

The varied forms of duplicated thumb in infants

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ABSTRACT

Hand polydactyly is a common congenital anomaly. It appears as a single disorder, or as a separate manifestation of a syndrome. In the Caucasian race, thumb duplication (preaxial polydactyly) is the most common form of hand polydactyly. The purpose of this study was to isolate cases of thumb duplication that were surgically treated during the time period 2008-2018, and to identify diagnostic pitfalls and therapeutic concerns. We studied twelve duplicated thumb cases and classified them according to their severity with Wasel's classification system. All cases were treated by the same surgical team under general anesthesia using pneumatic tourniquet. In a 2-8-year follow-up period, patients were evaluated according to subjective and objective criteria, emphasizing on functional and aesthetic outcomes. On an evaluation scale, results were satisfactory for ten patients (83.34%), while two patients presented moderate results. We analyze the reasons for the moderate results and look for ways of avoiding them. In conclusion, preoperative planning is considered necessary, given the diversity of cases. Choosing the right timing protects both the physician and the patient from practical difficulties. Clinical examination is the patient's first diagnostic approach, while simultaneously studying radiographs helps to identify the problem more accurately. The technique must follow the principles of plastic surgery.

KEY WORDS: preaxial polydactyly; duplicated thumb; congenital deformity; infants.

Introduction

Preaxial polydactyly is a very common congenital abnormality of the upper limb, which is clinically manifested by a duplication of the thumb, at a frequency ranging from 0.08 to 1.4 per 1000 healthy infant births [1,2]. Its reasoning has not yet been clarified, although an autosomal dominant inheritance

transfer was recognized in forms where the supernumerary thumb had three phalanges. This event appears to occur during the first eight fetal weeks as the entire upper extremity and hand develop. Several theories try to explain it as a possible fetal pathogenicity, but they are not reliable enough to be accepted. One of these theories claims that preaxial

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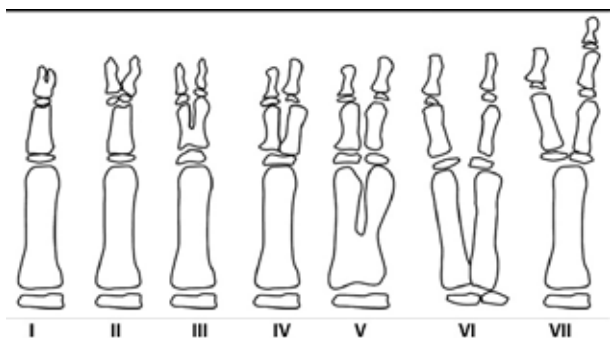


Figure 1. Wassel's classification system (1969).



Figure 2. Incomplete thumb duplication of distal phalanx (type I) a. clinical image b. radiography.

soderm and ectoderm during cell proliferation [3-5].

This condition may be isolated or may be associated with other manifestations of specific syndromes, such as acrocephalopolysyndactyly type 1 (Noack) and type 2 (Carpenter), Holt-Oram, Fanconi, Rubinstein-Taubi, or Down syndrome [6], brachydactyly, left lip-palate, imperforated anus, syndactyly, and vertebral anomaly.

Duplicated thumb cases are estimated according to Wassel's classification system [7], exclusively by radiological criteria, depending on the severity of the deformity and the type of phalanx and / or first metacarpal abnormality (**Fig. 1**).

Clinical and radiological evaluation are considered essential for evaluating the outcome of surgical intervention. Cheng et al proposed evaluation of the alignment of the remaining thumb, the range of motion of the interphalangeal and metacarpophalangeal joint, the stability, the interdigital space, the prominence at the point of resection of the supernumerary thumb, and the opposition ability of the remaining thumb [2]. Ogino et al consider as satisfactory parameters for assessing the evolution of surgical repair, the type of deformity, the type of surgery and surgeon's skill [8]. However, Kemnitz observed that the loss of complete thumb mobility is less important than previously thought [1]. Of greater

