Deformity and Pain in Foot of Neuromuscular Disease

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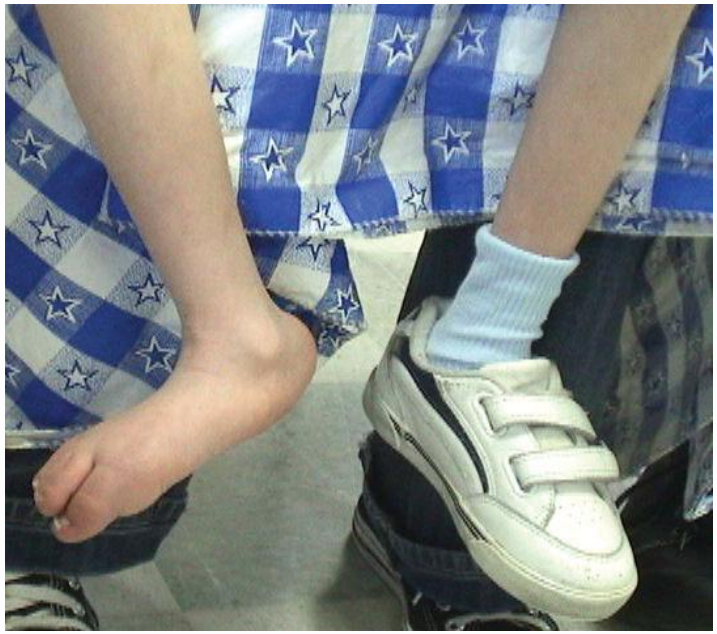
Neuromuscular Diseases & Foot

- Deformity : almost
- Pain : infrequent
- Non-ambulatory, Ambulatory



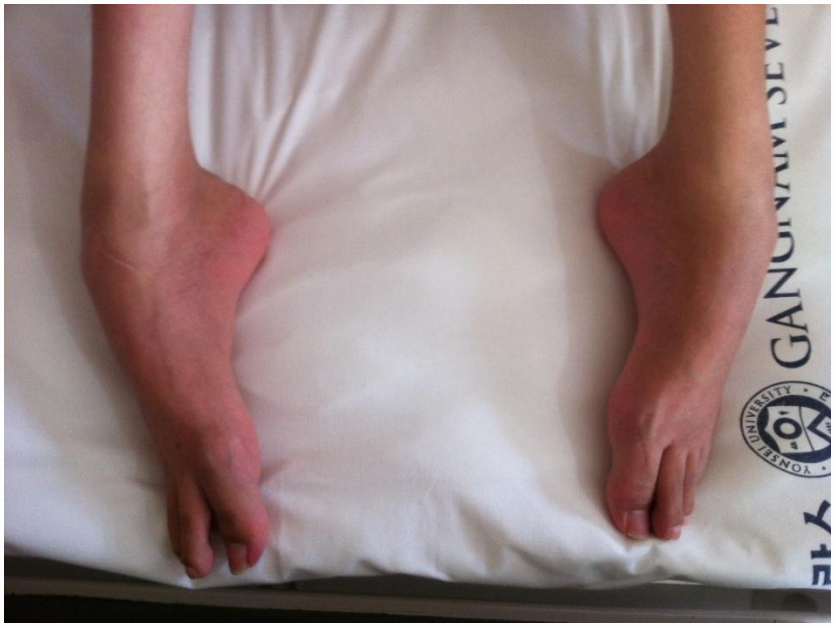
Neuromuscular Diseases in Children

-Genetic problems frequently associated

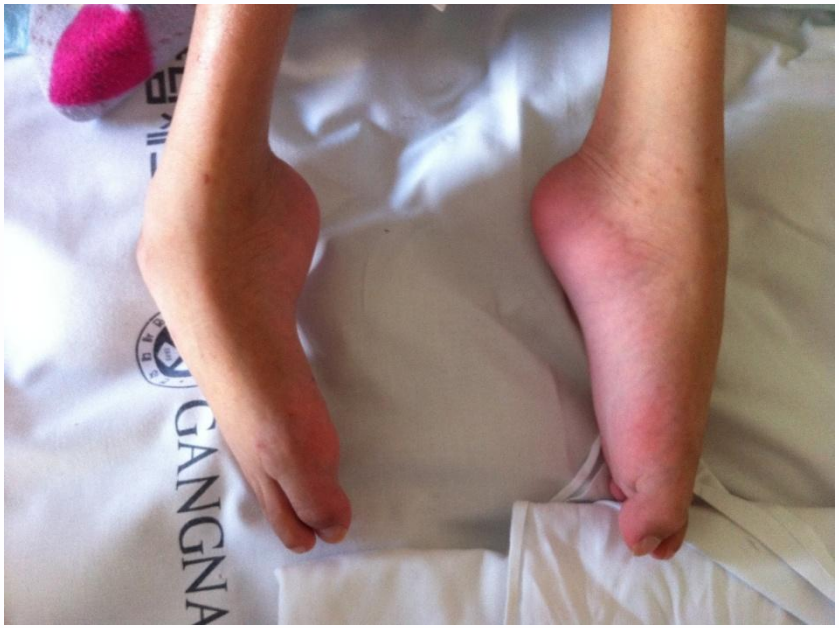
- Brain
  - injury or defined lesion in nervous system
  - developmental disdorder
- Spinal cord
  - congenital lesion (meningomyelocele,..)
- Peripheral nervous system
  - motor & sensory : hereditory
- Muscle disease

Muscle Disease

- Duchenne muscular dystrophy (DMD)
  - 1 / 3,500 Male birth
  - Invariable equinus deformity in DMD from 13 y/o
  - KAFO in children with DMD: >2/3 families were satisfied
- Metabolic myopathy



