# Cerebral Palsy

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# Definition

- Disorder of the development of posture and movement, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain
- It is the most common motor disability of childhood

# Definition

- 3 major criteria
  - A neuromotor control deficit that alters movement or posture
  - A non-progressive brain lesion
  - Brain injury either before birth or in the first year(s) of life

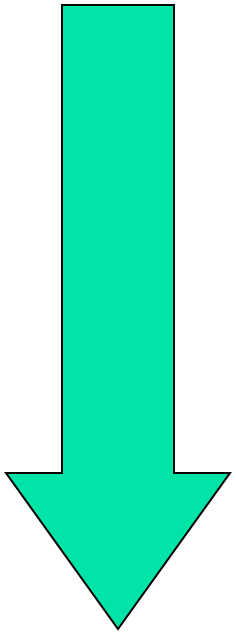
# What it is NOT

- Progressive
- Genetic/Hereditary
- Traumatic

# Cerebral Palsy

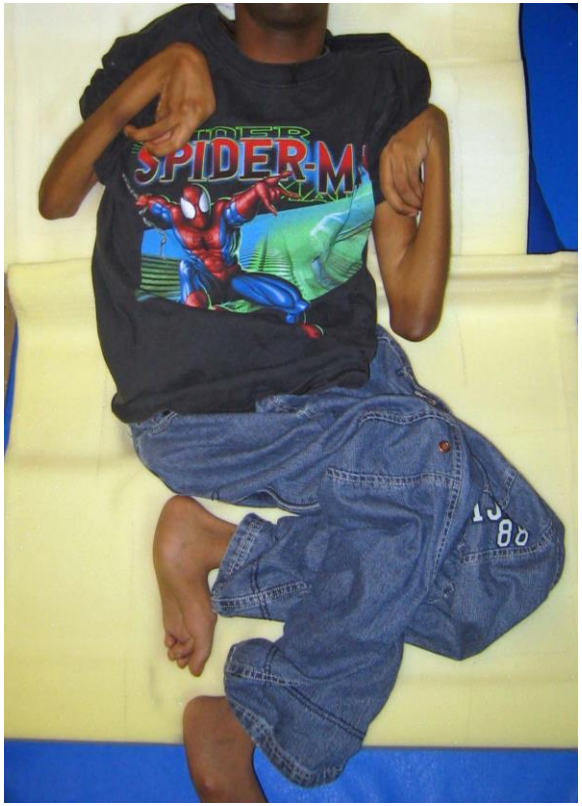
- Although the brain lesion in not progressive, the musculoskeletal pathology is certainly progressive

# Musculoskeletal progression in CP

| | | |
|---|---|---|
| Static (Brain lesion) |  | Spasticity and weakness |
| | | Spastic muscles don't grow as fast as bones |
| | | Fixed contracture |
| | | Bony torsion (twist) |
| Progressive (Musculoskeletal deformity) | | Joint instability |
| | | Dislocation or degenerative changes |

Graham HK, Eur J Neurol, 2001

# Progressive musculoskeletal deformities



# Incidence

- 2-3 per 1000 live births
- 700,000 children and adults with CP in USA
- Relatively constant despite medical advancements in maternal, perinatal and NICU care
- Possibly due to improving survival rates in very premature infants

# Risk factors for CP

- In the majority of cases in full term infants, the etiology is unknown

# Risk factors for CP

- Prematurity – <37 wks
  - Risk increases with increasing prematurity
- 34 weeks – 3 important developments for survival
  - Lungs are developed
  - Suck reflex has developed
  - Germinal matrix of the brain is mature; blood vessels less likely to "leak"
- CSHCS doesn't use gestational age for eligibility

# Risk factors for CP

- Low birth weight (< 2500 gm) or very low birth weight (<1500 gm)
  - Incident in premature and LBW infants is 40-150/1000 live births
  - 1/3 of children with CP had a birth weight <2500 grams
  - Incidence is 30 times higher if birth weight <1500 grams
- Children <999 gm (approx 2 lbs) are eligible for CSHCS

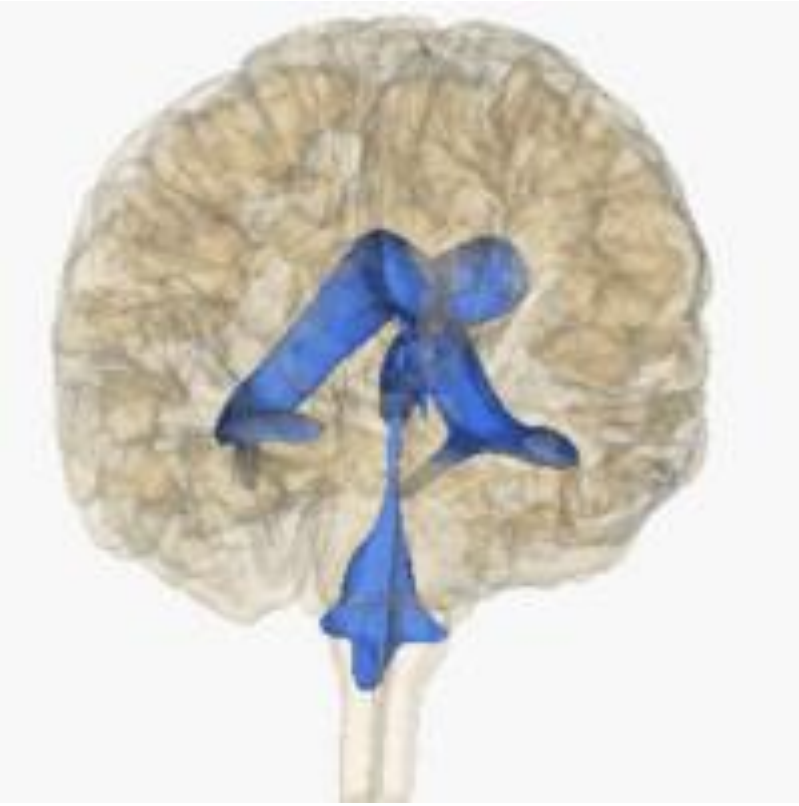
# Risk factors for CP

- Intraventricular hemorrhage
  - 90% of premies, 20% of full term infants with CP
- Intrauterine stroke

# Ventricular system of the brain

- 4 interconnected cavities in the brain where cerebrospinal fluid is produced
- Connected to central canal of spinal cord
- Tracts that control movement of the LE lie closest to the edge of the ventricle
- The bigger the bleed, the greater the brain damage

# Ventricular system of the brain



# Intraventricular hemorrhages

- Grade I – A small amount of blood; stays in the ventricle
- Grade II – A larger amount of blood; intraventricular; normal ventricle size
- Usually have no neurological sequella

# Intraventricular hemorrhages

- Grade III – Even larger amount of blood; intraventricular; ventricular dilation
- Grade IV – Blood spills outside of the ventricle and into the actual brain tissue
- Usually have neurological sequella, and this qualifies a child for CSHCS

