THUMB DUPLICATION

AUTHORS:

Dr. Mithun Pai

MS (Ortho); FNB (Hand & Microsurgery)

Assistant Professor, KMC, Manipal

Dr. Madhusudhan NC

DNB (Ortho); FNB (Hand & Microsurgery)

Consultant Hand Surgeon, St. Philomena's Hospital, Bangalore

Introduction

Thumb duplication or preaxial polydactyly is the most prevalent congenital anomaly of hand, with an incidence between 0.8 per 1,000 live births to 1 per 3000 live births[1]. Commonly being described as "duplication" the presentation can vary from a vestigial skin tag to a complete splitting type. However, it is important to understand that the two thumbs are not duplicated with equal size and function, rather neither is 'normal'. More commonly, one thumb is more anatomically and functionally developed than the other, leading to the introduction of the term ''split thumb''. Radial polydactyly originally was classified as a ''duplication'' by the International Federation of Societies for the Surgery of the Hand. It was thereafter reclassified as "malformation,'' which is a failure of axis formation, and/or differentiation of the radioulnar hand plate as described by Oberg, Manske, and Tonkin[2]. They classified anomalies based on developmental biology and pathogenesis rather than on morphologic features and described that, processes such as formation and differentiation occur together and not independently.

The duplicated thumb usually has both the sensory and motor units that work in concert with the entire hand, leading to minimal functional disability. Duplication alone is typically unilateral and sporadic, with only 20 percent being bilateral (Fig 1); however, duplication in the setting of a triphalangeal thumb is inherited in an autosomal dominant pattern[1].

Fig 1: Clinical picture depicting bilateral thumb duplication.

Embryology

The Zone of Polarizing Activity (ZPA) is an area of mesenchyme that contains signals that guide limb bud development. The duplication of thumb results from the failure of the hand plate to form or differentiate in the radioulnar axis. The sonic hedgehog protein expressed in ZPA promotes radio-ulnar differentiation. Abnormal expression of which along with other genes such as hox genes, bone morphogenic protein, and GLI3 gene are known to be involved in thumb duplication[2].

`

Classification

Wassel's classification (Fig 2 & 3) (Wassel-flat) is essential in evaluation and management of thumb duplication, which is based on the duplication level (phalanx/metacarpal) and the location (bone/joint). However, this classification curtails the limitation of assessing only the radiographic component of the duplication [3]. Intra-operative findings are often variable and might show cartilage fusion which is not visible in patient's radiographic images prior to gaining skeletal maturity. In addition to this, tendon abnormalities, disproportionate articular surfaces, and capsuloligamentous laxity add to the complexity.