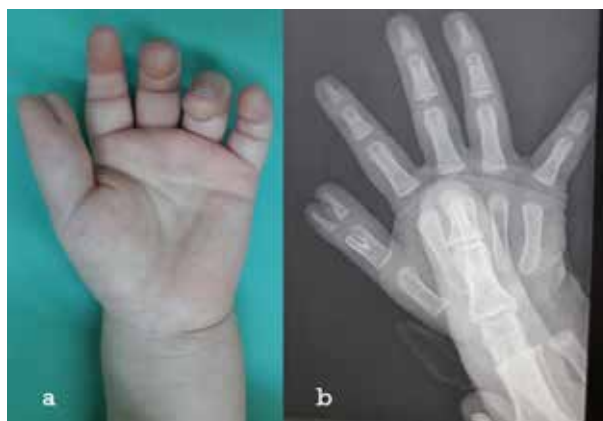


Figure 3. Incomplete thumb duplication of proximal phalanx (τύπος III) a. clinical image b. radiography, in which the mother's index finger is also obvious.

value seems to be the aesthetic problem created by the axial deviation.

The purpose of our study was to isolate the cases of thumb duplication that we treated surgically during 2008-2018 time period, to refer to our experience and to identify diagnostic pitfalls and therapeutic concerns. We considered important the subjective evaluation of the surgical outcome, based on a questionnaire completed by the patient's parents at follow-up.

Patients and Methods

From January 2008 to December 2018, we treated 92 cases of polydactyly in 43 hands and 49 feet in 69 patients. Twelve cases (7 boys and 5 girls) with an average age of 18 months (range 7 months - 4.5 years) related to duplicated thumb. According to the Wassel's classification, we operated 2 cases of type I (**Fig. 2**), 4 patients of type II, one patient of type III (**Fig. 3**), 4 patients of type IV with duplication of the proximal phalanx (**Fig. 4**) and a patient of type V (**Fig. 5**) with a bifid first metacarpal (**Table 1**).

The majority of cases (83.34%) were operated on over 12 months of age. At these ages, radiographs prove to be safe for bone structures, the risk of anesthesia is lower and manipulations more precise. Preoperatively, the clinical examination was performed in the presence of the parents, hands were photographed with their permission, and they were informed of the planned technique and expected results. Radiological examination was completed in each patient, both in the normal hand

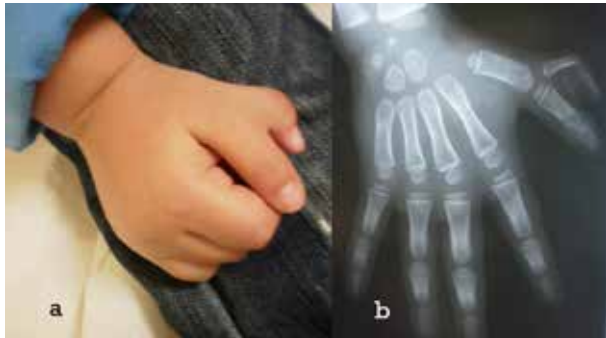


Figure 4. Complete thumb duplication of proximal phalanx (τύπος IV) a. clinical im-age b. radiography.



Figure 5. Clinical image of incomplete thumb duplication of first metacarpal (type V).

and in the pathological one.

All cases were treated under general anesthesia with the use of pneumatic tourniquet. In type I and II cases, special attention was paid to the nail splitting (Fig. 6). The Bilhaut-Cloquet technique was never used, although there were parents who had met it on the Internet and asked for it [9-11]. Also, no Kirschner wires were used to hold the desired axis during the first 3-4 postoperative weeks.