M/16, DMD



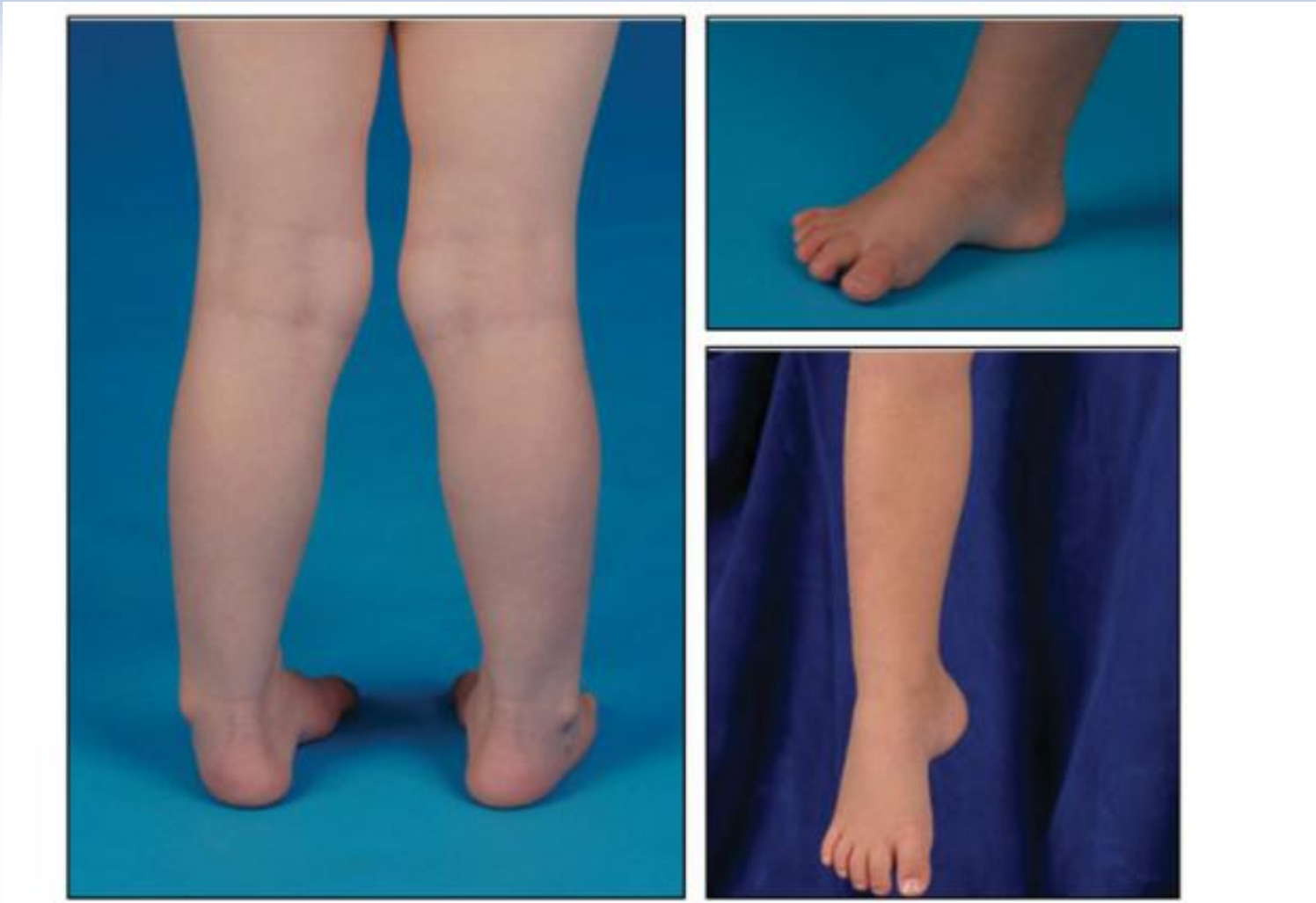
M/19, Glycogen storage disease

Peripheral nervous disease

- Guillain-Barré disease
  - weakness development from distal lower extremities

- *(Gupta et al. Disabil Rehabil 2010)*
  - acute peripheral polyneuropathy
  - affects motor, sensory, and autonimic ns. & spinal roots
  - early childhood/elderly
  - subject: 69 patients
  - residual deficits: 10% need walking aid at 2 years after onset
  - neuropathic pain (80%): limbs & back
  - foot drop (60%): almost bilateral – use AFO

    → need of AFO 34% at 1 year after

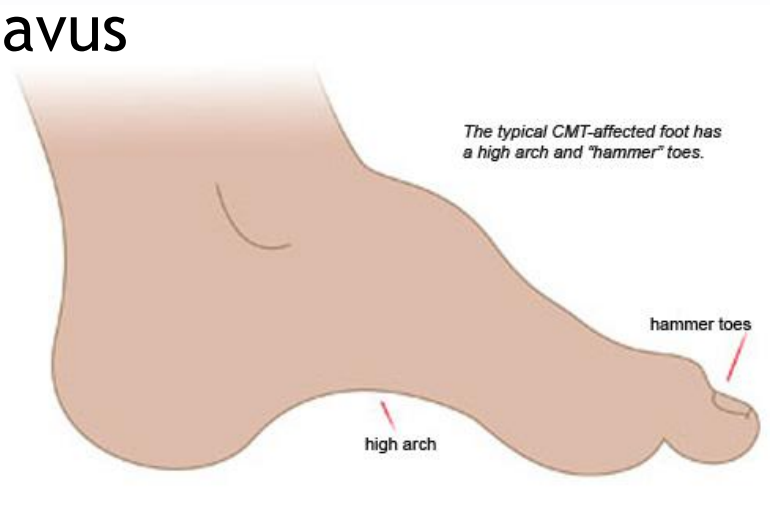


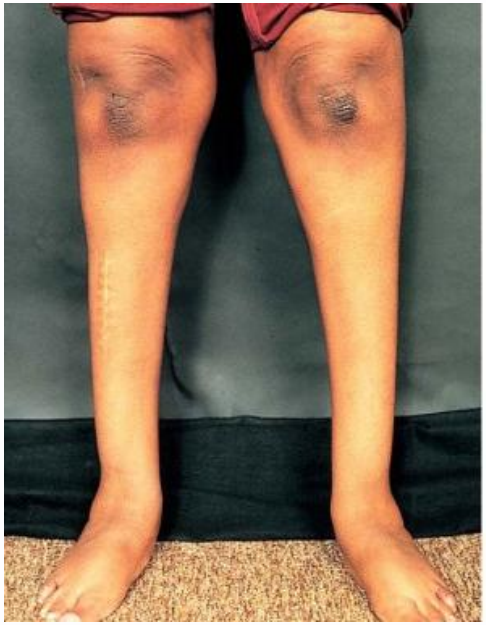
Early pes cavus in a 5-year-old child with CMT1A who scored an FPI of −5 on the right foot and −7 on the left foot.

Foot Posture Index : cavus =-12~-1/ normal = 0 ~ 5 / planus = 6~12

Peripheral nervous disease

- Charcot-Marie-Tooth (CMT)
  (hereditary motor-sensory neuropathy type I)
  - 1/2,500 births
  - heterogenous causes
  - recessive genes FGD4, PRX, MTMR2, SBF2, SH3TCs, GDAP1 : present early foot deformities with variable delay development (Baets, Brain 2011)
  - predominantly distal loss of strength and muscle atrophy in the leg
  - cavus





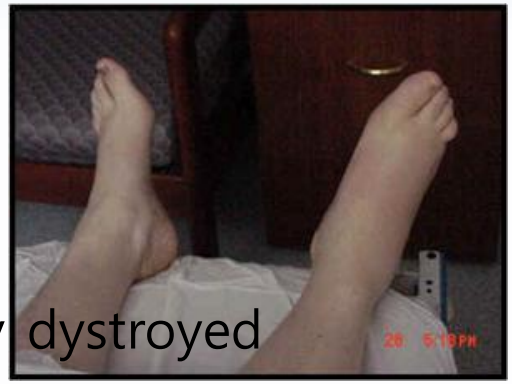
CMT & pain

- (Blyton et al. Neurology, 2011)
  - Type 1A: Autosomal dominant, large duplic. of chr. 17p
  - 81 children (mean 8.3 y/o, 2-16 y/o)
  - cramp
    - painful involuntary contraction of skeletal muscle
  - calf cramp in 32%, 1 child in toe
  - calf cramp associtated condition
    - older age
    - hand tremor
    - stronger foot inversion, eversion, dorsiflexion, plantarflexion
  - proposed mechanism of cramp
    - abnormal excitability of motor nerve terminals

