Congenital Hand Conditions

David Martineau, MD

10/13/23

Outline

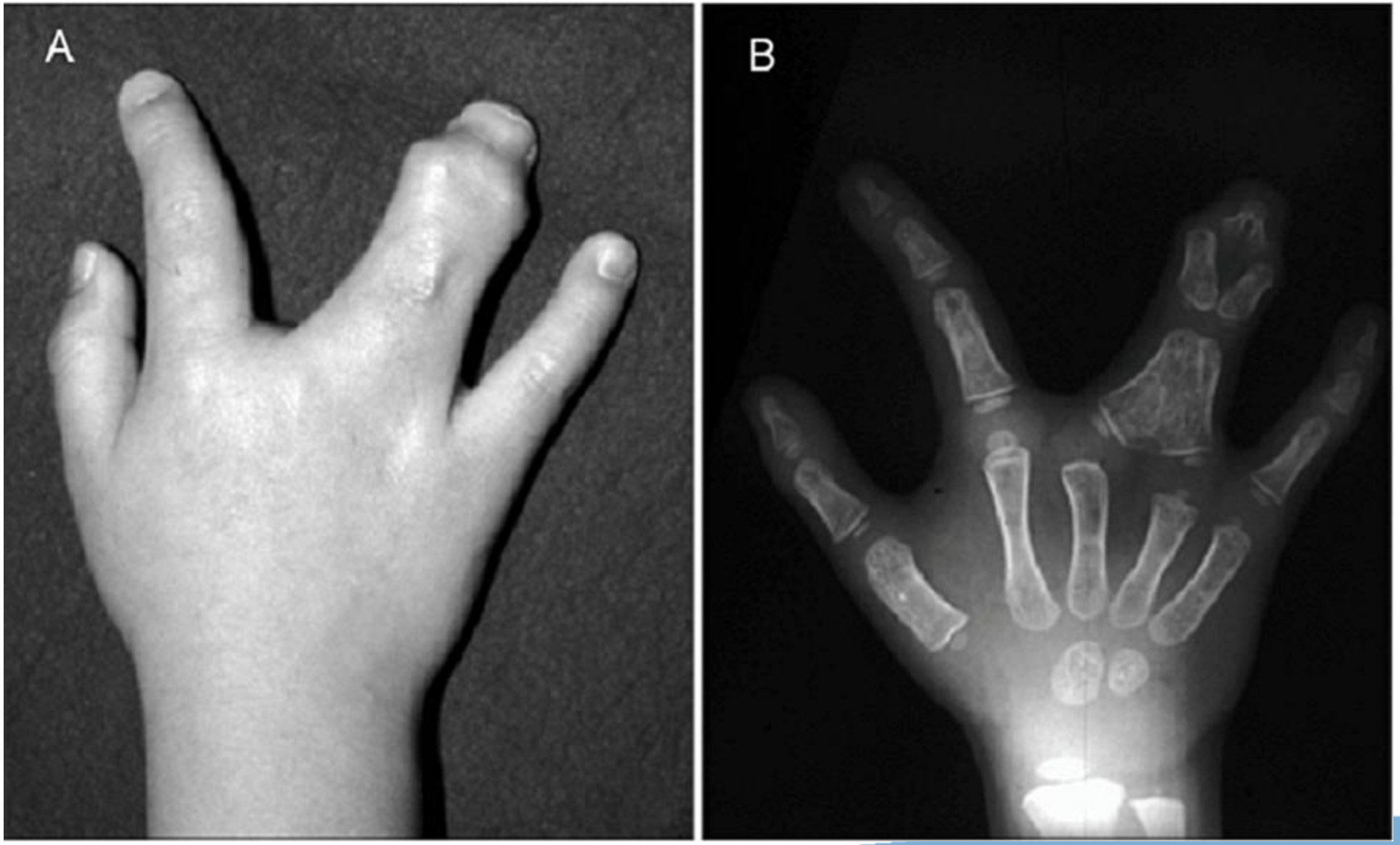
- Syndactyly
- Polydactyly
- Amniotic Band Syndrome
- Camptodactyly
- Clinodactyly
- Congenital Trigger thumb

Outline

- **Syndactyly**
- Polydactyly
- Amniotic Band Syndrome
- Camptodactyly
- Clinodactyly
- Congenital Trigger thumb

Syndactyly

- Derivation
  - *Syn*=together; *Dactylos*=digit
- Incidence
  - 1 in 2,000, the most common congenital hand deformity
- Etiology
  - failure of apoptosis of the digits
- Classification
  - Simple / Complex
    - soft tissue vs bony involvement
  - Incomplete / Complete



