Diploma Thesis

An analysis of commonly seen pathologies of the hand and foot in children

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Zusammenfassung

Hintergrund Pathologien der Hände und Füße bei Kindern stellen ein seltenes Krankheitsbild dar. Es ist von Bedeutung, die häufigsten angeborenen Anomalien zu kennen, um eine adäquate Therapie gewährleisten zu können. Das Ziel dieser Studie ist es, die häufigsten Pathologien der Hand und des Fußes und deren Therapie zu analysieren, sowie die Behandlungsresultate zu erfassen.

Methoden Alle Patienten mit Pathologien der Hände oder der Füße, die während einer 11-jährigen Periode (2000-2010) an der Universitätsklinik für Kinder- und Jugendchirurgie in Graz behandelt wurden, wurden der Van Schonhooven Klassifikation zugeordnet. Die häufigsten Pathologien waren der schnellende Daumen/Finger in 100 Fällen und die Polydaktylien in 86. Diese wurden genauer analysiert.

Ergebnisse Der schnellende Daumen/Finger betraf beide Geschlechter nahezu gleich häufig. In den meisten Fällen war der Daumen (85%) betroffen, in 83% der Fälle erfolgte eine Spaltung des A1-Ringbandes. Komplikationen nach dem chirurgischen Eingriff traten bei 10 Patienten auf, wobei bei 3 Patienten eine erneute chirurgische Intervention aufgrund eines unvollständig gespaltenen Ringbandes nötig war.

32 Patienten mit 53 ulnaren Polydaktylie, 29 radiale Polydaktylien bei 26 Patienten und 39 Patienten mit 53 Polydaktylien des Fußes konnten analysiert werden. Von 32 Patienten mit ulnarer Polydaktylie wurden 27 operativ behandelt, dabei mussten 9% einem erneuten chirurgischen Eingriff, aufgrund überschüssiger Narbenbildung, unterzogen werden. 96% der Patienten mit radialer Polydaktylie wurden operiert. Ein Patient musste aufgrund einer Deviation der distalen Phalanx erneut operiert werden. In den 39 Fällen von Polydaktylie des Fußes war der 5. Strahl in den meisten Fällen betroffen. 35 Patienten wurden operiert, postoperative Komplikationen waren selten, in 3 Fällen war eine erneute Operation aufgrund einer Achsabweichung nötig.

Schlussfolgerung Alle operativen Ergebnisse zeigen gute funktionelle und kosmetische Resultate. Auftretende Komplikationen können meist konservativ therapiert werden.

Schlagworte: **Schnellender Daumen, schnellender Finger, ulnare Polydaktylie, radiale Polydaktylie, Polydaktylie des Fußes**

Abstract

Background Congenital anomalies of the hand and foot in children are seldom seen. However it is important to be aware of the most common seen deficiencies to assure an appropriate therapy. Their etiology still remains unknown. However endogenic and exogenic factors are discussed to play a role in the development of these anomalies. The aim of this study is to analyze the treatment and outcome of the most commonly seen deficiencies of the hand and foot.

Methods Data were retrospectively collected at the Department of Pediatric and Adolescent Surgery at the Medical University of Graz. All patients treated with congenital deficiencies of the hand and/or foot during an 11 years period (2000-2010) were included. The patients were assigned to the different entities classified by Van Schoonhoven. The most commonly seen deficiencies of the hand and foot were the trigger thumb/finger in 100 cases and polydactylies in 86 cases.

Results Trigger thumb/finger affected boys and girls almost equally. Trigger thumb (85%) was more common than trigger finger. 83% underwent surgical splitting of the A1-pulley. In 10 patients minor complications occurred, a reoperation was needed in 3 cases due to an incomplete splitting of the A1-pulley. 53 ulnar polydactylies in 32 patients, 26 patients with 29 radial polydactylies and 39 patients with 53 polydactylies of the foot were analyzed. Out of 32 patients with ulnar polydactyly 27 underwent surgery, 9% needed a reoperation due to hypertrophic scaring. 96% of the cases with radial polydactyly underwent surgery, one patient needed a reoperation due to a deviation of the axis. Out of 39 cases with polydactyly of the foot 35 underwent surgery. Complications were rare. In 3 cases a second operation was performed due to a deviation of the axis.

