Congenital Hand Anomalies

Embryology
- Limb buds appear on 26th day
- Differentiation of upper limb into 3 segments (upper arm, forearm & hand) by 30th day
- 6th week – recognizable human form
- 12th week upper limb fully formed
- 14th week lower limb fully formed
- 20th week muscles and nerves developed to allow joint movement
- Congenital malformations begin with embryonal insults between 4th & 6th week, even if genetically determined.
- Genetic deformities are usually autosomal dominant.

Swanson Classification
- Failure of formation
  - Transverse
    - Proximal third of forearm
    - Absence of part of finger = symbrachdactyly
  - Longitudinal
    - Radial or ulnar club hand
    - cleft hand
    - thumb hypoplasia
- Failure of differentiation
  - Syndactyly
  - Acrosyndactyly
  - Camptodactyly
  - Clinodactyly
  - Arthrogryposis
- Duplication
  - Polydactyly
- Overgrowth
  - Macroductyly – stiff and unsightly giant finger; surgery rarely beneficial
- Undergrowth
  - Thumb hypoplasia
- Constriction bands
- Generalised skeletal abnormalities
  - Marfans \(\rightarrow\) spider hand
  - Achondroplasia \(\rightarrow\) trident hand
  - Down’s Syndrome \(\rightarrow\) short, curved little finger
**Radial Club Hand**
- Incomplete or absent radius: 1 in 100,000; 50% bilateral
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- 1 = short, 2 = hypoplastic, 3 = partial absence, 4 = complete absence
- Type 4 is most frequent when this condition exists
- Associations:
  - Absence of radial sided structures (extensors, radial artery, FCR)
  - Thumb hypoplasia
  - TAR syndrome = thrombocytopenia, absent radius
  - VATER = vertebral defects, anal + tracheal, esophageal atresia, renal defects
- Management:
  - Serial casting or external ring fixator to address soft tissue contractures
  - Then centralisation of carpus on ulna with pin stabilisation, providing no elbow extension contracture, and hand can reach mouth.
  - Radial extensor tendon release with transfer to ulnar side
  - Correct any thumb hypoplasia

**Thumb Hypoplasia**
- Varying degrees of intrinsic muscle defects
- 1st web space narrowing
- Ulnar collateral ligament instability
- 1st CMC joint instability → reconstruction not advised
- Treatment:
  - Salvage: web space deepening, UCL reconstruction & tendon transfers
  - Unsalvagable: pollicisation using index finger

**Madelung Deformity**
- Growth arrest of the ulnar and palmar portions of the distal radial physis → overgrowth and dorsal subluxation of ulna.
  - Whole carpus translates volar & ulnar resulting in “uncovering” of the lunate on x-ray.
- Adolescents present with wrist deformity and pain
- Treatment by distal radius joint levelling osteotomies with ulnar shortening.

**Syndactyly**
- Fused digits; 1 in 2000
- Partial or complete, simple (skin webbing) or complex (bone fusion)
- Middle and ring fingers most frequently affected
- Surgery:
  - Usually at 18 months
  - Earlier at 6 months if affecting thumb/index or ring/little fingers, to prevent deformity due to differential bone lengths of these digits.
  - Skin separation ± bone graft from ipsilateral groin

**Acrosyndactyly**
- Only tips of fingers are fused
Camptodactyly
- Flexion deformity of PIP joint
- Usually little finger affected
- Normal DIP joint distinguishes it from a boutonniere deformity
- Hereditary and often bilateral
- Splintage is main treatment.
- Associated with abnormalities of insertion of lumbricals or FDS.

Clinodactyly
- Sideways bent finger – usually little finger
- Delta deformity of middle phalanx, where epiphysis is along one side of the bone
- Treatment – corrective osteotomy

Arthrogryposis
- Intrinsic abnormality of motor nerves and muscles
- Severe weakness of hand with stiff, contracted joints
- Smooth skin
- No flexion creases on hand
- Treatment: splintage, tendon transfers or arthrodesis

Congenital clasped thumb vs. Congenital trigger thumb
- Clasped thumb has weak or absent extensor tendons
  - Requires splintage of extensor tendon transfer
- Triggering caused by stenosing tenovaginitis of FPL
  - Firm nodule over front of MCP joint
  - Spontaneous resolution possible in 1st year
  - Surgical release of pulley at MCP joint if persists.

Polydactyly
- Thumb polydactyly
  - Reconstructio using smaller radial-sided “extra” thumb to augment more robust ulnar-sided thumb.
  - Complication of reconstruction is MCP joint instability
- Little finger polydactyly
  - More common in black newborns
  - More likely to be well developed with a bony skeleton
  - Formal surgical ablation when older
  - If vestigial, may be simply tied off in the nursery
- Central polydactyly
  - Index, middle or ring fingers
  - Associated with syndactyly of extra digit, or cleft hand.

Cleft Hand
- A functional triumph and a social disaster
- V-shaped cleft in central portion of hand ± absence of a ray
- Likely 2° suppression of growth of a central ray in 7th week of development
- Treatment:
  - Closure of cleft with reconstruction of any syndactyly and deepening of 1st web space.