Cerebral Palsy

Kelly Pham, MD
What's New in Medicine
September 7, 2019

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

Financial disclosures

- There are no relevant financial relationships with commercial interests to disclose

Objectives

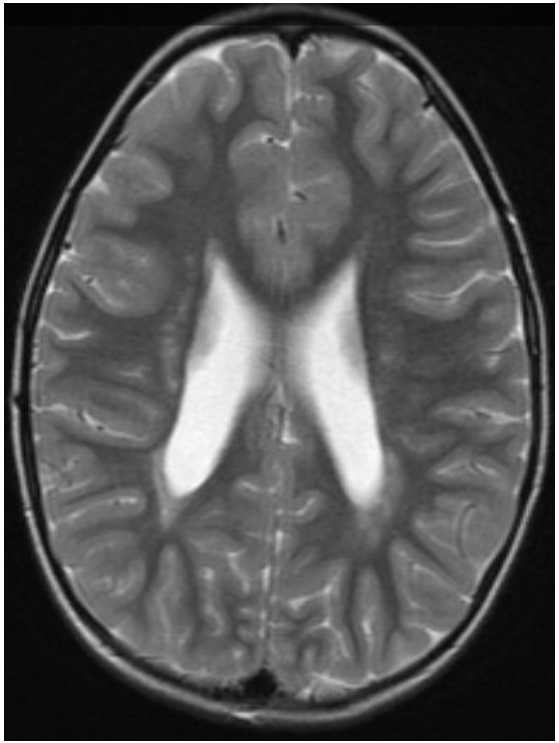
- To define and characterize CP
- To review impairments affecting gait
- To describe the classic gait patterns
- To review tone management treatment
- To describe orthopedic management

CP Basics

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

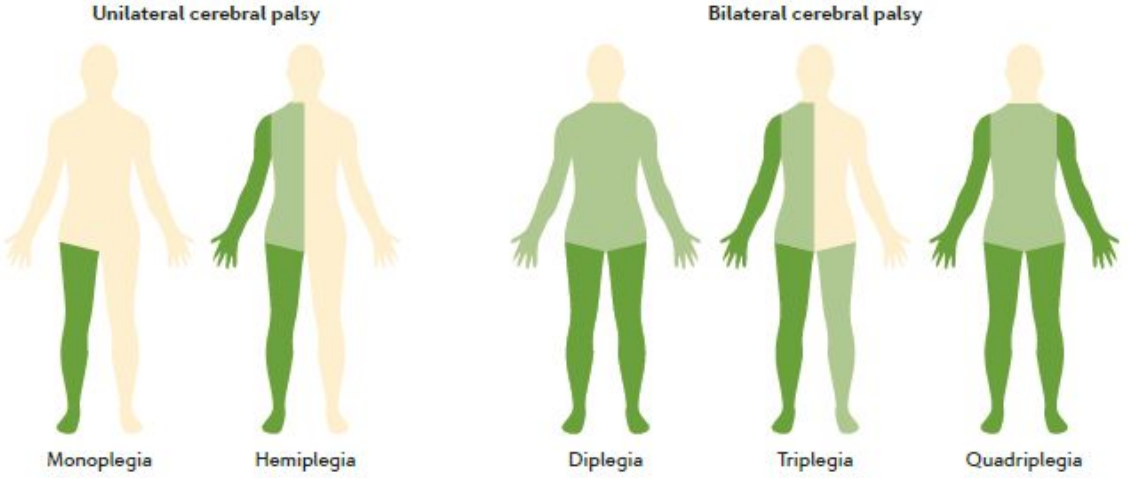
Definition

- Static lesion
- Developing brain
- Motor dysfunction



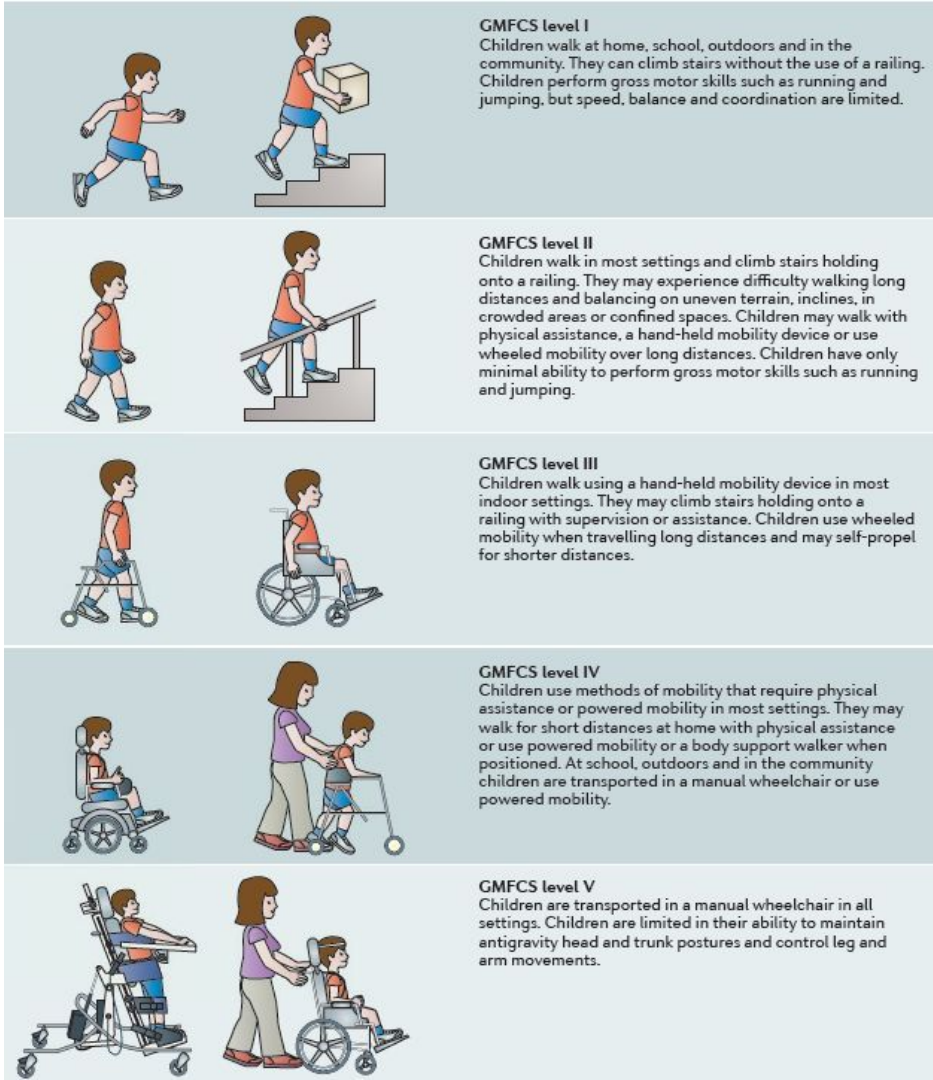
Characterization

- Topography
- Muscle tone/movement disorder
- Functional levels
- Etiology



Graham, Nature Reviews 2016

Gross Motor Function Classification System (GMFCS)



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 use 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 travelling 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.

Graham, Nature Reviews 2016

Impairments Affecting Gait

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

Muscle tone and Movement Disorder

- Spasticity
- Dystonia
- Chorea
- Athetosis
- Hypotonia

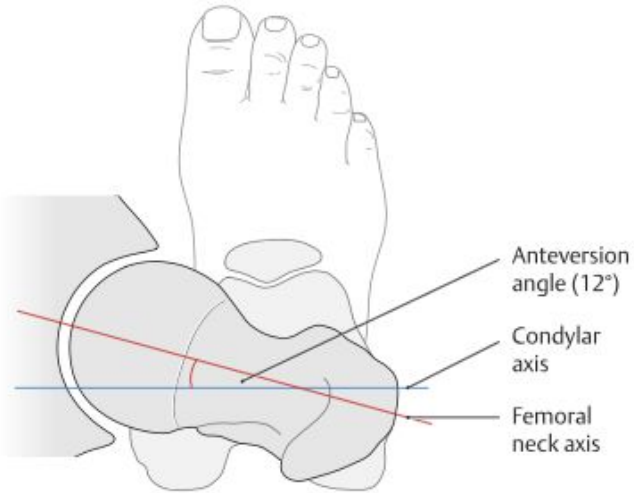
Weakness

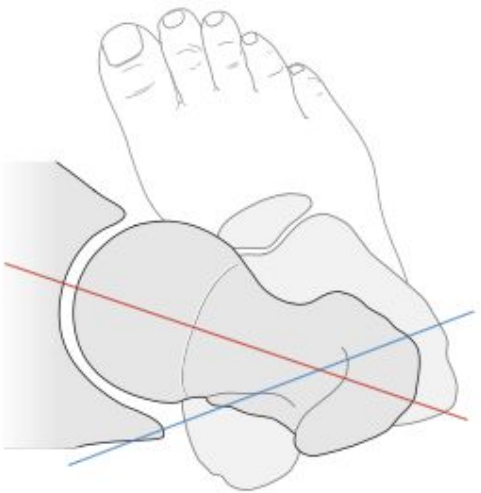
Motor Control

Contractures

Rotational Deformity

- Femoral anteversion
- Tibial torsion



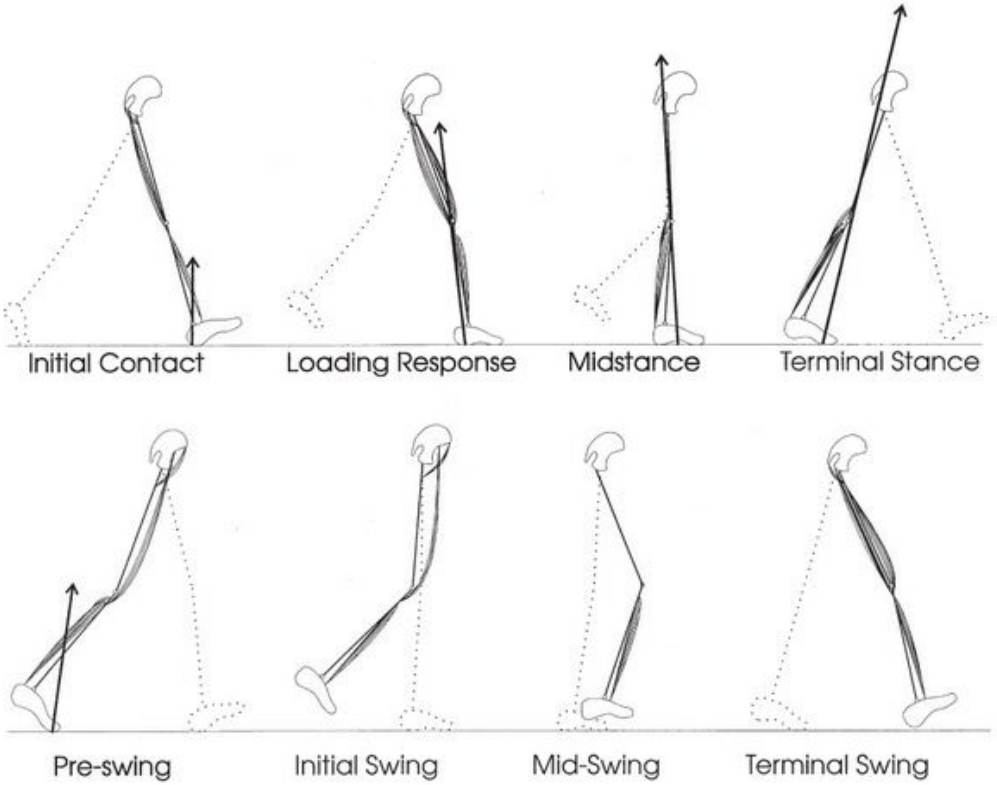


Schuenke, *The Lower Limb,* 2010.