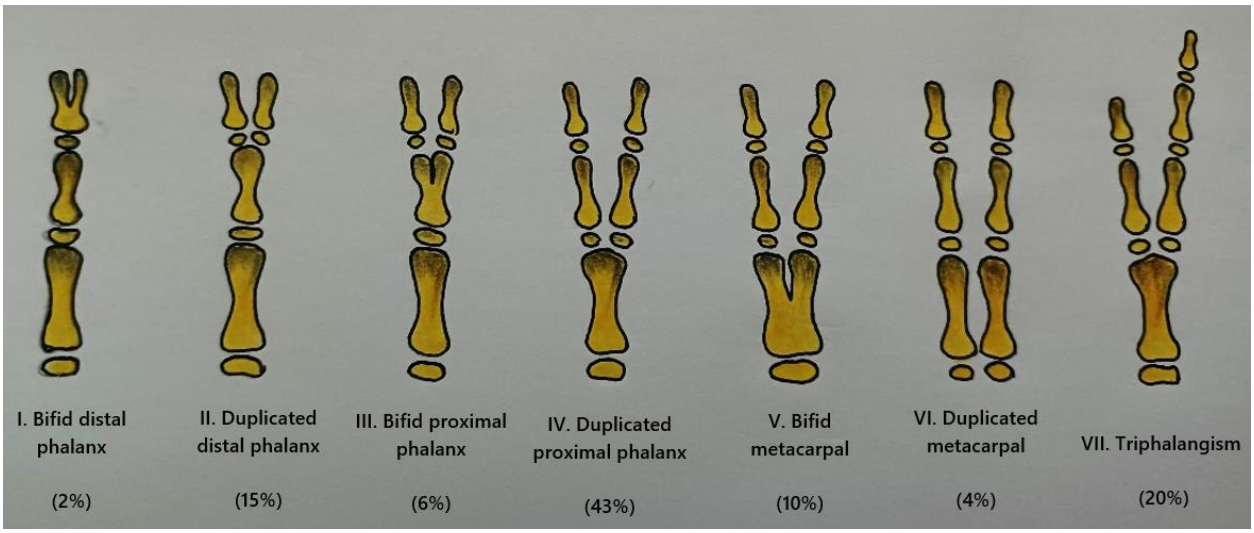

Fig 2: The classical Wassel's classification, Type IV being the most common type

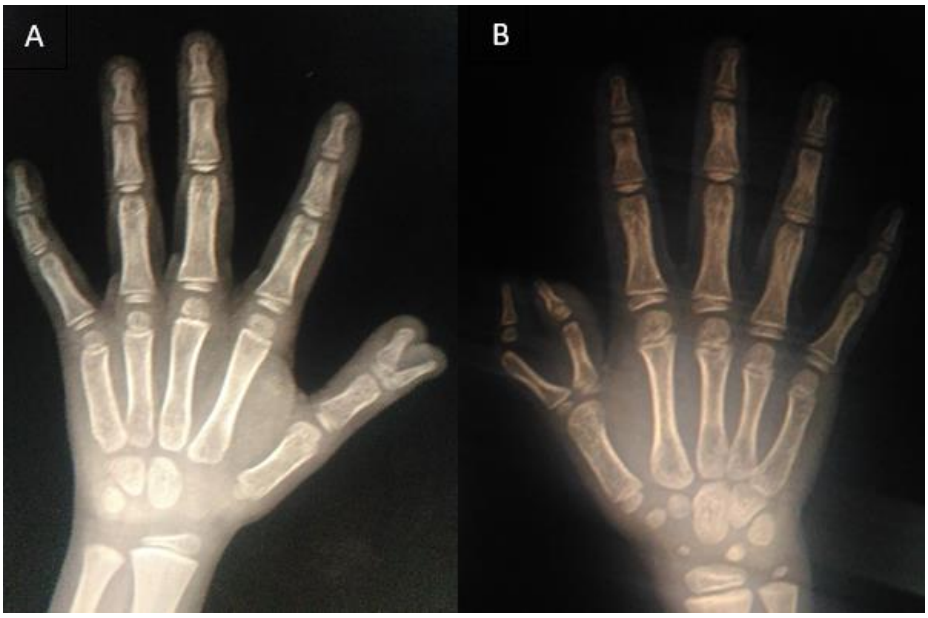

Fig 3: X ray showing type 2 (A) and type 4 (B) Wassel thumb

This was followed by a modification that aimed at including triphalagism and triplication in the early subdivisions of classification popularly known as **Rotterdam classification,** introduced in 2008 by Zuidam et al[4] (Fig 4).

Type I- Partial duplication distal phalanx
Type II- Complete duplication distal phalanx
Type III- Partial duplication proximal phalanx
Type **IV**- Complete duplication proximal phalanx
(Tph) Triphalangeal components
(T) Triplication
Type V- Partial duplication of metacarpal
Type **VI**- Complete duplication of metacarpal
(Tph) Triphalangeal components
(T) Triplication
Type VII- Partial duplication of carpals
Type VIII- Complete duplication of carpals

(***Marked in bold denotes the changes from the original classification***)

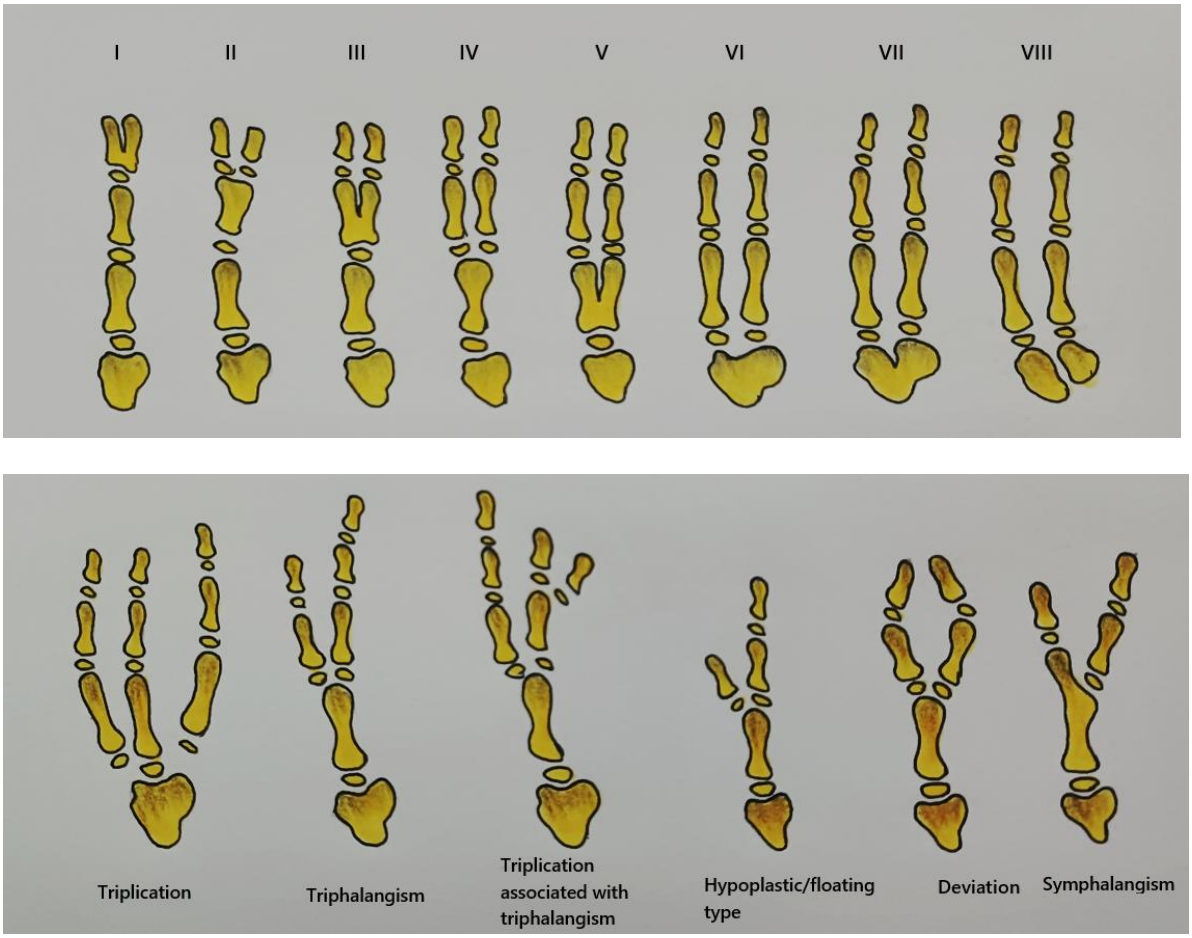

Fig 4: *The Rotterdam classification introducing triphalangism and triplication in the early subdivisions.*