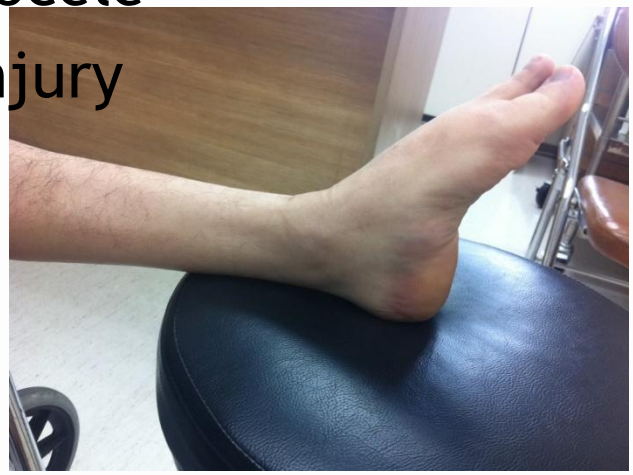
Peripheral nerve, dorsal root, spinal cord

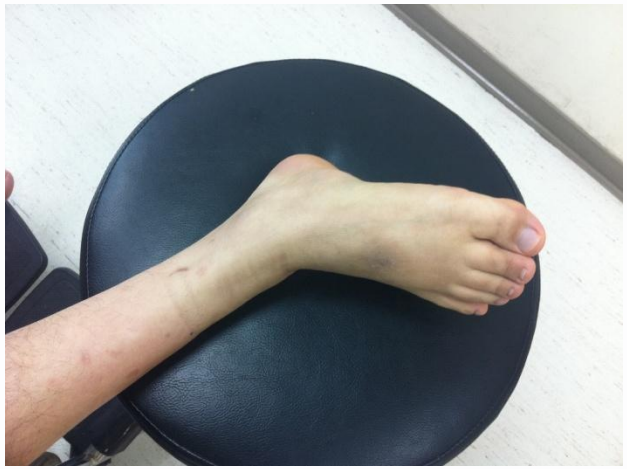
- Friedreich's ataxia
  - degenerative atrophy of the posterior columns of spinal cord: DRG, ppn, post.columm
  - autosomal recessive
  - most common earl-onset hereditary ataxia in Caucasians
  - ataxia, down going Babinski, areflexia
  - first symptom usually noticed around puberty
  - progressive limb ataxia after 10 ~ 15 years
  - scoliosis and pes cavus

Spinal cord level disease



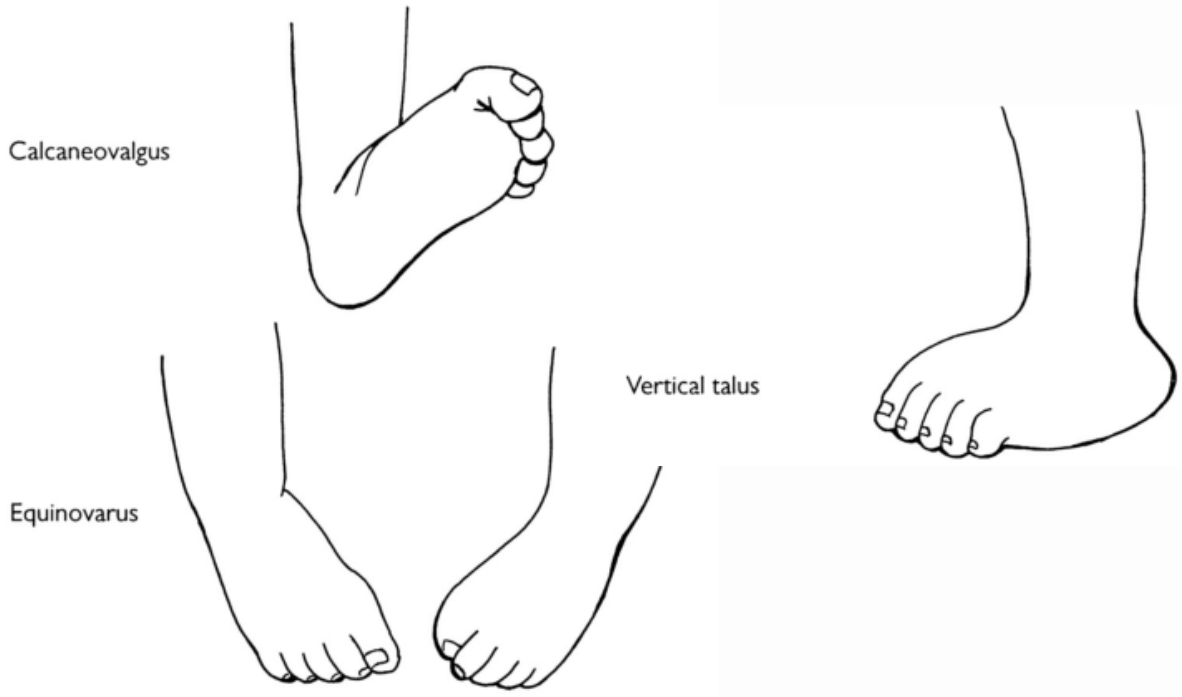
- Spinal muscular atrophy
  - autosomal recessive
  - 1/6,000 - 10,000
  - α motor neurons of ant. horn cell selectively dystroyed
  - type 2 onset 6 -18 months, type 3 >18 months
  - proximal and trunk weakness
- Meningomyelocele
- Spinal cord injury
- Poliomyelitis

