

## An unusual case of preaxial polydactyly of the hand (triplication of the thumbs)

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### Abstract

Preaxial polydactyly is the most common duplication pattern in white and Asian populations (1). It is a congenital anomaly with a wide range of manifestations. Current classification do not have the capacity to classify all different types of radial polydactyly. We describe here a very rare and unusual case of bilateral preaxial polydactyly (triplication) in a woman and report the operations results. We have not found similar case in the literature. Our case is unique and did not fit into the classification systems described for thumb polydactyly.

**Keywords:** Congenital Anomaly, Preaxial polydactyly, Thumb, Triphalangeal, Triplication.

### Introduction

The preaxial polydactyly represents a complete or partial duplication of the thumb. It is the most common duplication pattern in white and Asian populations, occurring in one per 3000 births (1-3).

The longstanding classification of thumb polydactyly was initially described by Wassel in 1969. Wassel's classification is the most common used classification. In this classification, the extent of duplication is defined and indexed according to whether the components are attached proximally (bifid) or completely separated (duplicated). Thumb duplication has been classified into seven types by Wassel (4). Various adaptations and modifications have been

added to this scheme, but the basic premise remains unchanged. In addition, there are some types of thumb duplication that defy classification (5-8).

We described here a rare and unusual case of bilateral preaxial polydactyly (triplication) with report of the result of operation in a woman.

In spite of searching through the literature we could not find any similar anomaly.

### Case report

A 22 year-old woman was seen for the assessment of congenital anomaly of the hands. She was right hand dominate and housewife. Family history was positive for congenital anomalies of the hand. She has four sisters and two brothers. One of her

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sister, one of her brothers, a grandmother and a grandfather has history of congenital hand anomalies. Except for deformity of the hands, the patient had no other anomalies and was in all other respect normal. Examination of the upper limbs showed no abnormalities of the shoulders, elbows, forearms or wrists.

At physical examination triplication of the thumbs with syndactyly and angular deformity was obvious bilaterally. Each thumbs had separate nail bed (Fig. 1). Radiologic examination showed a normal ulna and radial and carpal bone bilaterally. The X-ray examination revealed a triplication of thumbs bilaterally (Fig 2). She had bifid first metacarpal bilaterally. Both radial sides of thumbs were hypoplastic and each had two phalanxes. The ulnar side metacarpal had duplicated proximal phalanx with an extraosseous (or rudimentary phalanx) at radial side of each metacarpophalangeal joint bilaterally. Triphalangism was seen at radial and middle thumbs at right side and only at middle thumb at left side.

With good preoperative planning, surgery was advised to improve the thumbs appearances and function.

In the right side, two thumbs (radial and middle) were excised. The radialmost thumbs at the metacarpal base level and middle thumb at the MP joint level excised. Radial collateral ligament of the MP joint reconstructed and fixed with one pin for stability of the MP joint. We did not excise extraosseous at the MP joint. Delta phalanx (middle phalanx of the ulnar most thumb) excised and casuloplasty was done. Extensor tendon centralization and extensor tendon transfer (one's of the amputated thumb) for abductorplasty was done. Post operative recovery was uneventful (figure 3).

After one year, we operated the left thumb. At operation the radial and middle thumbs excised. Fusion of the MP joint with cortical bone graft (from amputated thumb) was done because of the MP joint was not reconstructable. The MP joint extraosseous excised and centralization of



Fig.1. Right thumb triplication photographs.



Fig 2. Left thumb triplication photographs.



Fig 3. Radiography of right and left thumb triplication.



Fig 4. Right side postoperative radiography.

the extensor tendon and abductorplasty were done. Post op recovery was uneventful (Fig. 4).

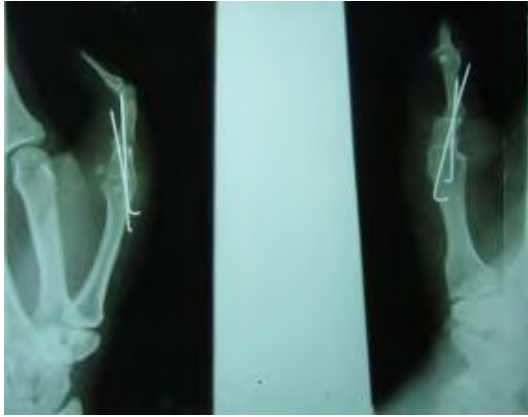


Fig5. Left side postoperative radiography.



Fig 6. Postoperative photography of bilateral triplication thumbs.

