RADIAL POLYDACTYLY: DOUBLE OR NOTHING?

Robert Dijkman

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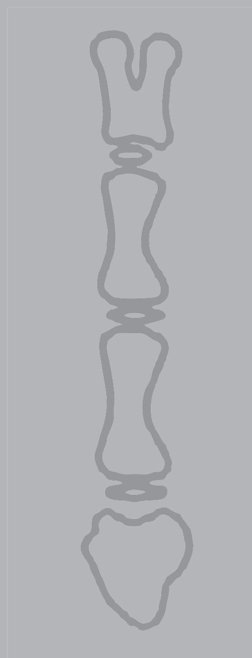

CHAPTER 1

General introduction

Derived from: Clinical presentation, surgical treatment, and outcome in radial polydactyly

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INCIDENCE

Thumb duplication or 'radial polydactyly' is one of the most common congenital upper limb anomalies ('CULAs') affecting the thumb. The incidence of radial polydactyly is estimated to be 2.3 per 10.000 live births.[1] Considerable regional and racial incidence variability has been suggested, although evidence is mainly limited to hospital-population-based studies.[2-4] Reportedly, the frequency of radial polydactyly relative to other congenital hand anomalies is highest in Asians.[5,6] In the Netherlands, the annual incidence of radial polydactyly approximates 60 – 80 patients, based on the incidence and the increase in adoption of 'special need' children from Asian countries. Given the importance of the thumb to everyday hand function,[7,8] most patients will undergo surgery to restore the functional anatomy, which is affected following abnormal embryological development of osseous structures and soft tissues.

GENETICS AND EMBRYOLOGY

Most radial polydactyly cases are unilateral and sporadic, although autosomal dominant inheritance may be observed in combination with triphalangeal thumbs [9] and Greig cephalopolysyndactyly.[10] While radial polydactyly typically occurs as an isolated anomaly,[11] the Online Mendelian Inheritance in Man ('OMIM') database shows it may present as part of over 125 distinct syndromes and phenotypical associations, including Holt-Oram syndrome, Townes-Brocks syndrome, Fanconi's anemia, etc.[12]

CULAs arise during upper limb development, which takes place between the fourth and eight week of gestation. During these four weeks, a fully functional hand is formed along three axes of development: the proximal-distal axis, the dorsal-ventral axis, and the anterior-posterior (or 'radial-ulnar') axis. Growth and differentiation of tissue along the axes is orchestrated through genetic and molecular signaling pathways, which arise from areas of specialized cells in the limb bud called 'signaling centres'.[13-15]

In the development of polydactyly, the radial-ulnar axis is the most important. This axis is formed along the Zone of Polarizing Activity ('ZPA'), where Sonic Hedgehog ('SHH') proteins regulate ulnarization and widening of the limb (Figure 1).[16] Disruption of SSH signaling pathways may result in radial polydactyly. Moreover, mutations of SHH and GLI3 are associated with various phenotypes of radial polydactyly [17-19] and triphalangeal thumb.[20-24] The complex genetic and molecular interactions result in a highly variable clinical presentation of radial polydactyly, ranging from a rudimental skin tag to very complex triplications of the thumb.

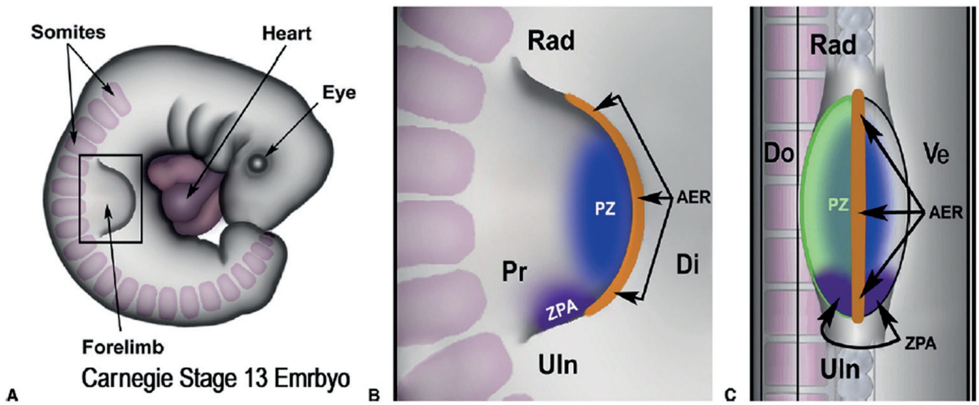

Figure 1: Limb development along three axes of growth and differentiation.

A. Carnegie Stage 13 Embryo displaying formation of the upper limb bud; B. Dorsal view of the developing limb bud; C. Lateral, end-on view of the developing limb; Rad = 'Radial'; Uln = 'Ulnar'; Pr = 'Proximal'; Di = 'Distal'; AER = 'Apical Ectodermal Ridge'; PZ = 'Progress Zone'; ZPA = 'Zone of Polarizing Activity'; Do = 'Dorsal'; Ve = 'Ventral'.

CLINICAL PRESENTATION

The clinical presentation of radial polydactyly is heterogeneous, ranging from a slightly broadened distal aspect of the thumb to complex synpolydactyly with three recognizable thumbs (Figure 2).

All osseous structures of the thumb may be affected, from the distal phalanx to the trapezium (or even to the scaphoid in very rare cases). Depending on the level of duplication, the interphalangeal joint ('IPJ'), metacarpophalangeal joint ('MCPJ'), and carpometacarpal joint ('CMCJ') of both thumbs may be normal, stiff, or hypermobile. Duplications at the MCPJ may move 'en bloc' due to cartilaginous or bony connection at the base of the basal phalanx. Abnormal joint surface, volar plate and collateral ligaments may result in hypermobility or deviation, especially at the IPJ of radial polydactyly cases where the tips of both thumbs converge distally.

Soft tissue anomalies such as eccentric insertions of the extensor pollicis longus ('EPL') and flexor pollicis brevis ('FPL') tendons may also contribute to distal convergence of both thumbs, and affect their active range of motion ('AROM'). When AROM is not observed clinically, flexion and extension creases at the palmar and dorsal aspects IPJ and MCPJ may be indicative of its presence (Figure 3). Thenar musculature may vary from normal to hypoplastic, especially in more proximal duplications (i.e., at the MCPJ and CMCJ). The same applies to the first web space, which may be narrow in more proximal duplications, but is usually adequate in the more distal duplications.

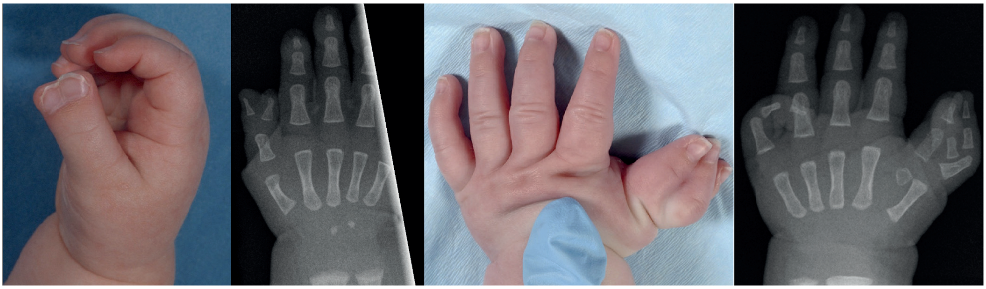

Figure 2: Case examples of the radial polydactyly spectrum. Note the apparently ‘simple’ broadening of the distal aspect of the thumb on the left, and the complex synpolydactyly with three thumbs on the right.

Figure 3: Effect of preoperative presence of an IPJ flexion crease on postoperative IPJ flexion. Note the better postoperative flexion of the IPJ in the thumb where a flexion crease was present preoperatively (photographs A and B).

A. Preoperative situation with an IPJ flexion crease on the ulnar thumb
B. Postoperative situation of the thumb shown in A, note the reasonable IPJ flexion
C. Preoperative situation without an IPJ flexion crease on the ulnar thumb
D. Postoperative situation of the thumb shown in C, note the absence of IPJ flexion.

Although radial polydactyly is often referred to as ‘thumb duplication’, it is characterized by hypoplasia of osseous structures and soft tissues. Instead of duplication or ‘doubling’ of tissue, both thumbs are invariably more hypoplastic than an unaffected (e.g., contralateral) thumb. This is most apparent in the nails and pulp of the affected thumbs, which are usually smaller compared to the unaffected contralateral side. While the degree of hypoplasia can be symmetrical, affecting both thumbs to a similar extent, the ulnar thumb is usually better developed than the radial thumb.[25]

Classification

In order to structure the highly variable clinical presentation, radial polydactyly cases are categorized using classification systems. Since soft tissue anomalies are harder to visualize, most classification systems are based on osseous configuration, which can be shown using x-rays. These classification systems play an important role in communication between specialists, in the evaluation of treatment outcomes, and in supporting clinical decision-making.

The spectrum of CULAs can be classified using the Oberg-Manske-Tonkin (‘OMT’) classification. In the OMT classification, radial polydactyly is classified as a malformation, under failure of axis formation/differentiation of the hand plate in the radial-ulnar axis (‘1B1’).[26] The OMT classification is based on our current understanding of molecular, genetic, and embryologic development of the upper limb, and has recently replaced the morphology-based Swanson classification [27] as the main classification for CULAs. In the Swanson classification (which was later adopted by the International Federation of Societies for Surgery of the Hand as the ‘IFSSH-classification’[28]), radial polydactyly was part of the ‘Duplications’ group.

The most widespread classification system for radial polydactyly is the Wassel classification (Figure 4).[2] It describes seven types of osseous configurations. Types I-VI represent distal-to-proximal levels of thumb duplication, while type VII represents radial polydactyly with a triphalangeal component. Although exact occurrence varies across different case series, the three most common types of radial polydactyly are type IV (30% - 46%), type II (9% - 25%) and type VII (7% - 32%).[29-33]

While easy to apply, the clinical relevance of the Wassel classification is limited by the inability to classify surgically important features of radial polydactyly (e.g., diverging components or hypoplasia). This has led to the introduction of many alternative classification systems, such as the Rotterdam classification,[34] which integrates elements of the Wassel, Buck-Gramcko,[35] and Upton[36] taxonomies, into an all-embracing classification system for radial polydactyly including triphalangeal components and triplications.

Since the Rotterdam classification is more complex than the widely applied Wassel classification, it is questionable whether both classification systems can be applied with equal reliability and consistency in a clinical setting. Furthermore, the clinical relevance of the expanded taxonomy of the Rotterdam classification is yet to be established.

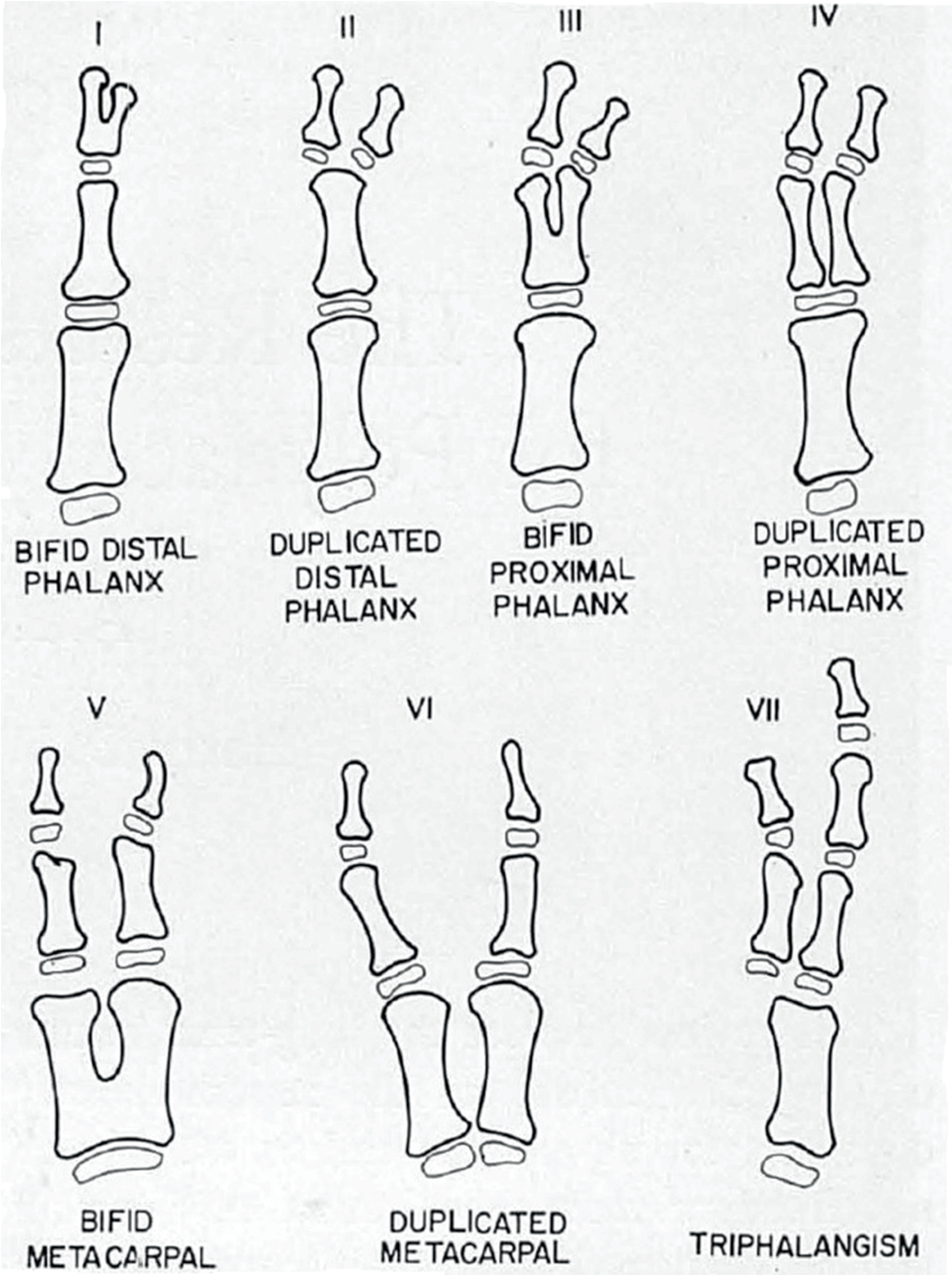

Figure 4: The Wassel classification for radial polydactyly.

SURGICAL TREATMENT

The aim of surgical treatment of radial polydactyly is to obtain a stable, mobile thumb of adequate size and appropriate shape.[25] An early report stated *"an extra finger or thumb is removed for cosmetic reasons. This operation requires no ingenuity and creates no problems"*.[37] This statement was soon refuted by the first follow-up studies, which demonstrated revision rates of 38% – 56% to correct joint instability and axial deviation.[38,39] As a result, the surgical approach to radial polydactyly has evolved considerably over the years.

In general, surgeons may choose between five different techniques to treat radial polydactyly, depending on the clinical presentation: resection and reconstruction, the Bilhaut procedure, simple ablation, on-top plasty, and pollicization. The latter three techniques are only applicable in rare and selected cases, and therefore beyond the scope of this thesis. The resection and reconstruction technique and the Bilhaut procedure are outlined in more detail below.

Resection and reconstruction

The most versatile and widely applied surgical treatment for radial polydactyly is the resection and reconstruction technique. The majority of cases can be treated using this technique, which is indicated whenever one of the extra thumbs is better developed than the other (with exception of the floating-type radial polydactyly). In most cases, the ulnar thumb is better developed and the radial thumb is resected. Resection of the radial thumb has the added benefit of preserving the ulnar collateral ligament, which plays a key role in stabilizing the MCPJ during pinch grip and prehension.[40] A typical case example where most aspects of the resection and reconstruction approach are utilized is the frequently occurring type IV, with a better-developed ulnar thumb (Figure 5).

The procedure starts with a dorso-lateral incision. Care is taken to conserve a sufficient amount of skin for later reconstruction of the pulp, and for creating flaps to interrupt linear scars. After attaining adequate exposure, the configuration of the EPL and FPL tendons can be evaluated. The extensors are often bifurcated, with a better-developed tendon towards the ulnar thumb in the majority of cases. The tendon insertion on the distal phalanx of the ulnar thumb can be situated too radially. Furthermore, the extensor hood can be confluent to palmar structures distal to the bifurcation. These anomalies are treated by: removal of the hypoplastic tendon at the bifurcation, reinsertion or transposition of the eccentric extensor tendon at the distal phalanx, augmentation of the extensor tendon using the extensor tendon of the removed thumb, and release of tendinous interconnections. The flexor tendons share similar characteristics and treatment with the extensor tendons. However, the pulley system may also be hypoplastic, absent, or out of line, warranting reconstruction to restore an anatomical pull of the FPL.

To allow resection of the radial thumb, both the thenar muscle insertion and the radial collateral ligament should be detached from the radial side of the proximal phalanx of the radial thumb. The muscle attachment and collateral ligament can be taken as a compound or as two separate structures. The radial collateral ligament is taken with a periosteal sleeve distally for later reinsertion as the radial thumb is resected.

After resection of the radial thumb, a broadened head of the MCPJ, which accommodated the extra thumb, is encountered. This is treated with a longitudinal osteotomy, without compromising the proximal radial collateral ligament attachment. Any residual ulnar deviation in the MCPJ can be addressed to using a closing wedge osteotomy. After bony alignment, the periosteal-collateral ligament sleeve can be reattached to the radial side of the proximal phalanx of the ulnar thumb, followed by thenar muscle reinsertion.

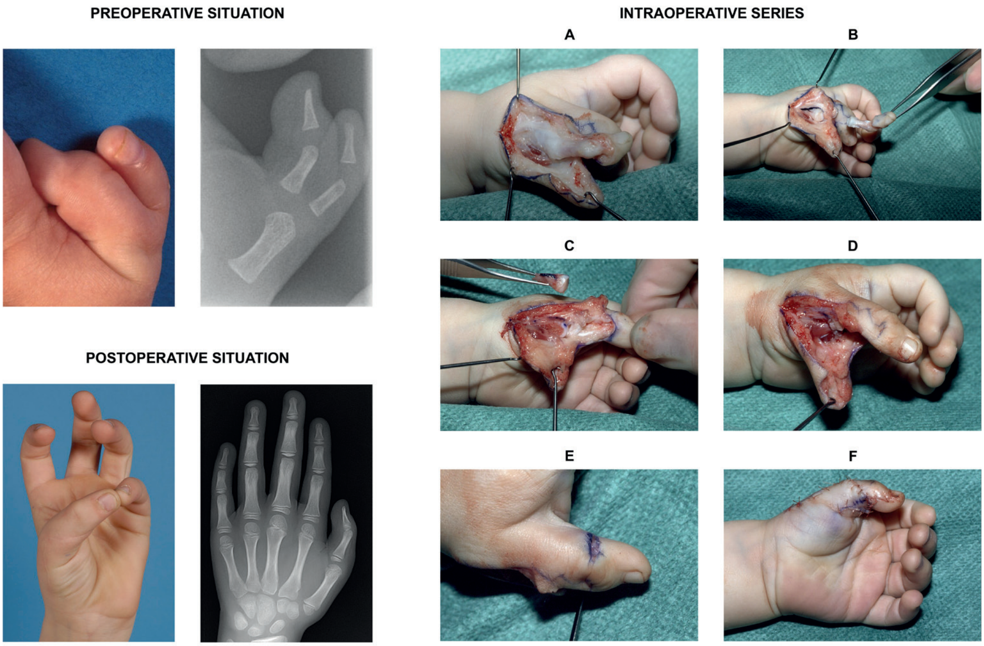

Figure 5: The resection and reconstruction procedure for a radial polydactyly type IV case.

Top left: preoperative situation of a radial polydactyly type IV case; Bottom left: postoperative situation after 8 years of follow-up; Right: intraoperative series; A. Incision; B. Resection of the radial thumb with preservation of the radial collateral ligament and intertwined thenar musculature. Note the markings for a longitudinal osteotomy of the broad first metacarpal head; C. Longitudinal osteotomy of the broad first metacarpal head, with preservation of the proximal attachment of the radial collateral ligament; D. Reinsertion of the radial collateral ligament at the base of the basal phalanx; E. In this specific case, residual IPJ deviation was corrected by reefing the lax ulnar collateral ligament; F. Y-to-V closure of the skin using a flap from the residual soft tissue.

Regarding the IPJ, deviation may be caused by joint anatomy, laxity of the (mostly) ulnar collateral ligament or eccentric insertion of the tendons. A closing wedge osteotomy, reefing of the ulnar collateral ligament, and centralizing the insertion of the involved tendons, respectively, can be used to restore the functional anatomy of the IPJ. A K-wire with a maximum diameter of 0.8 mm can be used to 'splint' the thumb internally.

The skin is meticulously sutured in a 'cut-as-you-go' fashion, creating a Y-to-V flap from the residual skin and pulp to provide enough bulk to ensure optimal pulp thickness, if appropriate. To prevent nail fold deformities due to linear scars, especially at the conjunction of the proximal and lateral nail fold, V flaps are inserted.[11,25,41]

The Bilhaut procedure

While radial polydactyly typically presents with one better-developed and one less-developed thumb, there are cases where both thumbs are of approximately the same size and shape. If the degree of hypoplasia is equally severe for both thumbs, and neither thumb seems adequate for reconstruction on its own, a Bilhaut procedure may be indicated. The Bilhaut procedure was originally described for radial polydactyly type I,[42] but can also be applied for types II-IV, and VII.[43,44] Moreover, some find it particularly well suited for the 'diamond-shaped' type IV radial polydactyly cases (Figure 6).[45]

Preoperative markings are longitudinal across (or along) the nails and zig-zag proximal to the nail fold. The aim is to combine bone, soft tissue, and nail of both thumbs, to retain a similar amount of tissue compared to the contralateral unaffected thumb. The procedure starts by elevation of the nails, followed by the distal-to-proximal excision of medial soft tissue and bone. Care is taken to preserve enough soft tissue to allow for tension-free coverage of the osseous structures underneath, and to preserve the insertions of the flexor and extensor tendons. The osseous structures are then meticulously aligned, paying close attention to the physes and especially the articular surfaces of both thumbs. Fixation is performed using interosseous cerclage wires or (PDS) sutures.

The distal phalanx should have a smooth dorsal surface, since irregularities in the closely adherent nail bed may cause a longitudinal nail ridge. The nail bed itself is meticulously repaired under magnification. If a Y-shaped EPL tendon was present, it is coapted before closing the skin. Finally, the nail or a piece of plastic is replaced between the nail fold and nail bed, to prevent adhesions during the healing process.

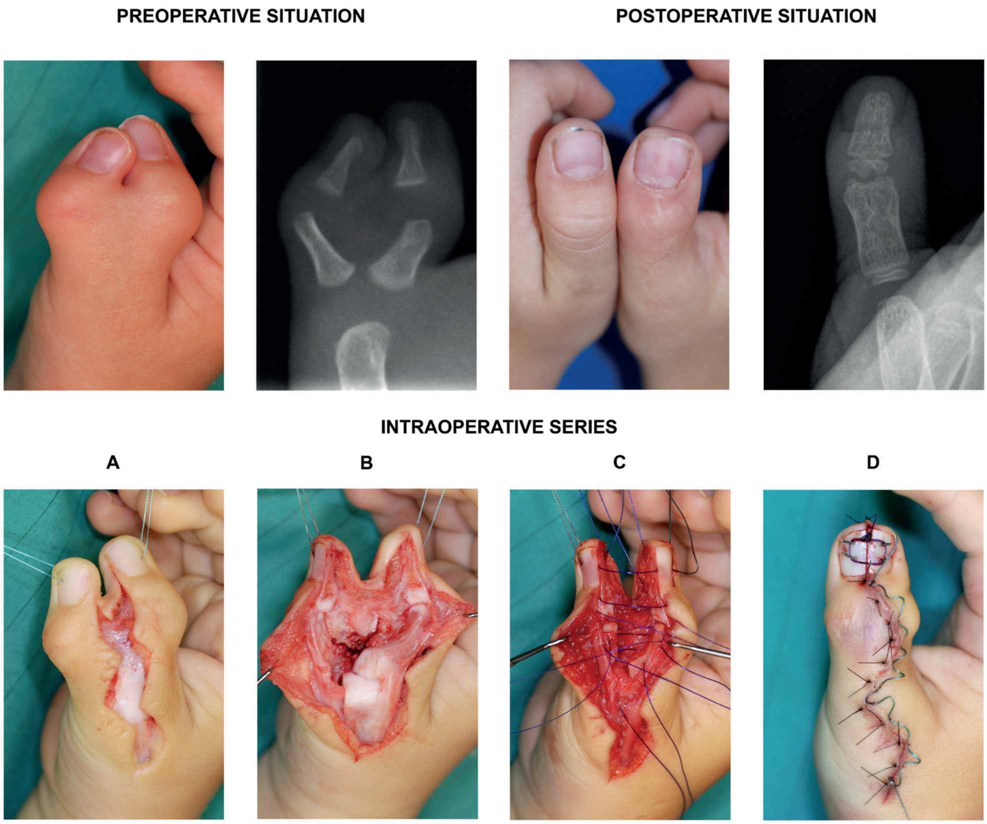

Figure 6: The Bilhaut procedure for a radial polydactyly type IV case.

Top: preoperative situation and postoperative situation after 4 years of follow-up; Bottom: intraoperative series; A. Incision; B. Resection of the central parts of the nail, soft tissue, and bone of each thumb; C. Coaptation of the lateral parts using transosseous PDS-sutures; D. Aspect at the end of operation.

Consensus and controversy of surgical treatment

Although evidence is limited to case-series and literature describing surgical technique, consensus exists on a number of issues. First, simple ablation of one thumb without adequate reconstruction of the remaining thumb is rarely indicated and can lead to unacceptably high revision rates.[38,39,44,46] Second, it is advisable to correct as much as possible during the first procedure, since secondary procedures can only improve 'poor' initial results to 'average' final results. [39,46,47] Third, approximately 20% of radial polydactyly cases will need secondary procedures,[30,48] with joint deviation and instability being the most common reasons for revision surgery.[30,46,47]

Despite this consensus, the effects of many other preoperative, perioperative, and surgery-related factors on postoperative outcome remain controversial.

Considering the preoperative situation, each radial polydactyly type presents with its own specific problems. However, it is unclear how AROM, stability, alignment, and strength are influenced by radial polydactyly type.[33,48,49] Moreover, it is unknown whether presence of aberrant deviating, hypoplastic, and triphalangeal components influences outcome.[48-50]

In addition to radial polydactyly (sub)type, little is known concerning other preoperative determinants of postoperative AROM. For example, preoperative absence of an IPJ flexion crease may be indicative of poor pre-existing range of motion, which is hard to improve surgically.[51] Moreover, abnormal configurations of flexor and extensor tendons may affect postoperative AROM.[25]

Controversy also exists regarding factors that could influence postoperative stability and alignment. For example, preoperative incongruent or hypoplastic joint surfaces and collateral ligament insufficiency are hypothesized to influence postoperative stability and alignment. However, it is unknown whether tendon rebalancing to correct line of pull, transverse osteotomies to realign the phalanges, and reconstruction of the collateral ligaments of the involved joints [25,48,49] are invariably beneficial, especially since extensive surgical correction of the tendon system and joint surfaces may lead to adhesions, which may impair the AROM of the IPJ in particular.[25]

Considering the perioperative situation, the age at which patients are first operated has been subject to debate. While some favor the technically challenging early surgery,[47,52-55] others advocate delay of the procedure until surgery is facilitated by growth of the anatomic structures.[2,33,41,56,57] Furthermore, although the relationship between increased volume and favorable outcomes has been well established in other surgical fields,[58,59] the extent to which experience of the surgeon determines outcome of surgery for radial polydactyly is unknown.

Considering surgical technique, both resection and reconstruction and the Bilhaut procedure are assumed to have specific advantages and drawbacks.[25] The resection and reconstruction technique presumably leads to smaller thumbs, which may be less stable, while preserving IPJ and MCPJ AROM, and producing less conspicuous scars. On the other hand, the Bilhaut procedure presumably leads to broader thumbs, which may be stronger and more stable, although IPJ and MCPJ AROM may be diminished, and esthetic results may fall short due to conspicuous scars of the thumb dorsum and thumb pulp, causing the characteristic nail ridge and longitudinal pulp crease.[39,44,60]

While modifications to the Bilhaut procedure help getting around some of its esthetic drawbacks,[29,45,61] the association between thumb strength and thumb stability in radial polydactyly patients remains uncertain.[30] However, although the functional and esthetic advantages of both procedures are subject to debate, there have been no comparative studies between resection and reconstruction and the Bilhaut procedure, despite the overlapping indications for both techniques.

OUTCOME ASSESSMENT

In 2002, the World Health Organization ('WHO') introduced a framework for evaluating the impact of disability on a person's overall functioning, called 'The International Classification for Functioning, Disability, and Health' ('ICF', Figure 7).[62] The ICF framework conceptualizes a person's level of functioning as the interplay between 'body function and structures', 'activity', 'participation', and 'contextual factors' (i.e. personal and environmental factors influencing health related quality of life ('HRQoL')).

Outcome assessment in radial polydactyly is presently focused on the 'body function and structures' domain of the ICF. Most long-term follow-up studies report AROM, stability, and alignment of the reconstructed thumb, [30,48,49] while some also report thumb strength, thumb appearance, and patient satisfaction. Items are often combined into a composite outcome assessment system to quantify an 'overall outcome'. The most widely applied composite outcome assessment system is the Tada system,[47] which combines AROM, stability, and alignment into a score (ranging 0 – 5 points) that represents poor-to-good overall outcome.

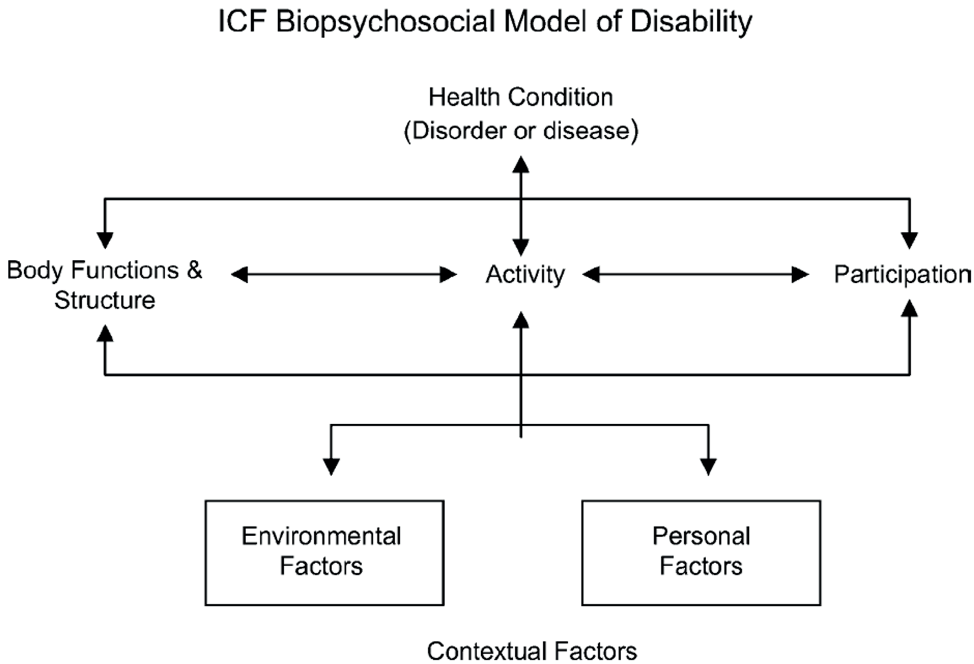

Figure 7: The ICF framework conceptualizes a person's level of functioning as the interplay between 'body function and structures', 'activity', 'participation', and 'contextual factors'.

A variety of other composite outcome assessment systems exist for radial polydactyly,[33,48,49,63,64] all of which weigh importance of the incorporated functional, esthetic, and patient-reported items differently. As a consequence, the conclusion on overall outcome may vary considerably depending on the applied assessment system and on the person evaluating the patient. At the same time, there is no evidence on the interobserver reliability of these systems. Moreover, the validity of presently available assessment systems is questionable, since there is no scientific support for the number of points per item, or, for the cut-off values per number of points in each item in these systems.

Concerning the other domains of the ICF, many different instruments exist to assess activity (e.g. manual ability),[65-68] participation,[69-71] and HRQoL,[72,73] but none of them are specifically designed for radial polydactyly patients. Consequently, little is known regarding the impact of radial polydactyly on outcome within the activity, participation, and HRQoL domains of the ICF framework.

AIM AND OUTLINE OF THIS THESIS

The aim of this thesis is to optimize treatment strategies for radial polydactyly by studying outcome within the ICF framework. This aim is pursued throughout three parts of this thesis, using data gathered in an international, multicenter, long-term follow-up project.

The first part of the thesis is focused on defining the main determinants of treatment outcome, which can be divided into studying preoperative factors and postoperative outcome assessment tools. In **Chapter 2**, the reliability, strengths, and shortcomings of the two most commonly used preoperative classification systems for radial polydactyly are investigated to determine which classification is best suited to meet current clinical and scientific demands. In **Chapter 3**, the reliability and validity of ten outcome assessment systems for radial polydactyly are compared in order to determine the state of the art in outcome assessment. **Chapter 4** describes how the best available outcome assessment system could be further optimized, by using long-term follow-up data from an international population to develop a valid, generalizable, and clinically-weighted outcome assessment system.

In the second part, the clinically-weighted assessment system developed in part one of this thesis was applied to study results of surgery in patients with radial polydactyly at the interphalangeal and metacarpophalangeal level. In **Chapter 5**, outcomes of the resection and reconstruction technique for radial polydactyly are described, and several important determinants of outcome are studied. Outcomes following the resection and reconstruction technique are compared to outcomes following the Bilhaut procedure in a matched comparative study in **Chapter 6**.

The third part of this thesis is focused on the relationship between outcomes in the body function and structures domain, and the other domains of the ICF-model. In **Chapter 7**, the impact of radial polydactyly on postoperative thumb strength is quantified, and the impact of strength on manual ability is studied. **Chapter 8** outlines the patient perspective on manual ability, participation, and HRQoL of radial polydactyly patients. These results are aimed at improving counseling in an outpatient clinic setting.

The general discussion in **Chapter 9** provides a comprehensive evaluation of the main findings of this thesis, as well as a number of directions for future research. Finally, the main findings of this thesis are summarized in English in **Chapter 10**, and in Dutch in **Chapter 11**.

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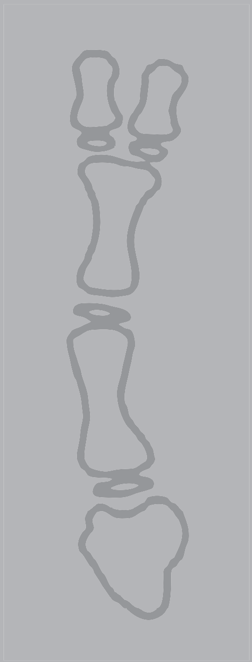
PART ONE:
CLASSIFICATION AND OUTCOME ASSESSMENT



CHAPTER 2

A multicenter comparative study of two classification systems for radial polydactyly

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ABSTRACT

BACKGROUND The aim of this study is to compare type occurrence and reliability of the Wassel and Rotterdam classifications for radial polydactyly.

METHODS We classified a large population of radial polydactyly patients from two European clinics using both classification systems, and compared the incidences of the different types to a population derived from a systematic literature review. We further assessed intra- and interobserver reliability of both classification systems in a test-retest design with seven observers, using Kappa statistics.

RESULTS Forty percent of the 520 cases with available x-rays could not be classified using the Wassel classification, while all cases could be classified using the Rotterdam classification. All unclassifiable cases had aberrant components; the majority were of the triphalangeal (63%), deviating (43%) or hypoplastic (39%) kind. Types III, IV and VI occurred more often using the Rotterdam classification. Intra- and interobserver reliability was comparable for both classification systems (K = 0.87 vs. K = 0.83 and K = 0.65 vs. K = 0.70). Types II and IV had the lowest reliability in both the Wassel and Rotterdam classification (K ranging 0.30 – 0.59). Aberrant components indicating deviation and hypoplasia had the lowest reliability in the Rotterdam classification (K ranging 0.23 – 0.45).

CONCLUSIONS The Rotterdam classification has broader classification possibilities and similar intra- and interobserver reliability compared to the Wassel classification. Although it is more complex and the aberrant components should be stricter defined to increase its clinical relevance, we recommend using the Rotterdam classification.

INTRODUCTION

In 1969, H.D. Wassel introduced a radiological classification system for radial polydactyly in which seven types of osseous configurations were described (Figure 1).[1] Since its introduction, the Wassel classification has played an important role in planning surgical intervention, evaluation of clinical outcome, and communication between congenital hand surgeons.[2-5]

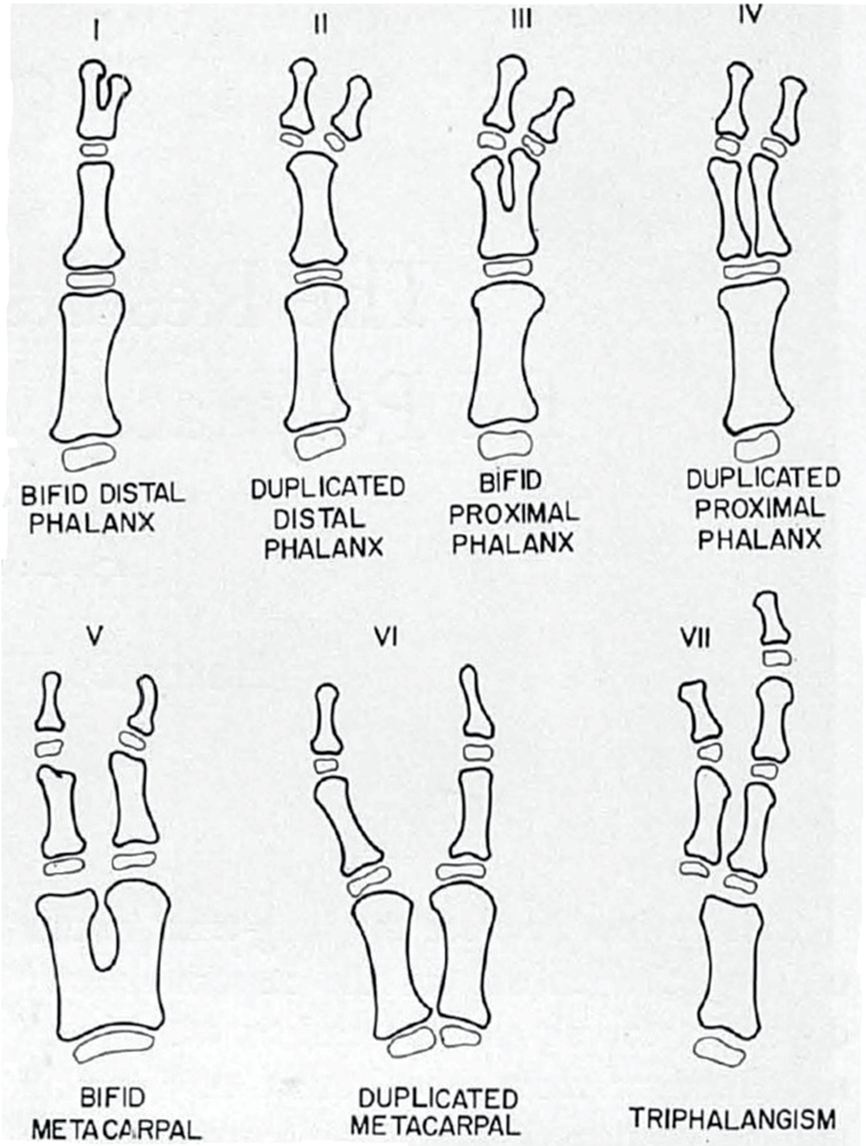

Figure 1: The Wassel classification.

