Treatment of congenital syndactyly

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Plan

• Generality
  • Classification
  • Epidemiology

• Indication and timing of surgery

• Principle of surgery
  • Patient’s setup
  • Commissure reconstruction
  • Digital separation
  • Resurfacing
  • Paronychial reconstruction
  • Post-op care

• Specific situations

• Complications

• Conclusion

→ Exclude from this presentation:
  • Symbrachydactyly
  • Amniotic band syndrome
Swanson / International Federation of Societies for Hand Surgery Classification - 1983

<table>
<thead>
<tr>
<th>Main Category</th>
<th>Subcategory</th>
<th>Diagnosis (Example)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Failure of formation (arrest)</td>
<td>Transverse longitudinal</td>
<td>Radial club</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cleft hand (typical/atypical)</td>
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<tr>
<td></td>
<td></td>
<td>Phocomelia</td>
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<tr>
<td>II. Failure of differentiation (separation)</td>
<td>Soft tissue</td>
<td>Arthrogryposis</td>
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<tr>
<td></td>
<td>Skeletal</td>
<td>Cutaneous syndactyly</td>
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<tr>
<td></td>
<td>Tumorous</td>
<td>Camptodactyly</td>
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<tr>
<td></td>
<td></td>
<td>Radioulnar synostosis</td>
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<td></td>
<td></td>
<td>Osseous syndactyly</td>
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<tr>
<td></td>
<td></td>
<td>Clinodactyly</td>
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<tr>
<td>III. Duplication</td>
<td>—</td>
<td>Mirror hand</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Polydactyly</td>
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<tr>
<td>IV. Overgrowth (gigantism)</td>
<td>—</td>
<td>Hemihypertrophy</td>
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<tr>
<td></td>
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<td>Macroductyly</td>
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<tr>
<td>V. Undergrowth (hypoplasia)</td>
<td>—</td>
<td>Brachysyndactyly</td>
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<tr>
<td></td>
<td></td>
<td>Brachydactyly</td>
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<tr>
<td>VI. Constriction band syndrome</td>
<td>Focal</td>
<td>Constriction band</td>
</tr>
<tr>
<td></td>
<td>Amputation</td>
<td>Acrosyndactyly</td>
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<tr>
<td></td>
<td></td>
<td>Intrauterine amputation</td>
</tr>
<tr>
<td>VII. Generalized</td>
<td>—</td>
<td>Achondroplasia</td>
</tr>
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<td></td>
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<td>Marfan's syndrome</td>
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</tbody>
</table>

Oberg-Manske-Tonkin classification - 2013

### 1) MALFORMATIONS

<table>
<thead>
<tr>
<th>a. Défauts de formation/différenciation axial de l'ensemble du membre supérieur</th>
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</thead>
<tbody>
<tr>
<td>I. axe proximo-distal</td>
</tr>
<tr>
<td>* brachymélie avec brachydactylie</td>
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<tr>
<td>* symbrachydactylie</td>
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<tr>
<td>* déficience transverse: amélie,</td>
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<tr>
<td>pseudarthrose claviculaire,</td>
</tr>
<tr>
<td>allongement/raccourcissement au-</td>
</tr>
<tr>
<td>dessus/au-dessous du coude, polignet,</td>
</tr>
<tr>
<td>1ère/2è rangée du carpe, metacarpe,</td>
</tr>
<tr>
<td>phalange proximale/intermédiaire/distale</td>
</tr>
<tr>
<td>* déficience intersegmentaire: phocomélie (totale/proximale/distale)</td>
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</table>

### 2) DEFORMATIONS

<table>
<thead>
<tr>
<th>a. Hypertrophie</th>
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</thead>
<tbody>
<tr>
<td>I. Totalité du membre:</td>
</tr>
<tr>
<td>* Maladie amniotique</td>
</tr>
<tr>
<td>* doigts à ressaut</td>
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</table>

### 3) DYSPLASIES

<table>
<thead>
<tr>
<th>a. Tumeurs</th>
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</thead>
<tbody>
<tr>
<td>I. Vasculaire:</td>
</tr>
<tr>
<td>* macrodactylie</td>
</tr>
<tr>
<td>* muscles de la main aberrants</td>
</tr>
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</table>

