How Cerebral Palsy Affects Muscles, Bones and Joints

CP-Achieve Webinar, November 30th 2022

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Disclosures

- National Health & Medical Research Council of Australia, CP-Achieve
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- Compliance with institutional requirements
- I have no financial disclosures

A short personal history

- Trained in London and Toronto in Children's Orthopaedics.
- First Clinician to use Botox for children with Cerebral Palsy, 1990.
- Needed a Gait Lab to measure the effects of Botox and surgery.
- Exchanged the patent on using Botox for first Gait Lab.
- In 1994 invited to set up the first Gait laboratory in Australia at RCH
- Hugh Williamson Gait Lab, Hip Surveillance, SEMLS surgery, SDR
- Retired from RCH October 31st 2022 after 28 years
- Professor of Orthopaedic Surgery UoM and 35 years of CP research.

How cerebral palsy affects muscles, bones and joints

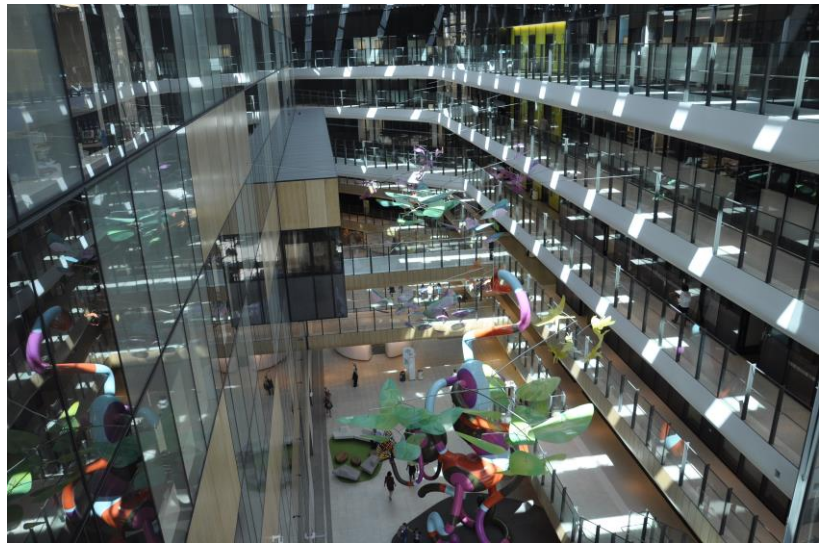
Cerebral palsy is by definition a condition which affects the brain, yet over time the effects on muscles (contractures) bones (twist or torsion) and joints (dislocation) can be profound. Population based studies from the Orthopaedic Department RCH, based on the Victorian Cerebral Palsy Register, have led to the identification of risk factors and causal pathways. These are very important for screening and guiding management. Musculoskeletal deformities can cause pain, impair function reduce participation and impair quality of life. The role of the Orthopaedic team is to identify problems at an early stage, when they are easier to correct. The long-term goals are to maintain the health of the musculoskeletal system in children and teenagers who have cerebral palsy and to prevent premature "wearing out" and arthritis.

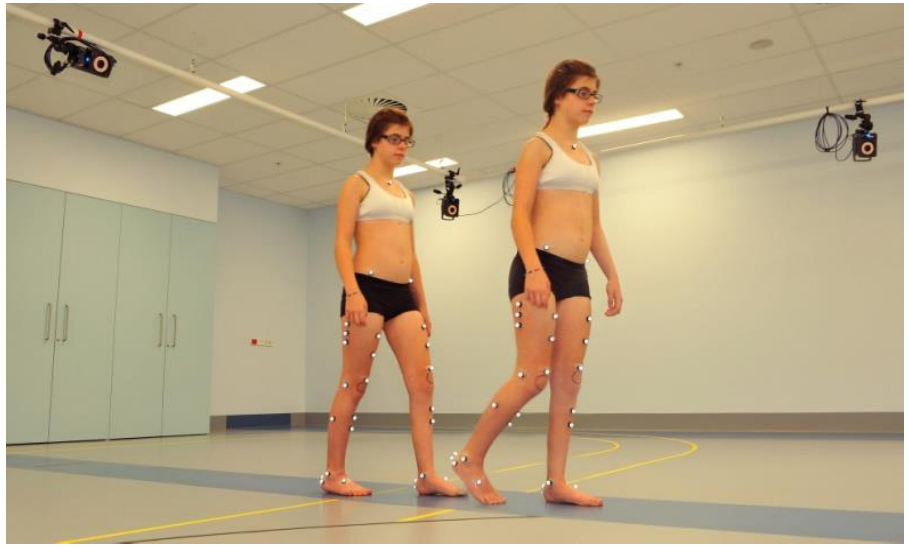
How cerebral palsy affects muscles, bones & joints

Today: general introduction- it's a big subject

1. Foot and ankle problems in children with hemiplegia
2. Knee problems in children with diplegia.
3. The Gait Laboratory and SEMLS surgery
4. Hip Surveillance and Hip Health
5. Spinal curvature, scoliosis and new surgery to straighten the spine.
6. Problems with feet, toes, orthotics and walking.
7. Botox update: when is it useful?

The Royal Children's Hospital, Melbourne





Thanks to: Hugh Williamson Gait Laboratory Team: Engineers, Physiotherapists and Surgeons

RCH CP Team: Understaffed and Over worked

- Gait Laboratory: Manager and Senior Physiotherapist: Pam Thomason
- Senior Engineer: Dr Elyse Passmore.
- Gait Lab Physiotherapists: Jess Pascoe, Annette O'Donnell, Jamie Kemp, Liz Heeps
- Nurse Co-Ordinator: Mela Harambasic
- Hip Surveillance Physiotherapist: Dr Kate Willoughby
- Surgeons: Professor Erich Rutz, Mr Abhay Khot: we need more
- Waiting lists are the longest in 30 years: Covid-19 and staffing issues

1. CP Definition: A *Neuromusculoskeletal* Disorder

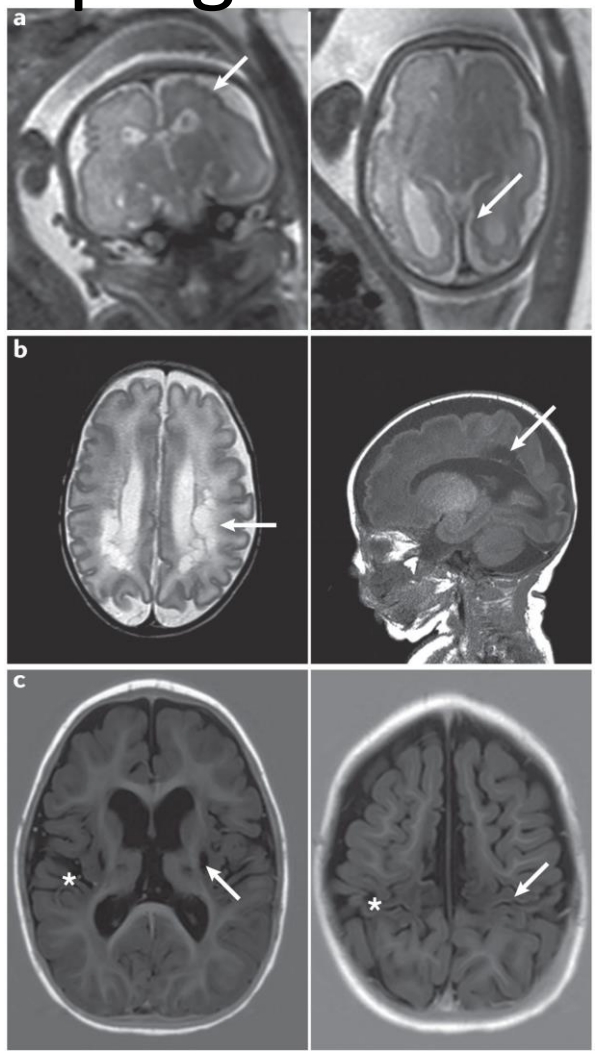
A group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.