Dr. Adrian E. Flatt first proposed a modified Wassel classification in 1977, excluding triphalangeal thumb.[6] Subsequently, a number of expanded classification systems have been proposed to accommodate the wide range of clinically relevant subtypes.[7-10] In 2008, Zuidam et al. introduced a nomenclature that integrated essential elements of these classifications into a single all-embracing taxonomy: the Rotterdam classification (Figure 2).[11] Though specifically designed to accommodate all types of triphalangeal thumb and thumb triplication, the Rotterdam classification can also give a clear description of joint involvement and aberrant components in all radial polydactylies.

Nevertheless, the Wassel classification remains the most widely used classification system in recent literature.[12,13] This might be because the Rotterdam classification appears too complex and the merit of its expanded classification possibilities is yet to be established. Also, it is unknown how consistently both classification systems can be applied based on the preoperative x-rays.

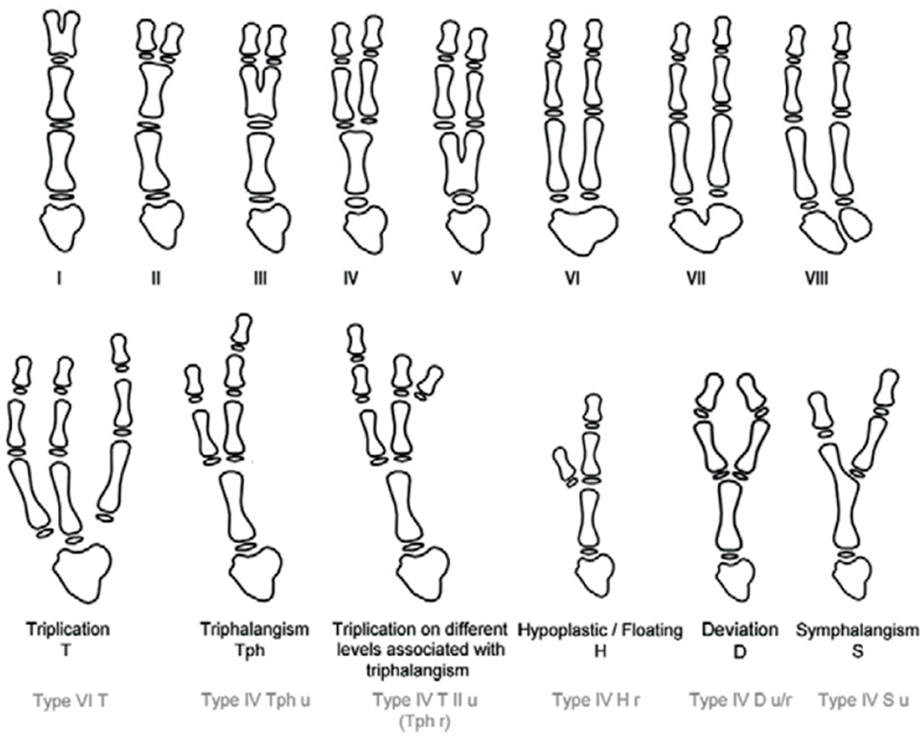

Figure 2: The Rotterdam classification.

Therefore, the aim of this study was twofold. First, we assessed the impact of the Rotterdam classification on the occurrence of the various types of radial polydactyly. To do this, we classified a large series of x-rays of patients with radial polydactyly using both the Wassel and the Rotterdam classification and compared our study population with a control population derived from medical literature. Second, we assessed the intra- and interobserver reliability of both the Wassel and the Rotterdam classification in a test-retest design.

MATERIALS AND METHODS

Patients and setting

This study took place in two large European congenital hand surgery departments: the Erasmus MC in Rotterdam, the Netherlands and the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. In the first part of this study, we investigated occurrence of the various subtypes of radial polydactyly. In the second part, we studied reliability of the Wassel and Rotterdam classification system with a test-retest experiment.

Type occurrence in the Wassel and Rotterdam classifications

In our study of type occurrence in radial polydactyly, a single investigator (RD) consecutively examined all available x-rays from both clinics, from the period 1980-2012. These x-rays were usually taken at an age of three to six months old. Both the Wassel and Rotterdam classifications were applied. The Wassel classification was applied using the exact definitions of seven types of radial polydactyly as described in the original article by H.D. Wassel in 1969.[1] If the type of radial polydactyly did not fit any of the abovementioned descriptions, the case was deemed "unclassifiable".

The Rotterdam classification consists of eight types of radial polydactyly and adds letters to describe aberrant components, followed by their location. It can be applied using the following four steps:

1. Annotate the most proximal level of duplication using numbers I-VIII from distal to proximal. Even numbers describe complete duplication, uneven numbers incomplete duplication. Types VII and VIII represent duplication of the carpal bones.
2. Count the number of thumbs. If there are two thumbs, proceed to step three. If there are three thumbs, add the capital "T" for "Triplication" followed by the level at which the more distal duplication occurs and its location (see step four).
3. Assess the presence of other aberrant components. If present, add the appropriate capital letter for each aberrant component: "Tph" for triphalangism, "D" for deviation, "H" for hypoplasia and "S" for symphalangism.
4. Indicate which of the digits show aberrant components. Use the letters "r" for radial, "m" for middle, "u" for ulnar digit or a combination of these letters if appropriate (for example:

"type IV D r/u" if both radial and ulnar digits deviate from the central axis in a complete duplication at the metacarpophalangeal level, Figure 3).

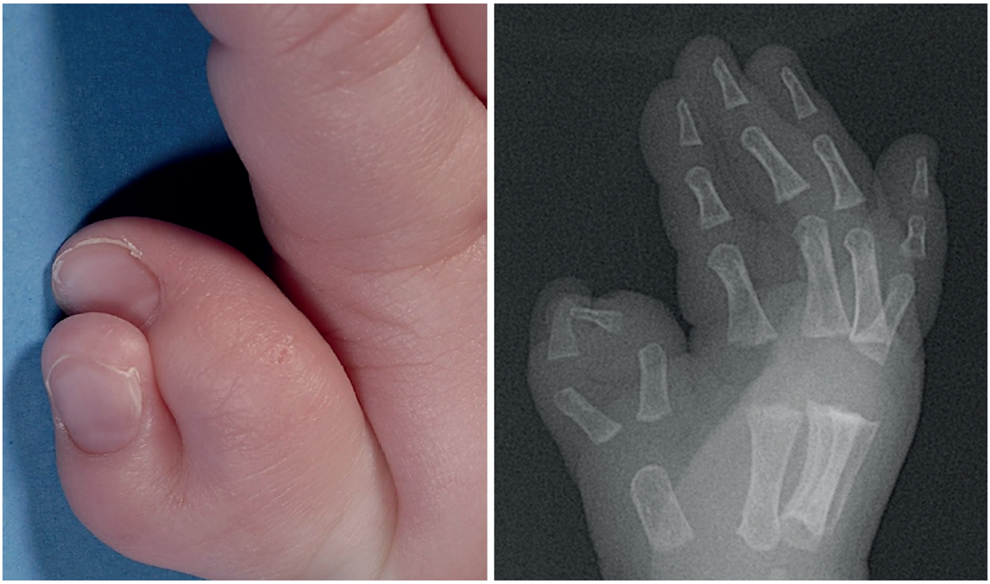

Figure 3: Example of a 'diamond shaped' or type IV D r/u radial polydactyly in the Rotterdam classification. There is a complete duplication at the MCP level, with a deviating aspect of both the radial and ulnar thumb.

In order to find a comparative historical cohort, a systematic search of medical literature in Embase and Medline was performed using the terms in Appendix 1. All available articles that applied the Wassel classification to describe their population were initially included. Case reports and articles that focused on a specific type of radial polydactyly were subsequently excluded.

Reliability of the Wassel and Rotterdam classifications

In the second part of this study we assessed the reliability of both classification systems. The principal observer (RD), who was not a hand surgeon and not involved in development of the Rotterdam classification, randomly selected 52 cases with available preoperative x-rays and photographs from the study population of 520 cases. These posterior-anterior x-rays and photographs were presented to seven observers: four congenital hand surgeons, a resident, a PhD candidate and one medical student. Two congenital hand surgeons were from Hamburg and two were from Rotterdam. The other observers were from Rotterdam. The observers were asked to classify the cases using both the Wassel and Rotterdam classification in three rounds, at two-week intervals. The first 'training' round [14] consisted of 12 cases, the next two 'classification' rounds of 40 cases. The same 40 cases were presented to the observers in a different order during the two classification rounds. Each observer had a protocol describing both classifications present during the experiments. Two observers from Rotterdam had been involved in development of the Rotterdam classification.

Statistical Analysis

We analyzed the occurrence of each type of radial polydactyly for both classifications in the study population and the literature cohort. The unclassifiable cases of the study population were further described using the Rotterdam classification. The presence of aberrant components (Rotterdam classification) was assessed in both classifiable and unclassifiable cases.

The reliability of both classification systems was assessed using Kappa statistics (K).[15] An average Kappa coefficient was calculated to represent the overall intra- and interobserver agreement of both classification systems. Intraobserver agreement was calculated using the Kappa coefficient between each observer's first and second classification readings. Interobserver agreement was calculated using the Kappa coefficient between the classification readings of the various observers. In the Rotterdam classification the reliability was assessed separately for each of the four steps. In addition, we assessed the intra- and interobserver reliability per subtype of radial polydactyly, using the classification assigned by RD as reference.

The relationship between reliability and years of experience in the field of congenital hand surgery of the observers was assessed using Spearman's correlation coefficient (r_s). We used IBM SPSS Statistics 21 for all analyses.

This study was done under approval of an accredited Medical Research Ethics Committee (MEC-2010-295) and in accordance with the declaration of Helsinki.

RESULTS

Type occurrence in the Wassel and Rotterdam classifications

A total of 633 cases of radial polydactyly were identified from the patient records, of which 520 (82%) had available x-rays. The Rotterdam population had significantly more female and bilaterally affected cases, whereas the right side was affected more often in the Hamburg population (Table 1).

Table 1: Population characteristics of the type occurrence study sample.

	Rotterdam	Hamburg	Total	p-value
Cases	301	332	633	
- Female*	157	137	294	0.04
- Male	144	195	339	0.06
Left hand affected	73	80	153	0.97
Right hand affected*	96	172	268	< 0.001
Bilateral*	132	80	212	< 0.001
Preoperative X-ray available	228	292	520	0.09

* Significant differences between Rotterdam and Hamburg study sample (χ^2).

The literature search yielded 854 articles, of which 163 met the inclusion criteria. One hundred and thirty-nine articles were further excluded from the analysis because they were cases reports or focused on a specific radial polydactyly subtype. From the remaining 25 articles,[1,3,4,11,16-36] we could pool 1723 cases of radial polydactyly to describe the frequencies reported in medical literature.

Figure 4 shows the occurrence of Wassel and Rotterdam types in our research population of 520 cases, together with the frequencies found in medical literature. Two hundred and eight (40%) out of our 520 cases and 96 (6%) out of 1723 cases from medical literature could not be classified using the Wassel classification. All cases could be classified using the Rotterdam classification. Due to the large proportion of unclassifiable cases, types III, IV and VI occurred more frequently when the Rotterdam classification was applied compared to when the Wassel classification was applied.

Aberrant components were present in 245 (78%) of the cases classifiable using the Wassel classification, with deviating components occurring most frequently (72%, Figure 5). All 207 (100%) cases that were unclassifiable using the Wassel classification had aberrant components and could be classified using the Rotterdam classification. The majority of these aberrant components were of the triphalangeal (63%), deviating (43%) or hypoplastic (39%) kind (Figure 6).

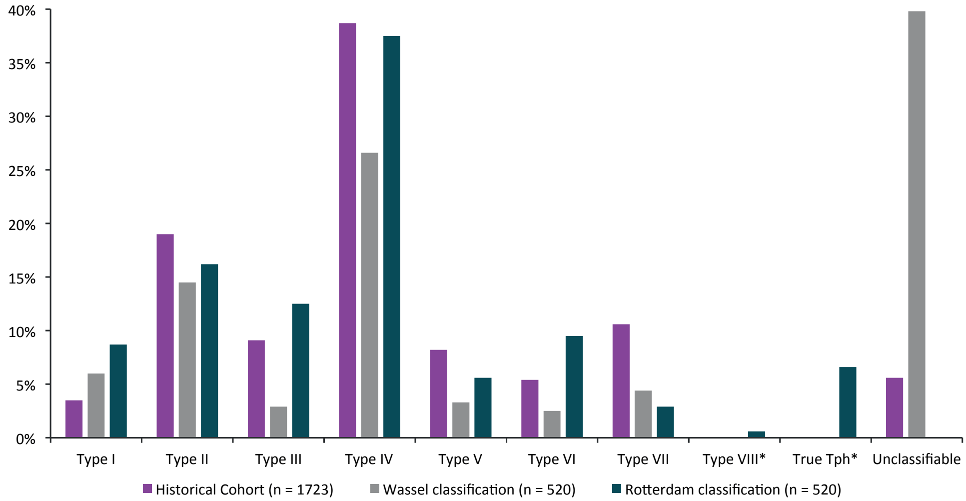

Figure 4: Distribution of radial polydactyly per type.

* Types only available in Rotterdam classification.

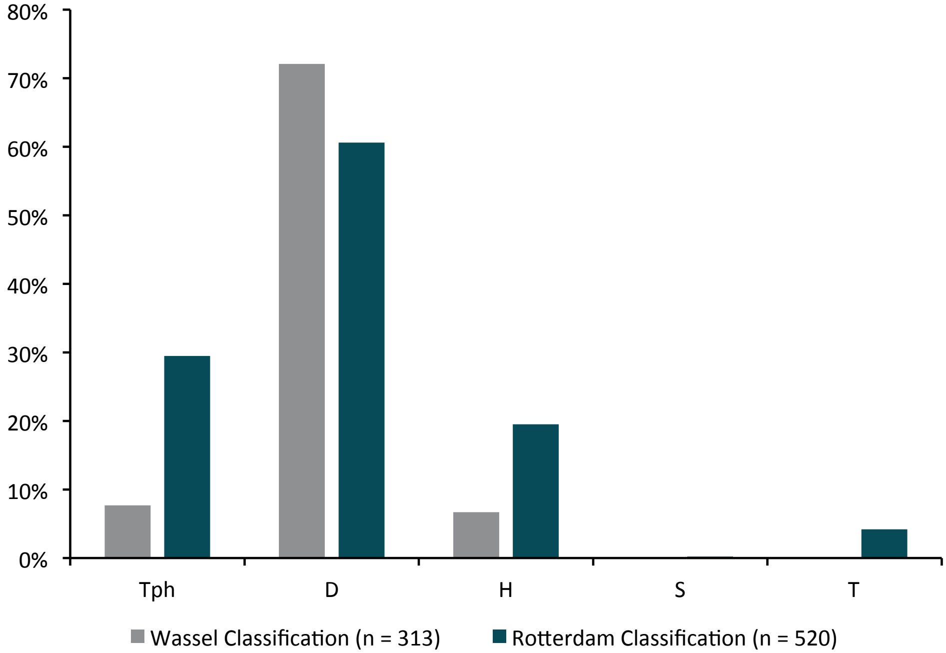

Figure 5: Distribution of aberrant components in classifiable cases when using the Wassel classification.

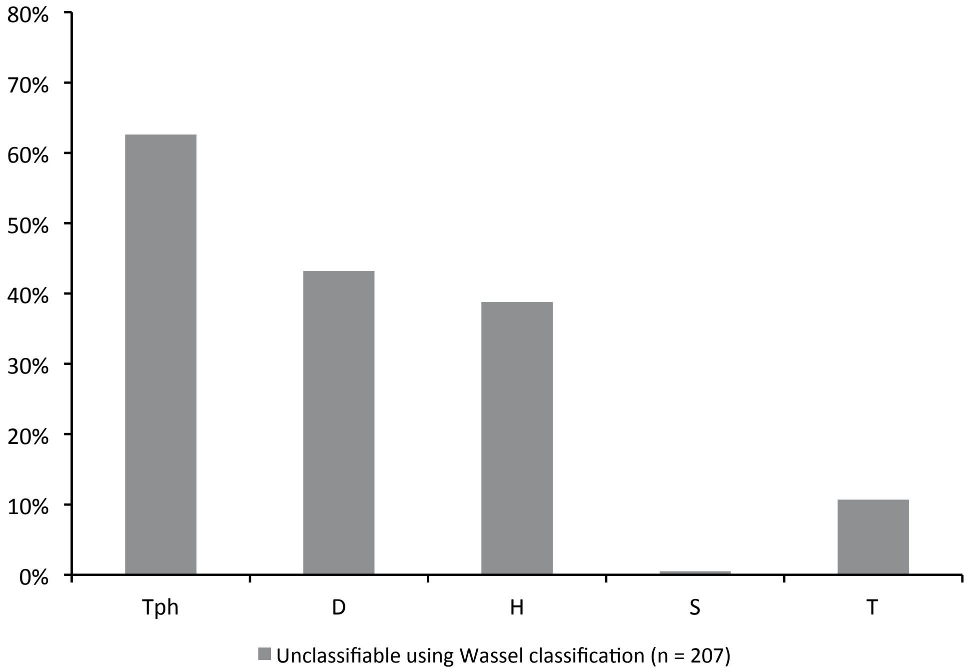

Figure 6: Distribution of aberrant components in unclassifiable cases when using the Wassel classification.

Reliability of the Wassel and Rotterdam classifications

Intraobserver reliability of both classification systems is shown in Table 2. The overall intraobserver reliability of the Wassel classification was similar to the average overall intraobserver reliability of the Rotterdam classification (K = 0.87 and K = 0.83 respectively). The observers from Rotterdam showed reliability comparable to the observers from Hamburg for both classification systems. In the Rotterdam classification, overall intraobserver reliability was lowest for deviating ("D") and hypoplastic ("H") aberrant components (K = 0.70 and K = 0.51 respectively), and when observers indicated both the radial and ulnar thumb had aberrant components ("r/u", K = 0.49).

Table 2: Intraobserver reliability (K)[i] of both the Wassel and Rotterdam classification.

	Rotterdam observers	**Hamburg observers**	**Overall**
Wassel classification	0.88	0.83	0.87
Rotterdam classification*	0.83	0.81	0.83
- Rotterdam classification step 1	0.86	0.89	0.87
- Rotterdam classification step 2	0.96	0.82	0.92
- Rotterdam classification step 3†	0.79	0.81	0.80
If "Tph" component indicated	0.87	0.90	0.88
If "D" component indicated	0.70	0.72	0.70
If "H" component indicated	0.52	0.48	0.51
If "S" component indicated	1.00	0.74	0.92
If no aberrant components indicated	0.78	0.84	0.80
- Rotterdam classification step 4†	0.72	0.71	0.72
If location indicated was "r"	0.68	0.62	0.67
If location indicated was "m"	1.00	1.00	1.00
If location indicated was "u"	0.68	0.66	0.67
If location indicated was "r/m"	1.00	1.00	1.00
If location indicated was "r/u"	0.47	0.54	0.49
If location indicated was "m/u"	1.00	1.00	1.00
If location indicated was "r/m/u"	0.93	1.00	0.95

* These results represent overall intraobserver reliability of the entire Rotterdam classification.
† These results represent overall intraobserver reliability of this step in the Rotterdam classification.

Interobserver reliability of both classification systems is shown in Table 3. The overall interobserver reliability of the Wassel classification was similar to the average overall interobserver reliability of the Rotterdam classification (K = 0.65 and K = 0.70 respectively). For the Rotterdam classification, interobserver reliability was comparable within (K = 0.70 and K = 0.71) and between (K = 0.70) the two clinics. As with the intraobserver reliability, the overall interobserver reliability was lowest when the observers indicated deviating and hypoplastic aberrant components, and if observers indicated both the radial and ulnar thumb ("r/u") had aberrant components (K = 0.42, K = 0.28 and K = 0.30 respectively).

i Kappa coefficients are interpreted as follows: 0.81 – 1.00 almost perfect agreement; 0.61 to 0.80 substantial agreement; 0.41 – 0.60 moderate agreement; 0.21 – 0.40 fair agreement; 0.01 – 0.20 slight agreement; ≤ 0.0 poor agreement.[15]

Table 3: Interobserver reliability (K)[i] of both the Wassel and Rotterdam classification.

	Rotterdam – Rotterdam	Hamburg – Hamburg	Rotterdam – Hamburg	Overall
Wassel classification	0.70	0.52	0.61	0.65
Rotterdam classification*	0.70	0.71	0.70	0.70
- Rotterdam classification step 1	0.78	0.78	0.79	0.79
- Rotterdam classification step 2	0.96	0.82	0.89	0.92
- Rotterdam classification step 3†	0.49	0.60	0.56	0.53
If "Tph" component indicated	0.78	0.62	0.75	0.76
If "D" component indicated	0.36	0.46	0.48	0.42
If "H" component indicated	0.21	0.43	0.33	0.28
If "S" component indicated	1.00	1.00	1.00	1.00
If no aberrant components indicated	0.45	0.63	0.54	0.56
- Rotterdam classification step 4†	0.57	0.63	0.57	0.57
If location indicated was "r"	0.43	0.57	0.46	0.45
If location indicated was "m"	1.00	1.00	1.00	1.00
If location indicated was "u"	0.47	0.54	0.52	0.50
If location indicated was "r/m"	0.74	1.00	1.00	0.89
If location indicated was "r/u"	0.28	0.38	0.30	0.30
If location indicated was "m/u"	1.00	1.00	1.00	1.00
If location indicated was "r/m/u"	0.81	1.00	0.93	0.82

* These results represent overall interobserver reliability of the entire Rotterdam classification.
† These results represent overall interobserver reliability of this step in the Rotterdam classification.

Type-specific reliability was lowest for both Wassel and Rotterdam types II and IV, with intraobserver reliability of K = 0.40 and K = 0.50 (Wassel) vs. K = 0.30 and K = 0.59 (Rotterdam). Interobserver reliability for types II and IV was lower still, with K = 0.33 and K = 0.34 for the Wassel classification vs. K = 0.19 and K = 0.34 for the Rotterdam classification. Deviating and hypoplastic components had intraobserver reliability of K = 0.45 and K = 0.37), and interobserver reliability of K = 0.25 and K = 0.23. If both radial and ulnar thumb had aberrant components intraobserver reliability was K = 0.38, and interobserver reliability K = 0.16.

There was no significant relationship between the experience of the observer in the field of congenital hand surgery and the reliability of either classification system (r_s = 0.60, p = 0.50 and r_s = 0.54, p = 0.16).

DISCUSSION

The aim of this study was to compare occurrence of the various types and reliability of both the Wassel and the Rotterdam classification systems for radial polydactyly. To do this we classified a large series of patients, systematically studied medical literature and conducted a test-retest experiment with seven independent observers.

Regarding the distribution of the different types within both classification systems, our first finding was a considerably larger proportion of the study population (40%) could not be classified using the Wassel classification compared to medical literature (6%). In contrast, we could classify all 520 cases (100%) using the Rotterdam classification. This indicates the Rotterdam classification is better suited to describe the entire spectrum of radial polydactyly, including triplications and triphalangeal thumb.

Second, we found the cases that were unclassifiable using the Wassel classification were predominantly classified as types III, IV and VI in the Rotterdam classification (Figure 4). Furthermore, cases that were unclassifiable using the Wassel classification more frequently had triphalangeal and hypoplastic components compared to classifiable cases (Figures 5 and 6). This suggests that presence of these components in Rotterdam types III, IV and VI is the main reason why cases do not fit the Wassel taxonomy.

Third, all cases unclassifiable using the Wassel classification, had aberrant components. The majority of these aberrant components were of the triphalangeal, deviating and hypoplastic (or 'floating') kind (69%, 42% and 39% respectively, Figure 6). These subtypes often warrant specific surgical approaches for different types of triphalangeal thumbs,[37,38] transverse osteotomies to correct angular deformity of deviating components[2,5] and simple ligation for thumb nubbins.[20] This finding underlines the potential of the Rotterdam classification to guide surgical treatment.

Regarding the reliability of both classification systems, we found good overall intraobserver reliability (K ranging 0.83 – 0.87) and fair interobserver reliability (K ranging 0.65 – 0.70) for both classification systems, with very small differences between the two clinics (Tables 2 and 3). Furthermore, no significant relationship could be found between reliability and years of observer experience. These findings indicate both classification systems can reliably be applied, regardless of experience with their use.

Interestingly, we found observers change their mind and disagree more often about patients with Wassel and Rotterdam types II and IV, and in patients with a deviating or hypoplastic component (Kappa ranging 0.19 – 0.45). This may be caused by difficulty interpreting the x-rays or by unclear definition of aberrant components.

One hundred and thirteen out of 633 cases (18%) did not have available x-rays and therefore could not be classified. However, since the 633 cases had a comparable medical history and a similar proportion of x-rays was missing in both clinics, selection bias is unlikely.

The large discrepancy in unclassifiable cases (6% in literature vs. 40% in our study) may be explained by our exclusion of papers that used the Wassel classification to study specific types of radial polydactyly, instead of their whole population. In addition, triphalangeal subtypes occurred more often in our sample, due to a genetic isolate in the Netherlands.[39] Consequently, the proportion of unclassifiable cases might be different in other parts of the world, as triphalangeal components occurred in 63% of the unclassifiable cases in our study.

Indeed, a single observer classified the whole series. Consequently, type occurrences might differ if other observers would have classified our entire population. However, we found reasonable interobserver reliability for both classification systems (Table 3), suggesting different observers would only mildly affect type occurrence.

The sample for the reliability questionnaires was drawn from the study population. Because RD found no unclassifiable cases using the Rotterdam classification on the study population, the Rotterdam classification lacked an 'unclassifiable' category in the reliability questionnaires.

In addition, the participants were asked to base their classification on a clinical photograph and an x-ray image. Because these were taken at an early age, bone development may have been insufficient to classify synostosis and aberrant components consistently. However, both classification systems are radiological and thus impeded equally in this respect. The experiment therefore resembled clinical practice.

The principal strength from an epidemiological point of view is the sample size, which allows for conclusions regarding radial polydactyly type occurrence, affected hands, and gender predominance in a large Northern European region. In addition, our study presents data on intra- and interobserver reliability of the most widely applied and the most all-embracing classification systems for radial polydactyly. While a number of other classification systems have been proposed, none of these studies provide reliability data.[7-10,40,41] Moreover, the accuracy of the Wassel classification with respect to intraoperative findings has been questioned in literature.[25,42,43] We feel a classification system intended for scientific or clinical use should provide data on reliability to facilitate interpretation of treatment outcome of various subtypes.

Another strength was the participation of observers from both Rotterdam and Hamburg in the reliability analysis of both classification systems. Intra- and interobserver reliability was comparable for the Rotterdam and Hamburg participants (Tables 2 and 3). Only two of the observers, both from Rotterdam, originally helped developing the Rotterdam classification. Moreover, the observers had clinical experience varying from a medical student doing a research internship on congenital hand disorders to experienced congenital hand surgeons. These features of our study design improve the generalizability of our results and correct for potential effects of familiarity of the observers for either classification system.

The Rotterdam classification has a number of advantages over the Wassel classification system. First, it is all embracing, which enables both clinicians and scientists to describe every kind of radial polydactyly, including triplications and triphalangeal subtypes. Second, it is unambiguous on level of duplication, while especially the description of type IV of the Wassel classification has been subject to debate.[11,44-46] Third, its description of aberrant components and their location allows for evaluation of type-specific outcome and may help guide surgical treatment. For example, it can facilitate comparing outcome in regular type IV versus type IV with a triphalangeal ulnar thumb and a relatively hypoplastic radial thumb (type IV Tph u H r), or results of the Bilhaut procedure versus ablation and reconstruction in types where both thumbs are equally hypoplastic (type IV H r/u).

The main drawbacks hindering widespread international use of the Rotterdam classification are its complexity and the fact that surgically relevant aspects like triphalangeal, symphalangeal or synostotic components may not be visible on x-ray at a young age.[5,16,42,43] Although based on the Wassel classification, the expanded taxonomy of the Rotterdam classification initially takes some time and effort to get familiar with. Our results show this can be achieved through a training session of 12 cases, which is available online.[14] To see how the Rotterdam classification system performs in practice, future studies should use it to describe treatment and outcome and define clinically relevant cut-off points for the amount of deviation and the degree of hypoplasia that mandate specific surgical techniques.

Other classification systems have been proposed that arrange the radial polydactyly spectrum based on surgical implications of each subtype.[41,44] However, these classifications are less descriptive, lack similarity to the Wassel classification, and also require interpretation of x-rays. Although clinical relevance of the aberrant components should be investigated further, we feel the Rotterdam classification has greater potential to fulfill the need for clear surgical subdivision of radial polydactyly.

Based on the results of this study, we recommend the Rotterdam classification be used for scientific evaluation of radial polydactyly. The results show it accommodates the entire spectrum of radial polydactyly while maintaining reliability equal to the Wassel classification, even when applied by inexperienced observers. Consequently, the Rotterdam classification paves the way to clear, type-specific description of treatment and outcome in radial polydactyly.

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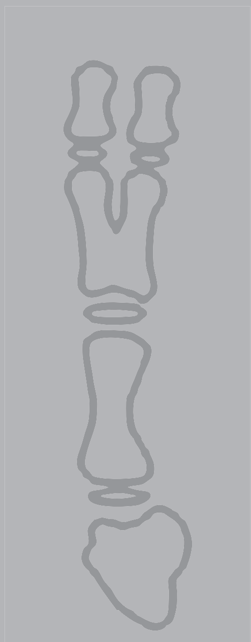
APPENDIX 1 LITERATURE SEARCH TERMS

Embase 828 (805 after undoubling)

((polydactyly/de OR polydactyl*:ab,ti OR hyperdactyl*:ab,ti) AND (radial* OR thumb* OR preaxial* OR (pre NEXT/1 axial*)):ab,ti OR (thumb* NEAR/3 (duplicat* OR triplicat* OR extra* OR addition* OR triphalangial* OR accessor* OR supernumerar* OR 'super numerary')):ab,ti) OR ((('Holt Oram syndrome'/de OR (((atriodigital OR 'heart hand' OR 'hand heart') NEAR/3 syndrome*) OR (holt NEAR/3 (Oram OR oran))):ab,ti) OR ('acrocallosal syndrome'/de OR ((greig OR acrocallosal OR 'acro callosal') NEAR/3 syndrome*):ab,ti) OR ('syndrome VATER'/de OR ((vater OR vacterl) NEAR/3 (syndrome* OR association*)):de,ab,ti) OR ('poland syndrome'/de OR (poland* NEAR/3 (syndrome* OR syndactyl* OR symbrachidactyl*)):ab,ti) OR ('Fanconi anemia'/de OR (Fanconi* NEAR/3 anemia*):ab,ti)) AND ('hand surgery'/exp OR ((hand* NEAR/3 surg*) OR handsurgery):ab,ti))

Medline 696 (49 after undoubling)

((exp polydactyly/ OR polydactyl*.ab,ti. OR hyperdactyl*.ab,ti.) AND (radial* OR thumb* OR preaxial* OR pre axial*).ab,ti. OR (thumb* ADJ3 (duplicat* OR triplicat* OR extra* OR addition* OR triphalangial* OR accessor* OR supernumerar* OR super numerar*)).ab,ti.) OR (((Holt Oram syndrome/ OR (((atriodigital OR (heart adj hand)) ADJ3 syndrome*) OR (holt ADJ3 (Oram OR oran))).ab,ti.) OR (acrocallosal syndrome/ OR ((greig OR acrocallosal OR acro callosal) ADJ3 syndrome*).ab,ti.) OR (VACTERL association/ OR ((vater OR vacterl) ADJ3 (syndrome* OR association*)).mp.) OR (poland syndrome/ OR (poland* ADJ3 (syndrome* OR syndactyl* OR symbrachidactyl*)).ab,ti.) OR (Fanconi anemia/ OR (Fanconi* ADJ3 anemia*).ab,ti.)) AND (exp hand/su OR ((hand* ADJ3 surg*) OR handsurgery).ab,ti.))



CHAPTER 3

Comparison of functional outcome scores in radial polydactyly

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ABSTRACT

BACKGROUND A wide range of outcome assessment systems have been used to describe the results of surgery and evaluate residual impairment in radial polydactyly. We conducted a study to determine which of these assessment systems should be considered superior for the most common types of radial polydactyly (types II and IV), based on interobserver reliability and validity.

METHODS Ten outcome assessment systems were selected from available literature. Three examiners independently evaluated 37 patients, aged 4 – 22 years with radial polydactyly. Patients were also asked to fill out two manual activity questionnaires. Interobserver reliability was determined using an intraclass correlation coefficient ('ICC'). Validity was assessed by correlating results from the outcome assessment systems with functional VAS scores, esthetic VAS scores and manual activity questionnaires.

RESULTS Thirty-seven patients visited the outpatient clinic, yielding 41 hands with radial polydactyly. All patients were evaluated by at least two examiners. Reliability was highest for the Japanese Society for Surgery of the Hand ('JSSH'), Cheng and Tada assessment systems (overall ICC's ≥ 0.70). The JSSH system had highest overall correlations (r_s 0.48 – 0.80 and 0.45 – 0.63) with functional and esthetic VAS scores. No significant correlations were found between the outcome scores and the results of the manual activity questionnaires at an average follow-up time of 94 months.

CONCLUSIONS Interobserver reliability was highest in the JSSH system. Furthermore, the JSSH showed superior correlations with both examiner- and patient-rated VAS scores for functional and esthetic outcome compared to the other nine assessment systems. The poor correlation between the outcome scores and manual activity questionnaires is in line with published literature. We would recommend the JSSH assessment method to be used for the scientific evaluation of radial polydactyly on the body structure and function level of outcome.

INTRODUCTION

Radial polydactyly is a relatively common and diverse Congenital Hand Difference ('CHD'). [1] Many studies have described results and complications of the complex surgical treatment of radial polydactyly. The postoperative evaluation methods applied in these studies range from mainly descriptive systems[2] to strict scoring forms.[3]

Early outcome studies in radial polydactyly often provided case-specific results of surgery.[4] In later years, postoperative assessment was structured, based on certain evaluation criteria[5,6] A comprehensive scoring system was first introduced by Tada et al. in 1983.[7] This commonly used [8-12] and frequently modified [13-16] system consists of range of motion, joint instability, and mal-alignment parameters. Consequently, the definition of outcome is solely based on functional parameters in the Tada scoring system. Other assessment systems include esthetic and subjective factors as important features of postoperative evaluation.[17-19]

Despite the large number of assessment systems and their frequent use, no evidence exists that these systems are reliable and valid indicators of overall surgical outcome or residual impairment, and whether they can be applied regardless of observer experience. In addition, patient and caregiver satisfaction and manual activity have become increasingly important determinants of outcome in contemporary research, but the relation between patient satisfaction, manual activity and functional and esthetic outcome in radial polydactyly has not been well described.

The purpose of this study was to determine which outcome assessment system is most suitable for postoperative evaluation of the most common types of radial polydactyly. We evaluated the interobserver reliability of ten outcome assessment systems that assess body structure and function. To evaluate the validity of these ten outcome assessment systems, we correlated the outcome scores with VAS scores for functional and esthetic results of three examiners and patients. Furthermore, we measured manual activity using questionnaires and correlated their results with scores of the outcome assessment systems. Although this paper does contain data describing overall outcome, it should be noted that our methodology was not designed to describe our long-term treatment results.

MATERIALS AND METHODS

Patients and examiners

We examined the patient registry for patients with radial polydactyly treated at our hospital between 1993 and 2011. Patients operated for Flatt[20] (or 'Wassel[4]') Types II and IV radial polydactyly, aged 4 years and older, and with a minimal postoperative follow-up of one year, were eligible for inclusion. Patients with severe psychomotor impairment due to multiple congenital anomalies and perinatal asphyxia were excluded from this study.

Two experienced congenital hand surgeons (CvN and SH) and one well-trained medical intern (RD) separately evaluated the patients upon their visit to the outpatient clinic. The intern was prepared for this project during several international clerkships with renowned congenital hand surgeons and was trained in conducting measurements in children by a pediatric hand therapist. Examiner RD was not involved in the surgical treatment of the patients and therefore an independent examiner, which allowed for an unbiased evaluation. This study was done under approval of an accredited Medical Research Ethics Committee (MEC-2010-295) and in accordance with the declaration of Helsinki.

Outcome assessment systems

Based on the available literature, ten clinical assessment systems were selected.[5-7,13-15,17-19,21] In this cross-sectional study the three examiners were completely blinded from each other's evaluation. Every examiner performed all standardized measurements necessary to compute the overall score of all outcome assessment systems for all patients, with exception of the strength measurements, which were conducted only once. Depending on the specific assessment system, measurements of the affected hand were compared to the unaffected contra-lateral hand. In case of bilateral involvement, normative values were used for comparison.

The content validity of each outcome assessment system was evaluated by consensus. All examiners were separately asked to indicate the seven items they felt were essential to assess outcome in radial polydactyly. After the first round, the authors agreed on six and disagreed on two items. In the second round, after reading each other's comments, the following items were considered determinants of content validity of an outcome assessment system for radial polydactyly by consensus: thumb alignment, interphalangeal (IP) and metacarpophalangeal (MCP) joint stability, combined IP and MCP range of motion, appearance of the nail and nail folds, prominence or residual deformity of the pulp and patient satisfaction. The assessment systems investigated in this study are shown in Figure 1.

Clinical Patient Evaluation

IP and MCP joint excursions, joint stability, and joint alignment were all measured using a handheld goniometer. The Pollexograph[22] and the Kapandji score[23] were used to measure palmar abduction and opposition. Tip, tripod, and key pinch strength were measured using a pinch strength dynamometer (Baseline, FEI, Irvington NY10533, USA). Thumb length was measured from the palpable base of the proximal phalanx to the tip of the thumb. Girth of the thumb was measured at the IP joint with the joint in neutral position. Pulp circumference was defined as the circumference at the level of the lunula. The width of the nail was measured at the lateral edges of the nail, where the lateral nail folds meet the hyponychium.