Chung et al.[5] in 2013 recognized the deficiency in intraoperative management, which did not correlate with radiological features. An alternative classification was proposed (Fig 5) based on the anatomic and morphologic features of the origin of the duplicated digit to guide in making surgical decision and to assess clinical outcomes.

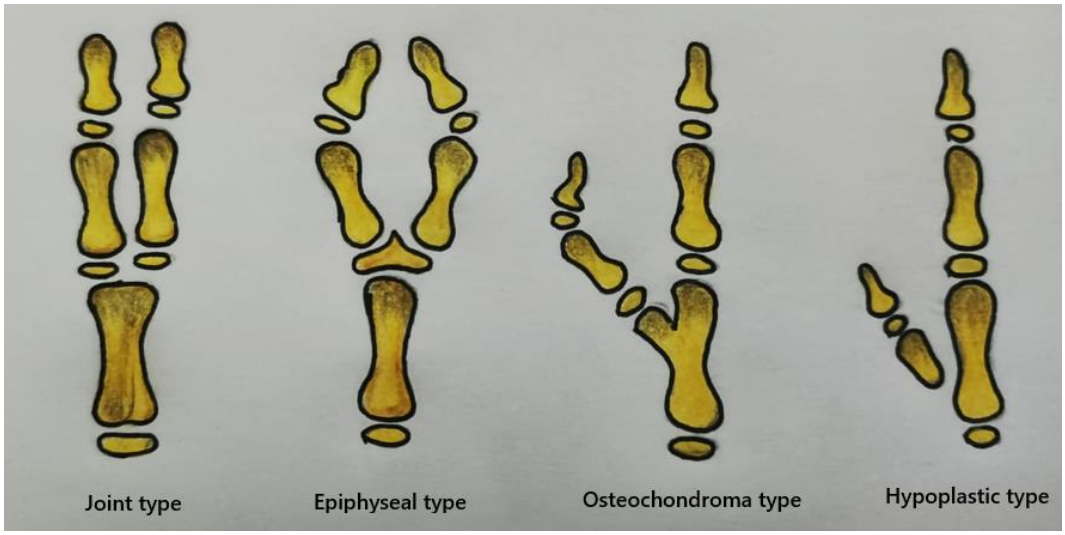

Fig 5: The Chung classification based on anatomic morphological features.

He described type 1 as the joint type where the extra digits have an independent articular surface, type 2 as the epiphyseal type where the extra digits share the epiphysis, type 3 as osteochondroma type where the origin of extra digit resembles the osteochondroma, and type 4 as hypo plastic-type where the extra digit is a mere soft tissue.

However, these classification systems are still not widely used because it is too complex to easily apply in the clinical practice (Zuidam et al.,2008) and unfamiliarity among the surgeons who have used the Wassel-Flatt classification system for a long time (Chung et al., 2013). Recently, two clinical classifications have been described to negate the shortcomings of Wassel's classification and aid in decision-making for surgical management.

Wang in 2021[6], described a classification system considering the alignment, size & shape, and development, which would help guide treatment and would be a helpful supplement for Wassel's classification. According to Wang the thumb duplication can be categorized as (Fig 6):

Type I- A well-developed thumb with nail width greater than or equal to 70% of that of the contralateral thumb.
Type IA Normal **alignment** of the main thumb
Type IB Abnormal **alignmen**t of the main thumb
Type II- Hypoplasia in both thumbs
Type IIA- The duplicated thumbs are equal in **size and shape**
Type IIB- The duplicated thumbs are unequal in **size and shape**
Type III- **Development** of the thumbs
IIIA- Each duplicate thumb is well-developed
IIIB- Stunted, polyploidy, and ulnar dementia etc.