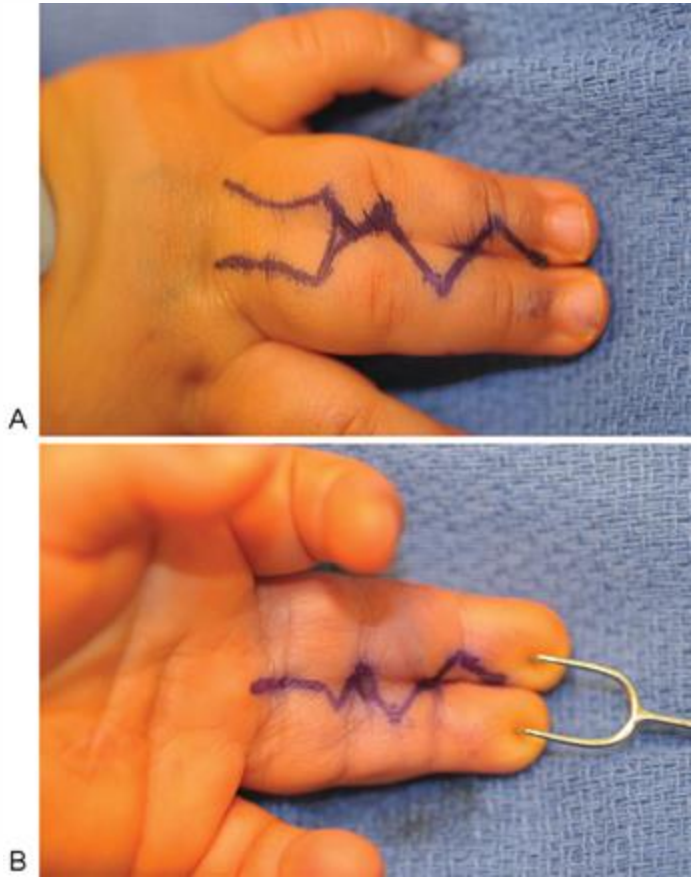
dayton children's

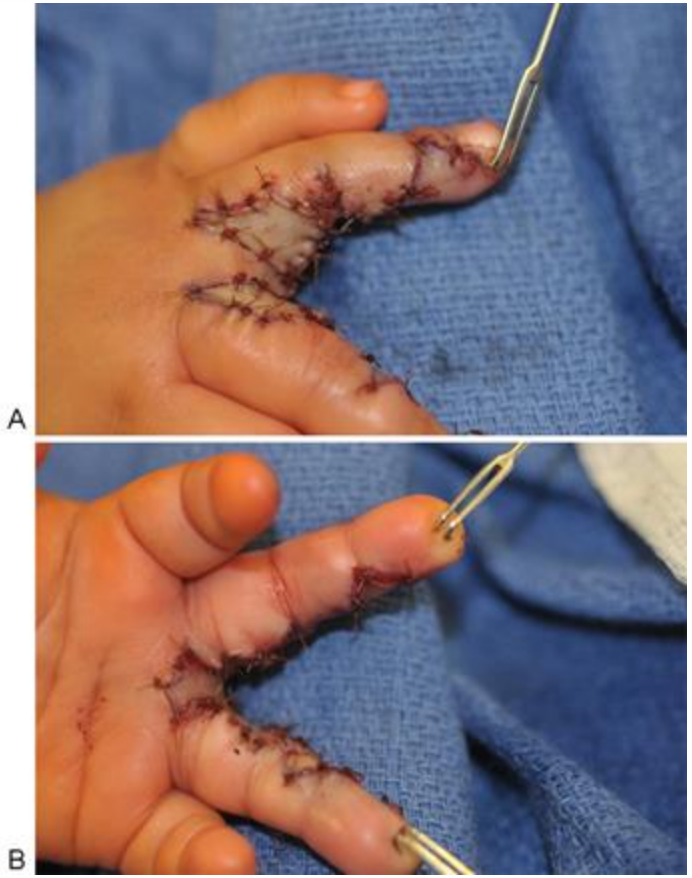
Management

- Nonoperative treatment
  - mild and incomplete
  - overall limited role for nonoperative treatment

- Operative treatment
  - Typically first line
  - Time to intervene - depends on the involved digits
    - Ring-Long; index middle
      - performed at about 1 year of age to mitigate effects of anesthesia
    - Border digits (e.g. thumb-index, or ring-small)
      - performed earlier at ~6 months due to length differential and inevitable tethering deformity
  - If Multiple digits involved
    - surgery to one side at a time, to preserve vascularity and digit perfusion

Management





Polydactyly

Baseball players are smarter than football players. How often do you see a baseball team penalized for too many men on the field?