The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.

Definition & Classification of Cerebral Palsy: Rosenbaum et al

2. “*non-progressive* disturbances (*lesions*) that occurred in the developing fetal or infant brain.”

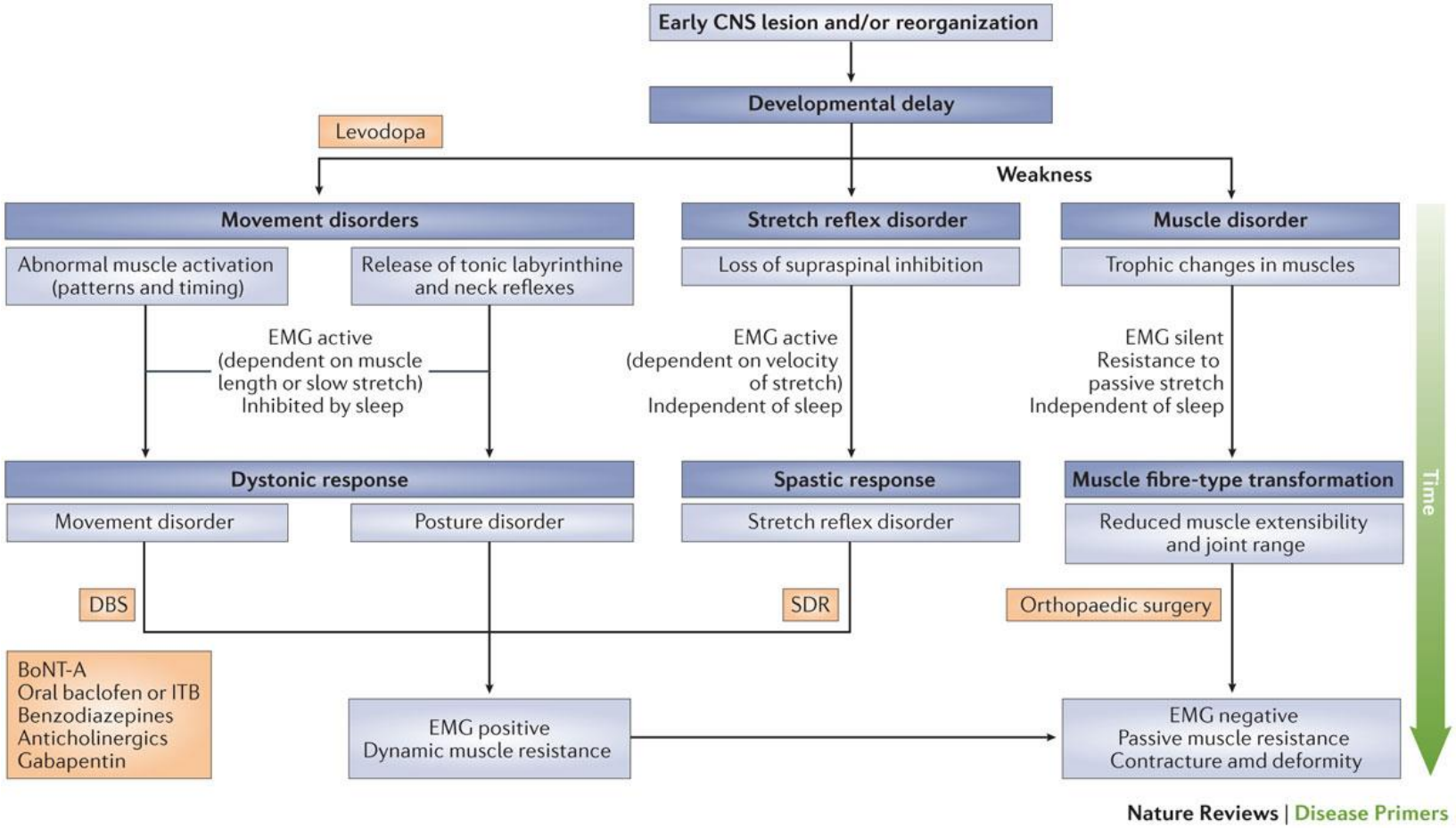
Static
encephalopathy



Nature Reviews | Disease Primers

Graham, H. K. *et al.* (2015) Cerebral palsy
Nat. Rev. Dis. Primers doi:10.1038/nrdp.2015.82

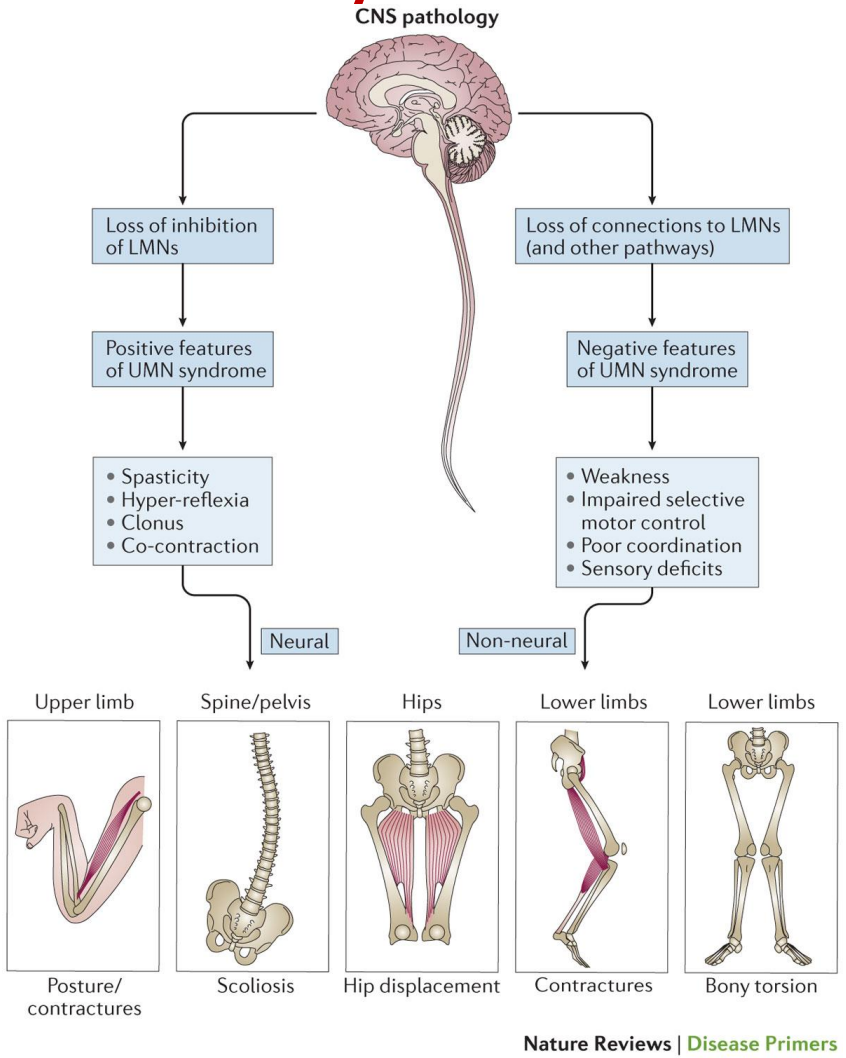
3. “A group of permanent disorders of the development of movement and posture”