Gait

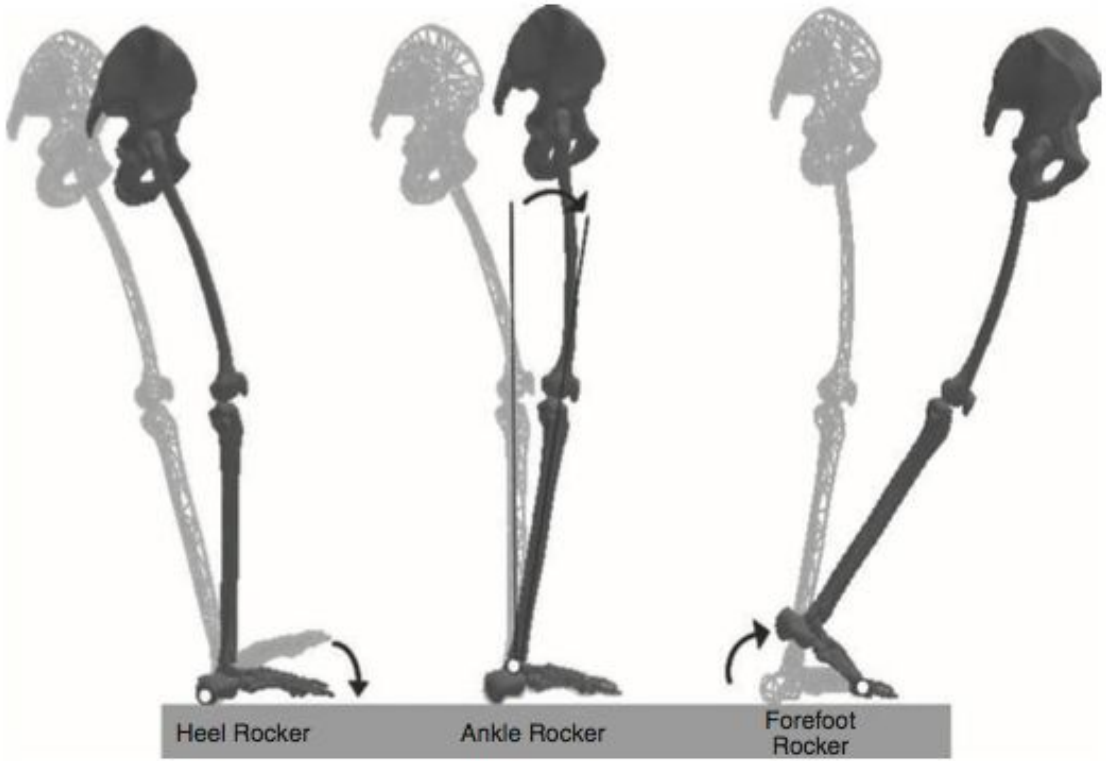
Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

Normal gait



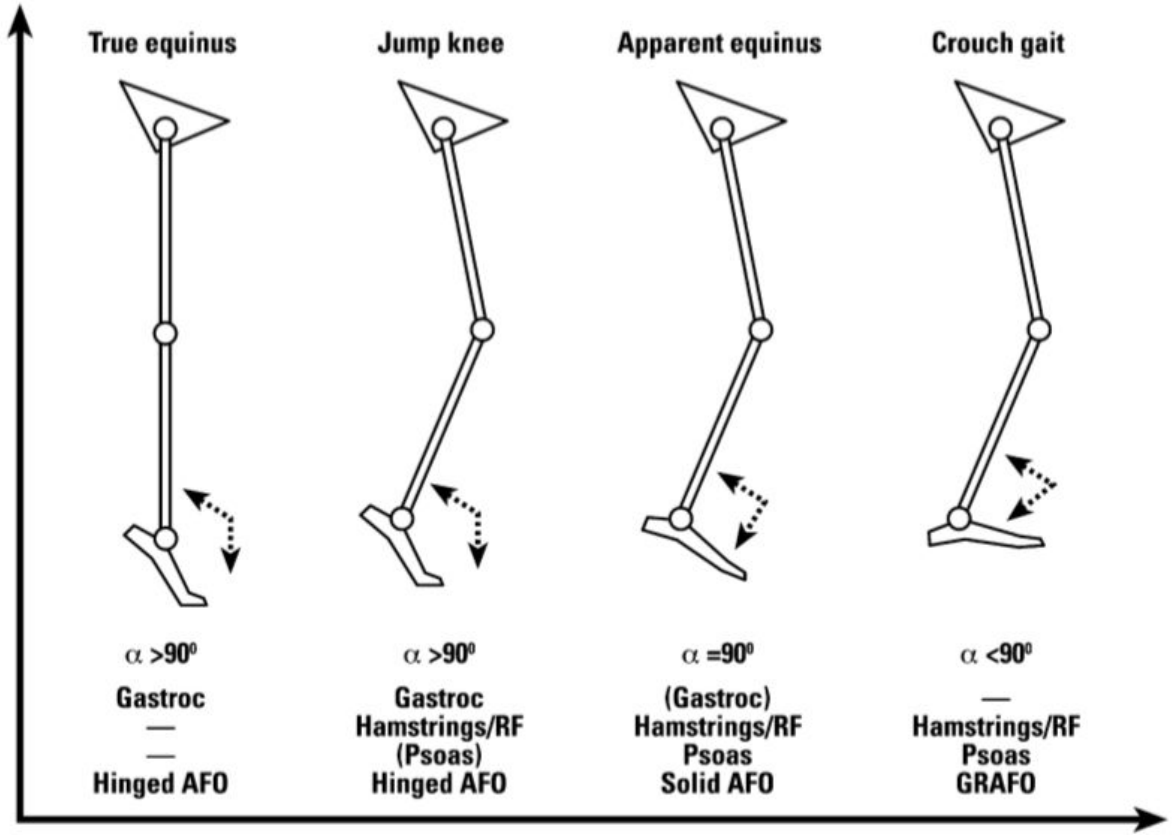
Pease and Bowyer, *Human Walking,* 2010.

Forward momentum in gait



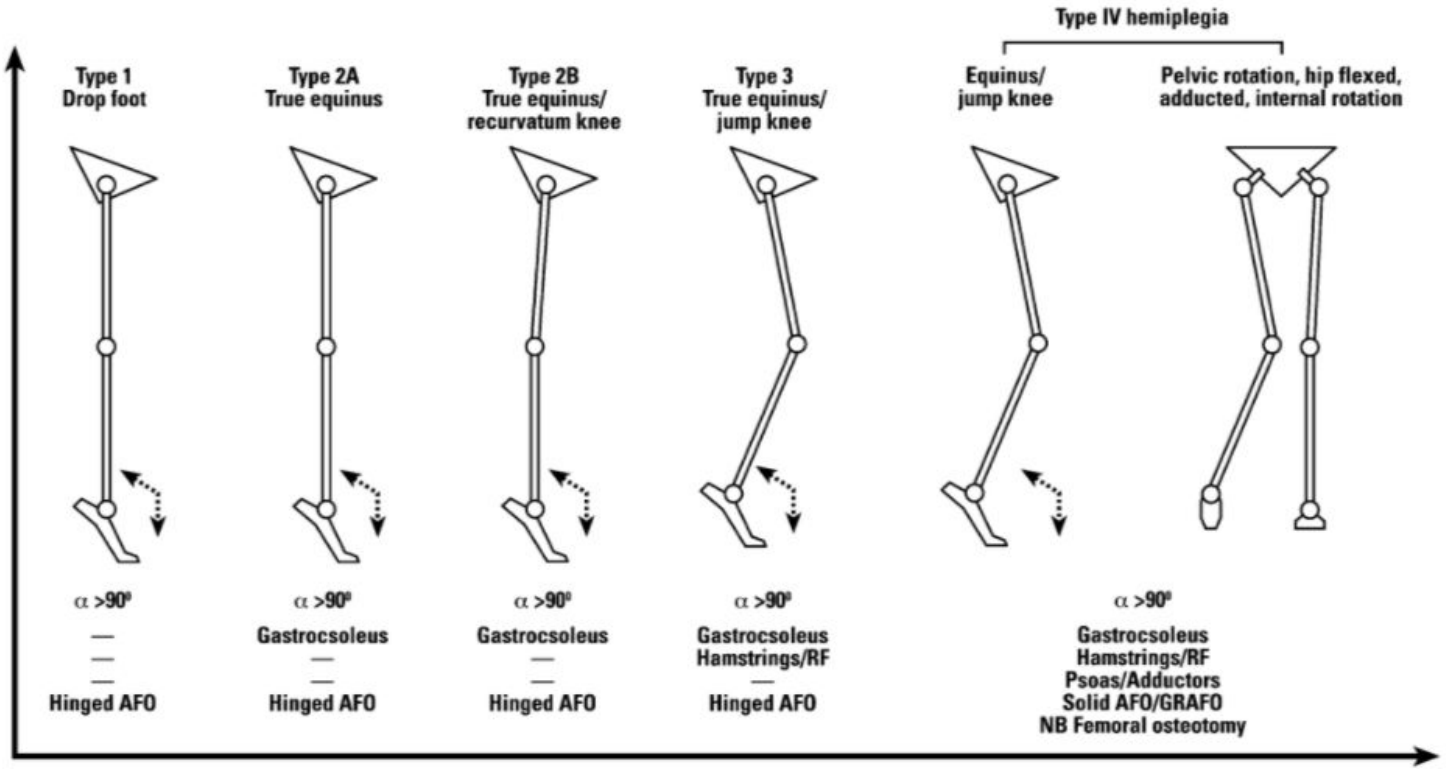
Carallo and Matthews, *Quantitative Assessment of Gait: A Systematic Approach,* 2015.