### 4) SYNDROMES

Epidemiology

• Incidence = 1 in 2000 to 3000 live births
• One of the most common congenital hand deformities
• 2/1 ratios in favor of males and of the Caucasian population
• 10% to 40% of patients have a family history of previous similar malformations
Epidemiology

• In isolated syndactyly:
  • both hands are involved in 50% of cases
  • The 3rd (50%) and 4th interdigital (30%) spaces are the most commonly affected

• In syndromic syndactyly: the 1st and 2nd web spaces are relatively more frequently involved

General examination

Assess:
• The whole affected limb and:
• Other limbs
• Chest wall
• Craniofacial areas
  → syndromic concomitant malformations
  (Poland, Apert’s syndrome...)
  → genetic counseling
Classification

• Complete or incomplete

• Simple, complex and complicated


Indications of surgery

- **Surgical management is indicated in most cases**
- Surgery is not indicated in complex cases without sufficient osteo-articular tissues to provide stable and mobile independent digits, since separation may worsen hand function

Timing of surgery

• Remains controversial

• Surgical program should end before three years of age

• Syndactyly between rays of different lengths induces angular deformities of the longer digit
  • Result in fixed camptodactyly and/or clinodactyly
  • $1^{st}$, $2^{nd}$ and $4^{th}$ interdigital spaces = before one year
  • $3^{rd}$ interdigital space = between 12 and 18 months


Timing of surgery

• Most authors advocate **staged separation surgeries when a finger is webbed on both sides**

• Syndactyly of all interdigital spaces will need two procedures:
  • first release of the first and third webs
  • and 3 to 6 months later the release of the second and fourth webs

Timing of surgery

• Some authors recently reported on single-staged release of all fingers
• Using dorsal gull-wing flaps
• Preoperative CT angiography to demonstrated at least one digital artery in all fingers

→ All were simple syndactyly


Principles of surgery

1. Patient’s setup
2. Commissure reconstruction
3. Digital separation
4. Resurfacing
5. Paronychial reconstruction
6. Post-op care
Principles of surgery

1. Patient’s setup
2. Commissure reconstruction
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Patient’s setup

- Ambulatory surgery
- General anesthesia
- Sterile tourniquet for access to the arm and elbow for skin graft harvest if necessary
- Optical magnification
- Anatomical landmarks using a skin-marker pencil:
  - metacarpal heads
  - longitudinal midlines of each digit
  - relation between dorsal and palmar flaps
- Measurements of proximal thickness of the conjoined digit
Principles of surgery

1. Patient’s setup
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Comissure reconstruction

Use of skin flaps is widely accepted

Second, third and fourth normal web spaces present with a 45° dorso-palmar slope

Commissure reconstruction: Incomplete syndactyly

- Three-flaps web plasties: Several designs have proposed over the years (MOSS 1990; OSTROWSKI 1991 BANDOH 1997...).
- If webbing is proximal to the proximal phalanx distal epiphysis: island flap and translate it proximally as in “V-Y”-plasty.

![Diagram showing three-flaps and island flaps for commissure reconstruction](image-url)
Ostrowski’s plasty

Commissure reconstruction: Complete syndactyly

Dorsal flaps


Palmar flaps

Combined dorsal and palmar flaps
Commissure reconstruction

• **Proximally-based dorsal flap** is the most commonly used technique
  • Starting proximally at the level of the metacarpal heads
  • Flap length is equal to the antero-posterior thickness of conjoined digits
  • About 1/2 ratio
  • Anastomosis digital artery and dorsal intermetacarpal artery
Dorsal flaps: Numerous shapes (pentagonal, hexagonal, bilobed, double wing-shaped, seagull wing–shaped...)