# Periventricular leukomalacia

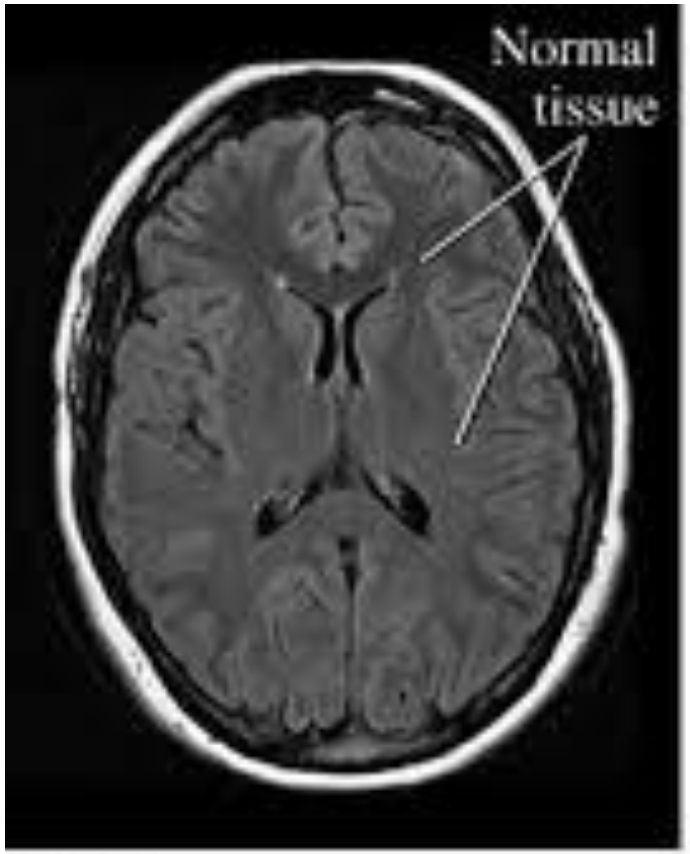
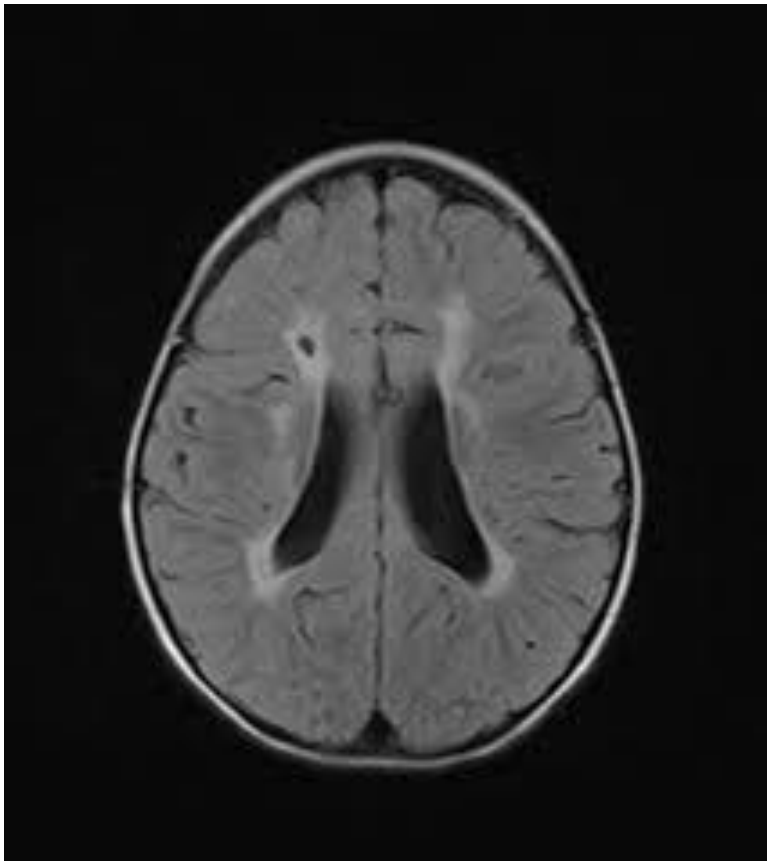



Figure 1



# Risk factors for CP

- Intrauterine infections/chorioamnionitis
  - TORCH
- Hyperbilirubinemia
  - Kernicterus – associated with dystonic CP and neurosensory hearing loss
- Multiple gestation
- "Vanishing twin" phenomenon
- Twin-to-twin transfusion

# Risk factors for CP

- Hypoxia?
  - <10% of children with CP have history of anoxia/hypoxia
  - Documented anoxia/hypoxia is a risk factor
    - Acidosis, bradycardia
    - Neonatal encephalopathy
    - Hypoxic ischemic encephalopathy (HIE) on MRI
      - Eligible for CSHCS
    - Respiratory difficulties, abnormal tone, seizures

# Anoxic brain damage

- Some parts of the brain are more susceptible to anoxia
  - Basal ganglia (dystonic cerebral palsy)
  - Auditory nuclei (neurosensory hearing loss)

# CP can be associated with:

- Cognitive impairment (50%)
- Seizures (50%)
- Learning disabilities
- Visual problems
  - Strabismus (75%), ROP, cortical blindness (HIE), hemianopsia (HP)
- Incontinence

# CP can be associated with:

- Speech delays / hearing problems
  - Sensorineural deafness in hypoxia, TORCH, kernicteris, bacterial meningitis
- GERD / Constipation / Failure to thrive
  - Aspiration pneumonia
- Orthopedic complications
  - Dislocated hips, scoliosis, joint contracture
  - Decreased bone density

# Functional Problems

- Gross motor/Mobility
- Learning
- Fine motor
- Feeding

# Classification of CP

- Type of movement disorder
- Anatomic distribution

# Type of movement disorder

- Spastic – 70-85%
- Dyskinetic (dystonic, athetoid) – 5-10%
- Ataxic – 5%
- Mixed – 10%
- Hypotonic – 3%
- Often overlapping/not clear cut

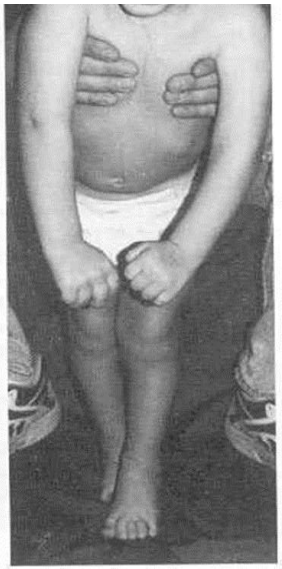
# Hypotonic/Ataxic

- Rare, therefore all children should receive a thorough diagnostic work-up for other neurological conditions