Graham, H. K. *et al.* (2015) Cerebral palsy
Nat. Rev. Dis. Primers doi:10.1038/nrdp.2015.82

4. The motor disorders of cerebral palsy are accompanied by *progressive* secondary musculoskeletal problems.

Progressive Musculoskeletal pathology (MSP)



Graham, H. K. *et al.* (2015) Cerebral palsy *Nat. Rev. Dis. Primers* doi:10.1038/nrdp.2015.82

The Victorian Cerebral Palsy Cohort Study: Birth Years: 1990-1992

J. Paediatr. Child Health (2005) 41, 479–483

Cerebral palsy in Victoria: Motor types, topography and gross motor function

Jason Howard,[1] Brendan Soo,[1,4] H Kerr Graham,[1,4,5] Roslyn N Boyd,[1,2,4] Sue Reid,[3,4] Anna Lanigan,[3,4] Rory Wolfe[6] and Dinah S Reddihough[3,4,5]

Royal Children's Hospital, Departments of [1]Orthopaedics, Neonatology[2], and Child Development and Rehabilitation[3], Murdoch Childrens Research Institute[4], Departments of Paediatrics, University of Melbourne[5], and Epidemiology and Preventitive Medicine, Monash University[6], Melbourne, Victoria, Australia

■ CHILDREN'S ORTHOPAEDICS

Proximal femoral geometry in cerebral palsy

A POPULATION-BASED CROSS-SECTIONAL STUDY

J. Robin, H. Kerr Graham, P. Selber, F. Dobson, K. Smith, R. Baker

1. 3 year inception cohort, from VCPR.
2. N=374 children with Cerebral Palsy
3. GMFCS, Motor Types, Howard et al, 2005
4. Hips and GMFCS, Soo et al, 2006
5. Scoliosis and GMFCS: Willoughby et al, 2021

HIP DISPLACEMENT IN CEREBRAL PALSY

BY BRENDAN SOO, MBBS, JASON J. HOWARD, MD, FRCS(C), ROSLYN N. BOYD, PHD, MSC(PHYSIOTHERAPY), SUSAN M. REID, MCLINEPI, ANNA LANIGAN, RN, RORY WOLFE, PHD, DINAH REDDIHOUGH, MD, FRACP, FAFRM, AND H. KERR GRAHAM, MD, FRCS(ED), FRACS

Investigation performed at the Royal Children's Hospital, Murdoch Children's Research Institute, University of Melbourne, Parkville, Victoria, Australia

Journal of Paediatrics and Child Health

doi:10.1111/jpc.15707

ORIGINAL ARTICLE

Epidemiology of scoliosis in cerebral palsy: A population-based study at skeletal maturity

Kate L Willoughby,[1,2] Soon Ghee Ang,[1,†] Pam Thomason,[2,3] Erich Rutz,[1,2,3] Benjamin Shore,[1,‡] Aaron J Buckland,[1] Michael B Johnson[1,2] and H Kerr Graham[1,2,3,4]

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1: Movement Disorder (Sanger T et al)

Movement Disorder 2005

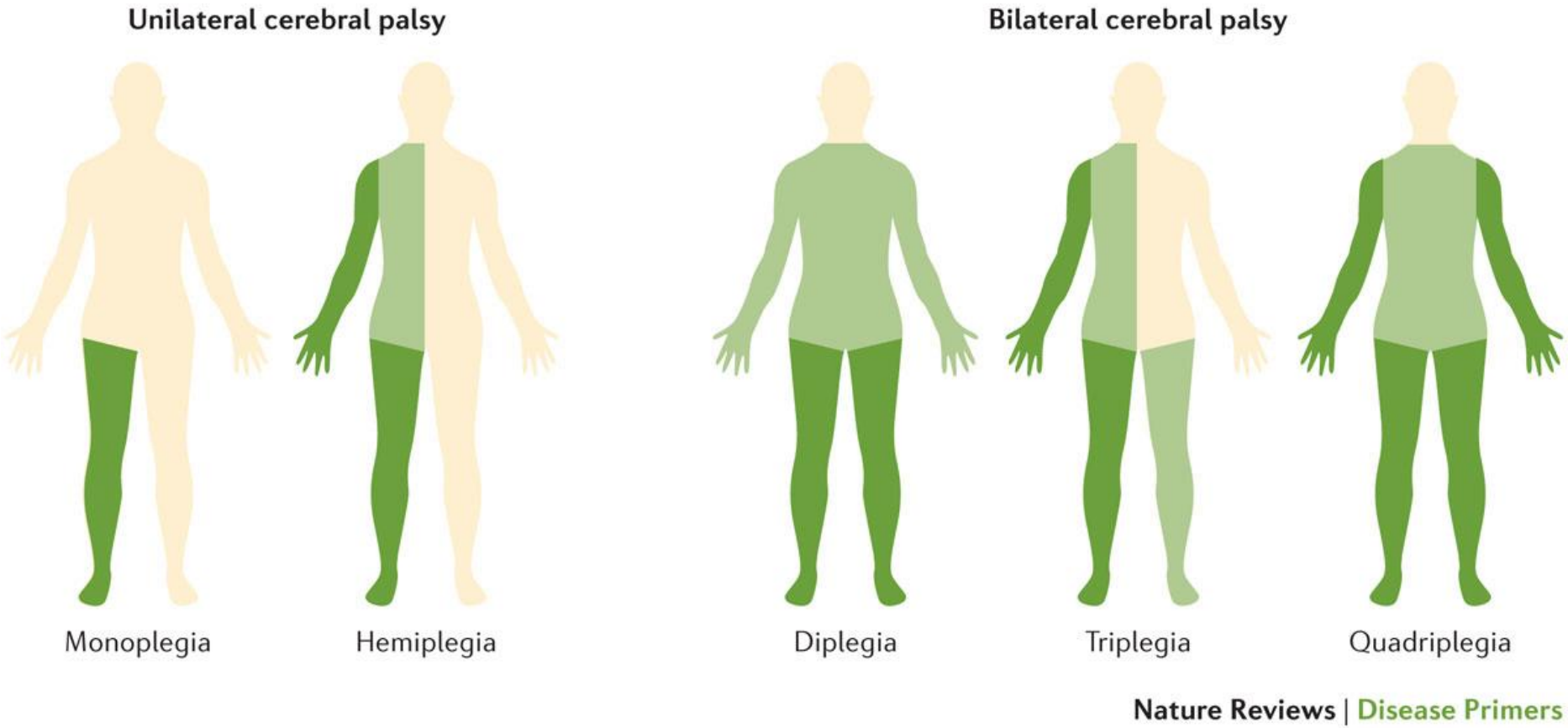
- Spastic 86%
- Mixed Hypertonia 7%
- Dyskinetic 2%
- Ataxia 3%
- Hypotonia 3%

Movement Disorder 2012

- Spastic 53%
- Mixed hypertonia 33%
- Dystonia 11%
- Ataxia 3%

First classified at registration, many sources.
Reclassified by small number of clinicians.

2: Topographical Classification in Cerebral Palsy



Graham, H. K. *et al.* (2015) Cerebral palsy
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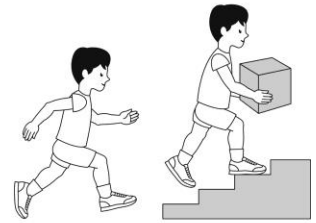
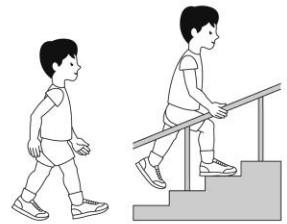
GMFCS for children aged 6-12 years: Descriptors and illustrations



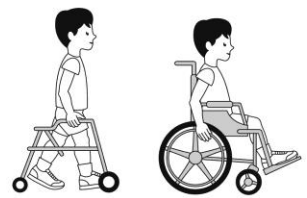
GMFCS Level I

Children walk indoors and outdoors and climb stairs without limitation. Children perform gross motor skills including running and jumping, but speed, balance and coordination are impaired.