-Jim Bouton

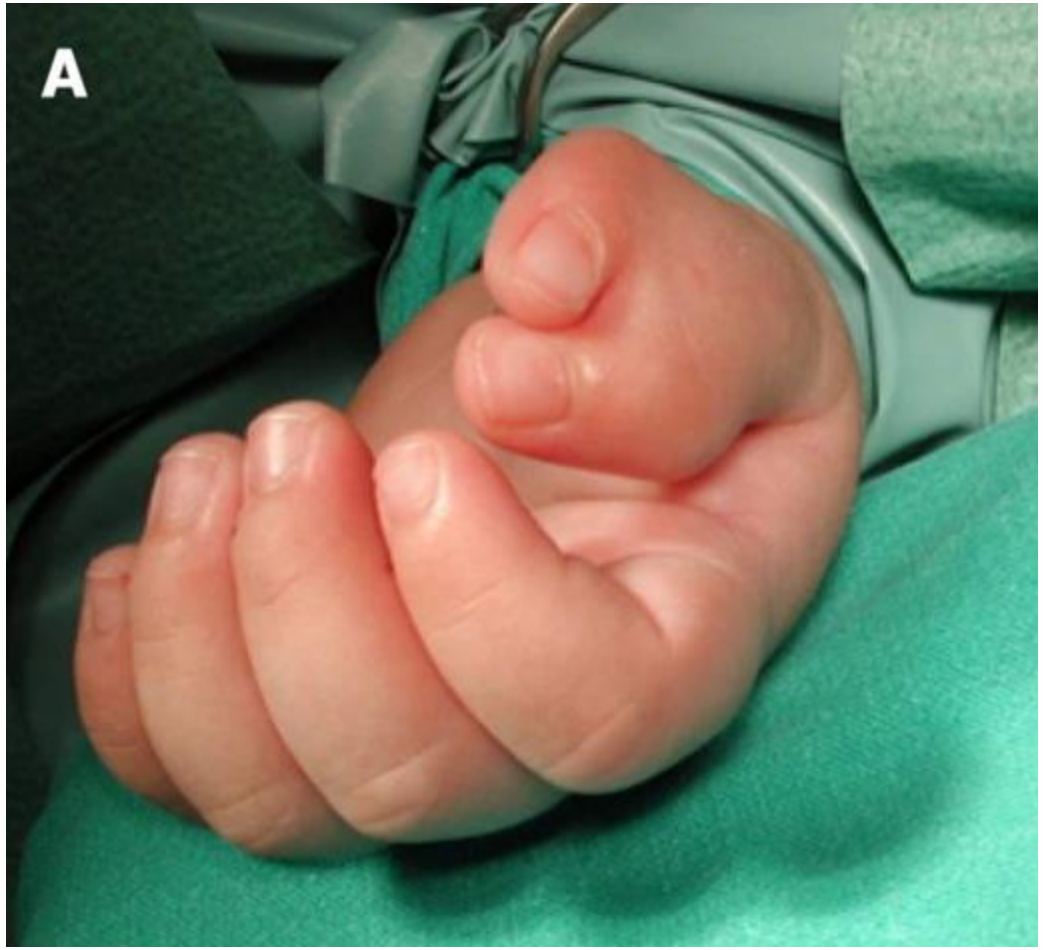
Polydactyly

- Incidence
  - 1 in 1,000

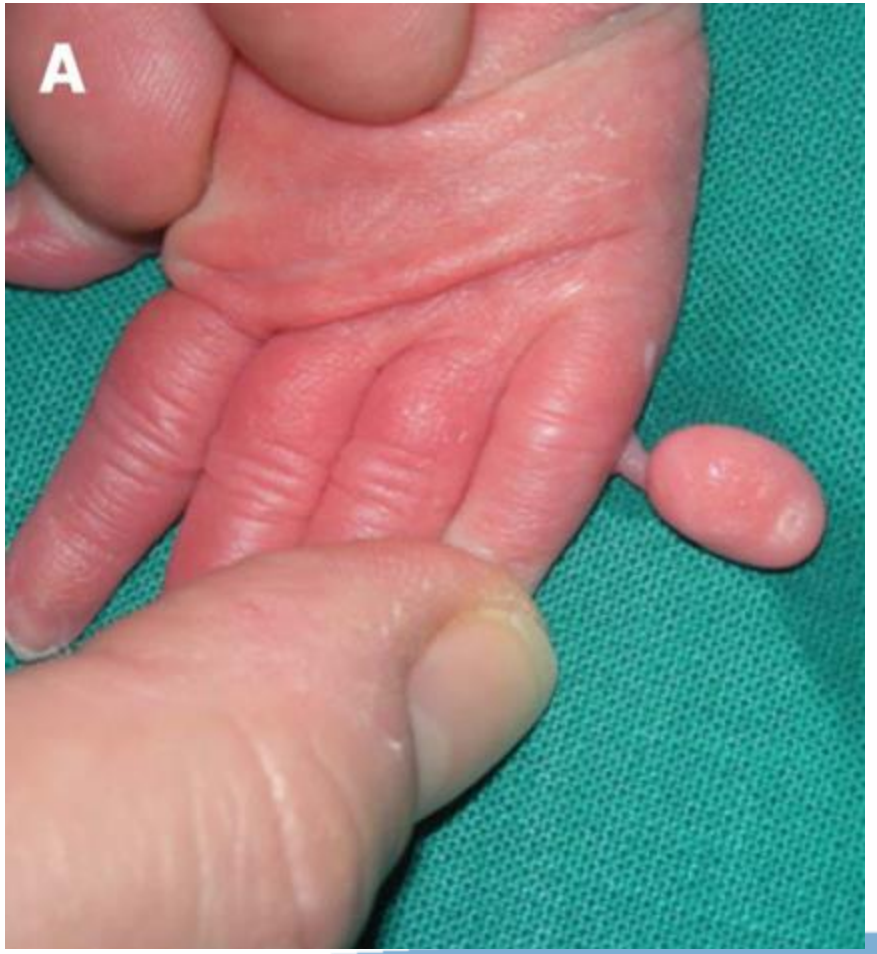
- Etiology
  - genetic mutation causing failure in limb bud development

- Types
  - Preaxial -Thumb Duplication
  - Postaxial -Small Finger Duplication
  - Central
  - Mirror image

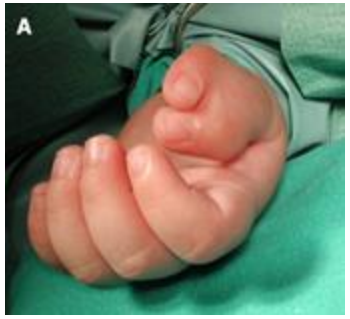
Pre Axial



Post Axial

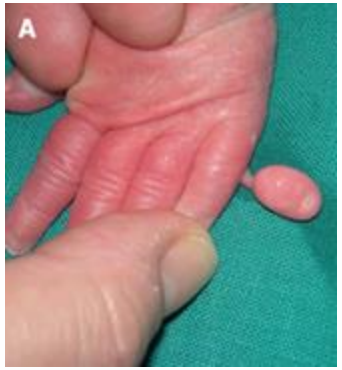


Pre Axial



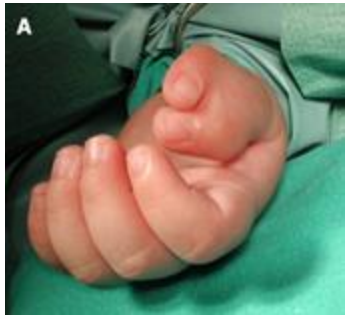
- Genetics
  - unilateral and sporadic
- Demographics
  - typically in Caucasians

Post Axial

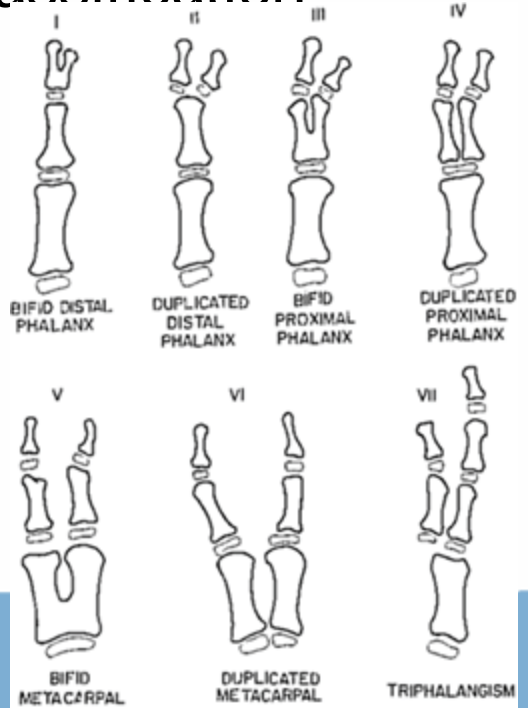


- Genetics
  - Autosomal dominant
    - chr19, chr13, Gli3 gene chr7
- Demographics
  - African Americans 10X > caucasians
  - if Caucasian, genetic work up to r/o chondroectodermal dysplasia or Ellis-van Creveld syndrome

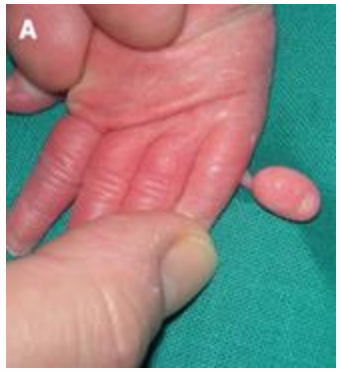
Pre Axial



- Classification



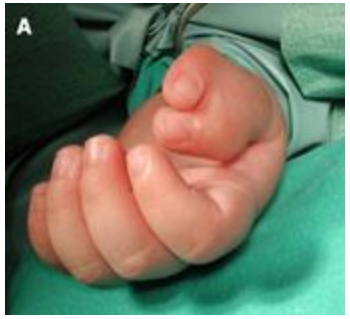
Post Axial



- Classification
  - Type A - fully formed digit
  - Type B - vestigial skin tag

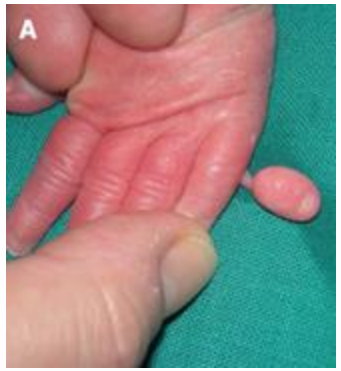
dayton children's

Pre Axial



- Management
  - virtually all require surgery, ideally at age 1-2 years old
  - typically ulnar thumb is preserved and the radial digit is excised