Common Gait Patterns: Spastic Diplegia



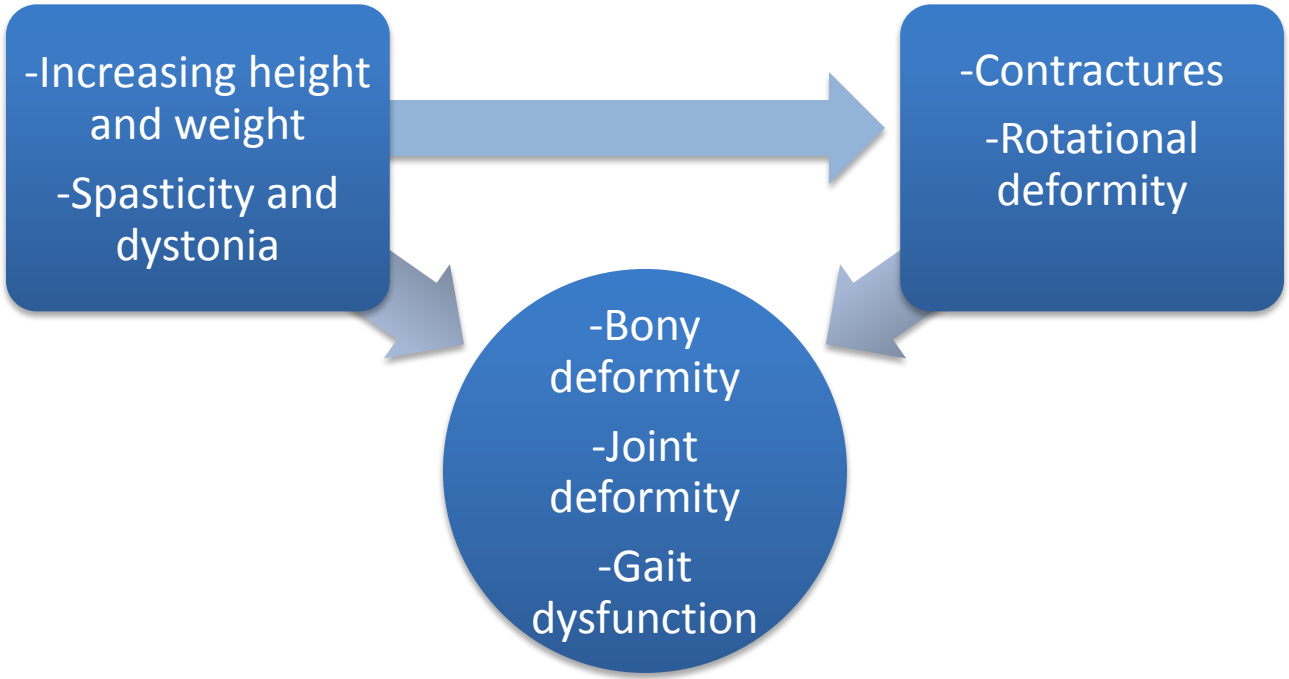
Rodda and Graham, Eur J Neurol 2001

Common Gait Patterns: Spastic Hemiplegia



Rodda and Graham, Eur J Neurol 2001

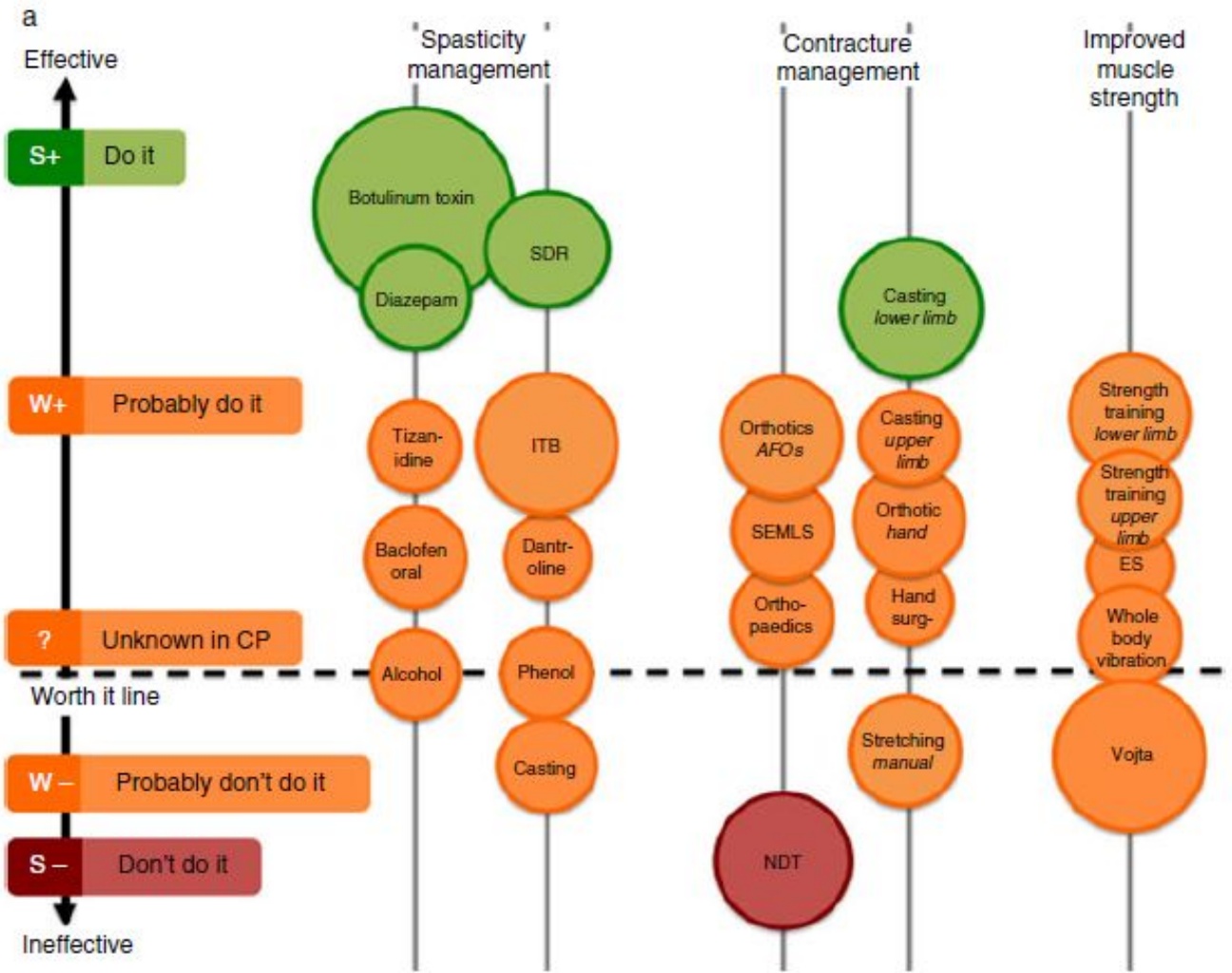
Aging with CP



Intervention

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

Evidence

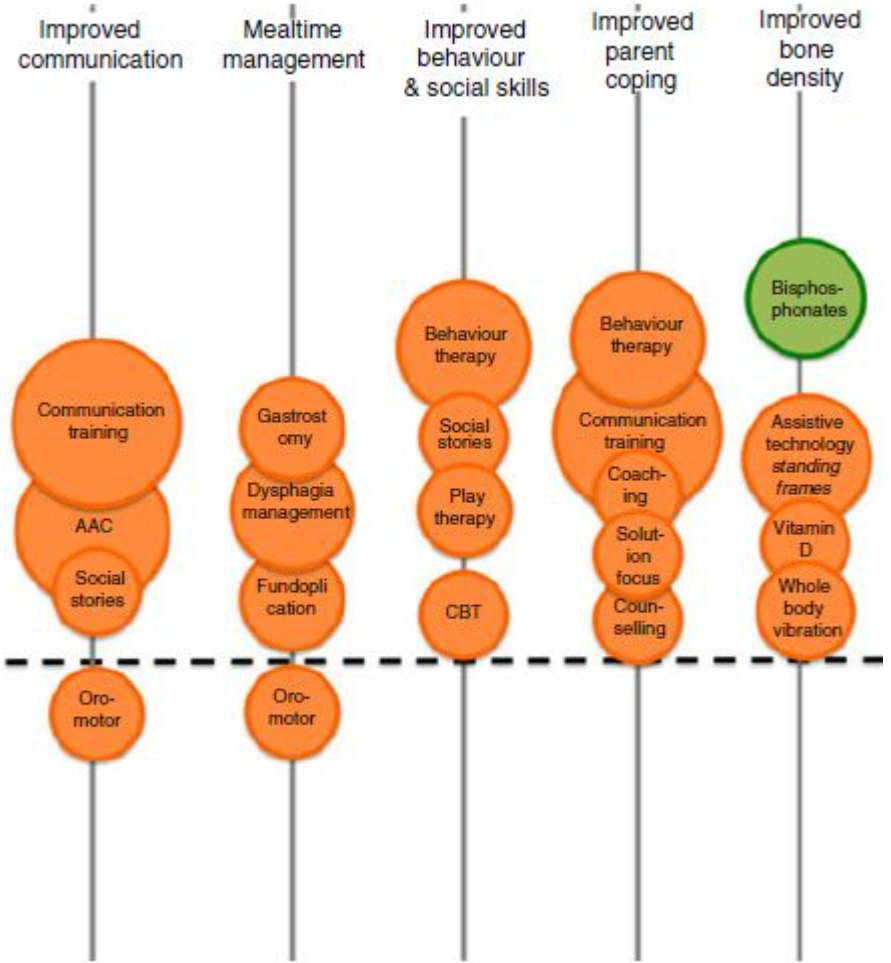


Novak, Dev Med Child Neurol 2013

Evidence

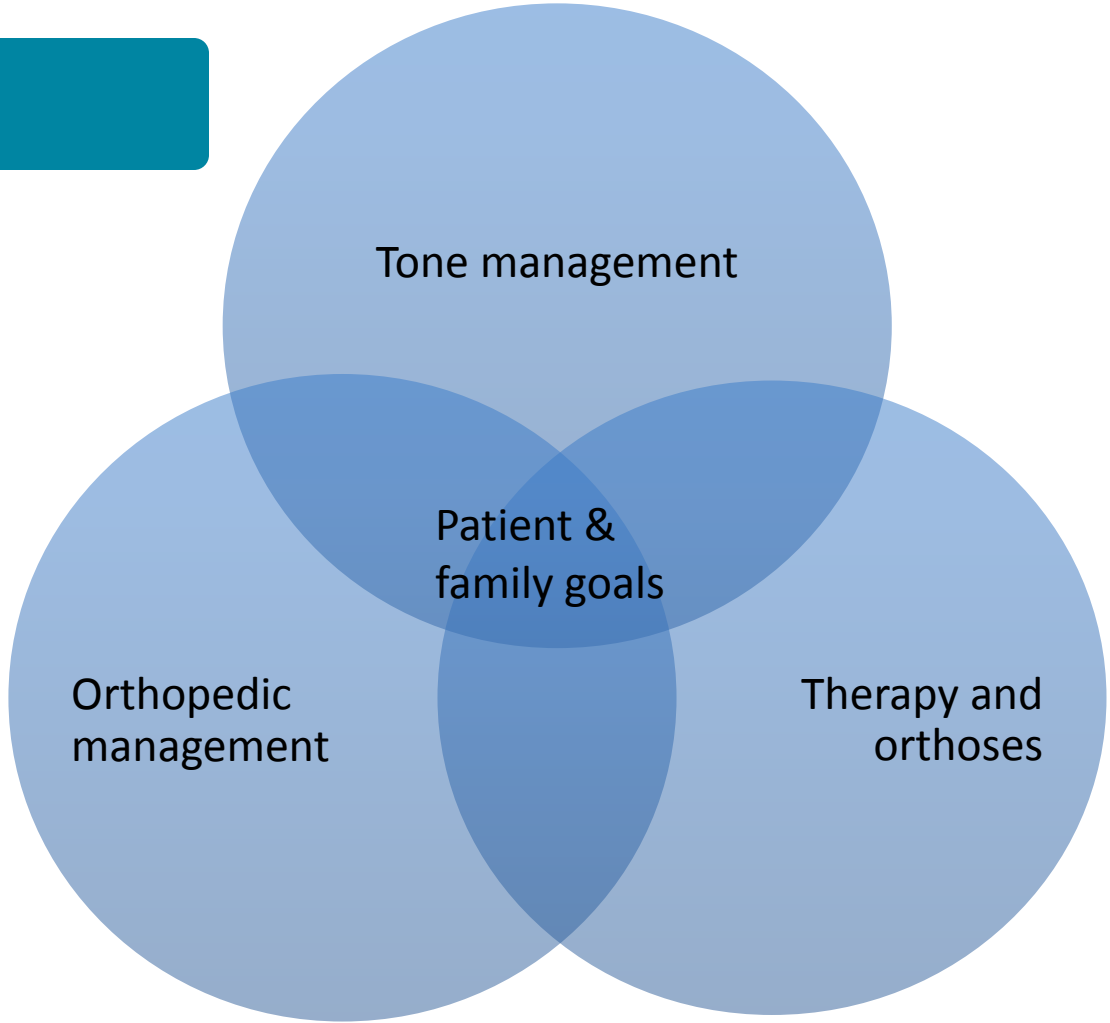
Novak, Dev Med Child Neurol 2013

Evidence



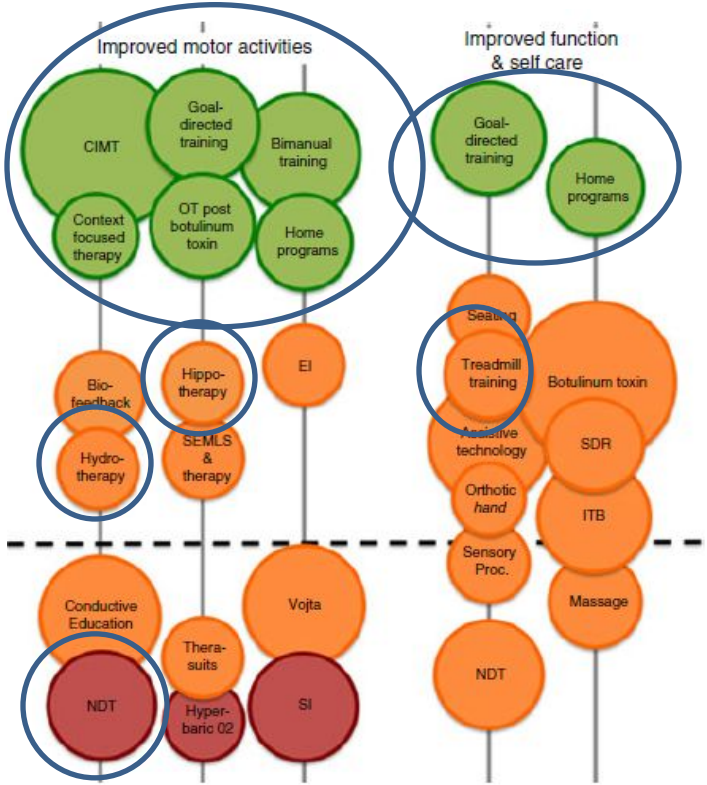
Novak, Dev Med Child Neurol 2013

Goals


Tone management
Patient &
family goals
Orthopedic
management
Therapy and
orthoses

Therapy

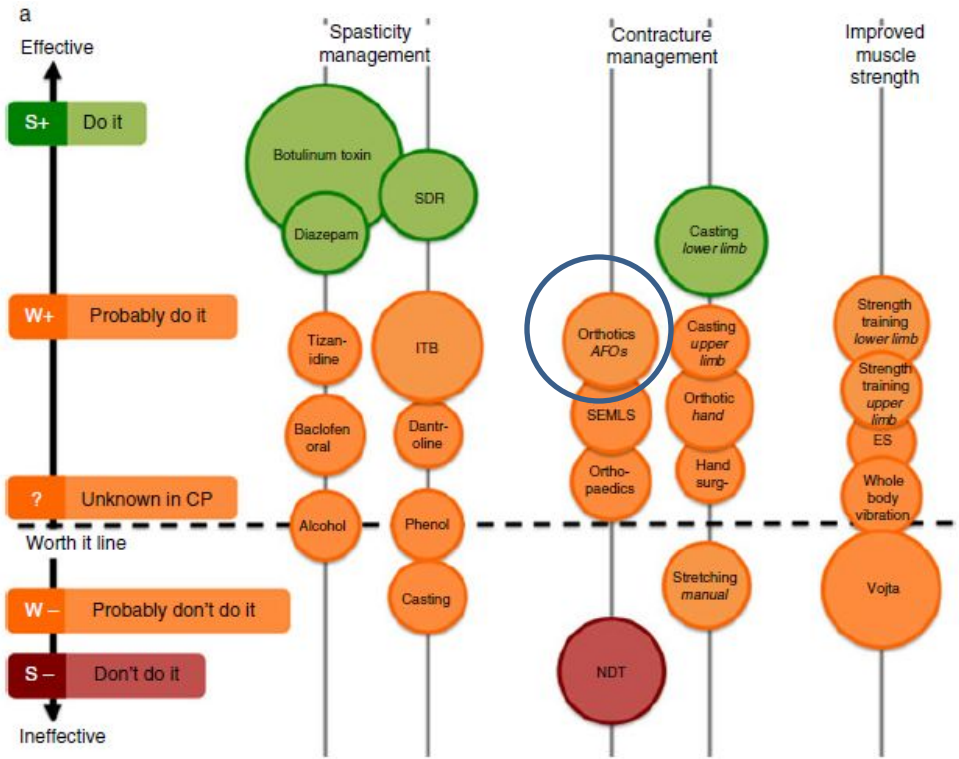
Therapy


Improved motor activities
Improved function & self care
CIMT
Goal-directed training
Bimanual training
Context focused therapy