	Tada[1]	Ogino[1]	Larsen[1]	Horii[2]	ALURRA[2]	Cheng[2]	JSSH[2]	Wood[3]	Dobyns[3]	Tuch[3]
FUNCTION										
Flexion / Extension*	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Stability*	✓	✓	✓	✓	✓	✓	✓		✓	✓
Alignment*	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Abduction							✓		✓	
Opposition						✓			✓	✓
Extension lag							✓	✓		
Strength			✓			✓			✓	✓
First web space						✓				✓
Overall function								✓		
ESTHETICS										
Nail*					✓	✓	✓		✓	
Pulp*					✓	✓	✓		✓	✓
Size					✓		✓		✓	✓
Scar							✓	✓		
Prominence*						✓	✓			✓
Overall appearance				✓				✓		
SUBJECTIVE EVALUATION										
Pain							✓			
Patient satisfaction*							✓			✓
SCORING										
Excellent							✓	✓	✓	n.a.
Good	✓	✓	✓	✓	✓	✓	✓	✓	✓	n.a.
Fair	✓	✓	✓	✓	✓	✓	✓	✓		n.a.
Poor	✓	✓	✓	✓	✓	✓	✓	✓	✓	n.a.
Scale	0-5	-1-5	0-9	0-7	0-24	0-30	0-20	n.a.	n.a.	n.a.
VALIDITY										
Content†	3/7	3/7	3/7	3/7	5/7	6/7	7/7	2/7	5/7	6/7

Figure 1: Summary of the reviewed outcome assessment systems.

1: purely functional outcome scores; 2: outcome scores evaluating function, esthetics and subjective aspects; 3: structured outcome criteria systems; *: essential item based on consensus; †: content validity based on consensus, number of essential items given; n.a. = "not available"

Examiners were asked to estimate whether the first web-space and thumb size were sufficient to allow acceptable function. In addition, examiners and patients or their caregivers were asked to assess functional and esthetic outcome on a VAS.[2,8,24,25] The functional VAS ranged from "no functional thumb at all" to "a perfectly functioning thumb". The esthetic VAS ranged from "so disfigured that the patient would prefer not to have a thumb at all" to "a perfectly-normal looking thumb".

The caregiver VAS score was only used if the child appeared to be too young to understand the question. For simplicity, we will refer to these results as "patient-rated" throughout the manuscript. Patients themselves were asked to assess pain on numeric rating scale with 0 representing no pain at all and 10 representing the worst pain imaginable.[26]

Questionnaires

Prior to their visit to the clinic, all patients were asked to fill out the ABILHAND-kids questionnaire.[27] Additionally, a modified Prosthetic Upper Extremity Functioning Index ('PUFI') questionnaire [28] was used in patients with a unilateral CHD. Both questionnaires quantify the child's manual ability by assessing ease of performance of everyday tasks.

Parents or caregivers of younger patients were asked to verbally or manually assist their child in completing the questionnaires when needed. No hard cut-off age was used to determine parent-assistance, though most children could independently fill out the questionnaires by the age of ten years.

Statistical analysis

All outcome scores were computed after clinical evaluation of the patient. Interobserver reliability of the outcome assessment systems was calculated with an intraclass correlation coefficient (ICC 2,1), using a two-way random single measure agreement model.[29] Spearman's correlation coefficient (r_s) was used to assess the relationship between the outcome assessment systems and functional and esthetic VAS scores. Additionally, Spearman's correlation coefficient was used to analyze correlation between the outcome assessment systems and results of the manual activity questionnaires.

RESULTS

We identified 211 patients with radial polydactyly from the patient registry, 58 (27%) of which met the inclusion criteria. Thirty-seven out of these 58 (64% response rate) visited the outpatient clinic. A total of 41 cases of radial polydactyly were evaluated. Examiner RD assessed all 41 cases. CvN and SH assessed 40 (98%) and 20 cases (49%) respectively. The average age at follow-up was 10 years (range 4 – 22); the average age at first surgery was 1.9 years (range 0 – 7). Patient characteristics of the studied population are shown in Table 1.

Table 1: Study population characteristics.

Characteristic	Participants	Non-Participants
N total population	37 (64%)	21 (36%)
Boy	17 (46%)	12 (57%)
Girl	20 (54%)	9 (43%)
N hands with radial polydactyly	41 (65%)	22 (35%)
Wassel type II	19 (46%)	9 (41%)
Wassel type IV	22 (54%)	13 (59%)
N patients with bilateral CHD	8 (22%)	3 (14%)
Radial polydactyly type II	3 (38%)	1 (33%)
Radial polydactyly type IV	1 (12%)	-
Radial polydactyly type II & IV	1 (12%)	1 (33%)
Contralateral thumb nubbin	1 (12%)	-
Contralateral thumb hypoplasia	1 (12%)	-
Contralateral radial club hand	1 (12%)	-
Contralateral syndactyly	-	1 (33%)
Mean number of surgeries (SD)	1.2 (0.6)	1.4 (0.7)
1 surgery	31 (84%)	15 (71%)
2 surgeries	5 (13%)	4 (19%)
> 2 surgeries	1 (3%)	2 (10%)
Mean age at first surgery in years (SD)	1.9 (1.8)	1.4 (2.1)
0 – 1 years of age	8 (22%)	7 (33%)
1 – 2 years of age	14 (38%)	9 (43%)
> 2 years of age	15 (40%)	5 (24%)
Range in years	0 – 7	0 – 10
Mean age at follow-up in years (SD)	10 (5.2)	13 (5.3)
< 7 years of age	11 (30%)	3 (14%)
7 – 17 years of age	22 (60%)	14 (67%)
≥ 18 years of age	4 (10%)	4 (18%)
Range in years	4 – 22	4 – 23
Mean duration of follow-up in years (SD)	9.3 (5.3)	11.9 (4.9)
< 5 years of follow-up	11 (30%)	2 (9%)
5 – 10 years of follow-up	11 (30%)	6 (29%)
> 10 years of follow-up	15 (40%)	13 (62%)
Range in years	1 – 22	3 – 21

CHD = "Congenital Hand Difference"

Figure 1 (above) also shows the items evaluated in each outcome assessment system. Seven systems (Tada, Ogino, Larsen, Horii, ALURRA, Cheng and JSSH) are designed to calculate an outcome score. The three remaining systems (Wood, Dobyns and Tuch) are combined evaluation criteria that define overall outcome. Each system contains 3 – 12 items. Three systems (Tada, Ogino and Larsen) do not evaluate esthetic aspects of outcome. Patient satisfaction was integrated as a determinant of outcome in the JSSH and Tuch system. Regarding content validity, we found the JSSH, Cheng and Tuch assessment systems encompassed the most items defined as essential by the examiners.

Results of the assessment of all patients are shown for each assessment system in Figure 2. While median outcome scores were fair to good, Figure 2 clearly illustrates that the conclusion on overall outcome heavily depends on the applied assessment system. For example, while approximately 75% of all outcomes were in the "good" to "excellent" range in the Cheng and Wood systems, less than 25% is considered "acceptable" using the Dobyns system. Figure 2 also contains information on the discriminative properties of the outcome assessment systems, as some systems classify the majority of patients in only one or two categories. Although not shown in Figure 2, a number of assessment systems were not sensitive enough to detect a difference between "poor" and "fair" outcome for one or more of the examiners. This was the case in the Larsen (CvN), ALURRA (SH), Cheng (RD & CvN) and Wood (CvN & SH) assessment systems. The Tuch system lacks a "fair" outcome category.

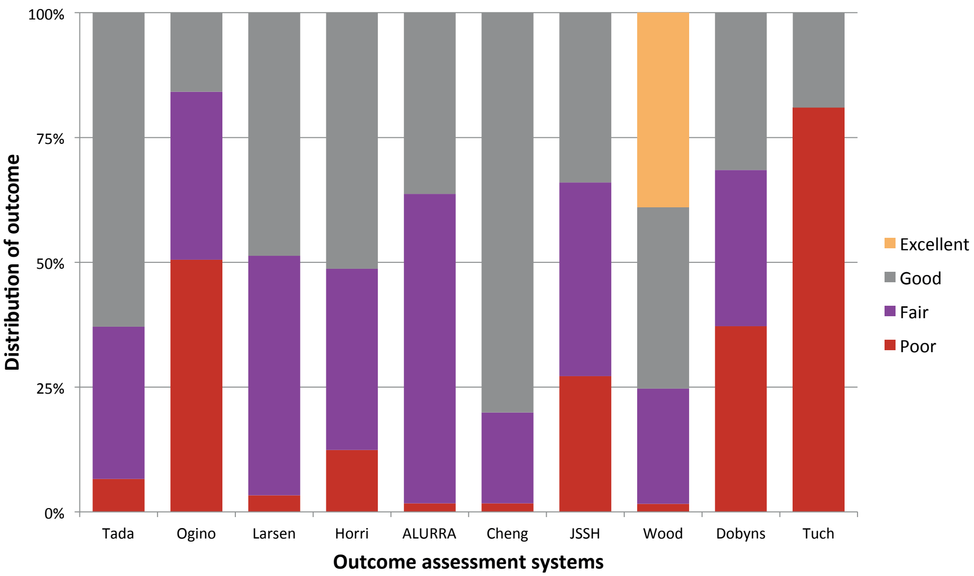

Figure 2: Distribution of outcome per system.

The interobserver reliability of the various outcome assessment systems is shown per assessment system in Table 2. The overall agreement between the examiners was highest for the JSSH assessment system, followed by the Cheng and Tada systems (overall ICC's > 0.70). The Cheng and Tada systems respectively had one and two ICC's < 0.70. Agreement was generally higher between both congenital hand surgeons when compared to the independent examiner.

Table 2: Intraclass Correlation Coefficients (ICC) for the various outcome assessment systems, for different combinations of examiners p < 0.05.

Examiners	Tada	Ogino	Larsen	Horii	ALURRA	Cheng	JSSH	Wood	Dobyns	Tuch
RD, CvN & SH	0.68	0.59	0.66	0.67	0.68	0.70	0.71	0.60	0.56	0.54
RD & CvN	0.62	0.57	0.71	0.64	0.65	0.70	0.77	0.34	0.56	0.45
RD & SH	0.76	0.47	0.69	0.67	0.58	0.64	0.73	0.57	0.58	0.49
CvN & SH	0.82	0.89	0.73	0.76	0.82	0.85	0.75	0.91	0.75	0.70

Regarding manual activity, patients scored a median of 41 out of 42 points (37.5 – 42.0 IQR) in the ABILHAND-kids questionnaire. They also scored themselves as able to complete all tasks (100%, 95.6% – 100% IQR) in the modified PUFI questionnaire. We found a weak correlation between age at follow-up and ABILHAND-kids score ($r_s = 0.36$, $p < 0.05$), but no significant correlation was found between age and PUFI score ($r_s = 0.32$, $p > 0.05$). No significant correlations were found between the various outcome assessment systems and results from the manual activity questionnaires (r_s ranging -0.33 – 0.43, $p > 0.1$).

Significant correlation coefficients were found between the examiner-rated VAS scores for function and esthetics (Figure 3) and the Horii, Cheng, JSSH, Wood and Dobyns outcome assessment systems. The strongest correlations were found between the VAS for functional outcome by SH and the Dobyns and Wood outcome assessment systems of the corresponding examiner ($r_s = 0.93$ and $r_s = 0.86$ respectively). However, the variability between the correlations was considerable in these systems (r_s ranging 0.33 – 0.93 and 0.34 – 0.86 respectively). Consequently, overall correlations were better for the Horii and JSSH systems (r_s ranging 0.47 – 0.75 and 0.48 – 0.80 respectively), indicating only moderately strong linear relations.

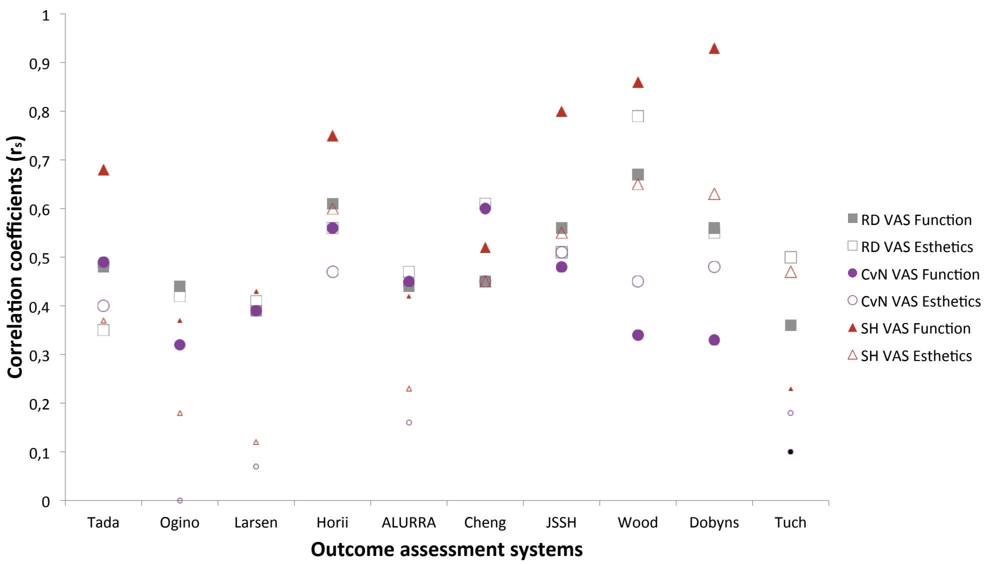

Figure 3: Correlations (r_s) between examiner-rated VAS scores of functional and esthetic outcome per examiner, per assessment system.

The larger markings represent statistically significant correlations ($p < 0.05$), the smaller markings are non-significant correlations ($p > 0.05$).

Correlation coefficients between the patient-rated VAS scores for function and esthetics, and the outcome on the different assessment systems (Figure 4), were absent to moderate. Overall, however, the strongest significant correlations were found for the Horii and JSSH systems (r_s ranging 0.44 – 0.58 and 0.45 – 0.63 respectively). The average significant correlations between patient-rated VAS scores and the outcome assessment systems were $r_s = 0.43$ for function and $r_s = 0.31$ for esthetics.

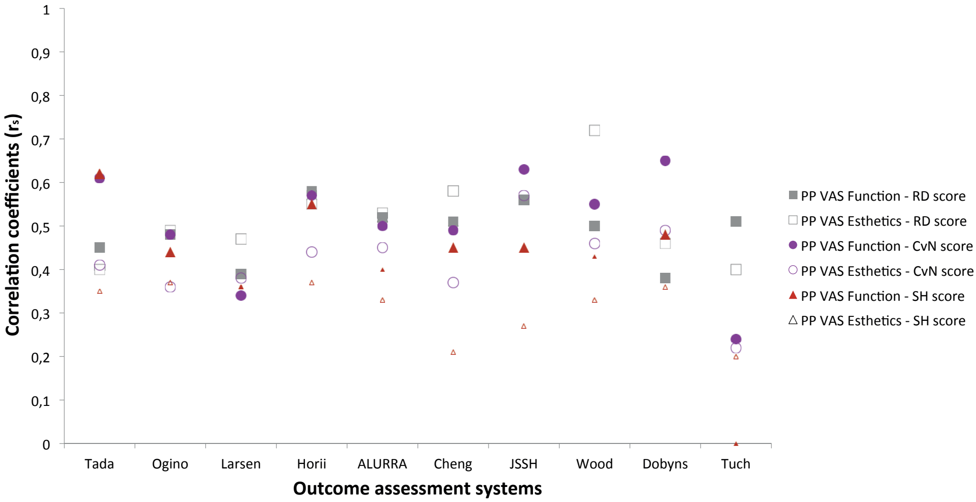

Figure 4: Correlations (r_s) between patient-rated VAS scores of functional and esthetic outcome per examiner, per assessment system.

The larger markings represent statistically significant correlations ($p < 0.05$), the smaller markings are non-significant correlations ($p > 0.05$). PP = "Patient/Parent".

DISCUSSION

The aim of this study was to determine which of the available assessment systems is best suited for postoperative evaluation of outcome in radial polydactyly. We found that conclusions on overall outcome can vary substantially within a single patient, depending on assessment system used. For example, the results of surgery shown in Figure 5 would be "excellent" if one applies Wood's criteria; "fair" using the JSSH assessment system; or "poor" according to Tuch's criteria.

Our results show that the JSSH assessment system has the most optimal properties. It has the highest overall interobserver reliability compared to the other nine assessment systems reviewed. Furthermore, it has relatively high correlations with VAS scores for function and esthetics. Additionally, it can differentiate between at least three outcome categories. Finally, it encompasses all important postoperative evaluation parameters (CME). Since we found no correlations between any of the assessment systems and manual activity questionnaires, these results did not contribute to selection of the most optimal system.

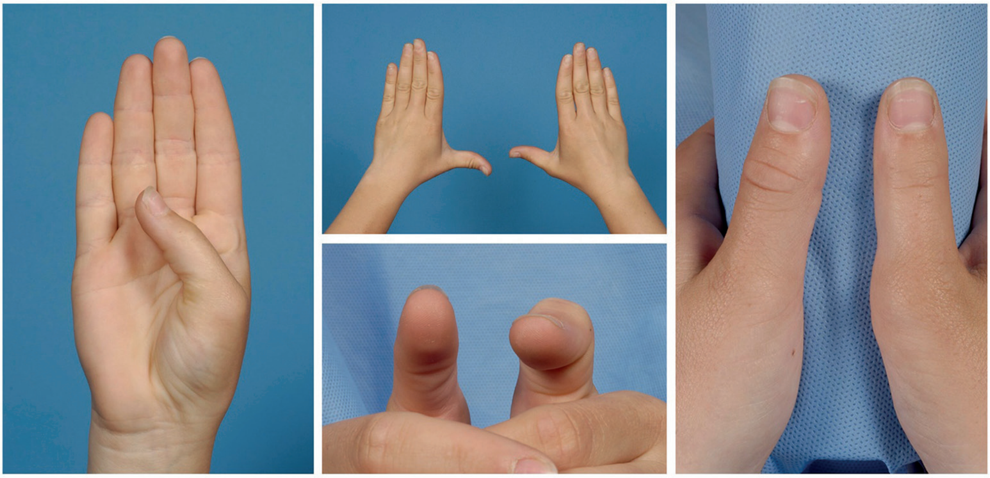

Figure 5: Postoperative situation after a Wassel type IV radial polydactyly on the right hand. Slight stability and mobility issues substantially influence the conclusion on overall outcome, which was rated as follows by a single examiner: Tada – fair; Ogino – fair; Larsen – fair; Horii – fair; ALURRA – fair; Cheng – good; JSSH – fair; Wood – excellent; Dobyns – good; Tuch – poor.

Although the Cheng, Tada and the JSSH assessment systems were the three most reliable methods to evaluate outcome (Table 2), the JSSH system has a number of additional assets. First, in contrast to the Cheng and Tada systems, the ICC was consistently larger than 0.70 for the JSSH system. Second, the overall correlations with functional and esthetic VAS scores were higher for the JSSH assessment system. Third, the Cheng system could not discriminate between the "poor" and "fair" outcomes in two examiners (RD & CvN), making it less suitable to detect improvement after revision surgery. Last, since preoperative patient characteristics influence postoperative residual impairment, in a solely functional system that emphasizes on joint mobility like the Tada score, a patient with poor preoperative joint motion will never achieve a good outcome. Moreover, the Cheng and Tada systems do not incorporate residual pain and patient satisfaction as determinants of outcome, while patient satisfaction is an especially important feature to our mind.

While the highest correlations with the examiner-rated VAS scores were found in two more subjective systems (i.e., Dobyns and Wood), the difference in correlation between the various examiners for these systems was large. For example, the correlation between the VAS of function and Dobyns' outcome assessment system was 0.93 for one hand surgeon and 0.35 for the other, suggesting these systems are less reliable.

The absence of significant correlations between any of the systems and the manual activity questionnaires can be explained by a majority of patients performing excellent in these questionnaires (a 'ceiling effect'). From our data, it cannot be concluded whether this results from radial polydactyly being a relatively mild functional impairment regardless of the success of the surgery, or that these questionnaires might not be sensitive enough to discriminate between good and poor results in thumb-related problems.

The main limitations of this study are the relatively small sample size and the fact that all three examiners did not evaluate every patient. Based on the originally described frequency [4] and total number of patients (i.e., 211), one expects an approximate of 122 patients to have Wassel type II or IV. The main criteria limiting inclusion to 58 patients were age 4 years and older, minimal postoperative follow-up of one year, and the relatively high occurrence of radial polydactyly with triphalangeal components in our population.[30] Nevertheless, we were able to show considerable differences in reliability, discriminative properties and correlation consistency between the different systems. Since this was also the aim of the study, we are confident that our sample size was adequate for this purpose. Increasing sample size might be of influence on correlations with questionnaires that failed to reach the significance level of $p < 0.05$ in the current setting. However, comparable studies have also found poor correlations between outcome involving body structures and function and questionnaires on manual activity or impairment.[24,25]

Another limitation is the validity of the ABILHAND-kids and PUFI questionnaires as measures of manual ability, as these questionnaires were only validated for use in pediatric cerebral palsy [27] and transverse upper limb deficiencies [31] populations. Though alternative measures of manual activity such as the Assisting Hand Assessment ('AHA') [32] exist, most of them are very time consuming and require specific expertise of hand therapists. In contrast, filling out a questionnaire and completing the JSSH assessment system takes about 15 minutes, which is more feasible during a busy outpatient clinic.

A possible third limitation is the restriction of our study population to the most prevalent types of radial polydactyly, i.e., types II and IV. Although this might impair the generalizability of the results, Wassel types II and IV have affected IP and MCP joints, making evaluation of alignment, stability, mobility, and strength more relevant. These items are incorporated in most outcome assessment systems.

One of the strengths of this study was the combined assessment of patients by two highly experienced surgeons and an independent examiner in the analysis of interobserver reliability. The independent examiner was qualified to conduct patient evaluation, but lacked the surgical experience of the two frequently collaborating congenital hand surgeons. This is a realistic research setting, because clinical research is often conducted either by observers that are in some stage of medical training or by physician assistants, due to the busy schedules of experienced consultants. Indeed, we did find considerable differences in interobserver reliability between the two congenital hand surgeons and the independent examiner in the majority of assessment systems. Although the surgical inexperience of the independent examiner may have lowered the overall agreement, we believe that the validity and generalizability of an assessment system is increased when it can be reliably applied irrespective of surgical experience in the field of congenital hand differences. Moreover, studies where the operating surgeon assesses the final outcome are prone to bias, making the JSSH system particularly valuable in designs with an independent examiner.

Another strength was that we related patient satisfaction on function and esthetics to outcome assessment of the three examiners. Our findings suggest poor-to-moderate correlations between doctor- and patient rated outcome, which is a well-known phenomenon.[2,6,8,24,33] Moreover, patient satisfaction is an important reason for seeking medical attention, a determining factor for performing revision surgery, and a substantial confounder in the outcome evaluation after revision surgery. We therefore believe that patient satisfaction should be an integral part of outcome assessment, as in the JSSH system.

A third strength was the systematic evaluation of validity of the outcome assessment systems. Validity evaluation usually includes assessment of content validity (i.e., whether the system measures all facets of the construct it's supposed to measure, here overall outcome in radial polydactyly); construct validity (i.e., whether the system measures or correlates with the construct that it claims to measure) and criterion validity (i.e., the extent to which results obtained by the system relate to concrete criteria in the 'real world'). In the absence of a golden standard to describe outcome of hand function in radial polydactyly, we assessed content validity based on consensus, construct validity based on correlations with VAS scores and criterion validity based on correlation with manual activity questionnaires.

A general limitation of the assessment systems is their item weighting. For example, the JSSH system assigns the same amount of points to MCP joint stability as IP joint flexion. Presently, these item weights are not based on their effect on overall hand function. Possibly, the JSSH system could benefit from a redistribution of points over the various items, based on their impact on overall hand function.

This study compares the reliability and validity of assessment systems designed to study outcome and residual impairment in radial polydactyly. The Tada score (or modified versions of this assessment system) has been applied most frequently to describe outcome in radial polydactyly. [7-16] However, the results of our study indicate that the JSSH system is better suited for this purpose. In addition to better reliability and overall correlations, the JSSH encompasses more items that we feel are relevant to describe overall outcome. We therefore recommend the JSSH assessment system for future scientific evaluation of outcome and residual impairment after surgery for radial polydactyly.

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CHAPTER 4

A clinically weighted approach to outcome assessment in radial polydactyly

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ABSTRACT

BACKGROUND Currently available outcome assessment systems for radial polydactyly are mainly based on expert opinion. The aim of this study was to develop an outcome assessment system based on clinical data.

METHODS We performed linear regression analysis on data from a multicenter study of 121 patients with radial polydactyly types II, IV and VII to develop a clinically weighted outcome assessment system. Items were weighted according to their influence on overall functional and esthetic outcome in the regression analysis.

RESULTS Active flexion, scar appearance and prominence at amputation site were the main items influencing overall functional and esthetic outcome (β=0.393, β=0.326, and β=0.288 respectively). Palmar abduction, metacarpophalangeal joint deviation and nail appearance influenced overall functional and esthetic outcome the least (β=-0.002, β=-0.104, and β=0.070 respectively).

CONCLUSIONS Our proposed assessment system for radial polydactyly reflects the way clinicians value individual aspects of outcome as determinants of overall outcome and helps guide future treatment and evaluation of outcome.

INTRODUCTION

Studies on radial polydactyly mostly focus on operative technique and treatment outcome.[1-4] Treatment outcome is often evaluated in a structured way using an assessment system.[5-8] It has been shown recently that the assessment system proposed by the Japanese Society for Surgery of the Hand (JSSH) has the best reliability and validity.[9] Despite these findings, modifications have already been proposed to further improve the JSSH system.[10]

In the original JSSH system (Figure 1),[11] 20 points are distributed over 13 items: seven functional, four esthetic and two subjective items. The functional items comprise two points each, leaving one point per item for the esthetic and subjective parts of the assessment system. Consequently, functional results contribute more than esthetic and subjective results to the total score in the JSSH system. This imbalance between the number of items and points in the functional, esthetic and subjective domains may be unfounded, since it is debatable whether radial polydactyly constitutes a mainly functional impairment.[12,13] In addition, it is unknown whether the items in the functional and esthetic domains only affect overall function and appearance respectively; they might affect both.[14,15] Moreover, although the points are distributed over up to three categories in each item, there is no apparent basis on which the categorical cut-off values were chosen. The dichotomization of esthetic items in particular seems arbitrary and unpractical.

The aim of this study was to develop a clinically weighted outcome assessment system for radial polydactyly.

Name	____________	Examinar	__________
Age	____________ years	Date	__________
ID	____________	Wassel type	__________
Sex	(M / F)	Dextrity	(yes / no)
Laterality	(R / L)		

FUNCTION

Alignment

IPj	5 or less	☐	2 points
	6-19	☐	1 point
	20 or more	☐	0 point
MPj	5 or less	☐	2 points
	6-19	☐	1 point
	20 or more	☐	0 point

Stability

IPj	10 or less	☐	2 points
	11-19	☐	1 point
	20 or more	☐	0 point
MPj	40 or less	☐	2 points
	39-59	☐	1 point
	60 or more	☐	0 point

Active flexion

IPj+MPj	90 or more	☐	2 points
	61-89	☐	1 point
	60 or less	☐	0 point

Extension lag

IPj+MPj	0	☐	2 points
	29 or less	☐	1 point
	30 or more	☐	0 point

Active palmar abduction

CMj+MPj	60 or more	☐	2 points
	31-59	☐	1 point
	30 or less	☐	0 point

COSMESIS

Size	good	☐	1 point
	poor	☐	0 point
Pulp/Nail	good	☐	1 point
	poor	☐	0 point
Scar	good	☐	1 point
	poor	☐	0 point
Prominence	good	☐	1 point
at MPj	poor	☐	0 point

SUBJECTIVE

Pain	good	☐	1 point
	poor	☐	0 point
Satisfaction	good	☐	1 point
	poor	☐	0 point

Sum ☐ points

Excellent	☐	20 points
Good	☐	17-19 points
Fair	☐	14-16 points
Poor	☐	0-13 points

Figure 1: The original JSSH assessment system for evaluation of overall outcome after surgery for radial polydactyly.

MATERIALS AND METHODS

Patients and measurements

This study was performed in the Sophia Children's Hospital in Rotterdam, the Netherlands and the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. Patients with Wassel types II, IV and VII radial polydactyly, aged 4 years and older and with a minimal postoperative follow-up of one year were eligible for inclusion. Five congenital hand surgeons and one PhD candidate (a physician) evaluated the patients.

All parameters of the JSSH system were documented by at least two independent observers. Angular deviation and active and passive ranges of motion (ROM) of the interphalangeal (IPJ) and metacarpophalangeal (MCPJ) joints were measured using a handheld goniometer, as was instability, which was measured whilst applying a lateral stress to the joint. Palmar abduction was measured using the pollexograph.[16]

In addition, the observers were asked to assess overall functional results on a VAS, ranging 0 – 100 points. The functional VAS anchors were "no functional thumb at all" to "perfectly functioning thumb". The overall esthetic results were also assessed on a VAS ranging 0 – 100 points, as were thumb size, pulp, nail, scar and prominence at amputation site. This VAS had anchors from "extremely ugly" to "looks perfectly normal".

Furthermore, patients were asked if they experienced any pain and whether they were satisfied with the overall outcome, their "yes or no" answers were documented.

Alternative outcome assessment system

The proposed alternative outcome assessment system was developed from the existing JSSH system using the following steps.

1. We separated the combined "nail/pulp" item. Because thumb pulp and nail can both be affected to a different extent, we hypothesized these items may separately influence overall esthetic outcome.
2. We added points to the esthetic and patient-reported items to improve the balance between these domains and the functional domain. At baseline, all esthetic and functional items had two points and three outcome categories, while the patient-reported items had three points each. Consequently, our proposed system has 30 points in total, while the JSSH system has 20.
3. We determined how each item from the JSSH system influences overall functional and esthetic outcome using regression analysis. Two separate regression analyses were done: one analysis containing the functional items; and another analysis containing the esthetic items. In the analysis of the functional items, the VAS score of overall functional outcome was the dependent variable, while the IPJ deviation, MCPJ deviation, IPJ instability, MCPJ instability, combined active IPJ and MCPJ flexion, extension lag, and palmar abduction

were the independent variables. For the analysis of the esthetic items, the VAS score of overall esthetic outcome was the dependent variable, while the VAS scores for thumb size, nail, pulp, scar, and prominence at amputation site were the independent variables. IPJ deviation, MCPJ deviation and size were added as independent variables in both analyses, because we hypothesized these parameters could influence both function and appearance. We also tested for interactions between items, to assess whether the impact of an item would depend on the value of other items.

4. We redistributed the 30 points over the items according to their impact on overall outcome, by using their weight and significance in the regression analyses.
5. Depending on the number of points each item was given in step four, we redefined the cut-off values for that parameter according to the distribution in our sample, using medians, tertiles and quartiles. Subsequently, these cut-off values were compared to item-related medical literature, to avoid penalizing outcome values that fall within the range for a healthy child.
6. We applied both the JSSH system and our proposed system to the entire study population and evaluated the distribution of overall outcome.
7. We also compared interobserver agreement, internal consistency and correlation between the existing JSSH system and our alternative system in our dataset.

Statistical analysis

The linear regression analyses were done including all items from the JSSH system ('forced entry method'), since we did not want to remove any original items or introduce new items into our proposed assessment system. Several regression analyses were done to determine how individual items influence overall functional and esthetic outcome, and whether an item should be included in the functional or esthetic domain of our proposed assessment system. The two regression models that could best predict overall functional and esthetic outcome (based on R^2 and Fisher's exact test) were selected.

After the regression analyses established the two optimal models for predicting overall functional and esthetic outcome and the appropriate domain for each item, we redistributed the points over all items based on their weight (standardized regression β-coefficients) in the two optimal regression models, meaning items with higher weight were given more points in our proposed assessment system.

To compare the JSSH system with our proposed assessment system, internal consistency[i] was assessed using Cronbach's α. In addition, interobserver reliability was assessed using an Intraclass Correlation Coefficient (ICC 2,1). The distribution of overall outcome was assessed using histograms, while normality assumptions in the linear regression analyses were tested

i The *internal consistency* expresses how well items in a system measure the same general construct (i.e., functional and esthetic outcome). It is often quantified using Cronbach's α, which ranges 0 – 1; the closer to 1, the better the internal consistency.

using the Kolmogorov-Smirnov test. Correlation between the two assessment systems, and between the observed and expected values of the regression analyses,[ii] were assessed using Spearman's correlation coefficient (r_s).

This study was approved by the Medical Research Ethics Committee (MEC-2010-295); informed consent was signed for all subjects.

RESULTS

One hundred and twenty-one (44%) out of the eligible 275 patients had complete follow-up and were included in the study; a total of 131 affected hands. Eighty-six patients (31%) could not be contacted, 60 (22%) were unable or unwilling to participate, two (1%) had died and six (2%) had incomplete follow-up data. Patient characteristics are shown in Table 1, together with the data used to design our proposed outcome assessment system.

Tables 2 and 3 show the regression coefficients for the functional and esthetic items. The explained variance[iii] of functional outcome (R^2 = 0.37) was much lower than for esthetic outcome (R^2 = 0.81). Of the functional items (Table 2), combined active IPJ and MCPJ flexion was the most important factor (β=0.393, p<0.001), whereas palmar abduction contributed least (β=-0.002, p=0.981). Although extension lag did not contribute significantly, the interaction term[iv] between active flexion and extension lag (β=0.569, p=0.032) indicated extension lag did explain part of the functional VAS score variance.

For the esthetic items (Table 3), scar appearance was the most important factor (β=0.326, p<0.001), while the nail appearance contributed least (β=0.070, p=0.165). IPJ deviation, MCPJ deviation and thumb size contributed more to esthetic outcome than to functional outcome (p<0.001). No significant interactions were found between any of the esthetic items, i.e., the impact of individual esthetic items on overall appearance did not depend on values of other esthetic items in the model (for an example of an interaction effect, see footnote IV).

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ii These correlations are indicators of regression model accuracy, i.e., how well the regression model predicts the observed data. Practically, you use the regression model to compute an *expected* set of outcome variables (e.g., overall esthetic outcome), which you than correlate with the same outcome variables *observed* in the dataset (i.e., 'reality'). The higher the correlation, the more accurate the regression model.

iii The *explained variance* is the proportion of variability of the overall outcome that can be predicted by the items included in the regression analysis. The higher the explained variance, the stronger the relation between the predictors and the outcome variable.

iv The *interaction term* is a combined item of active flexion and extension lag. The estimated effect of this combined item suggests that extension lag becomes a more influential determinant of overall functional outcome, when active flexion is also impaired.

Table 1: Patient characteristics and data used to build our proposed outcome assessment system.

PATIENT CHARACTERISTICS		
N total population	121	
Boy	63 (52%)	
Girl	59 (48%)	
N hands with radial polydactyly	131	
Wassel type II	50 (38%)	
Wassel type IV	63 (48%)	
Wassel type VII	18 (14%)	
	μ (SD)	Range
Age at follow-up*	9.7 (4.8)	4 – 30
Duration of follow-up*	9.7 (4.2)	1 – 22
Number of surgeries	1.2 (0.5)	1 – 6
OVERALL OUTCOME†		
VAS score for overall functional outcome	76.8 (18.5)	23 – 100
VAS score for overall esthetic outcome	72.3 (18.5)	13 – 98
FUNCTIONAL ITEMS‡		
IPJ alignment	8.3 (9.9)	0 – 50
MCPJ alignment	5.6 (8.8)	0 – 52
IPJ instability	4.3 (5.7)	0 – 28
MCPJ Instability	11.7 (7.6)	0 – 42
Active combined IPJ and MCPJ flexion	108.6 (29.9)	14 – 178
Combined IPJ and MCPJ extension lag	23.1 (22.6)	0 – 105
Palmar abduction	56.7 (8.9)	30 – 80
APPEARANCE ITEMS†		
VAS score for thumb size	78.6 (17.8)	12 – 100
VAS score for thumb pulp	77.0 (17.3)	9 – 99
VAS score for thumb nail	80.1 (17.4)	6 – 99
VAS score for scar appearance	79.6 (17.2)	9 – 99
VAS score for prominence at amputation site	75.5 (22.0)	11 – 99
PATIENT-REPORTED ITEMS§	N positive outcome	N negative outcome
Satisfaction with results	111 (85%)	20 (15%)
Pain of the thumb	101 (77%)	30 (23%)

IPJ = Interphalangeal Joint
MCPJ = Metacarpophalangeal Joint
* = in years
† = measured on a VAS ranging 0 – 100 points
‡ = measured in degrees
§ = reported as a "yes or no" answer.

Table 2: Regression coefficients of the various items predicting VAS score for overall functional outcome. The standardized regression coefficients were used to redistribute the points over the items in our proposed system.

REGRESSION ANALYSIS PREDICTING VAS FOR OVERALL FUNCTIONAL OUTCOME. $R^2 = 0.37$

	β	Sig.	Points in original JSSH system	Points in proposed system
AF	0.393	< 0.001	2	3
EL	-0.411	0.131	2	2
MCPJ instability	-0.085	0.300	2	2
IPJ instability	-0.059	0.452	2	2
Palmar abduction	-0.002	0.981	2	1
Interaction AF*EL	0.569	0.032		

β = standardized regression coefficient
Sig. = significance of the coefficient
JSSH = Japanese Society for Surgery of the Hand
AF = Combined Active IPJ and MCPJ Flexion
EL = Extension Lag
IPJ = Interphalangeal Joint
MCPJ = Metacarpophalangeal Joint.

Table 3: Regression coefficients of the various items predicting VAS score for overall esthetic outcome. The standardized regression coefficients were used to redistribute the points over the items in our proposed system.

REGRESSION ANALYSIS PREDICTING VAS FOR OVERALL ESTHETIC OUTCOME. $R^2 = 0.81$

	β	Sig.	Points in original JSSH system	Points in proposed system
VAS scar	0.326	< 0.001	2	3
VAS prominence	0.288	< 0.001	2	3
VAS pulp†	0.214	< 0.001	2	2
VAS size	0.213	< 0.001	2	2
IPJ deviation	-0.163	< 0.001	2	2
MCPJ deviation	-0.104	0.013	2	1
VAS nail†	0.070	0.165	2	1

β = standardized regression coefficient
Sig. = significance of the coefficient
JSSH = Japanese Society for Surgery of the Hand
IPJ = Interphalangeal Joint
MCPJ = Metacarpophalangeal Joint
† = Items combined in original JSSH assessment system.

Our proposed alternative outcome assessment system is shown in Figure 2. Based on the magnitude and statistical significance of the beta's, active flexion, scar appearance and prominence at the amputation site were given three points instead of two, whereas palmar abduction, MCPJ deviation and nail appearance were reduced to one point each. The other items were maintained at two points each, as in the JSSH system.