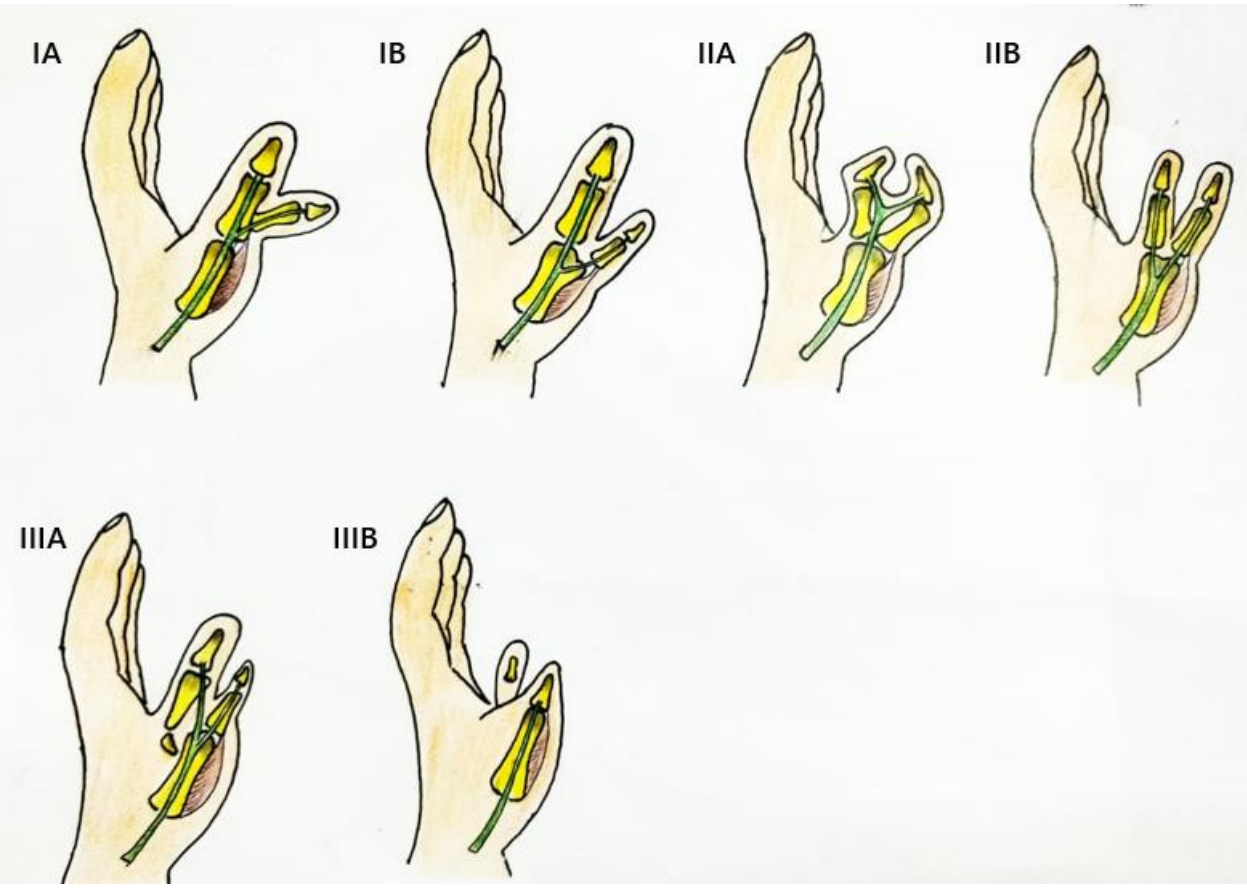

Fig 6: Schematic representation of classification described by Wang.

In the same year, Kim et al. (2021)[7] included the hypoplastic types and refined the definition of triphalangeal thumb without changing the main structure of the Wassel-Flatt classification system. He divided hypoplastic-type into hypoplastic distal type, where the extra digit is located at the distal phalanx level, and the hypoplastic proximal type where the extra digit is located proximal to the interphalangeal joint level. The thumb was classified as the triphalangeal type when the triphalangeal component exists on both side digits (Type VIIA) or the main digit (Type VIIB).

Pre-operative assessment

The thumb's range of motion, the status of thenar muscles, and the adequacy of the first web space are the main determinants of favorable surgical outcomes. Mobility is largely determined by the integrity of the carpometacarpal (CMC) joint, which is reasonably normal in more distal duplication. The thenar musculature may be hypoplastic with a narrower first web space in more proximal duplications.

One must also be familiar with the location of the tendons and their functionality. The flexor pollicis longus (FPL) and extensor pollicis longus (EPL), which are the workhorse for the thumb motion may be present in both thumbs[1]. However, the more functional thumb usually demonstrates better tendon quality and active range of motion. These tendons are notorious for having eccentric insertions, which can lead to distal convergence of the duplicated thumbs (Fig 7).

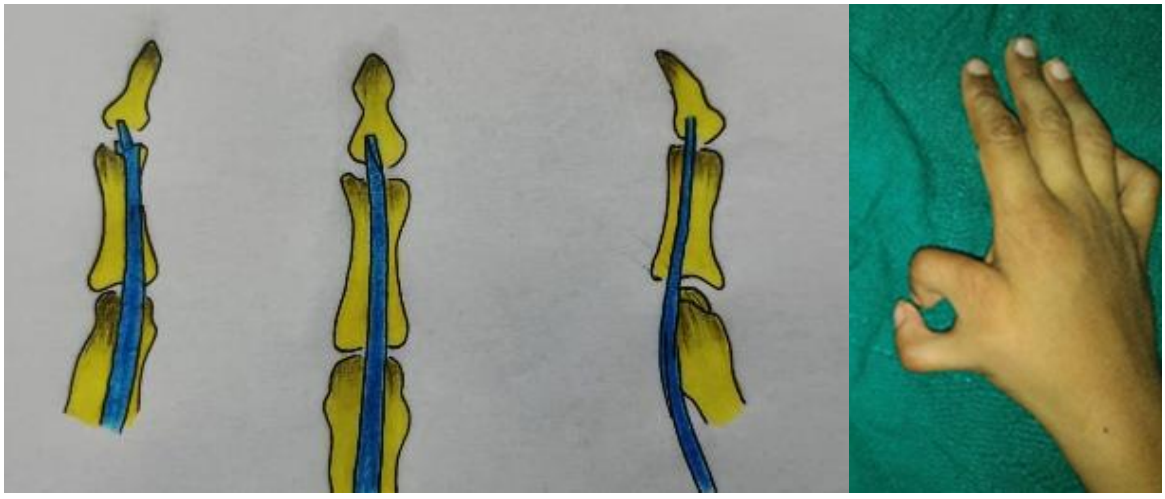

Fig 7: Schematic diagrams depicting dynamic deforming forces created by eccentric insertion of terminal tendon leading to divergent-convergent thumbs.