# Spasticity

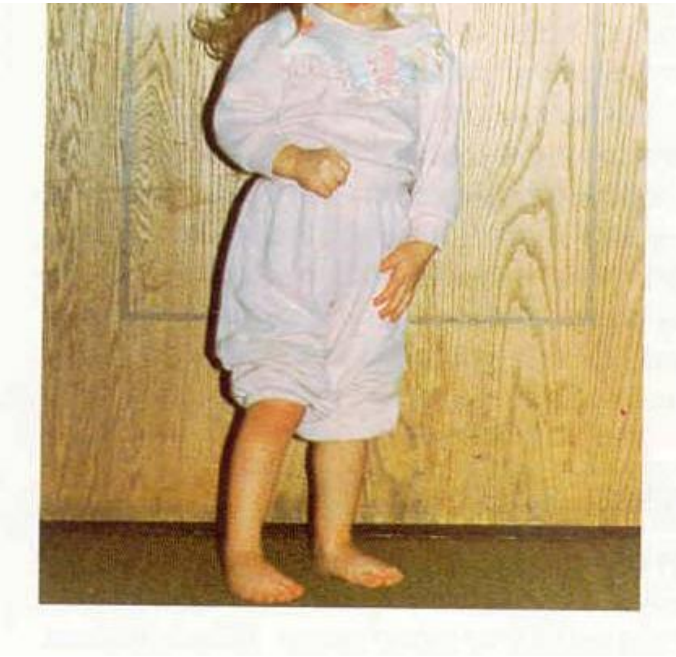
- Increased tone/resistance to movement
- Assessed by
  - Deep tendon reflexes
  - Passive mobilization
    - Movement through the full ROM should take less than one second

# Anatomic distribution of CP



Quadriplegia (32%)

Diplegia (24%)



Hemiplegia (30%)

# Spastic Quadriplegia

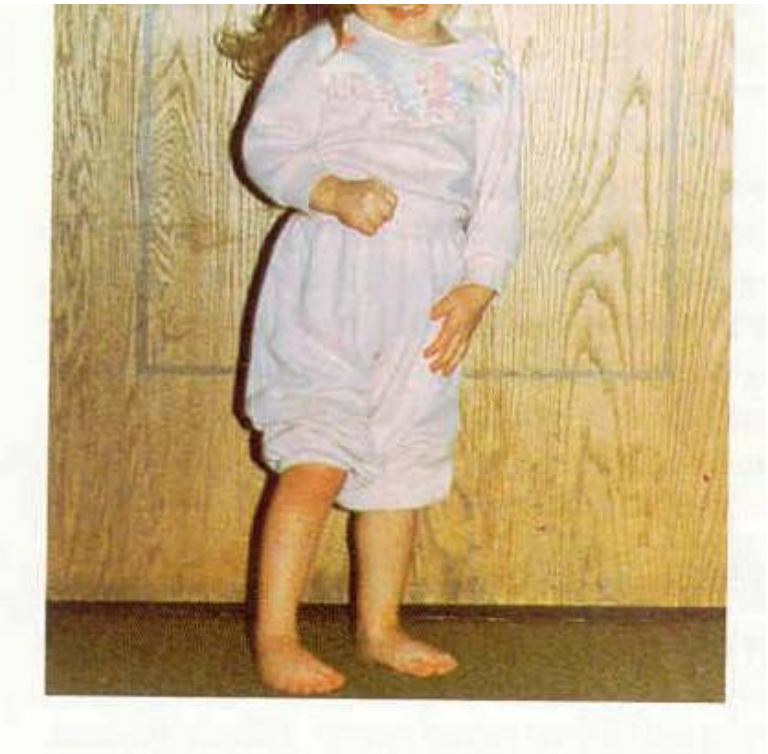
- All 4 extremities spastic; hypotonic trunk
- Grade IV IVH
- Majority with cognitive impairment
- Seizures in >50%
- High risk of aspiration pneumonia
- Highest risk for orthopedic complications (scoliosis, hip dislocation)
- 50% achieve minimal ambulatory skills with an assistive device

# Spastic Diplegia

- Lower extremities more spastic than upper extremities
- Grade III IVH
- 25-33% have seizures or cognitive impairment
- 80-90% are ambulatory with or without AD

# Spastic Hemiplegia

- One side affected, upper extremity more than lower extremity
- Right HP:Left HP = 2:1
- Full-term, intrauterine strokes involving MCA
- 50-70% have seizures
- 25% are cognitively impaired
- Leg length discrepancy
- Almost 100% ambulate, but late (not until 18-24 mo)
- Sensory deficits common



# Common Presentations of CP

- Delayed development of motor milestones
- Early handedness
- Persistence of primitive reflexes
- Presence of pathologic reflexes
- Failure to develop protective extension responses
  - Russman BS, etal, Spasticity 2002

# Diagnosis

- History, physical exam
- No laboratory tests are diagnostic
- EEG may be indicated
- Head imaging may be helpful
  - Ultrasound, MRI
  - >80% of kids with CP have abnormal findings
- BAERs if indicated

# Examination

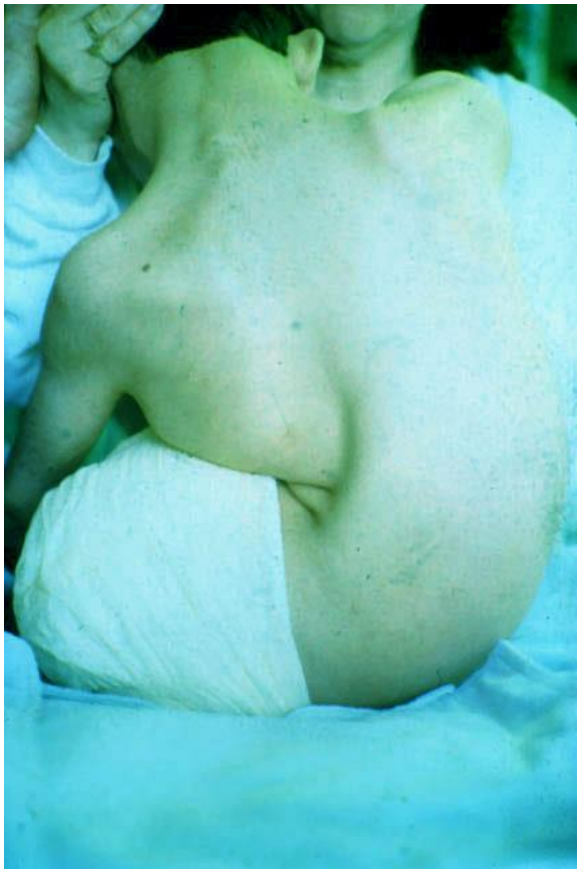
- Early hypotonia (especially trunkal)
- Spasticity in extremities, develops over time
- Brisk DTRs
- Clonus
- Upgoing plantar reflexes (Babinski)
- Synergistic movement patterns
- Joint contractures

