Congenital Hand Malformations

Plastic & Reconstructive Surgery
Post Graduate Course, April 2001

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History
History

- 1634 Ambroise Pare
- 1832 St. Hilaire – 1st classification
- 1847-1849 Velpeau, Vrolik
- 1892 Felizet – Surgery for Syndactyly
- 1960’s Thalidomide
- 1968 Swanson, Brsky, Ethin
Embryology

C-R: 13mm  18mm  30.7mm
Embryology

Mechanism of Anomalies
I. Failure of formation of parts (arrest of development)

A. Transverse deficiencies
   1. Amputations: arm, forearm, wrist, hand, digits

B. Longitudinal deficiencies
   1. Phocomelia: complete, proximal, distal
   2. Radial deficiencies (radial club hand)
   3. Central deficiencies (cleft hand)
   4. Ulnar deficiencies (ulnar club hand)
   5. Hypoplastic digits
II. Failure of differentiation (separation) of parts

A. Synostosis: elbow, forearm, wrist, metacarpals, phalanges
B. Radial head dislocation
C. Symphalangism
D. Syndactyly
   1. Simple
   2. Complex
   3. Associated syndrome

E. Contracture
   1. Soft tissue
      a) Arthrogryposis
      b) Pterygium cubitale
      c) Trigger digit
      d) Absent extensor tendons
      e) Hypoplastic thumb
      f) Thumb-clutched hand
      g) Camptodactyly
      h) Windblown hand
   2. Skeletal
      a) Clinodactyly
      b) Kirner's deformity
      c) Delta bone

III. Duplication

A. Whole limb.
B. Humeral segment
C. Radial segment
D. Ulnar segment- mirror hand
E. Digit
   Polydactyly- Radial (preaxial), Central, Ulnar (postaxial)
F. Epiphyseal
IV. Overgrowth
A. Whole limb
B. Partial limb
C. Digit
   1) Macrodactyly
      a) With associated vascular conditions
      b) with neurofibromatosis
      c) With bone or cartilage exostosis
      d) other

V. Undergrowth
A. Whole limb
B. Whole hand
C. Metacarpal
D. Digit
   1. Brachysyndactyly
   2. Brachydactyly
Classification (Swanson)

VI. Congenital constriction band syndrome
   A. Focal necrosis
   B. Amputation (intrauterine)

VII. Generalized skeletal abnormalities
   A. Chromosomal
   B. Madelung deformity

Classification (Swanson)

I. Failure of formation of parts (arrest of development)
II. Failure of differentiation (separation) of parts
III. Duplication
IV. Overgrowth
V. Undergrowth
VI. Congenital constriction band syndrome
VII. Generalized skeletal abnormalities
How Common Are Upper Limb Anomalies?

### Table 129-3. Relative Frequency (Percentage) of Anomalies Seen in Four Hand Clinics*

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Yokohama</th>
<th>Iowa</th>
<th>Hong Kong</th>
<th>Bangkok</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hand deficiencies</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar deficiency</td>
<td>0.01</td>
<td>0.11</td>
<td>0.02</td>
<td>0.01</td>
</tr>
<tr>
<td>Transverse deficiency</td>
<td>0.05</td>
<td>0.14</td>
<td>0.13</td>
<td>0.03</td>
</tr>
<tr>
<td>Camptodactyly (flexion deformity)</td>
<td>4.0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Constriction ring</td>
<td>0.1</td>
<td>0.11</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Note that in the foreign studies, duplications were not included. (Data summarized from Goldberg, M. J., and Bartoshesky, L. E.: Congenital hand anomaly: Etiology and associated malformations. Hand Clin., 7:409, 1995.)*

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Treatment Goal

Functional Independence & Esthetical acceptance
Non Surgical Treatment

Splinting
Light, Progressive- towards functional position.

