

Decision Making in Congenital Thumb Hypoplasia

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Thumb Hypoplasia can occur as a part of a broader spectrum of Radial longitudinal deficiencies or as an isolated anomaly. The thumb is very important for normal functionality of the human hand hence reconstruction of the hypoplastic thumb is one of the most satisfying operations in Hand Surgery. This write up will concentrate on the classification and management of a Thumb Hypoplasia that occurs as a congenital anomaly.

Syndromes or Associations With Radial Longitudinal Deficiency

Syndrome or Association	Characteristics
Holt-Oram syndrome	Heart defects, most commonly cardiac septal defects
Thrombocytopenia–Absent Radius (TAR)syndrome	Thrombocytopenia present at birth (may require transfusions) but improves over time. Thumb is always present
VACTERL association	Vertebral abnormalities, Anal atresia, Cardiac abnormalities, Tracheoesophageal fistula, Esophageal atresia, Renal defects, Radial dysplasia, Lower limb abnormalities
Fanconi anemia	Aplastic anemia not present at birth; develops at about 6 years of age
CHARGE syndrome	Coloboma of the eye, Heart defects, Atresia of the nasal choanae, Retardation of growth and/or development, Genital and/or urinary abnormalities, and Ear abnormalities and deafness

Diagnosis:

Depending on the presence or absence of any co-existing anomalies, the degree of deficiency on the affected side, thumb hypoplasia may be recognised early. When thumb hypoplasia is associated with profound radial deficiency diagnosis is made shortly after birth because the child's forearm is short and the hand deviated in a radial direction. Severe thumb hypoplasia is also recognized early on because there is marked asymmetry between the thumbs. Mild hypoplasia may go un-noticed for many years.

Diagnosis is made after physical examination findings and appropriate X-Rays. The presence or absence of Intrinsic and Extrinsic muscles as well as stability of the MCP and CMC joints of the affected thumb greatly determine the plan of reconstruction. Hence, assessment of these form an integral part of assessment of a child with thumb hypoplasia.

Classification and Treatment:

The *Blauth*¹ classification provides a good guideline for management.

Thumb Hypoplasia and Treatment Plan

Type	Findings	Treatment
I	Minor generalized hypoplasia	No treatment
II	Absence of intrinsic thenar muscles First web space narrowing Ulnar collateral ligament (UCL) insufficiency	Opponensplasty First-web release UCL reconstruction
III	Similar findings as Type II plus: Extrinsic muscle and tendon abnormalities Skeletal deficiency A: Stable carpometacarpal joint B: Unstable carpometacarpal joint	A: Reconstruction B: Mostly Pollicization
IV	Pouce flottant, or Floating thumb	Pollicization
V	Absence	Pollicization

In Blauth's classification, stability of the thumb CMC joint determines labelling the thumb as type A or B in type III hypoplasia and tilts the treatment option in favour of pollicisation instead of reconstructing the existing thumb if the CMC joint is unstable.

Timing of Surgery

We prefer to do the surgery around one and a half years of age. The aim is to have the child adapt and learn to use the reconstructed thumb for prehensile activities by the time he/she has to start going to school.

Type I

The thumb may look slim but no treatment is necessary.

Type II and III A hypoplasia has distinct findings that guide management. There is varying degrees of thenar muscle deficiency, thumb web narrowing and laxity of the Ulnar Collateral Ligament (UCL) of the MCP joint.

Reconstruction in type II and IIIA hypoplasia requires managing all the deficient elements. The narrowed thumb/index finger web space, MCP instability, and thenar muscle absence all require treatment. Thumb/index finger web space narrowing is treated with skin rearrangement and soft tissue release. A four-flap “Z”-plasty lengthens the tight skin and provides a rounded contour to the web space. An extremely tight web space may also require partial release of the adductor muscle and/or first dorsal interosseous muscle to obtain adequate breadth.

Augmenting/providing metacarpophalangeal (MCP) joint stability may require strengthening or reconstruction of the Ulnar Collateral Ligament using locally available soft tissue or tendon graft.