Post Axial



- Management
  - Type A
    - planned surgical excision at age 1-2 for type A
  - Type B
    - suture ligation ideally before age 1, can be done in the nursery
    - local anesthesia with bovie cauterization

dayton children's

Pre-axial Polydactyly Surgical Techniques

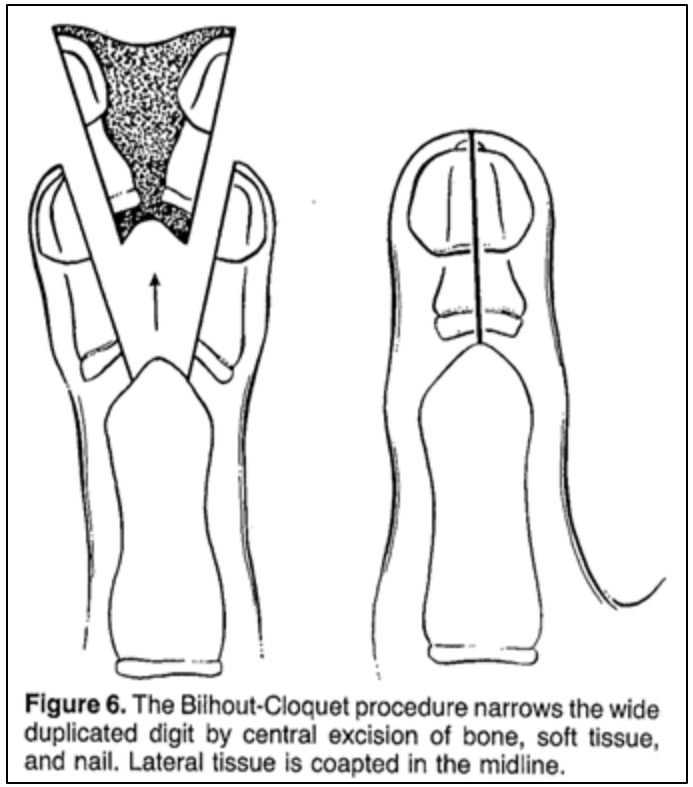
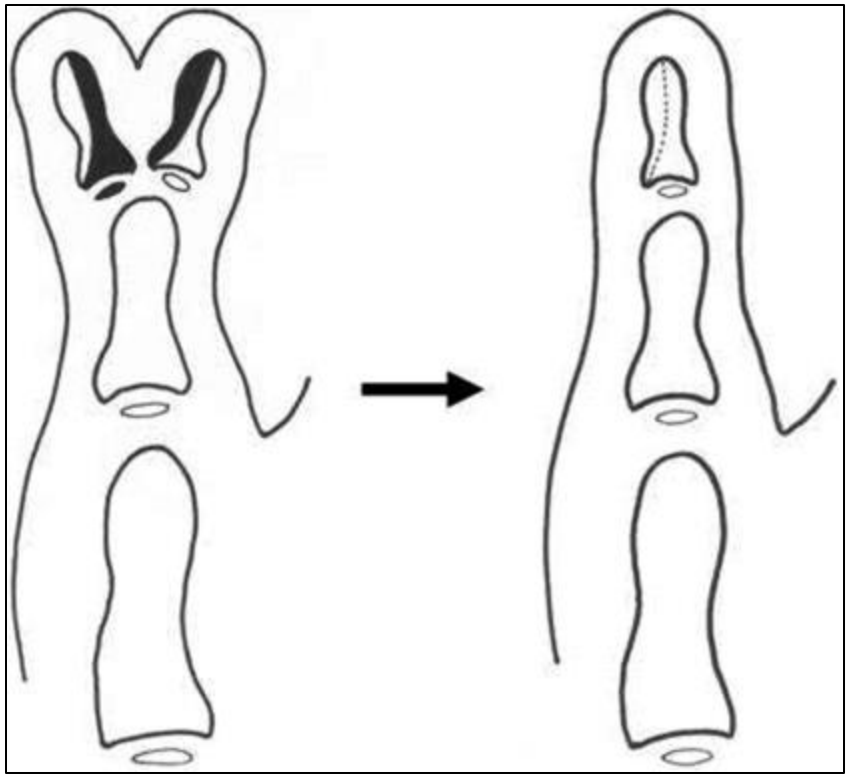
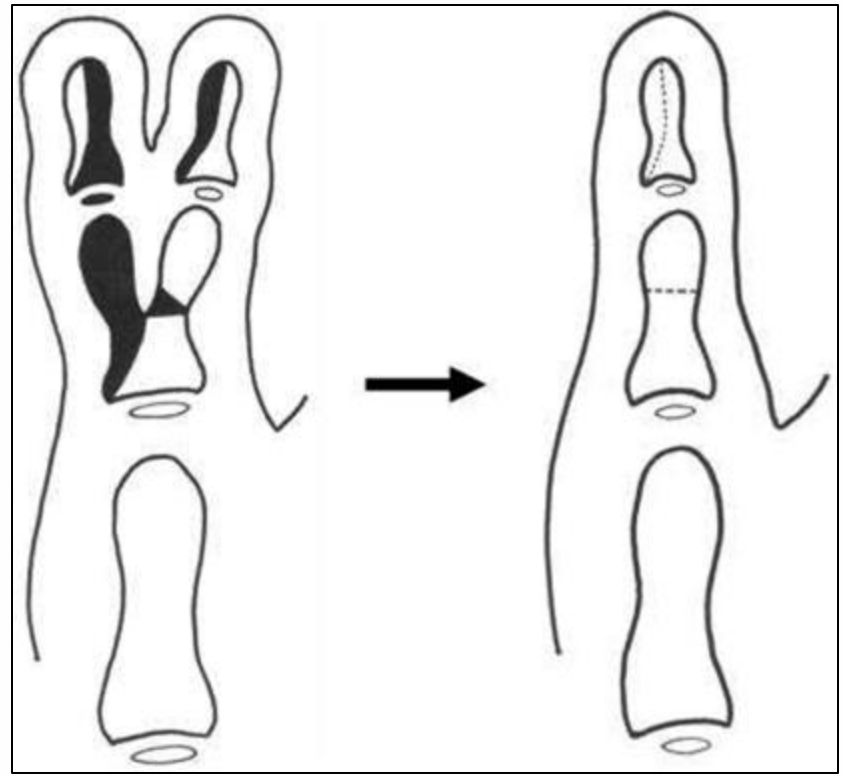