Proximally-based flaps

Island flaps

Commissure reconstruction

• Proximally-based dorsal flap: Numerous variations have been proposed:
  • hourglass flaps (GLICENSTEIN 1998, MALLET 2013)
  • trapezoid flaps (VEKRIS 2010)

Commissure reconstruction

- Proximally-based dorsal flap: Numerous variations have been proposed:
  - Dorsal omega flaps (DARCANGELO 1996) with palmar anchor

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Digital separation

- Matched or non matched broken-line incisions to elevate palmar and dorsal laterally-based interdigitating flaps
  - Avoid retractile scar formation
  - Avoid web creep
- Anticipate coverage of zone of interest (i.e., exposed bone or joint)

Digital separation

• Precautious dissection
  • from distal to proximal
  • from dorsal to palmar

• Release all fibrous connections

• Identify the neurovascular structures
  • If the division of the common interdigital artery is too distal, one of the two branches may be ligated
Digital separation

• Identify the neurovascular structures
  • provided that the contralateral digital artery is known to be intact
    • clamping test ++
    • Nerve: intraneural dissection
• Once all flaps are positioned tourniquet is released for hemostasis before grafting
Principles of surgery

1. Patient’s setup
2. Commissure reconstruction
3. Digital separation
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5. Paronychial reconstruction
6. Post-op care
Resurfacing - Controversy: with or without skin graft?

Most of the recent studies described satisfactory results with graftless procedures:

- Reduce operating time
- No donor site morbidity

Resurfacing is achieved through flap positioning and defatting, as well as leaving open the residual defects.


## Resurfacing: Controversy with or without skin graft?

- Recent prospective comparative study or retrospective comparative study = advantages in appearance and web creep in the group with skin graft

<table>
<thead>
<tr>
<th>Author</th>
<th>Date</th>
<th>Journal</th>
<th>Number of syndactylies</th>
<th>Skin graft</th>
<th>Mean follow-up Years)</th>
<th>Level of Evidence</th>
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<tbody>
<tr>
<td>Landi</td>
<td>2014</td>
<td>JHS Eur</td>
<td>26</td>
<td>Hyalomatrix</td>
<td>2</td>
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<td>Ni</td>
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<td>4.2</td>
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<tr>
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<td>16</td>
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<td>Sullivan</td>
<td>2017</td>
<td>JHS Am</td>
<td>Systematic review : 34 articles</td>
<td>Comparative graft/no graft</td>
<td>N/A</td>
<td>Therapeutic IV</td>
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<tr>
<td>Tian</td>
<td>2017</td>
<td>JHS Am</td>
<td>74</td>
<td>No graft in 21 out of 31 patients</td>
<td>1</td>
<td>Therapeutic IV</td>
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<tr>
<td>Dong</td>
<td>2017</td>
<td>Medicine (Baltimore)</td>
<td>35</td>
<td>No</td>
<td>4.6</td>
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<td>Yuan</td>
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<td>PRS</td>
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<td>JHS Am</td>
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<td>1 to 2</td>
<td>Therapeutic IV</td>
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<tr>
<td>Wang</td>
<td>2019</td>
<td>JHS Eur</td>
<td>28</td>
<td>Comparative graft/no graft</td>
<td>4.3</td>
<td>Therapeutic II</td>
</tr>
</tbody>
</table>

Failed graftless with Hyalomatrix
Donor site

• Full-thickness grafts:
  • Anterior Elbow crease
  • Wrist flexor crease
  • Medial part of the arm
  • Groin area
  • Retroauricular area
  • Abdomen...

→ Should match skin color

Principles of surgery

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5. Paronychial reconstruction
6. Post-op care
Paronychial reconstruction

- Complex syndactyly
  - fused phalangeal tufts
  - bony elements are exposed at the end of the release

- Pulp flaps:
  - 1: buck gramcko
  - 2: sugihara
  - 3: lundkvist
Paronychial reconstruction

Two laterally-based long and narrow triangular flaps
Principles of surgery

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Post op care

- Surgical wounds are closed without tension using 5.0 absorbable sutures
- The newly separated web space is maintained as opened as possible without compromising flap and/or digit vascularization
  - Most authors recommend to use only gauze to open to web space
- First dressing change is made 7 to 21 days after surgery

Post op care

• In complicated syndactyly, the newly separated web space can be maintained opened with an external fixator when needed.
Specific situations
Specific situations: First web space syndactyly

