Clubfoot
Congenital Talipes Equinocavovarvarus

Definition

- Congenital talipes equinocavovarvarus
  - Latin, talipes- talus, ankle + pes, foot ; equino-cavo-varus
- Congenital, contractural malalignment
- 4 sections
  - C- Cavus (high medial arch)- forefoot is plantarflexed in relation to the hindfoot
  - A- Adductus of the forefoot at the midfoot level
  - V- Varus/Inverted subtalar joint complex (Hindfoot is in varus)
  - E- Hindfoot equinus
Hindfoot varus positioning
Cavus foot position
Adductus position
Hindfoot in varus position
Types

- Idiopathic - normal child with clubfoot issues only
- Postural - resolves spontaneously with no Tx or with 1 to 2 casts
  - Can probably watch and see natural progression
- Neurogenic - related to myelomeningocele
- Syndromic - clubfoot seen with other anomalies as well

- Neurogenic and Syndromic are less likely to respond to non-operative techniques or tend to recur
Syndromes associated with clubfoot

- Arthrogryposis
- Constriction Bands
- Prune Belly
- Tibial hemimelia
- Mobius syndrome
- Freeman-Sheldon Syndrome - Whistling face
- Diastrophic dwarfism
- Larsen Syndrome
- Opitz Syndrome
- Pierre Robin Syndrome
1-2 per 1000 commonly quoted
- Variable by population groups
- Nordic (e.g., Sweden) - 0.93 to 1.5
- Asian - 0.6 per 1000
- Western Australia - 0.9
- Polynesians, Hawaiians, Maori - 6.8
What is the chance that it can happen in a family that already has a case of clubfoot?

- Occurs 17 times higher in first-degree relatives
  - Sharing 50% of genetic material - parents, children, siblings
- Occurs 6 times higher for second-degree relatives
  - Aunts, Uncles, grandparents, grandchildren, nieces, nephews, half-siblings
- Occurs same incidence as general population for third-degree relatives
  - Great-grandparents, Great grandchildren, Great uncles/aunts, First cousins

Wynn-Davies, JBJS (Br) 1964, Clin Orthop Relat Res 1972
Can this happen again in a family?

- Yes.
- Unaffected parents
  - One son with clubfoot - 1 in 40 chance of another son with clubfoot
  - But, subsequent daughter - unlikely to be affected
  - One daughter with clubfoot - 1 in 16 chance of a son with clubfoot and 1 in 40 chance of another daughter with clubfoot
- One affected parent and one affected child
  - 1 in 4 chance of another child having disorder
Etiology

- Heterogenous, Multifactorial inheritance
- Some suggestion of single gene, two alleles plus other un-identified factors
- Proposed theories
  - In utero molding
  - Primary muscle lesion
  - Primary bone deformity (germ plasm)
  - Primary vascular lesion
  - Intrauterine enteroviral infection
  - Developmental arrest
  - Primary nerve lesion
  - Abnormal tendon insertion
  - Retracting fibrosis
  - Abnormal histology
Environmental Factors

- Do they alter genetic expression?
- Cigarette smoking
  - Increased risk if smoking during pregnancy
  - Risk increased with higher number cigarettes smoked per day
  - Family history plus smoking - particularly higher
Genetics

- Chromosome 12q24.31 between NCOR2 and ZNF664- single nucleotide polymorphisms
  - Zhang TX et al, J Med Genet 2014 May

- Transcription factors and transcriptional regulators: PITX1
  - Dobbs, 2013, 2014
Signs

- Cavus
- Adductus (Inversion of subtalar joint - between talus and calcaneus)
- Varus
- Equinus
- Varying severity - flexible to rigid
Equinus and Cavus foot positions
Signs (continued)

- Single large (or Double) posterior ankle crease
- Empty heel pad sign - cannot feel calcaneus
- Transverse medial crease (midfoot)
- Palpable head of talus - dorsolateral over midfoot region just distal to ankle joint
- Smaller foot and calf – “clubleg”
Clubleg
Adductus component with curvature of lateral border of foot and medial crease
Signs (continued)

- LLD- greater than 0.5 cm
  - 18% if unilateral clubfoot
  - 4% if bilateral clubfoot
- May have higher internal hip rotation – 10 degrees or more
  - Why is this important?
  - Intoeing in a clubfoot patient may be due to hip rotation NOT recurrence of a treated clubfoot
- Watch for the syndromic and neurogenic patients
Classifications

- Dimeglio
- Pirani
- Most common referred to in literature
- Dimeglio is probably most reproducible
- Point systems with each feature summed up to give a composite score which reflects severity of the involved foot
- Other described classification schemes- Catteral, Harold and Walker, Ponseti and Smoley
DiMeglio Classification

Classification of clubfoot severity by Diméglio A. Equinus deviation B. Varus deviation C. Derotation D. Adduction.
Radiographic Studies

- Not required for diagnosis and management
- Poor reproducibility of foot positioning
- "Forced dorsiflexion" AP and lateral
  - Foot is held in maximum dorsiflexion while x-rays are taken
- In utero ultrasound
  - Recognition is 0.1% to 0.4%
  - Low false-negative
  - False-positive - 30 to 40%
  - Functional false-positive - correctible passively after delivery (Positional clubfoot)
Further intrauterine evaluation

- Controversial if amniocentesis is required with identified, isolated clubfoot deformity
  - To identify syndromic presentation
  - Recognize additional abnormalities
- Orthopedic consultation
  - For family counseling regarding etiology, treatment, prognosis
Pathoanatomy

- Malalignment of bones at the joints
  - How?
  - Partly deformation of the bones themselves
Natural History

- Untreated clubfoot
  - Rigid
  - Callus with bursa develops on the dorsolateral aspect
  - Hyperflexed midfoot
  - Extreme case - toes point backwards during ambulation
  - May be functional with a prosthesis
  - Pain more with hard floors and sidewalks
Untreated Clubfoot
Untreated clubfoot - dorsolateral callus
Untreated clubfoot x-ray
Treatment

- Non-operative Treatment
  - To achieve a plantigrade, supple, painless foot with normal appearance
  - To avoid special shoe wear
- Hiram Kite (Kite casting technique)
  - Poor long term results
  - Long-term immobilization
- Ignacio Ponseti (Ponseti casting technique)
- “French” Method
  - Inpatient admission
  - Serial strapping and physical therapy manipulations
Ignacio Ponseti

Started in 1940s

University of Iowa

Mid 1990’s began to be more accepted

Cooper and Dietz- long term, 34 year follow up of the Ponseti technique
Treatment- Ponseti technique

- Serial casting
- Average 4 to 8 casts
- Has been shown to be effective even up to age 9!
- Gentle manipulations/castings
- Starting even several months after birth may lead to similar outcomes
- Fiberglass shown to be superior to plaster of Paris for:
  - Durability, convenience, performance, ease of removal.
- Has been performed by lay individuals (no formal medical training)
Ponseti Casting

Note first cast: midfoot/forefoot is addressed first

Note final cast: hindfoot now dorsiflexed and in valgus position
Foot Abduction Orthosis and the Cast Knife

Adjustable bar for length

Cast knife for cast removal
Posteromedial Release
Bibliography

- [www.law.cornell.edu](http://www.law.cornell.edu)- 29 CFR 1635.3- Definitions specific to GINA.

Lovell and Winter Pediatric Orthopaedics, 7th ed., editors Stuart Weinstein, Jack Flynn

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