The supernumerary thumb was carefully excluded so that the skin flaps were sufficient to cover the surgical wound. The common neurovascular bundle was also preserved. In Type IV and V cases attention was paid to strengthening the extension of the remaining thumb by the use of connective tissue by the extensor mechanism of the excluded thumb, while the abduction was enhanced by the re-suturing of the abductor pollicis brevis using a slowly absorbable suture 5-0. When phalanx or metacarpal osteotomy was required, a thin

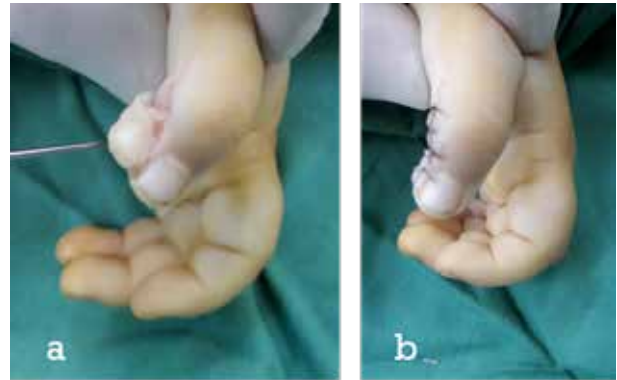


Figure 6. Careful separation of the nail for the treatment of duplicated thumb in type II (patient 3).



Figure 7. Prominence of soft tissues in a patient with duplicated thumb of type IV (patient 10) a. preoperative appearance b. surgical image c. postoperative appearance.

metal osteotome was used and bone wax was inserted to control bleeding.

Intraoperatively, the pneumatic tourniquet was removed, to check blood supply before suturing the skin with 4-0 or 5-0 nylon sutures. Gauze dressings, cotton and elastic bandages were applied in a boxing glove form. There were two visits to the outpatient clinics, the first for simple gauze changing and the second for suture removal. The hand was free of bandages at 3 weeks.

Follow-up lasted 2-8 years. At each reassessment, a special form was filled in with the patient's name, age at the time of surgery, Wassel's classification, surgical technique, aesthetic parameters (scarring, pulp or nail hypoplasia, axial deviation, phalangeal or metacarpal prominence), as well as functional parameters (hypo-sensitivity or numbness, ability to grasp small objects, thumb to index and thumb to little finger opposition). Grades 9-10 rated the score as satisfactory, 5-8 moderate and below 5 poor.

Results

In no case did the postoperative scar create a problem while the residual thumb pulp presented a satisfactory



Figure 8. Despite the excellent surgical technique in the treatment of duplicated thumb type IV (patient 8), the fair result could not be avoided due to prominent soft tissues and opposition inability of the remaining thumb.

image in all cases. On the contrary, nail hypoplasia to a degree that was aesthetically problematic was observed in two patients with type I duplicated thumb (Table 2). Slight axial deviation was found in one type II patient and in one with type V duplicated thumb. Prominence of the soft tissue, or the head of the first phalanx, or metacarpal, was found in one type II patient (Fig. 7) and two type IV patients.

The ability to grasp small objects was normal in all cases, as well as the opposition ability of the remaining thumb to the index finger. However, the opposition of the thumb to the little finger was problematic in one type II patient and two type IV patients, probably due to stiffness of the interphalangeal or the metacarpophalangeal joint.

Looking closely at Table 2, we observe that ten patients (83.34%) scored 9-10, with satisfactory results. One type IV patient (Fig. 8) was assessed with grade 8 and another type II patient with grade 7 (moderate results). The patient with the lowest score was the one with the highest age. In no case did the parents suggest re-operation to improve the aesthetic or functional outcome.

Discussion

In our study, all cases were unilateral. The presence of bilateral deformity would force us to look for a syndrome such as Townes-Brocks syndrome [12]. There was also no frequency difference according to gender, and most authors agree with this conclusion [1-5,7,8].

Most of the cases in this study belong to forms II and IV. There are other researchers who agree with this finding [13], although Naasan and Page claim that type VII is the most common [14]. However, Wassel's classification system is often inadequate to present details related to surgical design, or to describe common sub-

Patient	Sex	Age	Type
1	Male	7 months	I
2	Male	12 months	I
3	Female	15 months	II
4	Female	18 months	II
5	Male	4 y and 6 m	II
6	Female	12 months	II
7	Male	18 months	III
8	Male	20 months	IV
9	Female	22 months	IV
10	Male	17 months	IV
11	Female	10 months	IV
12	Male	11 months	V

Table 1. The patients of the study according Wassel's classification

types that require separate surgical treatment.

