C, fixed contractures only.

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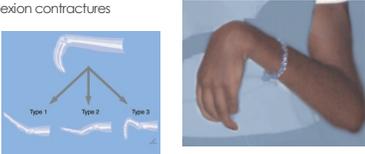
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### Upper Extremity Evaluation

- Wrist flexion contractures



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### Upper Extremity Evaluation

- Thumb in palm



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### Upper Extremity Evaluation

► Swan neck

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### Upper Extremity Evaluation

► Shoulder and elbow extension contractures

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### Lower Extremity Evaluation

Lower Extremities:	Right	Left	Brace evaluation: Bladder (leg braces) fit w/ without signs of skin irritation or breakdown?
Hips-Flex	110	110	0
Hips-Extnd	120	120	0
Hips-ABD (knee flexed)	45	45	0
Hips-ABD (knee flexed)	45	45	0
ankleDIP	90	90	0
Hips-Internal	70	70	0
Hips-External	45	45	0
Hips-Internal (prone)	70	70	0
Hips-External (prone)	45	45	0
Popliteal angle	40	40	0
Double leg popliteal angle	40	40	0
Knee extension	90	90	0
Ankle dorsiflex-Extension	15	15	0
Ankle dorsiflex-Flexion	15	15	0
Reclin-Ey	140	140	0
Thigh-Foot Angle	45	45	0
Metatarsal Axis	90	90	0

Condition of feet left:  right:   
 Complete block walk left:  right:

**Brace evaluation:** Bladder (leg braces) fit w/ without signs of skin irritation or breakdown?   
 0=Normal heel to toe progression.  1=NO (distal) yes.  2=Upper extremity protruding.  YES/NO   
 Ankle dip:  YES/NO   
 Tibiotalang limb:  YES/NO   
 Bariatric:   
 In bracer and shoes:   
**Neurology:**  
 Muscle tone:  normal in all activities.  
 Spasticity muscle rest:   
 Sensations apparent when to tight both over the dorsum and plantar aspect of the foot.  
 Strength:  NORMAL/ABNORMAL   
 Gait:   
 Balance:

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Lower Extremity Evaluation

► Hip extension

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Lower Extremity Evaluation

► Hip abduction – knees flexed, and knees extended

► Gracilis

► Adductors

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Lower Extremity Evaluation

► Hip rotation

► ER and IR

► Prone vs supine

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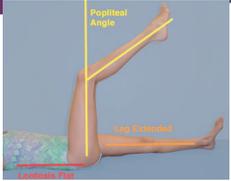
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Lower Extremity Evaluation

- ▶ Popliteal angle
  - ▶ Single
  - ▶ Double



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Lower Extremity Evaluation

- ▶ Knee extension
  - ▶ Knee flexion contracture vs popliteal angle



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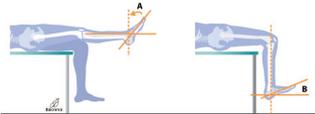
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Lower Extremity Evaluation

- ▶ Ankle dorsiflexion— knees flexed, and knees extended- "Silfverskiold test"
- ▶ Soleus
- ▶ Gastrocnemius

**Silfverskiold Test**



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Lower Extremity Evaluation

- ▶ Ankle dorsiflexion—knees flexed, and knees extended- "Silverskold test"
- ▶ Soleus
- ▶ Gastrocnemius

**Surgery for Spastic Contracted Gastrocnemius**

- Triceps surae muscle anatomy
- Gastrocnemius myofascial lengthening
- Complete release of gastrocnemius muscle to soleus tendon
- Myofascial lengthening of soleus muscle to lateral talar tendon
- Flexibility of the soleus muscle
- Release transfer of tendon to heel
- Measuring lengthening of tendon Achilles
- 2 lengthening

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Lower Extremity Evaluation

- ▶ Rectus Ely
- ▶ Spasticity
- ▶ R1 vs. R2

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Lower Extremity Evaluation

- ▶ Thigh foot angle
- ▶ Vs foot progression angle

Neutral = 0°

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Lower Extremity Evaluation

- ▶ Foot deformities:
  - ▶ Equinus
  - ▶ Hallux Valgus
  - ▶ EquinoPlanoValgus
  - ▶ EquinoCavovarus



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Lower Extremity Evaluation

- ▶ Foot deformities:
  - ▶ Hallux Valgus



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Lower Extremity Evaluation

- ▶ Foot deformities:
  - ▶ EquinoPlanoValgus



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Lower Extremity Evaluation

- ▶ Foot deformities:
  - ▶ EquinoCavovarus



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Lower Extremity Evaluation

- ▶ Muscle tone
  - ▶ Kind:
    - ▶ Abnormal tone – spasticity, hypotonia
  - ▶ Distribution:
    - ▶ Hemiplegic, diplegic, triplegic

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Lower Extremity Evaluation

- ▶ Selective muscle test → SCALE test quantifies selective motor control in CP
- ▶ Assessing the ability to perform specific movements of the ankle, knee, and hip, while ensuring that other joints remain stable.
- ▶ For example, the test might involve asking the patient to invert, evert, and then invert their ankle while maintaining knee extension.
- ▶ The test looks for signs of unwanted movements, such as movement of joints other than the one being tested, mirror movements (both ankles moving when only one is supposed to), or mass pattern movements (synergies).