# Orthopedic problems

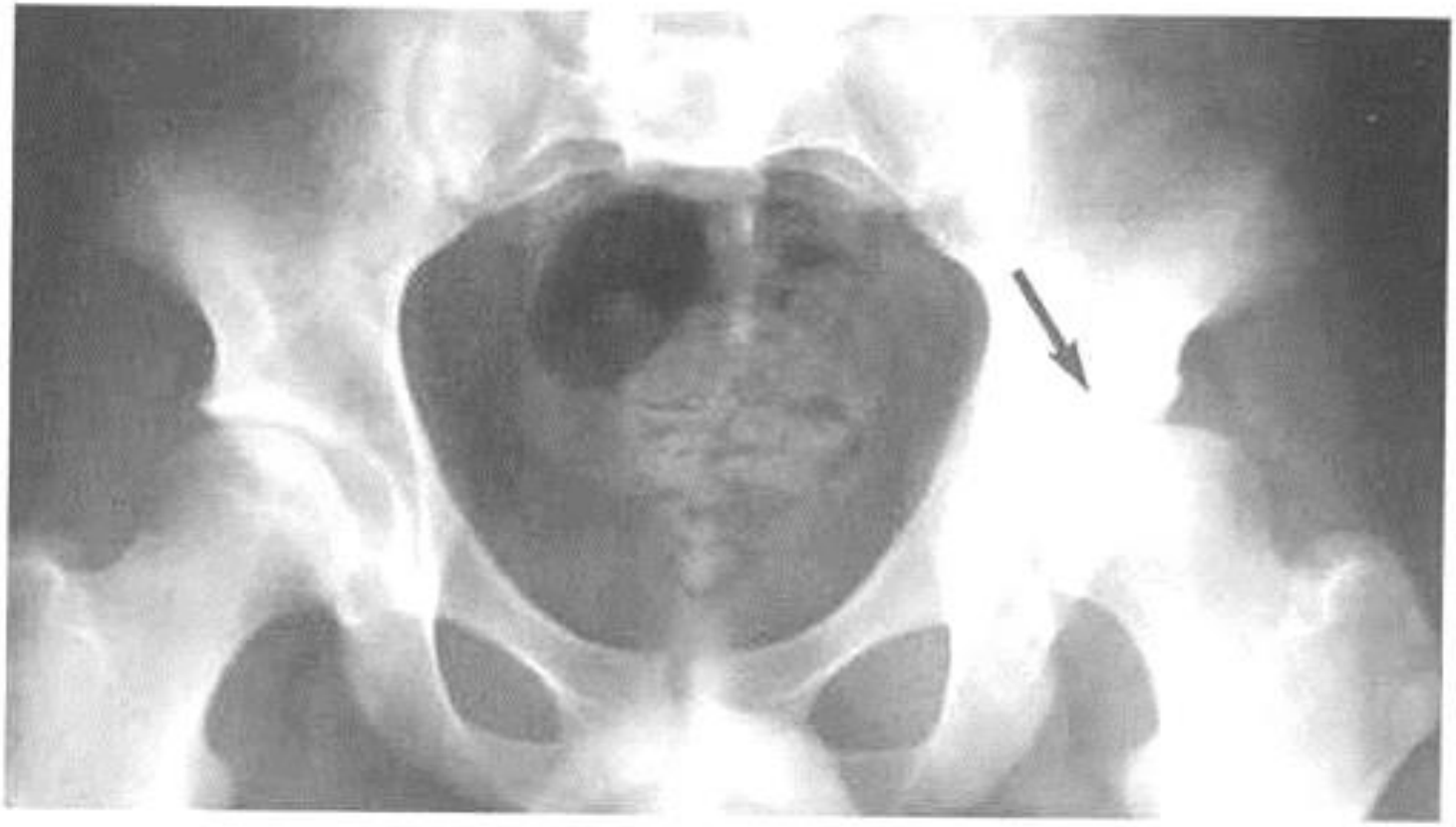
- Scoliosis
- Hip subluxation/dislocation
- Knee flexion contractures
- Patella alta
- Foot deformities

# Scoliosis

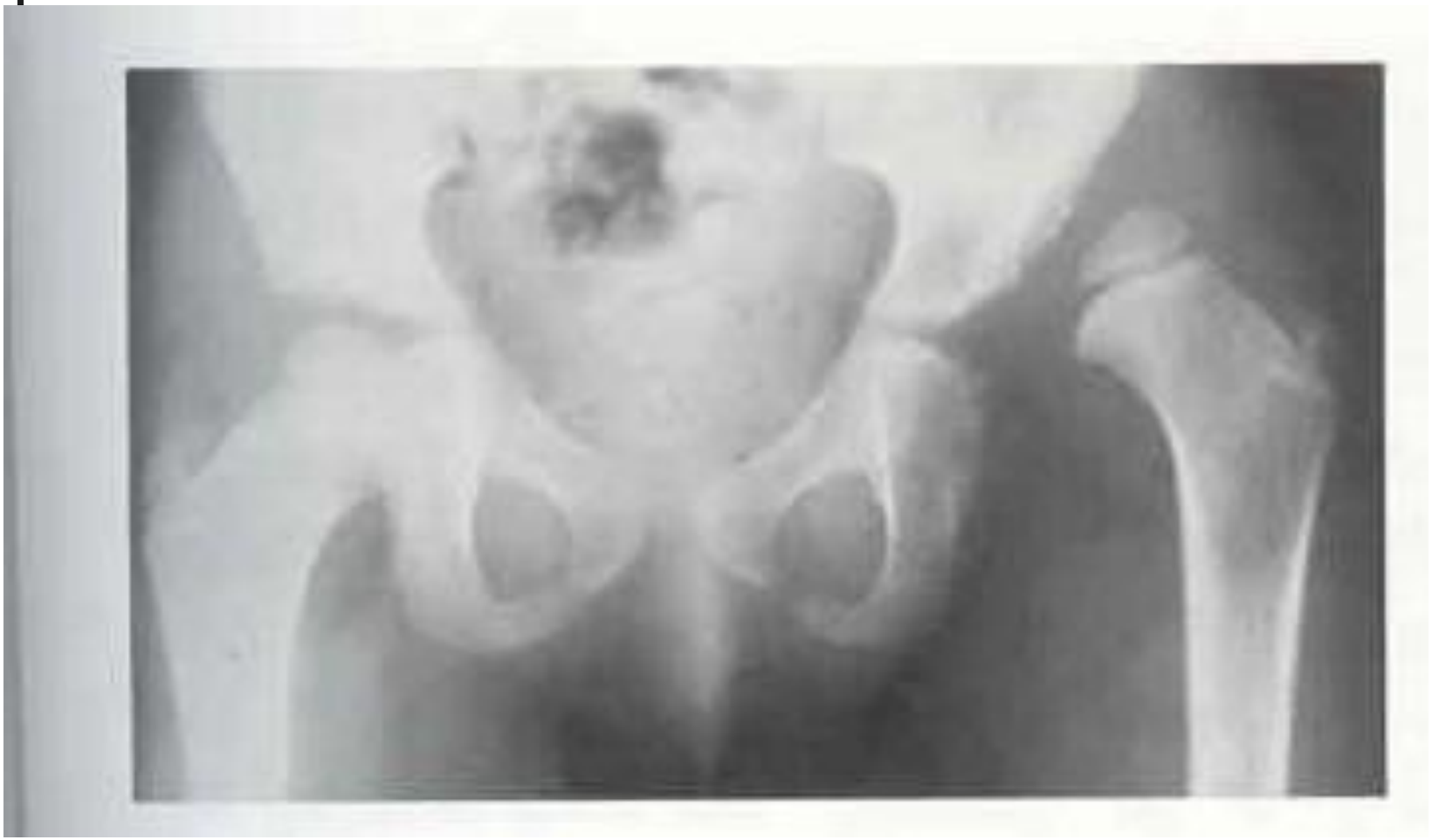
- Neuromuscular
- Risk increases with severity
  - 70% in quads
- Curves over 40 degrees tend to progress
- Complicated by skin breakdown, joint contractures, pelvic tilt