OT post botulinum toxin
Home programs
Goal-directed training
Home programs
Bio-feedback
Hippo-therapy
EI
Hydro-therapy
SEMLS & therapy
Seating
Treadmill training
Botulinum toxin
Assistive technology
SDR
Orthotic hand
ITB
Sensory Proc.
Massage
Conductive Education
Vojta
Thera-suits
NDT
Hyper-baric 02
SI
NDT

Orthoses

Cascade Orthotics

Orthoses

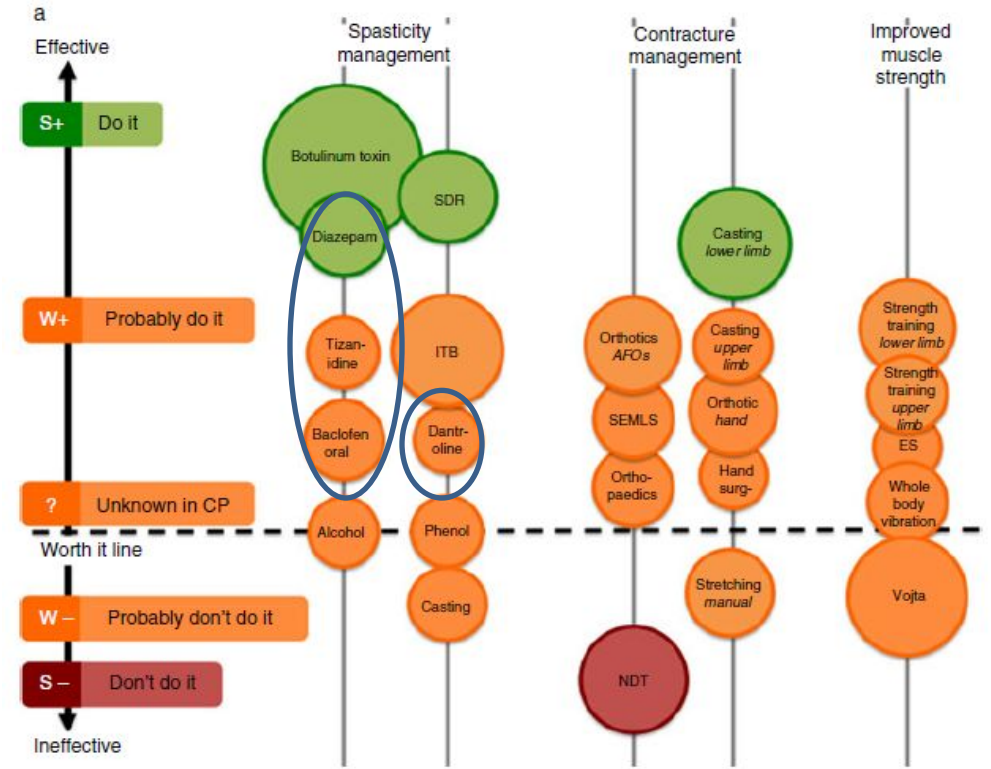

a
Effective
S+
Do it
W+
Probably do it
?
Unknown in CP
Worth it line
W –
Probably don't do it
S –
Don't do it
Ineffective
Spasticity management
Botulinum toxin
Diazepam
SDR
Tizan-idine
ITB
Baclofen oral
Dantr-oline
Alcohol
Phenol
Casting
Contracture management
Orthotics AFOs
Casting lower limb
Casting upper limb
SEMLS
Orthotic hand
Ortho-paedics
Hand surg-
Stretching manual
NDT
Improved muscle strength
Strength training lower limb
Strength training upper limb
ES
Whole body vibration
Vojta