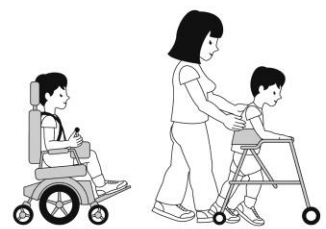
GMFCS Level II

Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces and with long distances.



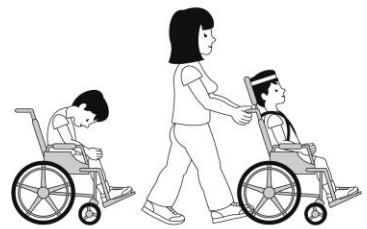
GMFCS Level III

Children walk indoors or outdoors on a level surface with an assistive mobility device and may climb stairs holding onto a railing. Children may use wheelchair mobility when traveling for long distances or outdoors on uneven terrain.



GMFCS Level IV

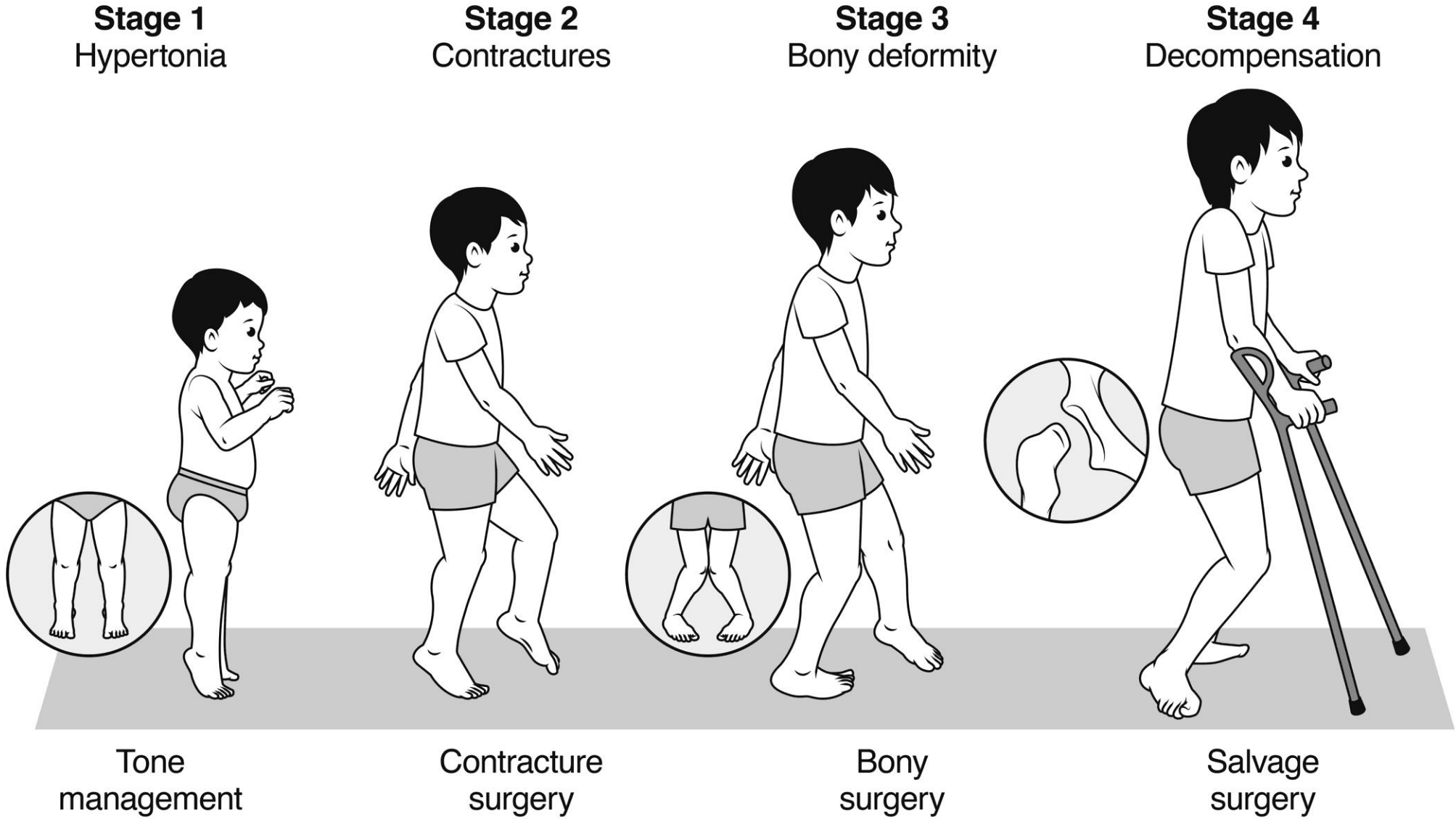
Children use methods of mobility that usually require adult assistance. They may continue to walk for short distances with physical assistance at home but rely more on wheeled mobility (pushed by an adult or operate a powered chair) outdoors, at school and in the community.



GMFCS Level V

Physical impairment restricts voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Children have no means of independent mobility and are transported by an adult.

Musculoskeletal pathology in cerebral palsy



LEVEL	STAGE 1 Hypertonia Birth to 4 to 6 years	STAGE 2 Contractures Age 4 to 12 years	STAGE 3 Bony deformity Age 4 to 12 years	STAGE 4 Decompensation Age 10 to adulthood
HIP	Flexion/adduction, posturing. Clinically: scissoring	Flexion/adduction contractures.	Increased FNA (>25°, hip IR > 2SDs internal 3DGA) Increased MP Acetabular dysplasia	Femoral head deformity, Acetabular deformity Loss of articular cartilage. Arthrosis
KNEE	Spastic knee flexion. Hamstring spasticity. Full knee extension	Hamstring contracture Increased pop. angle Full knee extension or knee FFD < 10°	Knee joint contracture Knee FFD: < 20° Mal-alignment: FNA+ETT	Patella alta Knee FFD > 20° Patellar fracture Arthrosis
ANKLE	Dynamic equinus. Corrects to DF > 0°	Fixed equinus. Ankle DF < 0°	Tibial torsion:ETT > 20° ITT <10°	Calcaneus, long heel-cord. Deformity of talus Arthrosis.
FOOT	Flexible varus or valgus postures.	Partially fixed/flexible varus with muscle imbalance and/or contracture.	Fixed/stiff equinocavovarus. Pes valgus with LAD.	Skin callosities and skin breakdown, fractures, Deformed tarsal bones Arthrosis.

