Triphalangeal Thumb; three is a crowd?

De triphalangeale duim; drie is teveel?

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Preface

"Iets uit de duim zuigen"

The thumb is of great importance in daily life. Etymologically speaking the thumb is seen as the *"swollen part"*, indicating the greater thickness compared to the fingers. The word derives from the Indo-European "tumere", meaning to swell. The phalanx derives from the Greek word "phalangos" meaning *" in the line of battle"*, indicating the position of the whole row of fingers joints. These historical etymological descriptions of the thumb and the phalanges already indicate the role of the thumb in the hand, working with the four fingers, not being one of them. The latin term for the thumb, pollux, is said to derive from polleo meaning "I am strong", hereby referring to the additional strength of the thumb over the fingers [1].

The best first sentence to start a thesis is probably not a sentence indicating lies and fabrications. However the saying in Dutch: "iets uit de duim zuigen", indicating that a person is making things up, has an interesting historical perspective. In old sagas, putting a thumb in the mouth was a way to gather more wisdom [2]. "Den vinger in den mond steken"; putting a finger or thumb in your mouth, is presented in mythology and sagas to acquire knowledge regarding a specific point. The thumbs in this thesis were used to gather knowledge, however none of the thumbs were used in the manner described above.

ONE

Introduction
Aim of the thesis
Update of literature

The only difference between me and a madman is that I'm not mad.

Salvador Dali

If anybody told you your thumb was different, how would you react?

You would probably laugh at them and tell them that their thumb is abnormal. Abnormality is determined by the perception of the majority about appearance and functional use. You would feel that your thumb was abnormal if you could not function in a proper way, or if the appearance would be different from the rest of the population. But what if your thumb appeared different, but was still functioning properly? Would you still think it was abnormal? Probably not to you or your surroundings. People with congenital 'abnormalities' deal with this question on a daily bases, we all think we are normal and the 'rest' is different.

What is so abnormal or different about a triphalangeal thumb?

The difference compared to the thumb we know as normal, is determined by the clinical presentation. A triphalangeal thumb is recognized by the extra phalanx located between the proximal and distal phalanx. The shape and size of this phalanx can differ from a very small delta type extra phalanx to a fully developed extra phalanx and consequently give the thumb a finger-like appearance. Besides the extra phalanx, a triphalangeal thumb can have aberrant intrinsic musculature, joint instability, inadequate positioning of the thumb, and many associated anomalies. Therefore clinical presentation has a broad spectrum of anomalies not only located in the hand. One could imagine that the fully developed finger-like thumb would easily be seen as aberrant and give impairments in daily life. The triphalangeal thumb with a very small extra phalanx, on the other hand, appears and functions better than the thumb with a fully developed phalanx. These thumbs with a small extra phalanx are less recognized or sometimes even overlooked.

Is it common?

If the triphalangeal thumb was common we probably would not see it as abnormal. Triphalangeal thumbs are relatively rare, with a general incidence of 1 in 25.000 live births [3] . Wood [4] in 1976 described 362 patients with 630 triphalangeal thumbs reported in literature until then. Buck-Gramcko [5] in his practice described from 1960 to 1990 147 patients with 239 triphalangeal thumbs, of which 92 were bilateral affected. Most patients with bilateral involvement have a positive family history for triphalangeal thumbs.

So what is the difference between a normal thumb and a finger?

Actually Galen [6] was the first to speak of a triphalangeal thumb, calling all thumbs triphalangeal. In his opinion, a normal thumb consisted of three phalanges, rather than a metacarpal and two phalanges. His theory was supported by the position of the epiphysis of the first metacarpal; in the metacarpals of the fingers the epiphysis is positioned distally, in the thumb metacarpal the epiphysis is positioned proximally as in the proximal phalanx. Moreover, the metacarpophalangeal joint of the thumb is restricted in its movements resembling the proximal interphalangeal joint of the finger, the metacarpophalangeal joints of the fingers have additional movement in the radio-ulnar plane [7]. Vesalius later argumented against the theory of the thumb consisting of three phalanges in his *Fabrica*, stressing that the hand should be classified as a thumb and four fingers, based on the unique function of the thumb [8].

How does it develop?

The triphalangeal thumb was first reported by Columbi in 1559, as mentioned by Kelikian [9] .The first known case was presented by Dubois [10] in 1826. In literature many different hypotheses have been presented regarding the etiology of the triphalangeal thumb, indicating the complex path of development of the hand and more specific the thumb. Frazer, as mentioned by Abramowitz [11], stated that the extra phalanx was the result of a failure of fusion of two elements normally forming the distal phalanx. This theory was supported by Wilkinson [12] and Abramowitz, who described that the anatomical basis of the flexor tendon in the thumb resembles the profundus tendon with rudiments of a flexor sublimus tendon. Joachimsthal [13] suggested a second theory, that regards the triphalangeal thumb as a duplication of the index finger with the absence of a normal thumb. The duplicated index finger was not functionally equivalent to a normal functioning thumb, as it lacked opposition. Lapidus and Guidotti [3] suggested that an arrested attempt of formation of a bifid thumb was the etiological base of the triphalangeal thumb. In their theory the "extra" phalanx was either the proximal or the distal phalanx. This arrested formation theory could explain the different developmental stage of the extra phalanx. In literature the finger-thumb discussion is also based on the position of the growth plate of the first metacarpal. While the normal position of the epiphyses in a thumb is on the proximal end, for the fingers the position is on the distal end. Flatt [14] stated that if the position of the epiphyses of the first metacarpal is distal, in a triphalangeal thumb, it is a non-opposable thumb. This type of thumb has a more finger-like appearance. Wood [15] suggested that congenital anomalies have a higher incidence of double epiphyses. In **Chapter two** and **three** the thumb versus finger discussion is further clarified by studies on the position of the growth plate in regard to the first metacarpal length.

Triphalangeal thumb has also been reported as a result of teratogen exposure. Fetal hydantoin exposure is most associated with a cleft lip, congenital heart disease, unusual facies and mental retardation. In literature three cases of triphalangeal thumb after fetal hydantoin exposure are reported [16]. Thalidomide can cause reduction defects of the extremities, besides hearing loss, blindness, duodenal atresia and malformations of the heart. Thalidomide ingestion between 45 and 50th days of gestation is associated with triphalangeal thumb [17].

Genetically the development of a triphalangeal thumb is highly correlated with radial polydactyly. Linkage analysis in a family near Rotterdam revealed a triphalangeal thumb gene to chromosome 7q36 [18] [19]. The morphogen sonic hedgehog (SHH) expands the zone of polarizing activity, located in the ulnar limb mesoderm, and develops the limb along the radio-ulnar axis. In mice and chickens ectopic radial SHH increases limb volume and creates radial polydactyly [20]. Mutations in the SHH regulatory region result in polydactyly or triphalangeal thumb [21]. Besides the clear SHH over expression more mutations in other genes, unrelated to the SHH pathway, are also connected to radial polydactyly and triphalangeal thumb.

What does it look like?

As mentioned before the triphalangeal thumb can have many different appearances. Physical examination of the thumb in small children is difficult, however it is of great importance. Valuable information regarding joint stiffness, passive range of motion, joint instability can be obtained. In addition to physical examination, preoperative radiographic images are necessary to assess the osseous structures. Besides the size and shape of the extra phalanx, additional information regarding the first metacarpal and present epiphyses can be obtained. After the first paragraphs the impression could have raised that some triphalangeal thumbs only have an extra phalanx and no aberrant musculature or joint abnormalities. In **Chapter four** we measured intrinsic musculature in all different types to discuss this more extensively.

So there are many different types of triphalangeal thumb?

Yes, as in most congenital anomalies the deformity is a developmental process gone bad at some point. It is a watershed of all the possible combinations in shape or size of the extra phalanx, in- and extrinsic musculature, joints, and ligaments. To facilitate communication and comparison many different classifications have been developed over the years. Most classifications are based on the shape of the extra phalanx, although some have incorporated soft tissue and muscle deformity. The most important and common classifications are those of Wood and Buck-Gramcko [4, 5]. To be complete all classifications presented in literature are summarized.

In 1907 Hilgenreiner [22], described different types of triphalangism - "hyperphalangie des daumens" -. He classified them into four types:

1. Incomplete hyperphalangism. The extra phalanx is not fully developed, has a triangular shape, and is small.
2. Incomplete not yet complete hyperphalangism. The extra phalanx is more developed compared to the first type. Hilgenreiner named it hyperphalangism with brachyphalangism.
3. Complete hyperphalangism. A fully developed extra phalanx, yet the first ray has a thumb like appearance. In these types it was likely that opposition was possible.
4. Complete hyperphalangism. A complete extra phalanx was present, although the first ray had no resemblance with a thumb, the so-called "five fingered hand". In these hands no opposition was present.

The words "incomplete not yet complete" makes this classification unusable in daily practice. The observer determines the "amount" of hyperphalangism, therefore the groups are not comparable between observers. Cocchi in 1952 [23] and later Swanson [24] presented a classification distinguishing two types: a brachymesophalangy form, with a rudimentary extra phalanx and secondly a dolichophalangy form, in which the thumb had a finger like appearance without opposition. Later Wood [4] added a third type: an extra phalanx not fully developed but with a thumb like appearance with opposition. Wood named the three types in order of development of the extra phalanx: Delta, Rectangular and Full. Nowadays Woods subdivision into three different types is the most common used classification for triphalangeal thumb. The shapes added by Wood (Delta, Rectangualar and Full) made the classification more suitable for daily practice. Still observer variation is present, but far less then compared to the classification of Hilgenreiner. Müller [25] in 1937 presented a classification based on teratologic sequence. He classified triphalangeal thumb into three types:

1. A rudimentary delta-shaped extra phalanx, or a small additional osseous part at the proximal end of the distal phalanx of the thumb.
2. An extra phalanx with the properties of a phalanx but without epiphyses.
3. An extra phalanx with epiphyses.

By classifying the triphalangeal thumb by teratologic sequence, Müller indicates the developing scale of triphalangeal thumb's extra phalanx, hereby discarding any possible anomalies of soft tissue, and intrinsic musculature. Buck-Gramcko [5, 26] in 1987 also designed a classification based on the teratologic sequence but subdivided six specific types based on treatment of the triphalangeal thumb. Buck-Gramcko stressed the importance and variance of the positions of the epiphyses, in the first metacarpal as well as the development and insertion of the intrinsic musculature. In his classification only the first types (type I to III) are opposable.

I. Rudimentary triphalangism: the distal phalanx has a long epiphysis without a well-segmented joint; sometimes a slight deviation is present.
II. Triphalangism with a short triangular middle phalanx: a wedge-shaped middle phalanx with deviation. The deviation is mostly to the ulnar side with otherwise normal anatomy of a thumb.
III. Triphalangism with trapezoidal middle phalanx: a transitional form is present with a relatively long thumb but with a short middle phalanx which is neither triangular (delta shaped) nor rectangular. The first ray has a thumb like appearance and the thenar muscles are present. The first metacarpal has two epiphyseal plates or a proximal epiphyseal plate; the normal position for a thumb. In that case a pseudo-epiphysis is often present at the distal end.
IV. Triphalangism with a long rectangular middle phalanx: the thumb has a finger like appearance ("five-fingered hand"). The thenar musculature is hypoplastic, so no opposition is possible.
V. Hypoplastic triphalangeal thumb: The thumb is hypoplastic, the joints are unstable and tendons are rudimentary. Often syndactyly with the index finger is present. These cases are often associated with other manifestations of radial deficiencies.
VI. Triphalangeal thumb associated with polydactyly: all possible varieties of a middle phalanx combined with radial polydactyly either on radial or ulnar side of the thumb. Cases with triplications are also classified as a type six.

With this classification Buck-Gramcko designed a more practical classification. Comparison amongst observers is more straightforward; however, the classification is still mainly focused on the extra phalanx. Since the combination of a triphalangeal thumb and polydactyly is often reported in literature, the classification by Wassel [27, 28] for radial polydactyly was adapted or subtypes were added. Wassel assigned a type VII for all different types of triphalangism independent of level of duplication. Wood [29] and Miura [30] added subtypes for a radial polydactyly with a duplication at the level of the first phalanx with a triphalangeal component (Wassel type IV). Wood divided the Wassel type VII into four subgroups depending of the position of the triphalangeal ray (radial, middle or ulnar), and added a type VII D if a triplicated thumb with triphalangeal components was present. Upton [31] added the suffix T, for triphalangeal, to the Wassel classification, to be able to classify the radial polydactyly with triphalangeal component for all levels of duplications. The position of the triphalangeal component was assigned by adding radial or ulnar in brackets after the level of duplication. Several other classifications for radial polydactyly incorporated the triphalangeal thumb in the classification [32, 33]. Besides all above-mentioned classifications, in our hospital, we still had patients with triphalangeal thumbs who would not fit in the current classifications. **In Chapter five** and **six** we discuss some of these cases and propose again a new classification for triphalangeal thumbs if combined with radial polydactyly. Thus allowing all possible types of triphalangeal thumbs to be classified without using a large "rest" group.

Can we do anything about it?

Treatment of the different types of triphalangeal thumb has varied over the years. Some authors advise no treatment [34], Kristjansen in 1926 advised more radical intervention, such as amputation of the distal phalanx with the nail complex [4]. Nowadays several treatment strategies are widely accepted for the simple triphalangeal thumbs as for the five fingered hand types.

For the less complex opposable triphalangeal thumbs, Beatson in 1897 advised the removal of the extra phalanx [35]. Milch [36] advised to perform the removal of the extra phalanx in early years of life because of the lower changes of capsular laxity or epiphyseal damage. He hereby mentions that operative treatment is contraindicated in adults, as it would give a poor functional outcome. Wood [4] suggested that performing an arthrodesis after removal of the extra phalanx in adults could have better functional outcome compared to the preoperative thumb that is too long. Cotta and Jager described the same operation, hereby mentioning the importance of correcting the joint capsule [37, 38]. Furthermore they advised a corrective sub-capital osteotomy of the basal phalanx if the joint was deviated. Wood stressed the importance of creating or recreating the collateral ligaments using surrounding soft tissue and periosteum, hereby giving stability to the created interphalangeal joint and preventing radial deviation. Wood also stressed the importance of the metacarpal epiphysis, and advised to destroy them when performing a metacarpal osteotomy, hereby preventing metacarpal overgrowth [4].

If the extra phalanx is larger, several types of osteotomies have been described. Flatt described a 'peg' shaped osteotomy for correction of length and deviation [14]. Peimer [39] adapted several types of osteotomies for his so called combined reduction osteotomy in which he performed a transverse osteotomy at the base of the distal phalanx, followed by an osteotomy at the base of the extra phalanx parallel to the proximal joint, hereby preserving the proximal interphalangeal joint and correcting possible deviation. In **Chapter seven** we will discuss and compare outcome of the osteotomy and the technique of removal of the extra phalanx.

For the more complex, nonopposable triphalangeal thumbs, Bunnell in 1944 [34] recommended no surgical interference, fearing the outcome would be worse compared to the preoperative function. It is generally agreed that patients with a nonopposable triphalangeal thumb benefit from surgical intervention. Many surgical procedures have been described for the correction of the position of the thumb through osteotomies. Barsky [40] and Bunnell and Littler (1959) advised an osteotomy of the shaft of the metacarpal to reduce and rotate the thumb, combined with deepening of the first web, hereby not correcting the triphalangeal components of the thumb but reducing the length. Scharizer [41] described a technique with excision of the first metacarpophalangeal joint with a first web deepening to achieve length reduction and to rotate and abduct the thumb. Furthermore they stressed the importance of combining the procedure with a flexor carpi ulnaris opponens plasty, creating adequate opposition. The flexor tendon was lengthened with the palmaris longus tendon and the pisiform bone was used as pulley. Cotta [37, 38] described a partial resection of the metacarpal joint, combined with shortening and arthrodesis on to the second metacarpal, hereby not correcting the triphalangeal component. Wood [4] described the possibility for correcting the deformity by performing a pollicization, however the insufficient amount of skin would limit the abduction. This problem could be overcome by creating a larger skin envelope with a dorsal skin rotation flap. The associated clinical malformations were seen by Wood as a major surgical problem. He advised early excision of polydactyly components, hereby preventing displacement and deviation. Also, in some cases, removal of the triphalangeal thumb could be preferred over removal of the more rudimentary biphalangeal thumb.

Several authors described the basic principles for correction of the triphalangeal thumb. Cotta and Jager [37] described three basic principles of treatment for the nonopposable thumb:

1. Decreasing the access length by shortening of the thumb.
2. Rotating and abducting the "finger" in a more opposable, "thumb-like" position, by performing a deepening of the first web, which they called "Phalangisierung".
3. Creating an opposable thumb by performing an opponens plasty.

Wood [4] described five separate surgical problems, for all types of triphalangeal thumb in order of importance:

1 The abnormally shaped extra phalanx
2 The associated clinical malformations; namely polydactyly
3 Five fingers in the same plane: the five-fingered hand
4 Narrow first web
5 Deficiency of the thenar musculature

In **Chapter eight** we discuss our results and surgical goals for the more difficult types of triphalangeal thumb.

Is there treatment algorithm available?

As mentioned above, triphalangeal thumb is a complex entity and it can be present in many different shapes, sizes and forms. As complex as it may be, basic principles as mentioned earlier are described [37, 38]. In our article of 2004 [42], not included in this thesis, we describe the methods used by the senior author in detail. For the small delta phalanx the procedure is as follows: 1) an excision of a transverse oval piece of skin, 2) resection of the extra phalanx with reconstruction of the ulnar collateral ligament at the new interphalangeal joint and 3) lengthening of the radial collateral ligament with a lengthening Z-plasty of the skin (Figure 1). If the extra phalanx is larger or of the trapezoidal type, excision of a transverse oval piece of skin, partial resection of the extra phalanx with correction of the angle and arthrodesis of the distal interphalangeal joint can be performed, gaining 1 to 1.5 centimeters length reduction (DIPRAD procedure) (Figure 2B). If a rectangular or full type is present, the

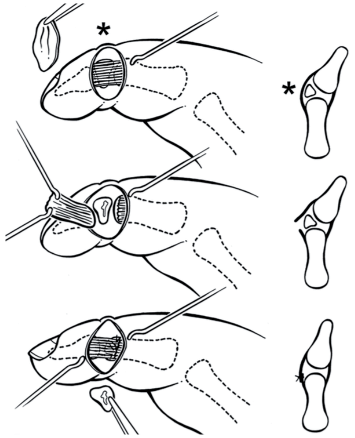

Fig. 1 A. Right Thumb : radial side

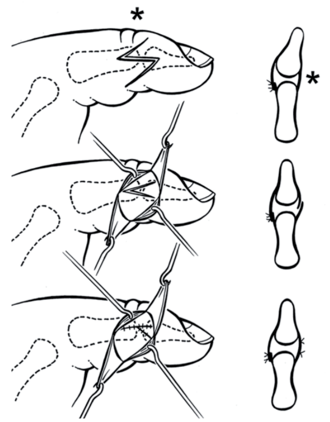

Fig. 1 B. Right Thumb: ulnar side (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

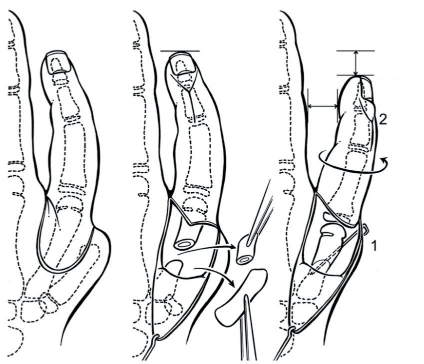

Fig. 2 A. Operation on metacarpal level (ROAMC1-procedure)

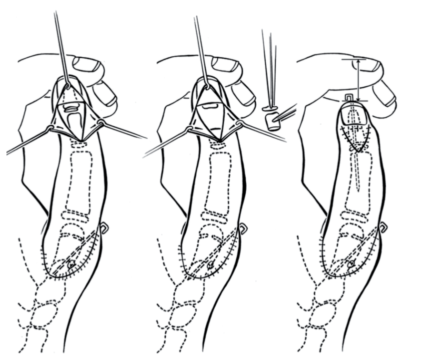

Fig. 2 B. Operation on phalangeal level (DIPRAD-procedure) (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

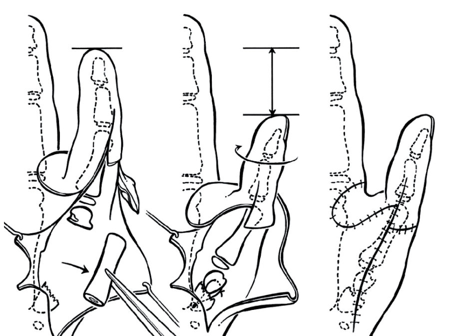

Fig. 3 A. Pollicisation : intraoperative view of metacarpal bone resection

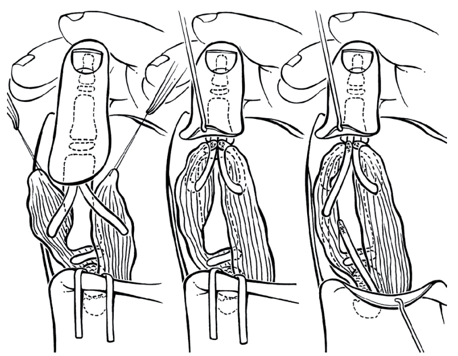

Fig. 3 B. Pollicisation: muscle and tendon reinsertion (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

carpometacarpal (CMC) joint of the first metacarpal is decisive for the type of procedure. If the CMC is reasonably developed, i.e., present and stable, a shortening, rotation and palmar abduction osteotomy at metacarpal level is performed to correct for position and length of the thumb (1 to 1.5 cm shortening (ROAMC procedure) (Figure 2A). Subsequently the extra phalanx is corrected through a partial resection of the extra phalanx with correction of the angle and arthrodesis of the distal interphalangeal joint [42] or proximal interphalangeal joint [43] Furthermore the extensor tendons are shortened as well as the intrinsic musculature. If the CMC joint is under developed a pollicisation is performed, according to Buck-Gramcko and Foucher, with adaptations to skin incisions and tendon reinsertions. Further adjustments regarding opposition, laxity of metacarpal joint of the thumb and first web deficiency can be necessary in individual cases (Figure 3). The patient is immobilized for four to six weeks, followed by instructions for movement by a hand therapist. If extensive surgery is performed, night splinting is continued for three months.

Do we need really need to do something about it?

Well, one could imagine the answer would be a loud yes, especially from the perspective of a hand surgeon. However some of the 'deformities' can be rather minor. In **Chapter nine** we discuss a group of adults with triphalangeal thumbs who had no surgical intervention.

AIM OF THE THESIS

The aim of this thesis was focused on the clinical outcome of the triphalangeal thumb, and on the many different aspects of the triphalangeal thumb that influence in these results. Therefore, this thesis can be subdivided into two parts: the aspects of the triphalangeal thumb in the preoperative assessment and the outcome of the performed surgical procedures on patients with triphalangeal thumbs.

Preoperative assessment and classification

The first focus of preoperative assessment was on the presence and position of the epiphyses of the first metacarpal. **Chapter two** reviews the different positions of the epiphyses and their relation with the length of the first metacarpal in triphalangeal thumb. In **Chapter three** the length of the first metacarpal is analyzed in relation to the different types of triphalangeal thumbs and the position of the epiphyses of the first metacarpal. In **Chapter four** intrinsic muscle strength of al the different types of triphalangeal thumb is measured individually. In **Chapter five** eleven cases of triplication or triplicated thumb combined with triphalangism are reported. **Chapter six** describes an adjusted classification that allows classification of all different types of triphalangeal thumb associated with radial polydactyly.

Treatment and long term follow up

The second part of the thesis is focused on clinical outcome and daily use of persons with a triphalangeal thumb. **Chapter seven** discusses clinical outcome of two different treatment strategies on removal of the extra phalanx in less complex triphalangeal thumb. **Chapter eight** discusses operative treatment for the specific types of triphalangeal thumb and addresses the possible procedures regarding their outcome. In **Chapter nine** the function of adults with untreated triphalangeal thumbs are evaluated and compared to the normal population.

Chapter ten includes the general discussion and future perspectives. In **Chapter eleven** a couple of years of research is summarized into a few pages of text.

UPDATE OF PRESENT LITERATURE

Although concepts of preoperative assessment and treatment strategies have not been altered in the past years, an update of published clinical literature is given. As the genetic research of triphalangeal thumb is not the scope of this thesis, publications regarding this topic are not discussed here.