### Discussion

Duplication of digits, or polydactyly is a common and conspicuous hand anomaly. Polydactyly is classified into three main categories: 1) preaxial, duplication of the thumb (bifid thumb), 2) central, duplication of the index, long or ring finger, and 3) postaxial, and duplication of the small finger (1).

Classifications are useful in guiding and evaluating surgery management and treatment (8). The long –standing classification of thumb polydactyly was initially described by Wassel in 1969, is practical and based on level of duplication. The classification starts from distal to proximal, in

which uneven numbers are bifid duplication, even numbers are complete duplication, and type VII refers to duplication involved with triphalangism. Whereas for "simple" radial duplication the Wassel classification is sufficient to classify all different types, for more complex duplications, triplications, or triphalangism, a more extensive classification is necessary. Wood and Miura created more subtypes for classifying triplication and triphalangism (5,6). In Wood's classification, type IV is subdivided into type A, where on the level of proximal phalanx both duplications have a triphalangeal component and type B has only a triphalangeal component on the radial side (5). Miura added type C with a triphalangeal component on the ulnar side (6). Wood also subdivided Wassel's triphalangism group (type VII) in to 4 subtypes:

- A. a triphalangeal ray originating at the level of the metacarpal on the ulnar side
- B. both sides have triphalangeal components
- C. the radial side is triphalangeal
- D. a central triphalangeal ray with nontriphangeal hyposplastic duplications on each side (also known as triplication) (5).

Zuidam et al recently attempted to address some concern with a more extended classification. This classification includes the carpal bones: Type VII represents partial bifurcation at the level of the trapezium, and type VIII represents complete bifurcation on the scaphotrapezium joint. Additional features include specific notations for triphalangeal (Tph), triplication (T), symphalangism (S), deviation (D), and hypoplastic or floating (H); the directional position of the deformities are assigned abbreviation: U (ulnar), M (middle), and R (radial).

Our case did not fit readily into any of the Wassel's groups. Whereas the left side deformity is compatible to Miura's VII D subgroups (without consideration of the MP joint rudimentary phalanx), but the right side deformity did not fit into any

subgroups of Miura's classification. For this reason it might be necessary to consider adding on the groups of Wassel and the Wood's-Miura's classification. Several type of radial polydactyly cannot be classified using the mentioned classification schemes. Zuidam et al. recently attempted to address some of these concerns with a more extended classification. With Zuidam et al classification, our case was triplication on different levels associated with triphalangism, rudimentary phalanx, hypoplastic and deviation. But other consideration such a syndactyly and extra osseous should be addressed. Our case had a extraosseous bone at the MP joint bilaterally. If we consider that a rudimentary or floating ray, exactly a non-reported four – thumb radial polydactyly was presented (tetraplication thumb).

Back–Gramko stated that the true triple thumbs is very rare and mentioned three cases known to him in 121 hands (112 patients) with radial polydactyly, which only nine patients had triplication.

The case described here dose seem to be a true triple (or perhaps tetraplication thumb). The rest of the hand was normal, rulling out any contribution from the index finger.

The case reported here did not fit into any of the Wassel groups and other classification. For this reason it might be necessary to consider adding another group to the radial polydactyly classifications.

Surgical correction of the radial polydactyly almost always is indicated, not only for the obvious cosmetic improvement, but also for better function. Surgical reconstruction generally is performed when the child is about 18 months old, but no later than 5 years old if possible. For type I and possibly type II bifid thumbs, a combination procedure (Bilhaut- Cloquet) is recommended. More proximal duplication requires excision of the most hypoplastic thumb, narrowing of the widened proximal articular surface, ligament reconstruction, intrinsic transfer and centralization of the extensor flexor and extensor tendon if necessary. In general the ulnar in most

thumbs should be preserved. Radial polydactyly combined with triphalangeal components requires detailed surgical procedures for reconstruction of joint surface, reduction of length, and positioning of the thumb.

Our patient was not presented at good age for surgery but for improvement of cosmetic and function, we advise her for reconstructive surgery. At operation, we save MP joint at right side and fused left MP joint, and then two extra thumbs excised bilaterally.

The results vary with the degree and complexity of the thumb duplication. Angular deformity, instability, size of thumb, scar of surgery, nail deformity and joint stiffness are the most frequent complications.

The results of our case were relatively good according to appearance, length, stability and function at follow up (18mo).

Our case is very rare and unusual radial polydactyly (triplication or tetraplication) and whereas the patients operated at 22 years old age but the result was good.

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