Conclusion The postoperative outcome of these anomalies is excellent regarding the functional and cosmetic appearance. The occurring complications are minor and in most of the cases conservative treatment can be realized.

Keywords: **Trigger thumb, trigger finger, ulnar polydactyly, radial polydactyly, polydactyly of the foot**

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1 Introduction

Congenital deficiencies of the hand and foot are rare conditions occurring approximately in 1 of 600 newborns. They are made up of a spectrum of clinically and radiologically different entities. In the etiology of congenital deficiencies of the hand and foot, endogenic factors (hereditary factors) or exogenic factors (e.g. thalidomide) may play a role. For the majority, however, the etiology is unknown. In some cases familial occurrence is described. The majority of congenital deficiencies of the wrist and hand are nowadays corrected surgically during the first years of life (1).

The most commonly seen deficiencies of the hand and foot are the trigger finger and/or thumb and polydactyly. Congenital deficiencies of the limbs imply restrictions for the concerned child, the appearance of the hand or foot is altered, the functionality is often limited and the social impact cannot be neglected. Therefore, treatment of these anomalies is essential in order to enable a normal and stable function of the concerned hand or foot.

1.1 Embryology

The development of the limbs begins at the 4th week of gestation with complex genetic and molecular signals being responsible for the differentiation of the upper and lower limbs. The limb buds occur around the 5th week of gestation and consist of a mesenchymal core covered by an ectoderm cap. The occurrence of the lower bud in the lumbal and upper sacral area is delayed about two days compared to the upper bud, appearing in the area of the cervical segments (C5-TH1). The ectoderm thickens to an apical ectoderm ridge at its distal end. This ridge acts as a signaling center and induces further growth of the buds from proximal to distal by release of important growth factors. The mesenchymal tissue formats, differentiates and migrates starting from proximal to distal (2–6).

Around the 6th week of gestation the hand and foot plates are formed by circular constriction of the distal limb buds. The separation of the digital rays is due to apoptosis. At this level five segments arise and further growth of the rays is induced. The zone of polarizing activity, a mesenchymal area, acts as a signaling center and induces the further growth of the buds in the anterior-posterior axis (3–5,7).

During the 7th week of gestation, the limbs rotate in different directions, the upper limb 90° laterally, so that the thumb lies laterally, and the lower limb around 90° medially, so that the big toe lies medially. The skeleton of the limb begins to form at the end of the 8th week of gestation by enchondral ossification. At the end of the 12th week primary ossification centers appear in every long bone and advance from the diaphysis to the end of the hyaline cartilage (3–5,7).

By the time of birth the diaphysis of the long bones is completely ossified, only the epiphyses are still made of cartilage. After a short time ossification centers start to appear in the epiphysis. Between the epiphyseal and diaphyseal ossification centers an epiphyseal plate of cartilage is preserved. This plate plays an important role for length growth of the bones. At the end of the length growth of the bones, between the age 18 and 23 years, the epiphyseal plate ossifies.

The development of the mesodermal musculature of the limbs commences around the 8th week of gestation (3,4,8).

Congenital limb anomalies are due to gene mutations during the embryonic growth caused by either genetic or environmental factors. The Swanson (2) classification is used to describe the different groups of anomalies occurring during embryogenesis. **Table 1** shows the Swanson classification of limb malformations.

Congenital Deformities
Failure of formation of parts
Failure of differentiation of parts
Duplication
Overgrowth
Undergrowth
Congenital constriction ring syndrome
General skeletal anomalies

Table 1: Classification of limb malformations according to Swanson (2)

1.2 Anatomy of the hand

1.2.1 Osteology

The hand consists of the carpus (wrist), metacarpus and the phalanges.

The wrist contains eight carpal bones, four are arranged in a proximal row and articulate with the lower arm; four in a distal row and articulate with the metacarpus distally and the proximal carpal bones proximally. The bones of the proximal row are named the navicular, lunate, triangular and pisiform, those of the distal row are named the greater multangular, lesser multangular, capitate and hamate (8–10).

