Congenital Disorders of the Hand

Overview and Classification
Classification

- Phocomèle
- Hemimèle
- Ectromèle

Isidore Geoffroy, Saint-Hilaire, 1837
Classification

- Kelikian
- Frantz & O’Rahilly
- Swanson
- IFSSH
Classification

I. Failure of Formation of Parts
II. Failure of Separation of Parts
III. Duplication
IV. Overgrowth
Classification

V. Undergrowth

VI. Congenital Constriction Ring Syndrome

VII. Generalized Skeletal Abnormalities
I. Failure of Formation of Parts

A. Transverse

B. Longitudinal
Longitudinal Absences

- Phocomelia
- Radial Club Hand
- Cleft Hand
- Ulnar Club Hand

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Longitudinal Deficiency

- PHOCOMELIA
- RADIAL (*Preaxial*)
- CENTRAL
- ULNAR (*Postaxial*)
Transverse Absences

Upper Limb

Forearm and Hand

Hand

Digits

Phalanges

Normal

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II. Failure of Differentiation

A. Soft Tissue Involvement
B. Skeletal Involvement
C. Congenital Tumorous Conditions
Syndactyly

Complex

Simple
A. Soft Tissue

1. Disseminated - Arthrogryposis
2. Shoulder
3. Elbow & Forearm
4. Wrist & Hand
   a. Cutaneous syndactyly
   b. Camptodactyly
   c. Thumb-in-palm deformity
   d. Deviated/deformed digits
B. Skeletal

1. Shoulder

2. Elbow - Elbow Synostosis

3. Forearm
   a. Proximal radioulnar synostosis
   b. Distal radioulnar synostosis

4. Wrist & Hand
   a. Osseous syndactyly
   b. Synostosis of carpal bones
   c. Symphalangism
   d. Clinodactyly
C. Tumorous Conditions

1. Hemangiotic
2. Lymphatic
3. Neorogenic
4. Connective Tissue
5. Skeletal
III. Duplication

1. Whole Limb
2. Humeral Segment
3. Ulnar Segment - Mirror Hand
4. Digit - Polydactyly
IV. Overgrowth

1. Whole Limb
2. Partial Limb
3. Digit - Macrodactyly
V. Undergrowth

1. Whole Limb
2. Partial Limb
3. Digit - Brachydactyly
VI. Congenital Constriction Ring Syndrome
VII. Generalized Skeletal Abnormalities
Diagnosis

- Whole body/multiple limb
- Entire Limb
- Too many fingers
- Too few fingers
- Short Fingers
Diagnosis

- Abnormal Thumb
- Too Big
- Bent Fingers
- Rigid Parts
- Combined Parts
- Neuromuscular Abnormalities
Whole Body/Multiple Limbs

- SYNDROMIC
- Apert & Craniofacial Syndromes
- Dwarfism
- Arthrogryposis
Entire Limb

- **Proximal Parts** - *Phocomelia*
- **Unstable/Fused Elbow** - *Ulnar Dysplasia*
- **Radially deviated hand** - *Radial Club Hand*
Too Many Fingers

- Ulnar - Ulnar Polydactyly
- Thumb - Radial Polydactyly
- Central - Central Polydactyly
- Many Fingers/No Thumbs - Mirror Hand
Too Few Fingers

- Single Digit Hand/Ulnar Side - Cleft Hand
- Single Digit Hand - Symbrachydactyly
Too Few Fingers

- Normal structures proximal/Bands/Edema/Other Limbs/Scarring - Amniotic Band
- Normal/underdeveloped proximal structures. Small/absent fingers/Nubbins - Symbrachydactyly
Too Few Fingers

- Absent/Small Thumb - Hypoplastic Thumb
- Combined/Missing Fingers/Deep Central Cleft - Cleft Hand
Too Few Fingers

- Two/Three Fingered Hand; Thumb Present - **Ulnar Dysplasia**
- Stubby, Empty Soft Tissue Sleeves; Fingers Partially or Totally Absent - **Symbrachydactyly**
Short Fingers

- Brachydactyly
- Brachymetacarpia
- Brachyphalangia
- Brachysyndactyly
Abnormal Thumb

- Small Thumb - Hypoplastic Thumb
- Segmented Thumb - Radial polydactyly
- Finger like thumb - Triphalangeal Thumb
Abnormal Thumb

- Angled Thumb - **Delta Phalanx**
- Thumb in same plane as fingers/too few fingers - **Ulnar Dysplasia**
- Thumb in same plane/Many fingers - **Mirror Hand**
Abnormal Thumb

- Tight Thumb - Trigger Digit, Spastic Hand/Arthrogryposis/Clasped Thumb
- Long Thumb - Triphalangeal Thumb/Radial polydactyly
Too Big

- Entire Limb - Hemihypertrophy
- Single Finger - Ollier, Klippel-Trenaunay-Weber/Vascular malformations/Hamartomatas
- Combined Digital Parts - Cleft Hands
Bent Fingers

- Flexed - Trigger, Camptodactyly, arthrogryposis
- Small Fingertip/Adolescent/Progressive - Kirner’s Deformity
- Angled - Delta Phalanx/Clinodactyly
Rigid Parts

- Rigid Forearm/Lack of Motion - Radioulnar Synostosis
- Rigid Elbow - Radiohumeral Synostosis/Ulnar Hemimelia
- Limited Elbow Motion - Congenital Radial Head Dislocation
Rigid Parts

- **Digit Stiff in Extension** - *Symphalangism*

- **Digit Stiff in Flexion** - *Contractures, Camptodactyly, Arthrogryposis*
Combined Parts

- Webbed Fingers - Syndactyly
- Unsegmented Finger Joints - Symphalangism, Brachysyndactyly
- Combined Digital Skeleton - Cleft Hand
- Lack of forearm Motion - RU Synostosis
Neuromuscular Abnormalities

- **Generalized Weakness** - Congenital Myopathies, Cerebral Palsy
- **Spasticity** - Encephalopathy, Cerebral Palsy
- **Intrinsic Muscle Weakness**/Tightness