The main surgical options for providing opposition in a hypoplastic thumb include transfer of Abductor Digiti Minimi (ADM) or Flexor Digitorum Superficialis (FDS) of ring finger. The use of the ADM was first described by **Huber** in the 1920s and has been suggested to be superior to the FDS transfer². In addition, it has been suggested that the appearance of the thenar eminence is also improved after an ADM opposition transfer.³

Technique of Huber Transfer: A midlateral incision is made on the ulnar border of the little finger proximal phalanx and extended proximally and radially to the distal

palmar crease. It then runs along the radial border of the hypothenar eminence and curves ulnarly as it crosses the distal wrist crease (Fig 1).



Fig 1: Skin markings for Modified Huber's Transfer in a case of Type 2 Thumb Hypoplasia.

The two ADM insertions (i.e., base of the proximal phalanx and extensor apparatus) are divided, and this muscle is then freed of soft tissue attachments by retrograde dissection toward its pisiform origin. When mobilizing the proximal portion of the ADM, great care must be taken to not damage its thin neurovascular pedicle, which is on its dorsoradial aspect.

Alternatively, this pedicle can be identified by exposing the ulnar nerve and artery proximally at the wrist and tracing them distally. Once the neurovascular bundle has been isolated, the transfer's length is increased by elevating the abductor origin from the pisiform while carefully retaining an attachment on the FCU tendon by dissecting a tendinous slip proximally. The transfer's only remaining soft tissue attachments are then its neurovascular pedicle and the FCU tendon. Next, a dorsoradial incision is made over the thumb MP joint and a wide subcutaneous tunnel is created between this incision and the area immediately proximal to the pisiform; this is easier if a third skin incision is made in the thenar crease at the base of the thenar eminence.

The ADM muscle is then turned through 180 degrees on its long axis to reduce the tension on its neurovascular bundle (as if turning the page of a book) [Fig 2], passed through the subcutaneous tunnel, and attached to the APB insertion. Because the

ADM muscle has only sufficient length to just reach the APB insertion, it is invariably attached under adequate tension. Postoperatively, the thumb is immobilized in a cast in full opposition for 4 weeks. The position of the wrist is not critical inasmuch as the transfer does not cross this joint.



Fig 2: Diagrammatic representation of the principle of Huber's transfer.



Fig 3: Immediate post-operative position and appearance of the thumb after a Modified Huber's transfer.



Fig 4: Post-operative follow up picture showing the thumb abduction.

Patients *without* a stable carpometacarpal joint, **those with type IIIB, IV, or V thumb hypoplasia**, are typically treated with pollicization.⁴⁻⁶

In case of a **type IIIB hypoplasia**, it may be difficult to get the parents to consent for a removal of the hypoplastic thumb and then Index Pollicization. In such cases, reconstruction using Metatarsal (including the head) or Iliac Crest graft has been done.⁷ Also, reconstruction using the non-vascularised toe phalanx transfer has been described.^{8,9} This reconstruction can entail multiple procedures after bony reconstruction, including Tendon Transfers/Grafting for FPL and EPL as well as Opponensplasty. Moreover, functionally, such a reconstructed thumb is almost always inferior to a well done pollicization. We would discuss the options with the parents and then choose the technique of reconstruction for each child.

Index Pollicization provides for a satisfactory reconstruction of the Thumb that is severely hypoplastic (type IIIB, IV, or V thumb hypoplasia).¹⁰ Pollicization provides a mobile and stable CMC joint to the new thumb and hence associated with better range of movement and resultant function. This is a technically demanding operation and there are many technical modifications described. Pollicization surgery as such does not need microsurgery (repair of the small vessels) however, microsurgical expertise is essential to dissect the small vessels in a child and to transfer the index finger with its intact vascularity and sensations to the position of the thumb. Magnification is highly

desirable especially when the operation is done in younger children. Detailed here is the technique practiced and preferred by the author.

Surgical Steps:

The Incision must allow for the Index transposition, metacarpal shortening, Intrinsic Muscle transfers and Thumb web reconstruction.

- Surgery is performed under appropriate Tourniquet control.
- Magnification is essential
- There are quite a few skin incisions described, author prefers the one proposed by Ezaki and Carter
- Palmar skin is incised and the radial neurovascular bundle is isolated
- Dissection proceeds in an ulnar direction to identify the common digital vessels to the index finger/long finger web space
- Proper digital nerves to the ulnar side of the index finger and the radial side of the long finger are isolated
- Proper digital artery to the long finger is ligated with a ligature clip
- First annular pulley of the index finger is incised
- Divide intermetacarpal ligament between index finger and long finger
- Dorsal skin incision is sharply elevated with preservation of dorsal veins
- The first dorsal and palmar interossei muscles are released with a portion of the hood
- Metacarpal index finger is shortened by a saw cut at the metaphyseal flare and a knife cut through the physis leading to an epiphysiodesis
- Index metacarpophalangeal joint (MCP) is fixed into hyperextension using nonabsorbable suture material passed through the epiphysis and dorsal capsule. This corrects the natural tendency of hyperextension at the index MCP joint.