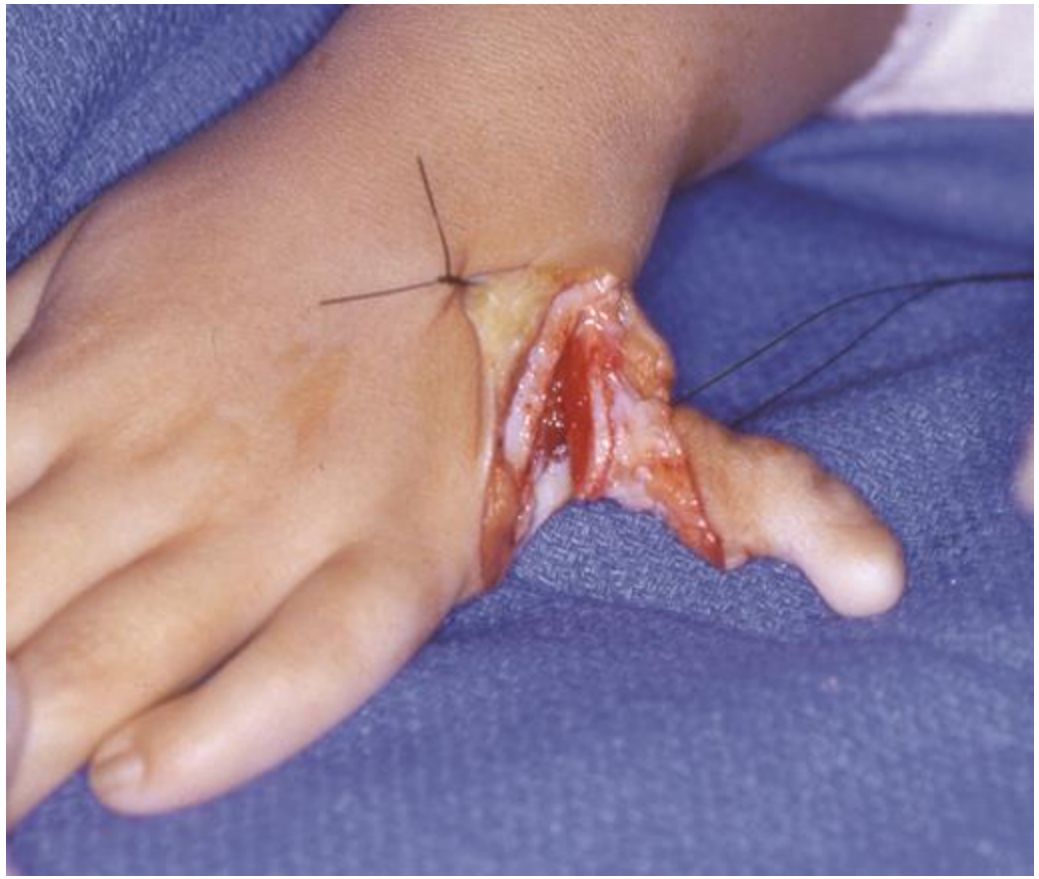
Figure 6. The Bilhout-Cloquet procedure narrows the wide duplicated digit by central excision of bone, soft tissue, and nail. Lateral tissue is coapted in the midline.





dayton children's

Post-axial Polydactyly Management



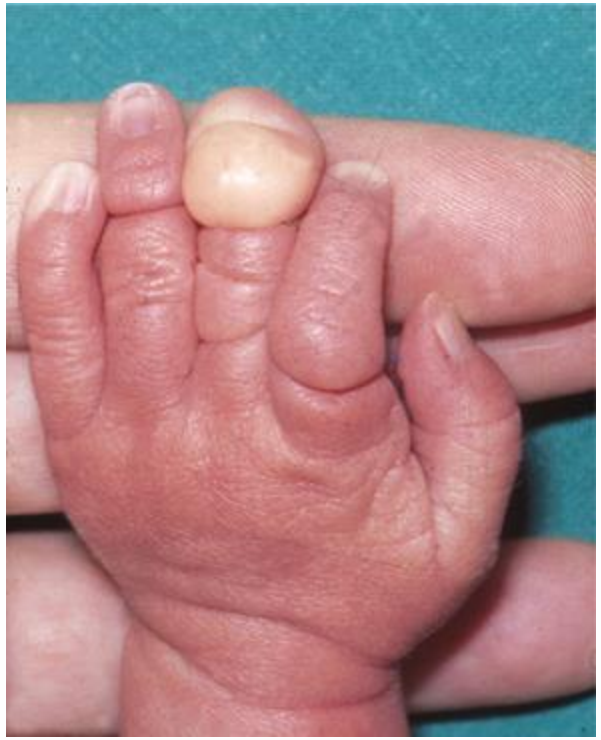
Amniotic band

Learn to differentiate between what is truly important and what can be dealt with at another time.

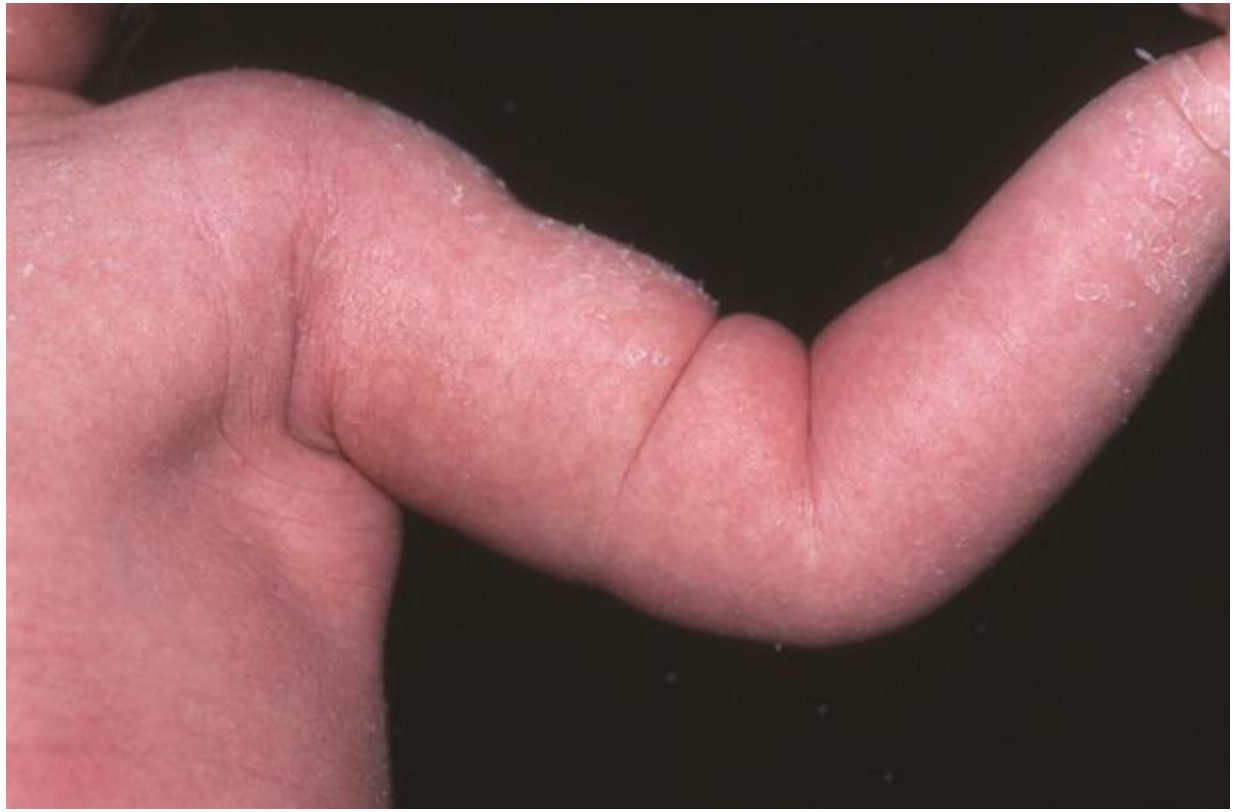
-Mia Hamm

Amniotic Band Syndrome

- Aka constriction band syndrome, aka amniotic constriction sequence
- Characterized by circumferential constriction of limbs or digits
- Etiology ?
  - vascular insult in utero
  - amniotic disruption leads to release of fibrous bands
- Constrictions may be complete or incomplete, ranging from incomplete simple bands to amputations
- Limb viability may be at risk