FUNCTION	Range	Points	APPEARANCE	Range	Points
Active IPJ + MCPJ flexion (°)	≥ 130	3	**Scar (VAS)**	≥ 95	3
	111 – 129	2		86 – 94	2
	91 – 110	1		71 – 85	1
	≤ 90	0		≤ 70	0
Extension lag (°)	≤ 20	2	**Prominence (VAS)**	≥ 95	3
	21 – 34	1		86 – 94	2
	≥ 35	0		66 – 85	1
				≤ 65	0
MCPJ instability (°)	≤ 20	2	**Size (VAS)**	≥ 90	2
	21 – 29	1		76 – 89	1
	≥ 30	0		≤ 75	0
IPJ instability (°)	≤ 5	2	**Pulp (VAS)**	≥ 85	2
	6 – 9	1		76 – 84	1
	≥ 10	0		≤ 75	0
Palmar abduction (°)	≥ 55	1	**Nail (VAS)**	≥ 85	1
	≤ 54	0		≤ 84	0
			IPJ deviation (°)	≤ 5	2
				6 – 14	1
				≥ 15	0
			MPJ deviation (°)	≤ 10	1
				≥ 11	0
PATIENT REPORTED					
Pain	Never	3	**Satisfaction**	Maximal	3
	When cold	2		Reasonable	2
	With use	1		Moderate	1
	Constant	0		Dissatisfied	0

Subtotal functional domain	/ 10 points
Subtotal appearance domain	/ 14 points
Subtotal patient reported domain	/ 6 points
Total outcome score	/ 30 points

Figure 2: Our proposed outcome assessment system based on clinical data from long-term follow-up studies.

IPJ = Interphalangeal Joint

MCPJ = Metacarpophalangeal Joint

° = in degrees.

With the exception of MCPJ instability, all cut-off values could be calculated based on the distribution of our follow-up data using quartiles, tertiles and medians. For MCPJ instability, most patients had values within the normal range (i.e. 0-20 degrees).[17] Therefore, MCPJ instability of 0-20 degrees was awarded two points, while the zero and one point cut-offs were based on the distribution of the remaining values.

Figures 3 and 4 show the distribution of outcomes in both the JSSH system and our proposed system. The overall outcome according to the JSSH was skewed, while the overall outcome according to our alternative system was symmetrical and approximately normal (μ=19.8, SD=5.1, p=0.194).

The internal consistency of our proposed system was Cronbach's α=0.64 versus Cronbach's α=0.56 for the JSSH system, while the interobserver agreement was ICC=0.57 versus ICC=0.65 respectively. Correlation between both systems was r_s=0.82. Correlation between the observed and predicted values was r_s=0.62 for the functional VAS scores and r_s=0.91 for the esthetic VAS scores.

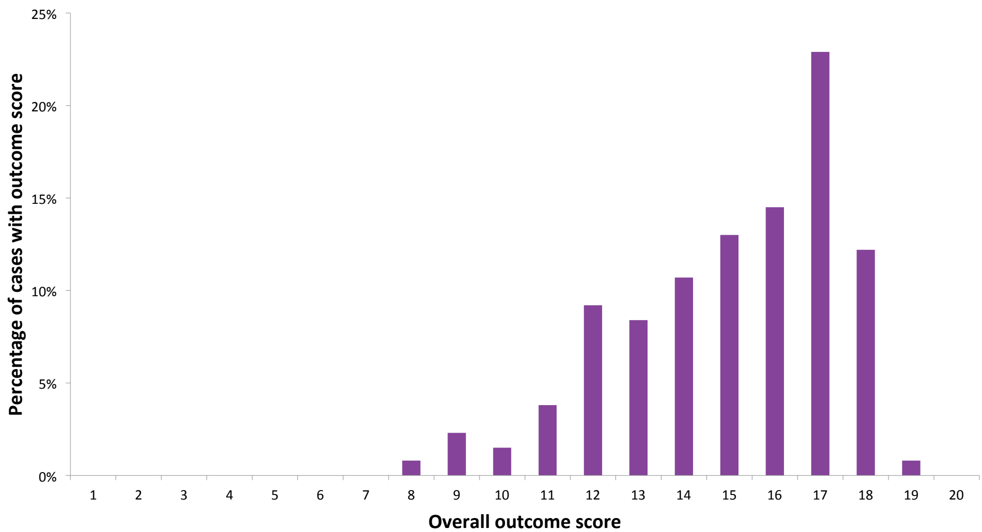

Figure 3: Distribution of overall outcome scores in the study sample using the JSSH outcome assessment system.

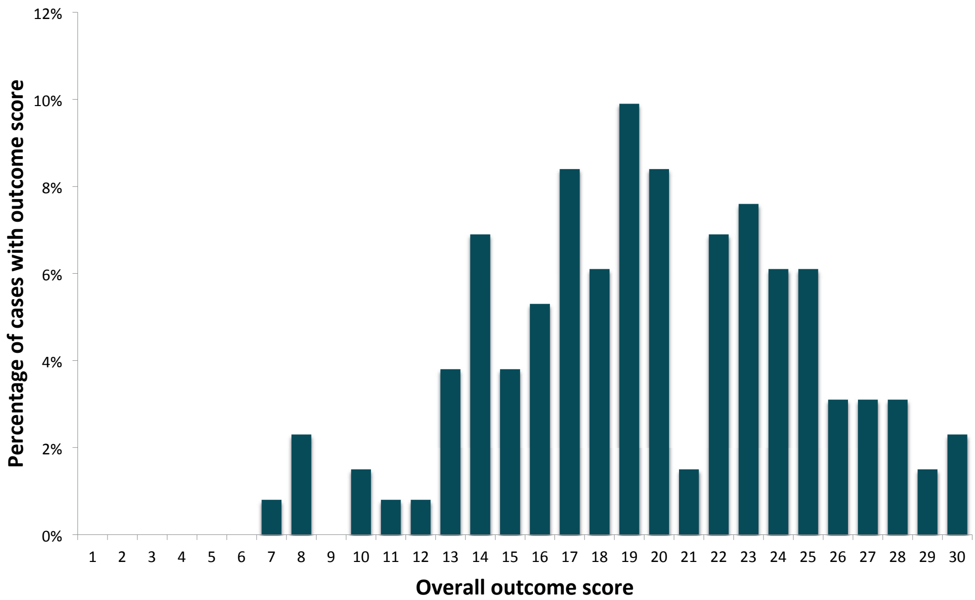

Figure 4: Distribution of overall outcome scores in the study sample using our proposed outcome assessment system.

DISCUSSION

In this study, a clinically weighted outcome assessment system for radial polydactyly was developed by optimizing the JSSH system, the most reliable and valid assessment system available,[9] using rigorous statistical analysis on follow-up data from two large congenital hand clinics.

We started by dividing the combined "nail/pulp" item from the JSSH system in two. Depending on radial polydactyly type and surgical technique, pulp or nail may show suboptimal results.[18-21] Our results show appearance of the pulp and nail affect overall appearance differently (Table 3), justifying separate evaluation of these items in our proposed system.

The appearance of the nail, scar, pulp, prominence and thumb size are assessed using a VAS in our proposed system, because subtle differences in these items are more likely to be detected by the VAS than by the dichotomized "good" or "poor" outcomes in the JSSH system. This could for example be relevant in evaluation of the pulp after a Bilhaut procedure versus the pulp after excision and reconstruction.[17] Such clinically appreciable differences are often difficult to quantify scientifically, which mandates use of a more sensitive scale.

The assessment of IPJ and MCPJ deviation as parameters affecting overall appearance rather than functional outcome is another fundamental change in our proposed system. Although tip-tip and lateral pinch were reported to affect overall hand function in a heterogeneous population of congenital hand differences,[22] we found deviation at the IPJ and MCPJ mainly affected the clinician-rated overall esthetic outcome (Tables 2 and 3). Therefore, these items were placed in the appearance domain, with a combined weight of three points.

The number of points per item was based on their weight in the regression analyses. Consequently, the points assigned to each item in our proposed system reflects how clinicians value the importance of that item as a determinant of overall outcome. Active flexion, scar, and prominence at the amputation site were the strongest determinants and were given three points, while palmar abduction, nail and MCPJ deviation appeared less important and were therefore given only one point in our alternative system. IPJ deviation, IPJ and MCPJ instability, extension lag and thumb size were given two points each, as in the JSSH system.

We also rescaled the cut-off values for all items in our alternative system, based on the distribution of our data. With exception of MCPJ instability, all cut-off values were supported by the literature on radial polydactyly.[5-8,12] Because most MCPJ instability values in our sample fell within a normal range, we rescaled this item by awarding two points to the normal value (i.e. < 20 degrees)[17] and placed the one-point cut-off value on the median of cases with > 20 degrees MCPJ instability (i.e. 30 degrees, Figure 2).

The scores for patient-reported items (satisfaction and pain) were changed from one out of 20 points each in the JSSH system, to three out of 30 points each in our proposed system. Regardless whether patient-reported outcomes constitute the 'gold standard' of post-operative evaluation, they are bound to influence the clinician's perception of outcome and decisions concerning revision surgery, warranting more weight and additional categories for these items.

The above-described weighting of patient-reported items is at odds with the general data-driven methodology of this study. Because patients may be dissatisfied with their thumb regardless of excellent function[23,24] analyzing patient-rated items as determinants of clinician-rated overall outcome would compromise valid conclusions on the effect of clinician-rated items. Ideally, patient-reported items would be weighted according to an 'overall patient reported outcome'. Unfortunately, to our knowledge, no such indicator exists for radial polydactyly. Although the weighting of the patient-reported items is a drawback of our proposed assessment system, the choice of clinically meaningful categories is an improvement over the existing JSSH system.

Use of VAS scores as proxy for overall functional[25] and esthetic[14] outcome may be unconventional. However, there are a number of arguments justifying their use in the present study. Ceiling effects[9,26] and poor correlation with functional parameters[9,27] (i.e. ROM, etc.) render manual ability questionnaires and manual ability tests unsuitable for the regression analyses in this study. Moreover, both VAS scores and the goniometer measurements are continuous scales, which facilitates interpretation. Nevertheless, our study shows the VAS is a poor construct to assess overall functional outcome, because little over one-third of its variability ($R^2 = 0.37$) could be predicted by generally accepted functional outcome parameters. Conversely, our results do support use of the VAS to assess esthetic results ($R^2 = 0.81$).

Restriction of the population to Wassel types II, IV and VII, and loss to follow-up may reduce the generalizability of this study. However, while the included types represent 78% of all radial polydactyly cases,[28] inclusion of cases with unaffected joints (i.e. types I, III and V) could have caused underestimation of joint related parameters. Moreover, our multicenter sample of patients and clinicians was of sufficient size to detect significant effects. Although a larger sample would allow for splitting the datasets into test and validation samples, the multicenter setting increases the generalizability of our results.[1,3-8,10,12-15,18-21,23,25,28-34]

This study provides unique insight into the way clinicians value functional, esthetic and patient reported factors as determinants of overall outcome leading to a clinically weighted assessment system. Our findings may also facilitate surgical decision-making. For example, since ROM proved a stronger determinant of postoperative functional outcome than MCPJ stability (β=0.393 versus β=-0.085 respectively, Table 2), surgeons faced with relatively hypoplastic type IV cases may be more inclined to resect one thumb and reconstruct the other (preserving ROM), rather than performing a Bilhaut procedure[21,35] sacrificing ROM for stability.

There are a number of other benefits to our clinically weighted system over the available arbitrarily constructed assessment systems for radial polydactyly.[5-8,10-12,25,32-34,36]. Since item weight and cut-off values represent clinical practice, there is less need for comparison with an unaffected contralateral hand,[10] which is particularly relevant in patients with bilateral involvement. [37,38] Moreover a 'normal' outcome (i.e., like an unaffected contralateral hand) is often unachievable due to preoperative problems e.g. reduced ROM and size.[15] While our proposed system and the JSSH system had similar internal consistency and interobserver reliability, outcomes were skewed towards the high range according the JSSH system, but normally distributed according to our proposed system (Figures 3 and 4). Consequently, our proposed system, 'the Rotterdam score', is less impaired by ceiling effects, and future researchers can report mean outcomes with standard deviations, which facilitates comparison and interpretation.

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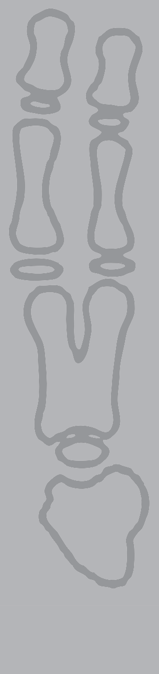
PART TWO: SURGICAL TREATMENT OF RADIAL POLYDACTYLY



CHAPTER 5

An international multicenter outcome study of radial polydactyly at the interphalangeal and metacarpophalangeal joint level

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ABSTRACT

BACKGROUND The aim of this international multicenter study was to investigate how patient characteristics and surgical technique influence outcome in radial polydactyly.

METHODS Patients with radial polydactyly types II, IV and IV with triphalangism ('IV-Tph') were evaluated using the validated Rotterdam score. Multiple determinants of outcome were studied, including polydactyly type, number of surgeries, age at operation, nail presentation, clinical deviation, tendon anomalies, surgical technique and surgeon experience.

RESULTS Of the 114 cases in this study, 94 (82%) were evaluated after single surgery with a mean follow-up of 7.8 years, and 20 (18%) cases were evaluated after multiple surgeries with a mean follow-up of 11.5 years. There were no significant differences in overall outcome between the specific types of radial polydactyly. Within the single surgery group, functional domain outcome was worse for type IV (1.3 points; p=0.005) compared to types II and IV-Tph. The multiple surgeries group had worse overall outcome (4.2 points; p<0.001), functional outcome (1.4 points; p=0.010), pain (1.1 points; p<0.001) and satisfaction (0.7 points; p=0.012) compared to the single surgery group. Tendon anomalies were more frequently treated in the worst outcomes. Overall outcome was better in patients primarily treated by dedicated congenital hand surgeons (2.7 points; p=0.014) compared to less specialized consultants.

CONCLUSIONS The most common types of radial polydactyly have similar overall outcome, although item- and domain-specific outcomes differ. Treatment of type IV remains challenging, especially in cases with tendon abnormalities, which are likely to influence outcome in a negative way. Treatment of radial polydactyly by dedicated congenital hand surgeons is associated with better outcome.

INTRODUCTION

The goal of surgical treatment of radial polydactyly is to create a single thumb of acceptable shape and size, which is stable but mobile enough to allow for unimpaired use and everyday functioning.[1] The variety of assessment systems exist to quantify achievement of this multifactorial goal, [2-11] are often designed for a particular study and rarely validated. These issues impede generalizable conclusions regarding effectiveness of surgical interventions in both scientific and clinical settings.

In addition to the challenges of valid outcome assessment, the sample heterogeneity in studies that describe outcome of all types of radial polydactyly[7,8,12-15] further impedes answering type-specific clinical questions. Of the studies that focus on specific types,[16-18] surgical techniques[19-21] or problems[22,23] in radial polydactyly, few demonstrate how patient characteristics and surgical techniques influence postoperative results. It would be beneficial to study results of surgery using a validated assessment system, in a homogenous but generalizable sample that covers the majority of radial polydactyly cases, to increase the evidence for type-specific approaches in radial polydactyly.

Therefore, we conducted an international, multicenter, long-term follow-up study to investigate how frequently-reported patient- and surgery-specific factors relate to outcome in patients with radial polydactyly at the interphalangeal (IPJ) or metacarpophalangeal (MCPJ) joint levels (i.e. types II, IV, and IV with triphalangeal components or 'IV-Tph' according to the Zuidam classification).[24] To evaluate outcome, we used an innovative, clinically-weighted and validated assessment system for radial polydactyly.[25] We studied the effect of surgery by comparing surgical procedures of our worst results to the surgical procedures of our whole sample. Subsequently, we explored specific relationships between patient characteristics, surgical technique, surgeon experience and outcome.

MATERIALS AND METHODS

This multicenter study was conducted at the Sophia Children's Hospital in Rotterdam, the Netherlands, and at the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. To create a fairly homogenous population, inclusion was limited to patients with radial polydactyly types II, IV and IV-Tph,[24] older than 4 years of age, and with a minimal follow-up of 2 years. Moreover, patients that were operated using the (modified) Bilhaut technique[26-29] and patients with unavailable operation reports were excluded from this study.

Patient-records were examined and radial polydactyly type (incl. aberrant components),[24] nail presentation (i.e. conjoined vs. separate), clinical deviation of the IPJ and MCPJ, tendon anomalies, age at operation and technical details of surgery were documented as potential determinants of postoperative outcome. In addition, we studied the association between having a dedicated (supervising) congenital hand surgeon, present during the procedure, and outcome. Dedicated congenital hand surgeons were defined as permanent members of either congenital hand team, in contrast to all other surgeons involved in the surgical procedures of our study population.

FUNCTION	Range	Points	**APPEARANCE**	Range	Points
Active IPJ + MCPJ flexion (°)	≥ 130	3	**Scar (VAS)**	≥ 95	3
	111 – 129	2		86 – 94	2
	91 – 110	1		71 – 85	1
	≤ 90	0		≤ 70	0
Extension lag (°)	≤ 20	2	**Prominence (VAS)**	≥ 95	3
	21 – 34	1		86 – 94	2
	≥ 35	0		66 – 85	1
				≤ 65	0
MCPJ instability (°)	≤ 20	2	**Size (VAS)**	≥ 90	2
	21 – 29	1		76 – 89	1
	≥ 30	0		≤ 75	0
IPJ instability (°)	≤ 5	2	**Pulp (VAS)**	≥ 85	2
	6 – 9	1		76 – 84	1
	≥ 10	0		≤ 75	0
Palmar abduction (°)	≥ 55	1	**Nail (VAS)**	≥ 85	1
	≤ 54	0		≤ 84	0
			IPJ deviation (°)	≤ 5	2
				6 – 14	1
				≥ 15	0
			MPJ deviation (°)	≤ 10	1
				≥ 11	0
PATIENT REPORTED					
Pain	Never	3	**Satisfaction**	Maximal	3
	When cold	2		Reasonable	2
	With use	1		Moderate	1
	Constant	0		Dissatisfied	0
Subtotal functional domain					/ 10 points
Subtotal appearance domain					/ 14 points
Subtotal patient reported domain					/ 6 points
Total outcome score					/ 30 points

Figure 1: Rotterdam score for radial polydactyly.

Outcome assessment

Overall-, domain-specific and item-specific outcomes were evaluated using the Rotterdam outcome assessment score for radial polydactyly (Figure 1).[25] Range of motion (ROM), instability and deviation of the IPJ and MCPJ were measured with a handheld goniometer, while palmar abduction was measured using the Pollexograph.[30] Appearance of the scar, residual prominence at the amputation site, thumb size, pulp, and nail were assessed using a VAS ranging from "extremely ugly" to "perfectly normal looking". In addition, patients were asked if they experienced any pain of the operated thumb and if they were satisfied with the overall results. A PhD candidate, not involved in the surgical treatment of the patients, gathered all outcome data.

Surgical technique

The standard surgical protocol used at the participating centers includes (1) removal of the more hypoplastic (usually) radial thumb via a dorso-lateral incision, (2) sparing the collateral ligament of the affected joint, (3) restoration of joint congruency by a longitudinal osteotomy or by shaping the joint surface, (4) closure of the joint capsule with repair of the collateral ligament to ensure joint stability, and (5) meticulous suturing of the skin (Figures 2-4).[1,31-34]

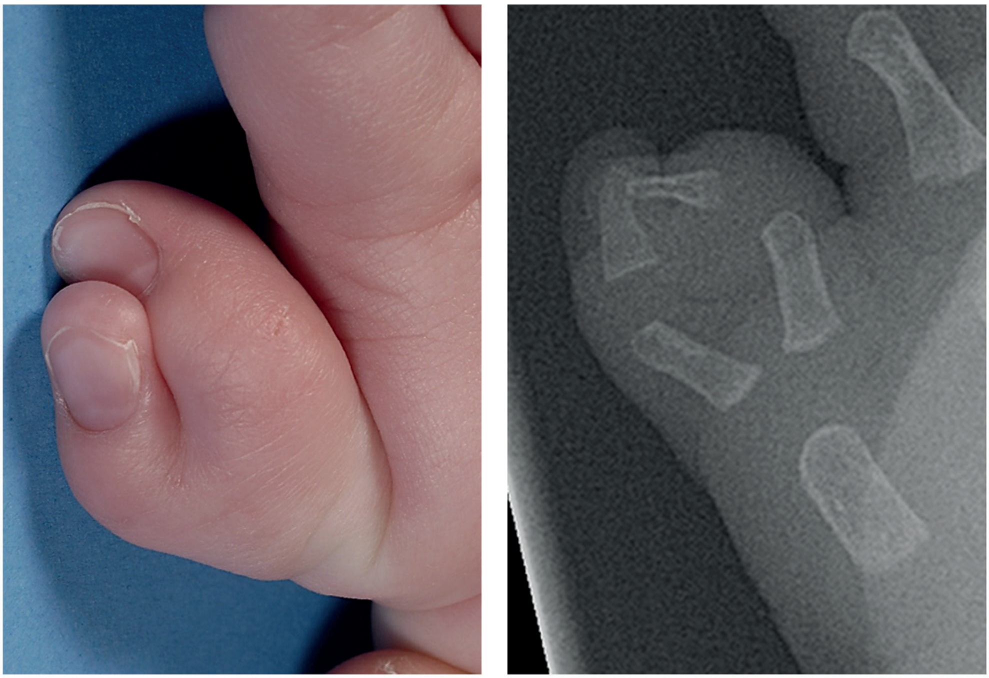

Figure 2: Preoperative situation in a 'Diamond shaped' radial polydactyly type IV-D r/u case.

Given specific circumstances, additionally indicated measures may be necessary to restore functional anatomy of the thumb. In case of abnormal tendon configurations, both flexor and extensor tendons may need to be centralized,[1,6,7,10,14,23,33-36] and aberrant tendinous interconnections devided.[1,14,33,35] A pulley reconstruction may be indicated in case of insufficient tendon sheath quality.[1,6,14,33] Significant deviation in the longitudinal axis may require corrective transverse osteotomies.[1,6-8,10,32-35] The available thenar musculature will need reattachment or transposition with the MCPJ collateral ligament.[1,3,6,14,15,32,33,35-37] Collateral ligament repair may require augmentation with a strip of volar plate or tendon graft from the resected thumb.[1,7,21,33]

The exceptional cases of radial polydactyly type II with a deltaphalangeal basal phalanx (54), may require bone grafting into an opening wedge osteotomy using bone from the removed thumb.[16] In the 'diamond shaped' type IV ('type IV-D r/u',[24] Figure 2), deviation at the IPJ level may need an osteotomy, and reefing of the joint capsule may help to further straighten the thumb and stabilize the IPJ.[4,6,34] If the triphalangeal counterpart is kept in type IV-Tph, surgical procedures were performed as described by Hovius et al.[38]

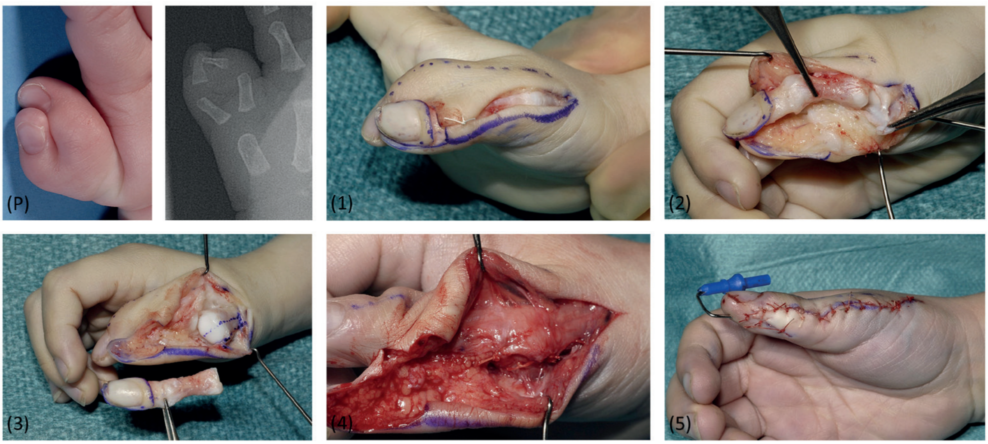

Figure 3: Standard surgical techniques for reconstruction of radial polydactyly.

(P) Preoperative situation

(1) Dorso-lateral incision over the radial thumb

(2) Sparing of the collateral ligament

(3) Markings for longitudinal osteotomy to restore joint congruency

(4) Closure of the joint capsule with repair of the collateral ligament to ensure joint stability

(5) Meticulous suturing of the skin.

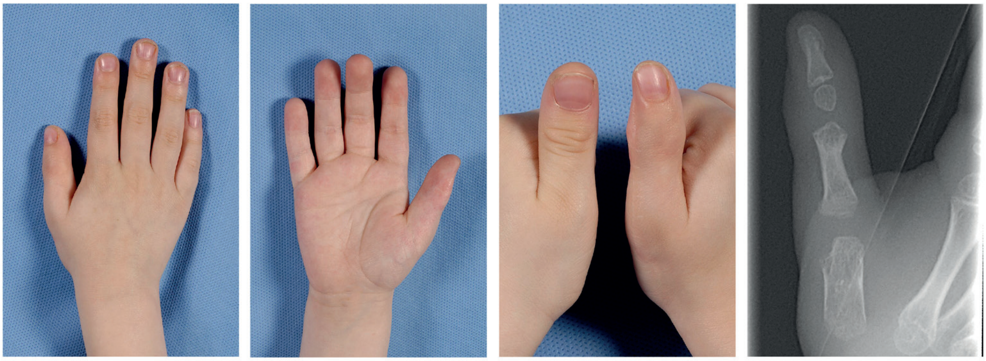

Figure 4: Postoperative situation of the case presented in Figures 2 and 3.

Statistical analysis

Throughout this study, we distinguished patients with outcome data after single surgery from patients with outcome data after multiple procedures in the treatment of radial polydactyly.

We investigated the relationship between primary surgical procedure and outcome, by examining the use of each before-mentioned surgical technique in our worst quartile (25%, or 'Q4') of outcomes, relative to their use in the entire sample. If this difference in use exceeded 10%, we studied mean differences in outcomes directly affected by that surgical technique in patients operated with and without its use.

Differences in continuous outcome parameters were analyzed using the Kruskal-Wallis and Mann-Whitney U tests, while Fischer's exact test was used for proportional outcome parameters, with statistical significance defined at $\alpha=0.05$.

RESULTS

Two-hundred-and-seventy-two patients were eligible for inclusion based on radial polydactyly type, age and duration of follow-up. Of these patients, 86 (32%) patients had changed address and could not be found, 51 (19%) were unwilling to participate, 2 (1%) had died and 5 (2%) had incomplete data. We further excluded 15 (5%) patients that were treated using the Bilhaut technique, and 4 (1%) because the operative report was missing. As a result, 109 patients (40%) or 114 cases could be analyzed in this study. Ninety-four (82%) of these 114 cases were evaluated after single surgery, while 20 (18%) had multiple procedures in their treatment of radial polydactyly (Table 1). Five (25%) of the multiple surgery cases were operated elsewhere prior to their referral.

Table 1: Patient characteristics of both the single surgery group and multiple surgeries group in our sample.

Single surgery group	Rotterdam	Hamburg	Total sample
	n (%)	n (%)	n (%)
Patients	43 (48%)	47 (52%)	90 (100%)
Girl	23 (53%)	21 (45%)	44 (49%)
Boy	20 (47%)	26 (55%)	46 (51%)
Cases	46 (49%)	48 (51%)	94 (100%)
Type II	23 (50%)	17 (35%)	40 (42%)
Type IV	18 (39%)	23 (48%)	41 (44%)
Type IV-Tph	5 (11%)	8 (17%)	13 (14%)
Bilateral CHD	8 (19%)	9 (19%)	17 (19%)
	μ (SD)	μ (SD)	μ (SD)
Age at surgery‡	2.1 (2.4)	1.3 (1.4)	1.7 (1.9)
Age at follow-up‡	8.9 (4.8)	9.3 (3.7)	9.1 (4.2)
Duration of follow-up‡	7.6 (4.6)	8.1 (3.8)	7.8 (4.2)

Multiple surgeries group	Rotterdam	Hamburg	Total sample
	n (%)	n (%)	n (%)
Patients	15 (80%)	4 (20%)	19 (100%)
Girl	10 (67%)	2 (50%)	12 (63%)
Boy	5 (33%)	2 (50%)	7 (37%)
Cases	16 (80%)	4 (20%)	20 (100%)
Type II	3 (19%)	1 (25%)	4 (20%)
Type IV	10 (62%)	2 (50%)	12 (60%)
Type IV-Tph	3 (19%)	1 (25%)	4 (20%)
Bilateral CHD	3 (19%)	2 (50%)	5 (25%)
	μ (SD)	μ (SD)	μ (SD)
Age at first surgery‡	1.5 (1.4)	0.5 (0.6)	1.3 (1.3)
Age at first revision surgery‡	6.3 (3.7)	3.8 (1.3)	5.8 (3.5)
Number of surgeries	2.4 (1.1)	2.0 (0.0)	2.3 (1.0)
Age at follow-up‡	12.1 (6.9)	15.0 (0.8)	12.7 (6.2)
Duration of follow-up‡	11.1 (7.3)	12.5 (2.1)	11.5 (6.5)

CHD = 'Congenital Hand Difference'
‡ = in years.

Table 2: Outcomes per radial polydactyly type, for both the single surgery group and multiple surgeries group in our sample.

	Single surgery group				Multiple surgeries group			
	Type II	**Type IV**	**Type IV-Tph**	**Overall**	**Type II**	**Type IV**	**Type IV-Tph**	**Overall**
	n = 40	**n = 41**	**n = 13**	**n = 94**	**n = 4**	**n = 12**	**n = 4**	**n = 20**
	µ (SD)	µ (SD)	µ (SD)	µ (SD)	µ (SD)	µ (SD)	µ (SD)	µ (SD)
Rotterdam score (0 – 30)†	21 (5)	21 (5)	19 (5)	21 (5)	19 (5)	15 (6)	20 (5)	17 (6)
Functional domain (0 – 10)†	8 (2)	6 (2)	7 (2)	7 (2)	7 (2)	5 (2)	7 (2)	6 (2)
†Patient rated domain (0 – 6)†	5 (2)	5 (1)	5 (2)	5 (2)	4 (3)	3 (2)	5 (2)	3 (2)
IPJ instability*	5 (5)	4 (5)	3 (3)	4 (5)	8 (9)	9 (9)	0 (0)	7 (8)
MCPJ instability*	11 (6)	14 (9)	12 (7)	12 (8)	8 (2)	15 (9)	9 (9)	13 (8)
Active flexion (IPJ+MCPJ)*	118 (25)	104 (24)	115 (38)	112 (27)	122 (24)	91 (25)	95 (49)	98 (31)
Extension lag (IPJ+MCPJ)*	16 (22)	27 (23)	28 (25)	23 (23)	22 (17)	32 (24)	24 (16)	28 (21)
Palmar abduction*	56 (9)	59 (7)	56 (9)	57 (8)	60 (7)	54 (10)	48 (13)	54 (10)
IPJ deviation*	8 (12)	6 (7)	9 (10)	7 (10)	12 (13)	12 (12)	6 (9)	11 (11)
MCPJ deviation*	0 (0)	10 (12)	13 (8)	6 (10)	0 (0)	10 (9)	9 (3)	8 (8)
VAS Scar†	83 (12)	84 (14)	82 (13)	83 (13)	80 (8)	73 (28)	78 (17)	75 (23)
VAS Prominence†	74 (23)	85 (16)	75 (18)	79 (20)	74 (21)	67 (22)	60 (34)	67 (24)
VAS Pulp†	79 (13)	80 (16)	82 (12)	80 (14)	79 (16)	79 (14)	84 (11)	80 (13)
VAS Nail†	81 (10)	88 (10)	87 (14)	85 (11)	79 (13)	88 (8)	88 (6)	86 (9)
VAS Size†	81 (15)	82 (15)	80 (12)	81 (15)	86 (15)	67 (31)	81 (13)	74 (26)
Satisfaction	85%	95%	85%	89%	75%	50%	100%	65%
Pain-free	80%	88%	77%	83%	50%	33%	75%	45%

IPJ = 'Interphalangeal joint'
MCPJ = 'Metacarpophalangeal joint'
VAS = 'Visual Analogue Scale, ranging 0 – 100'
† = in points
* = in degrees.

Overall outcome was similar between radial polydactyly types, both within the single surgery group ($p=0.489$), and within the multiple surgeries group ($p=0.201$).

Within the single surgery group, the functional domain score was significantly worse for type IV than for types II and IV-Tph (1.3 points; $p=0.005$). Furthermore, nail appearance was worse in type II than for types IV and IV-Tph (7 points; $p=0.005$) and prominence at amputation site was worse for types II and IV-Tph than for type IV (11 points; $p=0.032$). Types IV and IV-Tph had more MCPJ deviation (11 degrees; $p<0.001$) and extension lag (11 degrees; $p=0.030$) than type II.

Within the multiple surgeries group, types IV and IV-Tph had more MCPJ deviation than type II (9 degrees; $p=0.025$, Table 2).

The results in the multiple surgery group were significantly worse than in the single surgery group in terms of overall outcome (4.2 points; $p<0.001$), functional domain score (1.4 points; $p=0.010$), pain (1.1 points; $p<0.001$) and satisfaction (0.7 points; $p=0.012$,Table 2).

Surgical procedure and outcome

Regarding age at primary surgery, patients operated at an age older than 2 years had significantly better patient-reported domain scores (1.2 points; $p=0.036$) compared to patients operated before the age of 1 year. These differences were not found with regard to patients operated between ages 1-2 years. Furthermore, there were no significant differences in reoperation rate ($p=0.762$), overall outcome ($p=0.927$), or item-specific outcomes (p-values>0.10).

Five dedicated congenital hand surgeons carried out or supervised 86 of the 114 (75%) primary surgical procedures performed in our sample between 1984 and 2011. The remaining 28 (25%) primary surgical procedures were carried out by other hand surgeons and plastic surgeons. Both overall outcome and patient-reported outcome were significantly better (2.7 points; $p=0.014$ and 1.3 points; $p<0.001$) in patients who had been primarily operated by, or under supervision of one of the five dedicated congenital hand surgeons (Table 3).

Table 3: Patient characteristics and outcomes depending on presence of a dedicated congenital hand surgeon at primary surgery.

Patient Characteristics (n = 114)	DCHS operated	DCHS supervised	DCHS present	DCHS not present	Total sample
PATIENTS	63%	12%	75%	25%	100%
Type II	38%	43%	39%	39%	39%
Type IV	43%	57%	45%	50%	46%
Type IV-Tph	19%	0%	16%	11%	15%
Single surgery	88%	93%	88%	64%	83%
Multiple surgeries	12%	7%	12%	36%	17%
Age at first surgery < 1 year	14%	14%	14%	32%	18%
Age at first surgery 1 – 2 years	44%	72%	49%	39%	47%
Age at first surgery ≥ 2 years	42%	14%	37%	29%	35%
Aberrant components present	89%	71%	86%	64%	81%
Conjoined nail (type II only)	18%	7%	16%	29%	19%
Flexor tendon anomalies	31%	14%	28%	25%	27%
Extensor tendon anomalies	24%	7%	21%	32%	24%
	μ (SD)	μ (SD)	μ (SD)	μ (SD)	μ (SD)
Preoperative IPJ deviation*	27.4 (26.4)	13.3 (9.8)	25.0 (24.9)	0.0 (0.0)	25.0 (24.9)
Preoperative MCPJ deviation*	5.2 (11.6)	3.3 (10.0)	4.9 (11.2)	0.0 (0.0)	4.0 (10.4)
Age at first surgery‡	1.9 (2.1)	1.1 (0.7)	1.7 (1.9)	1.3 (1.5)	1.6 (1.8)
Age at first revision‡	6.8 (3.7)	2.0 (0.0)	6.3 (3.8)	5.4 (3.1)	5.9 (3.4)
Number of surgeries‡	1.1 (0.3)	1.4 (1.3)	1.2 (0.6)	1.4 (0.7)	1.2 (0.6)
Age at follow-up‡	8.7 (4.2)	10.0 (5.3)	8.9 (4.4)	11.8 (5.5)	9.6 (4.8)
Duration of follow-up‡	7.4 (4.1)	9.3 (5.6)	7.7 (4.4)	10.6 (5.6)	8.5 (4.9)
OUTCOME					
Rotterdam score (0 – 30)†	20.5 (5.5)	22.0 (4.2)	20.7 (5.3)	18.0 (4.3)	20.0 (5.2)
Functional domain (0 – 10)†	7.0 (2.0)	7.4 (2.1)	7.1 (2.0)	6.4 (2.1)	6.9 (2.0)
Appearance domain (0 – 14)†	8.3 (3.8)	9.5 (3.5)	8.5 (3.8)	7.8 (3.3)	8.3 (3.6)
Patient rated domain (0 – 6)†	5.2 (1.8)	5.1 (1.8)	5.2 (1.8)	3.9 (2.1)	4.8 (1.9)

DCHS = 'Dedicated Congenital Hand Surgeon'
IPJ = 'Interphalangeal joint'
MCPJ = 'Metacarpophalangeal joint'
† = in points
* = in degrees
‡ = in years.

In radial polydactyly type II, when comparing the primary surgical procedures in the worst quartile (25% of type II cases, or 'Q4') to the primary surgical procedures in all type II cases in the sample, we found that standard surgical protocol ('SSP') procedures 'longitudinal osteotomy

basal phalanx' (20%), 'IPJ collateral ligament repair' (13%), 'nail wall reconstruction' (18%) and additionally indicated measure ('AIM') 'extensor tendon centralization' (10%) were omitted more often in patients with poorer outcome ('Q4').
In radial polydactyly type IV, standard surgical protocol procedures 'longitudinal osteotomy first metacarpal' (10%) and 'thenar musculature reinsertion' (12%) were omitted more often in Q4, while additionally indicated measures 'IPJ collateral ligament repair' (17%), 'transverse osteotomy of the first phalanx' (21%), 'extensor tendon centralization' (12%) and 'pulley reconstruction' (12%) were done more often in Q4 to correct IPJ deviation, abnormal tendon insertions and deficient flexor tendon sheets, respectively.

With regard to radial polydactyly type IV-Tph, tendon abnormalities resulted in frequent use of additionally indicated measures 'flexor tendon centralization' (44%), 'extensor tendon centralization' (19%) and 'release of tendon adhesions' (17%) in the worst 25% of type IV-Tph cases (Table 4).

Table 5 shows the selection of above-mentioned surgical techniques, where the difference in item-specific outcomes between patients who were treated using these techniques, and those that were not, exceeded 10 degrees. For example, in type II cases where 'IPJ collateral ligament repair' was omitted on average had 10 degrees more postoperative IPJ deviation, compared to cases that were operated using this procedure from the standard surgical protocol. A more detailed accounting of these and other item-specific differences is shown in Appendix 1.