Afshar in 2011 discussed the origin of the five fingered hand, considering it a variant of mirror hand anomaly rather than a four fingered hand with a triphalangeal thumb, not further discussing assessment or treatment [44]

Several case reports were published [45, 46]; Gosk presented a case of a hand with eight fingers of which two had triphalangeal components. Ozturk (2012) corrected a radial polydactyly with triphalangeal component, by transposing the radial interphalangeal joint to the more-developed ulnar ray. Hereby creating a stable interphalangeal joint with adequate function and opposition. In a 16 years girl with triphalangeal thumb a partial resection of the extra phalanx with correction of the angle and arthrodesis of the distal interphalangeal joint was performed using a compression screw with good result. An useful addition to the arthrodesis technique however only possible in the more mature patients because of the size of the screw [47].

This thesis contains publications of which some have been published some years back, to our mind the opinions and conclusions of these publications are still valid and useful in present daily practice.

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Preoperative assessment and classification

TWO

The Effect of the Epiphyseal Growth Plate on Length of the First Metacarpal in Triphalangeal Thumb

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ABSTRACT

Purpose

Triphalangeal thumb (TPT) is characterized as a congenital difference of the thumb with an extra phalanx. Additional length of the thumb is mostly attributed to the extra phalanx. The influence of the position of the epiphyseal plates on growth in congenital hand anomalies is unclear. The purpose of this article is to compare the length of the first metacarpal in triphalangeal thumbs with the length of the same bone in normal thumbs, and to investigate the influence of the often aberrant metacarpal epiphyseal plates on the length of the triphalangeal thumb metacarpal during growth.

Methods

Positions of the epiphyseal plate and relative length of the first metacarpal were examined retrospectively in 37 TPT patients. Ratios of the measurements were calculated ($^{\text{Metacarpal II}}/_{\text{Metacarpal I}}$) and compared to a normal population. Subjects were divided in to three groups by position of epiphyseal plate (double, proximal and distal).

Results

The distally placed growth plate was the most common variety in 52%, the proximal position in 33% and double epiphyseal plates in 15%. All ratios of TPT patients were significantly smaller compared to a normal population, indicating a longer first metacarpal in this condition. First metacarpals in TPT with double epiphyses grew disproportionately more than in a normal population, while those with distal epiphyses grew disproportionately less than normal. First metacarpals with proximal epiphyses grew with the same rate as normal first metacarpals.

Conclusions

In this study population, the most common growth plate location was distal, whereas the most common in the normal population is proximal. The different positions of the epiphyseal plates correlate with difference in growth in patients with TPT.

INTRODUCTION

Triphalangeal thumb (TPT) is a condition with an additional phalanx of the thumb with a general incidence of 1 in 25.000 [1]. The additional length is mostly a result of the extra phalanx. However the distal phalanx is also mentioned to be of greater length then the other digits [2]. Length and growth potential of the first metacarpal are not well documented in TPT patients. In case of a five-fingered hand, a type of TPT, the first metacarpal is known to contribute to the additional length [3].

The growth of normal bones depends on epiphyseal growth plates. In the normal first metacarpal the epiphyseal plate is positioned at the proximal end. In the metacarpals II-V the epiphyseal plate is positioned at the distal end [4]. Deviant positions of the epiphyseal plates have been described earlier in the normal population [5, 6] (Figure 1). Incidences of double epiphyseal plates in the first metacarpal, in which a true epiphyseal plate is placed proximally and distally, are mentioned in a range from 1 to 50% [5-8]. For Silver syndrome and hyperthyroidism a higher frequency of double epiphyseal plates is mentioned [7, 9]. Double or deviant placed epiphyseal plates do not influence the definitive length of the metacarpals in a normal population [6, 10].

In patients with TPT, total length of the thumb (including the first metacarpal), is an important factor in functional outcome [11]. The additional length caused by the extra phalanx can be adjusted through the removal of the total extra phalanx. Furthermore the middle phalanx and distal phalanx can be adjusted to correct clinodactyly in combination with shortening if necessary [12-14]. In non-opposable thumbs a rotation-abduction osteotomy can be performed on the first metacarpal possibly with an opponensplasty.

If the position of the epiphyseal growth plate influences the final length of the first metacarpal we might have to consider adjusting the amount of reduction at the primary operation of the first metacarpal. The purpose of this article is to compare the length of the first metacarpal in triphalangeal thumbs with the length of the same bone in normal thumbs, and to investigate the influence of the often aberrant metacarpal epiphyseal plates on the length of the triphalangeal thumb metacarpal during growth.

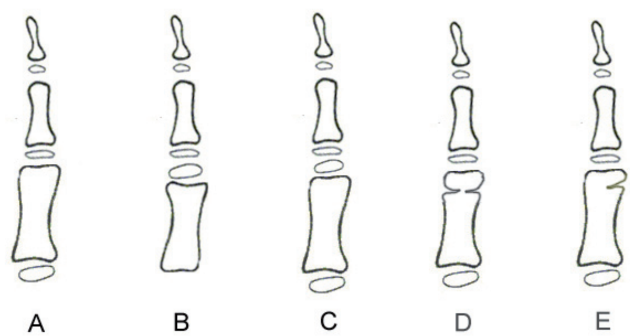

Figure 1. Schematic drawing of the different positions of the epiphyseal plates of the thumb: A) normal or proximal, B) distal, C) double, D) proximal with pseudo-epiphysis distal, E) proximal with notch distal.

MATERIAL AND METHODS

Charts of patients with Triphalangeal Thumb at the Erasmus Medical Center in Rotterdam in the period 1982 until 2003 were reviewed retrospectively. Most of the included subjects have a dominant inherited 7q36 deletion. This deletion is specific for TPT [15, 16]. Subjects with operations on first or second metacarpal, bone growth effecting disorders, index ray abnormality on x-rays, non-assessable x-rays or incomplete data were excluded. X-rays of the hands of the remaining patients where examined on number and positioning of the epiphyseal plates of the first and second metacarpal. In total 37 patients with TPT with a total of 59 affected hands were selected. In case of bilateral TPT with an equal position of epiphyseal plates, one hand was selected randomly by using a random number generator where the left hand was included if an even number was given and the right hand if an uneven number was given. After correction for a possible selection bias 42 hands were included. No differences were seen in distribution of sex and affected side. Patients with syndromes where equally spread amongst the three groups. The different types of TPT according to Wood's classification [17] were: Delta 22 (52%), Rectangular 7 (17%) and Full or Five fingered hand 13 (31%) (Table 1). In Five-fingered hands the first ray has a full extra phalanx, a narrow first web space, absent thenar muscles and is in the same plane as the fingers. The first ray can rather be considered as a digit than as a thumb. If the first ray has a trapezoid extra phalanx, a narrowed web space, hypoplastic or absent thenar muscles but is not in the same plane as the other rays, the first ray is considered a thumb.

Table 1. Characteristics of included TPT Patients

Total group 42 hands			
Growth plate position	**Double**	**Proximal**	**Distal**
N	6	14	22
Gender			
Male	3	5	11
Female	3	9	11
Affected Hand			
Left	3	5	11
Right	3	9	11
Shape extra phalanx1			
Delta	4 / 66%	10 /71%	8/ 36%
Rectangular	1 / 17%	3 / 21%	3 /14%
Full/ Five fingered hand	1 / 17%	1 / 8%	11 / 50 %
Syndromal2	1	1	2

[1] Classification according to Wood
[2] Poland syndrome (3x), Smith-Lemli-Opitz syndrome

After examination of the x-rays patients where divided into three groups: double, proximally placed or distally placed epiphyseal plates at the first metacarpal. Six patients had a double epiphyseal plate at the first metacarpal, fourteen at the proximal end and twenty-two at the distal end. Pseudo-epiphyses as well as notches were recorded but not taken into account because they do not contribute to growth in a normal first metacarpal [6, 10].

The first and second metacarpal of the affected hand were measured. The maximum length of the metacarpal was measured using the technique described by Garn [18]. (Figure 2) Absolute length could not be measured or calculated because of the unknown magnification factor of individual x-rays. To be able to compare the hands, measurements of the second metacarpal 'length' were divided by the 'length' of the first metacarpal (Metacarpal II/ Metacarpal I), thus creating a ratio. Garn described normal values of all hand bones of a population from the age of 2 to 18 years [18], Gefferth performed measurements in a population under the age of 15 months [19]. Ratios of the length of the second and first metacarpal were calculated of the measurements of the normal population as described by Garn and Gefferth (see figure 3). These data of the normal population were considered as a reference value for this study. For the age of 1 month to 2 years Gefferth's data was used and for the included subjects older than 2 year Garns data. Garn provided specific data for each year for 2 to 18 year whereas Gefferth used age periods of three years for the age from 2 to 18. The values are normalized to the values of Garn and Gefferth i.e. the normal values of Garn or Gefferth are subtracted from subject values on that specific age point. (See Table 2 and 3) Time of measurement varied because of the retrospective setting of this study.

The acquired data were analyzed for each individual subject over time, creating a longitudinal model, using a linear regression model with random slope and random intercept and Student's T-test. Data were compared between groups and with normal values on measured age points.

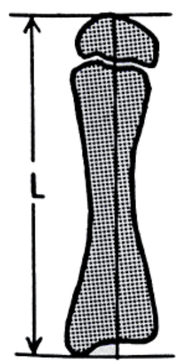

Figure 2. Measurement of the length of the metacarpal bone according to Garn. Length measurements made at right angles to the long axis and including the epiphyses, when separate.

Table 2: Measurements of first and second metacarpal published by Garn (1972). (m month, y year, MC1 first metacarpal and MC2 second metacarpal)

	Age (y)	Length MC1 (mm)	Length MC2 (mm)	Ratio (MC2/MC1)
Male				
	2	19.6	30.6	1.561
	3	22.0	34.5	1.568
	4	24.1	37.9	1.573
	5	26.7	41.6	1.558
	6	29.0	44.9	1.548
	7	30.9	47.7	1.544
	8	32.7	50.2	1.535
	9	34.4	52.6	1.529
	10	36.3	55.0	1.515
	11	38.2	57.3	1.500
	12	40.2	60.6	1.507
	13	42.5	63.3	1.489
	14	45.1	67.1	1.488
	15	47.6	70.6	1.483
	16	48.8	73.2	1.500
	17	49.5	74.2	1.499
	18	49.4	73.9	1.496
	Adults	49.6	73.7	1.486
Female				
	2	19.9.	31.3	1.573
	3	22.7	35.2	1.551
	4	24.8	38.2	1.540
	5	27.3	42.4	1.546
	6	29.6	45.6	1.541
	7	31.5	48.1	1.527
	8	33.5	51.2	1.528
	9	34.8	52.6	1.511
	10	37.4	56.6	1.513
	11	39.7	59.9	1.509
	12	42.0	63.2	1.504
	13	43.8	66.2	1.511
	14	44.4	67.4	1.518
	15	45.3	68.1	1.503
	16	45.0	68.6	1.524
	17	45.0	68.9	1.531
	18	44.6	67.5	1.513
	Adults	44.2	66.9	1.513

Table 3: Measurements of first and second metacarpal published by Gefferth (1972). (m month, y year, MC1 first metacarpal and MC2 second metacarpal)

	Age	Length MC1 (mm)	Length MC2 (mm)	Ratio (MC2/MC1)
Male				
	Newborn	9.4	14.5	1.543
	1-2 mo	10.6	16.4	1.547
	3-4 mo	11.9	17.0	1.429
	5-6 mo	12.3	19.0	1.545
	8-10 mo	13.5	20.3	1.504
	1 y	14.6	22.4	1.534
Female				
	Newborn	9.5	14.8	1.558
	1-2 mo	10.3	15.6	1.515
	3-4 mo	11.4	17.8	1.561
	5-6 mo	12.6	19.5	1.548
	8-10 mo	14.0	21.7	1.550
	1 y	15.6	24.5	1.571

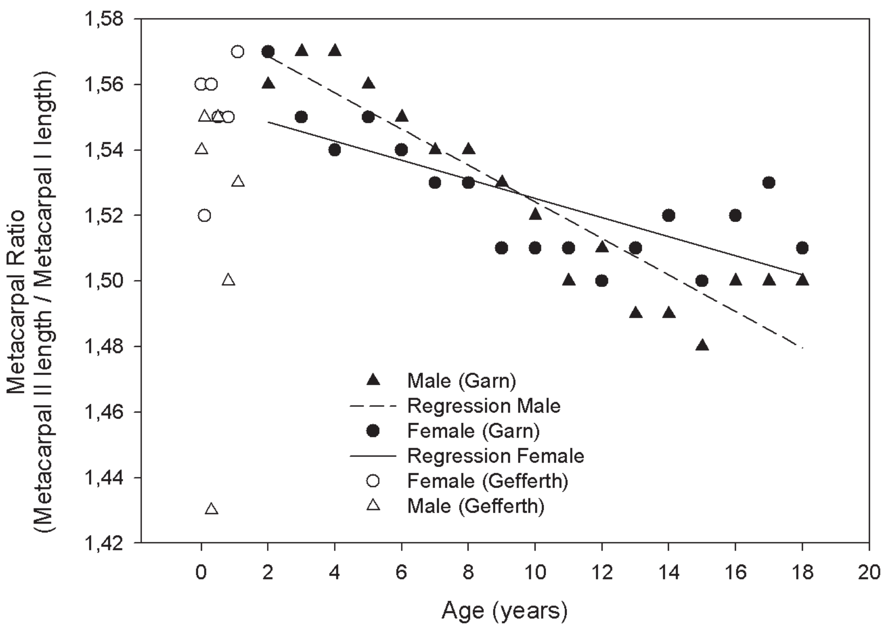

Figure 3. Ratios calculated of the measurements of first and second metacarpal provided by Garn and Gefferth.

RESULTS

The proximally placed epiphyseal plate, the position most common in the normal population, is not the most common presentation in TPT patients. The distally placed epiphyseal plate is the most common in TPT patients, i.e. 22 of 42 hands (52%). The distally placed epiphyseal plate is more commonly present in the so-called five-fingered hand. Measurements of the three different groups are plotted in figures 4A-C.

The ratios in all groups were significantly smaller then of the normal population ($p < 0.01$), in other words a longer first metacarpal. Ratios in the second group with the proximally placed growth plates, were close to the value in the normal population (according to Garn and Gefferth, see figure 4B). The regression line of this group lies parallel to the normal values showing no change of length in relation to normal values during aging (intercept –0.117 with an age factor 0.002; $p = 0.8$).

The first (double epiphyseal plate) and third group (distal epiphyseal plate) ratios have a negative value, meaning a smaller ratio than the ratio in the normal population. The regression line of the double epiphyseal plate group has a negative trend, thus during aging the ratio of the first to second metacarpal of the TPT patients decreases (intercept –0.200 with an age factor – 0.013; $p = 0.2$). This indicates an increase in length of the first metacarpal during growth in relation to the length of the first metacarpal in the normal population. Whereas the distal epiphyseal plate group has a positive trend, i.e. an increase of ratios (intercept -0.387 with an age factor 0.010; $p = 0.15$).

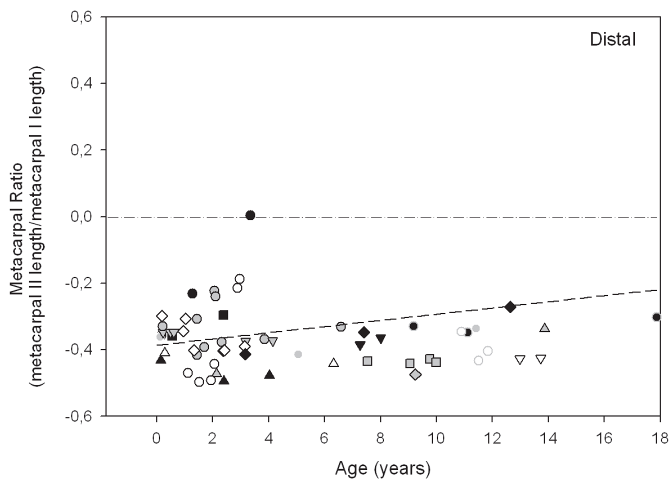

Figure 4A. Ratios and regression line plotted of the subjects with a distal epiphyseal plate. (circle, hexagon and square female, triangle male). By subtracting the normal values from the subject values, the normal values are visualized as the zero line (displayed as --------). During the growth period the additional length of the first metacarpal decreases and approaches normal length of first metacarpal if assumed that the second metacarpal is unaltered.

This indicates that the first metacarpal loses some of the additional length during growth compared to the length of the first metacarpal in a normal population. No significant gender differences were seen in the calculated ratios. The regression line of both male and female followed the regression line provided by Garn [18].

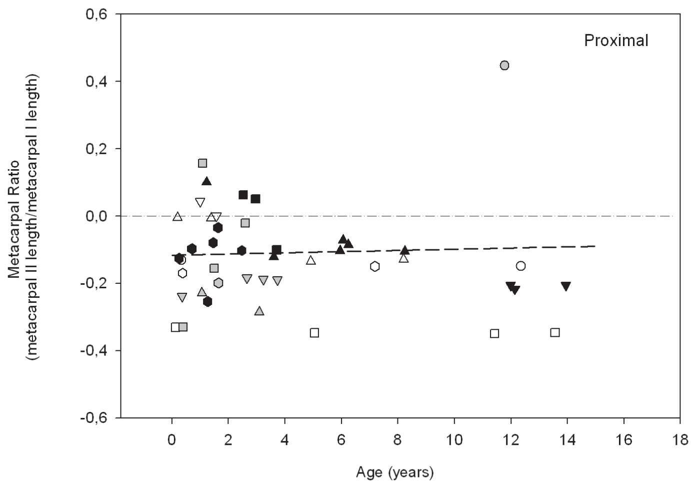

Figure 4B. Ratios and regression line plotted of the subjects with a proximal epiphyseal plate. (circle female, triangle, square and diamond male). By subtracting the normal values from the subject values, the normal values are visualized as the zero line (displayed as --------). During the growth period additional length of the first metacarpal compared to the normal value remains the same.

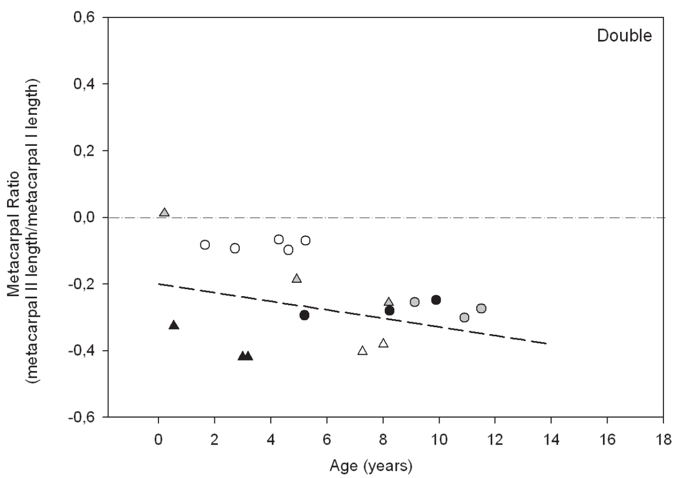

Figure 4C. Ratios and regression line plotted of the subjects with double epiphyseal plates. (circle female, triangle male). By subtracting the normal values from the subject values, the normal values are visualized as the zero line (displayed as --------). During the growth period the additional length of the first metacarpal increases compared to the normal values. If assumed that the second metacarpal is unaltered.

DISCUSSION

Retrospectively metacarpal length was measured longitudinal in 37 patients (42 hands) on hand X-rays expressed as ratios and compared with values of the normal population [18] [19]. The ratios calculated have the disadvantage when compared to absolute measurements, that the length of the second metacarpal in congenital hands is assumed unaltered. Length of the second metacarpal isn't altered by positioning of the epiphyseal plate [6], but length of the metacarpal and phalanges may be altered in syndromes. Reduction of metacarpal length has been described in Turner's syndrome, Down syndrome and idiopathic pseudo-Hypoparathyroidism [18]. Hypoplasia as well as hyperplasia of a complete ray haven been reported in Holt-Oram Syndrome [20]. In our study however the second metacarpal was assumed to be of normal length.The incidence of a double placed epiphyseal plate has been described to be ranging from 1 – 50 % [5-7]. This includes the report of Borovansky and Hnevkorsky [8] who found a 50% incidence, whereas others reported incidences between 1 and 3%. The criteria used by Borovansky et al and their studied population are unknown. All other studies were performed in normal populations. Wood suggested that congenital anomalies had a higher incidence of double epiphyseal plates in the first metacarpal. In our group we found 7.6 % double epiphyseal plates. Other than Borovansky's results we can conclude that TPT has a higher incidence of double epiphyseal plates compared to a normal population like Wood reported. The most common position for the growth plate of the first metacarpal is proximal; in our TPT patients 52% (22 of 42 patients) had a distally placed growth plate.

The position of the epiphyseal plate of the first metacarpal differs by type of TPT; distally placed epiphyseal plates occur more often with a full shaped extra phalanx or five fingered hand, as described by Flatt [21]. Flatt stated that the distally placed epiphyseal plate is only present in the non opposable thumb or five fingered hand. In our series the distally placed epiphyseal plate occurs also in the other variations of TPT in contrast to Flatt's description. Therefore it cannot attribute alone to the resemblance of an extra finger instead of a thumb.

Epiphyseal plates influence growth of bones; the position is of no effect in a normal population [6]. Regression lines of the ratios of the three different groups with TPT demonstrate a significant difference ($p < 0.01$) to normal values. In relation to each other the regression lines demonstrate a significant difference ($p < 0.01$) except for the relation between the double and proximally placed epiphyseal plate position ($p < 0.5$). Summarizing one could assume that position of the epiphyseal plate in TPT influences length.

The age dependent factor of the calculated regression lines indicates the amount of influence on growth. If the regression line approaches the regression line of the reference values ('zero-line') this represents a disproportionately less than normal growth of the first metacarpal. Which results in a decrease of the additional length during the growth period, if growth of the second metacarpal is assumed unaltered. This applies for the distally placed epiphyseal plate with an age dependent factor 0.01. If the regression line follows a negative path a disproportionately more than normal growth of the first metacarpal is present, thus the additional length of the first metacarpal will increase, as seen in the double epiphyseal plate group (age dependent factor -0.01) The age dependent factor for the proximal group follows the course of the reference values meaning a growth in the same rate as normal metacarpals, the additional length of the first metacarpal in this group remains the same during the growth period.

The above mentioned findings are applicable in the types of TPT were a bony reduction is required, namely the rectangular and full type. In cases with a double epiphyseal plate, with the positive trend, one must observe that the growth of the first metacarpal will increase over the years. In cases with a double epiphyseal plate an epiphyseodesis is worth considering, to determine exact timing and on which growth plate (distal or proximal) the epiphyseodesis should be performed, further research is necessary. So reduction of length when performing an osteotomy in cases with a double epiphyseal plate, must be larger then in cases with a proximal epiphyseal plate. For cases with a proximal epiphyseal reduction should be just enough to obtain the required length of the total thumb at operation. For cases with a distal epiphyseal plate the metacarpal reduction at operation should be more than the required length of the total thumb at operation, especially in the young patient.

We conclude that in all patients with TPT the ratio ($^{\text{Metacarpal II}}/_{\text{Metacarpal I}}$) is smaller compared to a normal population. In other words the length of the first metacarpal in TPT is longer compared to a normal population. Additional length of the thumb in TPT cannot be ascribed only to the extra phalanx but the first metacarpal participates as well. Length of the first metacarpal in TPT, in contrast to normal hands, is associated with by position of the epiphyseal plate.

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THREE

Implications for treatment of variations in length of the first metacarpal in different types of triphalangeal thumb

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SUMMARY

Abnormal function of the triphalangeal thumb is partially due to its extra length. This additional length is caused not only by the extra phalanx, but also by differences in the length of the first metacarpal. This study investigated whether the additional length of the first metacarpal is influenced by the growth plate location alone, or also by the type of triphalangeal thumb. In total, 59 hands in 37 patients with a triphalangeal thumb were examined for thumb type (delta 31, trapezoid nine and full type 19), growth plate location, and relative length of the first metacarpal. The first metacarpal in all three triphalangeal thumb types was significantly longer than that in a normal population. The length of the first metacarpal was related to the site of the growth plate. No direct influence of the triphalangeal thumb type on the length was found. These data suggest that a corrective procedure on the first metacarpal must be considered in all types of triphalangeal thumb.

INTRODUCTION

Triphalangeal thumb is a congenital anomaly of the thumb named after the extra phalanx in the thumb. The joints, ligaments, muscles, and tendons of the first ray may also show abnormalities. Wood (1976) classified the triphalangeal thumb according to the shape of the extra phalanx, distinguishing the delta type (a triangular shape), the trapezoid type (a rectangular shape) and the full type (a fully developed extra phalanx). These three types are illustrated in Figure 1.