1. **Movement Disorder**

2. **Topography**

3. **Function**

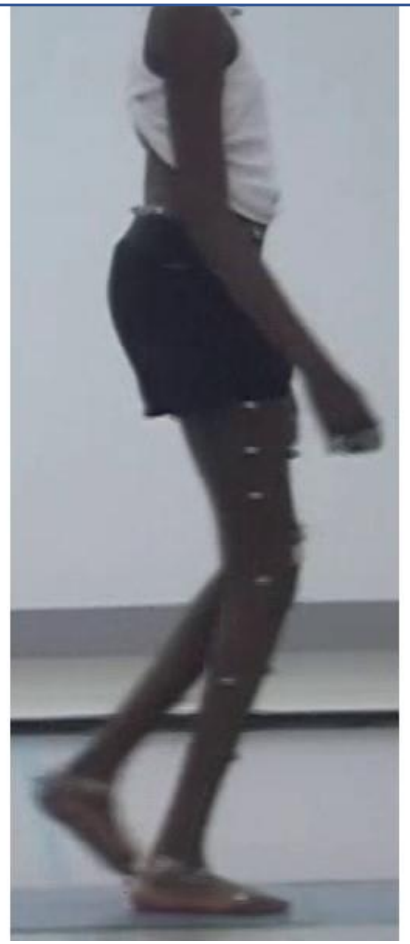
4. **MSP Stage**

CP Phenotype

Walking often gets harder as kids get older: Contractures increase Muscles do not grow as fast as the bone beside them

2016
Age 10

2017
11

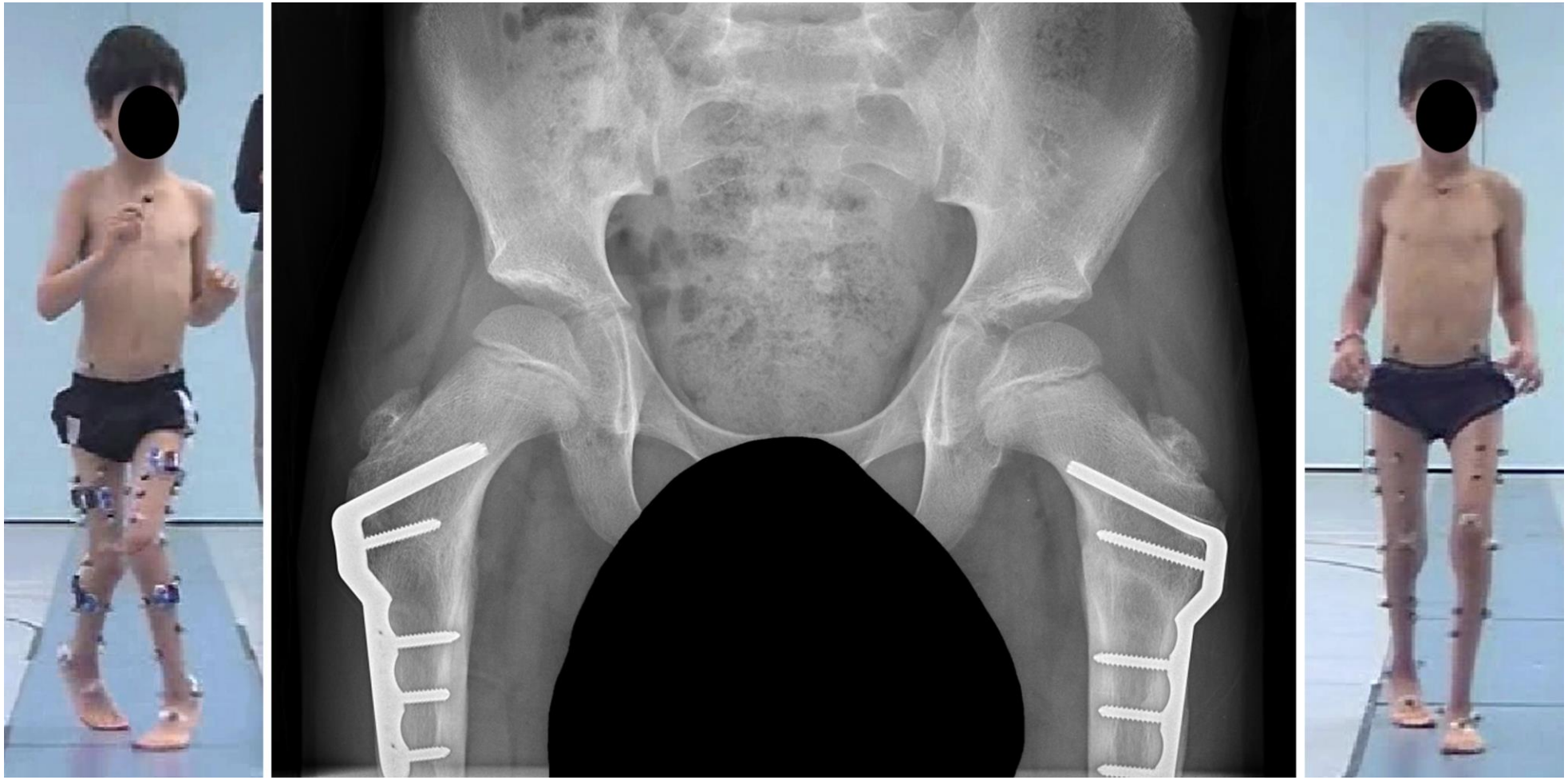


2018.
12

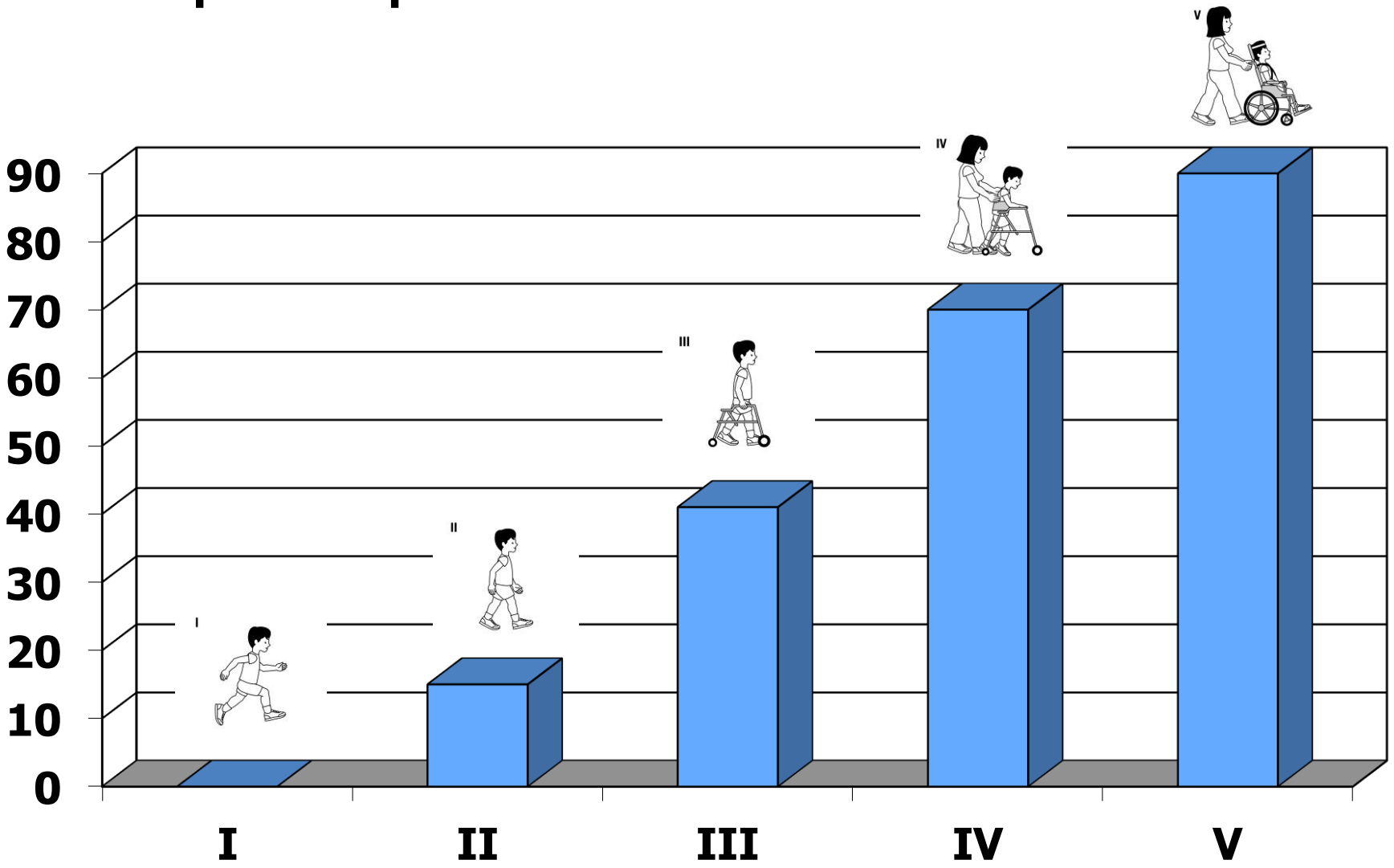
2020
13

Bony torsion: the long bones in children with CP may have twisting, femur (thigh bone) tibia (leg bone)