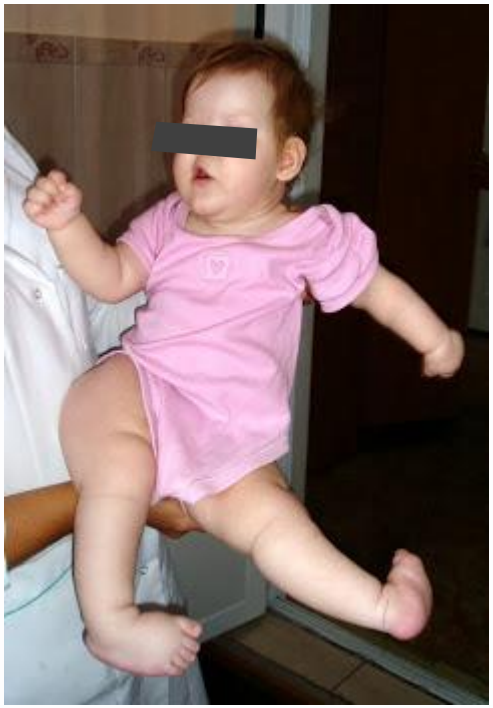




Level of lesion in spinal cord

- Foot manifestations are vary according to level of spinal cord lesion



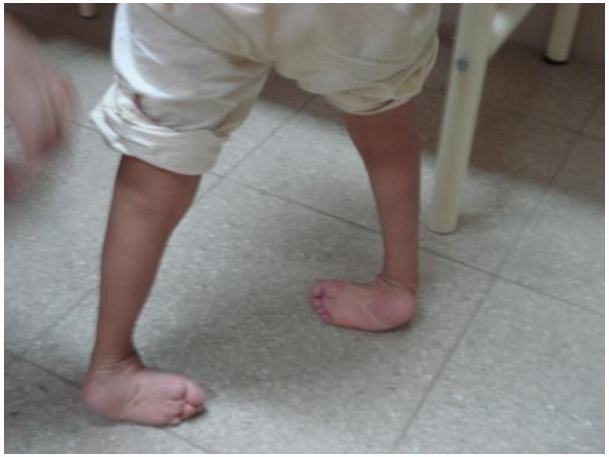


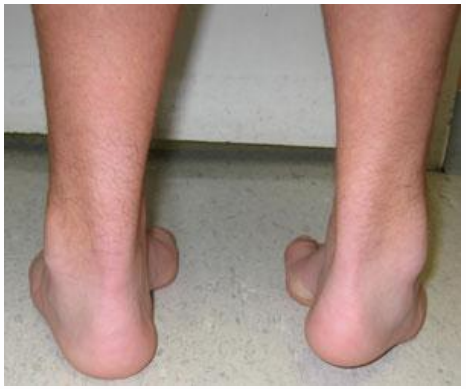
Cavus foot

- Etiology
  - CNS
    - Especially hemiplegia; spastic tibialis posterior
  - Spinal : Friedreich's ataxia
    - Myelodysplasia, syringomyelia, poliomyelitis, tetherd cord
  - Peripheral nerves
    - Guillain-Barre syndrome
    - HSMN, CMT

Cavovarus deformity

- Most common form
- Due to muscle imbalance
- Mild increase calcaneal pitch + forefoot plantarflexion deformity
- May be observed in CMT
  - CP
  - Friedreich's ataxia
  - Spinal dysraphism
  - poliomyelitis
- Toes clawing





Cavus foot

- Management
  - mainly orthotics
- Orthotics
  - For mild, non-progressive
  - Lateral forefoot and hindfoot posting
  - Large toe box shoes

Intervention for incresing ankle ROM in patients with neuromuscular disease (Cochrane)

- To resolve equinus
- RCT design
- 2 independent papers, 149 subjects
- CMT type 1A
  - no statistically or clinically significant differences for night splinting
- DMD
  - prednisolone use resulted improvements in strength and fucntion, but no difference in ROM

Brain-origin problems in foot & gait

Autism

- *Calhoun et al. Clin Biomech 2011:26*
  - 12 autism children: 22 control
  - reduced plantarflexor moments and increased dorsiflexion angles (hypotonia related?), decreased hip extensor moments
- *Barrow et al. J Child Neurol 2011: 26*
  - 954 ambulatory children
  - Autistic spectrum disorder
    - Persistent toe walking: 20.1%
    - Tight heel cords: 12.0%
  - Asperger syndrome
    - Persistent toe walking: 10.0%
    - Tight heel cords: 3.0%

Brain level problems

Cerebral palsy

- Brain lesion, non-progressive
- Typology
  - Spastic type
    - bilateral
    - unilateral
  - Dystonic type
  - Ataxic type
  - Choreoathetoid type

(Bax M, Goldstein M, Rosenbaum P, et al. Proposed definition and classification of cerebral palsy. *Dev Med Child Neurol*. 2005;47:571-576)

Global Delayed development

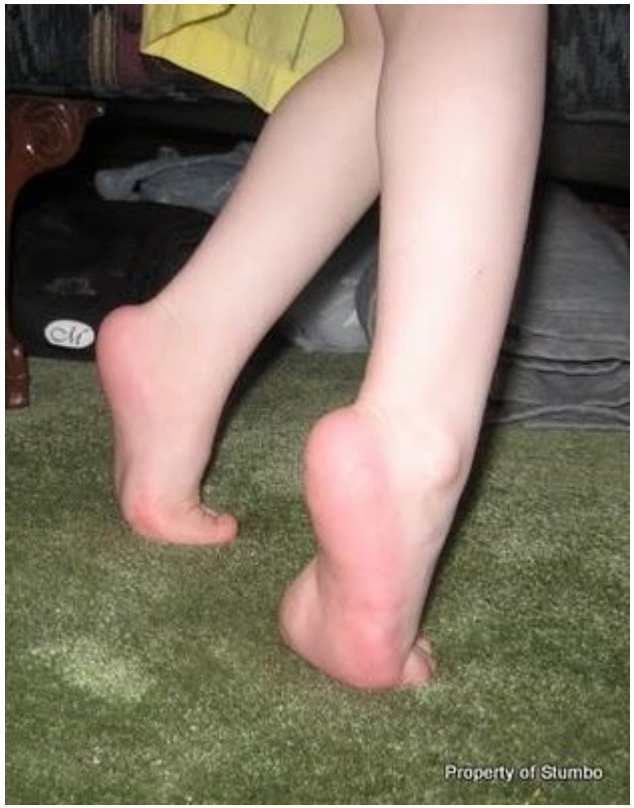
- Delay in ≥ 2 domains
  among gross/fine motor, speech/language, cognition, social/personal, ADL
- Variable degree of gross motor involvement
- Genetic syndromes
  - Down syndrome, autism,..
- Hypotonia & other tone presentation