# Hip subluxation



# Hip dislocation



# Risks for hip dislocation

- Scissoring pattern – adduction, often with hip flexion
  - Hip abduction < 35 degrees
  - Hip flexion contractures >20 degrees are risk factors
- Persistent coxa valga
- Excessive femoral anteversion
- Shallow acetabulum

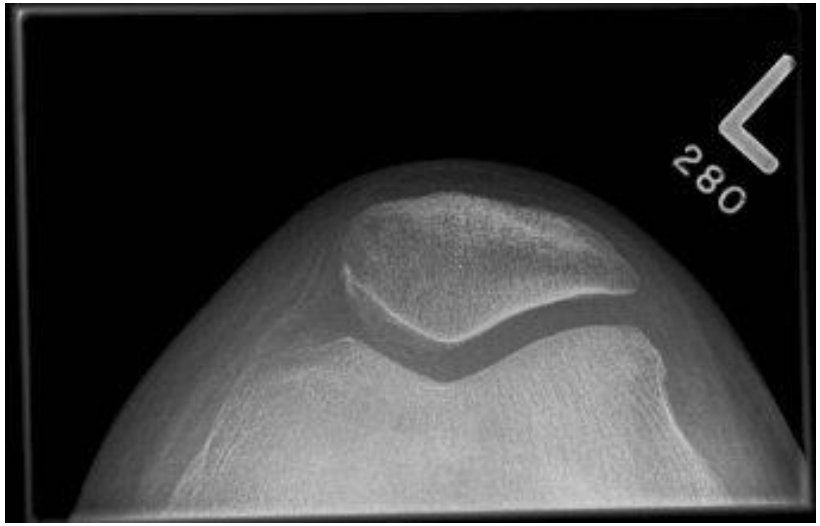
# Risks for hip dislocation

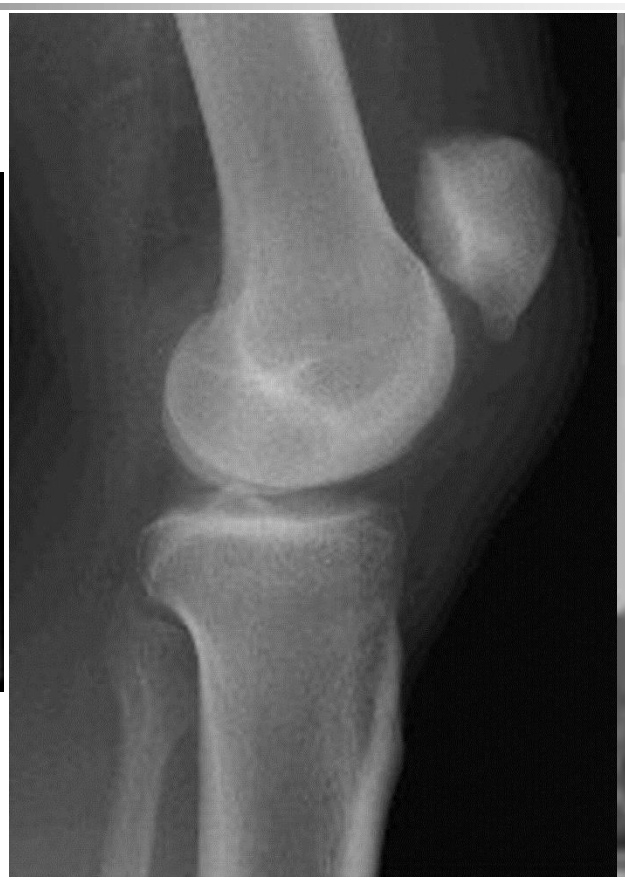
- Diagnosis
  - Quadriplegia – 80%
  - Dystonia – 40%
  - Diplegia – 20%
  - Hemiplegia – 1%
- Ambulatory Status
  - Non-ambulatory – 70-90%
  - Ambulatory – 0-40%
    - Graham, AACPDM presentation, 2006

# Knee problems

- Knee flexion contractures
- Patella alta
  - Abnormally high patella
  - In CP, caused by prolonged positioning in flexion or by overactive quadriceps in crouch gait
  - In adolescence, can be painful