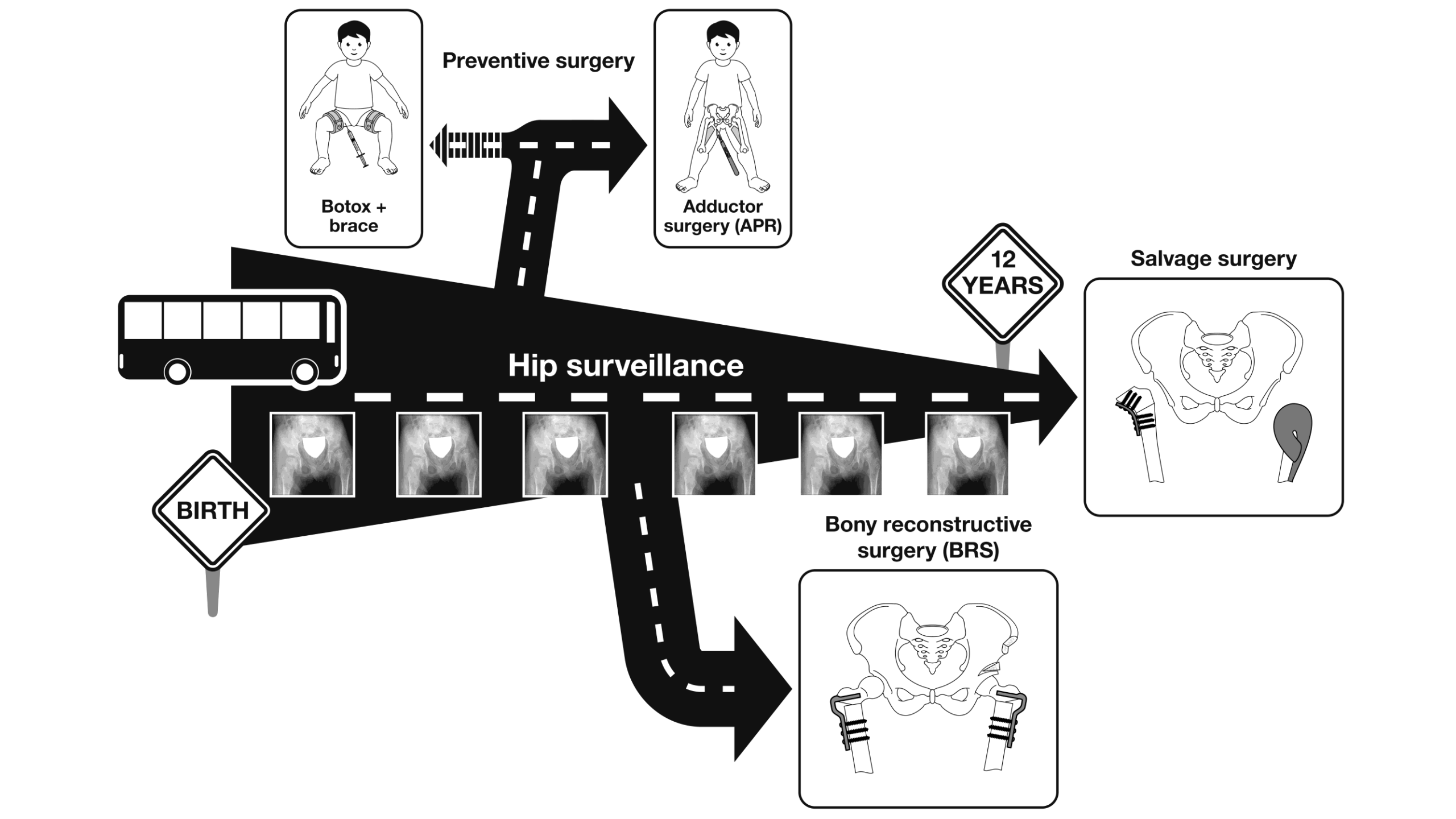
The most common knee problem is knees that face inwards, following the turned in hip.



Hip Displacement and GMFCS

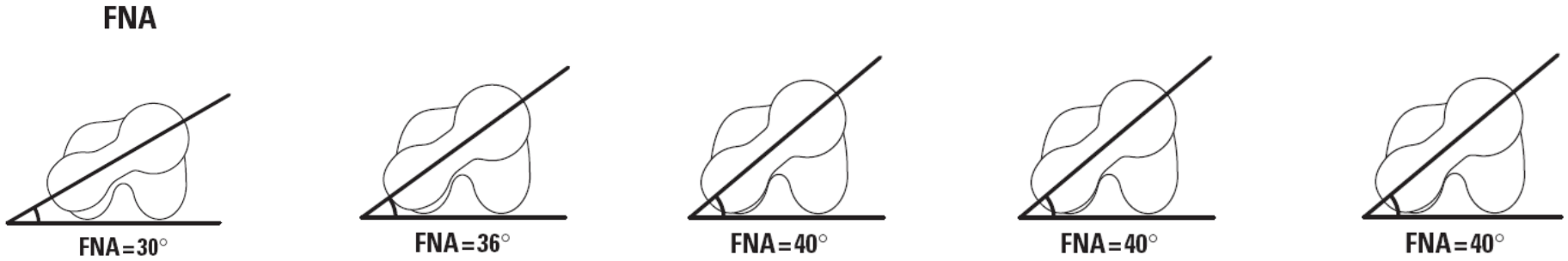


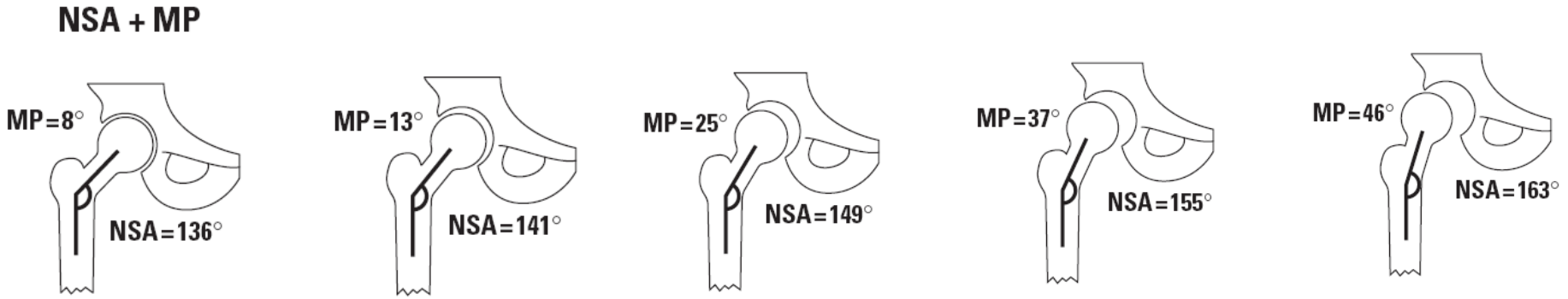
Soo, Howard et al., JBJS 2006


Botox +
brace
Preventive surgery
Adductor
surgery (APR)
12
YEARS
Salvage surgery
Hip surveillance
BIRTH
Bony reconstructive
surgery (BRS)