• Mild syndactyly:
  • “Z”-plasty
  • Four-flaps “Z”-plasty ++
• Dorsal advancement rotation flap
• Preliminary tissue expansion on the dorsum of the hand
• Transposition flap from the radial border of the index
• Severe contracture = free groin, forearm or lateral arm flap
First web space syndactyly: large advancement-rotation flap raised from the dorsum of the hand


Ghani HA. Modified dorsal rotation advancement flap for release of the thumb web space. J Hand Surg Br 2006;31:226-9
Complicated syndactyly
Polysyndactyly

• Can be associated with:
  • Clinodactyly
  • Brachydactyly
  • Symphalangism
  • Synostosis

• Ultrasound or magnetic resonance imaging may precise the tendon and neurovascular anatomies

• Problem with IP stiffness, joint instability, flexors/extensors tendon insertions....

Syndromes
Apert’s syndrome

- Autosomic dominant and mutations
  - Mutation of the fibroblast growth factor receptor type 2 gene (FGFR2)
- Severe complex syndactyly of the hands and feet
  - Complex syndactyly of the index, long, and ring fingers
  - Simple syndactyly between the ring and small finger
  - Incomplete first web space syndactyly
  - Radial clinodactyly of the thumb
- Abnormalities of the shoulder and elbow
- Craniostenosis, Mid facial hypoplasia, Hypertelorism
Apert’s Syndrome

• The goal of surgery is to complete separation of the digits and correct the thumb deformity before 2 years of age
  • preop soft tissue expansion (Lohmeyer)
• Reconstruction of an adequate first web space = first priority
• Thumb clinodactyly = corrective osteotomy (opening wedge)
• Release of the fourth and fifth metacarpal synostosis between the ages of 4 and 6 years

Poland’s Syndrome

- Attributed to disruption of the blood flow in the subclavian artery in the embryo
- Sternocostal head of pectoralis major agenesis
- Brachymésophalangy or une symbrachydactyly
  - Always unilateral
  - Smaller hand
  - Stiff fingers
  - Clinodactyly
Poland’s Syndrome

• Associated anomalies:
  • Hypoplasia of breast
  • Costal hypoplasia
  • Lung hernia
  • Scapula hypoplasia
  • Short axillary fold
  • Radioulnar synostosis
Complications
Complications

- Digital ischemia
- Flap necrosis
- Graft loss
- Wound dehiscence
- Delayed healing
- Superficial surgical site infections (most common)

→ about 2% of cases

Complications

- Web creep = distal translation of the commissure
- Palmar scar contractures with growth-related frontal and/or sagittal angulations
- Joint motion limitation
- Nail dystrophy
- Keloid formation
→ complexity of the malformation++

### Complications: Web-creep

<table>
<thead>
<tr>
<th>Flexion-extension deformity</th>
<th>Normal digit</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal digit</td>
</tr>
<tr>
<td>1</td>
<td>Finger can not be hyperextended</td>
</tr>
<tr>
<td>2</td>
<td>Reductible flexion deformity</td>
</tr>
<tr>
<td>3</td>
<td>Fixed finger deformity</td>
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</table>

<table>
<thead>
<tr>
<th>Web-creep</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Soft web. Abduction mirrors the adjacent web or equivalent controlateral web</td>
</tr>
<tr>
<td>1</td>
<td>No web advancement but thickening of the web with reduced span</td>
</tr>
<tr>
<td>2</td>
<td>Creep of web to 1/3 of the distance between base of the web and PIPJ crease</td>
</tr>
<tr>
<td>3</td>
<td>Creep of web to 2/3 of the distance between base of the web and PIPJ crease</td>
</tr>
<tr>
<td>4</td>
<td>Creep of web to the PIPJ crease</td>
</tr>
</tbody>
</table>

Palmar scar contracture and web creep
Results

- Dorsal Omega flap
- Palmar anchor
- Skin graft

Conclusion

• One of the most common deformity of the hand congenital deformity
• Initial examination must focus on the determining the syndromic or isolated
• Numerous techniques have been described
  • very little comparative data exists
• The physician has the choice regarding the technique to use
• Outcomes are most commonly satisfactory, provided that the syndactyly is simple and that basic operating rules are followed
Thank You

Thanks to Adeline Cambon-Binder for the drawings