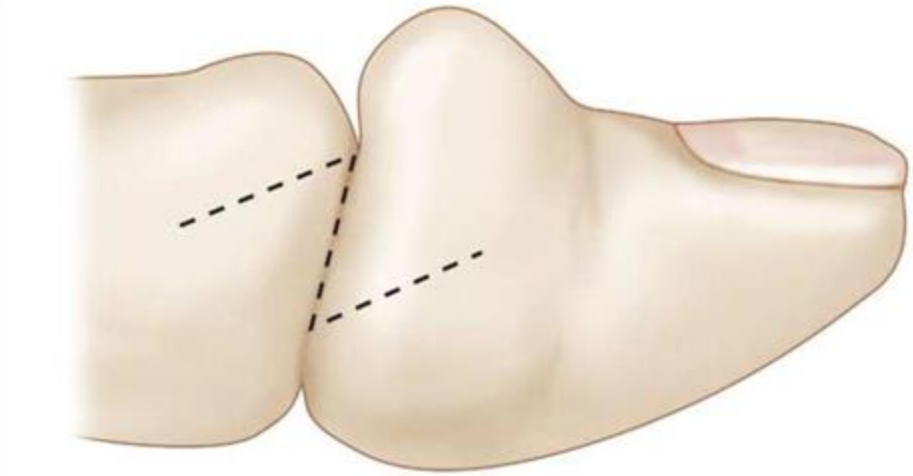
Amniotic Band Syndrome



Amniotic Band Syndrome - Management

- If diagnosed in utero and limb viability is threatened
  - poor prognosis - high rates of amputation
  - can consider fetoscopic vs in utero surgical release

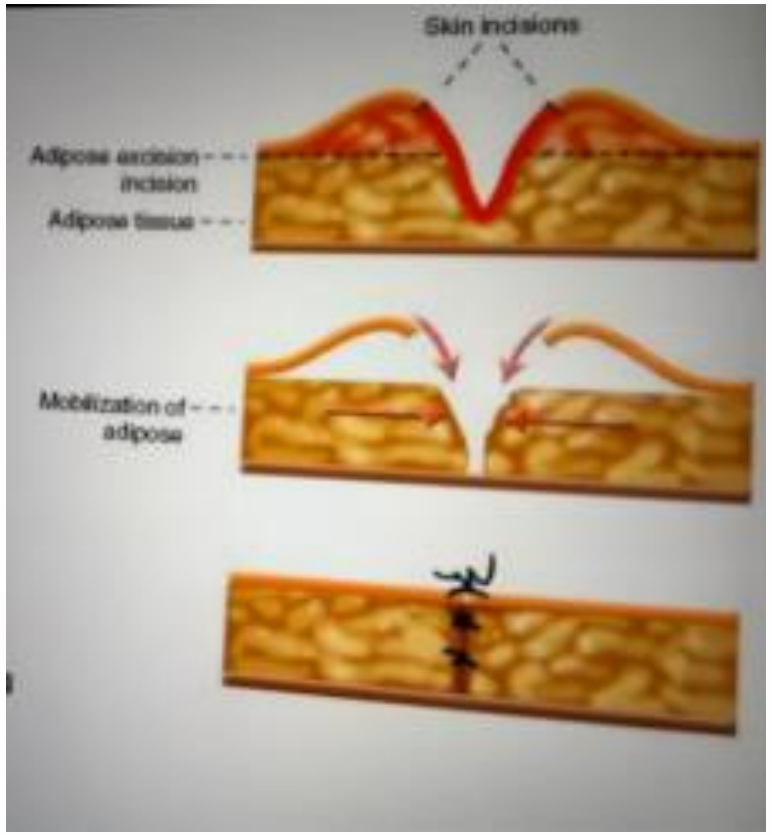
- Non-limb threatening constrictions
  - surgical release to improve function and cosmetic appearance
  - Z- or W-plasty technique



Amniotic Band Syndrome - Management

- If diagnosed in utero and limb viability is threatened
  - poor prognosis - high rates of amputation
  - can consider fetoscopic vs in utero surgical release

- Non-limb threatening constrictions
  - surgical release to improve function and cosmetic appearance
  - Z- or W-plasty technique



dayton children's

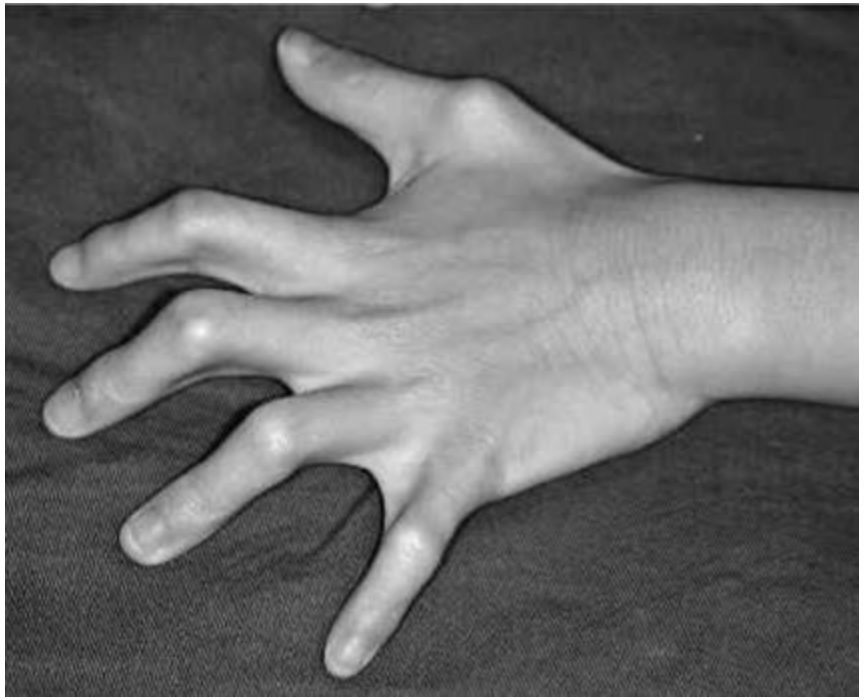
Camptodactyly

About the only thing that comes to us without effort is old age.