Hypertonicity and movement disorder management

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

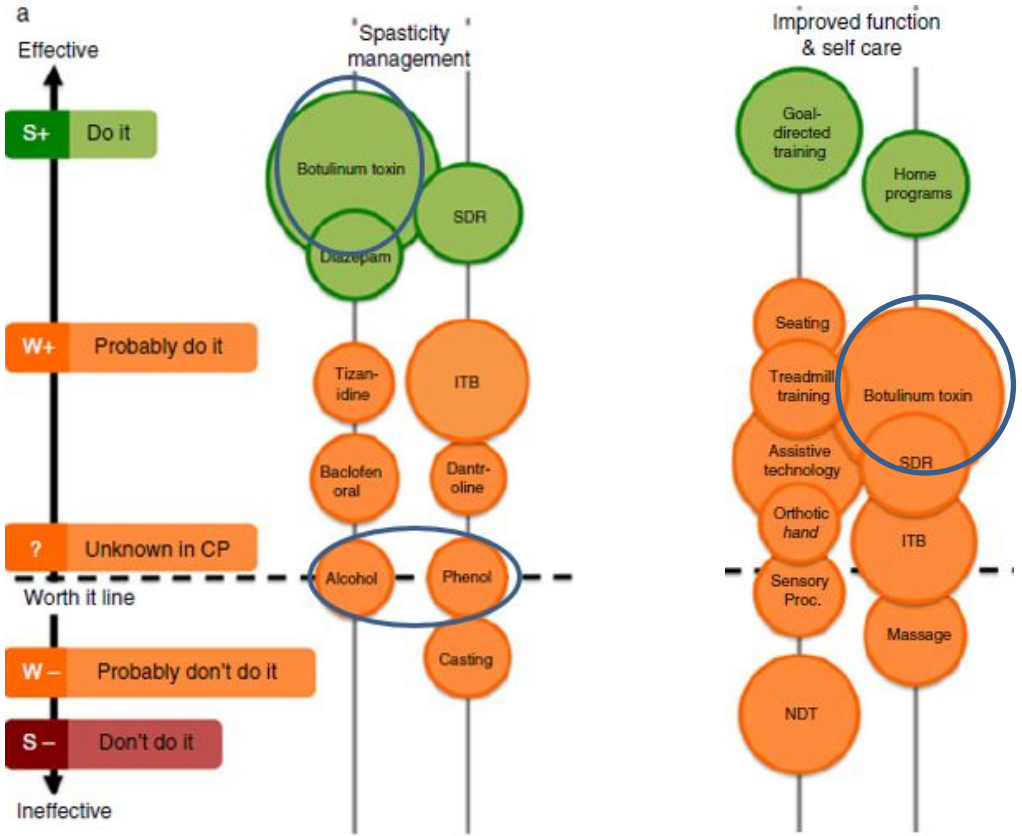
Enteral medications

- Baclofen
- Diazepam
- Artane
- Sinemet
- Dantrolene

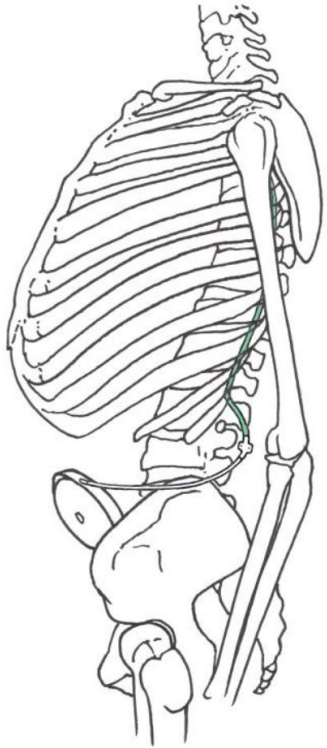


Chemodenervation and chemoneurolysis

- Botulinum toxin
- Phenol

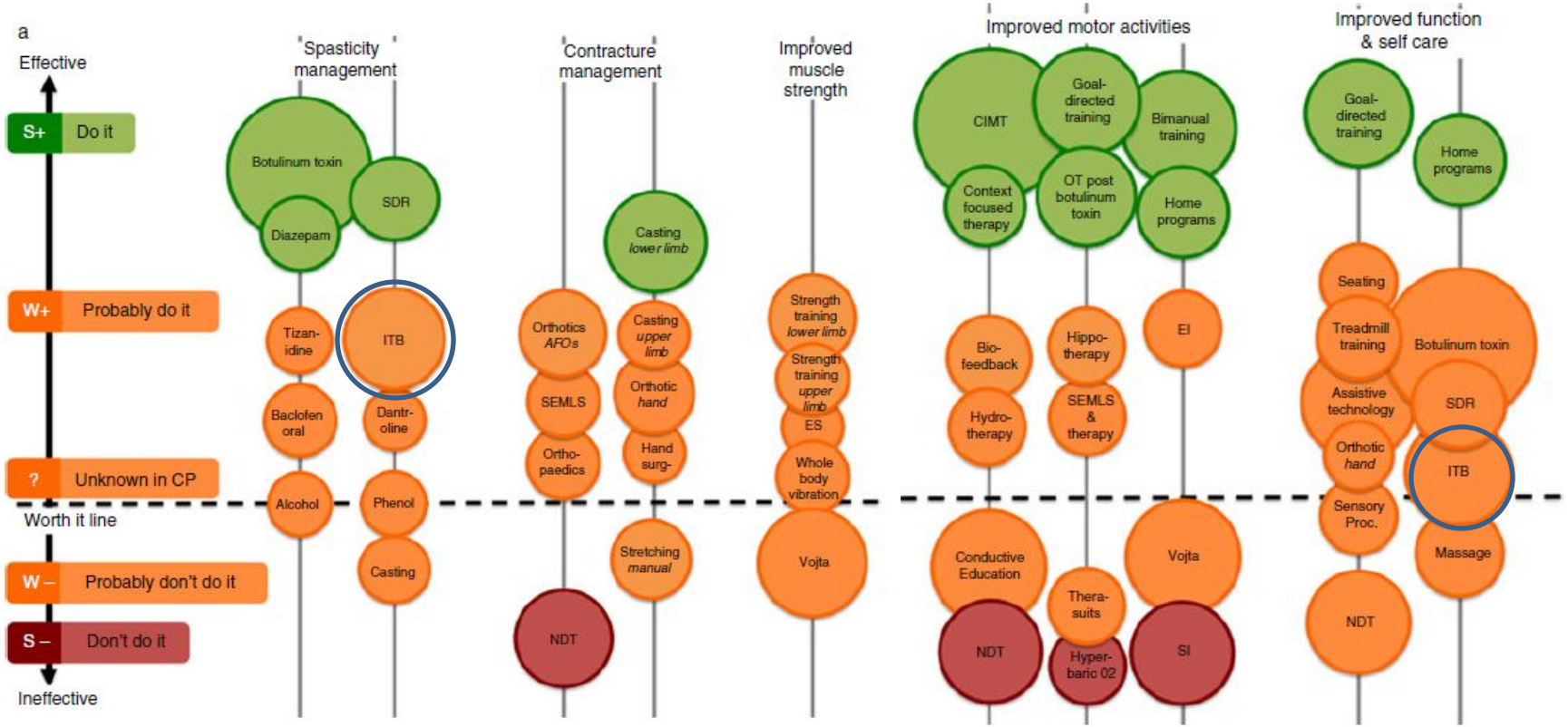


Intrathecal baclofen (ITB) pump



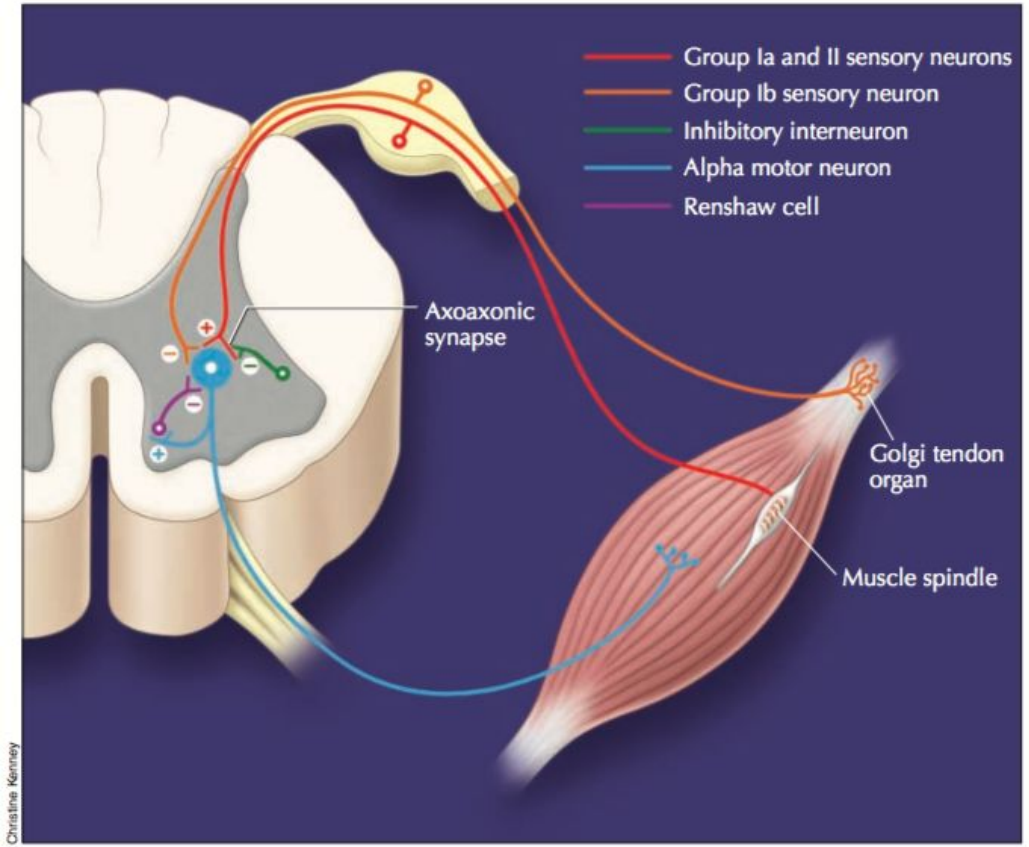
Medtronic

Intrathecal baclofen (ITB) pump



Selective dorsal rhizotomy (SDR)

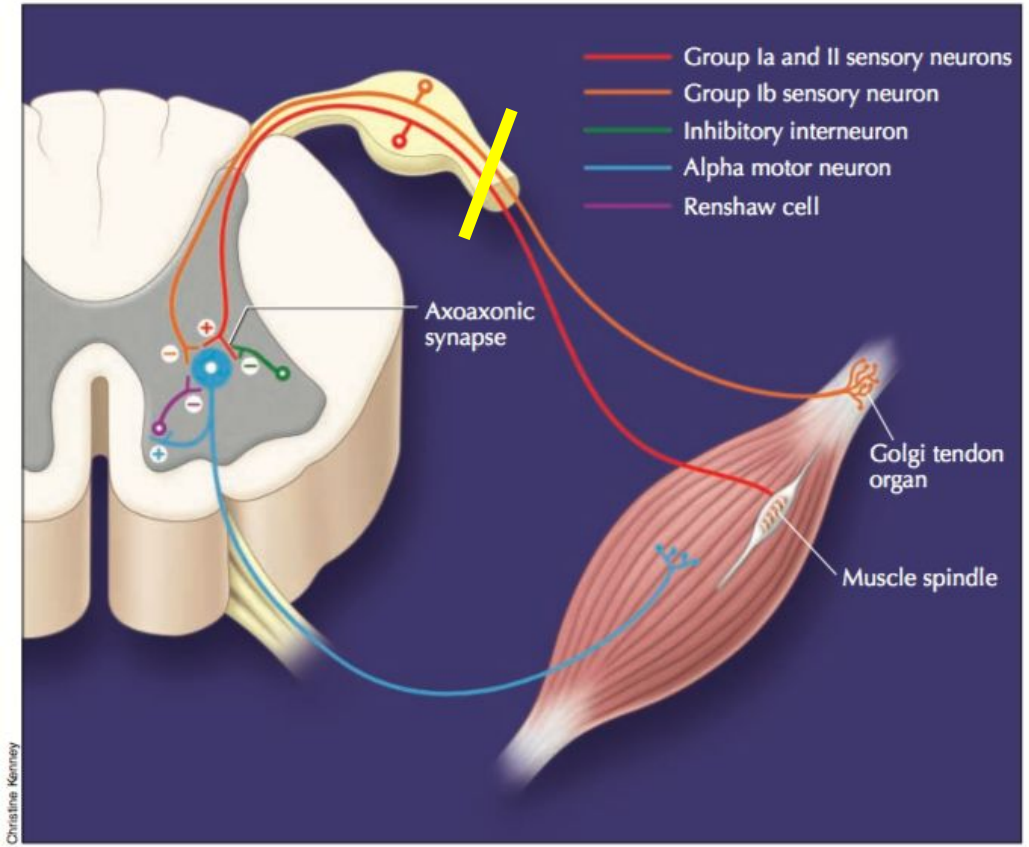
- How a stretch reflex works
  - Sensory input from the muscle
  - Integration of sensory information in the spinal cord
  - Output of information to the muscle
- How spasticity affects this
  - Loss of inhibition from the brain
    - over-activity of the muscle



Satkunam, CMAJ, 2003.