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Lower Extremity Evaluation

► Strength

Muscle Strength Grading Scale (Oxford Scale)	
0/5	No contraction
1/5	Visible/palpable muscle contraction but no movement
2/5	Movement with gravity eliminated
3/5	Movement against gravity only
4/5	Movement against gravity with some resistance
5/5	Movement against gravity with full resistance

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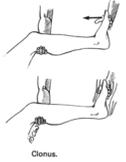
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Lower Extremity Evaluation

► Clonus = involuntary, sustained, rhythmic beating of ankle with the firm, passive stretch of the Achilles tendon

► Measured by beats: 1, 2, 3 etc.



Clonus.

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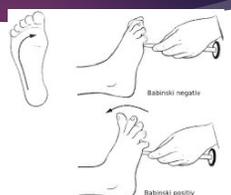
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Lower Extremity Evaluation

► Babinski



Babinski negative

Babinski positive

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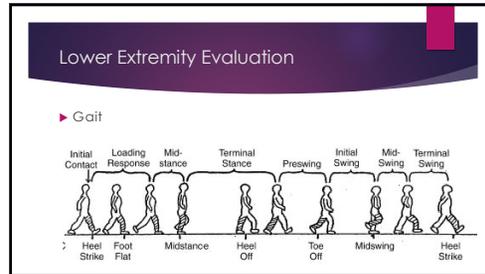
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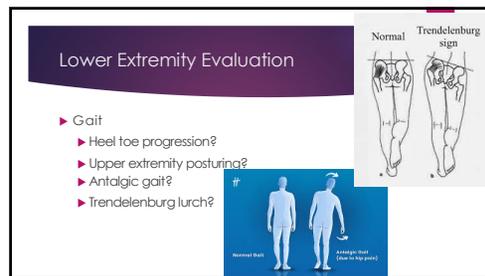
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### Red Flags – resources

- ▶ **\*\*Pediatrician \*\*\*** - one who knows them the best
  - ▶ Get parents, therapists, SW to weigh in- anyone who knows them from a daily basis
- ▶ **Neuro changes:**
  - ▶ Think shunt, hydrocephalus, baclofen, infection
  - ▶ NSGY, Neurology, urgent brain and spine imaging
- ▶ **General care**
  - ▶ Pulmonology
  - ▶ Cardiology
  - ▶ Endocrine
- ▶ **Tone**
  - ▶ Physiatry
- ▶ **Falls, Trauma**
  - ▶ Ortho

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### What can you do? REFER TO US

- **Physiatry:** maximize tone management → ASAP
- **PT and OT:** maximize mobility and strength → ASAP
- **Orthopedics:** hip surveillance, brace recommendations → by age two LATEST

**CARE PATHWAYS**

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### CP hip: AP Pelvis

- Supine
- Abduction/adduction: Neutral
- Hip rotation: Patellae up
- Neutral Pelvic Obliquity; Flattened lordosis
- AI: MP, NSA, HSA, AI

Standard positioning for AP Pelvic radiographs

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### NM hip dysplasia ≠ DDH

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### Hip Surveillance

**Hip surveillance** = process of monitoring and identifying the critical early indicators of hip displacement

**Hip displacement** = displacement of the femoral head laterally out of the acetabulum and is measured using a migration percentage (MP).

**Hip subluxation** = hip displacement where the femoral head is partially displaced from under the acetabulum while hip displacement refers to hip displacement where the femoral head is completely displaced from under the acetabulum.

**Reimer's Migration Index**

- Percentage of the femoral head NOT COVERED by the bony acetabulum
- Can be difficult to measure with pelvic dysplasia
- Error of measurement: -5%
- Surgical Indicators\* of what % vary: 30, 33, 40%...

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### Hip Displacement is Common and Silent

- 35% overall incidence
- Linear increased with GMFCS level

**HIP DISPLACEMENT IN CEREBRAL PALSY**

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### Hip Displacement is Common and Silent

- ▶ Non-ambulatory highest risk
- ▶ Spastic quadriplegia

Motor Type/Topographical Pattern	Present	Not Present	Total
<b>Spastic</b>			
Spastic quadriplegia	1 (5.0%)	27 (95.0%)	28
Spastic paraplegia	13 (52.0%)	12 (48.0%)	25
Spastic quadriplegia	85 (92.3%)	7 (7.7%)	92
Spastic	2 (20.0%)	8 (80.0%)	10
West	7 (23.3%)	23 (76.7%)	30
Ataxic	0 (0.0%)	9 (100.0%)	9
Hydrocephalus	4 (44.4%)	5 (55.6%)	9
<b>Total</b>	114 (25.3%)	329 (74.7%)	443

\*Hip displacement was defined as a migration percentage of >50%. The values are given as the number of children with the percentage in parentheses.