-Gloria Pitzer

Camptodactyly

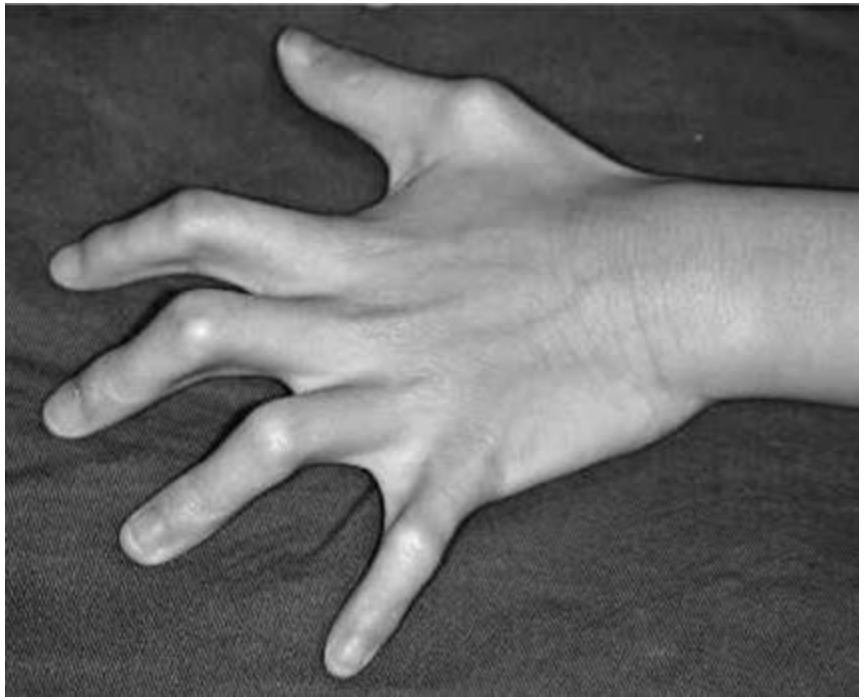
- Characterized by digit flexion deformity/contracture, typically of the PIP joints
  - nontraumatic
- Incidence - <1%
- Demographics
  - 66% bilateral
  - small finger MC



Camptodactyly Management

- Nonsurgical Treatment
  - Splinting and Stretching
  - definitive and first line tx for almost all cases

- Surgery
  - refractory to nonoperative tx and flexion deformity > 30 deg
  - Passive deformity
    - surgical release of involved structures (typicaly FDS)
  - Rigid deformity
    - arthrodesis



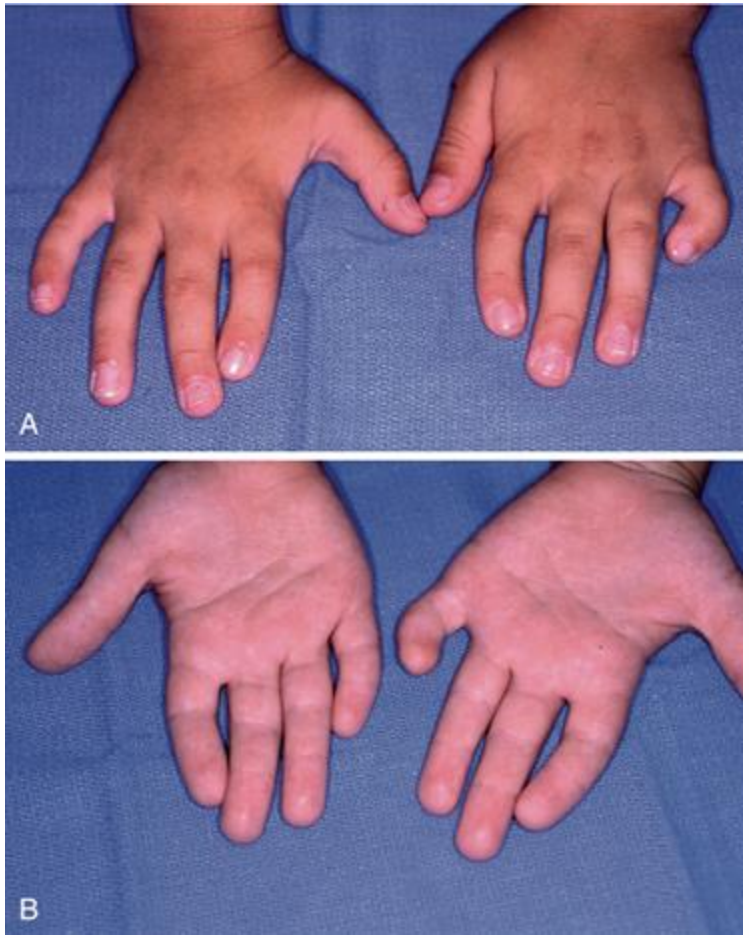
Clinodactyly

Do not let what you cannot do interfere with what you can do.

-John Wooden

Clinodactyly

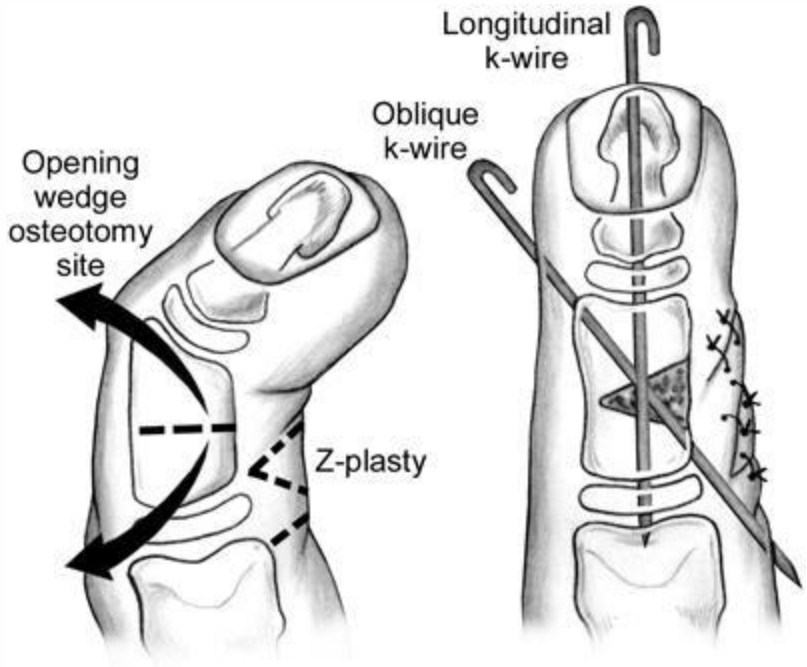
- Characterized by a congenital angulation in a finger in the radioulnar plane > 10°
  - MC presentation is radial angulation of the small finger middle phalanx
- Incidence - 3-19% of the general population
  - 25% of Downs Syndrome patients
- Genetics
  - autosomal dominant
- Typically is nonprogressive