# Patella alta

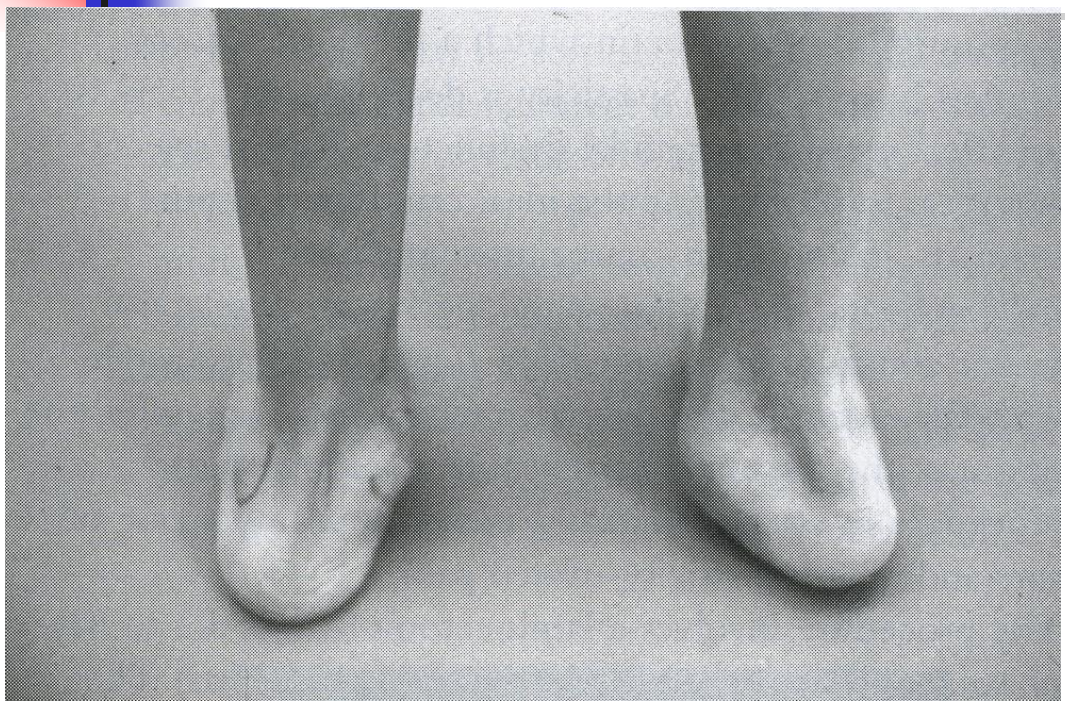


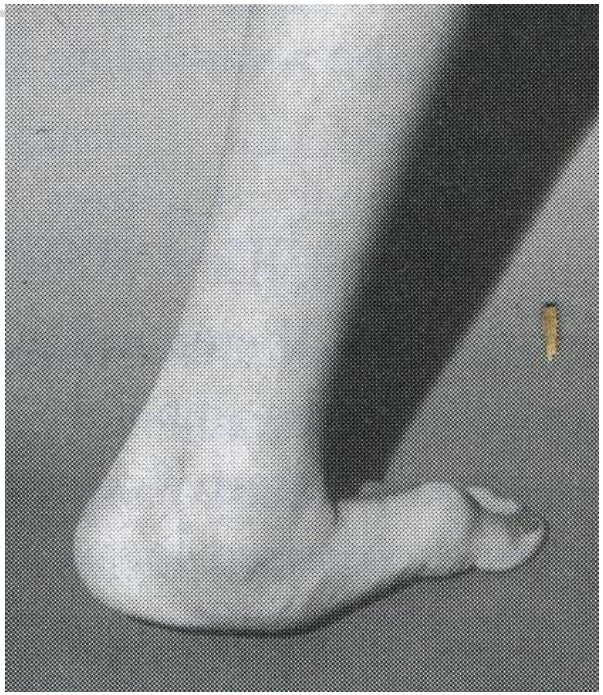



# Foot Deformities in Child with Cerebral Palsy

- Pronation
- Supination

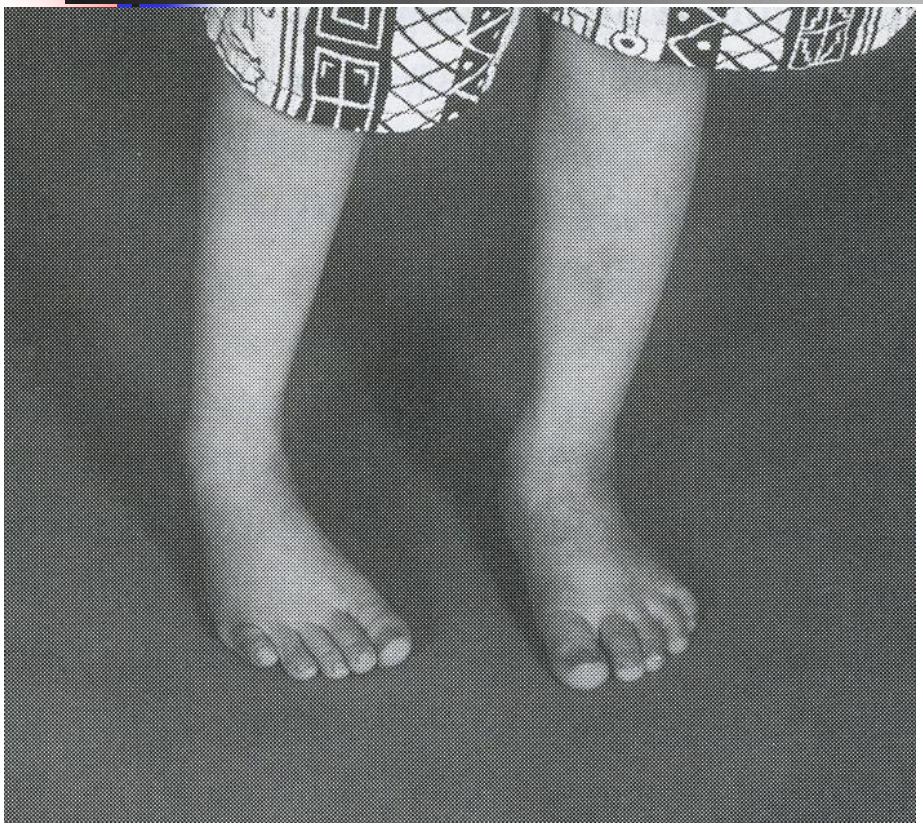
# Pronation in CP

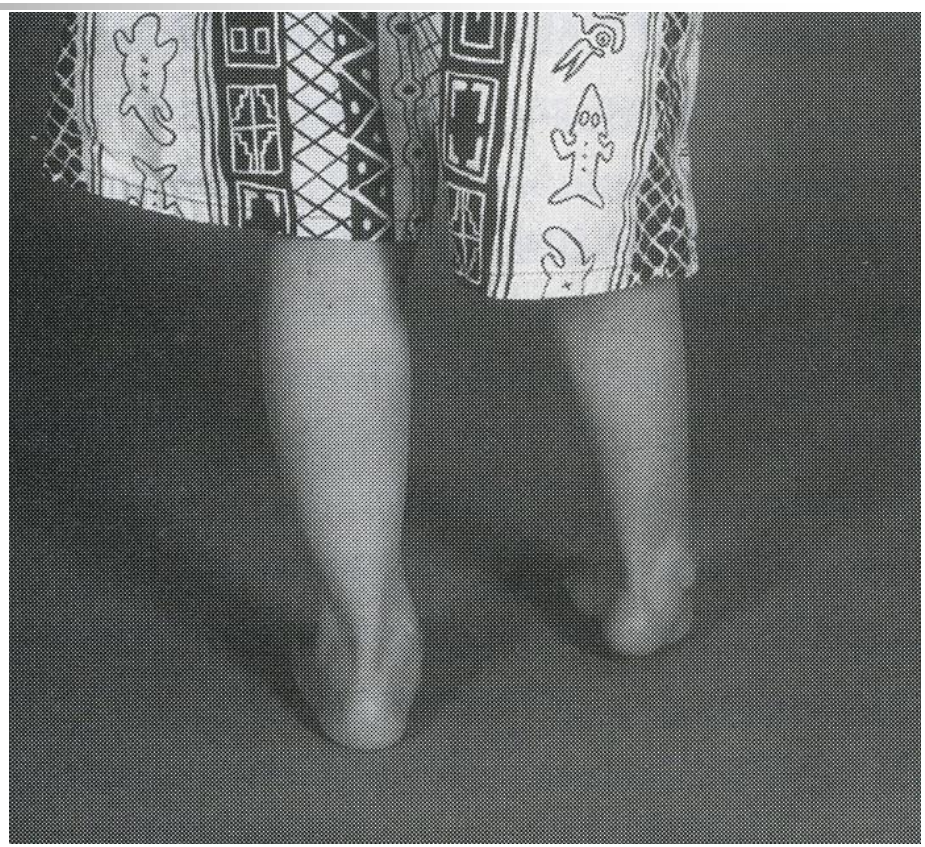




- From Dormans

# Supination Deformity in CP





- From Dormans

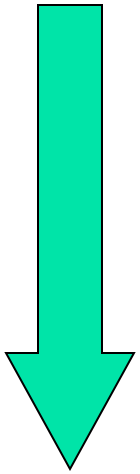
# Why don't all kids with CP qualify for CSHCS?