Delayed motor development

- Hypotonia - trunk

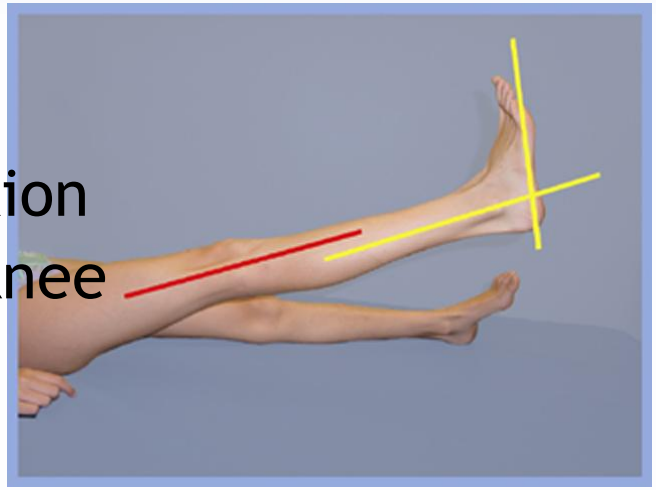
Equinus

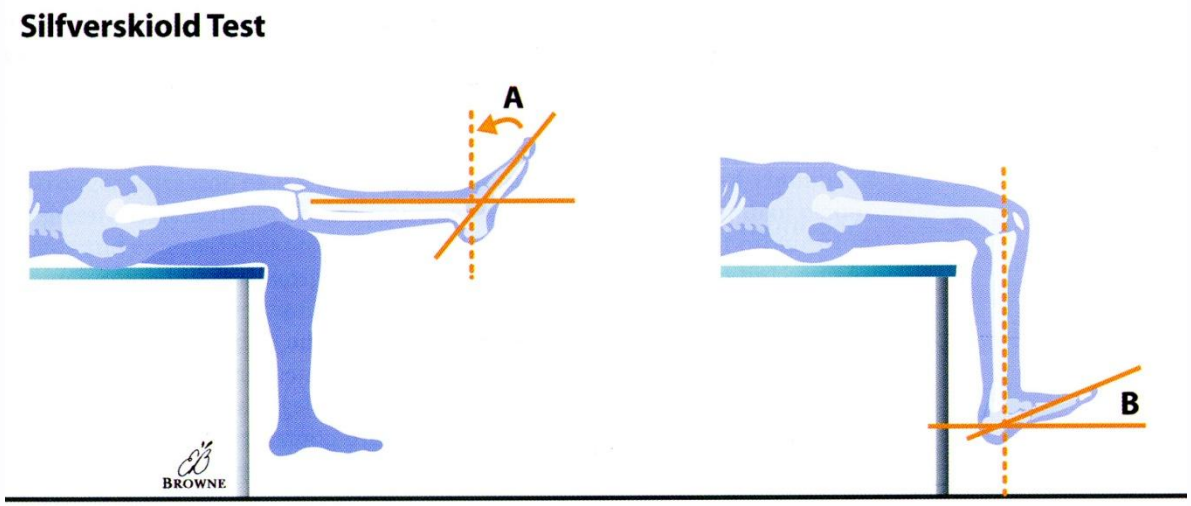
- Spastic bilateral type
- First deformity of spastic CP that gains attention
- Reason
  - Plantar flexor is 5 ~ 6 times stronger than dorsiflexor
  - Spasticity of total body, stretch reflex



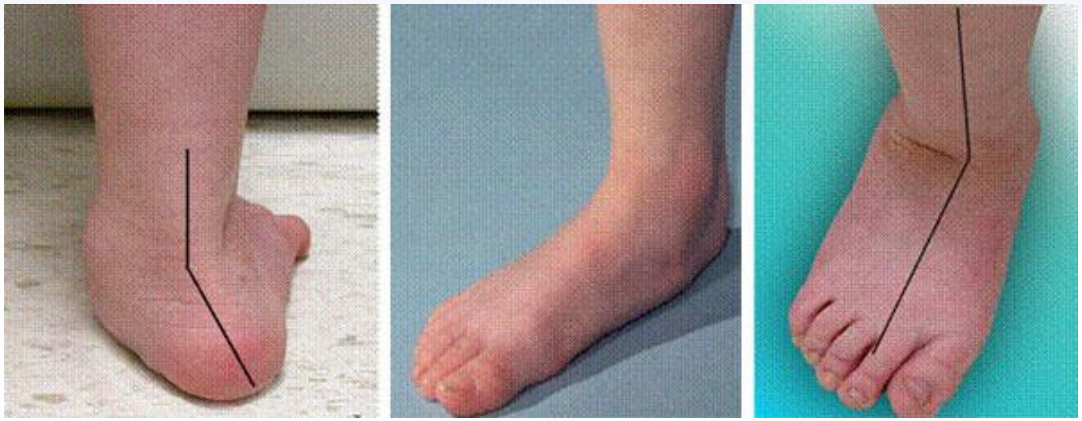
Equinus of Ankle

- Measure
  - Length of soleus : with knee flexion
  - Length of gastrocnemius : with knee extension





Ankle Valgus



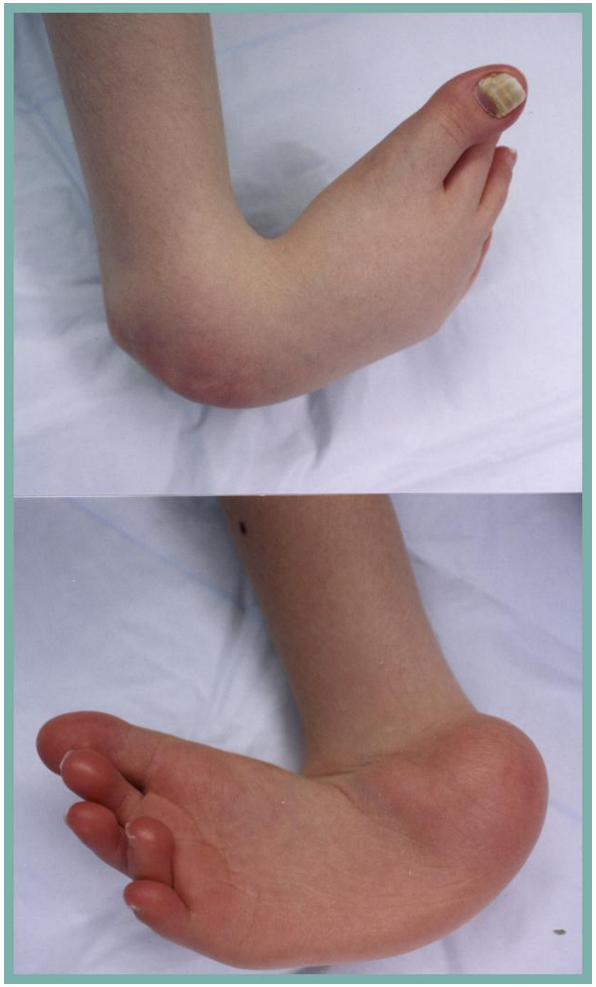
- External rotation planovalgus collapse
- Due to eccentric loading of ankle joint, partial growth arrest of lateral aspect of ankle joint
- Cause fibula to be shorter
- Complex rotational malalingnment of talus
- Gets worse during adolescent growth