See et al. (2012)

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### Established Hip Surveillance Program: AACPDM

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### GMFCS 1

- ▶ Clinical Assessment at 2 years (no Pelvic X-rays – different than Australia)
- ▶ Repeat @ 4 and 6 years
- ▶ If W & G Type IV hemi surveillance as GMFCS II

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### GMFCS 2

- ▶ Clinical assessment & AP pelvic Xray at 2 years
- ▶ Repeat clinical assessment at age 4 and age 8
- ▶ Repeat clinical assessment and AP Pelvis at age 6 and 10
- ▶ Discharge if MP<30% at age 10 (except for WGH Type IV)



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### GMFCS 3

- ▶ Clinical assessment & AP pelvic Xray at 2 years
- ▶ Repeat yearly until age 8
- ▶ Clinical assessment & AP pelvis every other year from 10 until skeletal maturity
- ▶ Discharge once skeletally mature and MP<30%



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### GMFCS 4 AND 5

- ▶ Clinical assessment & AP pelvic Xray at 2 yrs(or age at initial diagnosis)
- ▶ Repeat q6 monthly until age 4
- ▶ Repeat yearly until skeletally mature
- ▶ Discharge: when skeletally mature and MP<30%



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### Possible Hip Interventions

- ▶ Correction of muscle imbalance by adductor surgery alone has high failure rate (GMFCS the determining factor)
- ▶ Early reconstructive surgery (osteotomies) has high recurrence rate in <6 yo

(Shore et al. J Bone Joint Surg 2012)

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### Possible Hip Interventions

- ▶ RCT, multi-center adductor Botox and Swash brace for "Hips at Risk"
- ▶ BoNT-A q6m x 3y+ abduction brace
- ▶ 46 patients, bilateral spastic CP
- ▶ 40 preventive surgery (21 bot, 19 control)
- ▶ 18 reconstructive surgery (20 bot, 8 control)

**In children with bilateral spastic CP, early treatment with BoNT-A and hip abduction bracing does not reduce the need for surgery or improve hip development at skeletal maturity**

(Willoughby et al. DMCN, 2012)

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### Guided growth

- Abnormal proximal femoral geometry → acetabular dysplasia → hip instability
- Proximal femoral guided growth has shown some efficacy BUT most reports with older kids (>6 yo)

(Ulusaloglu et al. J Child Orthop 2022)

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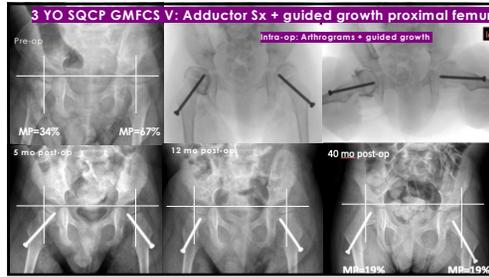
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### New Research on Guided Growth

- ▶ **Guided growth more responsive in younger children (<3 years-old)**
  - ▶ Higher growth rate
- ▶ **High rates of success at 2+ years follow-up**
  - ▶  $\Delta$ MP  $\geq$ 10% (improved, mainly  $\leq$ 3 years)
  - ▶  $\Delta$ MP <10% (no deterioration, for all patients included)
- ▶ **Greater MP improvement was seen w/ higher MP ( $\geq$ 40%) and longer follow-up**
  - ▶ Femoral neck shortening likely contributing

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### Guided Growth Indications

- ▶ **Primary treatment:** MP >40% to <70%, GMFCS IV-V, 18mo to 5-6 yo, +/- adductor spasticity.
  - ▶ Add traditional adductor, gracilis, iliopsoas releases if contractures present
- ▶ **Secondary treatment:** Rescue after VDRO, after time of implant removal.
  - ▶ Documented lateral tilting of physis and MP progression.
  - ▶ Perhaps beneficial for early VDROs as standard to prevent rebound but unknown at this point (risk of fracture at time of implant removal)

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**THANK YOU!**

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**EXTRA SLIDES**

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**CP – surgical goals**

- ▶ **Ambulatory (GMFCS 1-3)**
  - ▶ Improve gait
  - ▶ Upper limb appearance +/- function
- ▶ **Non ambulatory (GMFCS 4-5)**
  - ▶ Make care giving easier
  - ▶ Reduce pain
  - ▶ Improve upper limb hygiene/ function

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**CP – SEMLS**

**Most definitive evidence: RCT Thomson et al JPO**

- CP Group in Melbourne
- 19 matched children randomized, 11 surgical, 8 non-surgical
- Identical rehab
- 85 procedures (mean 8 per child)
- Statistical higher improvements in gait score

**Single stage multi-level surgery (SEMLS or MLS)**

- Muscle lengthening and transfers, and correction of all bony deformities in a single surgical session
- Single rehabilitative period
- Minimal immobilization
- Decreased rate of recurrence of deformities

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