Length is an important factor for adequate thumb function, since both increased length and shortness of the thumb makes it less suitable for use in daily life. In the triphalangeal thumb the length is increased as a result of an additional phalanx. However, besides the additional length caused by the extra phalanx, the metacarpal may also be longer. In the full type of triphalangeal thumb, the first metacarpal is longer compared to the first metacarpal in a normal population (Zguricas 1997). Furthermore, the length of the first metacarpal in the triphalangeal thumb is influenced by the position of the growth plate, independent of the type of thumb (Zuidam et al., 2006). More specifically, if the growth plate of the first metacarpal is placed distally or on both sides of the metacarpal, the metacarpal is longer compared to that in a normal population.

Although in current clinical practice the choice of intervention is generally based on the type of triphalangeal thumb, it is unclear whether there is a relation between the type of triphalangeal thumb and the length of the first metacarpal (with the exception of the full type of triphalangeal thumb). In standard operative treatment of the triphalangeal thumb, the first metacarpal is generally only shortened when the thumb is classified as a full-type triphalangeal thumb. In these cases the additional

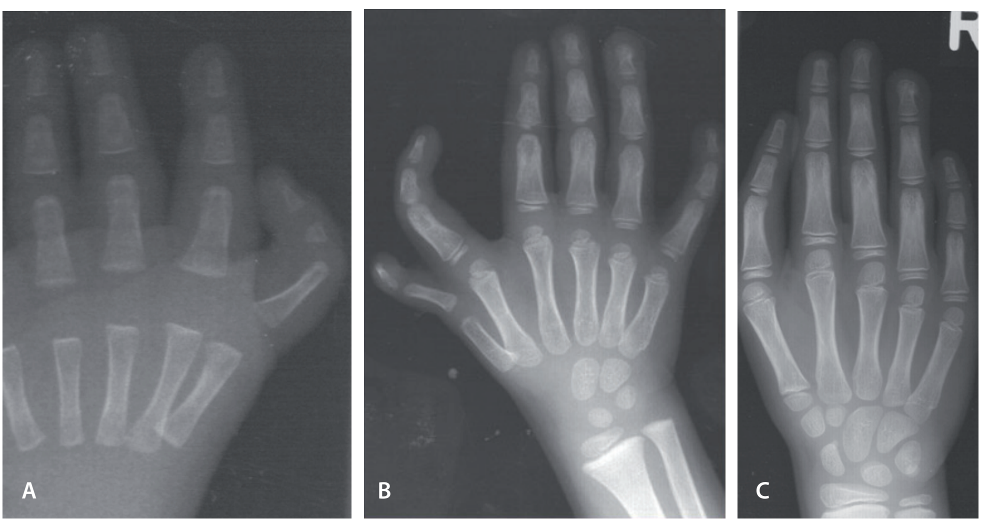

Figure 1. Radiographs illustrating the three types of triphalangeal thumb: A, the delta type (triangular shaped); B, the trapezoid type (rectangular shaped); and C, the full type (fully developed extra phalanx).

length of the first metacarpal is reduced by pollicization or by a combination of shortening of the extra phalanx and the first metacarpal. In the delta or the trapezoid types of triphalangeal thumb, length reduction is only obtained by removal or shortening of the extra phalanx and no operative procedures are done for any possible additional length of the first metacarpal (Upton and Shoen, 2000). The senior author has described shortening of the first metacarpal for the trapezoid type previously (Hovius et al., 2004). Looking at any relation between the type of triphalangeal thumb and length of the first metacarpal may indicate whether shortening of the first metacarpal should also be considered for the delta and the trapezoid type of triphalangeal thumb.

The aim of this study was to determine the influence of the different types of triphalangeal thumb on the length of the first metacarpal, irrespective of the location of the growth plate.

PATIENTS AND METHODS

Patient Sample

In the period 1982 to 2003, 51 patients with a triphalangeal thumb attended our Centre. Of these, 37 patients with 59 affected hands) were included in this study: 31 with a delta-shaped extra phalanx, nine with a trapezoid, and 19 with a full-type triphalangeal thumb (Table 1). Hands were classified as full-type triphalangeal thumb only if the first ray had a full extra phalanx, a narrow first web space, absent thenar muscles, and if the thumb was positioned in the same plane as the fingers, the so-called 'five-fingered hand' (Kozin, 2005). In Wood's classification, no distinction is made between an opposable and non-opposable thumb. In the present study, none of the full-type thumbs had opposition.

Patients were excluded if they had had operations on the first or second metacarpal, had disorders affecting bone growth, had abnormalities of the second metacarpal on radiographs, had non-assessable radiographs, or had incomplete data. Also excluded were patients with a condition which could influence the length of the first or second metacarpal, e.g. Turner's syndrome, pseudohypoparathyroidism, and Holt-Oram syndrome (Poznanski et al., 1972).

Table 1. Characteristics of the triphalangeal thumb in the study patients

Shape of extra phalanx	Delta	Trapezoid	Full	Total
N	31	9	19	59
Growth plate position				
Double	4	1	1	6
Distal	11	6	17	34
Proximal	16	2	1	19

Procedure and Measurements

The methods of measurement have been described previously (Zuidam et al., 2006) and are only briefly summarized here. Radiographs of each patient were assessed retrospectively and the location of the epiphyseal growth plate was identified as 1) proximal, 2) distal, or 3) double. The maximum length of the first and second metacarpals, including the growth plates, was measured perpendicular to the metacarpal axis (Garn et al., 1972). The absolute length of the first and second metacarpals could not be measured because of the unknown magnification factor on the radiographs. Therefore, we created a ratio by dividing the second metacarpal length by the first metacarpal length as measured on the images. Ratios calculated from a normal population were used as normal values (Garn et al., 1972; Gefferth, 1972). The measured at the corresponding age of the patient normal values were subtracted from the patients values, thereby setting the normal values at zero. Finally, the length of the first metacarpal in the triphalangeal thumb was calculated as a percentage of that of the normal population.

Statistical Analysis

Data were analysed as frequencies and percentages, or means and ranges. To detect possible differences between sub-groups, Student's t-test, one-way ANOVA and the Chi-squared tests were used. To study relationships between variables, Spearman's correlation was used. To determine if the type of triphalangeal thumb directly affects the first metacarpal length, or whether the additional length is an indirect influence as a result of growth plate location, a univariate analysis of variance was performed. Two-tailed probabilities $p < 0.05$ were considered statistically significant.

RESULTS

The length ratios ($^{\text{Index metacarpal}}/_{\text{Thumb metacarpal}}$) were significantly smaller for all types of triphalangeal thumb compared with normal values ($p<0.01$), indicating a longer first metacarpal in all types of triphalangeal thumb than in a normal population (Figure 2). Compared with the normal population, the mean additional length of the first metacarpal in the full type (-0.36, range -0.12 to -0.48) was similar to that in the trapezoid type (-0.35, range -0.26 to -0.45). The difference in ratio for the delta type was -0.22 (range 0.45 to -0.53).

Expressing the length of the first metacarpal in the triphalangeal thumb as a percentage of the length of this bone in a normal population indicated an increased length up to 154% for all three types (Figure 3). The mean increase for the delta type was 118% (range 77-154%), for the trapezoid type 131% (range 121-140%) and for the full type 131% (range 111-144%). When further subdividing the triphalangeal thumb types based on the position of the growth plate (distal, proximal and double), the additional length of the first metacarpal with a delta-shaped extra phalanx showed a significant difference for the different positions of the growth plate. If the growth plate was placed proximally, the first metacarpal had a mean length of 108% (range 77-120%), the distally-placed growth plate had

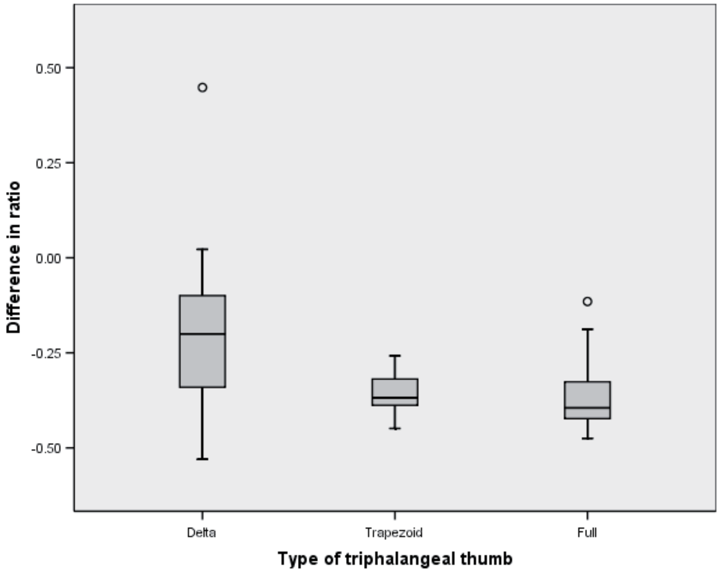

Figure 2. Box plots of the average of the differences in the metacarpal ratios triphalangeal and normal thumbs for each type of triphalangeal thumb. The plots show the median, upper and lower quartiles (box) and the lowest and highest observations (whiskers) for the three types of triphalangeal thumb. A lower ratio indicates a greater length of the first metacarpal.

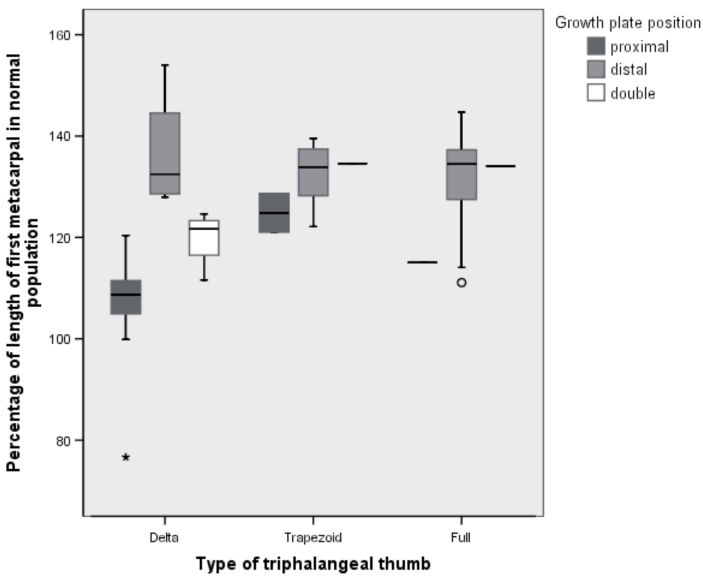

Figure 3. Box plots of the percentages of the first metacarpal length in triphalangeal thumb compared with the length of the first metacarpal in the normal population. The plots show the median, upper and lower quartiles and the lowest and highest observations for the three types of triphalangeal thumb, subdivided into groups according to the position of the growth plate. A higher percentage indicates a longer length of the first metacarpal.

a mean length of 136% (128-154%) and the mean for double growth plates was 120% (112-125%). No significant differences in length were found between the different growth plate positions for the trapezoid type and for the full-type triphalangeal thumb; in these small sub-groups.

The type of triphalangeal thumb, the growth plate position, and the length of the first metacarpal all showed significant correlations (Table 2). The position of the growth plate had a significant influence on the length ratio (p=0.02), independent of the type of triphalangeal thumb. The type of triphalangeal thumb, irrespective of growth plate location, had no significant influence on the length of the first metacarpal (p=0.08).

Table 2. Relation between type of triphalangeal thumb (TPT), growth plate position and additional length of the first metacarpal

Spearman's correlation[1]			
	Type of TPT	**Growth plate position**	**Additional length of first metacarpal**
Type of TPT	-	0.33 (p = 0.01)	-0.49 (p < 0.001)
Growth plate position	-	-	-0.56 (p < 0.001)
Chi-square test	Type of TPT		
	Growth plate position		p = 0.005
Univariate analysis of variance on additional length of first metacarpal			
Type of TPT	p = 0.083		
Growth plate position	p = 0.020		

[1]Spearman's correlations have a negative effect on the ratio as a result of the calculated difference in the metacarpal ratio which gives a negative result if the first metacarpal is longer in triphalangeal thumb.

DISCUSSION

We found a significantly longer length of the first metacarpal in all types of triphalangeal thumb compared with a normal population. No statistically significant relationship was found between the type of triphalangeal thumb and the length of the first metacarpal (p=0.08) It is possible that a larger sample size might indicate that the additional length of the first metacarpal is related to the growth plate location, and also to the type of triphalangeal thumb, at present this must remain a speculation

Although the growth plate of the first metacarpal is normally located proximally, abnormal positioning of the growth plate is not uncommon in patients with triphalangeal thumb. In the present study, a distally-placed epiphyseal plate was present in 89% in the full type of triphalangeal thumb, in 35% in the delta type, and in 67% in the trapezoid type. We have previously reported the influence of the location of the growth plate on the length of the first metacarpal (Zuidam et al., 2006). In our earlier study we found that the first metacarpal in triphalangeal thumbs was significantly longer compared with a normal population for all positions of the growth plate. However, metacarpals with a double or distally placed epiphyseal plate were longer. The current study has demonstrated a significant relationship between the type of triphalangeal thumb and the location of the growth plate

The extra length of the first metacarpal in comparison with the normal population in all types of triphalangeal thumb may affect the type of surgery. Mean values of the different types of triphalangeal thumb indicate an additional length of up to 54% over a a normal population, indicating a need to reduce the metacarpal length. However, it should be noted that, in the present study, one patient of the 31 in the delta group had a shorter first metacarpal (77% of the normal value). This finding, as well as the relatively large variation within the different types, indicates the importance of treating each patient on based on individual measurements of metacarpal length.

A limitation of this study is that metacarpal length could only be measured indirectly because of the unknown magnification factor in the individual radiographs. Ratios between the first and second metacarpal were calculated for each hand, assuming that the length of the second metacarpal in hands with a triphalangeal thumb is unaltered. To our knowledge, there are no reports on the influence of the triphalangeal thumb on the length of the other metacarpals or phalanges, other than those of the first ray, suggesting that this is a valid assumption.

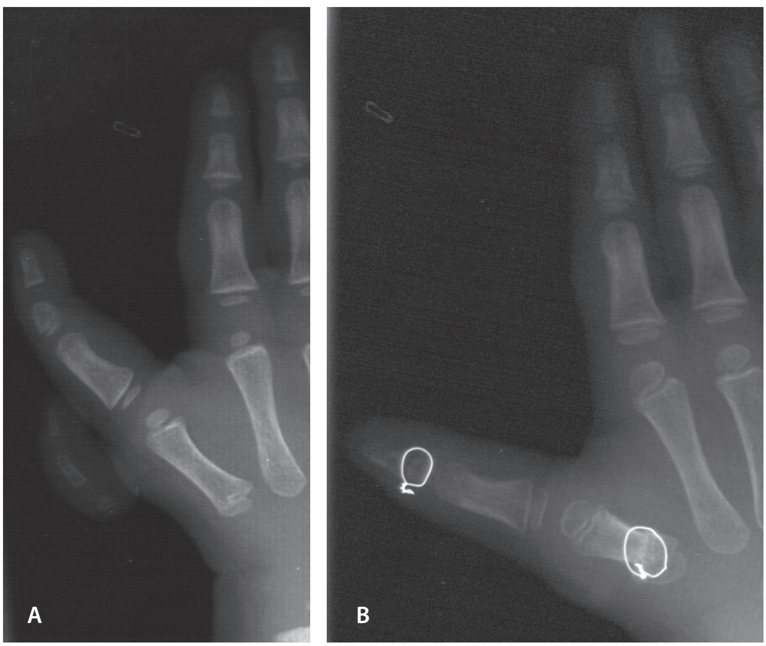

Figure 4. A; Preoperative radiographs of a patient with trapezoid type triphalangeal thumb and radial polydactyly. B; Postoperative radiographs after correction of the triphalangeal thumb by removal of the distal interphalangeal joint and extra phalanx and fixing the proximal interphalangeal joint to the distal phalanx (Peimer 1985). To reduce additional length of the first metacarpal a reduction arthrodesis of the first metacarpal has been done. The additional hypoplastic radial ray was removed in the same session.

Reduction osteotomy of the first metacarpal in addition to removal of the extra phalanx (Figure 4) is not often done in the triphalangeal thumb, particularly in the delta type. In patients with a full-type triphalangeal thumb, normal operative practice includes reduction of the first metacarpal either as part of the pollicization or by rotation-abduction-reduction osteotomy of the first metacarpal. However, since the present study reported an increased length of the first metacarpal in all three types of triphalangeal thumb, we suggest that a reduction osteotomy should be considered in all cases, depending on individual measurement of the length of the first metacarpal. Apart from a reduction osteotomy of the first metacarpal, an epiphyseodesis of the distal epiphyseal plate can be used to prevent extra growth in patients in whom double epiphyseal plates are present.

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FOUR

Thumb strength in the triphalangeal thumb

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ABSTRACT

Strength is regarded as normal, in patients with an opposable triphalangeal thumb. Our clinical impression is however, that intrinsic musculature is probably affected in all forms of triphalangeal thumb. Therefore we established the strength of 38 thumbs in subjects with a triphalangeal thumb. Patients were excluded if the intrinsic musculature was enhanced or if osteotomies of the first metacarpal were performed. On average, strength was statistically significant diminished for all thumb functions, up to 63% for opposition strength. Strength for the power grip was on average 70%. As shown by this study, strength of the musculature of the thumb is affected in all types of triphalangeal thumb. Although strength of the thumb is diminished, for the investigated group it is apparently sufficient in daily life, as they did not seek surgical enhancement. However reconstructive procedures enhancing intrinsic musculature must be considered in all types of triphalangeal thumb.

INTRODUCTION

Triphalangeal thumb is a congenital hand anomaly with a heterogenic occurrence, ranging from less complex with only an extra phalanx at the interphalangeal joint to a five-fingered hand with absent thenar musculature and therefore no opposition (Hovius et al., 2004, Upton and Shoen, 2000). In Buck-Gramcko's extensive classification for triphalangeal thumb, absent or hypoplastic thenar musculature is only assigned to the types with the larger transitional extra phalanges and to the long rectangular middle phalanx (Buck-Gramcko, 2002), which is comparable with Wood's Full type (Wood, 1976). In all others types no aberrant thenar musculature is mentioned, suggesting a normal thenar musculature is present.

In literature no involvement of intrinsic musculature in the 'Delta' type triphalangeal thumb, 'Trapezoid' shaped extra phalanx, 'short rudimentary phalanx' or 'short triangular bone' has been described. However our clinical impression was that intrinsic musculature is probably affected in more types of triphalangeal thumb. Therefore we established the strength of specific thumb functions in subjects with delta and trapezoid types of triphalangeal thumb.

PATIENTS AND METHODS

Patients Sample Characteristics

Between 1982 and 2006 a total of 60 patients with a triphalangeal thumb visited the Erasmus Medical Centre. Subjects with disorders affecting general strength, peripheral nerve injuries or other disorders disturbing hand strength were excluded. Secondly subjects who were not able to perform the required motion, as a result of joint limitations, stiffness or laxity were also excluded. Thirdly subjects who had been treated with a pollicisation, a first metacarpal reduction osteotomy, an opponens plasty, or any other form of tendon transfer to enhanced thumb function or reinsertion of intrinsic musculature were excluded as well. Lastly patients under the age of four years were excluded, because of the impossibility to perform the strength tests. Of the remaining 23 included patients, there were 7 male and 16 female, aged 7 to 64 years (mean 34 years); three persons were left dominant (Table 1).

Methods

Medical Data

All patient files were retrieved and patient characteristics, dominance, type of triphalangeal thumb, and surgical data were scored. Patients were classified by type of triphalangeal thumb using Wood's classification (Wood, 1976) of the shape of the extra phalanx: Delta, Trapezoid and Full type triphalangeal thumb in addition, we used Buck Gramcko's classification of the treatment options (Buck-Gramcko, 2002).

Table 1. Characteristics of patients with triphalangeal thumb operated or non-operated, without tendon transfer or reinsertion of intrinsic musculature

N = 23 H=38			
Gender			
Male			7 (30%)
Female			16 (70%)
Dominance			
Right			20 (87%)
Left			3 (13%)
Type of triphalangeal thumb			
Wood [5]		Buck-Gramcko [6]	
Delta	or	Rudimentary	14 (37%)
Trapezoid	or	Short Triangular	18 (47%)
		Larger Transitional	6 (16%)
Full	or	Long rectangular	0 (0%)
		Hypoplastic	0 (0%)
		Associated with radial duplication	5 (13%)
Treatment			
None			24
DEL			6
DIPRAD			8
PIPRAD			0

DEL: removal of extra phalanx if combined with reconstruction of the collateral ligaments, DIPRAD: distal interphalangeal joint combined reduction osteotomy and arthrodesis according to the technique described by Peimer for triphalangeal thumb [8], PIPRAD: proximal interphalangeal combined reduction osteotomy and arthrodesis ,variation of above mentioned technique.

Strength Measurements

Intrinsic hand strength was measured using the Rotterdam Intrinsic Hand Myometer (RIHM). The RIHM has proven to be a reliable instrument for strength measurements of the individual thumb and finger strength in healthy controls (Molenaar et al., 2008) and a number of patients groups (Selles et al., 2006) (Schreuders et al., 2004). Individual measurements were performed for the strength of opposition, flexion of the metacarpophalangeal joint of the thumb, anteposition, and radial abduction of the index finger. The mean of three repeated measurements was registered.

Maximal isometric grip, pinch and tip strength was measured using the Jamar Dynamometer and pinch gauge (Sammons Preston, Bolingbrook, IL, USA). Measurements with the Jamar dynamometer were performed with the handle in the second position and all persons were seated in the position suggested by the American Society of Hand Therapists, with the shoulder abducted, the elbow in 90 degrees of flexion, and the wrist in neutral position (Casanonva, 1992). The mean of three repeated measurements was registered.

Because of the wide spread in age in this group, and therefore wide variability in normal values, measurements for both instruments were presented in percentages of the age and sex specific reference values (1995, Mathiowetz et al., 1985, Mathiowetz et al., 1986, Molenaar et al., 2008, Schreuders and Selles, 2008)

Statistical Analysis

Data were analyzed as frequencies and percentages or means and ranges. To detect differences between the study group and the value of the normal population (100%) a one-sample T-test was used. A p-value of < 0.05 was accepted as statistically significant.

RESULTS

All strength measurements were significantly reduced when compared to age specific reference values. Mean strength for opposition was 63% (p 0,00)(SD 19, range 26 to 121). Mean strength for flexion of the metacarpophalangeal thumb was 62% (SD 16, range 26 to 97)(p 0,00). Anteposition of the thumb had a mean strength of 76% (SD 25, range 26 to 121)(p 0,00). Mean strength of radial abduction of the index finger was 88% (SD 26, range 48 to 160)(p 0,01) (Figure 1). Power grip had a mean strength of 69% (SD 33, range 22 to 142)(p 0,00). Strength of the pinch grip was 84% (SD 25, range 27 to 145)(p 0,00) and for key grip 73% (SD 19, range 21 to 107%)(p 0,00) (Figure 2). In patients which where affected unilateral (n =8), the hand with the triphalangeal thumb was compared to the non-affected hand, for all measurements the non-affected hand was stronger, although not statistically significant.

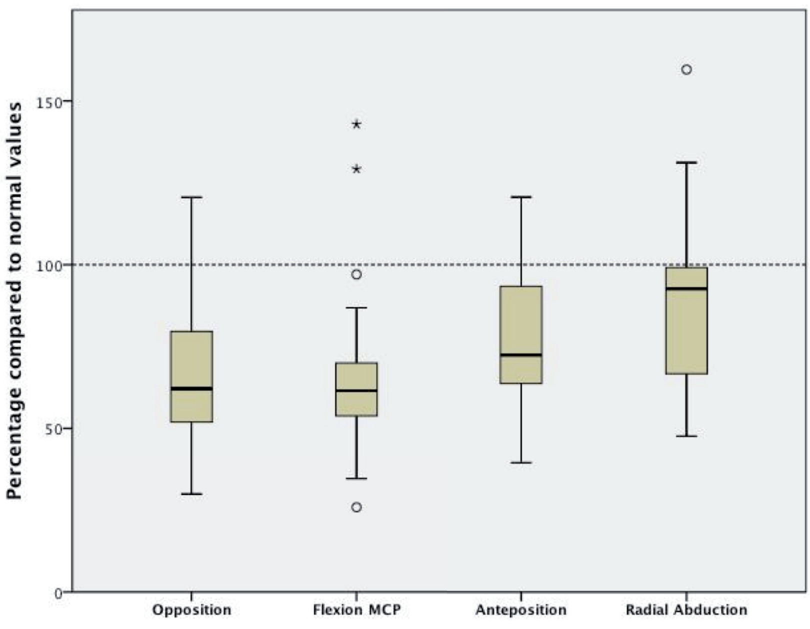

Figure 1. Boxplot of thumb strength in different directions as a percentage of the age-specific reference values. Measurements of the normal population are indicated as 100 percent (dotted line) (Molenaar et al., 2008, Schreuders and Selles, 2008). All measurements were statistically significant lower when compared to the reference values.