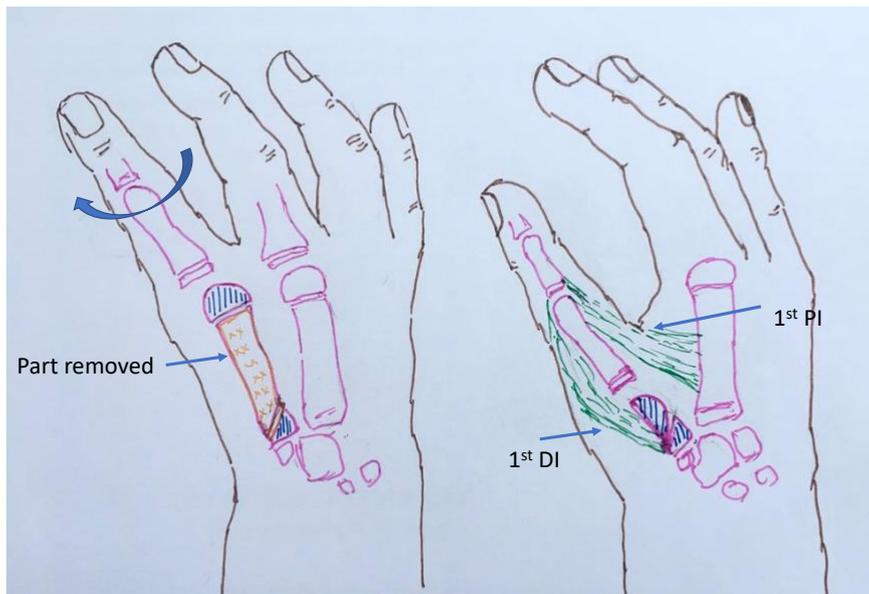


Figure 5: Scheme of Pollicization

- Kirschner wire drilled antegrade through the metacarpal epiphysis, into the proximal phalanx, and out the proximal interphalangeal joint
- Kirschner wire used as a joystick to align the index metacarpal into 45 degrees of abduction and 100 to 120 degrees of pronation
- Kirschner wire is drilled retrograde across the metacarpal base into the carpus
- Transfer the first dorsal interosseous to the radial lateral band and the first palmar interosseous to the ulnar lateral band about the PIP joint.
- So, the index finger dorsal interossei now becomes the 'new' abductor pollicis brevis and index palmar interossei becomes abductor pollicis brevis. The extensor digitorum of index becomes abductor pollicis longus and extensor indicis proprius would function as extensor pollicis longus of the 'newly' created thumb.
- Some surgeons may prefer to place sutures to take up any slack in the extensor tendon. However, the author has not found the need to do this either for the extensor or for the flexor tendon.
- Skin is closed with resection of any redundant skin and Kirschner wire is cut short.



Fig. 6: Type 4 Thumb Hypoplasia treated by Pollicization of the index finger



Post Operatively, a spica cast is applied. K wire is removed at 4 weeks and a forearm based thermoplastic splint is to be used for about 3 months after surgery.

Therapy is started with an aim on thumb usage. The initial goal is large object acquisition followed by acquisition of smaller objects and eventually fine pinch.

Pitfalls and tricks to avoid them:

1. Wrongly placed skin incisions can lead to difficulty in closure after the repositioning. This can be avoided by taking care about how the incisions are planned
2. Inappropriate Index Metacarpal Osteotomies can lead to longer than normal appearance of the reconstructed thumb. It is important to place the distal osteotomy (Usually this can be 'knifed' thorough the growth plate at this level) just subcapital.
3. Positioning the metacarpal head in full extension while fixing in the new location is important to have the right flexion/extension range in the reconstructed 'CMC' joint.

Conclusion:

Thumb Hypoplasia is a commonly encountered condition in Hand Surgery practice. Various treatment options are available and will have to be carefully chosen for each child.

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