The better-developed thumb is typically known to arise from the ulnar side and is the larger of the two in the majority of the cases. The duplicated thumbs usually have smaller nails and pulp than the contralateral side. It is also of utmost importance to compare the affected hand with the supposedly unaffected side as bilateral involvement should alert the examiner to a possible syndrome, such as Townes-Brocks syndrome[1] (Imperforate anus, abnormally shaped ears, and hand malformations that most often affect the thumbs).

In addition, in cases in which there is considerable IP joint stiffness and abduction of the thumb, the presence of a pollex abductus (Fig 8) should be considered. This abnormal connection between the flexor pollicis longus (FPL) and extensor pollicis longus (EPL) tendons has been reported in up to 20% of thumb duplications[2].

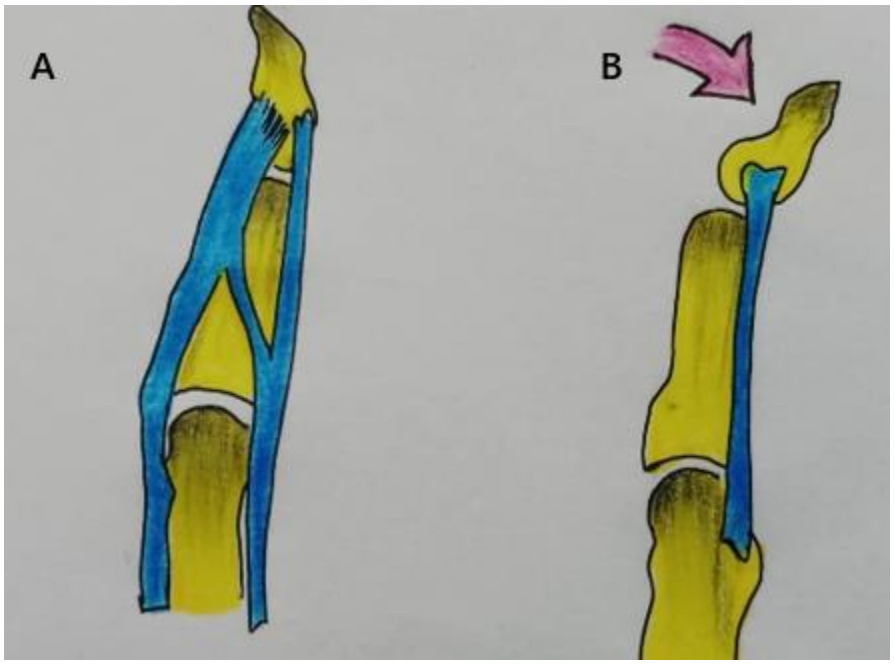

*Fig 8: Schematic diagram of the pollex abductus as seen from the radial **(A)** and dorsal **(B)** aspect. Note the resultant force acting on the IP joint.*

Coming to vascular supply Kitayama and Tsukada[8] have shown a digital artery to each thumb in 74% of cases. In 12% of cases, there were two arteries to the ulnar and one artery to the radial thumb. Two digital arteries supply each thumb in 9% of cases. However, it is essential to note that in 5 % of cases, there is a single digital artery typically to the more ulnar thumb. Hence caution is needed when excising the hypoplastic ulnar thumb.

Radiography usually provides adequate information regarding the pattern of involvement and is performed after one year of age or just before surgery. An X-ray of the opposite thumb is beneficial in comparing bone size and shape and assists in assessing joint deformity.

Saito et al. (2021)[9] introduced three-dimensional ultrasonography, which guided exploration of anatomy at the surgery to explain the forces that create the deformities in the ulnar of the two thumbs of a Type VI thumb duplication as a part of the abnormal insertion of thenar muscles and the extrinsic tendons.

Ultrasonography is proved to be beneficial in defining these anomalies, diminishing the extent of unwanted dissection. MRI is also known to have a similar role but is not used routinely due to anesthetic requirement.

Timing of surgery

Tip-to-tip pinch does not develop until up to 15 months of age[1]. Surgery may be safely and appropriately performed at 9 to 12 months of age[2]. The main goals of reconstruction are an improved appearance, stable, mobile, straight thumb, and the restoration and maintenance of function and pinch activity.

Surgical management

The index procedure should address all elements of the deformity if at all possible. This includes skin, extrinsic tendon, intrinsic muscle rebalancing, and longitudinal bone and joint realignment and stabilization. Simple ablative surgery alone in these cases can result in instability and malalignment of the remaining thumb[1].

Excision (Simple ablation)

Indications

1. Attachment only by a small soft tissue pedicle (Fig 9).
2. There is no bony connection between two thumbs.
3. The dominant thumb has good stability, range of motion, and appearance.

Despite the hypoplastic appearance of the common radial nubbin, the presence of a vascular pedicle within the soft tissue stalk is always a possibility, which should be identified and cauterized during ablation.