Table 4: Legend

pNII = Percentage of total type II cases in sample
pQ4II = Worst quartile (25%) of overall outcomes in type II cases
pNII – pQ4II = Percentile difference between use in total sample relative to use in worst quartile (25%) of outcomes in type II cases

pNIV = Percentage of total type IV cases in sample
pQ4IV = Worst quartile (25%) of overall outcomes in type IV cases
pNIV – pQ4IV = Percentile difference between use in total sample relative to use in worst quartile (25%) of outcomes in type IV cases

pNIVt = Percentage of total type IV-Tph cases in sample
pQ4IVt = Worst quartile (25%) of overall outcomes in type IV-Tph cases
pNIVt – pQ4IVt = Percentile difference between use in total sample relative to use in worst quartile (25%) of outcomes in type IV-Tph cases

IPJ = 'Interphalangeal joint'
MCPJ = 'Metacarpophalangeal joint'
n.a. = 'Not applicable'

Table 4: Documented primary surgery procedures per type of radial polydactyly, shown as the percentage of patients in which each procedure was performed.

	Single surgery group								
	Type II			Type IV			Type IV Tph		
	pN_{II}	$pQ4_{II}$	$pN_{II} - pQ4_{II}$	pN_{IV}	$pQ4_{IV}$	$pN_{IV} - pQ4_{IV}$	pN_{IVt}	$pQ4_{IVt}$	$pN_{IVt} - pQ4_{IVt}$
	n = 40	n = 10	%	n = 41	n = 14	%	n = 13	n = 4	%
STANDARD SURGICAL TECHNIQUES									
Longitudinal osteotomy basal phalanx	70%	50%	20%	7%	14%	7%	0%	0%	0%
Longitudinal osteotomy 1st metacarpal	n.a.	n.a.	n.a.	81%	71%	10%	69%	75%	6%
IPJ Collateral ligament repair	93%	80%	13%	12%	29%	17%	8%	0%	8%
MCPJ Collateral ligament repair	5%	0%	5%	95%	100%	5%	100%	100%	0%
Nail wall reconstruction	18%	0%	18%	n.a.	n.a.	n.a.	n.a.	n.a.	n.a.
ADDITIONALLY INDICATED TECHNIQUES									
Thenar musculature reinsertion	8%	10%	2%	76%	64%	12%	85%	100%	15%
Collateral ligament support	8%	0%	8%	12%	7%	5	15%	0%	15%
Transverse osteotomy 1st phalanx	35%	30%	5%	15%	36%	21%	8%	0%	8%
Transverse osteotomy metacarpal	n.a.	n.a.	n.a.	15%	21%	6%	54%	50%	4%
Flexor tendon centralization	20%	20%	0%	39%	36%	3%	31%	75%	44%
Extensor tendon centralization	20%	30%	10%	24%	36%	12%	31%	50%	19%
Tendon adhesiolysis	8%	10%	2%	5%	7%	2%	8%	25%	17%
Pulley reconstruction	5%	0%	5%	17%	29%	12%	0%	0%	0%
Fusion of P1 - P2 in type IV-Tph	3%	10%	7%	2%	7%	5%	8%	25%	17%
Fusion of P2 - P3 in type IV-Tph	n.a.	n.a.	n.a.	n.a.	n.a.	n.a.	31%	0%	31%
Bone graft	13%	10%	3%	5%	14%	9%	8%	0%	8%

Table 5: Selection of surgical techniques, where the difference in item-specific outcomes between patients who were treated using these techniques, and those who were not, exceeded 10 degrees.

	SP omitted / AI necessary	Directly affected outcome parameter	Outcome	Mean difference*
Type II (n = 40)				
Collateral ligament repair IPJ	SSP omitted (n = 3)	IPJ deviation	Worse	+ 10
Type IV (n = 41)				
Pulley reconstruction	AIM necessary (n = 7)	IPJ flexion	Worse	- 16
Type IV-Tph (n = 13)				
Extensor tendon centralization	AIM necessary (n = 4)	Extension lag	Worse	+24
Flexor tendon centralization	AIM necessary (n = 4)	IPJ flexion	Worse	- 28
		MCPJ flexion	Worse	- 12
Flexor-extensor tendon adhesiolysis	AIM necessary (n = 1)	IPJ flexion	Worse	- 50
		MCPJ flexion	Worse	- 11
		IPJ deviation	Better	- 10

IPJ = 'Interphalangeal joint'
MCPJ = 'Metacarpophalangeal joint'
SP = 'Standard Surgical Protocol procedure described in Methods section'
AI = 'Additionally Indicated Measure procedure described in Methods section'
* = in degrees

Preoperative patient characteristics and outcome

In radial polydactyly type II, transverse osteotomies were done for an average preoperative IPJ deviation of 33 degrees. Postoperative IPJ flexion was significantly diminished (23 degrees, p=0.026) in the deltaphalangeal or 'radially deviated' subtype[16] (Figure 5) compared to conventional type II. Furthermore, postoperative appearance of the nail, scar and pulp was similar in the conjoined and separate nail variants of type II radial polydactyly.

In radial polydactyly type IV an IV-Tph, transverse osteotomies were done for an average preoperative IPJ deviation of 26 degrees and for preoperative MCPJ deviation of 19 degrees. While patients with eccentric extensor tendon insertions had 9 degrees more postoperative extension lag, abnormal flexor tendon insertions significantly decreased IPJ flexion by 15 degrees (p=0.004). IPJ and MCPJ deviation was similar for patients with and without tendon anomalies. Finally, neither separate reinsertion of the thenar musculature, nor supporting the MCPJ collateral ligament with residual soft tissue significantly influenced MCPJ stability, MCPJ flexion or palmar abduction.

Although overall outcome was comparable for radial polydactyly cases with and without deviating, hypoplastic and triphalangeal osseous components, these aberrant components were more often operated by dedicated congenital hand surgeons (p=0.004) and were associated with a significantly lower reoperation rate (p=0.004) in our study.

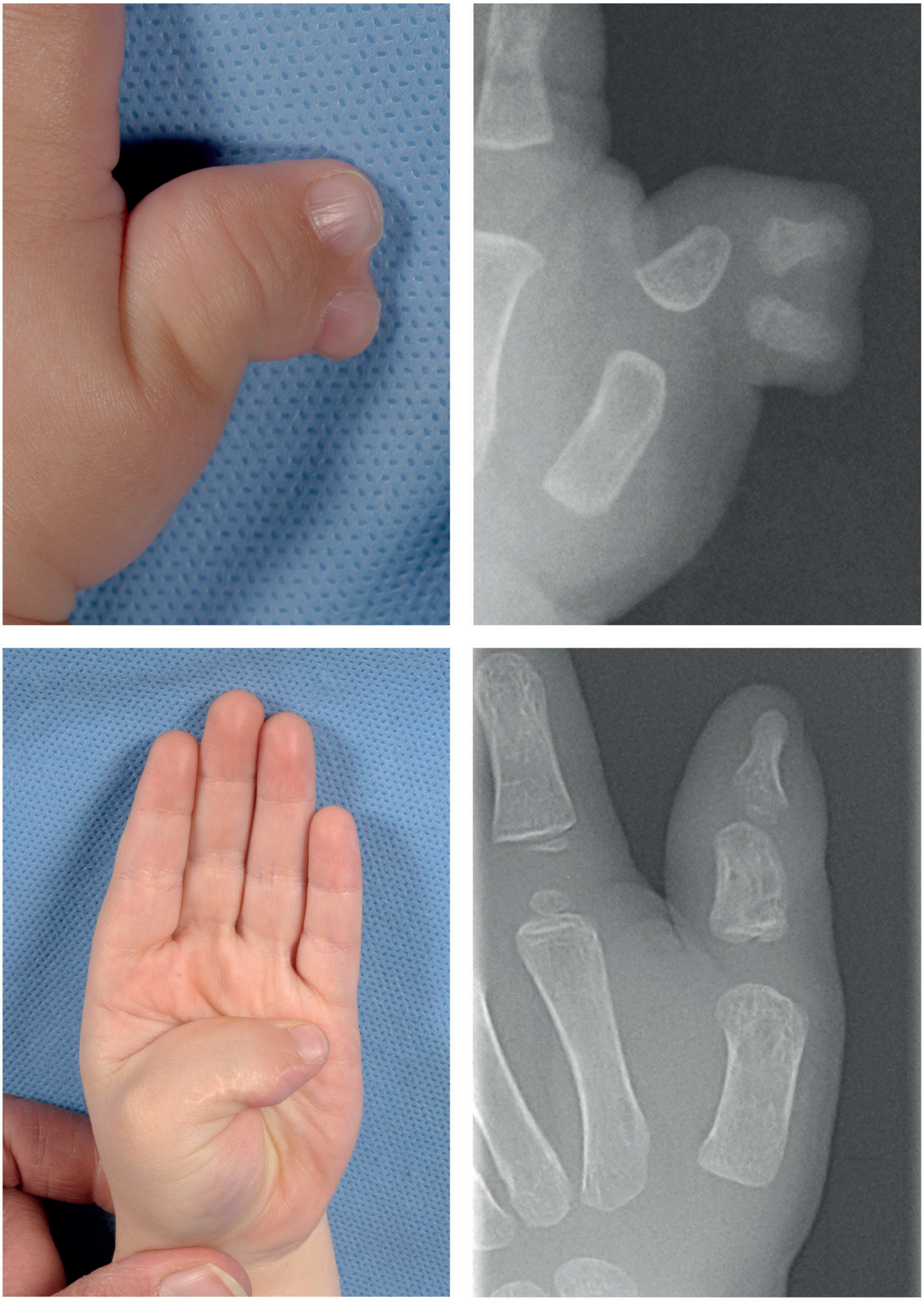

Figure 5: Preoperative (top) and postoperative (bottom) situation of a radially deviated radial polydactyly type II. Note the deltaphalangeal shape of the basal phalanx on the preoperative x-ray picture.

Reoperations

Twenty of the 114 cases (18%) required multiple surgeries to achieve acceptable outcome and were operated for a total of 35 amendable issues (Figure 6). Overall, the main reasons for reoperation were instability (34%) and deviation (17%). Absolute risks for developing deviation and instability following primary surgery, with a minimal follow-up of 1 year, are shown in Table 6. Deviation and instability together caused 67% of reoperations in type IV polydactyly. In the triphalangeal type IV-Tph, 25% of revision surgeries were done for reduced opposition and none for deviation. In type II, revision surgery was mainly carried out to improve appearance of the nail or nail wall (33%).

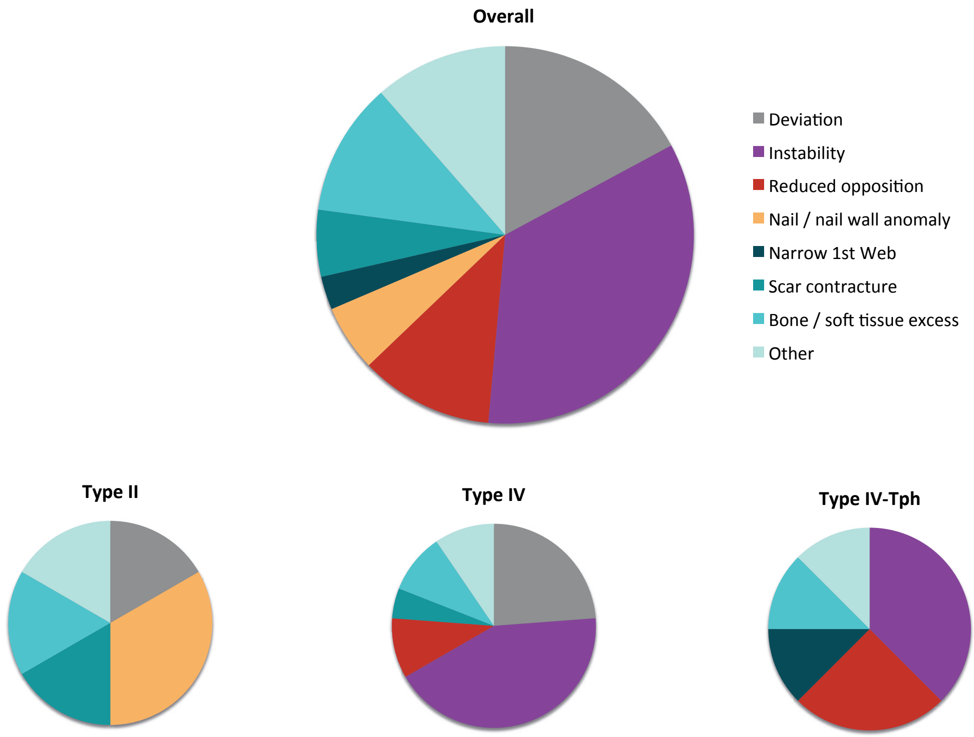

Figure 6: Amendable reasons for suboptimal outcome per type of radial polydactyly.

Table 6: Absolute risk of instability and deviation of the IP and MP joints following primary surgery and after a minimal follow-up of 1 year.

	10 – 20 degrees	**> 20 degrees**
Type II (n = 40)		
IPJ deviation	8 (20%)	4 (10%)
IPJ instability	6 (15%)	1 (3%)
Type IV (n = 41)		
IPJ deviation	13 (32%)	1 (2%)
IPJ instability	6 (15%)	1 (2%)
MCPJ deviation	9 (22%)	5 (12%)
MJ instability	23 (61%)	8 (19%)
Type IV-Tph (n = 13)		
IPJ deviation	5 (38%)	1 (8%)
IPJ instability	2 (15%)	0 (0%)
MCPJ deviation	9 (69%)	1 (8%)
MCPJ instability	9 (69%)	1 (8%)

DISCUSSION

This international, multicenter study shows no differences in overall outcome between the most common types of radial polydactyly, regardless whether evaluated after single surgery or multiple surgeries. However, on a domain-specific level, functional outcome was worse for type IV compared to types II and IV-Tph within the single surgery group. Focusing on item-specific outcome differences within the single surgery group, nail appearance was worse in type II, while MCPJ deviation and extension lag were significantly worse in types IV and IV-Tph (Table 2).

Patients with follow-up after multiple surgeries had significantly worse overall outcome, functional domain-specific outcome, patient satisfaction, and more frequently report pain, compared to patients with follow-up after single surgery. These findings correspond with other studies that suggest good outcome is difficult to achieve after a poor initial procedure[3,35,37,39] and underline the importance of a successful primary surgery (Table 2).

The results also show presence of a dedicated congenital hand surgeon during primary surgery is associated with significantly better overall and patient-reported outcome (Table 3). While experience seems an obvious prerequisite for the technically challenging early surgical correction of radial polydactyly,[3,8,14,19,40] delaying the surgery until growth facilitates the procedure has also been advocated.[7,12,31,36,37] However, since we found no difference in overall outcome between patients operated before the age of 1, between ages 1-2, or after the age of 2 years, we conclude experience in congenital hand surgery is a more critical determinant of outcome than age at primary surgery.

Closer analysis of surgical technique revealed poorer outcomes occurred more often when steps from the standard surgical protocol were omitted, and when additionally indicated measures were necessary to restore functional anatomy of the thumb. In type II for example, longitudinal osteotomies, collateral ligament repair, and nail wall reconstruction was often omitted in the 25% of cases with the worst outcomes (relative to all type II cases, Table 4). In types IV and IV-Tph, frequently performed correction of abnormal tendon configurations in the 25% worst outcomes suggests a negative effect of tendon anomalies (Table 4). Although ROM was often impaired in these cases, our results did not support the suspected negative effect[1,6,14,23,31,33-36,39] of eccentric tendon insertion on long-term postoperative joint deviation (Table 5).

Cases with deviating, hypoplastic, and triphalangeal components had favorable outcomes, presumably because they were mostly treated by dedicated congenital hand surgeons (Table 3). To communicate which cases require expert attention, stricter definition of deviating and hypoplastic components is necessary. Based on the deviation of MCPJ's in type IV cases requiring transverse osteotomy in our study, we suggest a minimal deviation of 20 degrees for classifying 'deviating components' (or 'D'). Consensus regarding classification of hypoplasia (or 'H') may be controversial, since a degree of hypoplasia is inherently associated with radial polydactyly. In cases with extra-capsular fibrous attachments or mere skin connecting the extra thumb to the dominant counterpart, collateral ligament repair may not be necessary, and classification as 'hypoplastic' seems justifiable.

Deviation and instability were the main reasons for revision surgery (Figure 6). It should be noted that the revision rate (n=20, 18%) includes five referred cases, who had had primary surgery elsewhere. However, both revision rate and reasons for revision correspond with other studies,[3-5,14,33-35,37,39] and referral of salvage cases to specialized centers represents clinical practice. Nevertheless, some patients in the single surgery group may require revision surgery later on, e.g., when the deviation becomes more apparent as the subcutaneous fatty tissue decreases with age.

Surgically, deviation and stability could be affected by collateral ligament repair, collateral ligament support, or osseous realignment. Since collateral ligament reconstruction was primarily performed in most cases (93%-100%), poor quality of collateral ligament and thenar musculature were possible factors in persisting instability. Regarding collateral ligament support, benefits of thenar musculature reinsertion to improve MCPJ stability[6,37] and palmar abduction,[15] and soft tissue augmentation to improve collateral ligament stability[1,21,33] could not be affirmed by evaluating the operative reports in our study. Regarding the osseous component of deviation and instability, transverse osteotomies should be considered as part of the standard surgical procedure, since they were only performed in 15-54% of cases (Table 4).

Cross-sectional outcome assessment and loss-to-follow-up are limitations of our study that may bias our results. A larger sample of prospective data would allow us to use parametric statistics to investigate causal relationships and to determine with more certainty which patient characteristics have little impact on outcome in radial polydactyly. These limitations notwithstanding, we were able to elucidate and quantify many clinically relevant associations between patient- and surgery-specific factors, and outcome, using non-parametric statistics in a cross-sectional design.

Moreover, the international, multicenter setting of this study, combined with the use of the validated and clinically-weighted Rotterdam outcome assessment score (Figure 1),[25] substantially increase the generalizability and validity of our results. Our findings are based on a large and homogenous study population,[3,4,10,11] with one of the longest follow-up durations in literature.[5,10,13,15,19] Consequently, the item-specific, domain-specific and overall outcomes in this study (Table 2) may be extrapolated to other clinics with a dedicated congenital hand surgery team, and are indicative for average expectable outcomes in the most common types of radial polydactyly.

In conclusion, this study shows experience in congenital hand surgery is associated with successful primary surgery, regardless of age at operation and case presentation. Successful extensive primary surgery is paramount, since this study shows patients requiring multiple surgeries to correct radial polydactyly have worse overall and functional outcomes, are less satisfied and more likely to experience pain. Deviation and instability are the main reasons for reoperation. While overall outcome is similar for the most common types of radial polydactyly, outcome may vary on an item- and domain-specific level. Successful treatment of type IV remains challenging, particularly since the congenital abnormalities of the tendon system, which occur more frequently in types IV and IV-Tph, are likely to influence outcome in a negative way.

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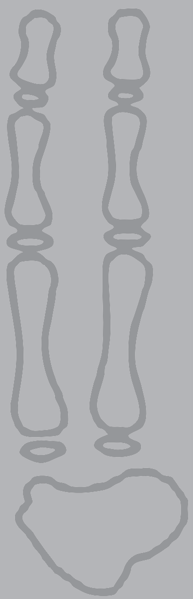
APPENDIX 1: Mean item-specific differences in outcomes directly affected by individual surgical techniques, per type of radial polydactyly.

Type II (n = 40)	SP/ AI	N_{II} (+)	N_{II} (-)	Use in $Q4_{II}$ – Use in N_{II}	Directly affected outcome parameter	Outcome	Mean difference
Longitudinal osteotomy IPJ	SP	28	12	-20%	IPJ deviation*	Worse	+ 4
					IPJ instability*	Worse	+ 5
					VAS size†	Worse	- 8
Collateral ligament repair IPJ*	SP	37	3	-13%	IPJ deviation	Worse	+ 10
					IPJ instability	Worse	+ 4
Nail wall reconstruction†	SP	7	33	-18%	VAS scar	Worse	- 4
Extensor tendon centralization*	AI	8	32	-10%	IPJ deviation	Worse	+ 1
					IPJ instability	Worse	+ 4
					Extension lag	Worse	+ 4

Type IV (n = 41)	SP/ AI	N_{IV} (+)	N_{IV} (-)	Use in $Q4_{IV}$ – Use in N_{IV}	Directly affected outcome parameter	Outcome	Mean difference
Longitudinal osteotomy MCPJ†	SP	33	8	- 10%	VAS prominence	Worse	- 5
Collateral ligament repair IPJ*	AI	5	36	+17%	IPJ deviation	Worse	+ 4
Thenar muscle reinsertion*	AI	31	10	- 12%	MCPJ instability	Same	0
Transverse osteotomy P1*	AI	6	35	+11%	IPJ deviation	Worse	+ 2
Extensor tendon centralization*	AI	10	31	+12%	IPJ deviation	Worse	+ 2
					IPJ instability	Worse	+ 2
					MCPJ deviation	Worse	+ 1
					MCPJ instability	Same	0
					Extension lag	Worse	+ 5
Pulley reconstruction*	AI	7	34	+12%	IPJ flexion	Worse	- 16
					MCPJ flexion	Worse	+ 5

Type IV-Tph (n = 13)	SP/ AI	N_{IVt} (+)	N_{IVt} (-)	Use in $Q4_{IVt}$ – Use in N_{IVt}	Directly affected outcome parameter	Outcome	Mean difference
Extensor tendon centralization*	AI	4	9	+44%	IPJ deviation	Worse	+3
					IPJ instability	Worse	+2
					MCPJ deviation	Better	- 1
					MCPJ instability	Worse	+1
					Extension lag	Worse	+24
Flexor tendon centralization*	AI	4	9	+19%	IPJ flexion	Worse	- 28
					MCPJ flexion	Worse	- 12
					IPJ deviation	Worse	+6
					IPJ instability	Same	0
					MCPJ deviation	Better	- 3
					MCPJ instability	Better	- 1
Flexor-extensor tendon adhesiolysis*	AI	1	12	+17%	IPJ flexion	Worse	- 50
					MCPJ flexion	Worse	- 11
					IPJ deviation	Better	- 10
					IPJ instability	Better	- 1
					MCPJ deviation	Worse	+4
					MCPJ instability	Same	0

IPJ = 'Interphalangeal joint'
MCPJ = Metacarpophalangeal joint'
SP = 'Standard Surgical Protocol procedure'
AI = 'Additionally Indicated Measure procedure'
VAS = 'Visual Analogue Scale, ranging 0 – 100'
$Q4_{II}$ = 'Quartile with worst overall outcome for radial polydactyly type II'
$Q4_{IV}$ = 'Quartile with worst overall outcome for radial polydactyly type IV'
$Q4_{IVt}$ = 'Quartile with worst overall outcome for radial polydactyly type IV-Tph'
N_{II} (+) = 'Total number of patients with radial polydactyly type II treated with this technique'
N_{II} (-) = 'Total number of patients with radial polydactyly type II treated without this technique'
N_{IV} (+) = 'Total number of patients with radial polydactyly type IV treated with this technique'
N_{IV} (-) = 'Total number of patients with radial polydactyly type IV treated without this technique'
N_{IVt} (+) = 'Total number of patients with radial polydactyly type IV-Tph treated with this technique'
N_{IVt} (-) = 'Total number of patients with radial polydactyly type IV-Tph treated without this technique'
* = in degrees
† = in points



CHAPTER 6

A matched comparative study of the Bilhaut procedure versus resection and reconstruction for treatment of radial polydactyly types II and IV

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ABSTRACT

BACKGROUND The aim of this study was to compare outcome of the Bilhaut procedure with outcome of conventional resection and reconstruction ('reconstruction') in radial polydactyly types II and IV.

METHODS Patients treated with the Bilhaut procedure were radiologically matched to patients treated with reconstruction. Evaluated outcomes included the Rotterdam score, pinch strength, and thumb size measurements. To objectify which esthetic outcome scores truly depend on surgical technique rather than observer opinion, evaluations by a blinded panel of 22 individuals with varying clinical experience were analyzed using a linear mixed regression model.

RESULTS The Bilhaut procedure reduced the risk of suboptimal outcome for MP joint instability in type IV radial polydactyly. Conversely, the Bilhaut procedure increased the risk of suboptimal scar appearance, residual prominence at amputation site, thumb size, and nail appearance. Tip-pinch strength was more significantly reduced after the Bilhaut compared to reconstruction, while pulp circumference and nail width exceeded 100% of the unaffected contralateral hand following a Bilhaut. There was no significant difference in active range of motion ('AROM') between both procedures. Nail appearance proved the only esthetic drawback of the Bilhaut procedure after adjustment for clinical experience.

CONCLUSIONS There is superior MP joint stability after the Bilhaut procedure for radial polydactyly type IV, but this does not result in the presumed benefit to thumb strength. In experienced hands, both procedures result in comparable thumb AROM. Nail appearance is the only esthetic aspect warranting specific attention when planning and performing a Bilhaut. Although a Bilhaut procedure that utilizes nail, soft tissue, and bone from both thumbs may not be worthwhile; there may still be indication for modifications that use the entire nail of one thumb.

INTRODUCTION

Surgeons specializing in congenital hand differences may apply a number of procedures in radial polydactyly patients in order to achieve a thumb of adequate mobility, stability, strength, and appearance.[1] In most cases, surgical treatment either consists of resecting the more hypoplastic thumb and reconstructing the more developed one ('reconstruction'),[2-4] or, combining relatively symmetrical parts of both thumbs into a single thumb: the 'Bilhaut procedure'.[5]

Of the two before-mentioned techniques, reconstruction is applicable in nearly all types of radial polydactyly and therefore most widely used.[6-8] Besides this versatility, reconstruction more likely preserves joint mobility. Drawbacks of reconstruction include the risk of joint instability and a smaller reconstructed thumb.[9-11] The Bilhaut procedure is classically indicated in cases were both thumbs are equal in size, but neither thumb seems sufficient for reconstruction on its own. Reported advantages of the Bilhaut procedure include joint stability, strength, and size, while joint stiffness, marked broadness, and conspicuous scarring of the nail bed and pulp are noted drawbacks.[12-14]

Numerous modifications to the original Bilhaut procedure[15-18] have been introduced to increase its applicability and ameliorate the above-mentioned drawbacks. Although these modifications have increased similarity between both surgical techniques and their indications, the key principles of the Bilhaut procedure remain unchanged. Despite the overlapping indications between two distinguishable treatments for radial polydactyly, there have been no comparative studies between the Bilhaut procedure and reconstruction.

The purpose of this matched study was to compare outcomes of the Bilhaut procedure to outcomes of reconstruction in patients with radial polydactyly types II and IV. Matching was done according to strictly prespecified preoperative radiological criteria. We evaluated the above-mentioned hypotheses regarding joint mobility, joint stability, thumb strength, and thumb size, and computed odds-ratios for suboptimal outcomes following both techniques. In addition to clinical evaluation, we compared esthetic outcomes, by presenting photographs to a blinded panel of experts, physicians, medical students, and lay people.

MATERIALS AND METHODS

This matched comparative study of the Bilhaut procedure versus reconstruction in treatment of radial polydactyly is nested within an international collaborative long-term follow-up project at the Sophia Children's Hospital in Rotterdam, the Netherlands, and at the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. Patients with a history of radial polydactyly types II and IV, a minimum age at follow-up of four years, and a minimum postoperative follow-up of one year, were included in this study. Patients without available x-rays and without operative reports were excluded. This study was approved by the Medical Research Ethics Committee (MEC-2010-295), and written informed consent was obtained for all participants.

Surgical procedure definitions and matching

We defined the Bilhaut procedure as surgery for radial polydactyly where parts of the nail, pulp, and bone from both thumbs are used to create a single thumb. If only one of the three before-mentioned elements from either thumb had not been utilized surgically, the procedure was considered too similar to the reconstruction technique to be labeled 'Bilhaut'. We defined reconstruction as a surgery for radial polydactyly where one entire thumb was preserved and reconstructed, with potential use of elements from the resected more hypoplastic counterpart.

The main matching criterion was preoperative osseous configuration, more specifically the level of duplication (i.e. type II and IV); degree of hypoplasia; and symmetry (Figure 1). Three observers (RD, CvN and SH) matched the preoperative x-rays of each Bilhaut case to four reconstruction cases until consensus was reached on optimal osseous similarity. Each Bilhaut case was matched to four reconstruction cases to optimize sample power.

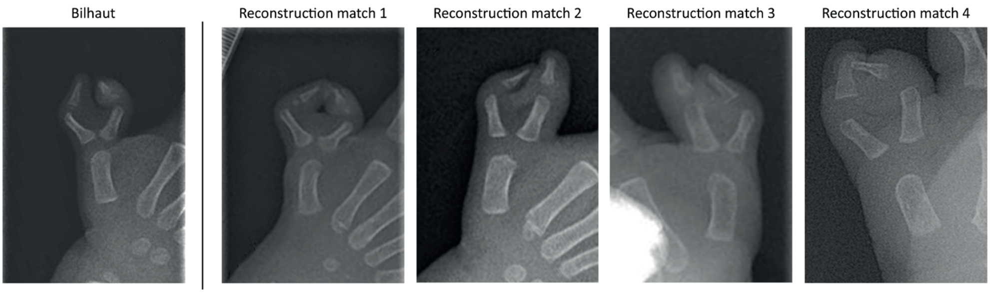

Figure 1: Example of matching based on osseous configuration.

Specialist outcome assessment

Outcome was evaluated using the Rotterdam outcome assessment score for radial polydactyly[19] (Figure 2), by a PhD-candidate (a physician) uninvolved in the treatment of the patients. Active range of motion (AROM), and deviation of the interphalangeal (IPJ) and metacarpophalangeal (MCPJ) joints were measured using a handheld goniometer. Instability was assessed by applying lateral stress to the joints. Palmar abduction was measured using the Pollexograph.[20]

Appearance of the scar, residual prominence at amputation site, thumb size, pulp, and nail, were separately quantified on a VAS ranging from "extremely ugly" to "perfectly normal looking". In addition, the same VAS was used by both clinician and patient to evaluate overall esthetic outcome. Furthermore, patients were asked if they experienced any pain and whether they were satisfied with the overall outcome.

FUNCTION	Range	Points	**APPEARANCE**	Range	Points
Active IPJ + MCPJ flexion (°)	≥ 130	3	**Scar (VAS)**	≥ 95	3
	111 – 129	2		86 – 94	2
	91 – 110	1		71 – 85	1
	≤ 90	0		≤ 70	0
Extension lag (°)	≤ 20	2	**Prominence (VAS)**	≥ 95	3
	21 – 34	1		86 – 94	2
	≥ 35	0		66 – 85	1
				≤ 65	0
MCPJ instability (°)	≤ 20	2	**Size (VAS)**	≥ 90	2
	21 – 29	1		76 – 89	1
	≥ 30	0		≤ 75	0
IPJ instability (°)	≤ 5	2	**Pulp (VAS)**	≥ 85	2
	6 – 9	1		76 – 84	1
	≥ 10	0		≤ 75	0
Palmar abduction (°)	≥ 55	1	**Nail (VAS)**	≥ 85	1
	≤ 54	0		≤ 84	0
			IPJ deviation (°)	≤ 5	2
				6 – 14	1
				≥ 15	0
			MPJ deviation (°)	≤ 10	1
				≥ 11	0
PATIENT REPORTED					
Pain	Never	3	**Satisfaction**	Maximal	3
	When cold	2		Reasonable	2
	With use	1		Moderate	1
	Constant	0		Dissatisfied	0

Subtotal functional domain	/ 10 points
Subtotal appearance domain	/ 14 points
Subtotal patient reported domain	/ 6 points
Total outcome score	/ 30 points

Figure 2: The Rotterdam outcome assessment score for radial polydactyly.

Tip-, tripod- and key-pinch strength were measured using a pinch strength dynamometer (Baseline; Fabrication Enterprises Inc., White Plains, New York). Each measurement was performed three times; the mean of these three maximum voluntary contractions was calculated to ensure measurement accuracy.

To evaluate thumb size, length was measured from the palpable base of the proximal phalanx to the tip of the thumb, while circumference was measured at the IPJ with the joint in neutral position. Pulp circumference was measured at the level of the lunula. Nail width was measured at the lateral edges, where the lateral nail folds meet the hyponychium. Thumb strength and thumb size measurements were compared to the unaffected contralateral hand.

Specialist, physician, medical student and laypersons esthetic outcome assessment

In addition to the above-mentioned outcome parameters, one hand surgeon specializing in congenital differences, eleven physicians, six medical students, and three laypersons independently assessed esthetic outcomes of the matched study population, using postoperative photographs (Figure 3).

All observers were blinded for the surgical intervention when using VAS scores to separately evaluate overall esthetic outcome, appearance of the scar, residual prominence at amputation site, thumb size, pulp, and nail. The VAS score anchors ranged from "extremely ugly" to "perfectly normal looking".

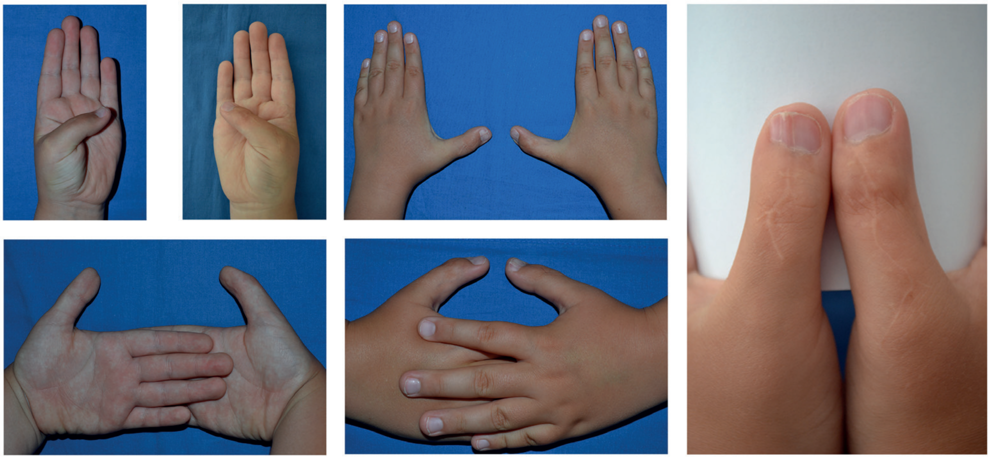

Figure 3: Example of photographs used to assess thumb appearance by a blinded panel of specialists, physicians, medical students, and laypeople.

Statistical analysis

To compare the continuous outcomes (i.e. Rotterdam score, VAS scores, thumb strength, and thumb size) between the matched Bilhaut and reconstruction cases, we used the Mann-Whitney U and Kruskal-Wallis tests, while categorical outcomes (i.e. satisfaction and pain) were compared using the chi-squared test.

To quantify the risk of suboptimal outcome after the Bilhaut procedure versus suboptimal outcome after reconstruction, we analyzed the matched data similar to a case-control study. Odds-ratios were calculated by dichotomizing results according to the Rotterdam score to optimal and suboptimal outcomes, and by defining both surgical techniques as exposures. In this case-control analysis, for all individual items, outcome was defined as suboptimal when it did not reach the maximum amount of points per item in the Rotterdam score.

To assess objectively which esthetic outcome scores (i.e. overall appearance, scar appearance, residual prominence at amputation site, thumb size, pulp, and nail) truly depend on surgical technique rather than observer opinion, we used a linear mixed regression model. In this model, procedure type was entered as a fixed effect, while adjusting for clinical experience (i.e. specialist, physician, medical student and layperson) and familiarity with results of similar cases (i.e. case number) as random effects.

RESULTS

One-hundred-and-sixteen cases from the collaborative long-term follow-up project met the inclusion criteria for radial polydactyly type, age, and duration of follow-up. Twenty-one (18%) cases were excluded due to missing or substandard x-rays, and four (3%) more due to missing operative reports. The remaining 91 (78%) cases contained eight (10%) Bilhaut procedure cases, and 83 (90%) reconstruction cases, prior to matching. We were able to match these eight Bilhaut cases (three type II (38%) and five type IV (62%) cases) to 32 reconstruction cases (12 type II (38%) and 20 type IV (62%) cases, Table 1).

Of the functional items in the Rotterdam score, MCPJ stability was 8 degrees better after the Bilhaut procedure compared to the matched reconstruction cases in type IV radial polydactyly ($p=0.019$), while the other functional items had similar outcomes for both techniques (Table 2).

Conversely, the esthetic items (i.e., scar appearance, thumb size, thumb pulp, and nail appearance) were significantly worse after the Bilhaut procedure than after reconstruction, with differences ranging 16–43 points ($p \leq 0.027$). Moreover, the Bilhaut procedure had a significantly worse appearance domain subscore (3.9 points; $p=0.002$) of the Rotterdam score, and a worse overall clinician-reported VAS score for overall appearance (18.7 points; $p=0.006$). The mean patient-reported VAS score for overall appearance was similar for both techniques.

The Bilhaut procedure reduced the risk of suboptimal outcome for MCPJ instability in type IV radial polydactyly (OR=0.8; 95% CI [0.6, 1.0], Table 3). Conversely, the Bilhaut procedure increased the risk of suboptimal scar appearance (OR=1.3; 95% CI [1.1, 1.6]), residual prominence at amputation site (OR=1.3; 95% CI [1.1, 1.6]), thumb size (OR=1.4; 95% CI [1.1, 1.7]), and nail appearance (OR=1.4; 95% CI [1.1, 1.9]).

Table 1: Patient characteristics of both the unmatched and matched sample of Bilhaut procedure and Resection and Reconstruction cases.

	Before matching	Study population		
	Unmatched Reconstruction	Matched Reconstruction	Bilhaut	Total
	n (%)	n (%)	n (%)	n (%)
Cases	83 (90%)	32 (80%)	8 (20%)	40 (100%)
Girl	39 (47%)	14 (44%)	4 (50%)	18 (45%)
Boy	44 (53%)	18 (56%)	4 (50%)	22 (55%)
Type II	29 (35%)	12 (38%)	3 (38%)	15 (38%)
Type IV	54 (65%)	20 (62%)	5 (62%)	25 (62%)
Affected hand				
Right	27 (33%)	22 (69%)	6 (75%)	28 (70%)
Left	56 (67%)	10 (31%)	2 (25%)	12 (30%)
Dominant	35 (42%)	12 (38%)	5 (63%)	17 (43%)
Bilateral CHD	22 (27%)	8 (25%)	3 (38%)	11 (28%)
Radial polydactyly	17 (20%)	6 (19%)	3 (38%)	9 (23%)
Outcome assessment after				
Primary surgery	71 (86%)	29 (91%)	7 (88%)	36 (90%)
Revision surgery	12 (14%)	3 (9%)	1 (12%)	4 (10%)
	μ (SD)	μ (SD)	μ (SD)	μ (SD)
Age at surgery†	1.5 (1.8)	1.4 (1.2)	1.0 (0.0)	1.3 (1.1)
Age at follow-up†	8.0 (3.6)	7.2 (2.8)	8.8 (3.2)	7.5 (2.9)
Duration of follow-up†	6.7 (3.3)	6.0 (2.8)	8.6 (3.1)	6.5 (3.0)
Number of surgeries	1.2 (0.4)	1.1 (0.3)	1.1 (0.4)	1.1 (0.3)

CHD = 'Congenital Hand Difference'
† = in years.