Selective dorsal rhizotomy (SDR)

- How SDR affects this
  - Decreases sensory input into the spinal cord decreasing abnormal muscle output

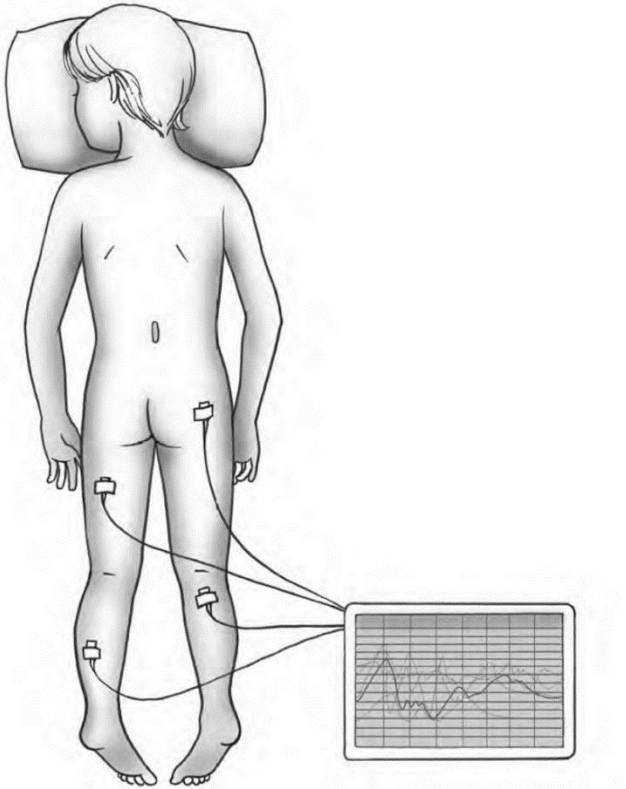


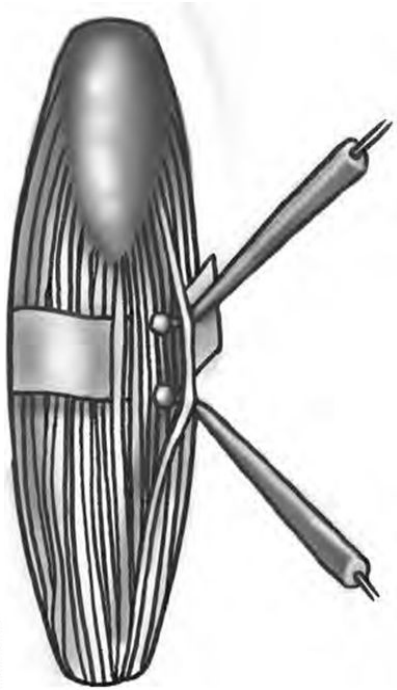
Satkunam, CMAJ, 2003.

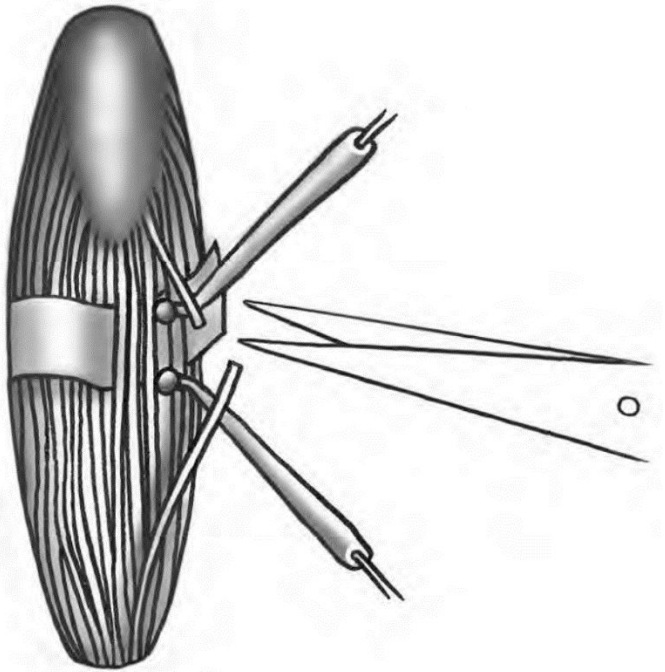
Selective dorsal rhizotomy (SDR)

- Selection criteria
  - Age
  - Strength
  - Motor control
  - Spasticity
  - Participation

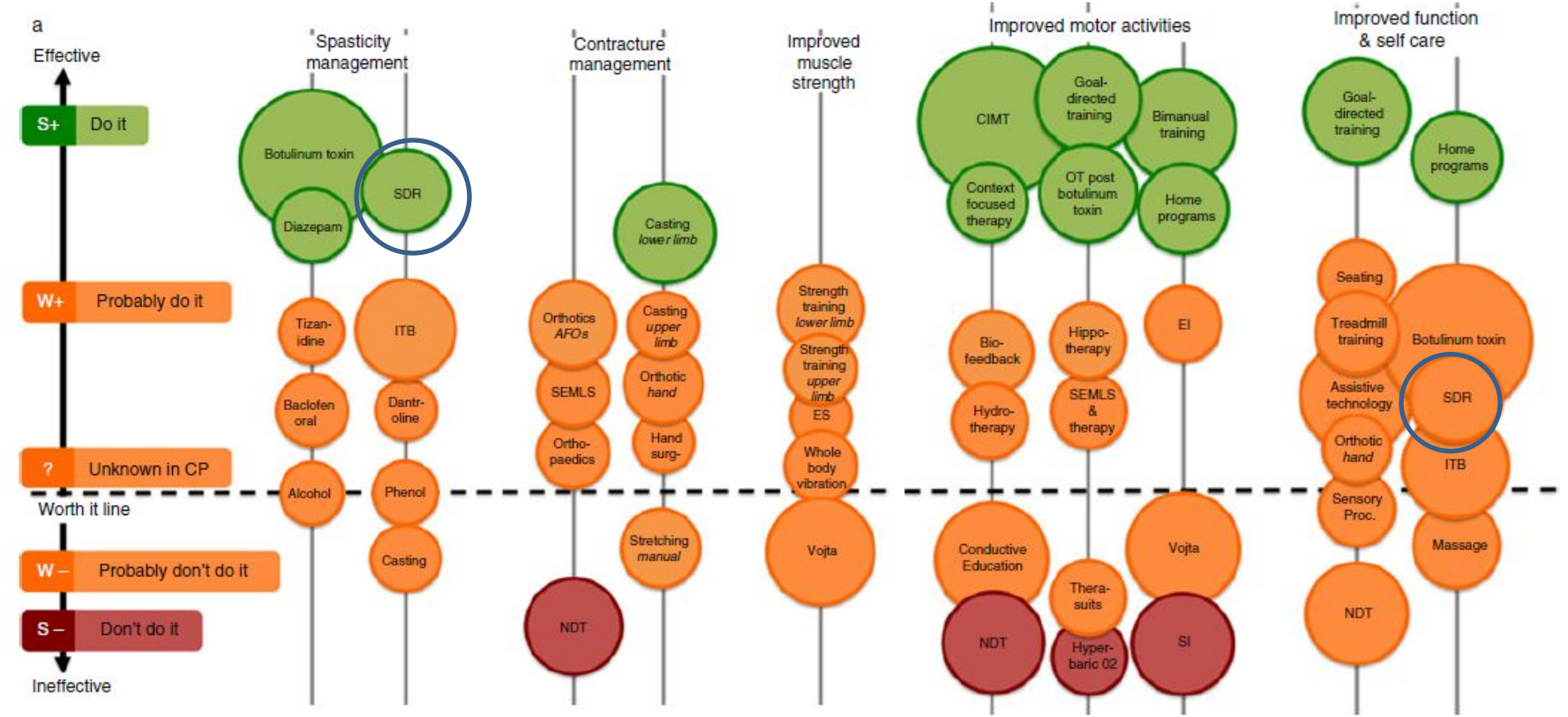
Selective dorsal rhizotomy (SDR)







Selective dorsal rhizotomy (SDR)



SDR Patient Case

- 4yo history of 30wk prematurity
  - Imaging: cystic changes per report from family
  - GMFCS III:
    - Reciprocal crawl
    - Ambulates with walker
    - Independent transfers on/off of toilet and to walker
    - Manual wheelchair for distances