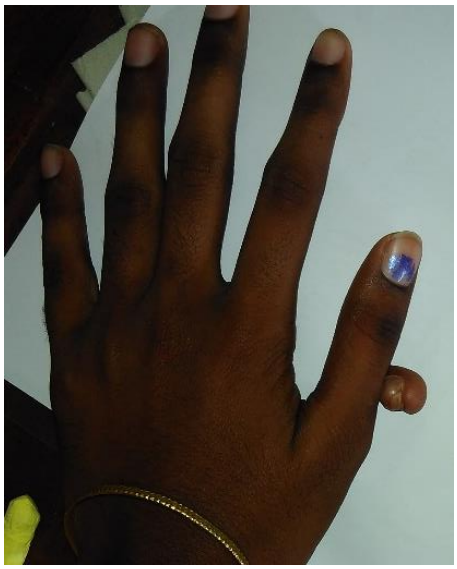

Fig 9: Clinical picture depicting skin tag attachment type of thumb duplication

Reconstruction

Indications

1. The unequal development of the thumb (Fig 10).
2. Bony or cartilaginous connection.
3. Share the same joint.

Fig 10: Clinical picture depicting unequal thumb sharing same joint.

Reconstructive Options

a) Arthroplasty of the MCP/IP joint raises the ligament periosteal flap and reconstructs the stable collateral ligament.

b) Partial head excision for unequal joint surface.
c) Corrective wedge osteotomy for angular deformity. (Fig 11)
d) Realignment of tendon insertion (FPL/EPL) in convergent-divergent deformity. (Fig 12)

Different skin incisions have been described based on the type of duplication, few of which are depicted in fig. 13

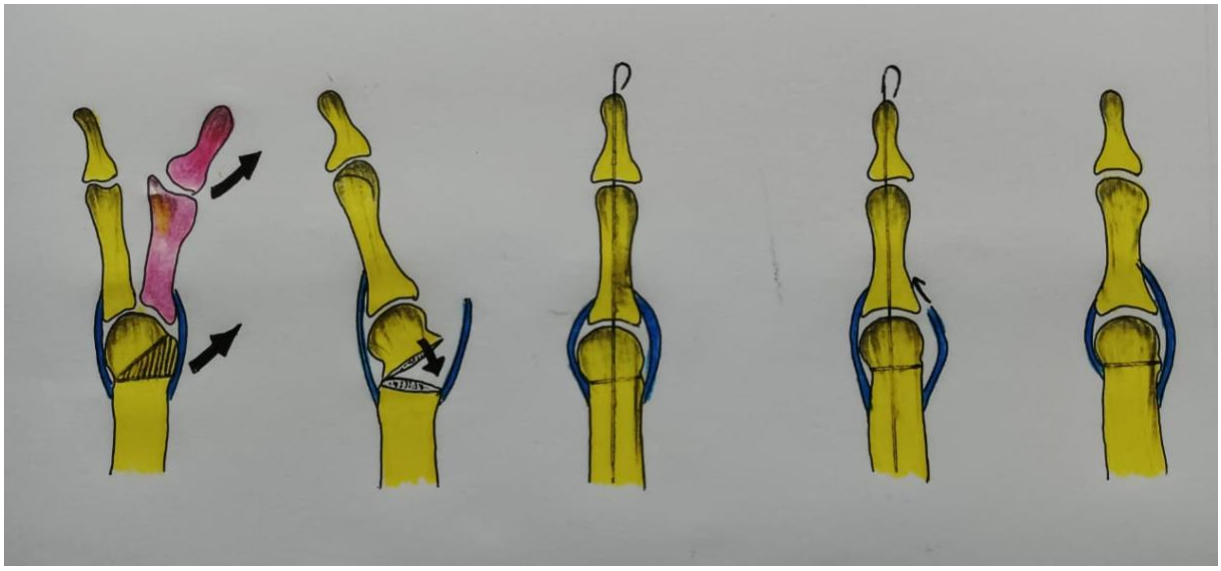

Fig 11: Schematic representation of corrective wedge osteotomy and ligamento-periosteal flap elevation and collateral reconstruction.

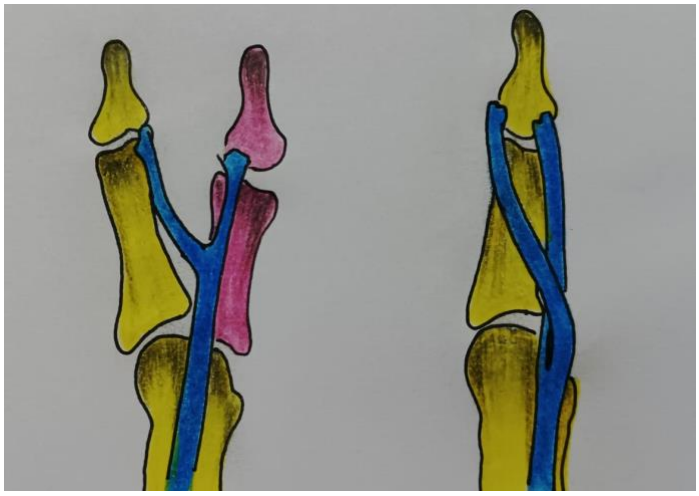

Fig 12: Schematic representation of realignment of terminal tendons thereby negating the deforming forces.