Robin et al, JBJS, 2008

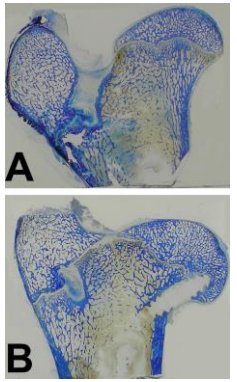
Proximal Femoral Geometry and GMFCS





Managing the hip in children, GMFCS IV and V

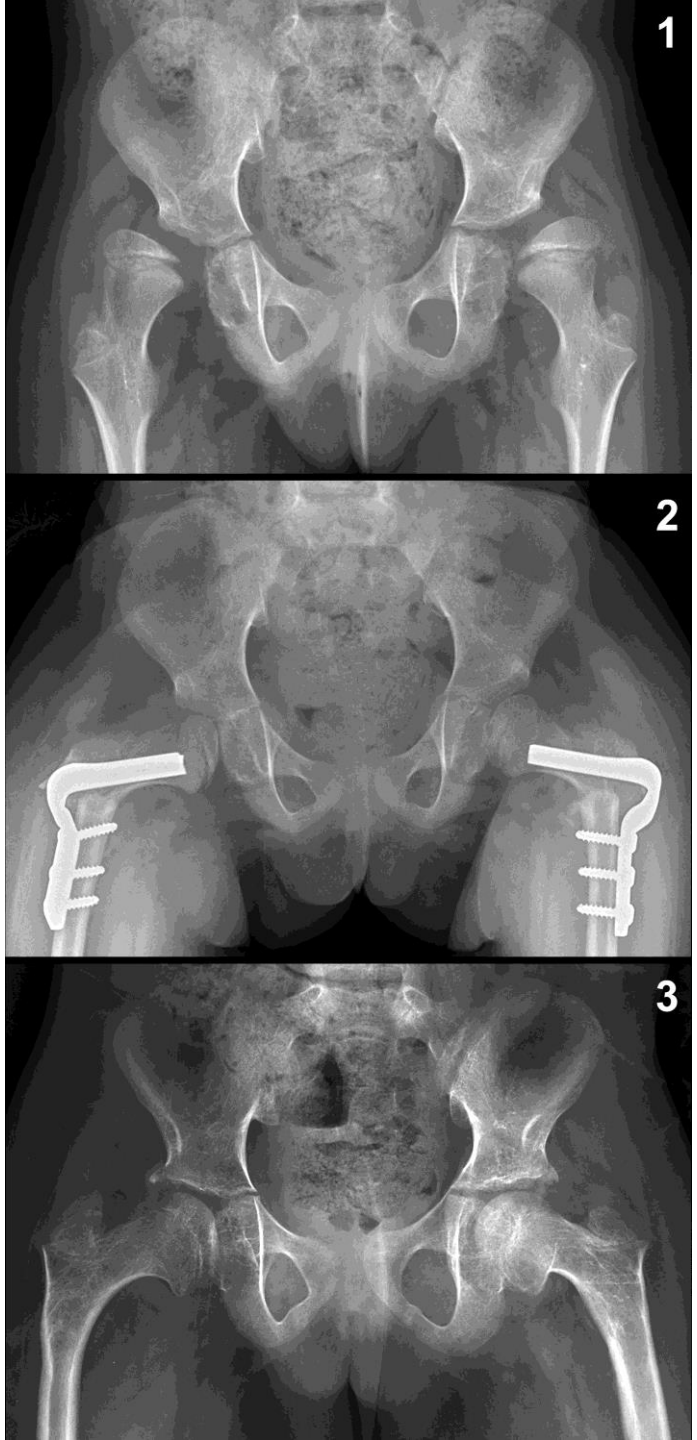
1. Specialized seating, sleep systems: good idea but lacks evidence.
2. Botox to adductors + Brace: does not work, 3 year RCT.
3. Adductor surgery: 80% success rate is now 80% failure rate
4. Guided growth: not as Primary treatment but good for after VDROs
5. VDROs: mainstay of management +/- Pelvic osteotomy



AU, 10 years: GMFCS IV

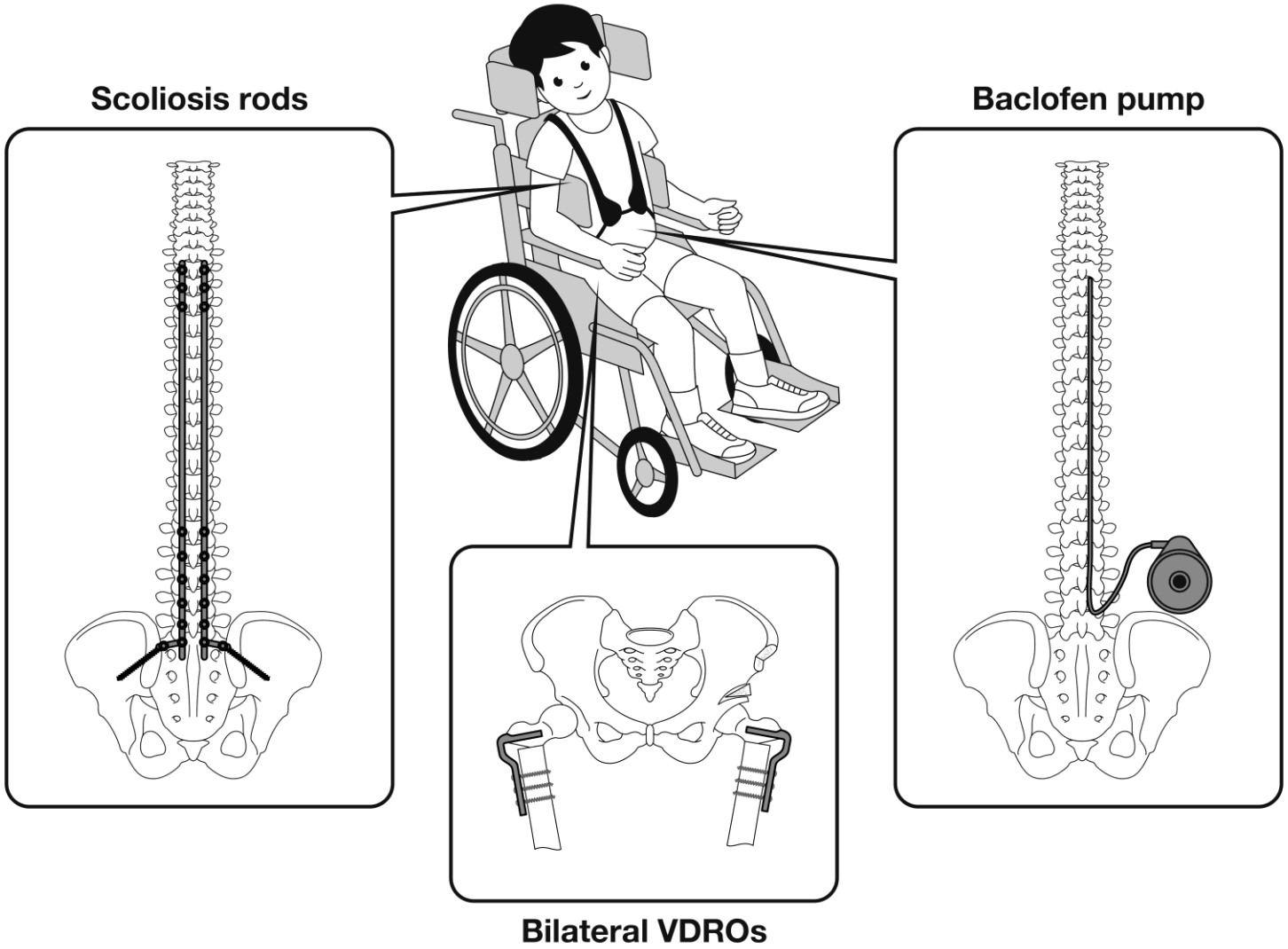
Bilateral hip pain, stiffness: missed hip surveillance