Table 2: Differences in outcome between the Bilhaut procedure and the Resection and Reconstruction technique.

	Bilhaut (n = 8)	**Reconstruction (n = 32)**		
	µ (SD)	µ (SD)	95% CI	p-value
Active IPJ + MCPJ flexion†	99 (46)	112 (25)	[-52, 25]	0.517
Extension lag†	21 (23)	25 (25)	[-25, 16]	0.753
IPJ instability†	3 (4)	4 (5)	[-6, 2]	0.539
MCPJ instability (type IV§)†	4 (5)	12 (6)	[-14, -2]	0.019*
Palmar abduction†	58 (6)	57 (8)	[-5, 7]	0.960
Scar††	64 (22)	80 (12)	[-27, -4]	0.027*
Prominence at amputation site††	70 (15)	77 (22)	[-24, 9]	0.095
Size††	67 (13)	82 (16)	[-27, -2]	0.010*
Pulp††	54 (29)	77 (16)	[-47, 1]	0.020*
Nail††	40 (26)	84 (11)	[-66, -22]	<0.001*
IPJ deviation†	10 (9)	8 (11)	[-6, 11]	0.377
MCPJ deviation (type IV§)†	7 (8)	7 (7)	[-8, 7]	0.717
Patient-reported pain‡	2.6 (1.1)	2.3 (1.3)	[-0.7, 1.3]	0.703
Patient-reported satisfaction‡	2.6 (1.1)	2.6 (1.1)	[-0.8, 0.8]	1.000
Rotterdam score‡	16.3 (4.3)	19.3 (4.8)	[-6.9, 0.7]	0.174
Functional domain score‡	7.1 (2.5)	6.9 (1.8)	[-1.3, 1.8]	0.778
Appearance domain score‡	4.3 (1.0)	8.2 (3.4)	[-5.3, -2.5]	0.002*
Patient-reported domain score‡	5.3 (2.1)	5.0 (1.8)	[-1.2, 1.8]	0.584
Overall esthetic outcome (CR)††	54 (19)	73 (21)	[-35, -2]	0.006*
Overall esthetic outcome (PR)††	80 (28)	74 (26)	[-15, 10]	0.415

CR = 'Clinician Rated'
PR = 'Patient Rated'
* = Significant difference between Bilhaut procedure and Resection and Reconstruction technique
§ = Bilhaut procedure n = 5; Resection and Reconstruction technique n = 20
† = in degrees
†† = measured using a VAS ranging 0 – 100 points
‡ = points in Rotterdam score.

Table 3: Risk of suboptimal outcome per item of the Rotterdam score.

	Bilhaut (n = 8)	Reconstruction (n = 32)	Risk of suboptimal outcome	
	n (%)	n (%)	Odds Ratio	95% CI
Active IPJ + MCPJ flexion	6 (75%)	22 (69%)	1.4	[0.2, 8.0]
Extension lag	3 (38%)	17 (53%)	0.5	[0.1, 2.6]
IPJ instability	1 (13%)	9 (28%)	0.4	[0.0, 3.4]
MCPJ instability (type IV§)	0 (0%)	1 (5%)	0.8	[0.6, 1.0]*
Palmar abduction	2 (25%)	10 (31%)	0.7	[0.1, 4.3]
Scar	8 (100%)	27 (84%)	1.3	[1.1, 1.6]*
Prominence at amputation site	8 (100%)	27 (84%)	1.3	[1.1, 1.6]*
Size	8 (100%)	21 (66%)	1.4	[1.1, 1.7]*
Pulp	6 (75%)	20 (63%)	1.8	[0.3, 10.4]
Nail	8 (100%)	17 (53%)	1.4	[1.1, 1.9]*
IPJ deviation	6 (75%)	15 (47%)	3.4	[0.6, 19.5]
MCPJ deviation (type IV§)	1 (20%)	4 (20%)	1.0	[0.1, 11.6]
Patient-reported pain	1 (13%)	7 (22%)	0.5	[0.1, 4.9]
Patient-reported satisfaction	1 (13%)	4 (13%)	1.0	[0.1, 10.4]

* = Significant difference between Bilhaut procedure and Resection and Reconstruction technique
§ = Bilhaut procedure n = 5; Resection and Reconstruction technique n = 20.

We found that tip-pinch was significantly weaker in the Bilhaut group (76% vs. 92% of the unaffected contralateral hand; p=0.024, Figure 4). Tripod- and key-pinch showed a similar trend. Regarding thumb size (Figure 5), pulp circumference was significantly larger after the Bilhaut procedure compared to reconstruction (113% vs. 91% of the unaffected contralateral hand; p=0.001), as was nail width (111% vs. 85% of the unaffected contralateral hand; p=0.004). IPJ circumference and thumb length were comparable for both surgical techniques.

The Bilhaut procedure had significantly poorer nail appearance according to all observer classes (differences ranging 18 – 26 points; p≤0.004), and significantly poorer pulp appearance according to laypersons (16 points; p=0.003), compared to reconstruction (Table 4).

After adjustment for clinical experience and familiarity of observers with results of similar cases, the Bilhaut procedure still showed significantly poorer nail appearance (β=-23.1; p<0.001) in the eyes of the entire blinded panel (Table 4). While the other esthetic outcomes differed significantly across observer classes for both techniques (p≤0.010), they all showed a negative trend with declining clinical experience of the observers (Table 4).

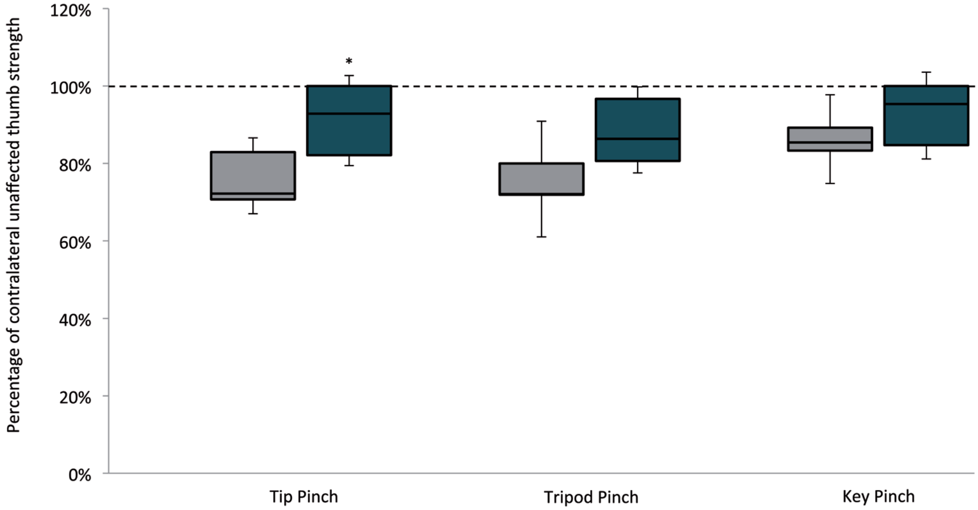

Figure 4: Thumb strength after the Bilhaut procedure versus Resection and Reconstruction cases, compared to the unaffected contralateral hand.

* = Significant difference between Bilhaut procedure and Resection and Reconstruction technique.

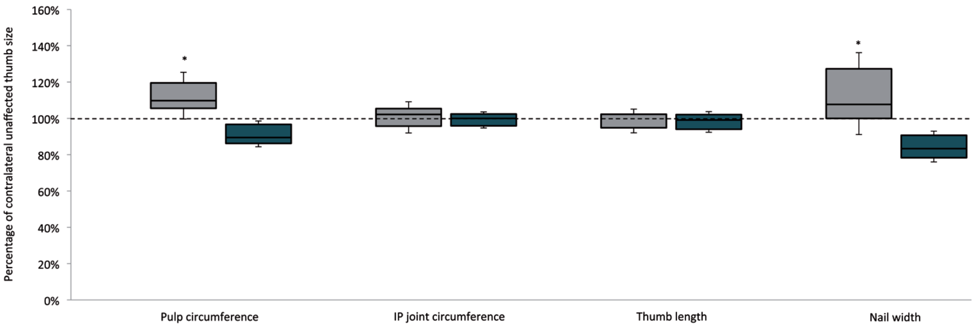

Figure 5: Thumb size after the Bilhaut procedure versus Resection and Reconstruction cases, compared to the unaffected contralateral hand.

* = Significant difference between Bilhaut procedure and Resection and Reconstruction technique.

Table 4: Mean VAS scores for overall esthetic outcome and appearance items of the Rotterdam assessment system, per observer class.

VAS score ranging 0 – 100 points	Specialists (n = 2)[§]		Physicians (n = 11)		Medical students (n = 6)		Lay persons (n = 3)		Overall (n = 22)		Linear mixed model analysis	
	μ (SD)		μ (SD)		μ (SD)		μ (SD)		μ (SD)		Effect of Bilhaut	
	Bilhaut	Reconstruction	Bilhaut	Reconstruction	Bilhaut	Reconstruction	Bilhaut	Reconstruction	Bilhaut	Reconstruction	β	P
Scar	75 (16)	81 (12)	60 (16)	67 (13)	53 (21)	63 (13)	59 (13)	63 (11)	60 (17)	67 (11)	-6.9	0.180
Prominence at amputation site	82 (10)	82 (16)	65 (11)	65 (12)	64 (16)	61 (15)	63 (7)	58 (13)	66 (10)	65 (11)	-0.46	0.920
Size	77 (9)	84 (11)	61 (10)	68 (11)	63 (13)	68 (11)	60 (5)	67 (10)	63 (9)	69 (10)	-7.5	0.065
Pulp	71 (18)	81 (12)	61 (13)	66 (9)	53 (13)	62 (11)	46 (12)*	64 (11)*	58 (12)	66 (9)	-4.9	0.216
Nail	59 (22)*	85 (10)*	46 (15)*	69 (13)*	43 (15)*	69 (14)*	46 (17)*	68 (12)*	46 (15)*	70 (12)*	-23.1†	<0.001†
Overall esthetic outcome	68 (15)	76 (19)	49 (15)	58 (16)	47 (15)	56 (17)	53 (9)	56 (11)	50 (14)	58 (15)	-8.9	0.133

§ = The specialist class consisted of one hand surgeon specialized in congenital hand differences and the principal investigator
* = Significant difference between Bilhaut procedure and Resection and Reconstruction technique within observer class ($P < 0.05$)
† = Significant difference between Bilhaut procedure and Resection and Reconstruction technique after adjustment for clinical experience ($P < 0.05$).

DISCUSSION

The aim of this matched study was to compare outcomes of the Bilhaut procedure to outcomes of resection and reconstruction ('reconstruction') in patients with radial polydactyly types II and IV. By applying strict prespecified criteria to define both surgical techniques and radiological similarity, we were able to create two comparable groups of patients and marginalize confounding by indication.

Our results support reported beneficial effects of the Bilhaut procedure on MCPJ stability in radial polydactyly type IV (Tables 2 and 3).[1,3,9-12,16,17] Furthermore, our results support reported unfavorable effects of the Bilhaut procedure on postoperative appearance of the thumb.[8,10,11,14,21-24] At clinical evaluation, scar appearance, residual prominence at amputation site, thumb size, and nail appearance showed increased risk of suboptimal results after the Bilhaut procedure.

Conversely, our results could not confirm beneficial effects of the Bilhaut procedure on IPJ stability,[1,3,9-12,16,17] or reported unfavorable effects on AROM of the IPJ and MCPJ.[3,6,8-12,14,16,17,21-25] Although our sample size may have been too small to detect these differences, the increased joint stability and AROM impairment following a Bilhaut procedure have only been reported in narrative reviews[1,3,10,11,22] and case series,[6,8,9,12,14,16,17,21,23-25] but have never been confirmed in a comparative study.

Thumb strength was less after the Bilhaut procedure compared with reconstruction (Figure 4). This suggests the statistically significant 8 degree-benefit in MCPJ stability after the Bilhaut procedure does not translate into a stronger thumb.[1,12,26] Since all patients had a minimum age at follow-up of four years, we found they were old enough to easily and accurately perform the strength measurements, which is supported by literature.[27,28] While thumb strength was consistently weaker for the Bilhaut procedure across all strength measurements, we were unable to match cases for preexistent soft tissue hypoplasia (due to the unavailability of criteria to quantify hypoplasia), which may entail some residual bias.

Pulp circumference and nail width were significantly larger after the Bilhaut procedure compared to reconstruction, while IPJ circumference and thumb length were similar for both surgical techniques (Figure 5). However, the lower VAS scores for thumb size (Table 2), the 40% increased risk of suboptimal size outcome (Table 3), and the disputable merit to thumb strength (Figure 3), indicate a larger thumb should not necessarily be considered a benefit of the Bilhaut procedure over reconstruction.[29]

Although most clinician-reported results in this study suggest the Bilhaut procedure is associated with poor esthetic outcomes (Tables 2 and 3), our linear mixed model analysis indicates only nail appearance is truly impaired by the Bilhaut procedure (by 23 points, Table 4), while the other appearance parameters are comparable for both techniques. By blinding 22 observers from surgical intervention, and by adjusting for their clinical experience and for familiarity with results of similar cases, we were able to objectify the effect of both surgical techniques on thumb appearance.

The small sample size resulting from our strict definition of both the operative techniques and radiological matching criteria is a limitation of this study. Radial polydactyly types II and IV were combined in the analysis to maintain power, except when comparing MCPJ instability, which is only warranted in type IV cases. While we found a number of clinically relevant differences between both procedures, a larger sample may have shown additional differences (e.g. in AROM), which this study was underpowered to detect. However, increasing sample size by applying more lenient definitions would only increase precision of our outcomes at the expense of comparability of the treatment groups, impeding the validity of the study.

The matched study-design is a considerable step forward from existing case-series.[6,8,9,12,14,16,17,21,23-25] By applying strict definitions of operative techniques, and radiologically matching four reconstruction cases to each Bilhaut procedure to optimize power, we attained two distinct and comparable treatment groups. The resulting comparison of both techniques affirmed benefits of the Bilhaut procedure to MCPJ stability,[1,3,9-12,16,17] thumb size,[1,3,6,9,11,12,16,26,30] and poor esthetic outcomes,[8,10,11,14,21-24] but refuted superior thumb strength[1,12,26] and poor AROM.[3,6,8-12,14,16,17,21-25]

Another strong feature of this study was the analysis of the matched data in a case-control fashion. By dichotomizing results according to the Rotterdam score to optimal and suboptimal outcomes, and by defining both surgical techniques as exposures, we were able to quantify risks of suboptimal outcomes for both procedures. This translates directly to clinical practice, since surgeons specializing in congenital hand differences may take the risks of suboptimal outcomes (Table 3) into account when counseling parents regarding indication for a Bilhaut procedure vs. reconstruction.

Moreover, the linear mixed model analysis of the effect of both surgical techniques on appearance helps putting esthetic preferences and surgical indication into perspective. Strong esthetic preferences of surgeons for one technique over the other (e.g. Tables 2 and 3)[6,14,22] could outweigh functional benefits in choice of treatment. However, considering the esthetic preferences of 22 observers with varying clinical experience, nail appearance was the only esthetic preference warranting specific attention during preoperative counseling and planning of the Bilhaut procedure.

All patients in our study had a classical indication for the Bilhaut procedure, i.e., two thumbs of similar size and shape, but neither sufficiently developed to be reconstructed on its own. Three out of five type IV Bilhaut cases had a 'diamond-shaped' presentation, and achieved better MCPJ stability compared to similar reconstruction cases. Therefore, angulation and slight size asymmetry of the thumbs do not preclude use of a (modified) Bilhaut procedure. Given the importance of nail appearance, we suggest only using modifications of the Bilhaut technique that aim to preserve the larger nail and its nail wall,[12,17,31] irrespective of nail size.[3,7,11,12,17]

Since multiple surgeons performed the Bilhaut procedure in our study and radial polydactyly is heterogeneous in presentation, minor technical variations were inevitable. However, all Bilhaut cases in this study were treated according to the same principles, utilizing nail, pulp, and bone from both thumbs. After careful alignment of the physes of both thumbs, joint congruency should always be assessed. In case of significant phalangeal size mismatch, joint congruency can be restored using osteotomies and by shaving the cartilage to correct any residual step-off. Our results suggest overcorrecting the width of the thumb, to ensure thumb size will match the unaffected contralateral thumb at a later age, may not be beneficial to postoperative appearance.

The results of this study underline greater MCPJ stability and thumb size may be achieved by using the Bilhaut procedure. However, these benefits do not translate into stronger thumbs, and esthetic results are more likely perceived as displeasing, even after adjustment for clinical experience of the observer. Therefore, a Bilhaut procedure that utilizes nail, soft tissue, and bone from both thumbs may not be worthwhile in cases where conventional reconstruction is possible. When attempting a Bilhaut-like procedure, effort should be made to respect the nail and nail wall. In addition, the risk estimates for suboptimal results associated with both procedures facilitate clinical decision-making and counseling parents.

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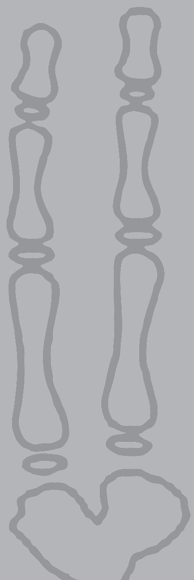
PART THREE: PATIENT-REPORTED OUTCOMES



CHAPTER 7

Thumb strength and manual ability in radial polydactyly types II and IV

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ABSTRACT

BACKGROUND The aims of this study were to quantify strength impairment in radial polydactyly type II and IV, to assess which factors affect thumbs strength in radial polydactyly, and to evaluate the impact of thumb strength impairment on manual ability in radial polydactyly patients.

METHODS Postoperative thumb strength was measured in patients with unilateral radial polydactyly type II and IV using a pinch gauge dynamometer and the Rotterdam Intrinsic Hand Myometer ('RIHM'). Strength of the unaffected contralateral side was used as the reference. Radial polydactyly type, hand dominance, thenar hypoplasia, number of surgeries, gender, age at follow-up, duration of follow-up, instability, and thumb size were evaluated as potential determinants of thumb strength. Manual ability was measured using the ABILHAND-kids questionnaire.

RESULTS In the 84 patients included in this study, thumb strength was impaired to 83% – 94% of the unaffected contralateral thumb in all measurements. Strength impairment was influenced by hand dominance, thumb size, and number of surgeries. Thumbs of radial polydactyly patients with an affected dominant hand were 10% – 23% stronger on average, compared to patients with an affected non-dominant hand, with exception of metacarpophalangeal joint flexion strength. Increased thumb size was positively correlated with pinch strengths, whereas increased number of surgeries showed a negative correlation. Forty-three (52%) patients attained the maximum score in the ABILHAND-kids questionnaire. While there was no correlation between relative thumb strength and manual ability, age at follow-up and duration of follow-up were correlated to manual ability.

CONCLUSIONS The relatively small but statistically significant strength impairment of radial polydactyly patients does not impair their manual ability. Strength is better in radial polydactyly cases with an affected dominant hand and in cases with relatively larger thumbs, while an increase in number of revision surgeries is associated greater strength impairment.

INTRODUCTION

Radial polydactyly is a relatively common Congenital Upper Limb Anomaly ('CULA') with a heterogeneous clinical presentation, affecting approximately 2.1 per 10.000 newborns.[1] It most often occurs at the metacarpophalangeal ('MCPJ') and interphalangeal ('IPJ') joint level (i.e., types IV and II, 40% and 16%, respectively),[2,3] and presents as a unilateral CULA in the majority of cases. However, up to 33% may be bilaterally affected, especially in combination with triphalangeal components.[4]

The most common surgical treatment for radial polydactyly consists of resecting the more hypoplastic radial thumb and reconstructing the better-developed, but inherently hypoplastic ulnar thumb. Outcomes are generally satisfactory in terms of thumb stability, mobility, and appearance.[5-7] However, residual hypoplasia of the reconstructed ulnar thumb tends to impair both postoperative thumb size and strength, although size is generally more amenable to surgical correction.[8-10]

While a number of studies have included strength measurements as part of outcome assessment, their findings have been inconsistent.[11-16] Furthermore, the strength measurements in radial polydactyly literature are limited to pinch strengths, and there is limited evidence on determinants of thumb strength in radial polydactyly, such as joint stability and hand dominance. Moreover, the clinical implications of thumb strength to manual ability of radial polydactyly patients have been subject to debate.[16,17]

The aims of this multicenter study were to quantify postoperative thumb strength in radial polydactyly, to assess which factors affect thumb strength, and to investigate the relationship between thumb strength and manual ability. Pinch and thenar musculature-specific strengths are measured in patients with unilateral radial polydactyly at the IPJ and MCPJ level (i.e., types II and IV). The impact of factors such as joint instability and dominance on thumb strength is assessed using univariate regression analysis. Furthermore, the relationship between thumb strength and manual ability is investigated using the ABILHAND-kids questionnaire.

MATERIALS AND METHODS

This international multicenter study was conducted at the Sophia Children's Hospital in Rotterdam, the Netherlands, and at the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. Patients with a history of radial polydactyly type II or IV, with a minimum age at follow-up of four years, and a minimum postoperative follow-up duration of one year, were eligible for inclusion. Patients with bilateral CULAs, triphalangeal components, and patients who had had a Bilhaut procedure or any opponens plasty were excluded to ensure a homogenous study sample. A physician uninvolved in the treatment of the patients gathered all data. This study was approved by the Medical Research Ethics Committee (MEC-2010-295) and written informed consent was obtained for all participants.

Strength measurements

Tip, tripod and key pinch strengths were measured using a hydraulic pinch dynamometer (Baseline, FEI, Irvington NY 10533, USA). The Rotterdam Intrinsic Hand Myometer ('RIHM')[18-20] was used to measure movement-specific strengths, i.e., thumb opposition, palmar abduction and MCPJ flexion strength. All measurements were performed in a standardized fashion: the pinch measurements in accordance with the ASHT guidelines,[21] the RIHM measurements using the RIHM protocol.[18] Each measurement was performed three times and the mean of these three measurements was used in the analysis.

Factors affecting thumb strength

A number of factors were hypothesized to affect thumb strength. Radial polydactyly type (i.e., II or IV), hand dominance, apparent thenar musculature hypoplasia, number of surgeries on the affected thumb, gender, age at follow-up, and duration of follow-up were all documented. Patients who were too young to determine hand dominance were considered right-handed, since more than 85% of the healthy population is right-handed.[22] In addition, instability of the IPJ and MCPJ were measured by applying lateral stress to the joint using a handheld goniometer. Thumb length was measured from the palpable base of the proximal phalanx to the tip of the thumb, and thumb circumference at the IPJ level with the joint in neutral position. Thumb size measurements were expressed relative to the unaffected contralateral side.

Manual ability

Manual ability was assessed using the ABILHAND-kids questionnaire.[23-25] The questionnaire measures the patient's capacity to perform 21 everyday tasks on a three-level scale: "easy", "difficult", or "impossible". Results of the questionnaire can be summarized using a logit score, representing the natural logarithm of the odds of successfully performing any task in the questionnaire (i.e., pass/fail ratio).[23] The logit score was calculated using an online tool designed for the ABILHAND-kids questionnaire.[26] Patients were stimulated to fill out the questionnaire by themselves, although children below the age of ten years tended to require help from their parents or caregivers.

Statistical analysis

The paired T-test was used to assess whether absolute strength of the affected thumb was significantly diminished compared to the unaffected contralateral thumb. Subsequently, thumb strength of the affected thumb was quantified as a percentage, using unaffected contralateral thumb as a reference.

Potential determinants of thumb strength were analyzed using univariate linear regression. In the univariate analysis, all six strength measurements (i.e., key-pinch, opposition etc.) were analyzed as dependent variables, and the abovementioned factors (i.e., radial polydactyly type, hand dominance etc.) as independent predictors. If a dichotomous predictor (e.g., hand dominance) variable consistently affected thumb strength across most univariate analyses, an additional analysis was done to compare impairment between patients with and without the predictor using Student's T-test.

The relationship between strength and manual ability (i.e., ABILHAND-kids logit score) was evaluated using Pearson's correlation coefficient

RESULTS

A total of 240 patients from the two participating centers were eligible for this study. Of the 240 eligible patients, 129 (54%) were lost-to-follow-up, 27 (11%) had to be excluded from the analysis, and 84 (35%) could be included. Of the 129 patients lost-to-follow-up, 77 (60%) had changed address and could not be reached, 50 (39%) did not want to participate in the study, and 2 (1%) had died. Of the 27 patients excluded patients, 13 (48%) had bilateral CULA, 11 (41%) had undergone a Bilhaut procedure, and 3 (11%) had had an opponens plasty. Of the 84 patients included in this study (Table 1), 83 (99%) had complete pinch strength data, 49 (59%) had complete RIHM strength data, and 82 (98%) had completed the ABILHAND-kids questionnaire.

Table 1: Patient Characteristics (n = 84).

Study population characteristics	
	n (%)
Patients	84 (100%)
Girl	41 (49%)
Boy	43 (51%)
Type II	35 (42%)
Type IV	49 (58%)
Hand dominance	
Left	16 (19%)
Right	64 (76%)
Not specified	4 (5%)
Dominant hand affected	38 (45%)
Thenar muscle hypoplasia	16 (19%)
	μ (SD)
Number of surgeries	1.2 (0.7)
Age at operation†	1.5 (1.8)
Age follow-up†	10.4 (5.0)
Duration follow-up†	9.1 (5.1)

† = in years.

Strength impairment

All thumb strength measurements in the affected thumbs were significantly reduced compared to the unaffected contralateral thumbs (p≤0.001, Figure 1). On average, tip, tripod, and key pinch strengths were significantly reduced to 94% (SD=15), 90% (SD=17), and 94% (SD=16) of the unaffected contralateral thumb, respectively. Strengths of opposition, palmar abduction, and MCPJ flexion, measured with the RIHM, were reduced even more, to 85% (SD=21), 83% (SD=26), and 88% (SD=24) of the unaffected contralateral thumb, respectively.

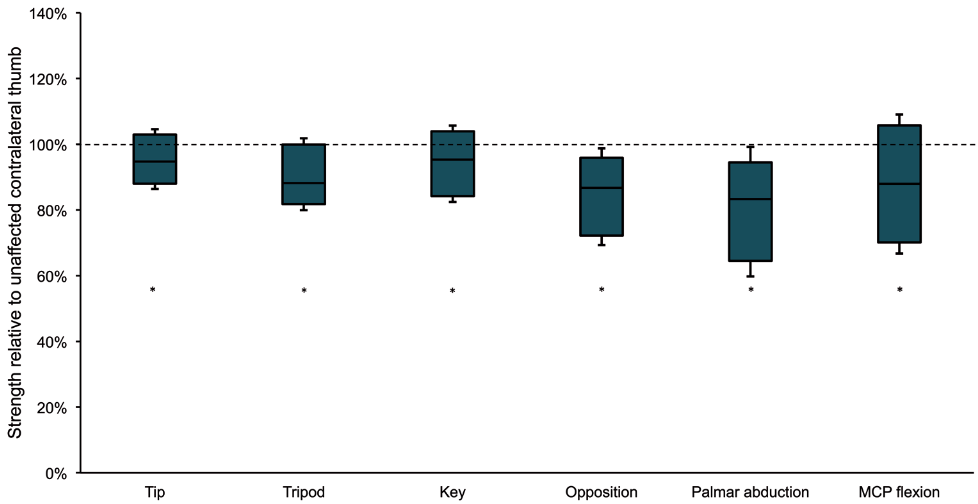

Figure 1: Strength of the affected thumb as the percentage of unaffected contralateral thumb strength.
* = significant difference (p < 0.05)

Factors affecting thumb strength

The univariate analysis revealed thumb strength impairment was influenced by whether the affected hand was the dominant hand, thumb size, and number of surgeries.

Radial polydactyly on the dominant hand was predictive for relatively stronger thumbs in all strength measurements (β ranging 9.7 – 22.8, p≤0.006) with exception of MCPJ flexion, which did not show this association (p=0.122). Thumbs of radial polydactyly cases with an affected dominant hand were 10% – 23% stronger, on average, compared to cases with an affected non-dominant hand (p≤0.006), with exception of MCPJ flexion strength (11%, p=0.122; Figure 2).

In addition, increased relative thumb length and circumference positively affected all pinch strength measurements (β ranging 0.6 – 0.8, p≤0.023). Conversely, increased number of surgeries negatively affected all pinch strength measurements (β ranging -5.2 – -8.8, p≤0.038). There were no significant differences in thumb strength between radial polydactyly types II and IV (Table 2).

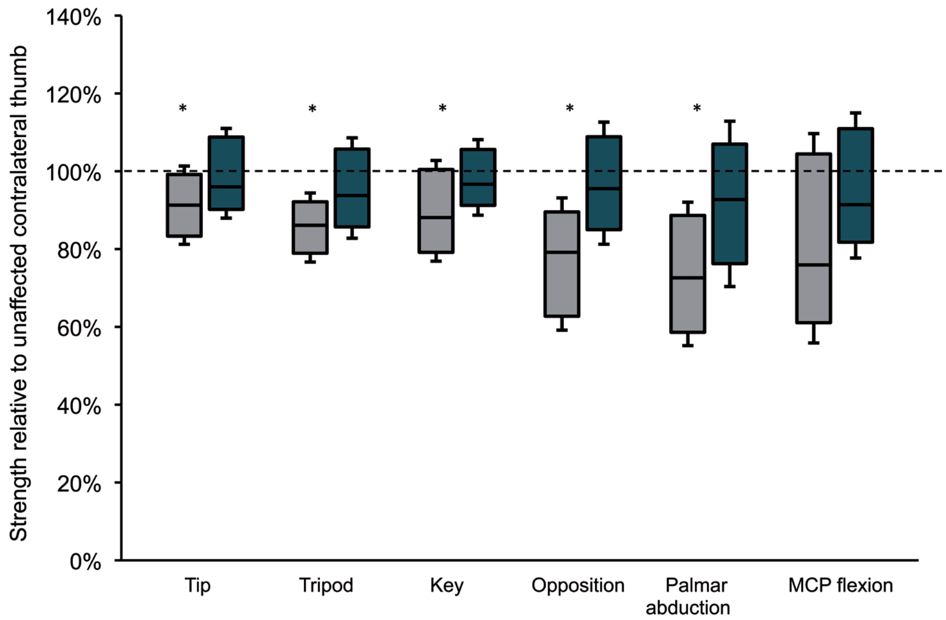

Figure 2: Thumbs strength of radial polydactyly cases on the non-dominant hand relative to strength impairment of radial polydactyly cases on the dominant hand.

* = significant difference ($p < 0.05$)

Table 2: Thumb strength relative to the unaffected contralateral side in radial polydactyly types II and IV. Note the absence of significant differences between both types.

	Type II n = 35	**Type IV n = 49**		
	µ % (SD)	µ % (SD)	95 % CI	p-value
Pinch measurements (n=83)†				
Tip pinch	96 (14)	92 (15)	[-2.5, 11.0]	0.224
Tripod pinch	93 (19)	87 (16)	[-2.5, 13.0]	0.180
Key pinch	97 (15)	92 (17)	[-2.4, 12.0]	0.188
RIHM measurements (n=49)‡				
Opposition	88 (21)	83 (20)	[-7.1, 17.0]	0.403
Palmar abduction	83 (23)	83 (27)	[-16.0, 15.0]	0.976
MCPJ flexion	96 (21)	83 (25)	[-0.7, 27.0]	0.061

† = in kg
‡ = in N.

Manual ability

Forty-three of the 82 (52%) patients who filled-out the ABILHAND-kids questionnaire attained the maximum score of 6.68 logits (median = 6.68, IQR[i] = 6.68 – 3.90; Figure 3). On average, 89% of the tasks in the questionnaire were considered 'easy' to perform, 9% were 'difficult', and 2% were 'impossible'. Of the top-three most difficult tasks to perform, two tasks mainly required bimanual dexterity (i.e., 'buttoning up trousers' and 'buttoning up a t-shirt or sweater'), and one task mainly required bimanual strength (i.e., 'opening a jar of jam').

There was no correlation between relative thumb strength and the ABILHAND-kids questionnaire score. However, absolute tip, tripod, and key pinch strengths did show as significant correlation (r ranging 0.44 – 0.53, $p<0.001$) with the ABILHAND-kids score. Furthermore, there was no relationship between ABILHAND-kids score and hand dominance ($p=0.881$), thumb length ($p=0.469$), thumb circumference ($p=0.459$), number of surgeries ($p=0.133$), or radial polydactyly type ($p=0.960$).

Of the other factors that were hypothesized to affect thumb strength, only age at follow-up and duration of follow-up were significantly correlated to the ABILHAND-kids score ($r=0.30$, $p=0.007$; and $r=0.32$, $p=0.004$, respectively). Patients who attained the maximum ABILHAND-kids score were 2.5 years older and had a 3.2 years longer follow-up, compared to patients who did not attain the maximum score ($p\leq0.020$).

i 'IQR' : Interquartile range equals the difference between the upper and lower quartile in a distribution of values, i.e., IQR = Q3 – Q1.

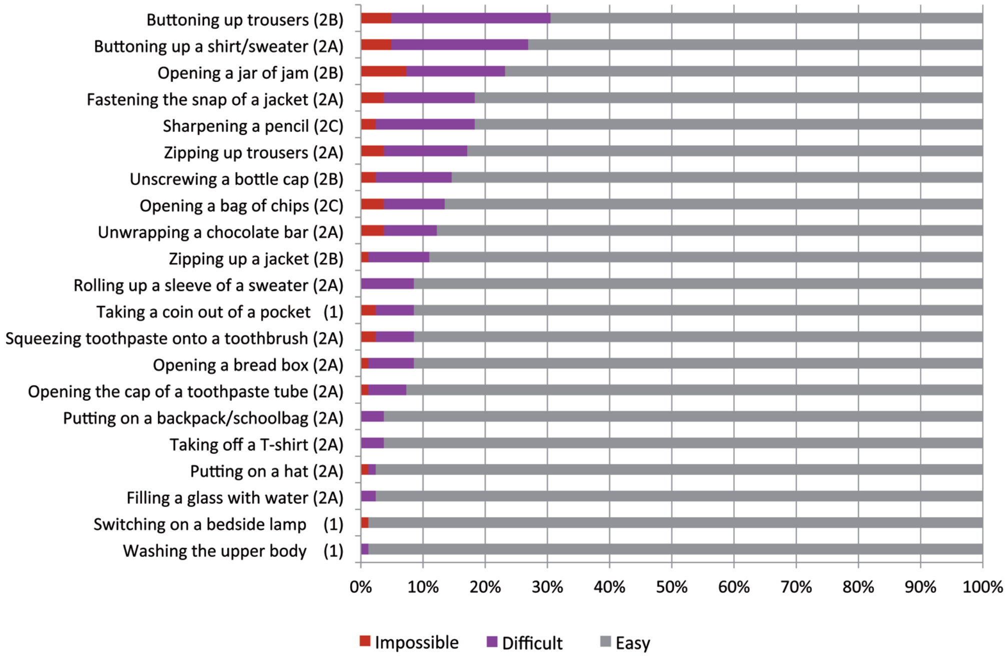

Figure 3: Results of ABILHAND-kids questionnaire.

1 = Unimanual activities

2A = Bimanual activities manageable with several unimanual steps

2B = Bimanual activities requiring stabilization with one hand and digital activity with the other

2C = Requiring digital activity from both hands.

DISCUSSION

The aims of this study were to quantify postoperative strength impairment in radial polydactyly type II and IV, to assess which factors affect postoperative thumb strength, and to evaluate the effect of impaired thumb strength on manual ability of radial polydactyly patients.

Thumb strength was significantly impaired compared to the unaffected contralateral thumb (Figure 1). On average, pinch strengths were reduced to 90% - 94%, while movement-specific strengths (i.e., opposition, palmar abduction and MCPJ flexion) were reduced to 83% - 88%. Although statistically significant, the findings in this study emphasize the minor strength impairment in radial polydactyly patients, [12,16,17] relative to strength impairment in other CULAs affecting the thumb such as triphalangeal thumbs[14] and thumb hypoplasia.[20,27]

The main factors affecting thumbs strength were hand dominance, thumb size, and number of surgeries. Thumbs of cases with an affected dominant hand were 10% – 23% stronger compared to cases where the non-dominant hand was affected (Figure 2). Since the majority of patients in this study were right hand dominant, this finding supports a similar effect of hand dominance on strength encountered in the general population.[28-31]

Relative thumb length and thumb circumference positively affected thumb strength (β ranging 0.6 - 0.8, $p \leq 0.023$), while number of surgeries showed a negative effect (β ranging -5.2 – -8.8, $p \leq 0.038$). These results indicate that thumb size should be taken into account at the primary surgery in order to optimize thumb strength. Postoperative thumb length and circumference are bound to be affected by preoperative hypoplasia, which may be indicative of coinciding hypoplasia of intrinsic and extrinsic thumb musculature. Therefore, surgically optimizing thumb size will not guarantee optimal thumb strength. For example, the Bilhaut procedure generally results in larger thumbs than the resection and reconstruction technique,[9,10] but a recent study showed thumbs are still weaker after a Bilhaut compared to resection and reconstruction.[32,33] The negative association between number of surgeries and thumb strength underlines the importance of successful primary surgery to prevent unnecessary revision procedures and poor outcomes. [5,6,34] At the primary surgery, all patho-anatomic differences should be corrected, especially those involving tendon insertion and alignment.

Other Other factors, such as radial polydactyly type, thenar musculature hypoplasia, gender, age at follow-up, duration of follow-up, IPJ and MCPJ instability, did not affect thumb strength. Contrary to findings in other studies,[11,12] thumb strength did not differ significantly between radial polydactyly types II and IV (Table 2). In addition, lack of clear definitions for thenar hypoplasia may have impeded valid documentation of this factor. This could explain why this study did not detect any effect of thenar hypoplasia on strength of thenar musculature functions (i.e., opposition, palmar abduction, and MCPJ flexion). Since the importance of collateral ligament reconstruction is widely appreciated, severe IPJ and MCPJ instability has become increasingly rare,[15-17,32] and did not affect thumb strength in this study.

When studying the relationship between manual ability and thumb strength in radial polydactyly patients, we found manual ability was not related to thumb strength, but was related to age at follow-up. Moreover, the majority of tasks that were rated as "impossible" or "difficult" required bimanual dexterity rather than strength (Figure 3). However, a minimum strength threshold may be necessary to easily perform all tasks in the questionnaire, given the significant correlation between absolute pinch strength and manual ability. Opponens plasty using abductor digiti minimi or flexor digitorum superficialis may improve both dexterity and strength in selected cases.