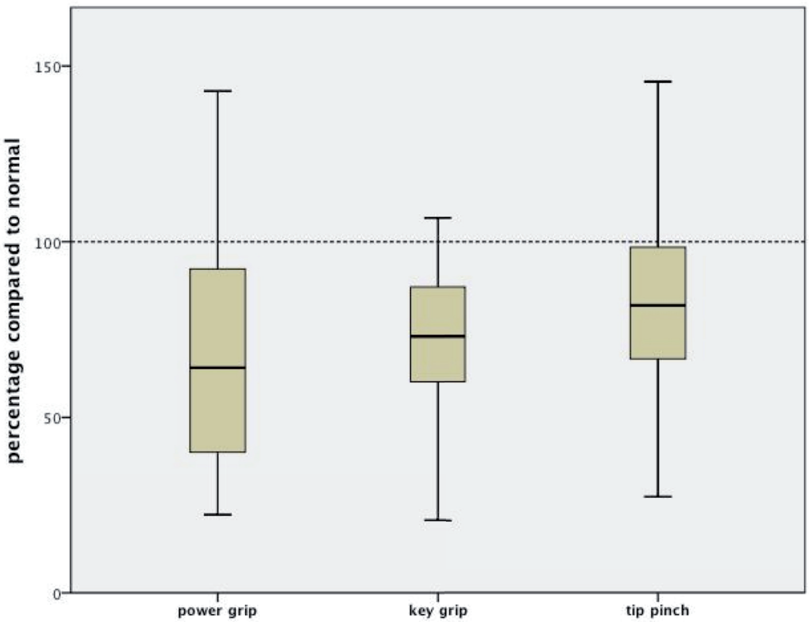

Figure 2. Boxplot of the power grip, key pinch and tip pinch as a percentage of the age-specific reference values. Measurements of the normal population are indicated as 100 percent (dotted line) (Mathiowetz et al., 1985, Mathiowetz et al., 1986). All measurements were statistically significant lower when compared to the reference values.

DISCUSSION

Our clinical impression was that strength of the thumb is affected in all types of triphalangeal thumb, besides the well known diminished strength in the five fingered hand. Therefore strength of the thumb in the 23 subjects with delta and trapezoid types of triphalangeal thumb was established. The strength of thumb and hand functions (opposition, flexion of the metacarpophalangeal joint, anteposition and radial abduction of the index finger), was significantly lower compared to a normal population. Strength of power grip, pinch and key grip was also significantly diminished.

Intrinsic hand function strength is difficult to measure; the Rotterdam Intrinsic Hand Myometer is the only capable and validated instrument for adults and children, although it has its limitations. One limitation is that is measures a function rather than a specific muscle. Another limitation of this study is the inclusion of patients who have been operated on the thumb, although no procedures enhanced or recreating intrinsic hand function were performed, alterations of the thumb have been performed. These procedures (e.g. removal of extra additional rays, correction of the extra phalanx) are not focused on enhancing muscle strength although a small effect cannot be excluded. To investigate a possible difference between the operated and non-operated group, a sub group analysis was performed. Both groups had statistically diminished strength for all measurements compared to a normal population (data not shown). Also older i.e. adults persons were included. These patients can have obtained diminished strength as a result of disuse of specific thumb functions. All adults however were affected bilaterally and the strength of both hands was diminished in all.

Triphalangeal thumb has a heterogenic presentation, from a small extra phalanx to a non opposable thumb, and everything in between. Therefore is it difficult to make classifications with clear cut-off points. Wood classified the triphalangeal thumb by the shape of the extra phalanx, Buck-Gramcko included besides the shape of the extra phalanx, also the associated duplications and the degree of development of the thumb; the hypoplastic type. The present classifications (Wood, 1976) classify the triphalangeal thumb on severity, however in both classifications the presence of absent or hypoplastic intrinsic musculature is only attributed to the more severe types (for Wood: Full type, and Buck-Gramcko: Larger transitional and long rectangular). A classification suitable for all types of triphalangeal thumb with their wide variety would probably be unusable.

This study enhances our clinical experience that the thumb function and intrinsic hand function strength is affected in all types of triphalangeal thumb. Although the strength of the different thumb functions is on average diminished in all types of triphalangeal thumb, it is apparently sufficient in daily life at least in the researched group. Persons with untreated triphalangeal thumbs probably have developed skills to perform daily life tasks with diminished strength. It is not necessary to recreate or 'enhance' intrinsic hand function in all patients with triphalangeal thumbs, although it must be considered in all the different variations and types.

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FIVE

A classification system of radial polydactyly: inclusion of triphalangeal thumb and triplication

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ABSTRACT

Purpose

Radial polydactyly is a congenital anomaly with a wide range of manifestations. Current classifications are not able to classify all different types of radial polydactyly when combined with triphalangeal components. The objective of this study was to test an adjusted classification and nomenclature that allows classification of triphalangeal components and triplication in radial polydactyly.

Methods

Patients from 1993 to 2006 with radial polydactyly (n=104) were identified from the hospital database with a total of 121 affected hands. All x-rays were carefully examined and classified according to the existing classifications for radial polydactyly and a modified classification. In the modified nomenclature Wassel's level of duplication is preserved. Type VII and VIII are assigned for partial or complete duplication of the carpal bones according to Buck-Gramcko. Triplication and triphalangeal components can be assigned to each type of radial polydactyly by suffices. Symphalangism, deviation, and hypoplasia can also be classified. Triplication on different levels of the thumb is classified by determining and including the different types of the original Wassel classification.

Results

Eighteen thumbs with triphalangeal components or triplication could not be classified according to existing classifications for radial polydactyly. After using the proposed classification, all patients could be classified.

Conclusion

We propose a modified classification that is practical and utilitarian for nomenclature of radial polydactyly and that may assist comparison of treatment outcomes and individual cases.

INTRODUCTION

Classifications are useful in guiding and evaluating surgical management and treatment. In radial polydactyly a classification based upon osseous anatomy, such as Wassel's classification [1], is most suited for planning surgical treatment. Wassel's classification, the most often used classification, is practical and based on level of duplication. The classification starts from distal to proximal, in which uneven numbers are bifid duplications whereas even numbers are complete duplications and type seven refers to duplications involved with triphalangism. While 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 [2] and Miura [3] created more subtypes for classifying triplication and triphalangism. In Woods classification, type IV is subdivided into type A, where on the level of the proximal phalanx both duplications have a triphalangeal component and type B with only a triphalangeal component on the radial side. Miura added type C with a triphalangeal component on the ulnar side. Wood also subdivided Wassel's triphalangism group (type VII) into four subtypes; A of 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, and D a central triphalangeal ray with non-triphalangeal hypoplastic duplications on each side (also known as triplication).

Buck-Gramcko and Behrens [4], and Blauth and Olason [5] created completely new classifications with additional subtypes for triphalangism and triplication, however neither have been used frequently. Several types of radial polydactyly cannot be classified using the abovementioned classification schemes. Especially complex forms of triplications with associated triphalangism cannot be classified in a detailed manner, demonstrating the shortcomings of the existing taxonomy and nomenclature of radial polydactyly. Several case reports have noted the same limitations [6-9].

Treatment of radial polydactyly combined with triphalangeal components require a more specific approach to joint structure and length of the thumb. Therefore we propose several modifications to Wassel's classification that result in a more complex taxonomy and more flexible nomenclature to facilitate communication, analysis and treatment of all known variations. The proposed classification is tested on our population of radial polydactyly and on the previously unclassifiable cases from literature.

MATERIAL AND METHODS

Material

One-hundred twelve patients with radial polydactyly visited our department between 1993 and 2006. Eight patients were excluded because they were operated elsewhere and therefore the original type of radial polydactyly could not be established. Seventeen of the remaining 104 patients had bilateral involvement of radial polydactyly, hereby obtaining a total of 121 affected hands. Eighteen hands (15%) could not be classified using the current classifications for radial polydactyly with triphalangeal components or triplication.

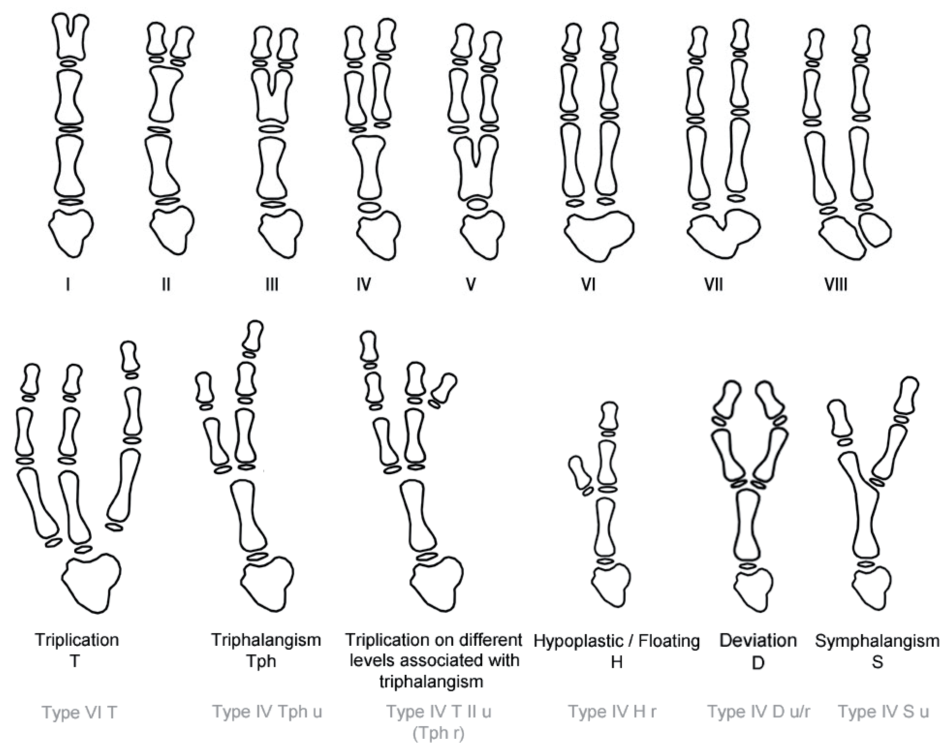
Method

A modified classification was developed in which the basis of Wassel's taxonomy and nomenclature was adopted, classifying the patient by level of duplication in the longitudinal direction, starting from the distal phalanx (I-VI). Wassel's type IV with duplication distal of the epiphyses of the proximal phalanx is in essence a type III and was therefore changed to a complete duplication of the proximal phalanx with duplicated epiphyses. To create a more logical and simple ranking based on level of duplication, Buck-Gramcko's carpal and intercarpal type were added [1, 4, 5]. A bifid duplication on level of the carpal bones was appointed type VII and a complete duplication type VIII (Figure 1). It should be noted that classification for duplication at the level of carpal bones is only possible after the ossification of the trapezium bone, starting at the fifth year [10].

Aberrant components were assigned using a specific abbreviation for each of these associated deformities, adapted from Upton's nomenclature for combined triphalangism and radial polydactyly [11] (see Figure 1). Triphalangism, a ray consisting of three phalanges, was assigned with "Tph". Thumb hypoplasia, a floating ray or rudimentary polydactyly with the suffix "H"; deviation from the longitudinal axis of the duplicates with the letter "D" and symphalangism, a condition with fusion of a joint, with "S". Designation of the associated deformity is followed by the position of the affected ray using "u" for the ulnar ray, "m" for the middle ray and "r" for the radial ray. For example, duplication at the level of the proximal phalanx with a triphalangeal component at the radial side, in Wood's modification a type IV B, would be classified as type IV Tph r.

Triplication was assigned with the abbreviation "T" (example given in figure 2). In cases with three thumbs not originating on the same part of the thumb, the most proximal level of duplication was indicated first using the Roman number for that specific type of duplication according to the Wassel classification, followed by the letter "T" (signifying thumb triplication). The second, more distal duplication was mentioned by the Roman numeral suitable for that specific level of duplication (example given in figure 3). Positions of the duplicated parts were designated by the same abbreviations used for the other associated deformities ("u" for ulnar, "m" middle, "r" radial).

In this study we classified all deformities for comparison according to the following three classifications: 1) the Wassel classification 2) Wood's and Miura's modification of Wassel's classification and 3) the modified newly proposed classification.



Level of Duplication		Abbreviations for associated deformities	
I	partial duplication distal phalanx	T	Triplication
II	complete duplication distal phalanx	Tph	Triphalangism
III	partial duplication proximal phalanx	H	Hypoplastic / Floating
IV	complete duplication proximal phalanx	D	Deviation
V	partial duplication metacarpal	S	Symphalangism
VI	complete duplication metacarpal		
VII	partial dupplication carpal bones	R	Radial
VIII	complete duplication carpal bones	U	Ulnar
		M	Middle

Figure 1. Suggested classification for radial polydactyly with overview of associated deformities and abbreviations. Level of duplication is assigned by Roman numbers starting with I at the distal phalanx and ending with VIII at the carpal bones. Partial duplication is assigned with uneven numbers and complete duplication with even. Abbreviations can be used for the different associated deformities: Tph: Triphalangeal, T: Triplication, S: symphalangism, D: Deviation and H: hypoplastic or floating. Triplication on different levels is classified by first assigning the most proximal duplication with the designated Roman number followed by the distal duplication.

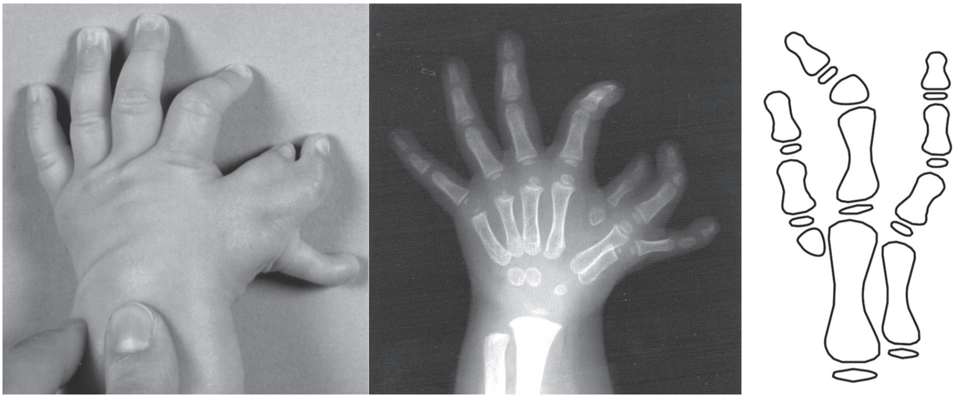


Figure 2. Typical example of the use of the modified classification: a triplicated left thumb. The thumb has triplication at the level of the first metacarpal (VI T), the ulnar ray is hypoplastic (H u) and the middle and radial ray are triphalangeal (Tph m/r). Therefore the deformity is classified as Type VI T (Tph m/r, H u).

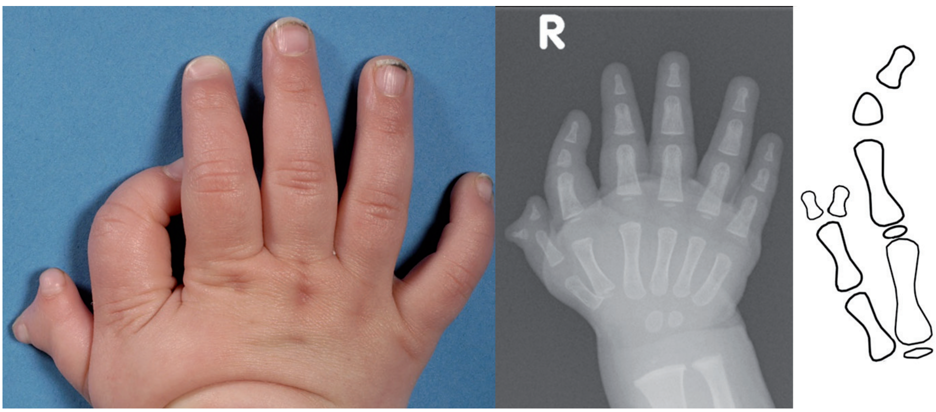


Figure 3. Example of triplication on different levels (T). The thumb has duplication at the level of the first metacarpal (VI); in addition duplication is present at the level of the distal phalanx of the radial ray (II). The ulnar ray has a triphalangeal component (Tph u). The thumb is classified as VI T II r (Tph u).

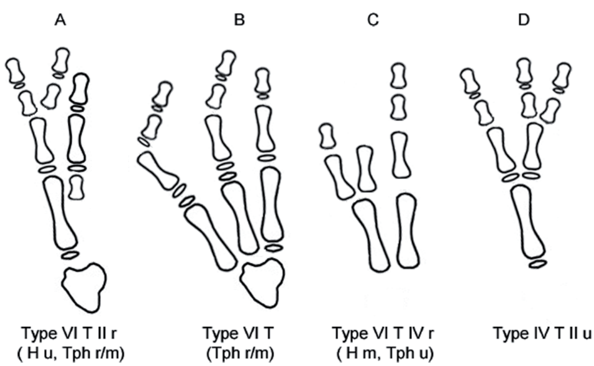


Figure 4. Schematic drawing of the case reports from literature classified using the newly proposed classification. A Atabay [7], B Yildirim [8], C Al-Qattan [9] and D Mennen [6]

RESULTS

Thirty-nine of the 121 hands had radial polydactyly associated with triphalangism (32%). Using the Wassel classification, all types of radial polydactyly could be classified. Triphalangism (type VII) was the most common type of radial polydactyly in current series (32%; Table 1). These 36 subjects had a variety of deformities: triplications, hypoplastic components and triphalangism. In Wood's modification of the Wassel classification, complete duplication at the level of the proximal phalanx was the most common type, subdividing the triphalangism group into subgroups with or without triphalangeal components (Type IV 30%). With this classification, eighteen of the 121 hands (15%) could not be classified.

The most common type using the newly proposed classification was type IV (29%) (Table 2). Nine patients had triplicated thumbs (suffix T). Two patients had partial duplication of the carpal bones (type VII) who were classified as type IV and VI using the existing classifications. In our series, no complete duplication of the carpal bones (type VIII) was present as described earlier by Blauth [5]. All eighteen subtypes could be classified into the proposed subgroups: fourteen into subgroups of type VI and four into type IV.

The presented cases in literature regarding triplication had duplications on different levels [6, 7, 9] or on the same level with different positions of the triphalangeal components compared to the Wood type 7D [8]. The specific anomalies could not be classified using Wood's adjusted Wassel classification. Using the proposed classification, all anomalies could be classified (Figure 4).

Table 1. Classification of patients with radial polydactyly according to the Wassel classification [1] and Woods and Miura's additions to the Wassel classification [2, 3]

Total N 104 Hands 121		
	Wassel	**Wood / Miura**
Type	N	N
I partial duplication distal phalanx	5 (4%)	5 (4%)
II complete duplication distal phalanx	20 (16%)	20 (16%)
III partial duplication proximal phalanx	7 (6%)	7 (6%)
IV complete duplication proximal phalanx	36 (30%)	36 (30%)
A triphalangeal on radial side	-	2 (2%)
B triphalangeal on both sides	-	2 (2%)
C triphalangeal on ulnar side	-	6 (5%)
V partial duplication of metacarpal	8 (7%)	8 (7%)
VI complete duplication of metacarpal	6 (5%)	6 (5%)
VII triphalangism	39 (32%)	-
duplication at level of the metacarpal and		
A triphalangeal on ulnar side	-	1 (1%)
B triphalangeal on both sides	-	5 (4%)
C triphalangeal on radial side	-	5 (4%)
D triplication; triphalangeal in centre, hypoplastic metacarpal on both sides	-	0
No classification possible	0	18 (15%)

Table 2. Classification of patients with radial polydactyly according to our modified classification

Type	N
I partial duplication distal phalanx	5 (4%)
II complete duplication distal phalanx	20 (16%)
III partial duplication proximal phalanx	7 (6%)
IV complete duplication proximal phalanx	35 (29%)
Tph triphalangeal components	13 (10%)
T triplication	1 (1%)*
V partial duplication of metacarpal	8 (7%)
VI complete duplication of metacarpal	5 (4%)
Tph triphalangeal components	17 (14%)
T triplication	8 (7%)*
VII partial duplication of carpal bones	2 (2%)
VIII complete duplication of carpal bones	0
No classification possible	0

* all triplications were associated with triphalangism

DISCUSSION

In our population, 32% of all subjects were classified as a type VII (triphalangism) using the Wassel classification, without any discrimination of the large variety of deformities amongst these subjects. Eighteen of 121 hands with radial polydactyly could not be classified using the current classifications for associated radial polydactyly and triphalangism. In our series, triphalangeal and triplicated thumbs are frequently present, probably because in the southwest region of the Netherlands a genetic isolate, a deformity located at chromosome 7q36, has been identified in patients with radial polydactyly and triphalangeal thumbs [12]. Radial polydactyly combined with triphalangeal components requires detailed operative procedures for reconstruction of joint surface, reduction of length and positioning of the thumb. Besides removal of the extra phalanx, distal or proximal interphalangeal joint removal with arthrodesis can reduce length. Regarding the positioning of the thumb, a rotation and reduction osteotomy of the first metacarpal can be performed. Therefore, triphalangeal components are an important factor in operative planning. A modified nomenclature for radial polydactyly was suggested to be able to discriminate between different types of radial polydactyly. In the modified classification, we adapted the basics of the Wassel classification in which level of duplication was assigned with Roman numbers, by adding abbreviations to further designate the specific deformities.

Using existing schemes, patients with radial polydactyly with complex types of triplication or triphalangeal components cannot be classified. Considering the relatively large number of subjects with radial polydactyly combined with a triphalangeal component (Wassel 27% [1]), Wood 11% [13], Buck-Gramcko 14% [4], and in our population 32%, Table 1), a more precise classification would be valuable to assist comparison [6-9].

In this study, a proposal for a modified classification and nomenclature was presented based on Wassel's [1] and Buck-Gramcko's [4] classification system and Upton's nomenclature for radial polydactyly associated with triphalangeal components [11]. Wassel's original type VII was substituted because it grouped deformities based on different taxonomic characteristics than the previous six types (i.e. associated triphalangeal components versus level of bifurcation. Designating type VII (and VIII) to duplication on a more proximal level, i.e. duplicated carpal bones, is more logical compared to the six other types. Triphalangeal components, triplication, hypoplasia or floating components [14],[15], deviation [15-18], and symphalangism [19] are reported associated deformities and should be categorized on a lower taxonomic level. Horii et al. [20] created a classification to include symphalangism and fibrous connections. The various sub-types that have been added to Wassel's groups by others to accommodate common patterns of associated deformities have been labeled by ways of a single letter [2, 3, 16, 20]. Correct identification of these subgroups is difficult, since the labels do not directly relate to the anatomy of the designated deformity. Upton proposed a more descriptive notation [11], designed to indicate triphalangeal components. In the present study, a similar yet more extended nomenclature system is proposed to label the various characteristics of radial polydactyly. With the proposed nomenclature, it is possible to classify complex deformities with radial polydactyly, but no new type or subgroup is implied every time a new anomaly is labeled. If such was the case, it would lead to a multitude of groups comprised of a single deformity. All patients of the published case reports could be classified using the proposed classification [6-9, 19].

Although thumb triplication (a condition where three partially or fully developed thumbs are present) is considered to be a complex degree of polydactyly, it was not assigned to a separate type. From a surgical perspective, thumb triplication can be considered a deformity consisting of two longitudinal duplications on the same level or on different levels of the first ray. Only careful evaluation of the surgical implications of the described anatomy might determine whether a new type is warranted or that a deformity should be grouped under a previously established type.

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SIX

Triplicated thumbs; a rarity?

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SUMMARY

Background

Triplication of the thumb is supposed to be a rare condition and a complex form of radial polydactyly. We encountered an unusual high number of triplicated thumbs at our unit. Is triplication of the thumb indeed a rare condition?

Methods

In our series from 1933 till 2005 121 patients with radial polydactyly were recorded. In nine subjects of this group triplicated thumbs either unilateral or bilateral were identified. This complex type of radial polydactyly occurs in various forms.

Results

A total of eleven triplicated thumbs are demonstrated, all cases were combined with triphalangeal components. Only one of these in total eleven triplicated thumbs could be classified according to the currently most used classifications. In all cases aberrant rays were excised, thumb length and alignment restored by osteotomies, joints were stabilised, tendons reinserted and nails and nail walls corrected if necessary. Also in all cases a correction of triphalangeal components was performed.