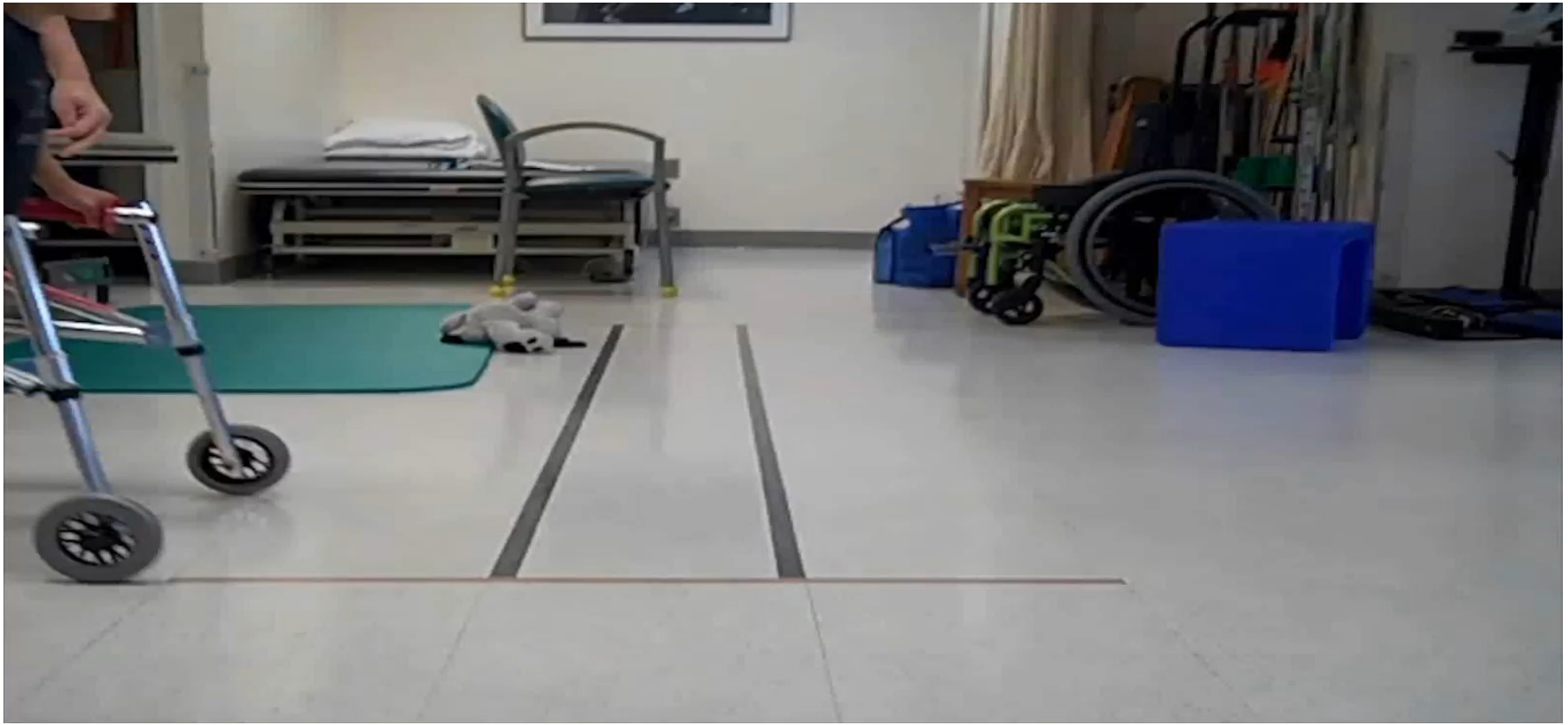
SDR Patient Case

- Range of Motion: Left Right

Hip E-F:	0-120^0	0-120^0
Hip Abd:	0-25^0	0-15^0
Popliteal:	-60^0	-60^0
Knee E-F:	0-150^0	0-150^0
Ankle DF with KE:	-5^0	-5^0

- Spasticity: 1/4 hip abductors, quads and ankle dorsiflexors, 3/4 hip adductors, hamstrings and plantarflexors
- Strength: unable to isolate hip flexion, 2+/5 hip abduction, 3/5 hip extension, knee extension and knee flexion, ankle DF and PF not isolated

SDR Patient Case – pre-op video



SDR Patient Case

- Recommended Intervention:
  - Selective dorsal rhizotomy
    - Strength
    - Isolated motor control
    - Spasticity
    - No dystonia
    - Rehabilitation potential

SDR Post-op Rehabilitation

- Intensive inpatient rehabilitation
  - Strengthening
  - ROM, stretching, serial casting
  - Retraining patterns of movement
- Intensive outpatient rehabilitation

SDR Patient Case – 1yr post-op video

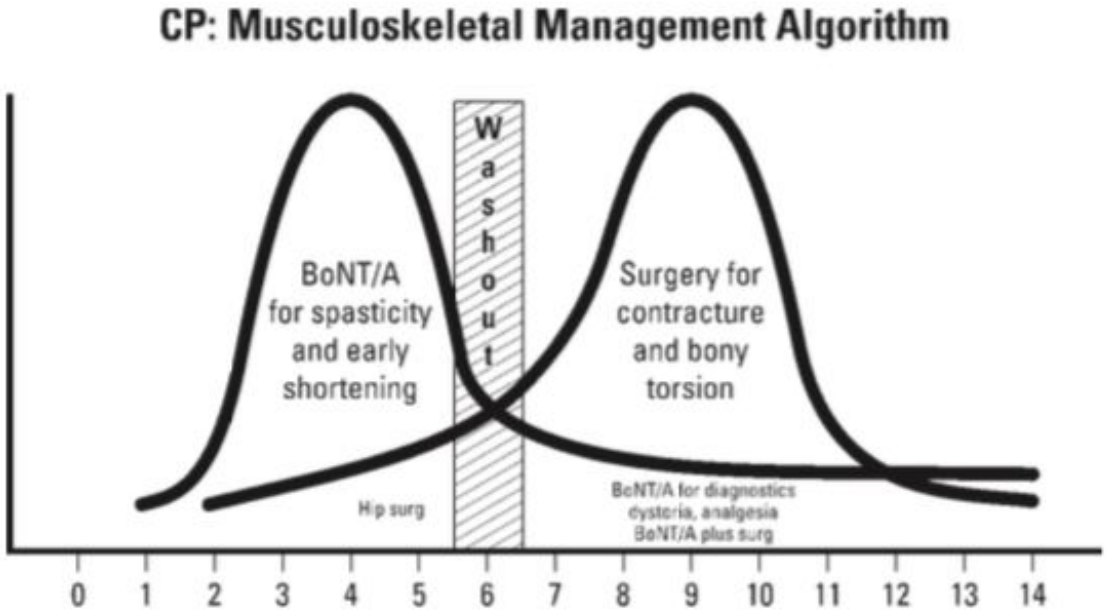
Orthopedic management

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

Contracture management

- Serial casting
- Botox injections

Contracture management



Graham, Thomason, Novacheck. *Cerebral Palsy,* 2014.

Contracture management

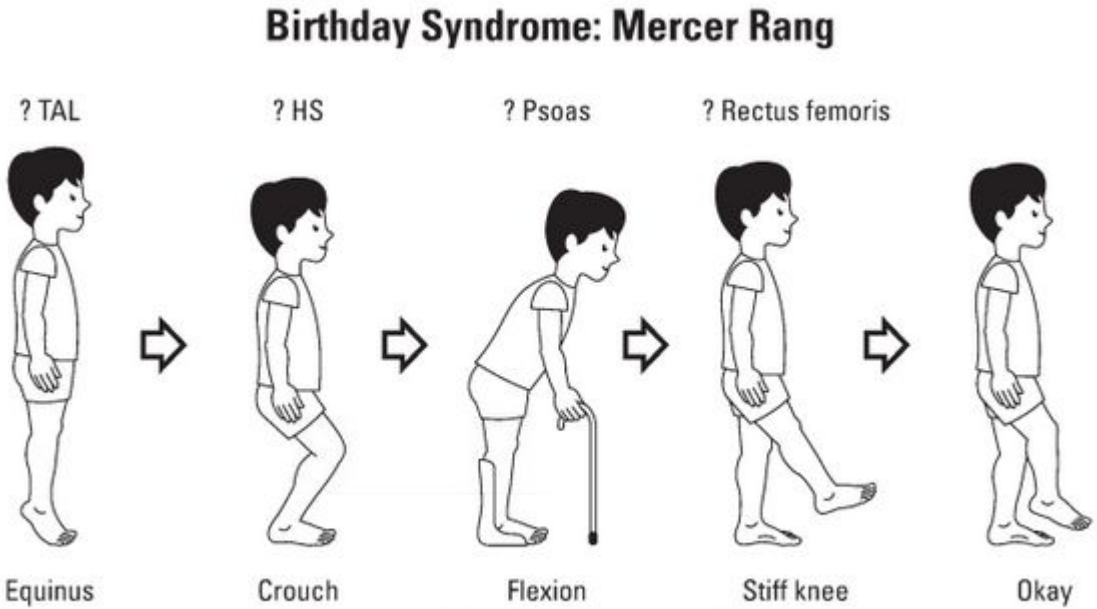
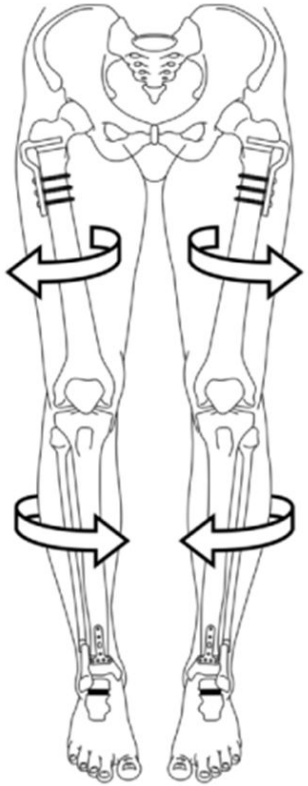


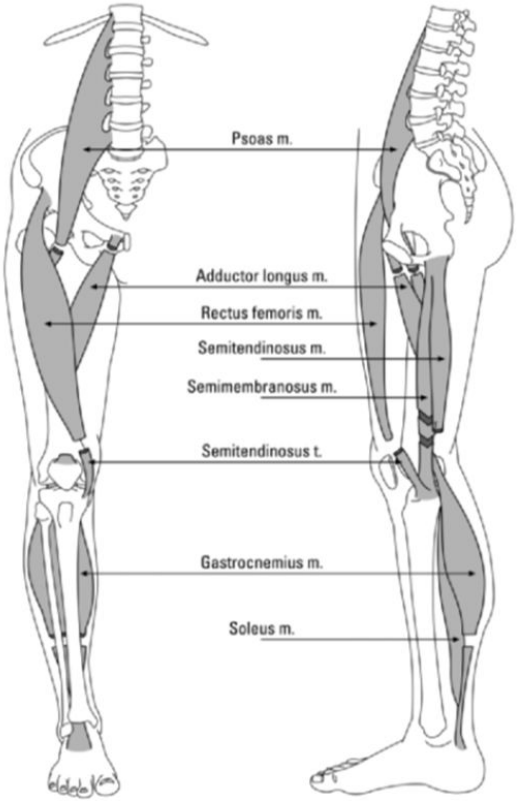
FIGURE 14-20. The Birthday Syndrome as described by Mercer Rang.

Graham, Thomason, Novacheck. *Cerebral Palsy,* 2014.

Contracture management

- SEMLS = Single Event Multilevel Surgery





Graham, Thomason, Novacheck. *Cerebral Palsy,* 2014.