Equinovarus

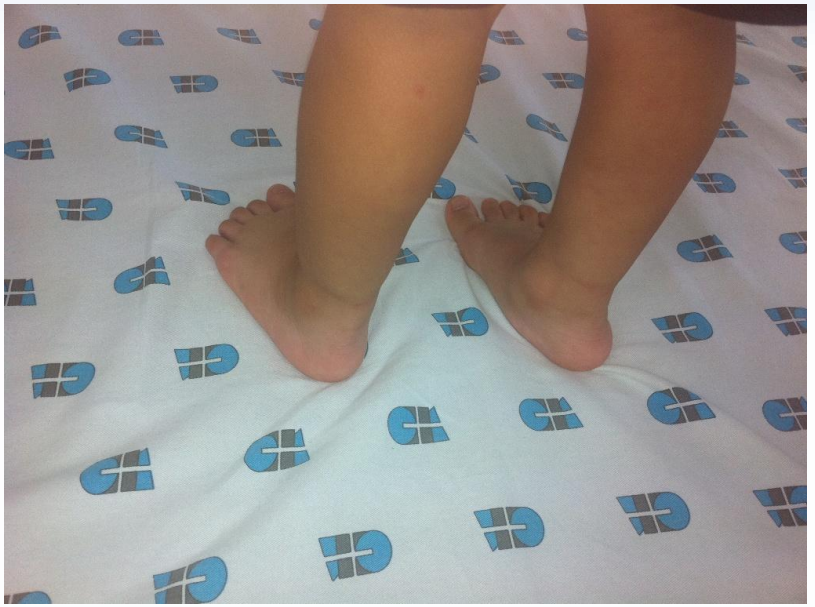
- Severe quadriplegia & hemiplegia

- Tibialis anterior & posterior

  : strongly pull



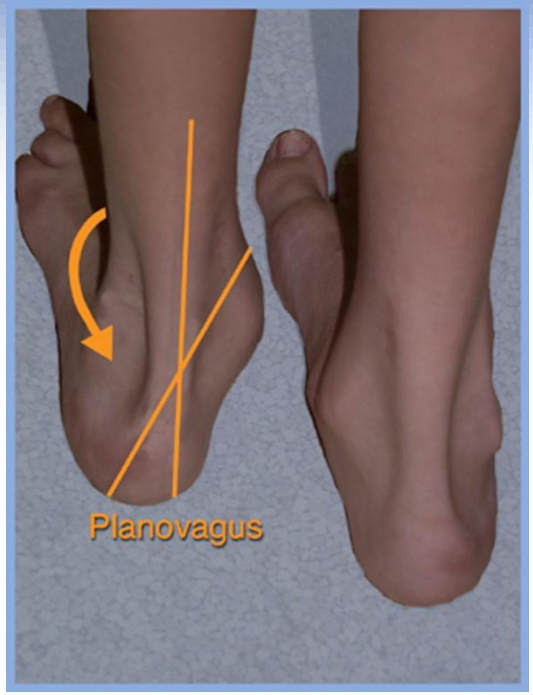
Planovalgus

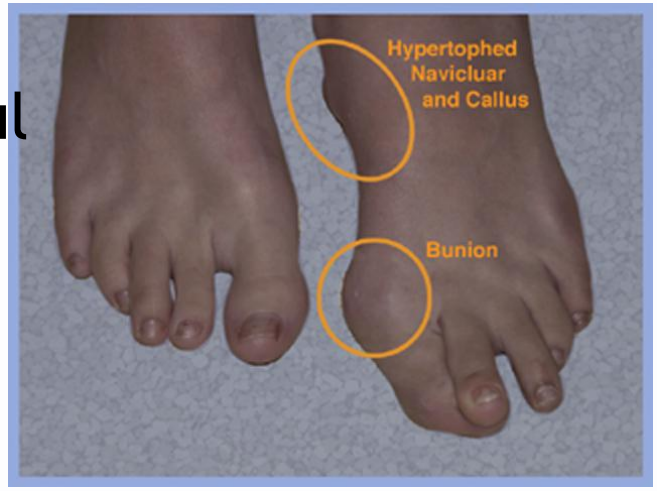


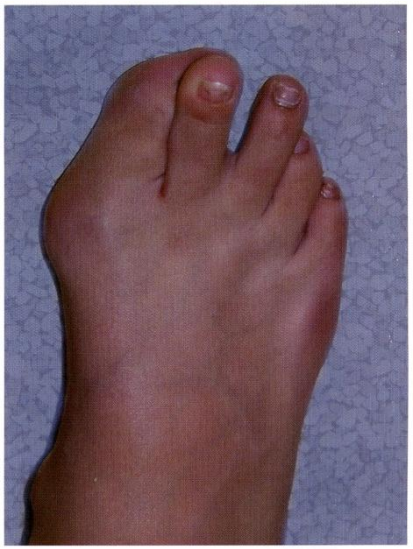
- Most common deformity
- Multifactorial etiology
  - High force enviornment
  - Toe-walking
- As collapse, navicular & head of talus become major weightbearing areas
- Often starts with toe-walking : high force on subtalar area

Planovalgus

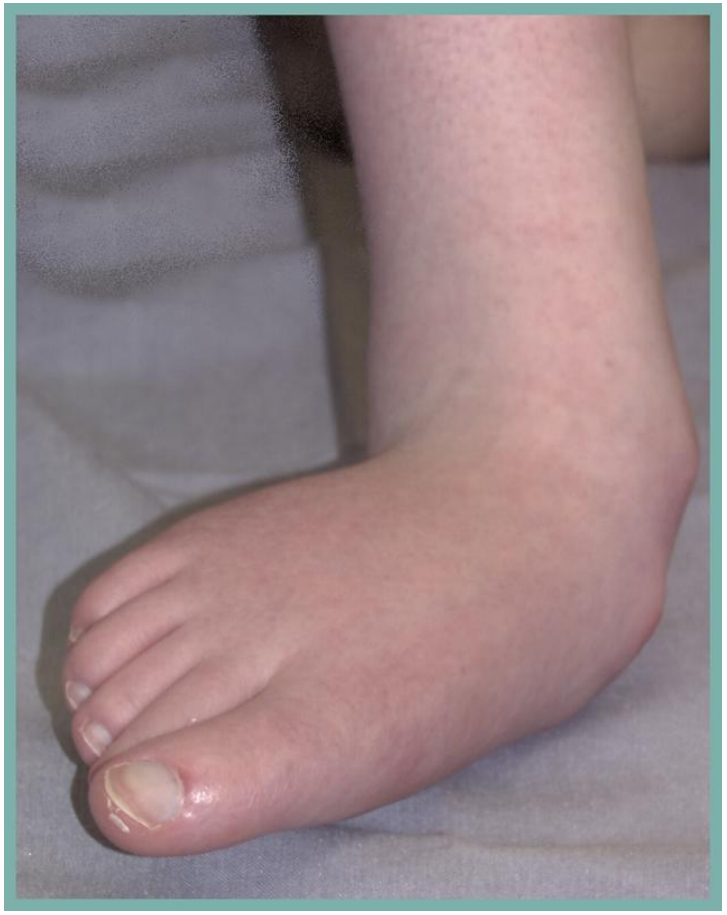
- As the child heavier, the force increases
  - joints start to collapse into external rotation, valgus → decreases the plantar flexion ability
- Often become painful
- Developing bunion