Incomplete data was the main limitation of this study. RIHM measurements of opposition, palmar abduction, and MCPJ flexion strengths were only available for 58% of the study population. This data was mainly missing for children younger than six years of age, which were often too distracted to get a reliable and valid reading. Consequently, there was insufficient RIHM data to support the positive effect of thumb size and the negative effect of number of surgeries on strength suggested by results of the pinch measurements. While normative data is available for both pinch [22,35,36] and RIHM strength measurements,[37] these data have a number of drawbacks. For one, the data is often restricted to specific age ranges (e.g., 4 – 12 year olds).[37] Furthermore, it has been suggested that normative data presents an overestimation of true strength,[36] and does not account for within patient differences.[12,17] Therefore, quantifying strength impairment as a percentage of the unaffected contralateral side may be considered a good alternative for comparison to reference values from medical literature.

Another limitation was use of the ABILHAND-kids questionnaire to measure manual ability. Being designed and validated for patients with Cerebral Palsy ('CP'),[23] the ABILHAND-kids questionnaire showed considerable ceiling effects, with over half the patients attaining the maximum score. While other measures of manual ability (such as the Assisting Hand Assessment etc.) are available,[38,39] these tests are very time-consuming and require specific expertise of hand therapists to administer. Moreover, the ceiling effects in the ABILHAND-kids questionnaire suggest most radial polydactyly patients experience only minor impairment in terms of manual ability. Therefore, it is unlikely more elaborate tests would lead to different findings.

Based on the results of this study, we conclude thumb strength may be significantly impaired in radial polydactyly from a statistical point of view, but this impairment does not entail clinically relevant reduction in manual ability. With respect to thumb strength, radial polydactyly patients with an affected dominant hand show favorable outcome. The relationship between thumb strength and thumb size is less straightforward, but larger thumbs are likely stronger. On the other hand, patients who need multiple surgeries in treatment of radial polydactyly may retain greater strength impairment. Therefore, the aim of primary surgery should be to correct all patho-anatomic differences, in order to prevent future revision surgeries. Since age is a more important determinant of manual ability than thumb strength in radial polydactyly patients, difficulties with performing everyday tasks are more likely related to dexterity than to strength. While some patients may experience trouble with tasks that require dexterity (e.g., buttoning their shirt) early on, radial polydactyly patients generally attain good to excellent manual ability.

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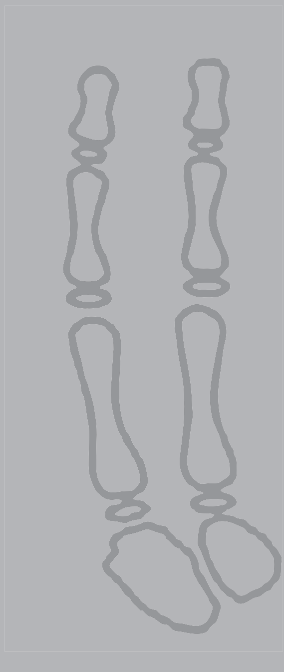

CHAPTER 8

The relationship between clinical evaluation, manual ability, participation, and quality of life in radial polydactyly patients

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ABSTRACT

BACKGROUND The aim of this study was to investigate the relationship between clinical assessment, manual ability, participation, and Health Related Quality of Life ('HRQoL') of patients with radial polydactyly types II, IV, and IV with triphalangeal components ('IV-Tph').

METHODS The Rotterdam system was used for clinical assessment. Patients were interviewed regarding satisfaction with thumb function and appearance, and filled out the ABILHAND-kids, CAPE, and HUI15Q questionnaires to quantify manual ability, participation, and HRQoL, respectively. Results were compared based on patient satisfaction with outcome, and correlation coefficients between the outcomes of clinical assessment, satisfaction, and the questionnaires were calculated.

RESULTS A total of 146 patients participated in this study, after a mean follow-up of 8.6 years. Patients rated their functional outcome better (95%CI 3 – 10; p=0.002) than, and the appearance of their thumb similar to clinicians. In terms of function, patients were mainly dissatisfied with impaired dexterity (n=3) and impaired strength (n=2). Regarding appearance, deviation (n=9) and a displeasing scar (n=4) were the main reasons for dissatisfaction. Manual ability was excellent for most patients, and related to patient-rated functional outcome (r_s=0.27). Patients who were dissatisfied with their outcome participated significantly less in 'Recreational activities' (95%CI 2.2 – 25.0; p=0.019), while 'Enjoyment' was positively correlated with patient-rated thumb appearance (r_s=0.43). The ABILHAND-kids score correlated with the 'Dexterity' attribute scores of the HUI15Q (r_s=0.21).

CONCLUSIONS While most radial polydactyly patients have excellent manual ability, participate in everyday activity, and enjoy good health, patient satisfaction with clinical outcome is related to these aspects. Primary surgery should aim to optimize dexterity, since manual ability is associated with HRQoL in radial polydactyly. Patient-rated thumb appearance is related to participation and enjoyment, which mandates consideration of revision surgery in radial polydactyly whose thumb appearance can be improved.

INTRODUCTION

Radial polydactyly is the most common congenital upper limb anomaly ('CULA') affecting the thumb, with a reported incidence of 2.1 per 10.000 live births in a Northern European population.[1] Surgical reconstruction of the thumb usually takes place before the age of two years. [2-4] Although there are many papers describing surgical technique and postoperative outcome, most studies are limited to assessment of thumb function and appearance from the surgeon's perspective. [5-9]

The literature that does touch on patient perception of outcomes is mainly focused on the 'body structures and function' domain of the International Classification of Functioning, Disability and Health ('ICF').[10] The World Health Organization ('WHO') introduced the ICF framework to conceptualize the interplay between body structures and function, activity, participation, and environmental factors to describe a person's functioning and health status (Figure 1).[11,12]

While literature describing postoperative 'body structures and function' is abundant, there is a paucity regarding postoperative manual ability, participation in everyday activities, and Health Related Quality of Life ('HRQoL') of radial polydactyly patients. Furthermore, the relationship between clinician- and patient-rated outcomes is unclear, and the relationship between outcomes in the other domains of the ICF framework is unknown for radial polydactyly patients.

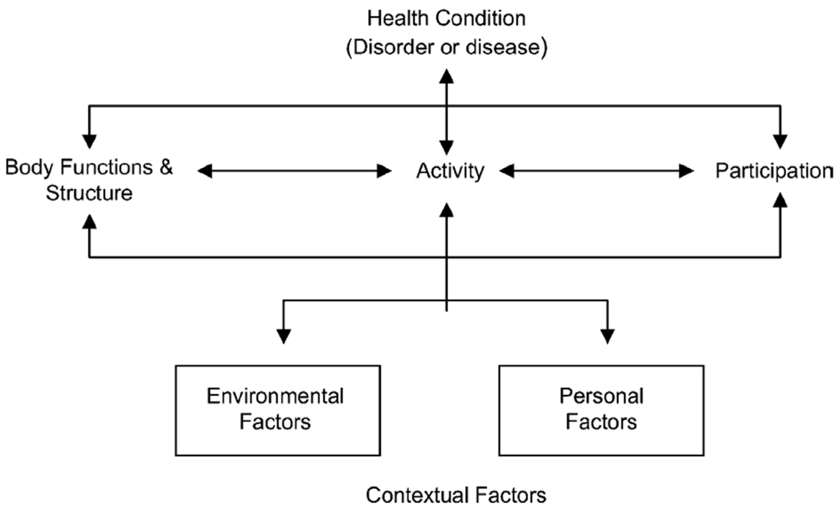

Figure 1: The ICF model of functioning, disability and health.

In this international multicenter study, the relationship between clinical assessment, manual ability, participation, and HRQoL of radial polydactyly patients is investigated, emphasizing on the patient perspective. Patients with radial polydactyly types II, IV, and IV with triphalangeal components ('IV-Tph') [13] were invited to the outpatient clinic for follow-up. In addition to clinical assessment, patient satisfaction was studied in depth, and patients were asked to fill out questionnaires designed to quantify manual ability, participation and HRQoL. The association between all domains of the ICF framework was studied by correlating the outcomes of clinical assessment, patient satisfaction, and questionnaires.

MATERIALS AND METHODS

This study was part of a multicenter long-term follow-up investigation at the Sophia Children's Hospital in Rotterdam, the Netherlands, and the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany. Between 2012 and 2015, all patients with radial polydactyly types II, IV, and IV-Tph, older than four years of age, and with a minimal postoperative follow-up of one year, where eligible for inclusion. Patient charts were reviewed in search of relevant medical history and comorbidities. A PhD candidate and a hand surgeon specialized in CULAs evaluated all patient during their visit to the outpatient clinic.

Clinical evaluation

The Rotterdam assessment score for radial polydactyly was used to quantify overall outcome. Combined active range of motion ('AROM') of the interphalangeal ('IPJ') and metacarpophalangeal ('MCPJ') joint were measured using a handheld goniometer, as were axial alignment and instability. Instability of the IPJ and MCPJ was assessed by applying lateral stress to the joints. Palmar abduction was measured using the Pollexograph.[14]

Subjective evaluation and interview

During the outpatient clinic visit, both the clinicians and the patients assessed overall functional and appearance of the thumb on a VAS with a range of 0 – 100 points. The functional VAS anchors were 'no functional thumb at all' to 'perfectly functioning thumb'. The overall appearance VAS anchors were 'extremely ugly' to 'looks perfectly normal'. In addition to the 'overall' VAS scores, the clinician also evaluated the appearance of thumb size, pulp, nail, scar, and prominence at amputation site on the abovementioned appearance VAS. Patients were also asked whether they were satisfied with their thumb function and appearance ('yes' or 'no'). Patients who were dissatisfied were asked which aspect of their thumb's function and/or appearance bothered them the most.

Manual ability, participation and HRQoL questionnaires

Manual ability was measured using the ABILHAND-kids questionnaire, a RASCH-built questionnaire that studies manual capacity on a three-point scale.[15] The questionnaire consists of 21 everyday tasks, and capacity is scored on a three-point scale ranging easy – difficult – impossible. The task-mix includes activities unimanual activities (1); bimanual activities manageable with several unimanual steps (2A); bimanual activities requiring stabilization with one hand and digital activity with the other (2B); and activities that requiring digital activity from both hands (3). By entering the answers into an online tool designed specifically for the ABILHAND-kids, a logit score is calculated.[16] This logit score represents the natural logarithm of the odds of successfully performing any task in the questionnaire (i.e., pass/fail ratio).

Participation was measured using the CAPE questionnaire.[17] The CAPE questionnaire consists of 55 everyday activities. It measures five dimensions of participation: diversity (i.e., participation in the activity in the past month), intensity (frequency of participation in an activity), with whom the child usually participates in the activity, where the activity usually takes place, and enjoyment (how much does the child like the activity?). The activities fall into one of five categories (i.e., recreational, physical, social, skill-based, and self-improvement activities), and into one of two domains (i.e., formal and informal activities). The results of the CAPE consist of separate scores for the different dimensions, activity categories, and activity domains in the questionnaire.

HRQoL was measured using the HUI15Q proxy-questionnaire, a validated generic questionnaire to measure HRQoL in children.[18] The HUI15Q consists of 15 questions that measure one of eight attributes: vision, hearing, speech, ambulation, dexterity, cognition, emotion, and pain. The answers to the questions translate into attribute scores. The attribute scores contribute to a composite 'multi-attribute' Health Utility Index (HUI) that represents a person's health state (ranging from death to perfect health), which is indicative of HRQoL. The Dutch version of the Mark-III formula was used to calculate the HUI in this study.

The questionnaire data was gathered during outpatient clinic visits or online for patients who were unable to visit the outpatient clinic but willing to participate. Patients were asked to fill out the questionnaires without help from their parents. If the child was to young to comply, parents were allowed to assist in answering the questions. Generally, patients older than 10 years of age were able to fill out the questionnaires by themselves.

Statistical analysis

The Wilcoxon matched-pair signed-rank test was used to compare differences between clinician-rated and patient-rated VAS scores for overall thumb function and appearance. The Mann-Whitney U and Kruskall-Wallis tests were used to compare clinical measurements and outcomes of questionnaires between patients who were satisfied it their results, and patients who were not. Spearman's correlation coefficient was used to express correlation between patient-rated and clinician-rated outcomes, and between results of the various questionnaires.

RESULTS

Between 2012 and 2015, 146 (53%) out of 274 eligible patients participated in this study. Of the patients lost-to-follow-up, 84 (66%) could not be contacted, 42 (33%) were unwilling to participate, and two (1%) had died. Data availability was variable for the participating patients: complete clinical follow-up data was available for 121 (83%) patients, while the ABILHAND-Kids, CAPE, and HUI15Q questionnaires were filled out for 138 (95%), 97 (66%), and 90 (62%) patients, respectively. Sixty-three (43%) patients had complete follow-up, with complete clinical and questionnaire data available (Table 1).

Table 1: Patient characteristics of both the entire study population and the complete follow-up population fraction.

	All participants	Clinical	AH-kids	CAPE	HUI15Q	Complete FU
	n (%)	n (%)	n (%)	n (%)	n (%)	n (%)
Patients	146 (100%)	121 (83%)	138 (95%)	97 (66%)	90 (62%)	63 (43%)
Girl	69 (47%)	62 (51%)	73 (47%)	53 (55%)	46 (51%)	34 (54%)
Boy	77 (53%)	59 (49%)	65 (53%)	44 (45%)	44 (49%)	29 (46%)
Cases	156	131	148	104	93	66
Type II	60 (39%)	49 (37%)	56 (38%)	38 (37%)	34 (36%)	24 (36%)
Type IV	75 (48%)	63 (48%)	71 (48%)	47 (45%)	50 (54%)	33 (50%)
Type IV-Tph	21 (13%)	19 (15%)	21 (14%)	19 (18%)	9 (10%)	9 (14%)
Comorbidity	47 (32%)	40 (33%)	45 (33%)	29 (30%)	23 (26%)	17 (27%)
Bilateral CHD	30 (21%)	26 (22%)	29 (21%)	19 (20%)	16 (18%)	12 (19%)
Dominant hand affected	66 (45%)	52 (43%)	61 (44%)	45 (46%)	39 (43%)	27 (43%)
Resection and reconstruction	139 (89%)	117 (89%)	132 (89%)	89 (86%)	83 (89%)	58 (88%)
Bilhaut	17 (11%)	14 (11%)	16 (11%)	15 (14%)	10 (11%)	8 (12%)
Satisfied with outcome	107 (72%)	102 (84%)	105 (76%)	76 (78%)	69 (77%)	56 (89%)
Dissatisfied with outcome	20 (14%)	19 (16%)	20 (14%)	13 (13%)	10 (11%)	7 (11%)
	μ (SD)	μ (SD)	μ (SD)	μ (SD)	μ (SD)	μ (SD)
Age at surgery‡	1.6 (2.1)	1.6 (1.8)	1.6 (2.1)	1.2 (1.2)	1.6 (1.8)	1.5 (1.3)
Age at follow-up‡	10.0 (4.9)	9.7 (4.8)	10.0 (4.9)	9.9 (4.5)	9.4 (4.8)	9.4 (5.0)
Duration of follow-up‡	8.6 (4.6)	8.5 (4.8)	8.6 (4.7)	8.6 (4.6)	8.0 (4.8)	8.2 (5.1)
Rotterdam score	19 (5)	19 (5)	19 (5)	20 (5)	20 (5)	21 (5)
VAS overall thumb function*	77 (18)	77 (18)	77 (18)	77 (19)	77 (18)	78 (19)
VAS overall thumb appearance*	73 (18)	73 (18)	73 (18)	75 (18)	73 (19)	76 (18)
VAS overall thumb function†	83 (15)	83 (14)	83 (15)	83 (15)	83 (15)	84 (15)
VAS overall thumb appearance†	72 (23)	72 (22)	72 (22)	76 (23)	74 (22)	78 (20)

CHD = 'Congenital Hand Difference'
‡ = in years.

Clinician-rated outcome and patient satisfaction

Nineteen (16%) of the 121 patients with complete clinical follow-up were dissatisfied with either thumb function (n=6), or thumb appearance (n=18), or both (n=5). These 19 patients had significantly worse outcomes in terms of Rotterdam score, and patient- and clinician-rated VAS scores, compared to patients who were satisfied with their outcomes (Table 2). In terms of functional deficit, patients were mainly dissatisfied due to impaired dexterity (n=3; 50%), and impaired strength (n=2; 33%). In terms of appearance, patients were mainly dissatisfied due to deviation of the thumb (n=9; 50%), and a displeasing scar (n=4; 22%).

Although patients on average rated their functional outcome significantly better than clinicians (95%CI 3 – 10; p=0.002, Table 1), there was no difference in how patients and clinicians rated appearance. The patient-rated VAS scores for function and for appearance were both positively correlated with the clinician-rated VAS scores (r_s=0.41 and r_s=0.28, respectively). Moreover, the Rotterdam score showed a positive correlation with the patient-rated VAS scores (r_s=0.36 and r_s=0.47, respectively).

Table 2: Overall outcomes of patients who were completely satisfied with their results, compared to patients who were dissatisfied with either functional or esthetic results of surgery.

	Satisfied (n = 102)	**Dissatisfied (n = 19)**		
	μ (SD)	μ (SD)	95% CI	p-value
Rotterdam score	20 (4)	14 (5)	[4.2, 8.5]	<0.001
VAS overall thumb function (CR)	79 (17)	64 (21)	[6.4, 23.8]	0.001
VAS overall thumb appearance (CR)	75 (17)	62 (21)	[4.2, 21.7]	0.004
VAS overall thumb function (PR)	85 (12)	71 (19)	[4.9, 23.7]	0.005
VAS overall thumb appearance (PR)	78 (17)	38 (19)	[31.6, 48.9]	<0.001

CR = Clinician-rated
PR = Patient-rated.

Patient-reported outcomes

Of the 138 patients who filled out the ABILHAND-kids questionnaire, 73 (53%) attained the maximum score of 42 points and 65 (47%) did not. In the 65 (47%) of patients who did not attain the maximum score, six of the top-ten most difficult tasks were of type 2B and 2C, which require precise digital activity of at least one hand (Figure 2). Overall, the ABILHAND-kids score was positively correlated with the patient-rated VAS score for functional outcome (r_s=0.27), and with age at follow-up (r_s=0.33). However, the ABILHAND-kids score was similar for patients who were satisfied with their outcomes and patients who were not (p=0.466).

In the 97 patients who filled out the CAPE questionnaire, 13 (13%) patients were dissatisfied with their outcomes. These 13 patients participated significantly less in 'Recreational activities'

(95%CI 2.2 – 25.0; p=0.019), compared to patients who were satisfied with their outcome. Dimension and domain specific scores were similar for patients who were satisfied with their outcomes and patients who were not (p≥0.069). However, the patient-rated VAS score for overall thumb appearance was positively correlated with the 'Enjoyment' dimension of the CAPE questionnaire (r_s=0.43), and with the 'Recreational activities' intensity score of the CAPE questionnaire (r_s=0.27).

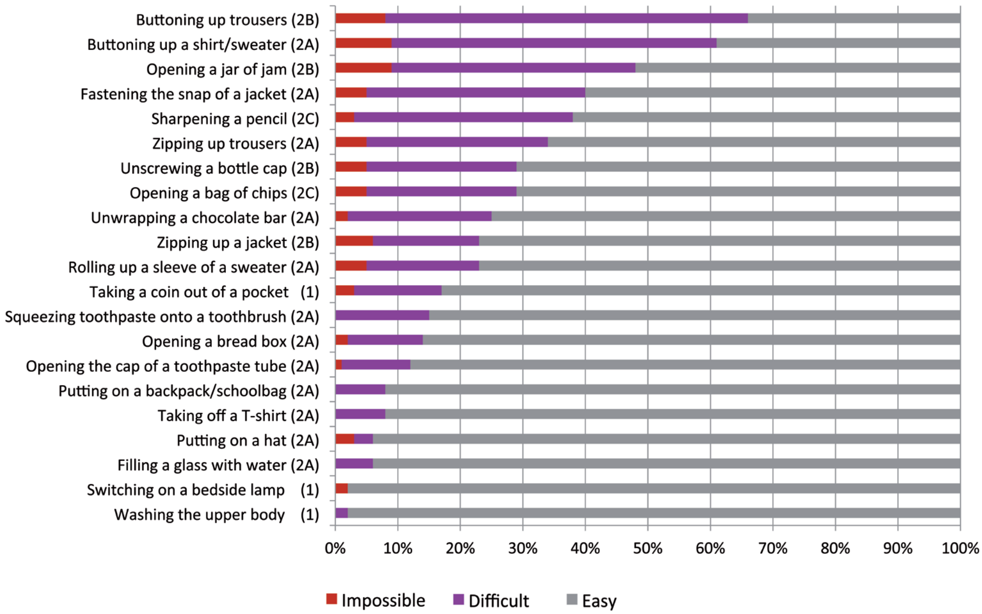

Figure 2: ABILHAND-kids questionnaire results of the 65 (47%) patients who did not attain the maximal score.

1 = Unimanual activities
2A = Bimanual activities manageable with several unimanual steps
2B = Bimanual activities requiring stabilization with one hand and digital activity with the other
2C = Requiring digital activity from both hands.

Results of the 79 patients who filled out the HUI15Q questionnaire are shown in Figure 3. The average HUI was 0.84 (SD=0.17). Both HUI and attribute scores were similar for patients who were satisfied with their outcomes and patients who were not (p≥0.214). Patient-rated VAS scores for functional outcome correlated with the 'Ambulation', 'Dexterity', 'Cognition', 'Emotion', and 'Pain' attribute scores (r_s ranging 0.23 – 0.34), and to the HUI itself (r_s=0.32).

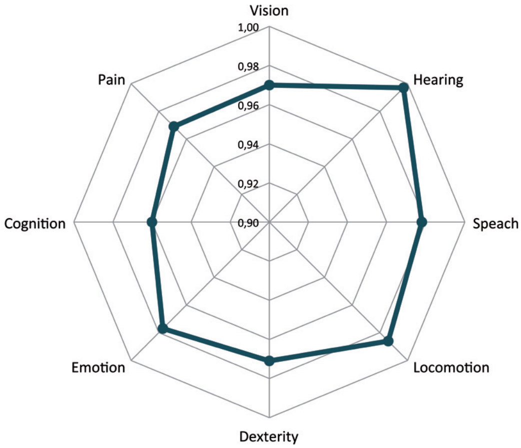

Figure 3: Radar plot of the different attribute scores of the HUI15Q in radial polydactyly patients.

The relationship between patient-reported domains of the ICF

When evaluating the relationships between outcomes of the various questionnaires, there was a correlation between the ABILHAND-kids score and participation in 'Physical activities' according to the CAPE (r_s=0.20). Furthermore, the ABILHAND-kids score correlated with the 'Dexterity' attribute scores of the HUI15Q (r_s=0.21). The 'Social activities' intensity score of the CAPE questionnaire correlated with the 'Speech' attribute scores of the HUI15Q (r_s=0.28), and participation in 'Informal activities' with the 'Speech' and 'Emotion' attribute scores (r_s=0.28 and r_s=0.26, respectively).

Comorbidity in our sample is described in Table 3. Patients with comorbidity reported significantly more manual ability impairment in the ABILHAND-kids questionnaire, compared to patients without comorbidity (95%CI -1.6 – -0,3; p=0.005). No such association was found for patient-reported VAS scores for thumb function and appearance, nor for participation and HRQoL.

Table 3: List of comorbidities in the total sample of 146 participating patients.

	Comorbidity *n* **= 47**
Cardiac	10 (21%)
Musculoskeletal	8 (17%)
Skin and soft tissue	7 (15%)
Urogenital	4 (9%)
Neurological	2 (4%)
Gastro-intestinal	1 (2%)
Pulmonary	1 (2%)
Syndrome	14 (30%)

DISCUSSION

The patient perspective on outcome of surgery in radial polydactyly is under-exposed in presently available medical literature. Therefore, the aim of this study was to shed light on the relationship between clinical assessment, manual ability, participation, and HRQoL of radial polydactyly patients.

Patients who were dissatisfied had objectively worse outcomes in terms of clinician-rated Rotterdam scores and VAS scores for function and appearance of the thumb (Table 2). Moreover, nine out of nineteen (47%) were dissatisfied due to residual deviation of the thumb and may need revision surgery. Therefore, it is advisable to correct all patho-anatomy with deviating potential (e.g., abnormal bone, instable joint surfaces, tendon anomalies, etc.) at the primary surgery [19] to prevent revision surgery, which only tends to result in modest improvements in outcome.[5,20-22]

On average, patients rated their functional outcome better than clinicians. Also, the patient-rated VAS scores for functional outcome were positively correlated to manual ability, which was excellent in the majority of cases. The tasks in the ABILHAND-kids questionnaire that were challenging for patients that did not attain the maximum score mostly required precise digital activity of at least one hand (type 2B and 2C; Figure 2). Since age was also positively correlated to manual ability, and the patients dissatisfied with dexterity were 5, 6, and 13 years old, respectively, it appears radial polydactyly patients rarely experience persistent problems with manual ability.[23,24]

Poor perceived thumb function or appearance might impact participation in everyday activities in radial polydactyly patients. This study shows dissatisfied patients participate less in recreational activities, compared to patients who are satisfied. They might also enjoy activities less if they are unhappy with thumb appearance. Although revision surgery may only lead to slight improvement of thumb function or appearance, its value in terms of participation may be substantial. Therefore, revision surgery may still be worthwhile if there is a realistic chance of improving the aspect of the thumb that bothers the patient the most.

The average HUI of 0.84 constitutes a relatively severe reduction in HRQoL,[25] comparable to, for example, pediatric asthma patients.[26] Since all outcomes were positive in general, and no significant association was found between HRQoL and comorbidity in this study, this might in part be explained by the fact the HUI15Q was a proxy-questionnaire. Parents tend to rate the HRQoL of their children lower than the children perceive themselves,[27,28] which is reflected by the relatively low 'Pain' and 'Dexterity' attribute scores in this study (Figure 3).

The main limitation of this study is missing data. Since response rates to the questionnaires were 62% – 95% and only 43% of patients had complete clinical and questionnaire follow-up, correlations between outcomes across the various domains of the ICF framework were weak. Moreover, lack of normative data impedes drawing conclusions on the manual ability and participation of radial polydactyly patients relative to the healthy population, which is beyond the scope of this study. While a complete dataset may have shown stronger relationships between clinical assessment, manual ability, participation, and HRQoL, the fact correlations were found in relatively small patient sample suggests the abovementioned domains are related.

Another limitation was the lack of sensitive and validated questionnaires to measure manual ability and participation. For example, the ABILHAND-kids was designed and validated for Cerebral Palsy ('CP').[15] Consequently, ABILHAND-kids scores showed significant ceiling effects in this study, suggesting radial polydactyly entails milder manual ability impairment than CP. Since the ABILHAND-kids could not discriminate between 'good' and 'poor' outcomes (i.e., between satisfied and dissatisfied patients, Rotterdam score, etc.).[23] Therefore, it would be beneficial to develop a questionnaire specific to CULAs affecting thumb function with better discriminative capabilities. While the CAPE and HUI15Q are validated for the pediatric population,[17,18] additional data on environmental factors (e.g., socio-economic status, education, etc.) is necessary to put the results into perspective.

These limitations notwithstanding, this study sheds light on aspects of outcome that affect radial polydactyly patients in everyday life. Although patient satisfaction may be difficult to improve, some aspects are amenable to surgical correction (e.g., thumb deviation) and may profoundly affect the patients overall wellbeing. Future studies should be directed at measuring the effectiveness of revision surgery to improve patient-reported outcomes, such as manual ability, participation, an HRQoL. Since perception of the aforementioned aspects may change depending on the age of the patient, it is important to actively follow-up patients until they reach maturity.

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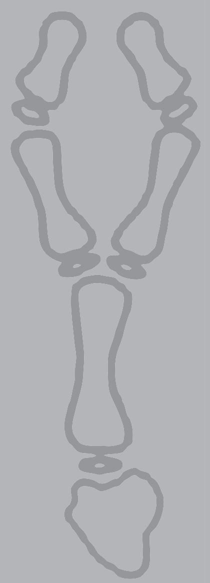


CHAPTER 9

General Discussion

Radial polydactyly is one of the most common congenital upper limb anomalies ('CULAs') affecting the thumb, and therefore likely to be encountered in the practice of many hand surgeons. However, with an estimated incidence of 2.3 per 10.000 live births, it is still a rare disease. Moreover, radial polydactyly has a very heterogeneous clinical presentation, impeding evidence-based consensus on treatment strategies. As a consequence, evidence on the optimal treatment of radial polydactyly is mainly limited to single-center or single-surgeon case series.

The aim of this thesis was to further optimize treatment strategies for radial polydactyly by studying outcome within the ICF framework, using data from an international, multicenter, long-term follow-up project. **Part one** of this thesis was focused on establishing the most reliable and valid classification and outcome assessment systems for radial polydactyly. In **Part two**, the established classification and outcome assessment systems were applied to study surgical treatment and the main determinants of postoperative outcome. The aim of **Part three** was to base the information on manual ability, participation, and quality of life, given during preoperative counseling of radial polydactyly patients, on outcome data.

The main findings are outlined and discussed below, followed by recommendations for clinical practice, and by stating future perspectives on radial polydactyly related research.

PART ONE: CLASSIFICATION AND OUTCOME ASSESSMENT

Classification

Clinical classification systems provide clinicians and scientists with a means to communicate by categorizing a clinical spectrum of cases into groups of patients with similar characteristics. For radial polydactyly, the Wassel classification is the most widely applied, while the more recently developed Rotterdam classification (**Chapter 2**, Figures 1 and 2) is the most all-embracing. In **Chapter 2** of this thesis, a comparison of both classifications revealed 40% of radial polydactyly cases do not fit any category in the Wassel classification, whereas all cases can be classified using the Rotterdam classification. The complexity of the Rotterdam classification does not affect its intra- and interobserver reliability, which is similar for both classifications.

Although classification systems could also support clinical decision-making by indicating the optimal surgical treatment for each category in the system, both abovementioned classifications for radial polydactyly fall short in this respect. The two main reasons for this shortcoming are the fact both classification systems are based on osseous configuration, and the fact that both classifications include several poorly defined categories.

Classifications based on osseous configuration are not fit to describe aspects of radial polydactyly that are less apparent on X-ray. Because the preoperative X-rays are usually made before the age of 2 years, the bones have not yet fully ossified (e.g., the physis and epiphysis, possible cartilaginous connections between adjacent phalanges). Moreover, part of the patho-anatomy cannot be visualized using X-ray (e.g., soft tissue abnormalities). Since the aforementioned structures do have surgical implications, it might be more beneficial to classify radial polydactyly cases according to indicated surgical treatment, rather than osseous configuration. Although such a classification system does exist, it is unknown whether the relationship between the categories in that system and the applied surgical treatment is generalizable beyond the clinic where the system was designed, as both case mix and surgical treatment may vary between clinics.

The fact that both the Rotterdam and Wassel classification include several poorly defined categories impedes valid comparison of outcomes between the categories in these systems. Several authors have emphasized the ambivalent definition of the level of duplication (especially in types I and II, and in types III and IV) in the Wassel classification, as well as the oversimplification of radial polydactyly with triphalangeal components (i.e., type VII). While the Rotterdam system is unequivocal regarding level of duplication, the deviating components ('D') and hypoplasia ('H') are poorly specified, which is reflected by poor reliability when classifying these components.

To add clinical relevance to the Rotterdam classification, **Chapter 5** contains suggestions for clear definitions of the abovementioned components based on their potential surgical implications. For deviating components, a minimal deviation of 20 degrees is suggested, based on the average angular deformity that was treated with transverse osteotomy in the study population. Hypoplasia ('H') is harder to define, since it is inherently associated with radial polydactyly. From a surgical point of view, cases with extra-capsular fibrous attachments or mere skin connecting the extra thumb to the dominant counterpart may be classified as 'hypoplastic', since collateral ligament repair might not be required in these cases and simple ablation may be sufficient.

Outcome assessment

There are numerous assessment systems to evaluate postoperative outcome in radial polydactyly patients. These assessment systems comprise different items and the importance of each item varies from system to system. Consequently, the conclusion on postoperative outcome varies substantially for a single radial polydactyly patient, depending on the assessment system used. To obtain an unbiased perspective of the effects of specific patient characteristics and surgical procedures on outcome, the assessment system with optimal reliability and validity needed to be established.

An analysis of agreement between three independent observers with varying clinical experience in **Chapter 3** showed the outcome assessment system designed by the Japanese Society for Surgery of the Hand ('JSSH') has the best interobserver reliability. Furthermore, the JSSH system has superior content validity, construct validity, and criterion validity compared to nine other outcome assessment systems.

In absence of a gold standard to describe hand function or overall outcome for radial polydactyly patients, the validity of each outcome assessment system had to be determined using several proxies. Content validity was established based on consensus between four investigators regarding the most important items of an assessment system, construct validity based on correlations with VAS scores for overall functional and esthetic outcomes rated by three investigators, and criterion validity based on correlation with a manual ability questionnaire. The construct validity of the studied assessment systems was particularly debatable, because the number of points per item and the cut-off values for a given number of points had been determined arbitrarily in each system.

The construct validity of the original JSSH outcome assessment system could be optimized using the data-driven approach described in **Chapter 4**. The importance of each item to overall functional and esthetic outcome was determined using linear regression analysis on data from an international, multicenter, long-term follow-up project. Based on the distribution of the data in the study population, cut-off values for the number of points within each item of the system were subsequently redefined, resulting in a clinically-weighted outcome assessment system for radial polydactyly: the Rotterdam score.

Compared to the original JSSH system, the Rotterdam score has greater construct validity, a similar internal consistency and interobserver reliability, a higher correlation with VAS scores for overall functional and esthetic outcome, and has better discriminative properties. Despite these advantages, the patient-reported domain score may still paradoxically influence overall outcome in cases where the patient is dissatisfied, despite excellent scores in the clinician-reported domains, and vice versa. Therefore, it is advisable to separately report the overall score, as well as the functional domain, appearance domain, and patient-reported domain scores.

PART TWO: SURGICAL TREATMENT OF RADIAL POLYDACTYLY

Surgical technique and determinants of outcome

Evidence on optimal surgical treatment of radial polydactyly and on determinants of postoperative outcome is presently limited to single-center case series and narrative reviews. Most studies cover all radial polydactyly types in a single paper, and discuss postoperative outcome using an assessment system that was specifically composed for the study by the leading investigators. Although these studies have substantially increased our understanding of the surgical treatment of radial polydactyly and its sequelae, evidence is still conflicting on many preoperative and surgery-related factors.

A multicenter effort to investigate optimal surgical treatment and determinants of postoperative outcome in a valid and generalizable way is described in **Chapter 5**. To retain a fairly homogenous sample, only patients with radial polydactyly types II, IV, and IV with triphalangeal components ('IV-Tph'), who were operated using the same resection and reconstruction (or 'reconstruction') principles were included. All cases were evaluated using the validated Rotterdam score, by an investigator who was uninvolved in the surgical treatment of the patients.

The main findings regarding optimal surgical treatment suggest poor outcomes occur less frequently in patients treated by the combination of: (1) removal of the worse-developed (usually) radial thumb, (2) sparing the collateral ligament of the affected joint, (3) restoration of joint congruency by a longitudinal osteotomy or by shaping the joint surface, (4) closure of the joint capsule with repair of the collateral ligament, and (5) meticulous suturing of the skin. Poor postoperative outcomes occur more frequently in patients with mal-alignment of the tendons.

Regarding other determinants of postoperative outcome, there is a positive association with experience in the field of congenital hand surgery. On the other hand, a negative association exists between multiple surgeries and postoperative outcome. Up to 18% of patients need secondary surgery, mostly to correct deviation (17% of reoperations in type II and 24% in type IV) and joint instability (43% of reoperations in type IV and 38% in type IV-Tph). These findings underline the need for centralization of care for congenital hand anomalies to hospitals that employ hand surgeons with the expertise to recognize and treat all patho-anatomic problems at the primary surgery.

Other suspected determinants, such as age at primary surgery and radial polydactyly type, did not affect outcome in our study. While these findings were supported by sufficient data, the study was still underpowered to find effects in a considerable number of other subgroup analyses, such as the hypothesized effect of a delta-shaped basal phalanx on postoperative deviation in radial polydactyly type II.

These limitations notwithstanding, the quantitative, statistical approach in this study contributes substantially to our understanding of determinants of outcome in radial polydactyly. Various abovementioned findings had been suggested in other retrospective and cross-sectional studies, but had never been supported by sufficient data or thorough statistical analysis. Nevertheless, to corroborate the causal relationships between these determinants and outcome, a prospective study is warranted.

The Bilhaut procedure versus reconstruction

The vast majority of radial polydactyly patients are operated using either the reconstruction technique or the Bilhaut procedure. In literature, specific benefits and drawbacks have been suggested for both procedures. Reconstruction may preserve active range of motion ('AROM') at the risk of joint instability and a smaller thumb size, while the Bilhaut procedure may yield more stable, larger, and stronger thumbs, at the risk of joint stiffness, marked broadness, and conspicuous scarring of the nail and pulp. Numerous modifications to the original Bilhaut procedure have been suggested to ameliorate the esthetic drawbacks of the procedure and expand its usefulness. Despite overlapping indications and suspected differences in outcome of both procedures, a comparative study has never been done.

To ensure a valid comparison between both procedures, two distinct treatment groups with comparable indications need to be defined. This can be achieved by strictly defining both surgical techniques and then matching cases treated with a Bilhaut procedure to similar cases treated with reconstruction. In **Chapter 6**, the Bilhaut procedure was defined as a surgery where nail, soft tissue, and bone from both thumbs are utilized to create a single thumb, and subsequently compared to cases treated with resection of one thumb and reconstruction of the other. Matching was done based on radiological similarity between cases.

The results of this matched comparative study indicate the above-defined Bilhaut procedure may not be worthwhile compared to reconstruction. Although thumbs are more stable after the Bilhaut, this stability does not result in stronger thumbs. In addition, the appearance of the nail proved unappealing in the eyes of a 22-observer panel consisting of experts, doctors, medical students, and laypeople. Although AROM was also reduced on average in the Bilhaut group compared to reconstruction, the study was likely underpowered to detect a significant difference.

Although **Chapter 6** revealed many clinically relevant differences between both procedures, the generalizability of the results is limited due to the strict definition of the Bilhaut procedure, and because the matched cases solely comprised two thumbs of similar size and shape. In practice, so-called ‘unbalanced’ radial polydactyly cases, which are classically treated with the reconstruction technique, occur more frequently. Moreover, modifications to the Bilhaut procedure can expand its use to cases with more asymmetric hypoplasia of both thumbs, and avert complications such as conspicuous scarring and nail deformity.