Clinodactyly Management

- Nonsurgical Treatment
  - Observation is first line for almost all cases
  - splinting is not necessary or effective

- Surgery
  - severe curvature causing bony deformity
  - presence of delta phalanx on x-ray



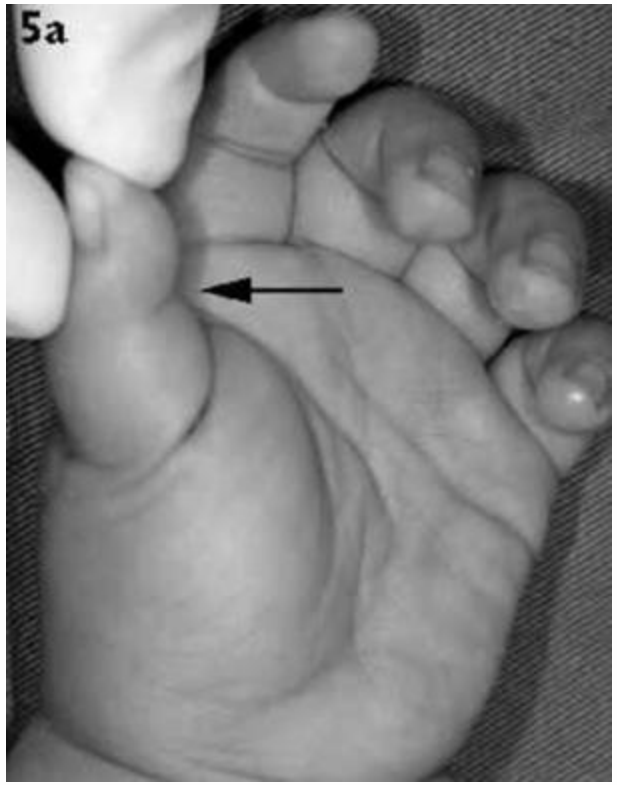
Congenital trigger thumb

Say you were standing with one foot in the oven and one foot in an ice bucket. According to the percentage people, you should be perfectly comfortable.

-Bobby Bragan

Congenital Trigger Thumb

- Presents with a flexion deformity to the thumb IP joint
- Most likely to be acquired within the first 6 months of life, “congenital” may be a misnomer
- Etiology
  - thickening of the flexor tendon
  - Notta node - nodular thickening of the FPL
- Bilateral in 30%
- Other causes of thumb IP joint flexion posturing
  - congenital clasped thumb, absent or aberrant extensor tendons, arthrogryposis, or spasticity



dayton children's

Congenital Trigger Thumb

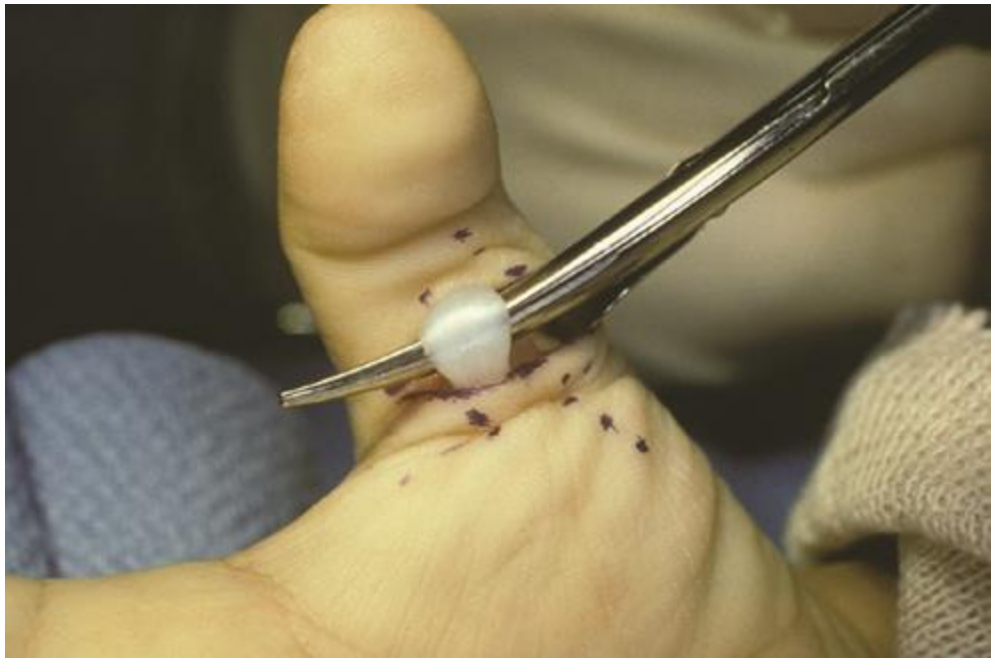
- Cohort of 78 patients with 93 trigger thumbs
- Results:
  - At 5 years from the initial visit, 32% of thumbs had resolved spontaneously
  - 43% elected to proceed to surgery

dayton children's

Congenital Trigger Thumb Management

- Nonsurgical Treatment
  - First line
  - Observation and surveillance
  - Stretching & splinting
    - difficult to maintain, but has been shown to be more effective than observation

- Surgery
  - fails to resolve by age 5
  - to prevent permanent IP flexion deformity, surgical release indicated before age 5



References

1. Mahindroo, Sonal, and Sean Tabaie. "Syndactyly in the Pediatric Population: A Review of the Literature." *Cureus* 15.3 (2023).
2. Bubshait, Dalal K. "A review of polydactyly and its inheritance: Connecting the dots." *Medicine* 101.50 (2022).
3. Ogino T, Ishigaki D, Satake H, Iba K: Free fat graft for congenital hand differences . Clin Orthop Surg. 2012, 4:45-57. 10.4055/cios.2012.4.1.45
4. Kyriazis Z, Kollia P, Grivea I, Stefanou N, Sotiriou S, Dailiana ZH. Polydactyly: Clinical and molecular manifestations. World J Orthop. 2023 Jan 18;14(1):13-22. doi: 10.5312/wjo.v14.i1.13. PMID: 36686282; PMCID: PMC9850794.
5. Light, Terry R. "TREATMENT OF PREAXIAL." *Hand clinics* 8.1 (1992): 161.
6. Oda, Takashi, Allison G. Pushman, and Kevin C. Chung. "Treatment of common congenital hand conditions." *Plastic and reconstructive surgery* 126.3 (2010): 121e.
7. Wolfe, Scott W., et al. *Green's operative hand surgery e-book*. Elsevier Health Sciences, 2021.
8. Ak, Chong. "Common congenital hand conditions." *Singapore Med J* 51.12 (2010): 965.
9. Hutchinson, Douglas T., Ajinkya A. Rane, and Anthony Montanez. "The natural history of pediatric trigger thumb in the United States." *The Journal of Hand Surgery* 46.5 (2021): 424-e1.


Cassidy BSN RN