- ☑ Chronic
- ☑ Sub-specialist-PM&R, Neuro, Ortho
- Severity-(here's the stopper!)
  - Therapy is not considered an indicator of severity
  - So we are looking for the need for interventions like equipment, spasticity medication, Botox injections, alcohol blocks, serial casting, surgery, etc

# Treatment options

- Therapy/Orthotics/ Equipment
- Oral medications
- Chemodenervation
  - Botulinum toxin/ Phenol blocks
- Neurosurgery
  - ITB
  - SDR
- Orthopedic surgery

- Least invasive



- Most invasive

# Therapy (very simplified!!)

- Occupational therapy
  - Fine motor skills, Activities of Daily Living
  - Equipment
- Physical therapy
  - Gross motor skills
  - Equipment
- Speech therapy
  - Language, communication
  - Augmentative communication devices

# Serial casting for joint contractures

- A series of casts is applied weekly to gradually stretch the muscle
- Might use Botox before
- Bracing or splinting afterward
- Preferable to a muscle lengthening surgery

# Bracing (Orthotics)

# Knee braces

# SWASH Orthosis

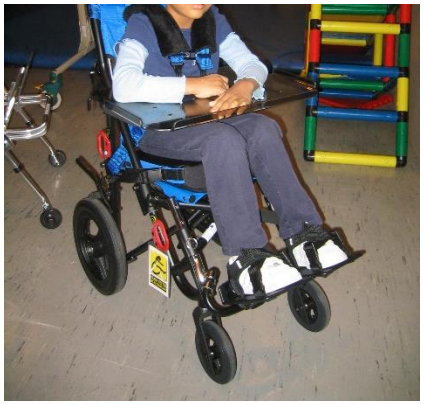
- Standing, Walking and Sitting Hip Orthosis
- Controls dynamic hip scissoring

# Other Equipment

- Bathseats/shower seats
- Strollers
- Wheelchairs
- Standers
- Walking assistive device
- Safe hospital beds
- Augmentative Communication devices

# Bath seats

# Strollers/Wheelchairs



# Standers

- Standing helps develop the hip joint
- Prevents contractures
- Improves bone density
- Standing 1 hour/day reduces hip dislocation by 60%
- Serve a different purpose than walkers

# Walkers

# Safe Hospital Beds

- It's easy to see how a child could get entrapped in a standard hospital bed
- The FDA identified 7 zones of entrapment
- Safer bed technology was developed

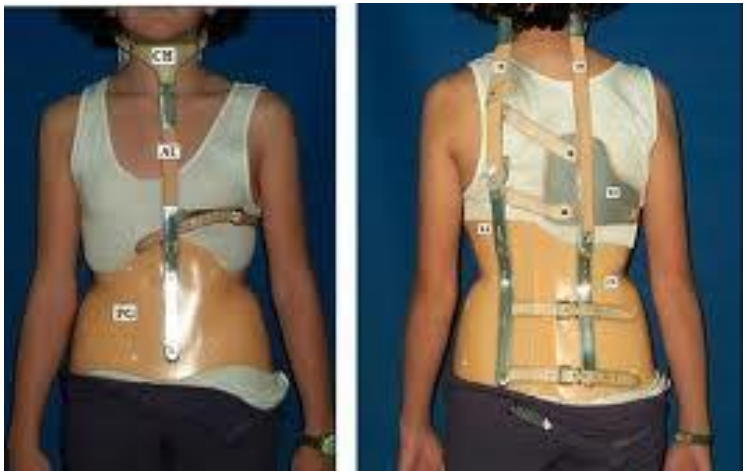
# Safe Hospital Beds

- Beds by George
- Sleep Safe
- Pedicraft

# Treatment of Scoliosis

- Bracing
- Surgery

# Bracing/positioning for scoliosis



# Treating spasticity

- If spasticity interferes with
  - Functioning
  - Positioning
  - Comfort
  - Care
- If spasticity is not useful (ie: transfers)
- If treatment is expected to provide improvement