Technical examples of potentially beneficial modifications to the Bilhaut procedure include use of osteotomies to realign joint surfaces after meticulous alignment of the joint surface in cases with osseous asymmetry, and utilization of a single (better-developed) nail and nail bed to improve postoperative appearance. Preoperative size of the nail may be less relevant, since postoperative nails size is rarely smaller than 75% of the contralateral hand after ten years of follow-up (**Chapter 6**, Figure 4).

PART THREE: PATIENT-REPORTED OUTCOMES

Strength in radial polydactyly

Radial polydactyly is inherently associated with hypoplasia of both affected thumbs. Although surgical correction is mostly satisfactory in terms of thumb stability, mobility, and appearance, residual hypoplasia tends to impair postoperative thumb size and strength. Since the preoperative patient presentation (e.g., degree of thumb hypoplasia, aspect of the thenar musculature, extrinsic tendon configuration, etc.) affects the surgeon’s ability to optimize postoperative thumb size and strength, radial polydactyly patients are likely to retain a degree of strength impairment.

While motion-specific strength, such as strength of opposition and palmar abduction, is particularly likely to be affected by of the specific patho-anatomy in radial polydactyly patients, medical literature is limited to pinch strength measurements. In addition, there is limited evidence on how potential determinants such as joint stability and hand dominance affect thumb strength in radial polydactyly patients. Furthermore, the extent to which strength impairment in radial polydactyly affects the patient's ability to perform everyday tasks is debatable.

The analyses in **Chapter 7** show strength of the affected thumb is reduced to 83% – 94% of the unaffected thumb in radial polydactyly patients. Although this is a significant impairment from a statistical point of view, 52% of patients report unimpaired manual ability, which is not correlated with thumb strength in radial polydactyly. On the other hand, there is a correlation between absolute pinch strength and manual ability, suggesting a minimum strength threshold is required to perform all everyday tasks with ease.

Patient-specific determinants of thumb strength in radial polydactyly are: hand dominance, thumb size, and number of surgeries. An affected dominant hand and increased thumb size are positively associated with thumb strength, while patients who need multiple surgeries in treatment of radial polydactyly tend to have weaker thumbs. The latter finding underlines the association between multiple surgeries and inferior outcomes (**Chapter 5**), and the importance of successful primary surgery.

The patient perspective

Medical literature on radial polydactyly is almost entirely focused on surgical technique and on outcomes that are directly influenced by surgical intervention, such as thumb alignment and joint stability. In practice, the majority of questions parents ask at the first outpatient clinic visit are related to other domains of the ICF framework, i.e., activity ('manual ability'), participation, and contextual factors influencing Health Related Quality of Life ('HRQoL'). To improve evidence-based counseling of radial polydactyly patients, **Chapter 8** provides a more holistic, patient-oriented perspective on treatment outcome, by integrating outcomes from all domains of the ICF framework.

Regarding the body function and structures domain, patients rate their thumb function better than clinicians, and their thumb appearance similar to clinicians. In terms of functional deficit, dexterity and thumb strength are the main reasons why patients are dissatisfied. In terms of appearance, patients are mainly dissatisfied with deviation of the thumb and with displeasing scars. The fact that clinicians also consider scar appearance a key determinant of esthetic outcome (**Chapter 4**), underlines the importance of meticulous suturing of surgical wounds (**Chapter 5**).

There is a positive correlation between patient-rated functional outcome and their manual ability. Although manual ability is generally excellent in radial polydactyly patients, they sometimes have trouble with tasks that require digital dexterity. In patients who are dissatisfied with their thumb function and dexterity, the surgeon may offer to perform an opponens plasty using abductor digiti minimi ('ADM') or flexor digitorum superficialis ('FDS'), in an effort to improve the patient's ability to more easily perform everyday tasks.

Patient participation in recreational activities and enjoyment scores are positively correlated to patient-rated esthetic outcome, which suggest perceived appearance of the thumb affects social functioning in radial polydactyly patients. Moreover, since patients rate their thumb appearance similar to clinicians, these findings also underline the relevance of esthetic outcome assessment (**Chapter 4**).

Regarding HRQoL, many attribute scores, such as dexterity, emotion, and pain, are positively correlated to patient-rated functional outcome. Moreover, deficits in dexterity affect health utility indices in almost half of radial polydactyly patients. While these findings suggest a perceived reduction in thumb function influences HRQoL, the questionnaire used to measure quality of life was a proxy version and proxies (e.g., parents) are known rate the affected child's HRQoL lower than these children would.

The abovementioned findings underline that patient perspective is importantly related to many factors within the ICF framework. However, the questionnaires used to measure manual ability, participation, and HRQoL may not be suitable to differentiate between good and poor outcomes, because they did not detect a significant difference between patients who were satisfied with their functional and esthetic outcomes, and patients who were not.

The findings described in **Chapter 8** may be helpful and reassuring when counseling patients and their parents. However, a comparative study is still necessary to assess the manual ability, participation, and HRQoL of radial polydactyly patients relative to the general population. Nevertheless, given the generally favorable outcomes in terms of hand function, appearance, and patient satisfaction, and given the incredible ability of children to adapt, radial polydactyly is unlikely to impede children from leading happy and fulfilled lives.

Recommendations for clinical practice

The findings in this thesis support a number of recommendations for optimal treatment of radial polydactyly.

At the first outpatient clinic visit, parents may be reassured isolated radial polydactyly constitutes a minor impairment in terms of hand function, manual ability, participation, and HRQoL in the majority of cases. However, up to 20% of patients will need secondary surgery at some point, most likely to correct residual deviation or joint instability, or to improve dexterity by optimizing thumb opposition.

The timing of surgery is arbitrary, and may take place between the ages of 1 – 2 years. However, it is paramount the child is operated by (or under supervision of) a hand surgeon with extensive experience in treating CULAs, preferably in a specialized center. All patho-anatomic differences should be addressed to at the primary surgery, to minimize the need for secondary procedures later in life.

From a surgical point of view, the reconstruction technique is indicated in the majority of cases. Adhering to the essential steps of the reconstruction technique (Section on 'Surgical technique and determinants of outcome', above) is likely to result in favorable outcomes. Although there may be indications for modified Bilhaut procedures, these are limited to cases with severe hypoplasia of both thumbs, since better results are likely achieved using the reconstruction technique, even in type IV D r/u (or 'diamond shaped') cases.

Postoperative dressings may include a small protective plaster cast for the thumb, and adhesive elastic bandage up to the elbow to ensure the child does not remove the bandages. In most cases, the short-term postoperative period is uneventful, and the child resumes full active use of the reconstructed thumb during the following 3 months. However, standardized follow-up should be continued until age 18, since functional complaints such as reduced dexterity, and esthetic considerations such as deviation of the joints may become more pronounced as the patient matures.

Future directions

Overall, the studies in this thesis have met the aims put forward in **Chapter 1**. Data from an international, multicenter, long-term follow-up project, helped to clarify potential determinants of outcome and the effects of available surgical techniques. Nevertheless, there are still a considerable number of unanswered questions.

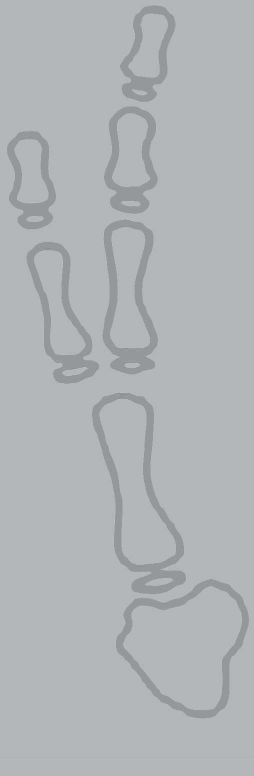
Regarding classification of radial polydactyly, it is debatable whether radiological taxonomies such as the Rotterdam classification remain feasible. Alternatively, it may be more useful to classify patients based on surgical implications. If radiological classification is preferred, one should consider optimizing the protocol regarding the directions of the X-rays taken. If surgical classification is preferred, the reliability and clinical applicability of the available classification systems needs to be investigated.

Concerning the radial polydactyly-specific outcome assessment, the analyses used to design the Rotterdam score would need to be repeated in a different population to see if the score is truly generalizable beyond the study population. In addition, a validated manual ability questionnaire specific to CULAs (involving the thumb) is needed, discriminating between good and poor outcomes. Moreover, all questionnaire data presented in this thesis need to be compared to data from a population of healthy subjects, to determine the true impact of radial polydactyly on manual ability, participation, and HRQoL.

Our study of effectiveness of surgical treatment and the main determinants of outcome was impeded by the cross-sectional study design, and by insufficient documentation of both surgical procedures and other (preoperative) determinants of outcome. Although we found numerous factors associated with outcome, we could not answer the question whether outcome in radial polydactyly is mainly determined by the preoperative situation, or by surgery. Since the only way to investigate causal relationships between determinants and outcome is through prospective studies, prospective studies should be the main focus of future congenital hand research.

A good example of how prospective studies may increase our understanding of a clinical course is the evaluation of strength in radial polydactyly. For instance, muscle volume is known to correlate with strength and can be measured using ultrasound. Measuring thenar volume before primary surgery, or measuring hypothenar volume before performing an ADM opponens plasty, may enable us to predict poor dexterity or successful opponens plasty in the long run.

Prospective studies require considerable investments in terms of time and resources. This is particularly applicable to radial polydactyly, which is a rare disease and requires a long duration of follow-up to draw meaningful conclusions. Before embarking on a prospective study, a number of factors should be carefully considered, including the population, the timing and number of follow-up measurements, and the determinants and outcomes documented at each follow-up moment. Furthermore, the only way to guarantee enough statistical power and generalizable findings is through pooling of data from (worldwide) international congenital hand clinics. A consortium with a clear and shared mission statement, and all the required expertise, would be needed to generate sufficient funding and to further propel evidence-based treatment of this patient group.



CHAPTER 10

Summary

Thumb duplication or 'radial polydactyly' is the most common congenital upper limb anomaly ('CULA') affecting the thumb. The clinical presentation of this highly heterogeneous patient population may vary from a subtle doubling of the nail lunula, to complex synpolydactyly with synostosis, intrinsic and extrinsic muscle anomalies, and joint instability. Therefore, defining an optimal treatment strategy for each individual case remains challenging.

The aim of this thesis was to improve treatment for radial polydactyly by studying outcome within the International Classification for Functioning, Disability, and Health ('ICF') framework. In pursuit of valid and generalizable findings, we gathered data in an international, multicenter setting, with long-term follow-up, and focussed on the most common radial polydactyly types (i.e., types II, IV, and IV with triphalangeal components or 'IV-Tph'). We started by defining the best classification and outcome assessment systems in **Part One**, proceeded to evaluate surgical technique and determinants of outcome in **Part Two**, and balanced these clinician-reported outcomes against patient-reported outcomes in **Part Three** of this thesis.

Part One: Classification and outcome assessment

Radial polydactyly is often classified using the Wassel classification. Many modifications have been proposed to expand classification possibilities and add surgical relevance to the Wassel classification. The recently introduced Rotterdam classification accommodates various subtypes of radial polydactyly that require specific surgical treatment, including triphalangeal thumbs and thumb triplication. Both classifications are compared in **Chapter 2**, to determine which is best suited to meet current clinical and scientific demands. A review of 520 radial polydactyly cases showed 40% could not be classified using the Wassel classification, while all cases could be classified using the Rotterdam classification. All unclassifiable cases had aberrant components (i.e., of the triphalangeal, deviating, or hypoplastic kind). Intra- and interobserver reliability was comparable for both classification systems (K 0.65 – 0.87). The most common radial polydactyly types (i.e., II and IV) had the lowest reliability in both classifications (K 0.30 – 0.59). In conclusion, both classifications have similar reliability, but the all-embracing taxonomy of the Rotterdam classification shows more potential when it comes to describing pre-operative situation and outcome in specific cases that require specific surgical treatment.

In **Chapter 3**, the reliability and validity of ten outcome assessment systems for radial polydactyly is compared. Three independent examiners evaluated 41 radial polydactyly cases to compare reliability. Validity was compared based on correlation with VAS scores for thumb function and appearance, and correlation with the ABILHAND-kids manual ability questionnaire. The outcome assessment system introduced by the Japanese Society for Surgery of the Hand ('JSSH') had the highest reliability (ICC's ≥ 0.70), and was most valid based on correlations with functional and appearance VAS scores ($r_s = 0.48 – 0.80$ and $r_s = 0.45 – 0.63$). Therefore, the JSSH system was the best available system to evaluate overall outcome in radial polydactyly at the start of this thesis.

Although the JSSH system had superior reliability and validity compared to other available systems, its design was based solely on expert opinion. **Chapter 4** describes the data-driven approach used to construct a clinically-weighted outcome assessment system, the Rotterdam score, using the items from the JSSH system. Linear regression analyses on data of 121 patients revealed active thumb flexion, scar appearance, and prominence at amputation site were the main items influencing overall thumb function and appearance in the eyes of clinicians. In the Rotterdam score, points were redistributed over the items based on their weight in the regression analyses, and the cut-off values for each number of points were defined by the distribution of data in the sample. The statistically supported design of the Rotterdam score ensures valid and generalizable outcome assessment, which reflects the way clinicians value individual items as determinants of overall outcome.

Part Two: Surgical treatment of radial polydactyly

After establishing the best ways for clinicians to classify and assess postoperative results in the first part of this thesis, **Chapter 5** aimed to determine how patient characteristics and surgical technique influence outcome. The Rotterdam assessment system was used to evaluate outcome in 114 cases, after a mean follow-up of ten years. There were 94 (82%) cases with follow-up data after single surgery, and 20 (18%) cases with data after multiple surgeries. Within the single surgery group, radial polydactyly type IV had worse functional outcome compared to types II and IV-Tph. The multiple surgeries group had worse overall outcome, functional outcome, pain, and satisfaction compared to the single surgery group. This underlines the importance of correcting as much as possible during the primary surgery, to prevent the need for revision procedures. In general, tendon anomalies had occurred more frequently in the worst 25% of outcomes. On the other hand, patients who were primarily treated by surgeons specialized in CULAs had better overall outcome that patients treated by less specialized consultants.

In **Chapter 6,** outcomes of the Bilhaut procedure were compared to outcomes of conventional reconstruction, in a matched sample consisting of radial polydactyly types II and IV. The Bilhaut procedure reduced the risk of suboptimal metacarpophalangeal joint ('MCPJ') instability outcome. On the other hand, increased MCPJ stability and thumb size following a Bilhaut procedure did not result in the presumed benefit to thumb strength. Regarding thumb appearance, the clinician found the Bilhaut increased the risk of suboptimal scar appearance, residual prominence at amputation site, thumb size, and nail appearance. However, a panel of 21 independent CULA specialists, doctors, medical students, and laypeople considered nail appearance the only esthetic drawback of the Bilhaut procedure. In conclusion, a Bilhaut procedure that utilizes nail, soft tissue, and bone from both thumbs may not be worthwhile in radial polydactyly cases where reconstruction is a viable option.

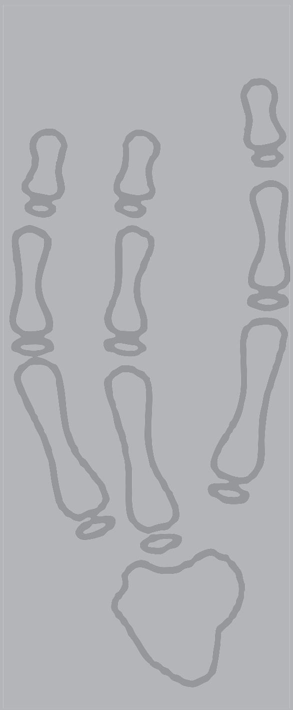
Part Three: Patient-reported outcomes

After focussing on the clinician's point of view in the first two parts, part three of this thesis elaborates on the point of view of the patient. In **Chapter 7**, strength impairment in radial polydactyly is quantified, the determinants of thumb strength are investigated, as well as the impact of thumb strength impairment on manual ability. In a sample of 84 unilaterally affected patients, thumb strength was impaired to 83% – 94% of the unaffected contralateral thumb. Thumbs of radial polydactyly patients with an affected dominant hand were 10% – 23% stronger on average, compared to patients with an affected non-dominant hand. Increased thumb size was positively correlated with pinch strengths, whereas increased number of surgeries showed a negative correlation. Forty-three (52%) patients attained the maximum score in the manual ability questionnaire, and there was no correlation between relative thumb strength and manual ability. Thus, we concluded that radial polydactyly patients have a relatively small but statistically significant strength impairment that does not seem impair their manual ability.

In **Chapter 8**, the clinician's assessment of thumb function and appearance is balanced against the patient's assessment of these aspects, as well as the patient-reported manual ability, participation, and Health Related Quality of Life ('HRQoL'). In a study with 146 participants, patients who were dissatisfied with either thumb function or appearance had lower clinician- and patient-rated functional and esthetic outcomes, compared to patients who were satisfied with their outcome. From a functional point of view, patients were mainly dissatisfied with dexterity and thumb strength. Regarding appearance, patients were mainly dissatisfied with deviation of the thumb and displeasing scars.

Patient-rated functional outcome was positively correlated with their manual ability, and with HRQoL attribute scores, such as dexterity, emotion, and pain. Although manual ability was generally excellent in radial polydactyly patients, they sometimes had trouble with tasks that require dexterity, especially at an early age. Patient participation in recreational activities and enjoyment scores are positively correlated to patient-rated esthetic outcome, which suggest perceived appearance of the thumb affects social functioning in radial polydactyly patients. Moreover, patients rate their thumb appearance similar to clinicians, which may play an important role in shared decision-making concerning revision surgery.

In summary, the Rotterdam classification and the Rotterdam score facilitate communication between clinicians and valid assessment of postoperative outcome, respectively. Surgical treatment can be challenging, especially in type IV, which may have worse functional outcomes than types II and IV-Tph. However, the resection and reconstruction principle should be preferred over the Bilhaut procedure whenever possible. It is paramount to correct all patho-anatomical differences at the primary surgery, in order to avoid the need for revision surgery, which is associated with poorer long-term outcomes. While radial polydactyly patients have reduced strength of the affected thumb, this does not impede their manual ability. Nevertheless, thumb function and appearance is likely to affect participation and HRQoL in radial polydactyly patients. All things considered, nothing stands in the way of doubling up on good outcomes at a table with experienced congenital hand specialists.



CHAPTER 11

Samenvatting

Dubbele duimen of 'radiale polydactylie' is de meest voorkomende aangeboren afwijking ('CULA') van de duim. De klinische presentatie is zeer heterogeen, en kan variëren van een subtiele verdubbeling van de lunula van de nagel van de duim, tot complexe synpolydactylie met synostose, afwijkende intrinsieke en extrinsieke musculatuur, en instabiele gewrichten. Het is dan ook taakstellend om de optimale behandelingsstrategie voor iedere individuele casus te bepalen.

Het doel van dit proefschrift was het verbeteren van behandeling van radiale polydactylie, door postoperatieve uitkomsten te bestuderen binnen de kaders van het International Classification for Functioning, Disability, and Health ('ICF') model. Met het oog op verkrijgen van valide en generaliseerbare bevindingen, verzamelden we data in internationaal, multicenter verband, met lange termijn follow-up, en richtten wij ons op de meest voorkomende typen radiale polydactylie (i.e., typen II, IV, en IV met triphalangeale componenten of 'IV-Tph'). We zijn begonnen met het definiëren van de beste classificatie systeem en uitkomst-beoordelingssystemen in **Deel Een**, vervolgens hebben we chirurgische techniek en determinanten van uitkomst bestudeerd in **Deel Twee**, en zetten we deze arts-gerapporteerde uitkomsten uit tegen patiënt-gerapporteerde uitkomsten in **Deel Drie** van dit proefschrift.

Deel Een: Classificatie en het beoordelen van uitkomst

Radiale polydactylie wordt vaak getypeerd met behulp van de Wassel classificatie. Er zijn vele aanpassingen op Wassel classificatie voorgesteld om meer diverse casuïstiek in te passen, en om de chirurgische relevantie te vergroten. De recent gepubliceerde Rotterdam classificatie omvat vele subtypen radiale polydactylie die specifieke chirurgische behandeling behoeven, zoals triphalangeale duimen en duim triplicaties. Beide classificaties worden met elkaar vergeleken in **Hoofdstuk 2,** om vast te stellen welk systeem het beste voldoet aan huidige klinische en wettenschappelijke vereisten. Een beschouwing van 520 radiale polydactylie casus bracht aan het licht dat 40% niet geclassificeerd kon worden in de Wassel classificatie, terwijl alle casus geclassificeerd konden worden in de Rotterdam classificatie. Alle niet-classificeerbare casus hadden aberrante componenten (i.e., van het triphalangeale, deviërende, of hypoplastische soort). De intra- en interbeoordelaar betrouwbaarheid was vergelijkbaar voor beide classificaties (K 0.65 – 0.87). De vaakst voorkomende typen radiale polydactylie (i.e., II en IV) konden het minst betrouwbaar worden geclassificeerd in beide systemen (K 0.30 – 0.59). Concluderend hebben beide classificaties vergelijkbare betrouwbaarheid, maar heeft de Rotterdam classificatie het meeste potentieel om de preoperatieve situatie en uitkomst te beschrijven van typen radiale polydactylie die specifieke chirurgische behandeling vergen.

In **Hoofdstuk 3** wordt de betrouwbaarheid en validiteit van tien uitkomst-beoordelingssystemen voor radiale polydactylie vergeleken. Drie onafhankelijke onderzoekers beoordeelden 41 radiale polydactylie casus om betrouwbaarheid te vergelijken. Validiteit werd vergeleken op basis van correlatie met VAS scores voor functie en uiterlijk van de duim, en correlatie met de ABILHAND-kids vragenlijst voor handvaardigheid. Het uitkomst-beoordelingssysteem van the Japanese Society for Surgery of the Hand ('JSSH') had de hoogste betrouwbaarheid (ICC's ≥ 0.70) en de beste validiteit gebaseerd op correlatie met VAS scores voor duim functie en duim uiterlijk ($r_s = 0.48 - 0.80$ en $r_s = 0.45 - 0.63$). Bij aanvang van dit proefschrift was het JSSH systeem het best beschikbare systeem voor het beoordelen van postoperatieve uitkomst bij radiale polydactylie.

Hoewel het JSSH systeem een betrouwbaar en valide instrument was in vergelijking tot andere beschikbare systemen, is het systeem volledig gebaseerd op expert opinion. **Hoofdstuk 4** beschrijft de data-gedreven methode die gebruikt werd om een klinisch gewogen uitkomst-beoordelingssysteem te construeren, de Rotterdam score. Hierbij vormden de items uit het JSSH systeem de bouwstenen. Lineaire regressie analyses van data van 121 patiënten toonde dat het actief buigen van de duim, uiterlijk van het litteken, en uitstulping ter plaatse van de verwijderde duim de belangrijkste determinanten waren van duim functie en duim uiterlijk in de ogen van clinici. In de Rotterdam score werd de puntenverdeling bepaald door het gewicht van items in de regressie analyses, en werd de afkapwaarde voor ieder aantal punten vastgesteld aan de hand van de dataverdeling in de steekproef. Deze statistische onderbouwing maakt de Rotterdam score tot een valide en generaliseerbaar uitkomst-beoordelingssysteem, dat weerspiegelt hoe clinici de waarde van individuele items voor algehele uitkomst zien.

Deel Twee: Chirurgische behandeling van radiale polydactylie

Na het definiëren van het beste classificatie systeem en uitkomst-beoordelingssysteem in het eerste gedeelte van dit proefschrift, wordt in **Hoofdstuk 5** beschreven hoe patiëntkenmerken en chirurgische techniek postoperatieve uitkomst beïnvloeden. De Rotterdam score werd gebruikt om uitkomst te beoordelen in 114 radiale polydactylie casus, na een gemiddelde follow-up van tien jaar. Er werden 94 (82%) casus beoordeeld met follow-up na één primaire procedure, en 20 (18%) casus na meerdere procedures. In de groep die beoordeeld werd na één primaire procedure, had type IV slechtere functionele uitkomsten dan typen II en IV-Tph. De groep die beoordeeld werd na meerdere procedures had gemiddeld slechtere algehele uitkomst, slechtere functionele uitkomst, meer pijn, en slechtere patiënt-tevredenheid dan patiënten uit de groep die beoordeeld werd na één primaire procedure. Dit onderstreept het belang van een uitgebreide primaire procedure, waarbij zo veel mogelijk gecorrigeerd wordt om toekomstige revisie procedures te voorkomen. Over het algemeen werden afwijkingen aan pezen van extrinsieke spieren vaker gezien in de 25% patiënten met de slechtste uitkomsten. Aan de andere kant hadden patiënten die primair geopereerd waren door CULAs gespecialiseerde chirurgen betere algehele uitkomsten dan patiënten die primair geopereerd waren door minder gespecialiseerde chirurgen.

In **Hoofdstuk 6** worden uitkomsten van de Bilhaut procedure vergeleken met uitkomsten van reconstructie, in een gematchte steekproef bestaande uit patiënten met radiale polydactylie typen II en IV. De Bilhaut procedure verminderde het risico op instabiliteit van het metacarpophalangeale gewricht. Aan de andere kan resulteerden betere stabiliteit van het metacarpophalangeale gewricht en de toegenomen omvang van de duim na een Bilhaut niet in het veronderstelde voordeel ten aanzien van duimkracht. Wat betreft uiterlijk van de duim hadden patiënten volgens de onderzoeker een hoger risico op suboptimale littekens, uitstulping ter plaatse van de verwijderde duim, afwijkende duim grootte, en nageldeformiteit. Echter, een panel van 21 CULA specialisten, artsen, geneeskunde studenten, en leken beschouwede nageldeformiteit het enige esthetische nadeel van de Bilhaut procedure. Concluderend worden Bilhaut procedures die nagel, weke delen, en bot van beide duimen combineren ontraden in casus waarbij conventionele reconstructie mogelijk is.

Deel Drie: Patiënt-gerapporteerde uitkomsten

Na uitgebreid te hebben stilgestaan bij het artsen-perspectief op behandeling en uitkomst bij radiale polydactylie, belicht deel drie van dit proefschrift het perspectief van de patiënt. In **Hoofdstuk 7** wordt krachtverlies van de duim bij patiënten met radiale polydactylie gekwantificeerd, en worden de determinanten van duimkracht onderzocht, alsmede de invloed van verminderde duimkracht op handvaardigheid. In een steekproef van 84 eenzijdig aangedane patiënten, was de duimkracht tot 83% – 94% verminderd ten opzichte van de gezonde zijde. Duimen van patiënten met een aangedane dominante hand waren gemiddeld 10% – 23% sterker ten opzichte van duimen van patiënten met een aangedane non-dominante hand. Duimomvang hier wel positief gecorreleerd met duimkracht, terwijl het aantal chirurgische ingrepen aan de duim een negatieve correlatie toonde. Drieënveertig (53%) patiënten behaalden de maximale score in de handvaardigheidsvragenlijst, maar er was geen relatie tussen handvaardigheid een relatieve duimkracht. Dit suggereert dat het kleine maar statistisch significante duimkrachtsverlies bij patiënten met radiale polydactylie geen invloed heeft op hun handvaardigheid.

In **Hoofdstuk 8** wordt de artsen-beoordeling van functie en uiterlijk van de duim uitgezet tegen de patiënten-beoordeling van deze aspecten, en tegen de patiënt-gerapporteerde handvaardigheid, participatie en Health Related Quality of Life (‘HRQoL’). Een studie met 146 deelnemers wees uit dat patiënten die ontevreden waren over het functioneren of het uiterlijk van hun duim daadwerkelijk slechtere functionele en uiterlijke uitkomsten hadden, volgens zowel arts als patiënt. Vanuit functioneel oogpunt waren patiënten met name ontevreden over verminderde handvaardigheid en duimkracht. Wat betreft uiterlijk waren standsafwijking en littekens van de duim het meest bezwaarlijk.

De patiënt-beoordeelde duimfunctie correleerde met handvaardigheid en met diverse HRQoL attributen, zoals handigheid, emotie, en pijn. Hoewel handvaardigheid van radiale polydactylie patiënten over het algemeen uitstekend was, hadden ze soms moeite met activiteiten die fijne motoriek vereisen, met name op jonge leeftijd. Participatie in recreatieve activiteiten en plezier-scores waren positief gecorreleerd met de patiënt-beoordeling van uiterlijk van de duim. Dit suggereert dat perceptie het duimuiterlijk gerelateerd is aan sociaal welbevinden bij radiale polydactylie patiënten. Daarnaast beschouwen patiënten het uiterlijk van de duim vaak hetzelfde als de arts, hetgeen van belang kan zijn bij beslissingen rondom het doen van revisie chirurgie.

Samenvattend faciliteren de Rotterdam classificatie en de Rotterdam score respectievelijk communicatie tussen clinici en valide beoordeling van postoperatieve uitkomst. Chirurgische correctie blijft uitdagend, met name bij type IV, welke een slechtere functionele uitkomst kan hebben dan typen II en IV-Tph. De Bilhaut procedure dient te worden vermeden indien reconstructie ook mogelijk is. Het is essentieel om bij de primaire ingreep alle patho-anatomische verschillen te corrigeren, in de hoop dat revisie-chirurgie in de toekomst onnodig zal zijn. Hoewel patiënten met radiale polydactylie verminderde duimkracht hebben, ondervinden ze daar geen hinder van in het dagelijks leven, al is duim functie en uiterlijk waarschijnlijk wel van invloed op hun participatie en kwaliteit van leven. Onder aan de streep staat niets in de weg van dubbel inzetten op goede uitkomsten aan een tafel met gespecialiseerde chirurgen.

APPENDICES

List of publications
PhD portfolio
Dankwoord
Curriculum vitae

LIST OF PUBLICATIONS

Comparison of functional outcome scores in radial polydactyly
Dijkman RR, van Nieuwenhoven CA, Selles RW, Hovius SE
J Bone Joint Surg Am. 2014 Mar; 96(6): 463-70.

Letter to the Editor regarding "Prognostic value of age and Wassel classification in the reconstruction of thumb duplication"
Dijkman RR, van Nieuwenhoven CA, Hovius SE
J Child Orthop. 2014 May; 8(3): 289-90.

A multicenter comparative study of two classification systems for radial polydactyly
Dijkman RR, van Nieuwenhoven CA, Selles RW, Habenicht R, Hovius SE
Plast Reconstr Surg. 2014 Nov; 134(5): 991-1001.

Clinical presentation, surgical treatment, and outcome in radial polydactyly
Dijkman RR, van Nieuwenhoven CA, Hovius SE, Hülsemann W
Handchir Mikrochir Plast Chir. 2016 Feb; 48(1): 10-17.

A matched comparative study of the Bilhaut procedure versus resection and reconstruction for treatment of radial polydactyly types II and IV
Dijkman RR, Selles RW, Hülsemann W, Mann M, Habenicht R, Hovius SE, van Nieuwenhoven CA
J Hand Surg Am. 2016 May; 41(5): e73-83.

A clinically weighted approach to outcome assessment in radial polydactyly
Dijkman RR, Selles RW, van Rosmalen J, Hülsemann W, Mann M, Habenicht R, Hovius SE, van Nieuwenhoven CA
J Hand Surg Eur. 2016 Mar; 41(3): 265-74.

Thumb strength and manual ability in radial polydactyly types II and IV
Dijkman RR, Leicher RG, Selles RW, Mann M, Hülsemann W, Hovius SE, van Nieuwenhoven CA
Submitted to J Hand Surg Am.

An international multicenter outcome study of radial polydactyly at the interphalangeal and metacarpophalangeal joint level
Dijkman RR, Selles RW, Mann M, Hülsemann W, Habenicht R, Hovius SE, van Nieuwenhoven CA
To be submitted to J Hand Surg Eur.

PHD PORTFOLIO

Name PhD student: Robert Richard Dijkman
PhD period: 2012 – 2016
Erasmus MC department: Plastic and Reconstructive Surgery and Hand Surgery
Promotor: Prof. dr. S.E.R. Hovius
Copromotors: Dr. C.A. van Nieuwenhoven & Dr. R.W. Selles

1. PhD training

Courses in methodology and statistics	Year	Workload
NIHES Master of clinical epidemiology	2012 – 2014	1.960 hours

Courses in didactic skills and research integrity		
BKO Individualized supervision course	2015	3 hours
BKO Teach the teacher - Module I	2015	15 hours
Erasmus MC research integrity course	2014	3 hours
Good Clinical Practice (GCP/BROK) course	2012	30 hours

Oral presentations		
A clinically weighted approach to outcome assessment in radial polydactyly – 10th World Symposium on Congenital Malformations of the Hand and Upper Limb, Rotterdam, The Netherlands	2015	20 hours
Radial polydactyly: The Bilhaut procedure vs. Resection and Reconstruction – NVPC Fall meeting, Amsterdam, The Netherlands	2014	20 hours
Comparison of ten outcome systems for evaluating treatment of radial polydactyly – 12th IFSSH Meeting, New Delhi, India	2013	20 hours
Comparison of nine outcome systems for evaluating treatment of radial polydactyly – 17th FESSH Meeting, Antwerp, Belgium	2012	20 hours
Comparing classifications for radial polydactyly: The change in distribution for the different subtypes – 17th FESSH Meeting, Antwerp, Belgium	2012	20 hours
Comparison of nine outcome systems for evaluating treatment of radial polydactyly – 23rd EURAPS Meeting, Munich, Germany	2012	20 hours
Comparing classifications for radial polydactyly: The change in distribution for the different subtypes – 9th World Symposium on Congenital Malformations of the Hand and Upper Limb, Dallas, Texas, USA	2012	20 hours

Poster presentations		
Comparing classifications for radial polydactyly: the change in distribution for the different subtypes – International Federation of Societies for Surgery of the Hand (IFSSH), New Delhi, India	2013	20 hours

(Inter)national conferences and seminars		
10th World Symposium on Congenital Malformations of the Hand and Upper Limb, Rotterdam, The Netherlands	2015	30 hours
19th FESSH Meeting, Paris, France	2014	24 hours
12th IFSSH Meeting, New Delhi, India	2013	30 hours
17th FESSH Congress, Antwerp, Belgium	2012	24 hours
23rd EURAPS Meeting, Munich, Germany	2012	24 hours
9th World Symposium on Congenital Malformations of the Hand and Upper Limb, Dallas, Texas, USA	2012	30 hours
NVPC	2012 – 2015	24 hours
NVvH	2012 – 2015	6 hours
Kortjakje	2012 – 2015	12 hours

Grants and funding		
Esser foundation research grant (10 month extension)	2015	n.a.
Nuts Ohra foundation research grant (€170.000)	2012	60 hours

Skillslab		
Microsurgical training	2012 – 2015	290 hours

2. Teaching activities

Lecturing		
2nd year medical school musculoskeletal elective: anatomy of the hand and forearm	2012 – 2015	30 hours
2nd year medical school musculoskeletal elective: acute and chronic conditions of the hand	2012 – 2015	30 hours
3rd year medical school minor 'From Head to Hands'	2012 – 2015	30 hours

Skills		
Dutch course in microsurgery (microsurgery coach)	2012 – 2015	40 hours
International course in microsurgery (microsurgery coach)	2012 – 2015	60 hours

Supervising master thesis		
S. van Dam	2013	60 hours
B. Leicher	2015	60 hours

3. Organizing activities

Conferences and symposia		
10th World Symposium on Congenital Malformations of the Hand and Upper Limb	2015	300 hours
24th Esser Course: "Ins and Outs of Nose Reconstruction."	2014	50 hours
22nd Esser Course: "What's New in Breast Reconstruction."	2014	50 hours
21st Esser Course: "Wide Awake: A Live Surgery Event."	2013	50 hours
20th Esser Course: "Masterclass Neuropathic Pain."	2013	50 hours
19th Esser Course: "To the base of the thumb. The CMC Joint."	2013	50 hours
18th Esser Course: "Bend… or break. Tendon injuries of the hand."	2012	50 hours

Workshops		
Local flaps in plastic surgery	2014	2 hours
Tenden repair	2013	2 hours
Basic nerve reconstruction	2013	2 hours

DANKWOORD

"Alleen kun je niets, je moet het samen doen." Hoewel ik zelf geen voetballer ben, raakt dit een-tweetje tussen Cruijff en mij de kern ieder promotietraject. In de zes jaren waarin dit proefschrift vorm kreeg hebben meer mensen mij gesteund, geïnspireerd, of anderszins geholpen dan ik op kan noemen. Graag bedank ik hier iedereen die mij hielp dit proefschrift af te ronden.

Prof.dr. S.E.R. Hovius, beste prof, ik weet nog goed dat ik al twee jaar onderweg was, pas één artikel had gepubliceerd, maar wel bezig was met het organiseren van een aantal symposia en een stranddag. U belde mij (al fietsend) om mede te delen dat ik naast het onderzoek een full-time NIHES master zou volgen, die in uw optiek de 'gouden rand' om dit proefschrift ging worden. Wat heb ik veel van u mogen leren. We zien beiden in iedere uitdaging een mogelijkheid, dank dat u beide zag in mij.

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Dear dr. Habenicht, thank you for the warm welcome to both your Wilhelmstift and your home in Hamburg. Even more so, thank you for making me feel ***at*** home there. The way you modestly carry unparalleled clinical expertise is an example to me personally. Learning from you was one of the greatest privileges of my PhD period. I hope we will see each other in the future.

Dear dr. Hülsemann, dr. Mann, dr. Winkler, thank you for your support throughout this crazy "Doppeldaumen" project and for your hospitality during my stay in Hamburg. Es hat gut geklappt!

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De staf en assistenten van de plastische chirurgie van het Erasmus MC, in het bijzonder Michiel en Jan, dank voor jullie hulp en advies, zowel op de werkvloer als daarbuiten. Ik ben blij de opleiding te doen met onze te gekke groep assistenten en trots het vak te mogen leren aan onze afdeling. Ik hoop jullie allemaal weer te zien op het feest!

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CURRICULUM VITAE

Robert Richard Dijkman was born on September 16th, 1986 in Leidschendam, the Netherlands. After graduating from the Bonnefanten College in Maastricht in 2004, he studied Biomedical Sciences at Utrecht University. In 2005, he switched to studying medicine at the Erasmus University in Rotterdam and graduated with honours 2012. That year, he started the PhD project resulting in this thesis at the Department of Plastic and Reconstructive Surgery and Hand Surgery at the Erasmus MC University Medical Center in Rotterdam (Prof. Steven Hovius), in close collaboration with the Catholic Children's Hospital Wilhelmstift in Hamburg, Germany (Dr. Rolf Habenicht). In 2014, he attained a Master's degree in clinical epidemiology at the Netherlands Institute for Health Sciences (NIHES). After working as a house officer in the fall of 2014, he was admitted into the plastic surgery training program at the Erasmus MC University Medical Center in Rotterdam (Dr. Leon van Adrichem). He is currently completing his two-year general surgical training at the St. Antonius Hospital in Nieuwegein, the Netherlands (Dr. Djamila Boerma). He lives in Utrecht with Madeleine Bergers and their son, Philip.