Conclusions

In literature triplications are a rarity presented by single case reports. In our series triplicated thumbs are not so rare probably because in the southwest region of the Netherlands a genetic isolate, a deformity located at chromosome 7q36, has been identified with radial polydactyly and triphalangeal thumbs Treatment for the presented triplicated thumbs was based on the same general principles as for less complex forms of radial polydactyly. In essence to assemble useful elements of the separate (partial) thumbs to reconstruct one functioning, stable thumb.

INTRODUCTION

Radial polydactyly, a congenital condition of the hand with a supernumerary thumb, is a common form of polydactyly; however more complex forms like triplication of the thumb are rare. Radial polydactyly, including triphalangeal thumb and triphalangeal thumb-poly-syndactyly syndrome are mapped to chromosome 7q36 [1-4]. In mouse models up regulation of the signaling molecule Sonic Hedgehog (SHH) leading to ectopic expression at the anterior margin of the limb bud, is appointed as the molecular basis of radial polydactyly [5]. Radial polydactyly has a wide range of presentation and complexity ranging from a bifid distal phalanx, to a complete duplication of the first ray with involvement of the carpal bones. The developmental status of the supernumerary radial digit can vary from a rudimentary finger, consisting solely of soft tissue, to a full finger containing all phalanges. Wassel categorized all thumb polydactylies in seven different groups [6]. This classification was later expanded with additional subtypes to be able to register radial polydactyly with triphalangeal components [7]. Additionally, however less common features like triphalangism, symphalangism, and deviation add to the complexity. Triplication, a condition where the first ray consists of three thumbs, is very rare, case reports reported in literature are sparse. All reported subjects however are in combination with triphalangism [8-10], and two in combination with radial club hand [11, 12]. Nine patients with eleven triplications of the thumb are presented to demonstrate the various forms of presentation of this rare condition of radial polydactyly. The aim of this paper is to discuss the various encountered forms of complex radial polydactyly and their basic surgical management.

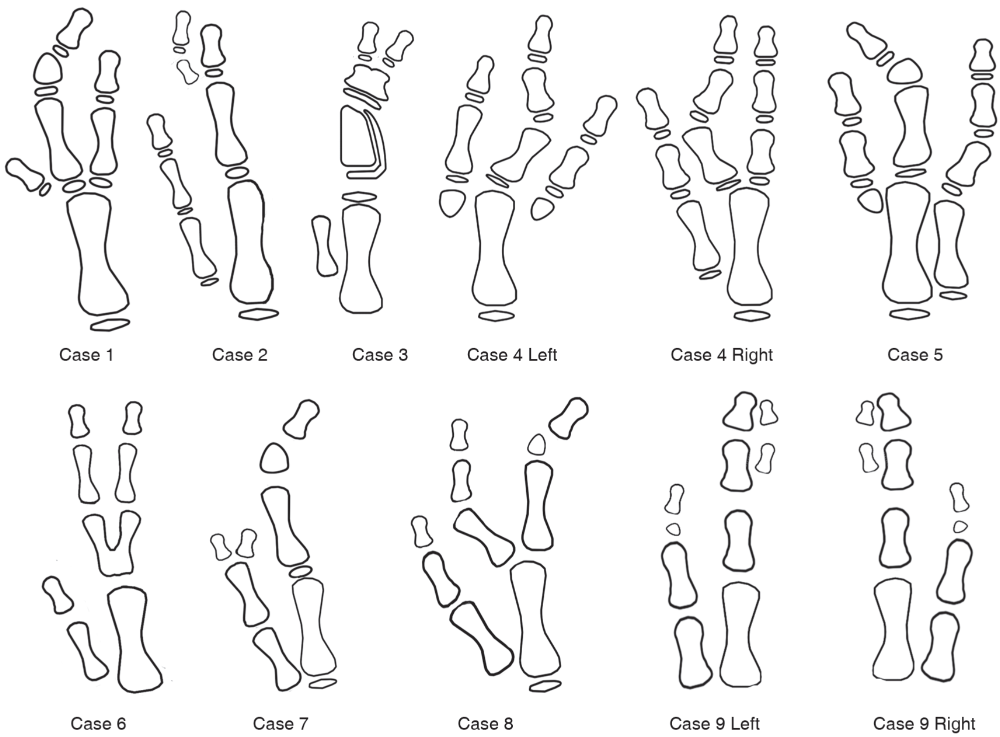

Figure 1. Illustrations of the 11 triplicated thumbs.

CASE REPORTS

Case 1

A three months old boy visited our multidisciplinary outpatient clinic for congenital differences of the upper extremities for assessment of polysyndactyly of both hands. Except for the limb deformities no other congenital deformities were present. Family history was positive for polydactyly. The left hand showed a triplication of the thumb, consisting of a hypoplastic radial thumb, a triphalangeal intermediate thumb and an ulnar biphalangeal thumb (Figure 1). All three rays shared one common metacarpal. The two ulnar thumbs featured a simple complete syndactyly. Examination of the rest of the hand revealed a rudimentary ulnar polydactyly; Type-B [13] and a complete syndactyly of the fourth and fifth ray. The triplication could not be classified by one of the classifications. The deformities on the right hand demonstrated besides an ulnar polydactyly, a Wood Type-VIIA radial polydactylOperative procedures: At the age of six months he was operated on his left hand; the rudimentary radial and biphalangeal ulnar ray were removed. The delta shaped extra phalanx of the middle ray was excised and the collateral ligaments of the interphalangeal (IP) joint were repaired. In the same session the rudimentary ulnar polydactyly was excised and a fourth web syndactyly was dissolved. A second procedure was performed at the age of four year, where a reduction osteotomy to reduce the length of the first metacarpal with reinsertion of the intrinsic musculature was performed. The radial collateral ligament was tightened to adjust the metacarpophalangeal (MCP) instability. A Huber opponensplasty was performed to enhance opposition at the age of seven year.

Case 2

At the outpatient clinic a boy at the age of six months was assessed with a radial polysyndactyly of the right hand. He had no other congenital deformities and no family history of polydactyly was present. Radiological examination revealed a triplicated thumb, consisting of a radial hypoplastic biphalangeal thumb and an ulnar thumb which was duplicated in the interphalangeal joint (Figure 1). The radial side of the ulnar thumb had two distal phalanges and the ulnar side had only one. These two sides were connected by a complete simple syndactyly. Two first metacarpals were present. The deformity could not be matched to any subtype of a type seven polydactyly [7].

Operative procedure: At the age of three years the radial component of distal phalanx of the ulnar ray was removed and stabilisation of IP joint with the accessory extensor tendon of the radial component was performed. The total rudimentary radial ray was excised; the accessory extensor pollicis longus of the rudimentary radial ray was used to restore stabilisation of the metacarpophalangeal (MP) joint.

Case 3

A three year old girl was assessed with a radial polydactyly of both hands. There were no other congenital anomalies and the family history was positive for triphalangeal thumb. At physical examination three thumbs were observed on the right hand and two thumbs on the left. The right hand consisted of a radial hypoplastic thumb and a partially duplicated ulnar thumb. The left hand showed a radial hypoplastic thumb and a single ulnar thumb. X-ray examination of the right hand revealed a hypoplastic radial digit, an ulnar thumb with a complete duplication of the distal phalanx, a bifurcated middle phalanx, a delta proximal phalanx, and two first metacarpals (Figure 1). The right thumb deformity as a whole could not be classified according to existing classification systems. The left hand showed a duplicated middle phalanx (i.e. bracketed epiphyses/kissing delta phalanges), with both duplicates articulating with a single proximal and a single distal phalanx; Wood Type VII-A deformity.

Operative procedures: at the age of three and a half years a surgical correction of the right thumb was planned. The ulnar distal phalanx was excised; an osteotomy of the mid phalanx was performed to reduce the broadness, the collateral ligaments of the IP joint were repaired. A corrective osteotomy of the proximal phalanx was performed with removal of the ulnar placed growth plate and reinsertion of the intrinsic musculature. A correction osteotomy at the proximal interphalangeal joint, with arthrodesis was realized at later age (nine years).

Case 4

A three months old boy was presented with a radial polysyndactyly and ulnar polydactyly on both hands. He had no other congenital deformities. There was a family history of polydactyly. He had thumb triplication on the right hand, consisting of two ulnar triphalangeal thumbs and a radial biphalangeal thumb. There were two metacarpals, the ulnar one was fully developed and the radial one was rudimentary. There was a complete simple syndactyly of all three thumbs. The patient had a rudimentary ulnar polydactyly and a complex syndactyly with conjoined distal phalanges of the fourth and fifth digit. The thumb deformity of the right hand could not be classified according to existing classification systems. (Figure 1 and 2A)

The left hand also featured a triplicated thumb. X-ray examination showed a triphalangeal thumb accompanied on either side by biphalangeal duplicates. The ulnar thumbs had a partial simple syndactyly and the radial biphalangeal thumb was independent. One fully developed metacarpal was present with two very small bony remnants of a metacarpal on either side. The left triplicated thumb could be classified as Wood type VII-D. (Figure 1 and 3A)

Operative Procedures: the right hand was operated at the age of one year. The radial and ulnar rays were excised and the MP joint stability was restored using the accessory tendons of the excised rays. The middle ray was reduced to a biphalangeal thumb through a distal interphalangeal (DIP) joint reduction osteotomy and arthrodesis (DIPRAD) [14]. In the same session a fourth web was created (Figure 2B).

The left hand was treated at the age of two years. A stable biphalangeal thumb was created by excision of the radial and ulnar ray, stabilisation of MP joint with accessory tendons, and reduction of the triphalangeal component by a DIPRAD procedure. The syndactyly was resolved by creating a fourth web (Figure 3B).

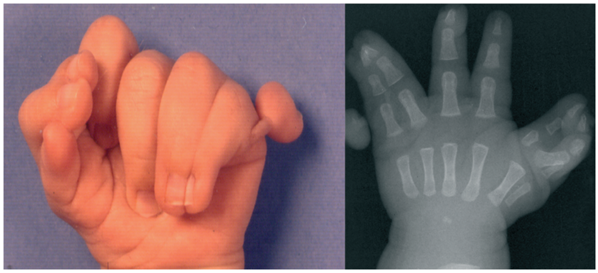

Figure 2A. Preoperative picture and x-ray of right hand of case 4

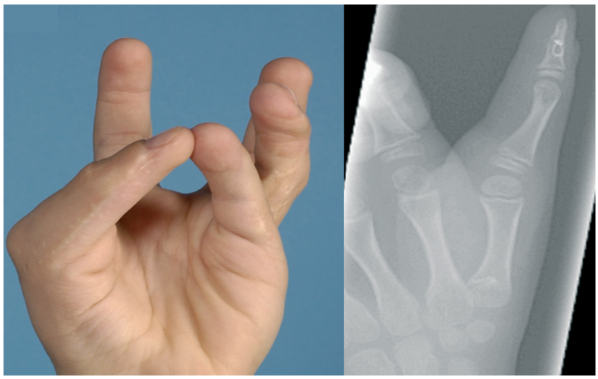

Figure 2B. Postoperative results of right hand.

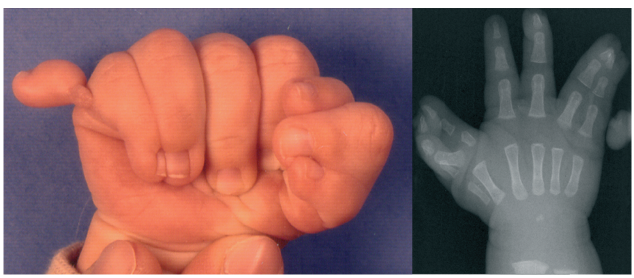

Figure 3A. Preoperative picture and x-ray of left hand of case 4

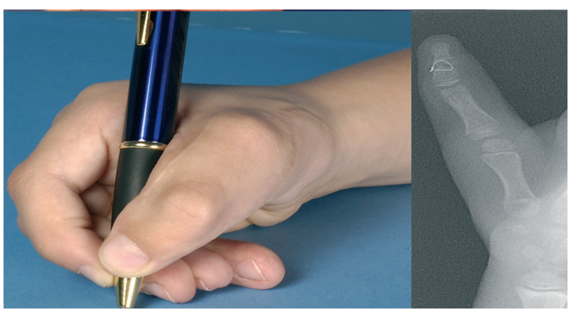

Figure 3B. Postoperative results of left hand.

Case 5

A four months old girl was referred with a triplication of the left thumb. Her left elbow demonstrated hypoplasia of the radial head and ulna; there was no family history of thumb deformities. The left hand showed a triplication of the first ray at the metacarpal level of which one was very rudimentary. The two radial thumbs were triphalangeal, and the ulnar thumb was biphalangeal (Figure 1). The two ulnar thumbs had a simple and partial syndactyly. The thumb deformity as a whole could not be classified according to existing classification systems.

Operative procedures; at one year of age the ulnar ray and hypoplastic radial ray of the left thumb were removed, and the MP joint was stabilized using the intrinsic musculature. The additional phalanx of the remaining middle ray was removed and the IP joint stabilized. A Huber opponens plasty with revision of the radial collateral ligament was performed at the age of five.

Case 6

A four months old girl was recently presented at our outpatient clinic with bilateral radial polydactyly. She had a congenital club foot on the right and gastro-esophageal reflux; there was no family history of hand deformities. The right hand had a bifurcated first metacarpal (type V); the left hand demonstrated a triplication with duplication at metacarpal level and a bifurcated ulnar proximal phalanx. Both ulnar thumbs were triphalangeal with incomplete syndactyly (Figure 1). The radial thumb only showed a proximal remnant on the x-ray. Existing classifications were not suitable for the deformity. She is on the waiting list for operation.

Case 7

A one year old girl was referred with bilateral radial polydactyly with triphalangeal components. No other congenital deformities were present; there was a positive family history of triphalangeal thumb. The left hand had duplication at the level of the distal phalanx (type 2), the right hand had a triplicated thumb with a duplicated first metacarpal, the ulnar ray of the thumb was triphalangeal and the radial ray had a duplicated distal phalanx (Figure 1).

Operative procedure: At the age of two, an operative correction was performed. The radial ray was excised including the duplicated distal phalanges. A pollicisation of the ulnar ray with stabilisation using the intrinsic musculature was performed to create an adequate thumb.

Case 8

A seven months old girl visited the outpatient's clinic with radial polydactyly of both hands. The right hand had a triplication of the thumb; a floating hypoplastic biphalangeal radial ray and a Wassel type IV duplication of the radial ray of which both were triphalangeal (Figure 1 and 4A). On the left hand a hypoplastic radial ray was present, the proximal phalanx demonstrated a 'kissing delta phalanx' [15], with the distal phalanges co-joined.

Operative procedure: She was operated at the age of two and a half year. The delta phalanx of the

Figure 4A. Preoperative picture and x-ray of right hand of case 8.

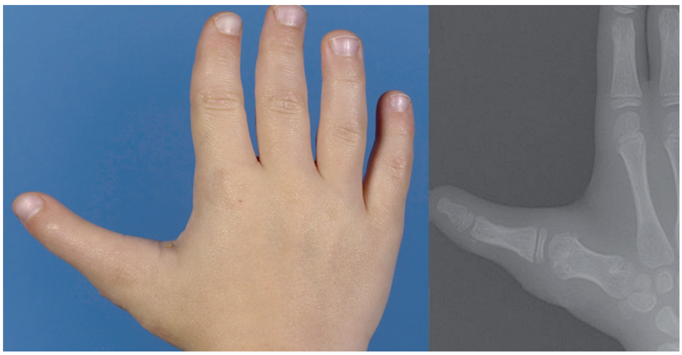

Figure 4B. Postoperative results.

ulnar thumb was removed and a correction of the joint surface of the ulnar proximal phalanx with restoration of IP joint stability was performed. The floating hypoplastic radial ray and radial ray at the level of the MP joint were both excised; the joint surface of the ulnar first metacarpal was corrected. Using the accessory tendons of the excised thumbs stability of the MP joint was created (Figure 4B).

Case 9

A one month old boy with bilateral radial polydactyly was referred to the outpatient clinic. The left side had a triplication of the thumb with triphalangeal components; a hypoplastic triphalangeal radial ray and an ulnar ray with a type II polydactyly. The right thumb had the same type of triplication compared to the left side (Figure 1).

Operative procedure: Operative corrections on the left thumb were performed at the age of three months. The complete hypoplastic radial ray and the two distal phalanges of the ulnar ray were removed; the middle ray was reduced to a biphalangeal thumb through a distal interphalangeal (DIP) joint reduction osteotomy and arthrodesis (DIPRAD), and positioned as a thumb using a rotation-abduction osteotomy of the first metacarpal. In the same session a similar procedure was performed for the right side.

DISCUSSION

Triplication of the thumb is supposed to be rare with only a few cases reported in literature [8-11]. Other authors found only one case of triplication of the thumb in large polydactyly series, [7, 16]. In our series over the last 12 years (1993 till 2005) out of 121 radial polydactyly hands, eleven triplicated thumbs in nine patients were registered (9%). As in previous literature all eleven thumbs had triphalangeal components, indicating the relation between triphalangism and triplication. The high amount of patients with complex radial polydactyly at this department must be due to the high quantity of triphalangeal thumbs in the southwest region of the Netherlands. Research on this genetic isolate has revealed the genetic locus to be on chromosome 7q36 [1].

Classification of triplicated thumbs is only possible for triplication at metacarpal level by Wood's subdivision of Wassel type VII. Wood subdivided type seven by four different categories with triphalangeal components; type VII D is a central triphalangeal thumb with on both sides a duplication of the metacarpal, no further information on the different joints is provided. Buck-Gramcko [16] subdivided triplication of the metacarpals by the different positions of triphalangeal components. Blauth and Olason [17] created a classification were the degree of duplication could be assigned, no additional information on variables; e.g. level of duplication is given in this classification. Upton's descriptive classification [18] of radial polydactyly combined with triphalangeal components does not allow classification of triplicated thumbs. Ten of the eleven presented thumbs could not be classified according to current classifications, because the duplications are on different levels. In literature only Yildirim's case report [11] can be classified according to the VII D type radial polydactyly.

Basic principles of the treatment of above mentioned triplicated thumbs are the same as for less complex radial polydactyly types; proper alignment of the thumb along the longitudinal axis, stabilisation of joints, tendon balancing, nail and nail wall correction and creating sufficient thumb size [19]. Furthermore the always present triphalangeal components create additional procedures regarding alignment and removal of the extra phalanx. Also growth and position of the epiphyseal growth plates of the first metacarpal have to be observed as the first metacarpal is known to add additional length to the thumb in Triphalangeal Thumb (TPT), therefore a reduction osteotomy of the first metacarpal has to be considered [20]. Proper alignment of the thumb in TPT frequently requires a considerable amount of rotation and abduction osteotomy of the first metacarpal with reinsertion of the intrinsic musculature; in the less complex cases of radial polydactyly these considerations are not necessary. Removal of the extra phalanx in combination with restoration of the joint stability is necessary in cases with delta or small trapezoid phalanx for instance through a reduction arthrodesis at the distal interphalangeal joint [21]. In cases with a larger trapezoid or full type extra phalanx with no opposition an other treatment is required: a reduction-rotation osteotomy of the first metacarpal or a pollicisation can be performed [14, 18].

In summary triplication of the thumb occurs in a higher frequency if the studied population contains many triphalangeal thumbs. In our series this was up to 9% of all hands with radial polydactyly. It is an entity with various expressions in regard to level of duplication, joint stability, hypoplasia, and triphalangism as presented in the nine patient reports. Except one all the other ten triplicated thumbs could not be classified according to existing classifications. Treatment of the triplicated thumb is based on the same principles as other radial polydactylies. However the often coexisting triphalangeal components require adequate correction. For instance for the additional length a reduction osteotomy of the first metacarpal, a rotation abduction osteotomy for proper alignment, and removal of the extra phalanx or a proper pollicisation procedure of the best triphalangeal thumb can be performed.

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Treatment and long term follow up

SEVEN

Outcome of two types of surgical correction of the extra phalanx in triphalangeal thumb: is there a difference?

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SUMMARY

The surgical strategy of treatment of the opposable triphalangeal thumb is correction of the radio-ulnar deviation, reduction of the additional length and joint stabilization. The commonest procedures are: 1) removal of the extra phalanx and stabilization of the remaining joint; and 2) a combined reduction osteotomy with resection of the distal joint followed by arthrodesis. We treated 20 patients (33hands). In 17 hands the extra phalanx was removed and in 16 hands we used the combined osteotomy procedure of distal joint removal and arthrodesis. None of the patients in either group had an unstable interphalangeal joint. The mean radial or ulnar deviation in the interphalangeal joint was respectively 5^0 degrees and 9^0. Mean active flexion in the interphalangeal joint was respectively 35^0 and 46^0 in the two groups. Results for both procedures are similar for both objective measures and self-rated function and activities of daily living. Either surgical approach seems reliable.

INTRODUCTION

Triphalangeal thumb is a congenital anomaly of the thumb named after the extra phalanx in the thumb. The shape of the extra phalanx can present in various forms, ranging from a small rudimentary bone to a completely developed phalanx. Wood (1976) classified the triphalangeal thumb according to the shape of the extra phalanx, distinguishing the delta type (a triangular shape), the trapezoid type (a rectangular shape) and the full type (a fully developed extra phalanx). Buck-Gramcko (1987) suggested a broader classification into to six types, based on joint mobility, web space and intrinsic and extrinsic muscle abnormalities.

The most common type is the opposable triphalangeal thumb with a small extra phalanx varying in shape. This thumb has normal carpo- (CMC) and metacarpophalangeal (MP) joints, with normal or slightly hypoplastic musculature and with a normal first web space. Sometimes the MP joint can be hyper mobile or unstable. The surgical goals are to correct the extra length, to restore the malalignment while maintaining growth potential, to stabilize the joint and to maintain as much range of motion as possible. A procedure where the extra phalanx is removed provides these goals. In the same procedure the collateral ligaments are shortened or lengthened on both the radial and ulnar sides as necessary (Flatt, 1977, Hovius et al., 2004, Wood, 1976). The main disadvantages of this procedure are the possible damage to the joint surfaces, resulting in loss of motion or stiffness of the interphalangeal joint as well as possible joint instability as a result of inadequate correction or reconstruction of the collateral ligaments.

An alternative procedure to remove the extra phalanx is a combined reduction osteotomy (Peimer, 1985). For this procedure a larger extra phalanx has to be present combined with at least one adequately functioning joint. In this procedure a transverse osteotomy is performed at the base of the distal phalanx is followed by an osteotomy at the base of the extra phalanx parallel to the proximal joint. After adequate alignment a K-wire is placed across the osteotomy site. The combined osteotomy preserves the proximal joint and reduces the deviation, without interfering with the 'new' interphalangeal joint. The main disadvantage is possible malalignment as a result of inadequate osteotomy planning or poor execution.

Upton states that the results of the removal of the extra phalanx are less predictable if the children are older than four years and therefore a reduction osteotomy is advised (Upton and Shoen, 2000). Many complications have been described following excision of the extra phalanx, such as incomplete correction, joint deviation, joint instability, and joint stiffness (Flatt, 1977, Wood, 1976). For the combined reduction osteotomy, far fewer complications are reported and joint instability or incomplete resection, have not been reported. A few cases with insufficient reduction require an additional metacarpal reduction osteotomy and one non-union has been reported (Jennings et al., 1992). Based on this, one could assume that the combined reduction osteotomy is preferred over the simple excision, thereby deferring the surgical procedures to a later age when the extra phalanx is large enough to perform the osteotomy.

The aim of this study was to evaluate retrospectively the medium-term outcomes of both procedures.

PATIENTS AND METHODS

Patients

From 1982 to 2003, 51 patients with a triphalangeal thumb attended our centre. Thirty-one patients were excluded as they had long rectangular or full extra phalanges and therefore regarded as more severe triphalangeal thumbs which require different procedures. The remaining 20 patients with 33 affected hands were included in this study: 18 had a delta-shaped extra phalanx; and 15 had a trapezoid extra phalanx. All patients were tested for flexion in the existing interphalangeal joints which was present. As a result of the young age of the patient no exact measurements were performed. In 17 children the extra phalanx was removed; in 16 a combined reduction osteotomy was performed. The senior author assigned the thumb to a specific procedure. No clear indications were established to assign a patient to one particular procedure. However the feasibility of an osteotomy and therefore the size of the extra phalanx was an indicator. With the retrospective nature of the study and the non-scaled radiographs the exact size of the extra phalanx of each patient in the preoperative setting was impossible to determine accurately. The minimal age at follow up was chosen as 6 years, so that all children would be able to perform and understand the examination and tasks completely. The mean postoperative follow-up was 7 (range 3 to 17) years. The mean age of removal of the extra phalanx was 3 (range 1 to 4) years and for the combined reduction osteotomy 3 (range 1 to 8) years.