SEMLS Patient Case 1

- 5yo history of 30wk prematurity
  - Imaging: unavailable
  - GMFCS III:
    - Bunny hops and knee walks at home
    - Walks short distances with posterior walker
    - Manual wheelchair in the community

SEMLS Patient Case 1

- Range of Motion: Left Right

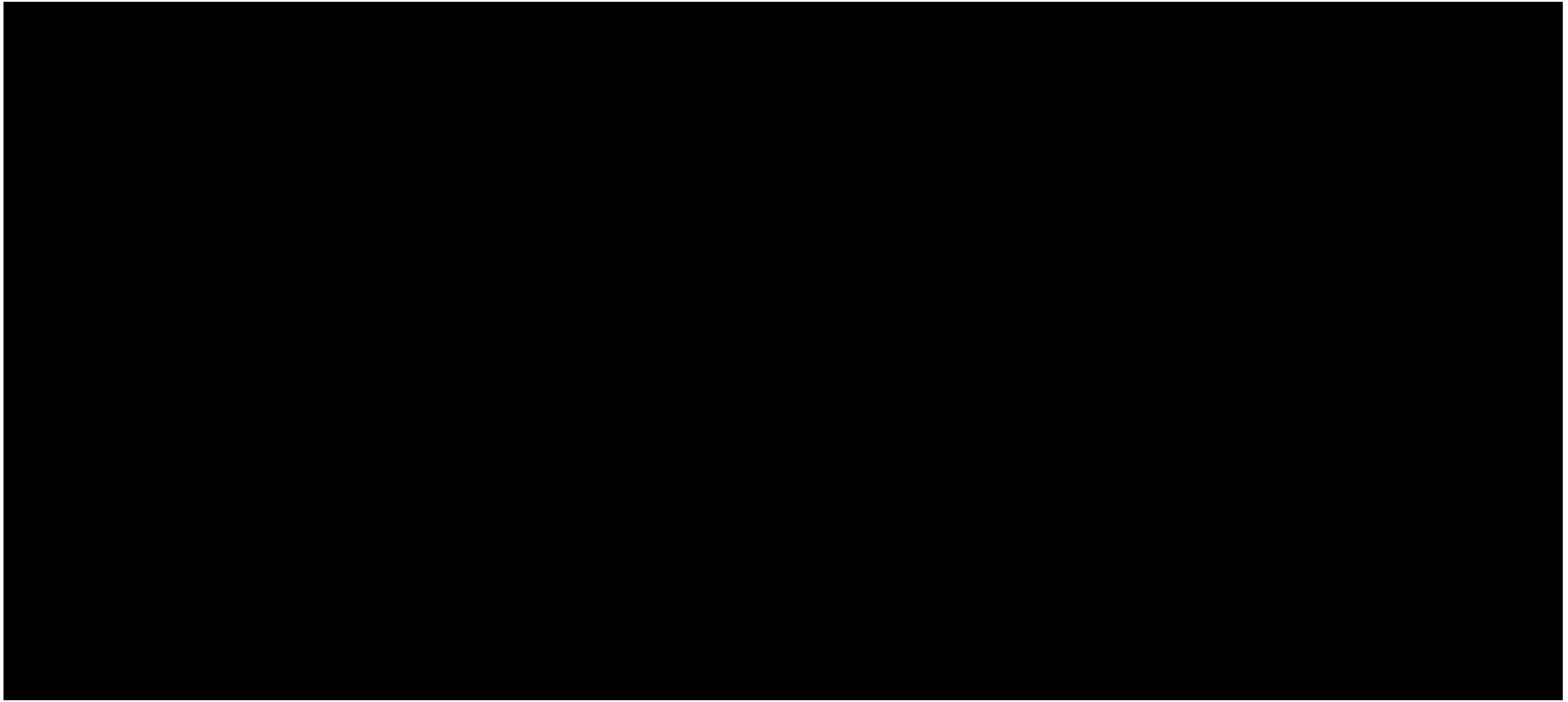
Hip E-F:	-30-120^0	-30-120^0
Hip Abd:	0-20^0	0-20^0
Hip IR:	0-60^0	0-60^0
Hip ER:	0-40^0	0-40^0
Popliteal:	-70^0	-60^0
Knee E-F:	-15-140^0	-15-140^0
Ankle DF with KE:	-30^0	-30^0

- Spasticity: Grossly grade 2/4 on MAS hip add, hamstrings, plantarflexors
- Strength: Good underlying strength though difficult to assess in a 5yo

SEMLS Patient Case 1

- Recommended Intervention:
  - Soft tissue lengthening of
    - Bilateral hip adductors
    - Bilateral iliopsoas
    - Bilateral medial hamstrings
    - Bilateral tendo-achilles

SEMLS Patient Case 1



SEMLS Patient Case 2

- 8yo history of 27wk prematurity
  - Imaging: Intraventricular hemorrhage (IVH)

- GMFCS III:
  - Crawls at home
  - Walks with tripod canes in school and home
  - Manual wheelchair in community

SEMLS Patient Case 2

- Prior interventions:
  - Therapy (community, school, hippotherapy)
  - Baclofen
  - Serial casting
  - Botulinum toxin injections

SEMLS Patient Case 2

- Range of Motion:

	left	right
Hip E-F:	-35-110^{0}	-30-120 ^{0}
Hip Abd:	0-15^{0}	0-20 ^{0}
Hip IR:	0-25^{0}	0-50 ^{0}
Hip ER:	0-30 ^{0}	0-20 ^{0}
Popliteal:	-90 ^{0}	-90 ^{0}
Knee E-F:	-30-140 ^{0}	-20-140 ^{0}
Ankle DF with KE:	-20 ^{0}	-20 ^{0}

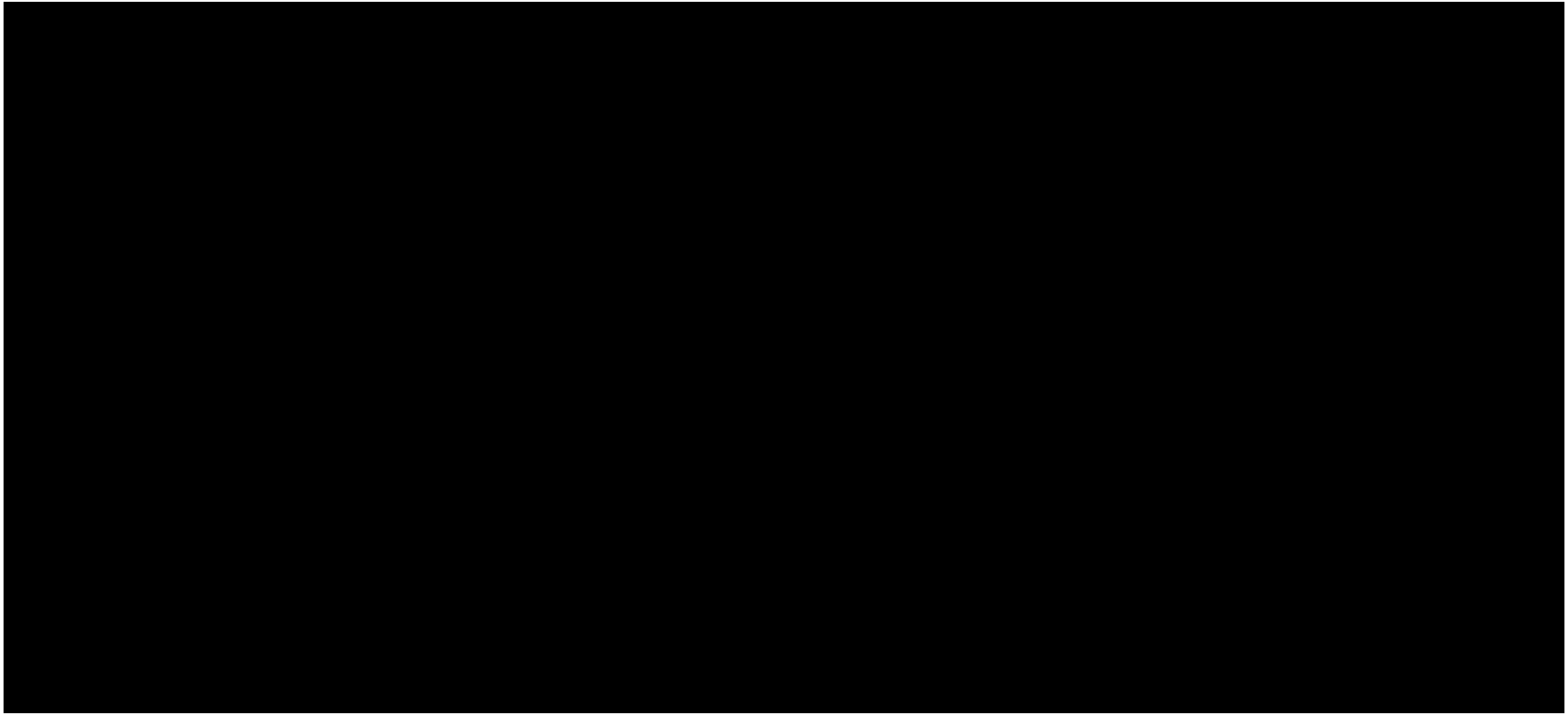
- Tone: Grossly grade 2/4 on MAS hip add, hamstrings, plantarflexors

- Strength: good underlying strength fighting against considerable contractures.

SEMLS Patient Case 2

- Recommended procedure:
  - Soft tissue lengthening
    - Bilateral hip adductors
    - Bilateral iliopsoas
    - Bilateral medial hamstrings
    - Bilateral tendo-achilles
    - Bilateral peroneus brevis
  - Bony procedure
    - Bilateral calcaneal lengthening

SEMLS Patient Case 2



Summary

- Impairments affecting gait
  - Hypertonicity and movement disorder
  - Weakness
  - Motor control
  - Contracture
  - Rotational deformity
- Typical gait patterns for diplegia and hemiplegia
- Intervention based on goals
  - Therapy
  - Orthoses
  - Tone management
    - Enteral medications
    - Botox and phenol injections
    - ITB
    - SDR
  - Orthopedic management
    - SEMLS

Thank you

Seattle Children's®
HOSPITAL • RESEARCH • FOUNDATION

References

- Carallo JJ, Matthews DJ. Quantitative Assessment of Gait: A Systematic Approach. In Alexander MA, Matthews DJ, Murphy KP. *Pediatric Rehabilitation Principles and Practice. 5th Edition.* New York, NY: Demos Medical; 2015.
- Graham HK, Rosenbaum P, Paneth N, Dan B, et al. Primer: Cerebral Palsy. *Nature Rev.* 2016.
- Graham HK, Thomason P, Novacheck TF. Cerebral palsy. In Weinstein SL, Flynn JM. *Lovell & Winter's Pediatric Orthopaedics. 7th Edition.* Philadelphia, PA: Lippincott Williams & Wilkins; 2014.
- Novak I, Mcintyre S, Morgan C, Campbell L, et al. A systematic review of interventions for children with cerebral palsy: state of the evidence. *Dev Med Child Neurol.* 2013; 55: 885-910.
- Pease WS, Bowyer BL. Human walking. In: Frontera WR, Delisa JA, Gans BM, Walsh NE, Robinson LR, et al. *DeLisa's Physical Medicine & Rehabilitation Principles and Practice. 5th Edition.* Philadelphia, PA: Lippincott Williams & Wilkins; 2010.
- Rodda J, Graham HK. Classification of gait patterns in spastic hemiplegia and spastic diplegia: a basis for a management algorithm. *Eur J Neurol.* 2001;8(5):98-108.
- Satkunam LE. Rehabilitation Medicine: 3. Management of adult spasticity. *CMAJ.* 2003; 169 (11): 1173-9.
- Schuenke M, Schulte E, Schumacher U. The Lower Limb. In: Ross LM, Lampert ED. *Atlas of Anatomy General Anatomy and Musculoskeletal System.* Stuttgart, Germany: Thieme; 2010.