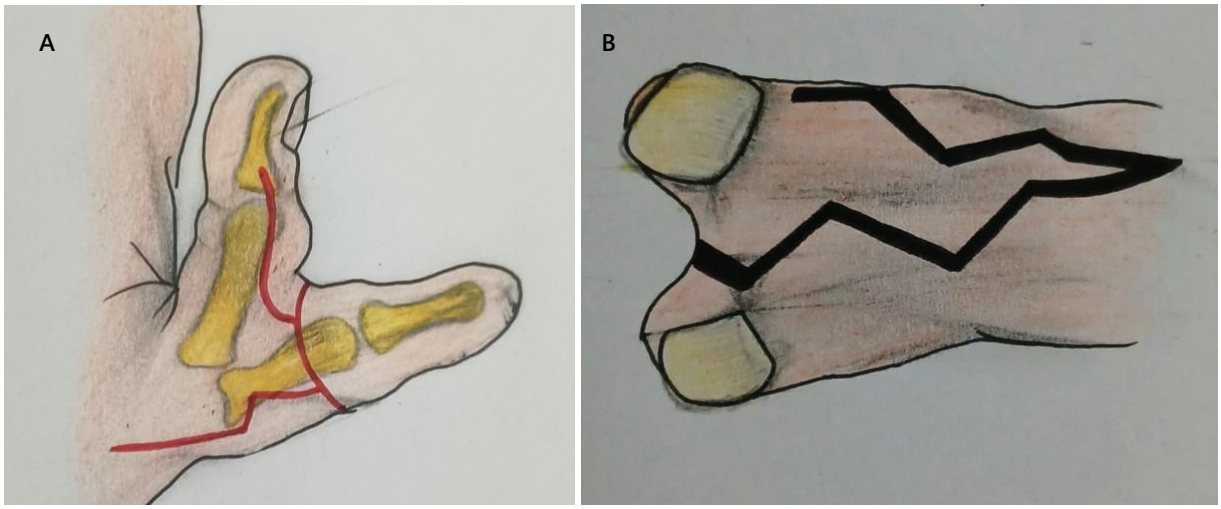

Fig 13: Schematic diagram of the radial zigzag incisions with circumferential racquet extension around base of hypoplastic radial thumb used for a Wassel IV reconstruction (A). Converging dorsal and volar incisions, preserving the nail plate, nail bed, and eponychium for soft tissue reapproximation (B).

Bilhaut-Cloquet procedure[10] (Fig 14)

Indicated when the two duplicates are equal in size and length (Fig 15), but both are hypoplastic or have a zigzag deformity with instability. The central parts of the duplicates are excised, and the outer elements are combined to form the new thumb. Since the nails are shared, a split-nail appearance is expected.

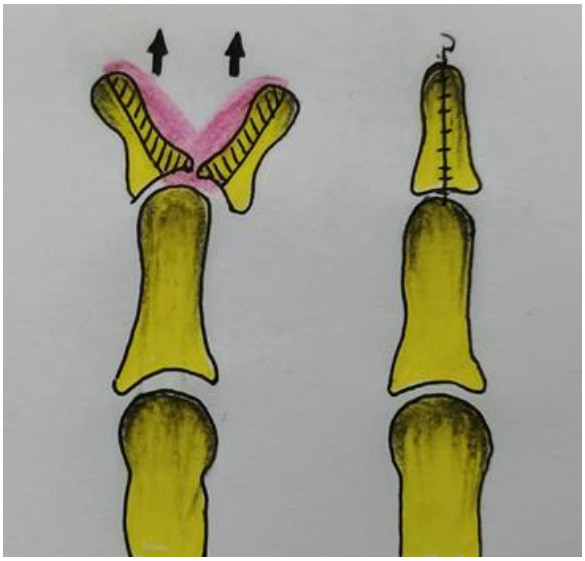

Fig 14: Schematic representation of Bilhaut – cloquet procedure.

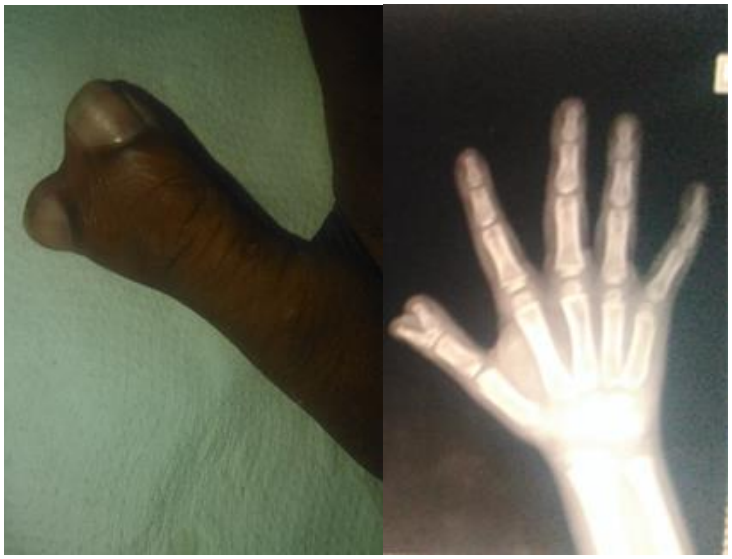

Fig 15: Clinical picture showing duplicates with equal size but are hypoplastic.

Modified Bilhaut-Cloquet procedure[1] (Fig 16)

Done for the same indications as the Bilhaut-Cloquet procedure, but one of the duplicates has a more giant nail (Fig 17). Here some longitudinal components of both thumbs are combined. The larger nail is retained, and the hypoplastic nail is discarded. Perhaps the most common combination is to use the nail and terminal phalanx of the better thumb, combining this with a part of the terminal phalanx of the lesser thumb. This requires unequal bony excision at the distal phalanges. Post resection the autologous tissue becomes available for grafting. They are made to provide mechanical support by various transpositional techniques. The raised flaps augment the retained thumb to make it more cosmetically comparable to the uninvolved side.