Surgical procedures

The procedure consisted of removal of the extra phalanx and adjustment of the collateral ligaments on both the radial and ulnar sides. If necessary a Z-plasty lengthened the skin and additional skin was excised on the expanded contra lateral side when apparent deviation existed (Figure 1). A K wire (various sizes were used) stabilized the interphalangeal joint for four to six weeks.

In the combined reduction osteotomy a transverse osteotomy was performed at the base of the distal phalanx, followed by an osteotomy at the base of the extra phalanx parallel to the proximal joint via a dorsal Y-incision. Following alignment, the skin was inserted in a V-shaped fashion. Surplus skin and soft tissue were excised. The bone ends were aligned and fixed with a K wire through the interphalangeal joint for 4-6 weeks (Figure 2); in eight cases the proximal part of the extra phalanx was long and solid enough the K wire was not passed through the residual interphalangeal joint. The osteotomies were by choice performed with a scalpel; if the bone was too hard a hand-held osteotome was used. In both procedures, the extensor tendons were shortened. Additional correctional procedures including web space deepening or thumb metacarpal length shortening were performed in the same session if necessary. The thumbs were immobilized in plaster until K wire removal. Guided active and passive motion started after immobilization in both procedures.

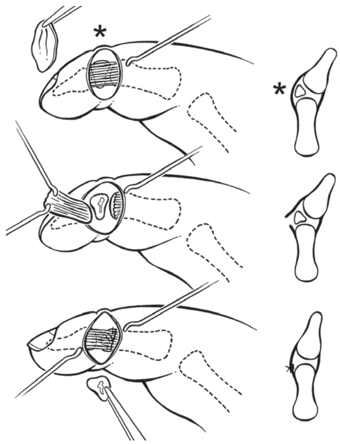

Figure 1. The extra phalanx is removed and the remaining joint is stabilized. (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

Figure 2. A combined reduction osteotomy with resection of the distal joint followed by desis ,after proper aligment, is performed. Hereby preserving the proximal interphalangeal joint. (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

Methods

To assess thumb function particularly of the interphalangeal (IP) joint, the first author examined all hands in a standardized manner. Stability of the IP joint was tested manually in the radio-ulnar plane. Lateral movement of the joints was measured with a hand held goniometer, recorded in degrees and marked as unstable if more than 5^0 of deviation to stress was possible (Ogino et al., 1996). Patients were asked if pain was present at rest or during motion or when pressure was applied. All thumbs were assessed on present nail abnormalities. All patients were asked to score function and appearance using a visual analogue scale, ranging from 0 (very poor/ugly) to 10 (very good/beautiful). To assess function during activities of daily life the ABILHAND-kids questionnaire was used. The ABILHAND-kids is a questionnaire created for children with cerebral palsy and has been validated to measure manual ability in children (Arnould et al., 2004). The questionnaire consists of 21 questions concerning mostly bimanual tasks. Children are asked to score if a task is easy, difficult or impossible to perform. Total scores range from 0 to 42, with 42 being the best possible score.

Statistical Analysis

Data were analysed as frequencies and percentages, or means and ranges.
Institutional review board approval was acquired for this study.

RESULTS

In total 20 patients were operated, in 17 thumbs the extra phalanx was removed; in 16 thumbs a combined reduction osteotomy was performed. There was no interphalangeal joint instability in either group. After a combined reduction osteotomy, five of 16 thumbs had ulnar or radial deviation in the interphalangeal joint and three of 17 after removal of the extra phalanx. No active or passive flexion or extension in the interphalangeal joint was present in two of the 17 hands after removal of the extra phalanx and one of 16 in the osteotomy group (Table 1).

Table 1. Function measurements

	Removal of extra phalanx N = 17	**Combined reduction osteotomy N=16**
Pain	0	2
Nail deformities	2	0
Joint instability	0	0
No deviation	14/ 17	11 / 16
Deviation if present in degrees *mean (range)*	21 (15-25)	18 (5-25)
Interphalangeal joint flexion *mean (range)*	35 (0-85)	46 (0-90)

Table 2. Appearance and function in daily life

	Removal of extra phalanx N = 17	**Combined reduction osteotomy N =16**
	mean (range)	
Appearance (VAS)	5.8 (1.5 – 9.7)	4.8 (1.2 – 9.7)
Function (VAS)	8.1 (5.1 – 10)	7.4 (0.2 – 10)
Daily life questionnaire		
ABILHAND-kids (0-42)	38 (34- 42)	39 (36-42)

The stiffness in the interphalangeal joint in one patient was due to a recognized technical failure during operation. In this specific case no release of the short and tight radial collateral ligament was performed. On clinical assessment the thumb was too long postoperatively in three patients and an additional reduction osteotomy of the first metacarpal was performed as a secondary procedure. When pressure was applied to the interphangeal joint, two children with a combined reduction osteotomy complained of pain. On inspection, two children showed a deformity of the nail, both after removal of the extra phalanx.

The visual analogue scale (VAS) scores for function and cosmesis were similar in both groups (Table 2). On the ABILHAND-kids questionnaire on activities of daily living the patients who had removal of the extra phalanx scored a mean of 38 (range 34 to 42) out of maximal 42. The patients who had an osteotomy had a mean score of 39 (range 36 to 42).

DISCUSSION

Several surgical procedures for correction of the additional length and deviation with maintaining adequate joint movement have been reported earlier. 'Simple' removal of the extra phalanx with tightening or reconstruction of the collateral ligaments has been advised for the younger children up to one year Flatt (1977). Several complications have been reported for this intra-articular procedure, such as joint instability, joint stiffness, insufficient range of motion, incomplete resection of the extra phalanx, and secondary deviation (Kozin, 2005, Peimer, 1985). Wood reported 10 of 18 hands had deviation present postoperative, probably as a result of ligament instability (Wood, 1976). In our series, fewer complications occurred; we found no joint instability, minor deviation (up to 25 degrees) in only three of the 17 patients, and inadequate interphalangeal joint movement (less than 25 degrees flexion) in five of 17 children.

In patients with a larger extra phalanx, a correction osteotomy can be performed; Flatt described a 'peg' shaped osteotomy for correction of length and deviation in such cases (Flatt, 1977). Peimer adapted several techniques for his combined reduction osteotomy in which he corrected the malalignment and additional length by removing the distal interphalangeal joint, at the same time leaving the proximal interphalangeal joint untouched.. With a mean of 11-year follow up, Jennings reported very good results for interphalangeal joint flexion (mean 63 degrees, range 35 to 100) (Jennings et al., 1992). They reported only a few minor complications occurred: one split nail and one case with 10 degrees ulnar deviation in a group of 13 patients with 15 involved hands In our series, the range of motion was less good than the results of Jennings et al.: four children of 16 had inadequate (<25 degrees) or no interphalangeal joint flexion, although the mean flexion for the complete group (46 degrees) was comparable. In our correction osteotomy group, more children had postoperative deviation (5 0f 16), although clinically it was not significant (mean for total group 9 degrees).

Although the triphalangeal thumb is named after the extra phalanx, the skeletal anomaly can be more extensive, even for the cases with a small extra phalanx. Earlier we described the influence of the growth plates of the metacarpal on the length of the thumb (Zuidam et al., 2006). In Jennings series two patients out of 15 required an additional metacarpal reduction osteotomy because the combined reduction osteotomy did not adequately reduce the additional length of the thumb. In our series, in three an additional reduction osteotomy of the first metacarpal was necessary.

In the earlier procedures performed by the senior author (SH), procedure selection was based on age and the type of the extra phalanx: younger patients with a smaller extra phalange underwent a phalanx removal while older patients and patients with a more trapezoidal phalanx sustained a reductive osteotomy. Over the years, in younger patients, even if they had a trapezoidal phalanx, the phalanx was still resected. In addition to selecting patients at a younger age, the technique following removal of the extra phalanx changed over the years by performing more correction on the radial and ulnar side. On the tight side, which was mostly radial, ligament and skin were released. On the surplus side, which was mostly ulnar, ligament and skin were tightened. This study is therefore not a comparison amongst two groups, rather a description of outcomes of the techniques that can be used in the same patient population.

Following resection or corrective osteotomy, the thumb can still be too large, i.e. the tip of the thumb is at or distal to the PIP joint of the index finger side. In those patients, the first metacarpal was shortened and corrected with intrinsic tightening in the same operation (Zuidam et al 2006).

Function is difficult to measure in young children with congenital hand malformations, which is a general problem in studies on surgery in congenital hand malformations. As a result, deficits in preoperative active flexion of the interphalangeal joints are almost impossible to measure since children are typically operated at a very young age. In our retrospective study, this problem is one of the main methodological limitations of this study.

In conclusion the suggestion in literature that the reduction osteotomy is superior to the 'simple' removal of the extra phalanx is not confirmed by the present study. Results of both procedures from the point of view of the interphalangeal joint are similar for functional measures as well as for self-rated function and activities of daily life. Therefore, in our opinion, there is no need for delaying surgery for the small extra phalanx in young children since in all these cases removal of the extra phalanx can be performed and procedures are exchangeable. However proper patient selection is necessary. To fully compare both procedures a prospective study should be conducted. It should also be noted that removal of the extra phalanx or combined reductive osteotomy is not always sufficient for length reduction in all cases and a reductive osteotomy of the first metacarpal should be combined if necessary.

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EIGHT

Surgical corrections for re-positioning and shortening of the full type triphalangeal thumb; long term outcome

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SUMMARY

Purpose

Triphalangeal thumbs present in many different variations; probably the most challenging types to correct as a surgeon are the 'five fingered' hands. These types can be differentiated among others in the development of the carpometacarpal joint; the joint can be a normal saddle joint type or a 'finger-like' joint with inadequate movement for a functional thumb. In this study the long-term outcome of two different surgical procedures to correct triphalangeal thumbs positioned in the same radio-ulnar plane of the fingers is presented.

Methods

Six patients (seven hands) following pollicisation and seven patients (nine hands) following rotation abduction and shortening osteotomy of the first metacarpal with removal osteotomy of the extra phalanx, were submitted to a standardized examination. Strength measurements were performed for opposition and flexion in the first metacarpophalangeal joint, and visual analogue scales were used for functionality and appearance.

Results

Both procedures, i.e. pollicisation and rotation abduction osteotomy, had an adequate opposition according to the Kapandji scoring (average respectively 6.1 and 6.8). Strength for opposition was diminished for both groups compared to a normal population (48% and 64%). Subjective measurements for functionality were on average 5.7 and 6.7. Scores for appearance were 5.5 and 7.6.

Discussion

Outcome for both surgical corrections proved adequate for function in daily life. Procedures are not interchangeable; the correct procedure is chosen based on the appearance of the first carpometacarpal joint.

INTRODUCTION

The five fingered hand is a well known entity in which the thumb looks like a finger because it contains three phalanges and is positioned in the plane of the fingers. This triphalangeal thumb is a developmental disorder; therefore the malformation can occur in many degrees of severity. The middle phalanx can be fully developed or can be more or less trapezoidal with more or less clinodactyly (Wood 1976). The "thumb" can be nearly parallel to the index, lying in the same plane of the hand, having a finger-like appearance or can be more abducted becoming more a long thumb. The metacarpal bone is always longer than a normal first metacarpal. The amount of additional length is depending. on the position of the growth plate. The growth plate can be either proximal, distal or both proximal and distal (Zuidam et al. 2006) . Polydactyly can be present on several levels from distal to proximal even with two CMC 1 joints. Regarding the extrinsics a full extensor mechanism can be developed like in a normal finger. The flexor tendons can be single or with a fully developed deep and superficial flexor tendon. The thenar muscles can be hypoplastic to nearly absent, often rudimentary tendons are present with hypoplastic muscles. As a result of this the MP joint can be instable. The adductor pollicis muscle can vary from nearly normal to very hypoplastic. The first web can be inadequate. Adequate palmar abduction and real opposition with opposing thumb nail to little finger nail is mostly not possible. However pseudo-opposition is nearly always possible. The 'thumb' can reach the small finger but not in the right plane. In unilateral cases it is mostly a sporadic condition. In patients with a bilateral triphalangeal thumb it is mostly an autosomal dominant treat.

Not only the five fingered hand but also a thumb with a trapezoidal shaped extra phalanx can be partly positioned in the plane of the fingers as well and can also lack sufficient intrinsic function and adequate first web. Therefore, both types may benefit from an extensive correction, besides solely removing the extra phalanx, which are challenging for the hand surgeon.

In literature, two surgical techniques are advocated for correction of these types of triphalangeal thumb (Upton and Shoen 2000; Hovius et al. 2004). The most used and well known technique is a pollicisation where most of the first metacarpal is excised. The other technique is a rotation, abduction and reduction corrective osteotomy of the first metacarpal combined with distal interphalangeal joint resection with reduction of bone of mainly the middle phalanx followed by an arthrodesis. The handling of the carpometacarpal joint is the major difference between both procedures. In pollicisation the CMC 1 joint is 'recreated' while in the corrective osteotomy the carpometacarpal joint is unaltered.

In our institution both procedures have been used for patients with triphalangeal thumb. While the number of patients of both groups is limited and does not allow statistical comparison, to our knowledge, both procedures have never been compared in the same study using similar outcome measures. Therefore, the aim of this article is to describe long term follow-up of patients operated with both methods and to provide recommendations for the technique to be used.

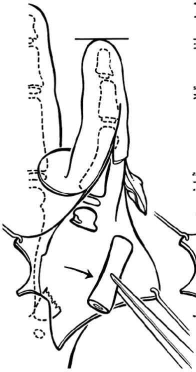

Figure 1A. Pollicisation: intraoperative view of metacarpal bone resection

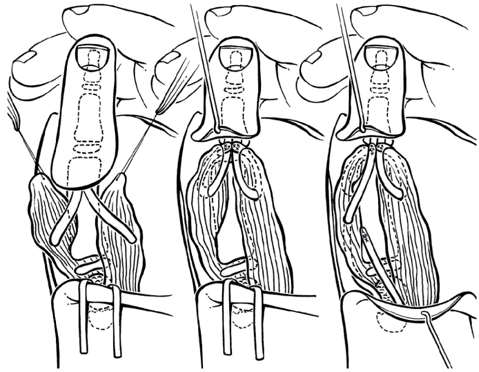

Figure 1B. Pollicisation: muscle and tendon reinsertion (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

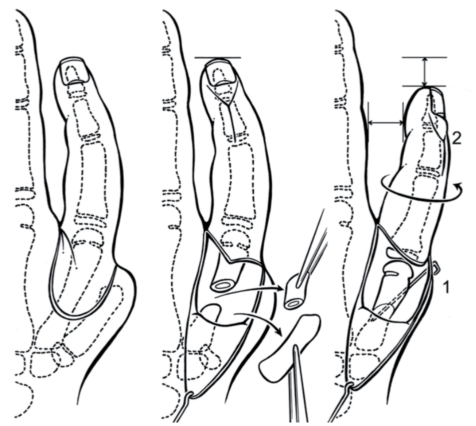

Figure 2A. Operation on metacarpal level (ROAMC1-procedure)

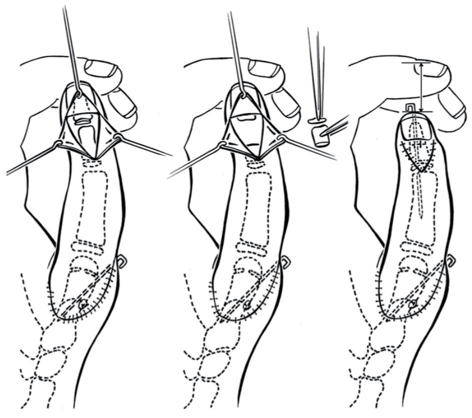

Figure 2B. Operation on phalangeal level (DIPRAD-procedure) (reprint from Hovius et al, Treatment of triphalangeal thumb, 2004)

PATIENTS AND METHODS

Patient Sample

In the period 1982 to 2013, in total 74 patients with triphalangeal thumb attended our centre. Of this group we selected patients with full type or trapezoid type with the finger in the same plane of the hand. Subjects under the age of four years were excluded because of the impossibility to perform the strength measurements. Subjects with disorders affecting general strength, peripheral nerve injuries or other disorders disturbing hand strength were excluded as well. Nine patients with sixteen affected hands were willing to participate in this study. Seven hands underwent pollicisation and nine had a rotation abduction correction osteotomy combined with distal interphalageal joint resection was performed. Age at operation ranged from one to seven years. The mean follow up was seven years (range three to eleven) and of the selected cases seven patients (78%) were female.

Surgical procedures

The pollicisation procedure was performed according to the description by Buck-Gramcko (Buck-Gramcko 1971) with modifications described by Foucher (Foucher et al. 2004; Hovius et al. 2004) (Figure 1). For the rotation abduction correction osteotomy, the first metacarpal was shortened, rotated and abducted palmarly. The extra phalanx was excised by a reduction osteotomy of the middle phalanx and arthrodesis of the distal interphalangeal joint was performed (Peimer 1985). The amount of reduction was determined by shortening the thumb up to the proximal interphalangeal joint of the index finger, when placed longitudinally next to the index finger. Extensor tendons and intrinsic tendons were shortened and repositioned, even when they were hypoplastic (Figure 2). If intrinsic musculature was not sufficient, an opponens plasty was performed in a later procedure.

Post-surgery rehabilitation was equal for both procedures; the thumb was immobilized for six weeks. In the corrective osteotomy group Kirschner wires were removed after six weeks. Afterwards a custom-made removable forearm splint was applied and mobilization was started under guidance of a hand therapist.

Measurements

To assess hand function objectively, all hands were examined in a standardized manner. The interphalangeal, metacarpophalangeal and carpometacarpal joint were manually tested for stability in the radio-ulnar plane. Joints were noted as unstable if deviation to stress was more than 5 degrees for the interphalangeal joint and more then 20 degrees for the metacarpophalangeal joint (Ogino et al. 1996). Opposition was measured using Kapandji's scale for opposition (Kapandji 1986), ranging from 0 (tip of the thumb on lateral aspect of the proximal phalanx of the index finger) to 10 (tip of the thumb reaches the distal palmar crease at the base of the little finger).

Intrinsic hand strength was measured using the Rotterdam Intrinsic Hand Myometer (RIHM) (Schreuders et al. 2006). Individual measurements were performed for the strength of opposition, flexion of the metacarpophalangeal joint of the thumb, and anteposition. The mean of three repeated measurements was registered. Measurements were calculated as percentages of reference values, corrected for the specific age group (Molenaar et al. 2008).

To evaluate hand function and appearance during daily life, all patients were asked to score function and appearance using a visual analogue scale, ranging from 0 (very poor/ugly) to 10 (very good/ beautiful). To assess function during activities of daily life, the ABILHAND-kids questionnaire was used. The ABILHAND-kids is a questionnaire created for children with cerebral palsy (Arnould et al. 2004; Penta et al. 1998). The questionnaire consists of 21 questions concerning mostly bimanual tasks. Children are asked to score if a task is easy, difficult or impossible to perform. Total scores range from 0 to 42, with 42 being the best possible score.

Statistical Analysis

Data were analysed as frequencies and percentages, or means and ranges. No statistical comparison analysis was performed as a result of small sample size (seven versus nine). Results were compared if possible as frequencies and means.

RESULTS

All subjects of both groups were able to perform opposition; mean Kapandji score for the pollicisation group was 6.1 and for the metacarpal osteotomy group 6.8. In both groups an opponensplasty was performed in a later stage if necessary, five in the pollicisation group and two in the metacarpal osteotomy group. Mean opposition strength for the pollicisation group was 48% of normative data, and for the metacarpal osteotomy group 64%. In both groups, one subject was not able to perform flexion in the metacarpophalangeal joint; mean flexion strength in the first metacarpophalangeal joint the pollicisation group was 49% and for the metacarpal osteotomy group 50% (Table 1). No instability of the interphalangeal and carpometacarpal joints was found in both groups. One subject had an instable metacarpophalangeal joint in the metacarpal osteotomy group, while no metacarpophalangeal joint instability was found following pollicisation.

Patient-rated evaluation of functionality were good for both groups; mean visual analogue scale score were 5.7 for the pollicisation group and 6.7 for the metacarpal osteotomy group. Results of the ABILHAND questionnaire for functionality also illustrated high functional scores: 35 and 40 of a maximum score of 42, respectively. Visual analogue scale scores for appearance were 5.5 for the pollicisation group and 7.6 following osteotomy of the first metacarpal and removal osteotomy of the extra phalanx (table 2).

Table 1. Function and strength measurements for both operative procedures

	Pollicisation (n=7) mean (SD)	Metacarpal osteotomy (n=9) mean (SD)
Kapandji score	6.1 (1.9)	6.8 (2.6)
Active Flexion of joint *in degrees*		
Interphalangeal	59 (34)	62 (24)
Metacarpophalangeal	42 (32)	44 (36)
Joint instability		
Interphalangeal	0	0
Metacarpophalangeal	0	1
Carpometacarpal	0	0
Strength in % (SD)		
Opposition	48.2 (16.8)	64.4 (18.5)
Flexion MP joint	49.4 (17.1)	50.4 (11.7)

Table 2. Patient-rated visual analogue scale scores and ABILHAND scores for both operative procedures

	Pollicisation (n=7) mean (SD)	Metacarpal osteotomy (n=9) mean (SD)
Functionality visual analogue scale score (0-10)	5.7 (3.3)	6.7 (2.3)
Appearance visual analogue scale score (0-10)	5.5 (2.6)	7.6 (2.1)
Abilhand questionnaire	35 (4.3)	40 (3.2)

DISCUSSION

Triphalangeal thumbs with the thumb in a finger-like position can present as a spectrum varying from the full type triphalangeal thumb with a less mobile and hypoplastic carpometacarpal joint to the trapezoid type combined with an aberrant position of the thumb and mostly a more adequate, but never a normal carpometacarpal joint. The carpometacarpal joint in triphalangeal thumb can vary from a nearly functional saddle joint, to a finger-like carpometacarpal joint, which can only move in the palmar to dorsal plane. In polydactylies with a fully developed triphalangeal thumb even two CMC 1 joints can be present, making the choice which CMC 1 joint should be preserved not easy, although the ulnar thumb and CMC1 joint are mostly more developed. As patients are mostly operated at a young age to correct the anomaly, the assessment of the development of the CMC joint is difficult.

Many authors have advocated to do a formal pollicisation in case of a five fingered hand. The reasoning being that the thumb resembles a finger and should be regarded and treated as if it were a finger with a CMC joint only moving in the dorsal to palmar plane. Therefore, as in the four fingered hand, the most radial 'finger' is pollicised. The difference however with the four fingered hand is the presence of a 'thumb' or rather a 'fifth finger with a CMC joint with a varying mobility'. The main disadvantage of leaving the original CMC1 joint intact is the inability to pronate in the thumb following correction

In this retrospective study patients have received more pollicizations in the beginning of the senior author's career. As the senior author encountered adult patients with unoperated five fingered hands with a rather powerful stable grip a change was made to preserve the original CMC 1 joint under the assumption the power grip would stay more stable and could be sustained longer. Therefore the reduction and abduction osteotomy of the first metacarpal was performed more often.

In this study, long-term outcome of two procedures to correct triphalangeal thumbs with the thumb in the same plane as the fingers are presented. The number and variety of patients together with the fact this study is not randomized makes comparison between the two techniques impossible. In both procedures results for function in daily life were adequate, although thumb strength was diminished (up to 49%). In both groups two opponens plasties were performed; strength and function of these patients were comparable with the rest of the group (data not shown). Good function is also represented by the high ABILHAND questionnaire scores and visual analogue scale scores, hereby indicating that pollicisation and osteotomy of the first metacarpal and reduction osteotomy of the extra phalanx and distal arthrodesis are both adequate procedures for the indicated anomalies.

Pollicisation in triphalangeal thumbs is not different from a pollicisation in other congenital anomalies (Foucher et al. 2004). Other outcome studies on pollicisation in the congenital hand have demonstrated the same functional results as in the present study (Clark et al. 1998; Staines et al. 2005). El-Karef described outcome of the reduction osteotomy of the first metacarpal and removal of the extra phalanx in 15 hands (El-Karef 2004). The difference with our technique is that the arthrodesis in El-Karef's study is performed at the PIP joint while we performed the arthrodesis at the DIP joint. The results for function as well as appearance were comparable with our study. In his series, in all hands, an opponensplasty was performed in the same stage or in a second stage. Adequate opposition was present in all patients. We performed an opponensplasty in two of the nine hands. All nine had an adequate and functional opposition, both in terms of mobility (mean Kapandji score of 6.8) and strength (64% of the normal population). The necessity of an opponensplasty remains to be decided based on the function and strength of an individual patient.