Occupational Therapy
teaching of movement patterns

Physiotherapy
Passive and active range of motion

Parents Education

Psychological support

Timing of Surgery

Early
• Tethering of adjacent structures- Irreversible angulation of skeleton
• Better and earlier correct brain development
• Anatomic adaptation of reconstructed part with growth and external forces
• Psychology (child & parents)

Late
• Larger hand
• More precise operation
• More adequate functional evaluation
• Eye-hand coordination not fully developed until age 2-3

Other
• Other deformities and general condition
• Surgeon’s experience and preferences
Specific Anomalies

Polydactyly

**Thumb Duplication** (pre-axial polydactyly)
Incidence: ~ 1/10,000. Race:B=W
Genetic: Sporadic, AD, AR

**Post Axial**
Commonest congenital anomaly
Incidence: 1/300-1/3000. Race:B>W
Genetic: AD, Sporadic

**Central**
rare
Usually with syndactyly
Polydactyly

Duplicated Digit
Type I- Extra soft tissue only
Type II- Bone, tendon and cartilage
Type III- Completely developed with own metacarpal (rare)

Surgical Principles
• Type I: Simple amputation
• Type II:
  • Choose the component to amputate by observing the child play
  • Conservation of usable structures. Extensor or flexor tendons from ablated segment may be used to reinforce remaining tendons or redirect balance forces
  • Accurate establishment of the longitudinal skeletal axis
• Type III: Ray Amputation
Polydactyly
Duplicated thumb

Incidence = 1:1000 births
Look for Cardiac anomalies & Fanconi anaemia.

Thumb Polydactyly
Principles of Operative Management:

• The goal is Opposable thumb with stable pinch (especially important: UCL of MCP)
• Accurate establishment of the longitudinal skeletal axis
• Reduction in number of elements

Usually Esthetic Problem
Thumb Polydactyly

Principles of Operative Management

• Choose the component to amputate by observing the child play

• Conservation of usable structures. Extensor or flexor tendons from ablated segment may be used to reinforce remaining tendons or redirect balance forces
Syndactyly

**Complete** or **Incomplete**: (complete= entire length of digit is affected)

**Simple** (skin only) **Complex** (bone involvement) or **complicated** (bizarre disorganization)

**Syndactyly**

Incidence = 1:2000 births

Can be associated with many anomalies:

**Craniofacial syndromes**: Apert Syndrome (acrocephalosyndactyly) Carpenter, Pfeiffer, Mobius, Pierre-Robin

**Chromosomal**: Trisomy 13, 14, 21, Partial trisomy 10q, Triploidy syndrome.

**Other syndromes**: Cornelia de Lange, Frazer, Poland
Syndactyly

PreOp Examination:
• Third space (foot-second) most frequently involved.
• Differential movement between digits
• Shape and contour of nails
• Individual Allen's tests to each involved digit
• Radiographs to rule out tethering of digits

PreOp Planning:
• Pierce, ligate, divide webs - not acceptable
• Tissue expanders - high complication rate
• One side of digit should be operated on at a time, in order to avoid vascular insufficiency
• With multiple syndactyly attention is first directed to border digits
• The usual strategy is to preserve as much regional skin as possible on the radial side of the involved digits (to maximize pinch sensation) and saving FTSG for ulnar side of the affected digit
• Very early operation - no need for skin graft
• Vickers - no skin graft, extensive defatening
Syndactyly

PreOp Planning:

- Flap for commissure, proximally based, volar/dorsal
- Scars planned to minimize late contractures
- Full thickness grafts for exposed areas
- Timing - before 6 months if complex or when growth will be impaired by uneven fingers. Otherwise at least one year.
- Planning - three dimensional reconstruction. Web wider distally, slope 35-45° proximo- dorsal to disto- volar. Circumference of two combined digits smaller than two separate ones.
- Multiple webs - minimize number of operations

Syndactyly

PreOp Planning:

- Skin incisions
  - Dorsal flap for web avoid scar at the commissure.
  - Reciprocal dorsal & palmar flaps that will interdigitate with each other, usually skin grafts are needed at the base of the fingers.
- Keep/ reconstruct finger tips
- Reconstruct nail margins
Syndactyly

Complications & pitfalls

Digital necrosis - Operate one side of a finger at a time, check vasculature when in doubt