In an attempt to solve this problem, Hung et al [15] propose for Wassel's Type IV a new classification with four subtypes to avoid possible post-operative complications: IV/A (hypo-plastic supernumerary finger), IV/B (supernumerary finger with ulnar deviation), IV/C (divergent duplication), and IV/D (convergent duplication). For cases where coexists a supernumerary thumb with three phalanges (Type VII), Wood [16] proposes four subtypes: VII/A (radial hypoplastic triphalangeal finger); VII/B (consisting of two complete triphalangeal thumbs); VII/C (ulnar hypoplastic triphalangeal finger); VII/D (triphalangeal finger accompanied by a hypoplastic ulnar digit).

Chung et al [17] adopt a new classification, based on the anatomical features of the deformation, with the aim of facilitating surgical times and avoiding complications: type I (joint type) where the supernumerary thumb has its own joint; type II (single epiphyseal type) where both thumbs share the same epiphysis; type III (osteochondroma-like type) where the supernumerary thumb resembles to osteochondroma; type IV (hypo-plastic type) where the supernumerary thumb is associated with the normal one via soft tissue. In a series of 159 cases, 134 duplicated thumbs (84%) were treated with excellent results, 17 (11%) with moderate and 8 (5%) with poor results.

Recently, Wassel's classification system has been im-

Patient	Scar	Pulp	Nail	Deviation	Prominence	Hypoesthesia	Numbness	Pinch	Thumb-index	Opposition
1	-	-	+	-	-	-	-	yes	yes	yes
2	-	-	+	-	-	-	-	yes	yes	yes
3	-	-	-	-	-	-	-	yes	yes	yes
4	-	-	-	-	-	-	-	yes	yes	yes
5	-	-	-	+	+	-	-	yes	yes	no
6	-	-	-	-	-	-	-	yes	yes	yes
7	-	-	-	-	-	-	-	yes	yes	yes
8	-	-	-	-	+	-	-	yes	yes	no
9	-	-	-	-	-	-	-	yes	yes	yes
10	-	-	-	-	+	-	-	yes	yes	yes
11	-	-	-	-	-	-	-	yes	yes	no
12	-	-	-	+	-	-	-	yes	yes	yes

Table 2. Treatment results in twelve patients based on subjective and objective criteria, regarding aesthetic and functional parameters.

proved with the Rotterdam classification, which consists of 8 types and uses letters to indicate anomalous elements and their location, such as thumb with three phalanges or triple-thumb variants [18,19]. In a study [20] where 520 cases were examined, only 60% were estimated with the Wassel's classification, whereas they could be evaluated with the Rotterdam classification at 100%.

The patient's age on the day of surgery positively or negatively affects the postoperative outcome. Proper timing should be the first care of the Surgeon. However, there is controversy among the researchers. Although some claim the deformity to be repaired at ages younger than 6 months, most recommend surgery later, at around 12 months, when the anesthetic risk is lower, the patient may progressively improve hand functionality, and before social effects occur which would adversely affect his confidence [21]. Cabrera Gonzáles et al recommend that surgery be performed at ages 7-12 months, based on their experience with the least complications [22]. Dobyns et al recommend the age of 6-18 months [6], while Goffin et al suggest that the ideal age is between the first and second year [23]. Excellent results appear to arise when surgery is performed under the age of 3 years, with the ultimate goal of preventing postoperative deformities. This is probably the reason for the moderate outcome in our patient aged 4 years and 6 months.

The extensor tendon of the supernumerary thumb strengthened the action of the extensor of the remaining thumb in type II cases in our study, but we did not move the flexor tendon centrally as described by Miura [24], nor did we use the extensor tendon of the index finger to stabilize the interphalangeal joint and correct the extension deficit as described by Kawabata et al [25]. Our view was that we should not seek to improve axis deviations, as suggested by Goffin et al [23], in order not to disrupt the growth plate of the epiphysis and not to limit joint mobility.