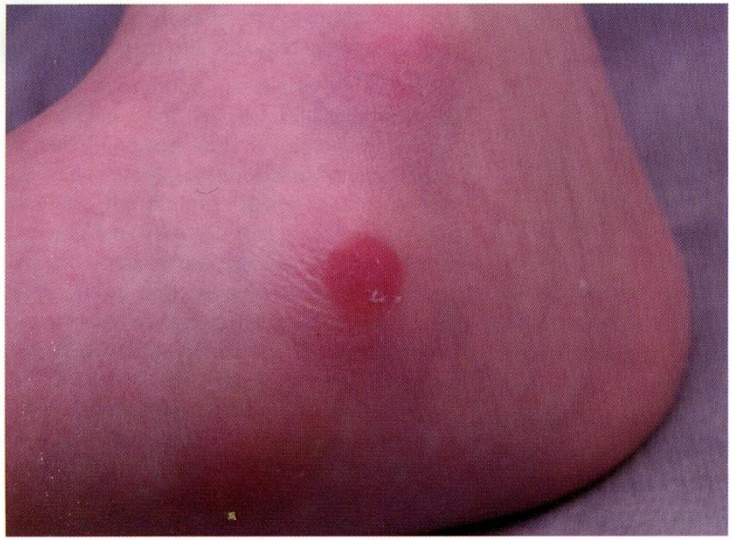




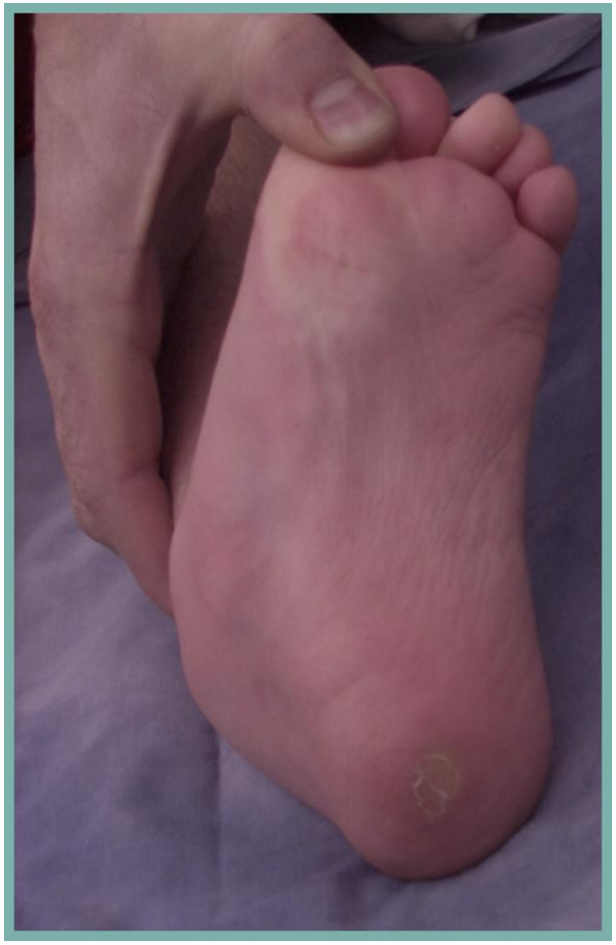


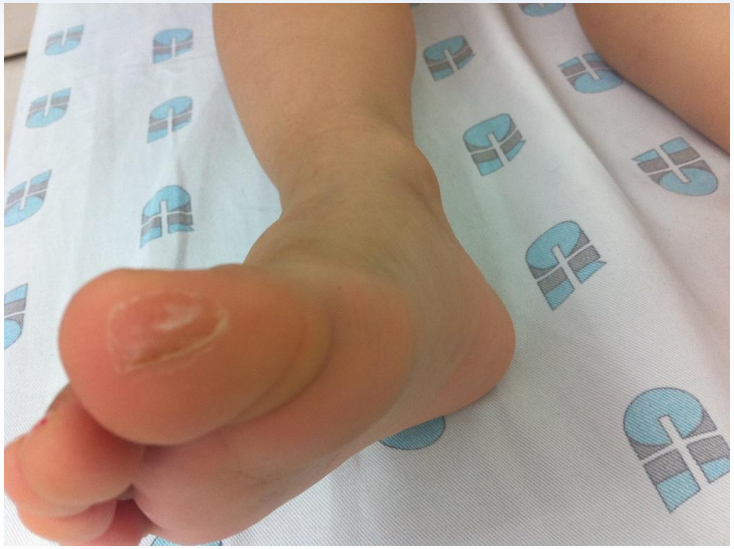
Severe case of Planovalgus

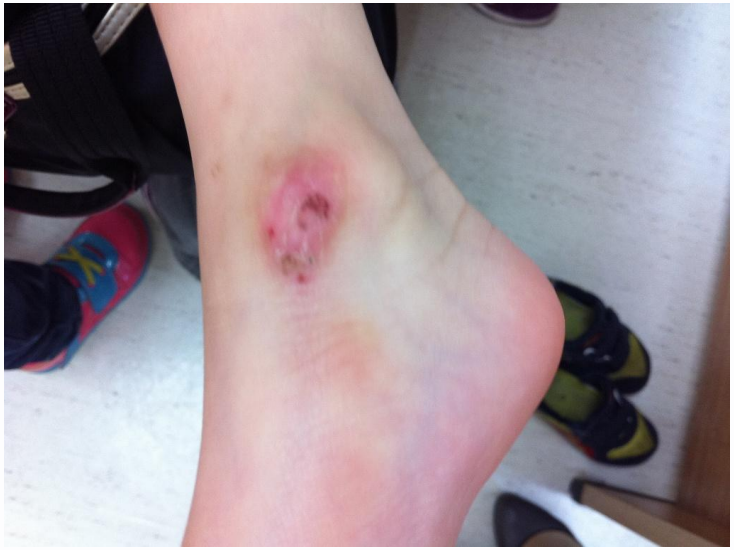




Calluces from chronic posture

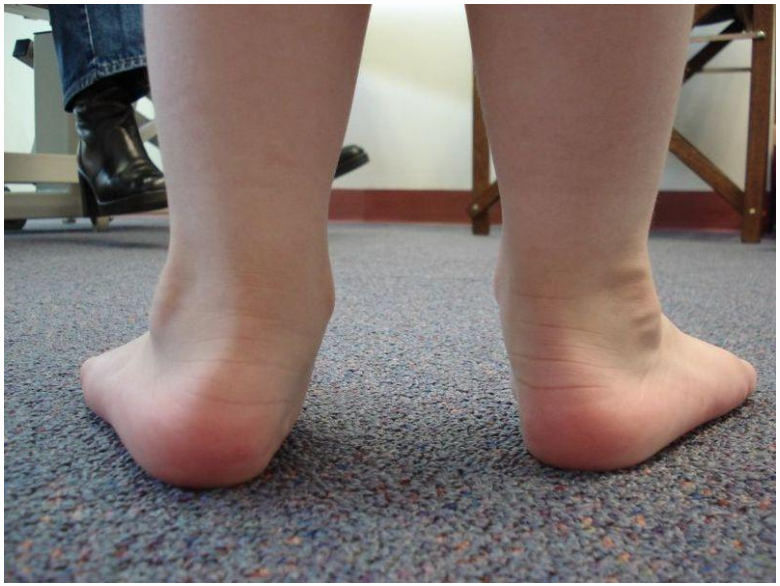


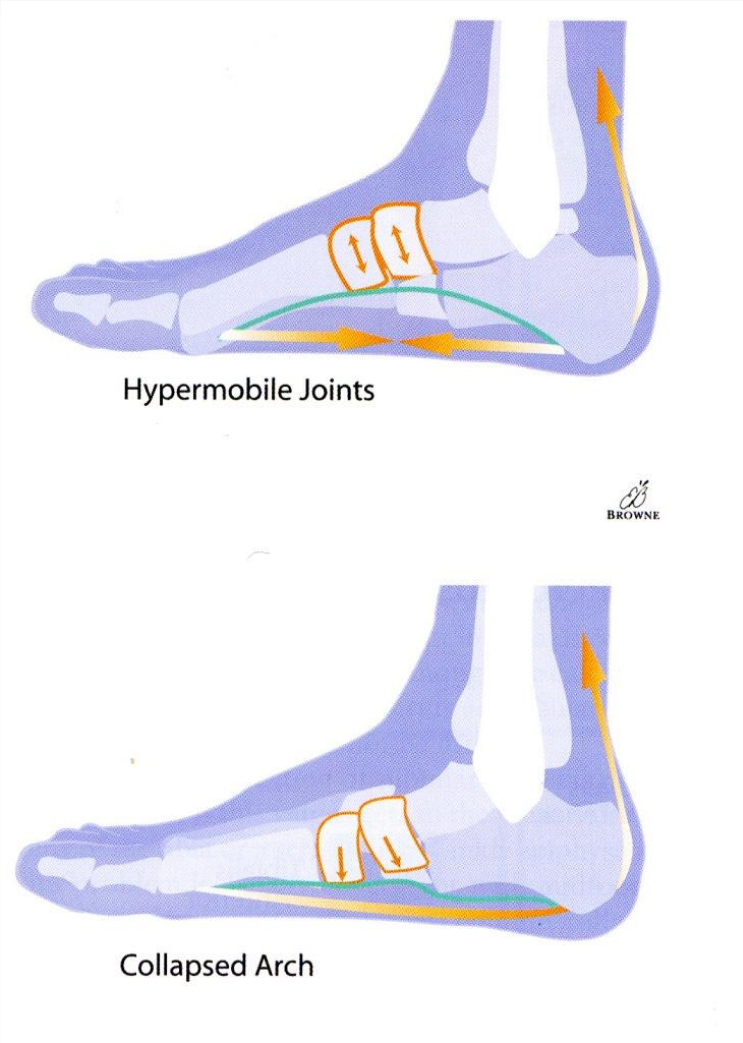




Flat foot

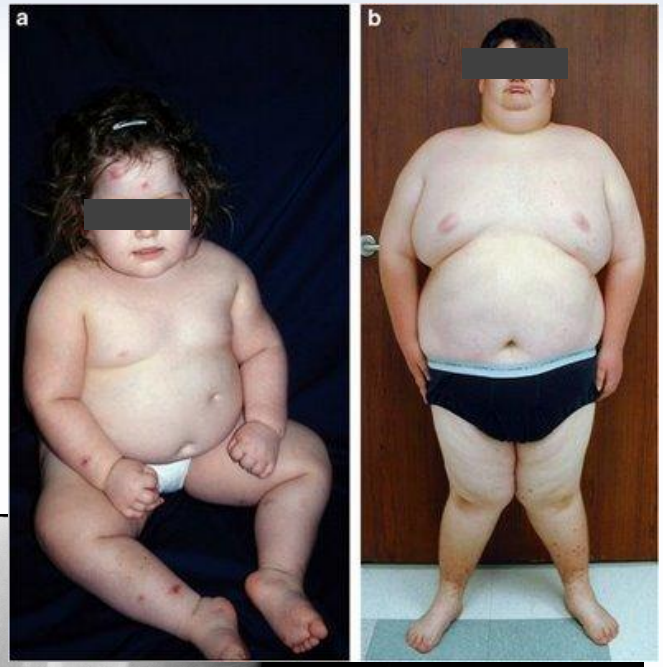
- Most hypotonic children
- Pronation
  - most frequent compensation for soft tissues or osseous deformities