Graft loss - Prevent shearing or local infection

Web creep - Planning of scar, loss of palmar position

Nail deformity - Lack of support, tension of flaps, abnormal growth
Constricting Ring Syndrome

- Wide range of clinical presentation
- Treatment varies with severity of deformity, number of functional digits and presence or absence of thumb

Main principle – Excision of annular groove and Z-plasty
Trigger Digit

Congenital stenosis at A1 pulley. Often bilateral. **30% may resolve spontaneously by 1 year.** Attempt splinting but **aim to correct surgically by 3 years.**

Hypoplastic Thumb

<table>
<thead>
<tr>
<th>Blauth Grade</th>
<th>Description</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Short thumb, hypoplastic thenar muscles</td>
<td>Augment intrinsics</td>
</tr>
<tr>
<td>II</td>
<td>Grade I + Adducted MPJ</td>
<td>Soft tissue Z-plasty</td>
</tr>
<tr>
<td>III</td>
<td>Deficient metacarpal Abducted thumb</td>
<td>Augment/ bone graft/pollicisation</td>
</tr>
<tr>
<td>IV</td>
<td>Floating thumb</td>
<td>Pollicisation</td>
</tr>
<tr>
<td>V</td>
<td>Absent thumb</td>
<td>Pollicisation</td>
</tr>
</tbody>
</table>
Hypoplastic Thumb

Policization

Radial clubhand

• Absence or hypoplasia of pre-axial structures:
  • radius
  • radial carpus (scaphoid, trapezium, trapezoid)
  • Thumb - may be floppy (pouce flottante) or absent
• May occur in association with visceral anomalies (CVS/GI/GU), VATER or blood dyscrasias e.g. Fanconi’s syndrome.
• Most common in the right hand
• Bilateral in 50% of cases.
**Radial clubhand**

<table>
<thead>
<tr>
<th>Type</th>
<th>Features</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Short</td>
<td>address thumb</td>
</tr>
<tr>
<td>II</td>
<td>hypoplastic</td>
<td>individualised</td>
</tr>
<tr>
<td>III</td>
<td>partially absent</td>
<td>centralisation</td>
</tr>
<tr>
<td>IV</td>
<td>totally absent</td>
<td>centralisation</td>
</tr>
</tbody>
</table>

![Diagram of normal and radial clubhand types](image)
Radial clubhand

- Treatment is difficult - mild cases - strapping & manipulation;
- severe cases - correction of wrist deformity (Centralisation) by fusing ulna to carpus or circular frame & provision of a thumb (Pollicisation) using Buck-Gramko method (transfer, shortening & rotation of index finger).
- Abnormal distribution of nerves & arteries must be appreciated prior to surgery.

Ulnar Clubhand

Note the difference between this anradial club hand. In ulnar club hand the corpus remains articulated to the distal radius.
**Ulnar Clubhand**

This is not associated with systemic disorders as is the radial club hand but can be associated with other musculoskeletal deformities, especially hand deformities. Treatment may include conversion to a one bone forearm.

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**Central Deficiency**

(cleft hand / lobster claw hand / ectrodactyly)

Central absense of at least one digit other digits may fuse (syndactyly)

Typical Deformity = Autosomal Dominant; often bilateral; more common in boys & may involve the feet

Atypical Deformity = sporadic; associated with cardiac & GIT abnormalities
Phocomelia

Phocomelia presents a problem primarily to the prothetist and engineer, and the surgeon rarely intervenes (Aitkin & Franz, 1960).

Deviated Digits

**Camptodactyly**
- Familial soft tissue abnormality with deviation in the sagittal plane. Commonly involves the little finger causing a flexion contracture at the PIPJ.
- May be ass. with Dupuytren's, Marfan's, arthrogryposis or other genetic syndromes.
- Few good surgical procedures exist.
- 2 stage correction (with ex-fix applied at the time of FDS extensor transfer) has good results.

**Clinodactyly**
Skeletal abnormality causes deviation in the lateral plane. Usually involves the little finger and is caused by a trapezoidal middle phalanx. Surgical correction for cosmesis only.