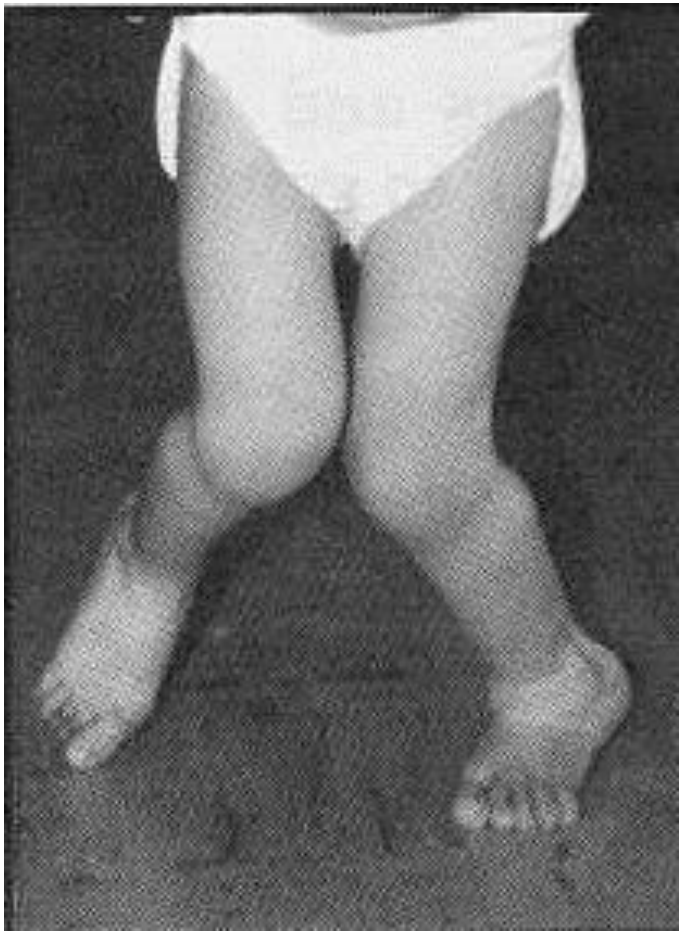
# Treating spasticity

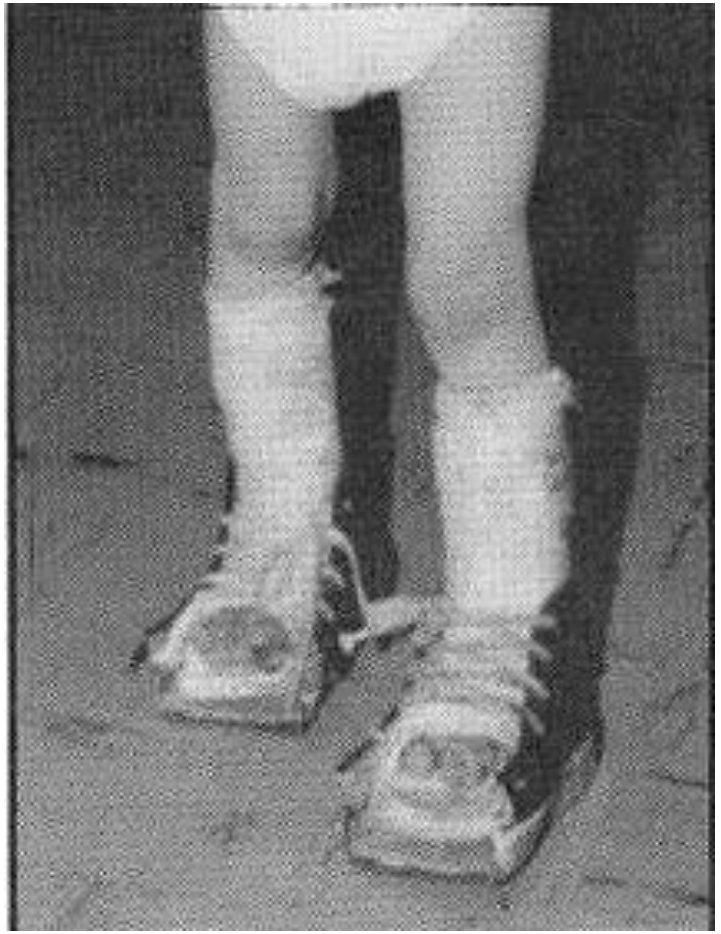
- Positioning
- Oral medication
- Chemodenervation
- Surgery

# Orthotics/Positioning

- Bracing / Splinting
  - Positioning – biomechanical alignment is key!
  - Consider skin tolerance and wearing time

# Effects of Biomechanical Alignment on Spasticity





From Cusick

# Oral medications

- Treat systemic spasticity, but have systemic side effects
- Most common side effect is drowsiness

# Oral medications

- Benzodiazepines (Valium, Klonopin)
- Baclofen (Lioresal)
- Dantrolene sodium (Dantrium)
- Tizanidine (Zanaflex)
- Clonidine (Catapres)

# Chemodenervation

- Injectable therapy which results in local muscle weakening
- Temporary and titratable

- Botulinum toxin
- Phenol or ethyl alcohol

# Botulinum toxin

- Temporarily weakens a muscle (3 months)
- Creates a "window of opportunity"
  - Adjunct to serial casting, intense therapy
- Don't need to use anesthesia (vs alcohol blocks)
  - Topical anesthetic

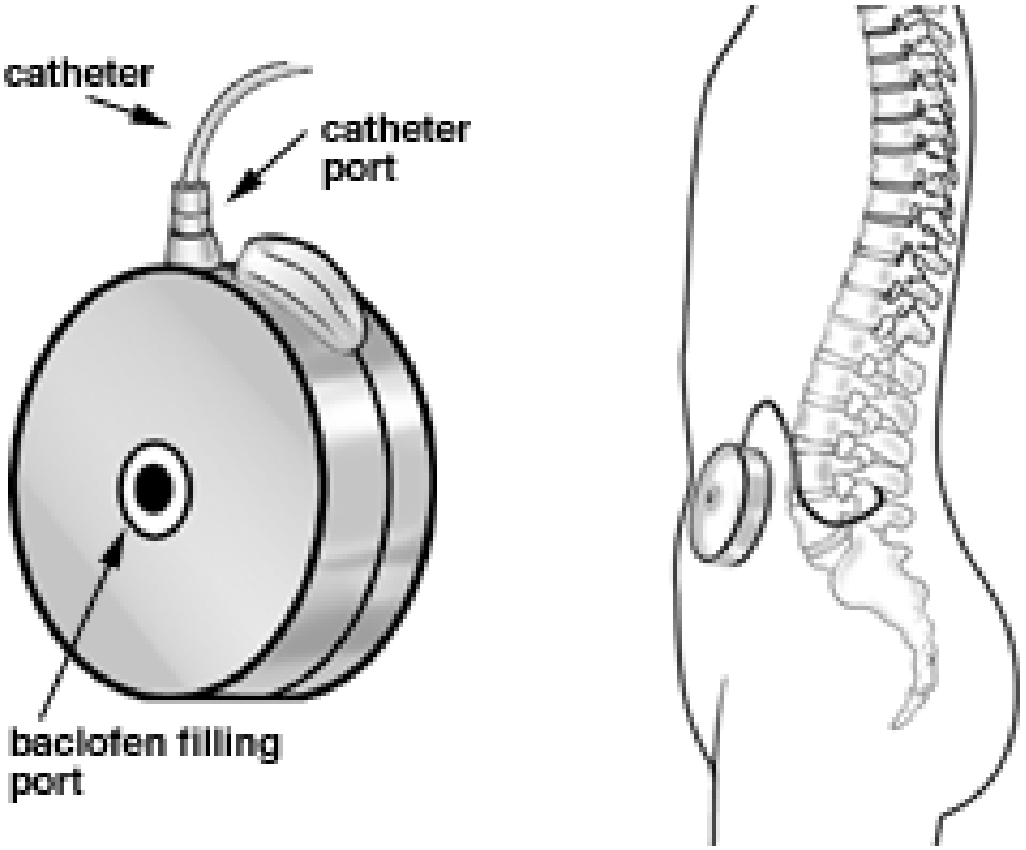
# Phenol/Ethyl Alcohol Injections

- Motor nerve block or motor point block
  - Can only be used on motor nerves (not sensory)
  - Obturator and musculocutaneous nerves
- Causes axonal protein denaturation
- Results usually last 6-12 months
- Done under anesthesia

# Surgical Treatments

- Intrathecal Baclofen Pump
- Selective Dorsal Rhizotomy
- Orthopedic surgeries

# Intrathecal baclofen pump




intrathecal baclofen pump system

# Intrathecal Baclofen Pump

- Implantable, programmable pump, controlled by telemetry
- Baclofen – dosing significantly less that oral dose, so "no" side effects
- Medication stable in pump up to 6 months
- Lots of flexibility in dosing
- Reduces risk of hip dislocation

# Selective Dorsal Rhizotomy

- EMG guided sectioning of afferent nerve rootlets from L2-S2
- Interruption of reflex arc
- Often “unmasks” underlying weakness
- Not flexible dosing
- Not done (locally) as much as ITB pump

# Orthopedic Surgery

- Lengthening Procedures (Muscular)
  - Tendo-achilles lengthening
  - Hip adductor lengthening
  - Hamstring lengthening
  - Selective Percutaneous Myofascial Lenghtening (Percs); New Jersey

# Orthopedic Surgery

- Rotational surgeries (Bony)
  - Varus Derotation Osteotomy (VDRO)
  - Tibia / fibula osteotomy

# Contact me with questions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