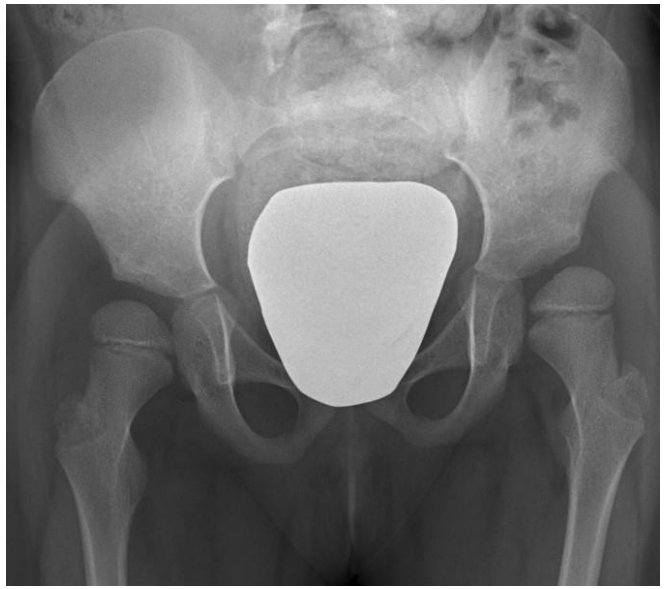
Goals: pilgrimage (sports wheelchair) on Camino with family/support crew to Santiago de Compestela

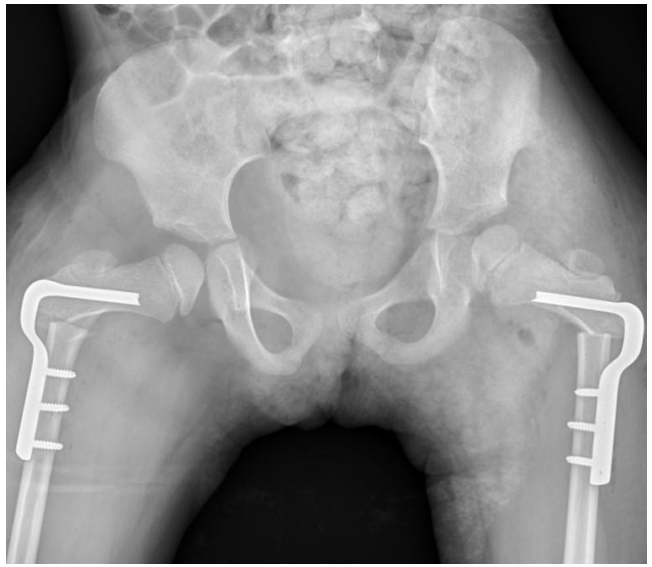


Some children with CP have difficulty sitting: Combinations of hip dislocation and scoliosis

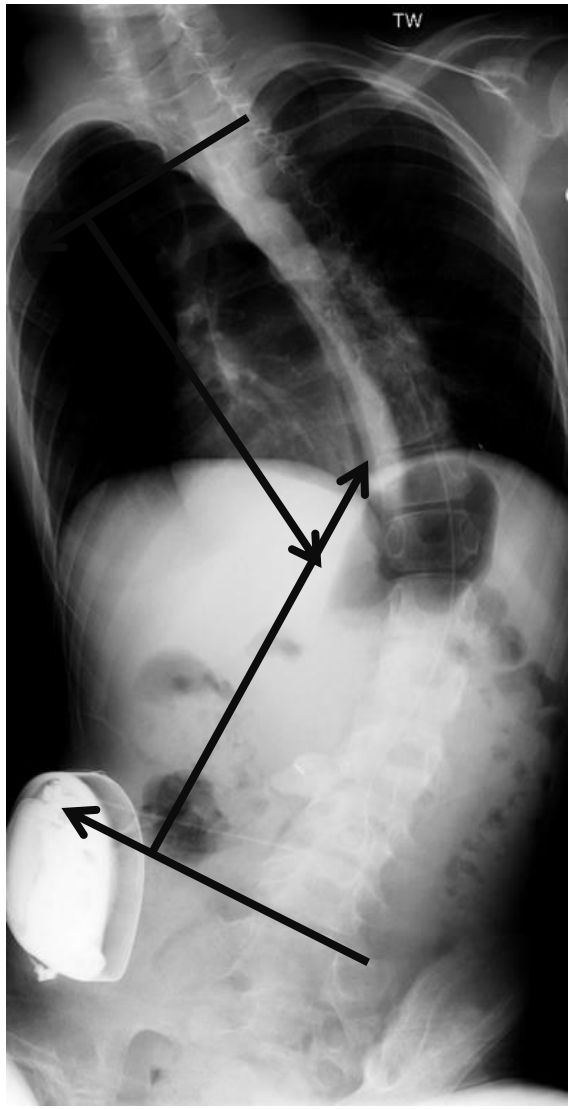
How much technology/hardware is needed at GMFCS V?

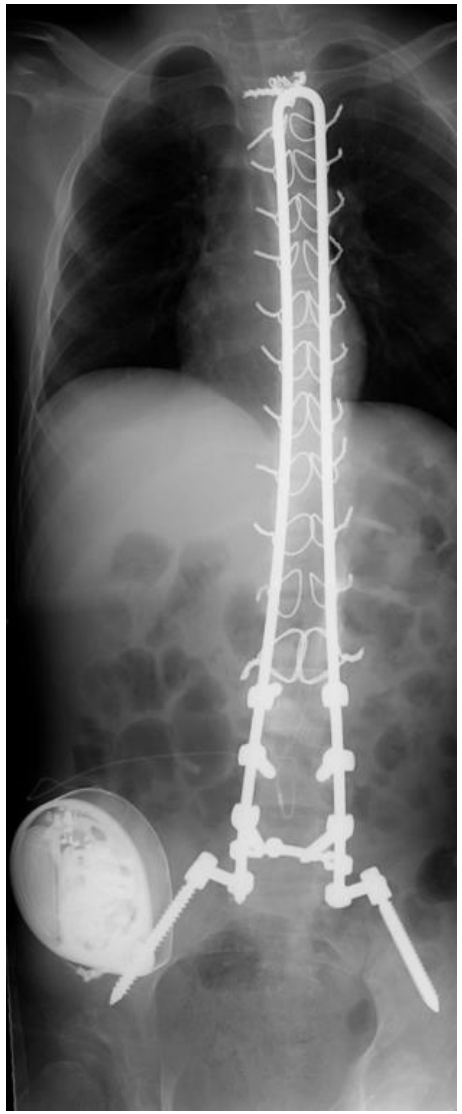






14 years. GMFCS Level V; ITB Pump: Cobb Angle Before Scoliosis Surgery = 68º, flexible MSP Stage 3





How cerebral palsy affects muscles, bones & joints

What should we cover in more detail, next year?

Please let Deb Markelis or me know.

1. Foot and ankle problems in children with hemiplegia
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4. Hip Surveillance and Hip Health
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Thank you for your kind attention

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