EL-Karef (El-Karef 2004) favours the reduction osteotomy combined with removal of the extra phalanx over the pollicisation, because of the possible stiffness of the interphalangeal joints. During pollicisation the proximal interphalangeal joint is used to create a metacarpophalangeal joint. If preoperative stiffness is present, good range of motion and adequate strength of the new metacarpophalangeal joint is difficult to achieve. While in triphalangeal thumb both interphalangeal joints can be stiff, the distal interphalangeal joint is more often affected. In our series, preoperative range of motion for both joints was adequate. The preoperative assessment of the joint mobility remains vital for the post-operative outcome. As the interphalangeal joints tend to be more stiff, it is however more stable compared to the metacarpophalangeal joint. The metacarpophalangeal joint shows more laxity. In our series, 1 of 16 (6%) patients had an unstable metacarpophalangeal joint post-operative, as in 27% in El-Karef series. In a pollicisation, this potentially unstable metacarpophalangeal joint is changed in to a carpometacarpal joint and the more stable proximal interphalangeal joint is used as the new metacarpophalangeal joint.

Both procedures are not exchangeable as a result of the functional limitations and additional correction of the carpometacarpal joint. However, the reduction osteotomy of the first metacarpal joint in patients with a triphalangeal thumb with a functioning, although hypoplastic joint is an adequate procedure for restoring position and function of the first ray. However, if the metacarpophalangeal joint is lax, a volar plate plasty or sesamoidesis can be performed. Although not statistically significant due to a limited number of patients the trend is definitively in favour of preserving the original CMC1 joint, therefore our first choice remains a rotation abduction reduction osteotomy at the metacarpal and a reduction arthrodesis at the DIP joint, partly removing the extra phalanx.

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NINE

Evaluation of function and appearance of adults with untreated triphalangeal thumbs

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ABSTRACT

Purpose

Triphalangeal thumb is a congenital malformation determined by an additional phalanx of the thumb. While surgical treatment of this condition is common practice, in the past this was not generally advised. Therefore, a population with an untreated triphalangeal thumb is still present. The purpose of this study is to compare function and appearance of adults with an untreated triphalangeal thumb to a normal population.

Methods

Twelve adults with 23 hands with an untreated triphalangeal thumb, unilateral or bilateral, were examined using objective measurements (thumb movement, joint instability, pain, and strength) and subjective measurements (Visual Analogue Scale (VAS), DASH, and SF-36).

Results

Objective measurements showed no limitations in range of motion or in grip and pinch strength. In addition, no joint instability was found in the interphalangeal joints. However, 5 thumbs had instability in the metacarpophalangeal joint. Strength of the thumb in anteposition was diminished to 64% compared to a normal population. Opposition was diminished to 62% and metacarpophalangeal joint flexion strength to 61%. The patients scored lower compared to a normal population for the domain of social functioning in the SF-36; the DASH showed no differences. VAS scores for appearance of the thumb were scored low (2.2 out of 10) by the adults, in contrast to VAS scores for function (7.7).

Conclusion

The examined group of adults with an untreated triphalangeal thumb had adequate thumb movement. Thumb strength was diminished for all specific thumb functions (anteposition, opposition, and thumb flexion), as low as 55%. Self-rated scores indicate that patients perceived their functionality as good. The appearance, however, was rated much lower, implicating a dislike of the thumb by the patients. This indicates that the main impact of an untreated triphalangeal thumb in daily functioning may not be the diminished function but rather the dissimilar appearance.

INTRODUCTION

Triphalangeal thumb is a congenital malformation determined by an additional phalanx of the thumb. The real incidence is unknown but up to 1:25.000 live births is reported [1]. Triphalangeal thumbs can vary considerably in appearance, from an additional delta phalanx to a non-opposable full length thumb (Figure 1 and 2). The extra phalanx at the interphalangeal joint level can result in problems with joint stability, deviation in the radial ulnar plane, and additional length. In addition, misalignment can occur as a result of the aberrant shaped extra phalanx [2]. Intrinsic musculature [3], the first metacarpophalangeal joint [4], and carpometacarpal joints can be aberrant as well. Wood [5] classified triphalangeal thumb by the shape of the extra phalanx (delta, trapezoid and full). Buck-Gramcko [6] presented a more extensive classification, incorporating additional factors (e.g. intrinsic musculature, web-space, and length) to plan the surgical procedure. The extended Wassel classification can be used to further classify combined radial polydactyly and triphalangism, if present [7].

Surgical treatment [8-12] is generally described to be focused on restoring function [13], although the effect on appearance of the thumb is also often considered important. In the past, however, this was generally not advised [14, 15]. Furthermore, variations of the deformity were not always recognized as a handicap in daily life. Therefore, a population with an untreated triphalangeal thumb is still present. This population with untreated triphalangeal thumbs does not always seek medical treatment, which may indicate that the thumb function may be more or less adequate in daily life. However, to our knowledge, the function in daily life of adults with an untreated triphalangeal thumb has never been studied. Therefore, the purpose of this study was to compare function and appearance of adults with untreated triphalangeal thumbs to a normal population.

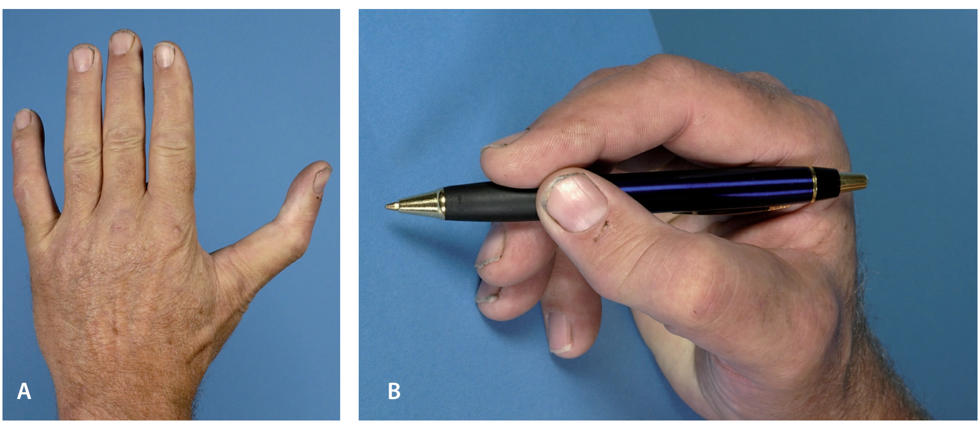

Figure 1 A-B. Conventional pictures of a patient with bilateral triphalangeal thumb A. Left hand dorsal view, showing extra length of the thumb. B Dominant hand writing, showing the deviation of the thumb.

Figure 2. Radiograph of a patient with bilateral untreated triphalangeal thumb. Radiograph of left thumb in radial abduction showing a rectangular type triphalangeal thumb.

Table 1. Characteristics of total group of untreated triphalangeal thumbs.

N = 12	H= 23		N	
Gender	Male		3 (25%)	
	Female		9 (75%)	
Dominance	Left		0	
	Right		12 (100%)	
Type of triphalangeal thumb				
	Wood [5]	Buck-Gramcko [6]		
		Rudimentary	-	0
	Delta		0	-
		Short Triangular		4
	Trapezoid		22	-
		Larger Transitional		11
	Full		1	-
		Long Rectangular	-	1
		Hypoplastic	-	0
		Associated with radial duplications		7

N= number of adults, H= hands

PATIENTS AND METHODS

Sample Characteristics

We included a total of 17 parents or grandparents with untreated triphalangeal thumbs, accompanying there treated children with triphalangeal thumb during a visit to our center for a research study, of which 12 participated in this study. Mean age was 48 years (range 36 to 63 years), 75% was female and all persons were right dominant. One person had unilateral involvement; all others had bilateral triphalangeal thumbs, creating a total group of 23 hands.

The triphalangeal thumbs were classified using Wood's classification [5] by shape of the extra phalanx; delta, trapezoid and full type triphalangeal thumb and by Buck Gramcko's classification by treatment options [6] (see Table 1).

Objective Measurements

To assess hand function objectively all hands were examined in a standardized manner. Measurements and execution of the measurements were selected to maximally allow comparison with reference data from a normal population described in the literature.

Deviation in the radial-ulnar plane was measured in degrees for all interphalangeal and metacarpophalangeal joints that were present. These joints and the carpometacarpal joints were examined for pain and joint instability. Joint instability was tested manually. Since healthy joints also have a certain laxity, following Onigo et al. [16], we defined joint instability as more than 5 degrees deviation to stress in the lateral plane of the interphalangeal joint or more than 20 degrees deviation to stress in the lateral plane of the metacarpophalangeal joint.

Range of motion in the interphalangeal and metacarpophalangeal joint was measured using a standard goniometer. In addition, we measured the maximum active radial abduction, anteposition, and retroposition of the thumb. Radial abduction was measured as the maximum angle between the first and second metacarpal in the plane of the palm. Anteposition (palmar abduction) of the thumb was measured as the maximum angle between the first and second metacarpal of the thumb perpendicular to the plane of the palm [17]. Retroposition was measured as the distance in centimeters from table to top of the thumb, with the hands flat on the table and the thumb maximally lifted [18]. Kapandji's method for scoring opposition [19] was used, ranging from 0 (tip thumb on lateral aspect of proximal phalanx of index finger) to 10 (tip thumb reaches distal palmar crease at base little finger).

Grip strength and key (lateral) pinch strength were examined using a JAMAR dynamometer according to the protocol described by the American Society of Hand Therapists [20]. Intrinsic hand strength was measured using the Rotterdam Intrinsic Hand Myometer (RIHM) in a standardized protocol [21]. The RIHM can measure the strength of the individual fingers and thumb in different directions and can, in some situations, measure intrinsic hand musculature in isolation. Individual RIHM measurements were performed for (1) thumb opposition strength, primarily an opponens pollicis muscle function,

(2) flexion strength of the metacarpophalangeal joint of the thumb, primarily a flexor pollicis brevis muscle function, (3) anteposition (palmar abduction) strength of the thumb, primarily a function of the abductor pollicis brevis muscle, and (4) radial abduction strength of the index finger, a function of the first dorsal interosseus muscle.

For all strength measurement, the mean of three repeated measurements was registered. Besides strength measurements, all movements were also graded in task performance (0 not able to perform, 1 can fulfill task, 2 fulfills task in a different manner, 3 minor adaptation, 4 normal technique)[22]. If the subject was not able to perform a specific measurement the strength was scored as 0. All measurements were compared to a normal population correcting for age, gender and dominance.

Subjective Measurements

All persons were asked to score their own thumbs for functionality and appearance using a Visual Analogue Scale, ranging from 0 (very poor/ugly) to 10 (very good/beautiful). Scores were measured for both hands separately. Furthermore, patients were asked to fill out the Disability of the Arm, Shoulder and Hand (DASH) questionnaire [23] to measure impairment and limitations in activity and participation. The DASH scales range from 0 to 40, with lower scores indicating a higher or better performance level. The DASH measures overall arm and hand function. Overall health status was assessed using a generic health questionnaire: the Medical Outcome Study 36-item short form health survey (SF-36). The SF-36 contains 8 different item scales: Physical Functioning, Role Physical, Bodily Pain, General Health, Vitality, Social Functioning, Role Emotional, and Mental Health. Scales range from 0 to 100, with higher scores indicating higher level of wellbeing.

Table 2. Specific ROM measurements of the untreated triphalangeal thumbs.

N=12 H=23					
		mean	**minimum**	**maximum**	**SD**
Deviation in lateral plane	MP (deg)	0	0	0	0
	IP	20	0	45	15.4
Opposition		8	4	10	1.8
Radial Abduction (deg)		80	45	109	15.0
Anteposition (deg)		67	48	90	12.5
Retropostion (cm)		4.1	2.0	5.5	0.9
Flexion	MP	61	45	75	8.9
	DIP	9	0	60	18.5
	PIP	67	0	95	28.5
Extension	MP	19	0	45	15.5
	IP	3	0	20	13.0
		H			
Joint instability	MP	5 (22%)			
	IP	0			

N= number of adults, H= hands, MP = metacarpophalangeal joint, DIP= distal interphalangeal joint, PIP= proximal interphalangeal joint, deg = degrees, cm = centimeters.

RESULTS

No specific joint pain was present in any of the subjects. In addition, there was no deviation in the radio- ulnar plain in the metacarpophalangeal joint in any of the subjects. However, deviation in the interphalangeal joints, which is zero in a healthy population, was present in 18 hands (78%), all in ulnar direction. Mean degree of deviation was 20 degrees (SD 15, range 0 to 45) (see Table 2). The standardized x-ray examination showed besides the triphalangeal thumb three persons with carpometacarpal joint arthrosis (13%) (Figure 3).

Range of motion of the triphalangeal thumb in the different directions was not reduced (see Table 2). On the contrary, mean active radial abduction was larger compared to a normal population [24]. Measurements for retroposition had a mean of 4.1 centimeters (SD 0.9), the mean of anteposition was 67 degrees (SD 12.5). No joint instability was found in the distal or proximal interphalangeal joint. However, 5 thumbs had instability in the metacarpophalangeal joint with an excursion up to 30 degrees. None of the adults had an instable carpometacarpal joint of the thumb.

While some persons had diminished opposition, most adults of the group could reach a thumb-finger pinch up to the proximal interphalangeal crease of the little finger (8 in the Kapandji evaluation) [19]. Flexion and extension range of motion in the metacarpophalangeal, distal and proximal interphalangeal joints were not significantly reduced. However, impaired movement, both actively and passively, was always present in one of the additional interphalangeal joints. In 20 thumbs (87%) the distal interphalangeal joint was insufficient. In resting position, nine persons had the thumb in a flexed position without extension (one metacarpophalangeal joint, eight interphalangeal joint), up to 40 degrees.

Figure 3. Radiograph of a patient with bilateral untreated triphalangeal thumb. Radiograph of left thumb in radial abduction showing a trapezoid type triphalangeal thumb with arthrosis of the carpometacarpal joint.

Grip strength of the dominant hand for the total group had a mean of 23 kilograms, for the non-dominant hand 21 kilograms. Key or lateral pinch could not be performed by one person for both hands because of the length and deviation of the thumb; in this case a zero was scored. Mean key pinch for the dominant and non-dominant hand was 6.2 and 5.7 kilograms, respectively. One person could not perform anteposition at all. The strength of anteposition was 45 Newton for the dominant hand and 44 Newton for the non-dominant hand. Opposition strength could not be measured in one person, because of the lack of opposition; in this subject, a zero was scored. The group had a mean opposition strength of 58 and 46 Newton. All persons were able to flex their metacarpophalangeal joint, the measured strength for the dominant hand was 61 Newton.

The mean value for the Visual Analogue Scale scores regarding self-rated function for the dominant hand was 7.7 (SD 2.5), and for the non-dominant hand 8.1 (SD 2.2). Visual Analogue Scale scores for appearance were respectively 2.2 (SD 1.9) and 2.6 (SD 2.8). Results of the DASH questionnaire had a mean of 8 (SD 13) with a range for the total group from 0 to 38. The results of the SF-36 were for mental health 84.0 (SD 17.0), vitality 64.1 (SD 22.8), bodily pain 87.9 (SD 23.0), general health 72.5 (SD 21.1), physical functioning 87.7 (SD 21.7), role limitations due to physical problems 88.6 (SD 30.3), role limitations due to emotional problems 81.8 (SD 34.5), and for social functioning 64.7 (SD 20.9).

Table 3. Strength measurements presented by force direction and dominance in absolute forces

Movement		**Force**			
	Side	**mean**	**minimum**	**maximum**	**SD**
Grip strength (Kg)	Dom	23	9	43	10.1
	Non	22	7	47	12.8
Key/Lateral grip*(Kg)	Dom	6.2	0	11.2	2.8
	Non	5.7	0	10.0	2.6
Abduction index finger (N)	Dom	36	17	71	15.1
	Non	30	15	56	11.8
Anteposition* (N)	Dom	45	0	76	20.8
	Non	44	0	68	20.5
Opposition thumb* (N)	Dom	58	0	142	33.6
	Non	46	0	95	24.2
Flexion MCP thumb (N)	Dom	61	40	109	17.7
	Non	51	23	68	12.3

Dom = dominant hand, Non = non dominant hand, N = Newton, Kg = kilograms, SD standard deviation.
*For opposition as well as for anteposition and key grip one person was not able to perform the specific movement or task. Strength was therefore scored as 0 for these specific subjects.

DISCUSSION

Twelve adults (23 hands) with untreated triphalangeal thumbs were examined using objective measurements and subjective evaluation. Our study is, to our knowledge, the first to report on function of patients with untreated triphalangeal thumbs. At present, there is only a small group of these adults in the Western world. In earlier literature, surgical treatment for triphalangeal thumb was not advised [14, 15]. Nowadays, surgical treatment is general practice. Many different surgical procedures have been described for the existing variety of triphalangeal thumbs [3]. As a result of limited availability of subjects, the main limitation of this study is the small power (n = 12). Another limitation is a possible bias in group selection. The included subjects did not seek surgical intervention, probably because of a good hand function. One could imagine that subjects with poor function seek intervention more actively.

Thumb movements of the examined group of untreated triphalangeal thumb were comparable with measurements from literature in a normal population, the Kapandji score for opposition was respectively 8 for the examined group and 8-10 in a normal population. The measurements for anteposition were respectively 67 degrees versus 40-80 degrees, retroposition respectively 4,1 versus 3,5 to 7,0 centimeters, and radial abduction 80 versus over 50 degrees [17, 24, 25]. However, two adults

Table 4. Self-reported evaluation of function and appearance using a Visual Analogue Scale score, the Short Form 36 (SF-36) and the Disability of the Arm, Shoulder and Hand (DASH)

Movement		
N=12	**mean**	**SD**
VAS Function		
Dom	7.7	2.5
Non	8.1	2.2
VAS Appearance		
Dom	2.2	1.9
Non	2.6	2.8
Sf-36		
Social Functioning	64.7	20.9
Mental Health	80.4	17.0
Energy Vitality	64.1	22.8
Bodily Pain	87.9	23.0
General Health	72.5	21.1
Physical Functioning	87.7	21.7
Role limitation due to Physical problems	88.6	30.3
Role limitation due to Emotional problems	81.8	34.5
DASH	8	13

N= number of adults, Dom = dominant hand, Non = non dominant hand, SD = standard deviation,VAS= visual analogue scale score, 10 optimal score, SF-36= short-form 36, 100 optimal score, DASH= Disability of the Arm, Should and Hand, 0 optimal score.

were not able to perform one of the tasks (one anteposition and one opposition). Joint movement of present interphalangeal and metacarpophalangeal joint was adequate, although instable metacarpophalangeal joints were present in 22% of the population. Outcome of the general strength measurements (grip strength and key grip) are comparable to a normal population. Grip strength was on average reduced compared to the mean of the normal population [24]: for the dominant hand grip strength was 77% of normal standard (range 38 to 130%) and for the non dominant hand 79% (range 30 to 143%). Overall, mean key pinch strength was higher than for the normal population: dominant hand 109%, and non-dominant mean hand 108%. This increased key grip strength could be a result of adaptation to the diminished strength of the other thumb functions. In contrast to the grip strength and key grip, strength was diminished for all specific intrinsic thumb functions. Anteposition strength of the dominant hand was diminished to 64% and for the non-dominant hand to 68%. Opposition strength was reduced to 62% and 50%, respectively, with again one subject that could not perform any opposition strength. Metacarpophalangeal joint flexion strength could be performed by all subjects and was reduced to 61% and 55% of the normal populations. The three adults with arthrosis of the carpometacarpal joint had no significant difference for strength and range of motion compared to the total group.

The adults with untreated triphalangeal thumbs indicated that functionality is good suggesting that an untreated triphalangeal leads to only limited disability in daily life. The appearance, however, was rated much lower, implicating a dislike of the thumb by the involved group. This indicates that the main impact of an untreated triphalangeal thumb in daily functioning may not be diminished function but probably the dissimilar appearance. Therefore the overall health scores (SF-36) were diminished for social functioning only. The group with untreated triphalangeal thumbs scored a mean of 64.7 (SD 21.0) compared to a normal population 84.0 (SD 22.4) [26].

For a good overall function of the thumb not only anatomical function should be optimized but also the thumb and hand should be easily used in all circumstances. However it must be said that there are groups with strong religious beliefs, who have no problems with their anomaly as well as for appearance as for function. In the studied group most of the patients hide their involved hand. Therefore, not only psychosocial functioning may be disturbed but also normal use when they are not alone.

In summary, objective measurements showed no limitations in range of motion in this group of adults with an untreated triphalangeal thumb. Hand strength was equal compared to a normal population. However, thumb strength was diminished in the different directions. The adults with untreated triphalangeal thumbs scored their own thumb low for appearance but much higher for functioning. We suggest that the triphalangeal thumb should therefore be interpreted as an entity which requires surgical intervention if desired by the patient to optimize function but, maybe more importantly, to improve appearance, hereby giving the patients a more "usable" thumb.

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TEN

Discussion and future perspectives

'It is better, of course, to know useless things than to know nothing'

Seneca

This chapter discusses the results of our research on outcome of the triphalangeal thumb as a whole. Firstly the questions asked in the introduction are answered with the results of the previous chapters. Furthermore the limitations of this thesis are discussed and future perspectives are presented.

What is so abnormal or different about a triphalangeal thumb?

The triphalangeal thumb is an uncommon congenital abnormality of the thumb. The extra phalanx is only a small aspect of the deformation, which has a much wider presentation. The thumb as a whole can be altered; joints, ligaments, nails, intrinsic and extrinsic musculature, phalanges and metacarpals can all be deformed besides the present extra phalanx.

The population of this thesis was similar to other reported cases in literature. The preoperative assessment of the thumb, which can be very hard to perform in young children, remains imperative for a good treatment strategy. Based on the findings in this thesis, however, some additional guides for this difficult preoperative assessment can be given.

1. The position and presence of possible additional growth plates are an indication of additional length or growth potential of the first metacarpal. As described in **Chapter two**, first metacarpals with double growth plates are larger compared to metacarpals with only one growth plate. Besides the additional length present at time of the measurements, the double growth plate provides an additional growth potential during the years. If the growth plate is positioned at the distal end of the metacarpal, the metacarpal is also longer, however the additional growth during the years as seen with the double growth plates, is not present. Although smaller, additional length is also present in cases with the growth plate on the proximal side of the first metacarpal, which is the normal side in children. The position of the growth plate has a significant relationship with the type of triphalangeal thumb, and therefore indirect with the length of the first metacarpal as presented in **Chapter three**.

Based on these findings, therefore, we conclude that in clinical practice the position is of growth plate is important for establishing the necessity of a reduction osteotomy of the first metacarpal. Unfortunately, the exact amount of reduction needed in specific cases depends on good judgment rather than on solid scientific evidence.

2. Strength and therefore function can be diminished in the triphalangeal thumb, as many authors earlier described for the five-fingered hand. As mentioned earlier, the thumb as whole can be altered even in the less complex triphalangeal thumb types. In **Chapter four** we described diminished strength in the delta and trapezoid type of triphalangeal thumb. Average opposition strength was 63% of the strength in a normal population. The diminished strength of the triphalangeal thumb also lowered grip strength of the hand, which was 69% of the normal population.

Our findings on diminished thumb strength indicate that, in clinical practice, strength-enhancing procedures can be necessary in all types of triphalangeal thumb. Because determining the exact amount of thumb strength can be tricky in very young children, we suggest that an opponens plasty can be delayed until it becomes clear that strength is importantly diminished rather than absent.

So there are many different types of triphalangeal thumb?

Congenital anomalies, including triphalangeal thumb, can have all different kinds of appearances. The shape of the extra phalanx alone is not a 'standard' shape, it rather is a pallet of shapes and sizes. In addition, radial polydactyly can occur in addition to the triphalangeal component and the deformed thumb can be a part of a larger syndromal presentation. The additional thumb components also have a wide variance in appearance. The presence of rather simple radial polydactyly components can vary to several complex components resulting in triplications of the thumb. In **Chapter five** we present several cases of triphalangeal thumbs with three thumbs; triplication, from our population. Although more challenging to correct, the main focus remains the same as in less complex radial polydactyly with triphalangeal thumb; removal of additional rays, reconstruction of joint surface, reduction of length and adequate positioning of the thumb.