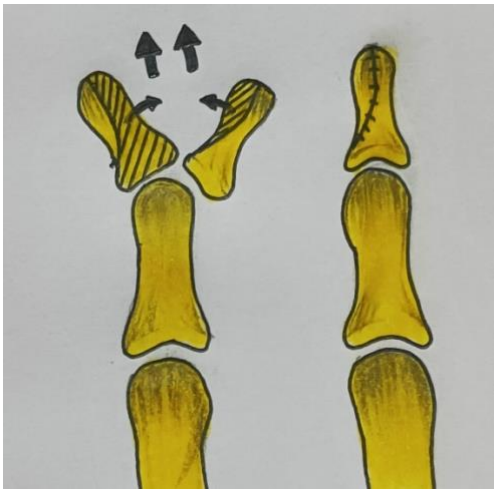

Fig 16: Schematic representation of Modified Bilhaut – cloquet procedure.

Fig 17: Clinical picture with one of the duplicates having a giant nail.

On-top plasty[11]

Indicated in duplications in which one thumb is proximally (Fig 18), and the other thumb contains a better pulp and nails distally. One thumb is put on top of the other. On top plasty, also known as *Acral transposition* is effective in combining the better parts of both thumbs. This may be combined with a modified Bilhuat-Cloquet procedure. It is the reconstruction by Combining the Distal Radial and Proximal Ulnar Thumbs.

Fig 18: Clinical picture with one of the duplicates being less developed and is proximal.

Ray transfer

Here is the duplication at the metacarpal level (Fig 19). One thumb will have a well-developed metacarpal and the other a well-developed distal part.

Fig 19: Clinical picture with duplication at metacarpal level.

Atypical cases

Atypical cases include Triphalangism, Triplication and Ulnar Dimelia. The methods stated above are applicable. The surgeon must aim for a CMC joint that is appropriate in size and shape, supports a solid skeleton distally, and results in a thumb with optimal mobility and size and shape. During evaluation one must decide which digit, or combination of digits, will give the most effective thumb. The construction of an appropriate CMC joint is the key to success. For thumb reconstruction, the best digit is chosen. This digit may have the skeleton of a finger or a transitional metacarpal with proximal and distal physes in some cases.

Two Inadequate Thumbs

It is uncommon to be encountered with a situation in which both thumbs' CMC joints are insufficient. However, when this is accompanied with extreme distal hypoplasia and instability, no reconstruction is likely to result in a functional thumb. Although many find it difficult to accept

but removing both the hypoplastic thumbs, which are non-functional, to make a four-finger hand and then performing a pollicization of the index finger may provide a better appearance and function.

Complications

Nail deformity is the common complication in type 1 duplication, in type 2 & 4 duplications there is a greater risk of stiffness and instability post-surgery. Type 3 has got good prognosis as it does not involve the joint. Type 5 needs reinsertion of the thenar muscles which may lead to reduced opposition power.

To summarize, thumb duplication, like many other congenital hand anomalies, could present as a wide spectrum of deformities and the surgeon should be aware of the challenges its correction may pose. Effective management requires a thorough understanding of the pathological anatomy of duplication and ability to correct all of them at a single sitting to get a thumb which is of good shape, size, stable and has adequate mobility to provide good function.

References

1. Tonkin MA. Thumb duplication: concepts and techniques. *Clin Orthop Surg*. 2012;4(1):1-17.

2. Watt AJ, Chung KC. Duplication. Hand Clin. 2009 May;25(2):215-27

3. Manske MC, Kennedy CD, Huang JI. Classification in brief: the Wassel classification for radial polydactyly. Clin Orthop Relat Res. 2017, 475: 1740–6.

4. Zuidam JM, Selles RW, Ananta M, Runia J, Hovius SE. A classification system of radial polydactyly: inclusion of triphalangealthumb and triplication. J Hand Surg Am. 2008, 33:

5. Chung MS, Baek GH, Gong HS, Lee, HJ, Kim J, Rhee SH. Radial polydactyly: proposal for a new classification system based on the 159 duplicated thumbs. J Pediatr Orthop. 2013;33:190–196.

6. Wang Y, Wang ZT, Wang DH, Zhang LS, Hu Y. A radial polydactyly classification system used for surgical planning. J Hand Surg Eur. 2021a, 46: 378–83

7. Kim JK, Al-Dhafer BAA, Shin YH, Joo HS. Polydactyly of the thumb: a modification of the Wassel–Flatt classification. J Hand Surg Eur. 2021, 46: 346–51

8. Kitayama Y, Tsukada S. Patterns of arterial distribution in the duplicated thumb. *Plast Reconstr Surg*. 1983;72:535–542

9. Saito S, Tsuge I, Yamanaka H, Morimoto N. Soft tissue abnormalities in Wassel Type VI radial polydactyly: a detailed anatomicalstudy. J Hand Surg Eur. 2021, 46: 352–9

10. Tonkin MA, Bulstrode NW. The Bilhaut-Cloquet procedure for Wassel types III, IV and VII thumb duplication. J Hand Surg Eur Vol. 2007 Dec;32(6):684-93

11. Al-Qattan NM, Al-Qattan MM. On-top and side-to-side plasties for thumb polydactyly. *Int J Surg Case Rep*. 2017;39:88-92.