The goal of the procedure should be to provide a stable and flexible thumb of sufficient size and shape. Stability and size are related to strength, both for strong and fine grip [26]. Thumb mobility depends on the integrity of the carpometacarpal joint, which is guaranteed in duplicated thumb types I, II, III and IV but not in types V and VI. The mobility of the metacarpophalangeal and interphalangeal joints, although considered essential for proper thumb function, is of less importance.

The ingenuity of the various authors to overcome complex problems during surgery is remarkable. It is a rule that both thumbs have both flexor pollicis longus (FPL) and extensor pollicis longus (EPL), but the more functional thumb has better tendons and a wider range of active motion. However, the eccentric adhesion of these tendons poses a problem that is seeking its solu-

tion. Also the shape of the remaining thumb is often not the desired one. Pulp and nail are also smaller than normal in most cases [13].

Chang et al [27] propose transport of the duplicated thumb, provided that none of the thumbs is superior to the other, where one having a better central section and the other a better peripheral section. According to the authors, this technique is simple, safe, and effective for remaining thumb functionality in type IV cases.


Various techniques have been proposed for subtype IV/D (convergent duplication). Abid et al recommend a modified Bilhaut-Cloquet technique, in cases where both thumbs are hypoplastic, with their divergence at the level of the metacarpophalangeal joint and convergence at the level of the interphalangeal joint [28]. The purpose is to avoid the usual complications of the classic Bilhaut-Cloquet technique (coaptation of equal parts of bone, soft tissue, and nail tissue after resection of the central segment of the duplicated thumb), like nail dystrophy, axial deviation and instability. Hung recommends the use of an epiphyseal segment of the proximal phalanx with insertion of the abductor pollicis brevis tendon into the radial side of the epiphyseal proximal phalanx of the ulnar thumb [29]. Xu Yun-lan et al support the ablation of the radial thumb and reconstruction of the ulnar thumb by a series of soft tissues procedures, including FPL rebalancing [30].

We did not use the Bilhaut-Cloquet technique because we considered it useless for the specific cases we had to deal with. This kind of operation presents a real challenge, as long as thumb duplication is absolutely symmetrical. However, it often leads to nail splinting [9-11,13,31]. An innovative technique to avoid this complication was described by Back et al in 2008, modifying the Bilhaut-Cloquet technique. Indications of this technique were concerning cases of type II or III, where both thumbs were symmetrical and the nail size was less than two-thirds of the normal thumb of the other hand, or smaller than the index finger size in patients with bilateral deformity [32].

Regarding the evaluation method used in this study, we preferred subjective and objective criteria, giving importance to the appearance and functionality of the remaining thumb. In 1998, Cohen was the first to classify residual deformities after duplicated thumb repair [5]. In the same year, Mih [33] distinguished residual deformities into three categories: joint abnormality (stiffness, deviation, and instability); bone abnormality (angled bone growth and presence of a delta-bone); soft tissue abnormality (reduction of the first space, hypoplasia of the thenar muscles and anomalous insertion of the flexor and extensor tendons).

We are pleased with the results of our research. However, the cases we faced were relatively simple and less complex. A disadvantage of our study is the short follow-up as well as the small number of patients. A study that will include the cases we have described, along with other new ones, is the next challenge for the future.

Conclusion

Preoperative planning in the case of duplicated thumb treatment is considered necessary given the diversity of the deformity. The day before surgery, decisions are made about the surgical approach, the creation of flaps and how to protect the thumb which will be preserved, while informing parents. Choosing the right timing protects both physicians (orthopedist and anesthesiologist) and patients of appropriate age to redefine the functionality of their hand. Clinical examination is the first diagnostic approach in any case. It is recommended to take a hand photograph as a reminder of the original thumb image, which can help parents appreciate the value of the end result. The study of x-rays helps to identify the problem more accurately. The technique must follow the principles of plastic surgery *lege artis*. 

Conflict of interest

The authors declare no conflicts of interest.

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