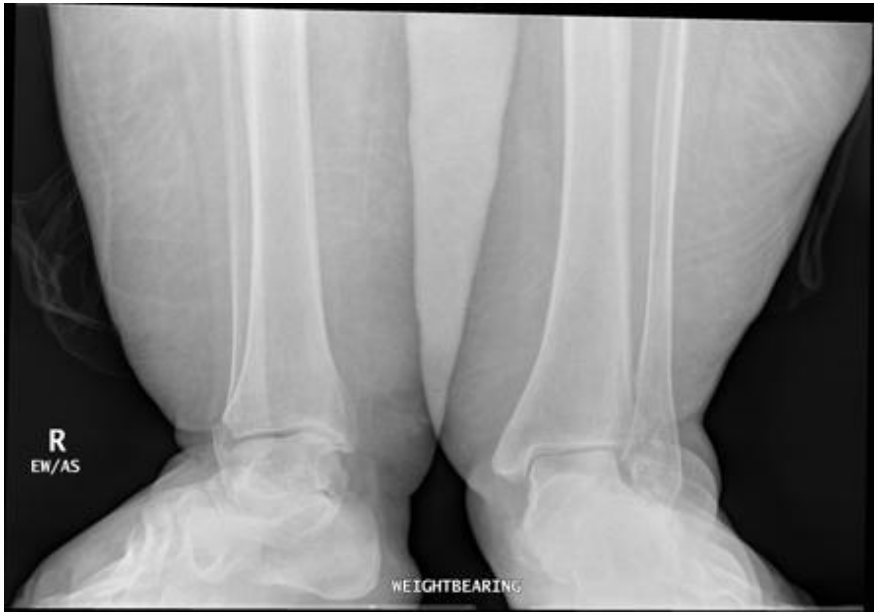


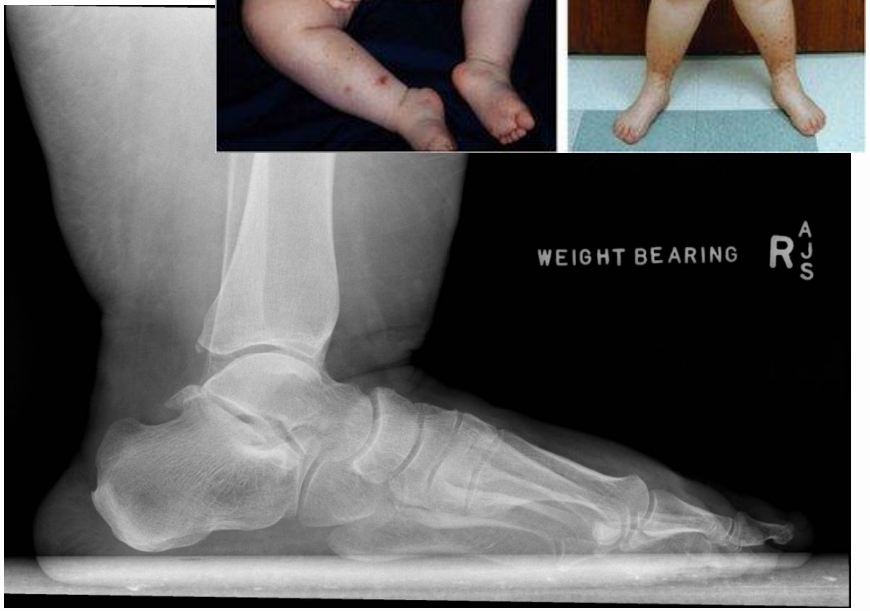


Pradder-Willi syndrome

- Rare genetic disorder
  - chromosome 15(q13-15)
- Pronation of feet
- Low tone, short stature







Down syndrome

- flat pronated feet
  - decreased ankle ROM
  - wide base of gait
  - inadequate heel contact
  - poor foot clearance during swing
- common foot conditions
  - hallux abduction, valugus
  - plantar fasciitis
  - clawing toes
  - shoes irritation in narrow shoes