We as clinicians try to squeeze all these different sizes and shapes into classifications to facilitate communication and surgical protocols. Many different classifications are present in daily literature for radial polydactyly as well as for triphalangeal thumb. Some authors combined classification of both anomalies because of the often simultaneous presentation. The cases described in chapter five could not be classified using the known classifications. Therefore, in **Chapter six**, an altered classification is presented. The presented classification focuses on the present anomalies; e.g. triphalangeal component, deviation of the joint, position of duplicature of bones. With the altered classification does not divide the anomalies into subtypes, it rather facilitates the nomenclature.

In clinical practice complex and extensive classifications are not always useful, because they often do not provide a surgical protocol or plan. In our minds classifications are more a tool to record the anomaly in exact and precise manner to compare in research or confer with others.

The different types of triphalangeal thumb and their broad presentation are further pointed out in **Chapter four**. In this chapter, strength of 38 hands with delta or trapezoid type triphalangeal thumb in terms of precision grips (key grip, tip pinch) and power grip, as well as specific strength measurements of the thumb: opposition, MP joint flexion, anteposition and radial abduction strength. We found that all measurements strength were statistically diminished compared to a normal population. However, at the same time, the range of strength values is broad and the strength may generally still be sufficient for daily life. Therefore no direct surgical recommendations for the opposable triphalangeal thumb group can be made. Opponens plasty or other strength enhancing procedures can be necessary in the individual patient.

Can we do anything about it?

Off course we can! The treatment options for the different types of triphalangeal thumb are described in the introduction. In **Chapter seven**, we described in more detail two techniques for correction of an extra phalanx in the opposable triphalangeal thumb: removal of the extra phalanx and stabilization of the newly created joint versus the combined reduction osteotomy with resection of the distal joint followed by arthrodesis. In this chapter, outcome is described in 33 affected hands. The main goal of the chapter was not two compare the two techniques because the patients receiving both techniques were not similar. For example, patients with a small extra phalanx are not suitable for the combined reduction osteotomy technique. Therefore a selection bias will always be present when comparing patients and techniques. However, patients with an average to large extra phalange are suitable for both techniques. In the chapter, no differences between the two techniques are seen in functional measurements (range of motion mean, joint deviation, instability) while minor differences in subjective appearance and function are found in favor of the removal of the extra phalanx, although not statistically significant. These minor changes may be as a result of the degree of abnormality; the combined reduction osteotomy technique will be performed more often in the patients with the larger extra phalanx. This group with a larger extra phalanx has probably a more abnormal thumb, although this not measurable in a preoperative setting.

In **Chapter eight** the treatment options for the non-opposable triphalangeal thumbs are described. In this chapter, long term follow up of two techniques used on 33 hands are studied; firstly removal of the extra phalanx and stabilization of the newly formed joint and secondly the combined reduction osteotomy with the resection of the distal joint followed by desis. Both techniques have a good subjective score for function with visual analog scale scores of 8.1 versus 7.4 out of a possible 10, however subjective appearance scored less with respectively 5.8 and 4.8. So one can concluded that both techniques are provide a good outcome in opposable triphalangeal thumbs.

The combined reduction osteotomy is inappropriate for the smaller extra phalanges because it is technically demanding or sometimes impossible, therefore techniques are difficult to compare. Both techniques are suitable for the larger extra phalanges, although the prospective setting does not allow comparison of two groups with these larger phalanges.

Do we really need to do anything about it?

In **Chapter nine** we describe patients with triphalangeal thumbs without surgical intervention. Flexion was reduced in interphalangeal and metacarpophalangeal joints, the most in the distal interphalangeal joint (mean 9 degrees, range 0-60). All of these 23 hands had diminished strength for thumb movements (opposition, MP joint flexion, anteposition, and radial abduction); therefore we would call these 23 hands aberrant or 'different' compared to normal standards. However, despite strength loss, function in daily live measured with the DASH scores were comparable to a normal population (8 out of a possible 40). The subjects reviewed the function of their thumbs on a visual analog scale as good (7.7 out of 10). So, in their opinion, their hands were useful in their daily life. Does this mean that their daily life is adapted to the function of their hands or that the function is sufficient? The answer to this question is not something we investigated in this thesis. However, one could assume that their daily life is adapted to the function of their hand. Average function must still be possible so it is probably a bit of both. On the other hand, for appearance, they score a 2.2 out of a possible 10 on a visual analog scale. Although no comparative measurements for esthetics of the hand in a normal population are available, this score is very low. Another question arises regarding the appearance of the hand: does this low score on appearance of the hand influence the daily function in a social environment. In chapter ten, subjects also filled out a Short Form (SF) 36 questionnaire. They scored significantly lower on the sub score social functioning (64.7, SD 20.9) compared to a normal population (84.0, SD 22.4). However the sub score 'role limitations regarding functional problems' were actually higher in the investigated group (88.6 vs 76.4). These outcomes are difficult to explain; it is not easily said the appearance of their hand i.e. thumb, refrains them from using the hand in daily life or in public, although the sub score for social functioning is lower.

So the answer to this question depends on the demands of the patient. Primary goals of intervention in the triphalangeal thumb, as mentioned in the introduction, dependent on the specific deformity. Opposition of the thumb is a basic function in daily life, although patients without opposition develop an alternative grip: the scissor grip. This creates a usable grip in daily life, though not as powerful as precision grips (key, pinch, tripod). So the answer to the above mentioned question could be: "no not really". The best possible answer in our opinion, remains "if the patients asks for an improvement in function or esthetics".

LIMITATIONS

No limitations in any research, let alone in a research on congenital anomalies would be a miracle. Since this thesis is no miracle is has its limitations;

The studied population with a triphalangeal thumb is very heterogeneous in regard to presentation, type of anomaly and comorbidity. Combined with the small amount of patients, presents us researchers with an impossibility of comparing larger groups or creating randomized controlled trials.

In addition to above-mentioned limitations, final outcome of congenital anomalies changes during growth until patients reaches adult life. Besides the growing up process of the patient, the surgeon changes the intervention strategy with minor or major changes or amendments.

FUTURE PERSPECTIVES

People will be different, hands will be different, thumbs will be different, all depending on the perception of society and the individual itself. Will there be a need for intervention in triphalangeal thumbs? Of course! A non-opposable thumb is improved with enhancing position and motion, although high level evidence to substantiate this opinion is hard to find, even in this thesis. For all congenital anomalies an individual approach is necessary and the surgeon must always explain and discuss the possibilities of intervention with the patients and its caregivers. This thesis has shown that many different surgical interventions are possible and with good results. The choice of procedure will be a shared decision with patient, parents and surgeon, supported by a hand therapist and rehabilitation specialist.

In an ideal world, patients could be tested and classified regarding type of triphalangeal thumb, so that in the future we would be able to treat every patient similarly and based on the highest level of evidence all over the world. Unfortunately, or luckily, this will never happen; unfortunately for the surgeon who likes to categorized every patients and treat them accordingly, therefore the surgeon needs to be well informed with the different types of clinical presentations and interventions options and apply on the material at hand. Would either of the above be better for the patient? Probably not, the battle between the wish for cookbook surgery and the adage " cut as you go" will be beneficial for the patient. Especially while congenital anomalies will always be a watershed of specific anomalies, although this watershed can be a small nightmare for every starting researcher.

Summary

A triphalangeal thumb is recognized by the extra phalanx located between the proximal and distal phalanx. The shape and size of this phalanx can differ from a very small delta type extra phalanx to a fully developed extra phalanx and consequently give the thumb a finger-like appearance. Besides the extra phalanx, a triphalangeal thumb can have aberrant intrinsic musculature, joint instability, inadequate placement of the thumb, and many associated anomalies. Therefore clinical presentation has a broad spectrum of anomalies not only located in the hand. This thesis is focused on the clinical outcome of triphalangeal thumb, and on the many different aspects of triphalangeal thumb influencing these results. We subdivided it into two parts: the aspects of the triphalangeal thumb in the preoperative assessment and the outcome of the performed surgical procedures on patients with triphalangeal thumbs.

Preoperative assessment and classification

The first focus of preoperative assessment was on the presence and position of the epiphyses of the first metacarpal. **Chapter two** reviews the different positions of the epiphyses and their relation with the length of the first metacarpal in triphalangeal thumb. The distally-placed growth plate was the most common variety in 52%, the proximal position in 33% and double epiphyseal plates in 15%. All ratios of TPT patients were significantly smaller compared to a normal population, indicating a longer first metacarpal in this condition. First metacarpals in TPT with double epiphyses grew disproportionately longer than in a normal population, while those with distal epiphyses grew disproportionately less than normal. First metacarpals with proximal epiphyses grew with the same rate as normal first metacarpals.

In **Chapter three** the length of the first metacarpal is analyzed in relation to the different types of triphalangeal thumbs and the position of the epiphyses of the first metacarpal. In total, 59 hands in 37 patients with a triphalangeal thumb were examined for thumb type (delta 31, trapezoid nine and full type 19), growth plate location, and relative length of the first metacarpal. The first metacarpal in all three triphalangeal thumb types was significantly longer than in a normal population. The length of the first metacarpal was related to the site of the growth plate. We didn't find any relation between type of triphalangeal thumb and final length of the thumb

In **Chapter four** intrinsic muscle strength of all the different types of triphalangeal thumb was measured individually. Our clinical impression was that intrinsic musculature is probably affected in all forms of triphalangeal thumb. Therefore we established the strength of 38 thumbs in subjects with a triphalangeal thumb. On average, strength was significantly diminished for all thumb functions, up to 63% for opposition strength. Strength for the power grip was on average 70%. Although strength of the thumb is diminished, for the investigated group it is apparently sufficient in daily life, as they did not seek surgical enhancement. However, reconstructive procedures enhancing intrinsic musculature must be considered in all types of triphalangeal thumb

In **Chapter five** eleven cases of triplication or triplicated thumb combined with triphalangism are reported. Only one of these eleven triplicated thumbs could be classified according to the currently most used classifications. In all cases aberrant rays were excised, thumb length and alignment restored by osteotomies, joints were stabilized, tendons reinserted and nails and nail walls corrected if necessary. Also in all cases a correction of triphalangeal components was performed.

Chapter six describes an adjusted classification that allows classification of all different types of triphalangeal thumb associated with radial polydactyly. Patients from 1993 to 2006 with radial polydactyly (n=104) were identified from the hospital database with a total of 121 affected hands. All x-rays were carefully examined and classified according to the existing classifications for radial polydactyly and a modified classification. In the modified nomenclature Wassel's level of duplication is preserved. Type VII and VIII are assigned for partial or complete duplication of the carpal bones according to Buck-Gramcko. Triplication and triphalangeal components can be assigned to each type of radial polydactyly by suffices. Symphalangism, deviation, and hypoplasia can also be classified. Triplication on different levels of the thumb is classified by determining and including the different types of the original Wassel classification. Eighteen thumbs with triphalangeal components or triplication could not be classified according to existing classifications for radial polydactyly. After using the proposed classification, all patients could be classified. We propose a modified classification that is practical and utilitarian for nomenclature of radial polydactyly and that may assist comparison of treatment outcomes and individual cases.

Treatment and long term follow up

The second part of the thesis is focused on clinical outcome and daily use of persons with a triphalangeal thumb. **Chapter seven** discusses clinical outcome of two different treatment strategies on removal of the extra phalanx in less complex triphalangeal thumb. The procedures are: 1) removal of the extra phalanx and stabilization of the remaining joint; and 2) a combined reduction osteotomy with resection of the distal joint followed by arthrodesis. We treated 20 patients (33 hands); In 17 hands the extra phalanx was removed and in 16 hands we used the combined osteotomy procedure of distal joint removal and arthrodesis. None of the patients in either group had an unstable interphalangeal joint. Arthrodesis was performed when the extra phalanx was larger or when the child was older. The mean radial or ulnar deviation in the interphalangeal joint was respectively 5 degrees and 9 degrees. Mean active flexion in the interphalangeal joint was respectively 35^0 and 46^0 in the two groups. Results for both procedures are similar for both objective measures and self-rated function and activities of daily living. Either surgical approach seems reliable.

Chapter eight discusses operative treatment for the specific types of triphalangeal thumb and addresses the possible procedures regarding their outcome. The long-term outcome of two different surgical procedures to correct triphalangeal thumbs positioned in the same radio-ulnar plane of the fingers is presented. Six patients (seven hands) following pollicisation and seven patients (nine hands) following rotation abduction and shortening osteotomy of the first metacarpal with removal osteotomy of the extra phalanx were submitted to a standardized examination. Both procedures, i.e. pollicisation and rotation abduction osteotomy, had an adequate opposition according to the Kapandji scoring (average respectively 6.1 and 6.8). Strength for opposition was diminished for both groups compared to a normal population (48% and 64%). Subjective measurements for functionality were on average 5.7 and 6.7. Scores for appearance were 5.5 and 7.6. Outcome for both surgical corrections proved adequate for function in daily life. Procedures are not interchangeable; the operative procedure is chosen based on the appearance of the first carpometacarpal joint.

In **Chapter nine** the function of adults with untreated triphalangeal thumbs are evaluated and compared to the normal population. Twelve adults with 23 hands with an untreated triphalangeal thumb, unilateral or bilateral, were examined using objective measurements (thumb movement, joint instability, pain, and strength) and subjective measurements (Visual Analogue Scale (VAS), DASH, and SF-36). Objective measurements showed no limitations in range of motion or in grip and pinch strength. Strength of the thumb in anteposition was diminished to 64% compared to a normal population. Opposition was diminished to 62% and metacarpophalangeal joint flexion strength to 61%. VAS scores for appearance of the thumb were scored low (2.2 out of 10) by the adults, in contrast to VAS scores for function (7.7). The examined group of adults with an untreated triphalangeal thumb had adequate thumb movement and self-rated scores indicate that patients perceived their functionality as good. The appearance, however, was rated much lower, implicating a dislike of the thumb by the patients. This indicates that the main impact of an untreated triphalangeal thumb in daily functioning may not be the diminished function but rather the dissimilar appearance.

The triphalangeal thumb is a complex entity with different appearances. In preoperative assessment the position of the growth supplies information on the growth potential of the first metacarpal, even in the less complex types. The intrinsic muscle strength can also be affected in all types; which can be diminished up to 63%. The outcome of the described procedures for the complex and more simple triphalangeal thumbs, are interchangeable comparing results. However these procedures are not interchangeable for individual patients, because all of them demand specific conditions of the thumb to be present. In this thesis several subjects concerning the triphalangeal thumb are clarified regarding preoperative analysis and surgical procedures, however in practice it remains a challenging diagnosis.

Samenvatting

De triphalangeale duim is herkenbaar aan de extra falanx tussen de proximale en distale falanx. De vorm en grootte van deze falanx kan erg verschillen. Er kan alleen een extra falanx aanwezig zijn, dan wel is de extra falanx compleet ontwikkelt en heeft de duim het uiterlijk van een vinger. Naast de extra falanx kan de intrinsieke musculatuur veranderd zijn, de gewrichten instabiel, de duim anders geplaatst zijn en meerdere geassocieerde afwijkingen aanwezig zijn. Daarom kan de klinische presentatie erg wisselen en kunnen er ook afwijkingen buiten de hand aanwezig zijn. Dit proefschrift is gericht op de klinische uitkomsten van de triphalangeale duim en op de factoren welke deze resultaten kunnen beïnvloeden. Het proefschrift is onderverdeeld in twee delen: preoperatieve inventarisatie en de uitkomst van verschillende chirurgische interventies bij patiënten met een triphalangeale duim.

PREOPERATIEVE BEOORDELING EN INDELING

De eerste factoren welke preoperatief beoordeeld werden is de aanwezigheid en positie van de groeischijf van de 1e metacarpaal. In **hoofdstuk twee** worden de verschillende posities van de groeischijf beoordeeld ten opzichte van de lengte van de eerste metacarpaal bij een triphalangeale duim. De groeischijf was meestal gepositioneerd aan de distale zijde (52%), proximaal in 33% en aan beide zijden in 15%. De ratio's van de lengte van de tweede metacarpaal gedeeld door de lengte van de eerste metacarpaal waren significant kleiner vergeleken met een normale populatie. Dit betekent dat de eerste metacarpaal langer is in de groep met een triphalangeale duim. Indien de groeischijf aan beide zijden van de metacarpaal aanwezig was, groeide deze metacarpaal over de jaren ook harder. Indien de groeischijf zich aan de proximale zijde bevond was de groei vergelijkbaar met de normale eerste metacarpaal.

Hoofdstuk drie beschrijft de lengte en groeischijfpositie van de eerste metacarpaal in relatie met het type triphalangeale duim. In totaal werden 59 handen (in 37 patiënten) met een triphalangeale duim onderzocht op type (delta 31, trapezoïde 9 en volledig type 19), positie van de groeischijf en lengte van de eerste metacarpaal. De metacarpaal van de duim was in alle drie de typen significant langer vergeleken met een normale populatie. De lengte van deze metacarpaal was gerelateerd aan de positie van de groeischijf. Er was geen direct verband tussen het type triphalangeale duim en de lengte.

In **hoofdstuk vier** is de kracht van de intrinsieke spieren van de duim van alle typen triphalangeale duim gemeten. In de praktijk was ons opgevallen dat deze spieren weleens aangedaan zouden kunnen zijn bij alle drie de verschillende typen. Voor deze studie werd de kracht van de duimen en handen bepaald in 38 patiënten met een triphalangeale duim. De kracht was significant verminderd voor alle duim functies, tot 63% voor de kracht bij oppositie ten opzichte van normaal. De kracht van de vuistgreep was 70% van normaal. Alhoewel deze kracht aanzienlijk verminderd is, is dit blijkbaar voldoende om in het dagelijkse leven te functioneren, aangezien geen van de patiënten een hulpvraag hadden op dit gebied. Desondanks zullen interventies waarbij de intrinsieke kracht van de duim verbeterd kan worden bij alle typen triphalangeale duim overwogen moeten worden.

Hoofdstuk vijf laat elf patiënten zien met een triplicatie van de duim, waarbij er ook een triphalangeale component aanwezig is. Slechts één van deze elf duimen kon worden ingedeeld volgens de bestaande classificatie systemen. Bij alle patiënten werden de afwijkende stralen verwijderd en lengte van de duim en richting gecorrigeerd middels osteotomieën. Daarnaast werden de gewrichten gestabiliseerd, pezen geïnsereerd en nagels met nagelwal gecorrigeerd, daar waar nodig. Bij alle patiënten werd de triphalangeale component gecorrigeerd.

Hoofdstuk zes beschrijft een aangepaste classificatie waarin alle verschillende typen triphalangeale duim geassocieerd met radiale polydactylie passen. Alle patiënten van 1993 tot 2006 met een radiale polydactylie (n=104), totaal 121 handen, werden geanalyseerd. De röntgenonderzoeken van de duimen met een radiale polydactylie werden geclassificeerd volgens de bestaande en de nieuw beschreven classificatie. In de nieuwe indeling is de nomenclatuur van Wassel ten aanzien van het niveau van duplicatie behouden. Types VII en VIII zijn toegevoegd voor een partiële of complete duplicatie van de carpale botten zoals beschreven door Buck-Gramcko. Triplicatie en triphalangeale componenten kunnen worden beschreven aan alle typen door middel van achtervoegsels. Symphalangisme, deviatie, hypoplasie kunnen ook op deze manier worden ingedeeld. Triplicaties op verschillende niveaus worden ingedeeld door het toevoegen van de hoogte op basis van de Wassel classificatie. Achttien duimen met triphalangeale componenten of triplicatie konden niet worden geclassificeerd met de bestaande classificaties terwijl met de aangepaste indeling alle duimen geclassificeerd konden worden.

BEHANDELING EN LANG TERMIJN RESULTATEN

Het tweede gedeelte van dit proefschrift is gespitst op klinische uitkomsten en het dagelijks handgebruik van personen met een triphalangeale duim. **Hoofdstuk zeven** vergelijkt twee procedures voor de minder complexe triphalangeale duimen waarbij alleen een extra falanx aanwezig is. Bij de eerste techniek wordt de extra falanx verwijderd en het nieuwe gewricht gestabiliseerd, bij de tweede ingreep wordt het distale interfalangeale gewricht verwijderd door middel van een osteotomie en arthrodese. In total werden er 20 patiënten (33 handen) behandeld. Er werd 17 maal de eerste techniek uitgevoerd en in 16 handen werd de gecombineerde osteotomie met arthrodese uitgevoerd. De keuze voor de arthrodese met osteotomie werd gemaakt indien de extra falanx groter was of de patiënt ouder. In beide groepen waren de gewrichten stabiel. Gemiddeld was de radiale deviatie in het interfalangeale gewricht respectievelijk 5 versus 9 graden. Actieve buiging in het gewricht was 35 versus 46 graden. De resultaten voor beide groepen zijn vergelijkbaar voor zowel objectieve maten als zelf score systemen voor functie en activiteiten. Beide technieken zijn betrouwbaar en tonen een goede functionele uitkomst.

In **hoofdstuk acht** worden de uitkomst van twee verschillende operatie technieken beschreven voor het corrigeren van het type triphalangeale duim waarbij de duim in hetzelfde vlak als de vingers geplaatst is. Zes patiënten (zeven handen) waarbij een pollicisatie verricht is en zeven (negen handen) patiënten na een rotatie abductie en verkortingsosteotomie van de eerste metacarpaal met het verwijderen van de extra falanx via een osteotomie, werden onderzocht. Beide procedures hadden postoperatief een adequate oppositie (Kapandji score 6.1 vs 6.8). De kracht van de oppositie was verminderd in beide groepen vergeleken met een normale populatie (48% en 64%). Subjectieve metingen voor functionaliteit waren respectievelijk 5.7 en 6.7. Voor het uiterlijk waren deze scores 5.5 en 7.6. De uitkomst van beide ingrepen laat een voldoende functie zien voor gebruik in het dagelijks leven. Beide procedures zijn niet uitwisselbaar; een functionerend duimbasis gewricht is noodzakelijk voor de procedure met de osteotomieën van de eerste metacarpaal.

Hoofstuk negen laat de functie van een groep van 12 volwassen mensen zien met een triphalangeale duim (totaal 23 handen), terwijl deze wordt vergeleken met de normale populatie. Objectieve metingen (beweging, gewricht instabiliteit en pijn) toonden geen beperkingen, kracht in antepositie was tot 64% verminderd, terwijl power grip en pinch kracht niet beperkt waren. Oppositie kracht was verminderd tot 62% en kracht bij het buigen in het metacarpofalangeale gewricht eveneens verminderd (61%). Visuele analoge schaal scores voor uiterlijk scoorden zeer laag (2.2 uit een maximale 10), scores voor functie scoorde veel hoger (7.7). De beperking van volwassen patiënten met een triphalangeale duim in het dagelijkse gebruik ligt dan waarschijnlijk ook meer bij het uiterlijk dan bij de daadwerkelijke functie.

De triphalangeale duim is een complexe entiteit met een grote differentiatie in presentatie. Bij de preoperatieve analyse is de positie van de groeischijf van belang voor de groei van de eerste metacarpaal, ook bij de meer simpele types. De intrinsieke musculatuur kan eveneens bij alle types aangedaan zijn , met een krachtsvermindering tot 63%. De uitkomsten van de beschreven operatie technieken zijn vergelijkbaar, de technieken zijn echter niet uit wisselbaar voor individuele patiënten, aangezien alle technieken bepaalde voorwaarden van de bestaande duim eisen. In dit proefschrift worden enkele punten van de triphalangeale duim verhelderd, de praktijk blijkt vaak iets weerbarstiger.

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  ER Staff
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- Basic hand surgery / LISA course 2013 - 2015
  Residents program General surgery
- Around the Wrist 2011
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- New Advances in Peripheral Nerve Surgery 2009
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Three ladies is a crowd?? No way!

Curriculum vitae

Jelle Michiel Zuidam was born on the first day of the year 1976 in Sittard, the Netherlands. After graduating from Stella Maris College in Meerssen (1994), he started his medical career in Amsterdam at the Vrije Universiteit. After some years doing research, he started his plastic surgery training with two years of general surgery (Dr Wibo Weidema, Ikazia Ziekenhuis Rotterdam) continued with four years of plastic surgery in the Erasmus Medical Centre (Prof. Dr S.E.R. Hovius). In 2011 he visited the Martin Singer Hand Unit in Het Groote Schuur hospital in Cape Town, South Africa (Dr. Michael Solomons). In 2012 he completed the Dutch national hand fellowship (Erasmus Medical Centre, Rode Kruis Ziekenhuis Beverwijk, HAGA Ziekenhuis Den Haag, AMC Amsterdam and Xpert Clinic). He passed the european board of hand surgery handsurgery diploma examination (FESSH) in Antwerp, Belgium in 2012. In the same year he passed the EBOPRAS examination. In 2013 he started working as a plastic/hand surgeon in the Erasmus Medical Centre and Franciscus Gasthuis in Rotterdam. He is living with three lovely ladies (Daniëlla, Elin and Kiki)