The metacarpus is made up of five long bones, numbered consecutively from laterally to medially. Each bone consists of a body, a base and a head.

There are three phalanges for each finger and two for the thumb (8–10).

1.2.2 Joints

The wrist-joint connects the wirst to the lower arm. The joint is a condyloid articulation and is formed by the distal end of the radius, the articular ulnocarpal disc and the navicular, lunate and triangular bones. The joint is strengthened by the palmar and dorsal radiocarpal and the ulnar and radial collateral ligaments (8–10).

The flexor tendons cover the wrist-joint on the palmar side and the extensor tendons dorsally. The condyloid articulation permits all movements except rotation. Movements of the wrist are always combined movements of the proximal and distal row (8–10).

The midcarpal joint consists of the proximal and distal carpal bones articulating with each other. The socket is made of the navicular, lunate and triangular bone articulating with the hamate and capitate. On the radial side the navicular articulates with the greater and lesser multangulars. The midcarpal joint is also strengthened by various ligaments (palmar and dorsal ligament; ulnar and radial collateral ligament) and allows flexion/extension and abduction/adduction.

The first carpometacarpal joint connects the first metacarpal with the greater multangular and is a joint of reciprocal reception. The articulation is saddle-shaped allowing great freedom of movement. The permitted movements of this joint are flexion/extension, abduction/adduction as well as circumduction and opposition (8–10).

The other four metacarpal bones articulate with the carpus in an arthrodial joint. Palmar, dorsal and interosseous ligaments link the bones to each other. The motion in these joints is restricted and only slight gliding of the articular surfaces are possible (8,9). The four metacarpal bones also articulate with each other and are connected by dorsal, palmar and interosseous ligaments.

The metacarpus is linked to the phalanges by metacarpophalangeal joints. These joints are condyloid articulations and are formed by the rounded heads of the metacarpal bones and the cavities of the proximal ends of the first phalanges. Only the thumb represents another type of joint. A palmar and two collateral ligaments as well as deep transverse metacarpal ligament strengthen the joint. The possible movements in these joints are flexion/extension, adduction/abduction and circumduction (8,9).

The interphalangeal articulations are the joints between the proximal, medial and distal phalanges and are hinge-joints. Each joint has a palmar and two collateral ligaments and the dorsal aponeurosis of the extensor muscles. In some cases a sesamoid bone may exists on the thumb. The possible movements are extension and flexion of the fingers (8–10).

1.2.3 Ligaments

The ligaments of the carpus are divided into three layers (superficial, middle and deep). The superficial layer consists of the retinaculum flexorum covering the carpal tunnel, forming two sheaths on the front of the wrist; one for the flexor digitorum profundus and superficialis and the other for the pollicis longus. The retinaculum extensorum consists of 6 compartments for the extensor tendons on the back of the wrist. The middle layer comprises the palmar, dorsal and collateral ligaments deriving from the radius, ulna or from the wrist. The deep layer connects the carpal bones with palmar, dorsal and interosseous ligaments (8,9).

The palmar aponeurosis embeds the muscles of the palm and consists of a central, a lateral and a medial portion.

The dorsal aponeurosis is formed by the interosseous muscles and it is located on the dorsal side and connected to the periost.

On the palmar side of the digits gliding sheaths of the flexor muscles can be found. These sheaths act as a guiding device and allow the tendons to glide. The outer layer is enhanced by annular and oblique ligaments. **Table 2** shows the different pulleys and their respective location on the digit. The annular ligaments are numbered A1-A5 pulleys, the oblique ligaments C1-C3 (**Figure 1**) (8–10).

Pulley	Location
A1	Metacarophphalangeal-joint
A2	Proximal phalanx
A3	Proximal Interphalangeal-joint
A4	Middle phalanx
A5	Distal Interphalangeal-joint
C1	Between A2 and A3
C2	Between A3 and A4
C3	Between A4 and A5

Table 2: Location of the different pulleys

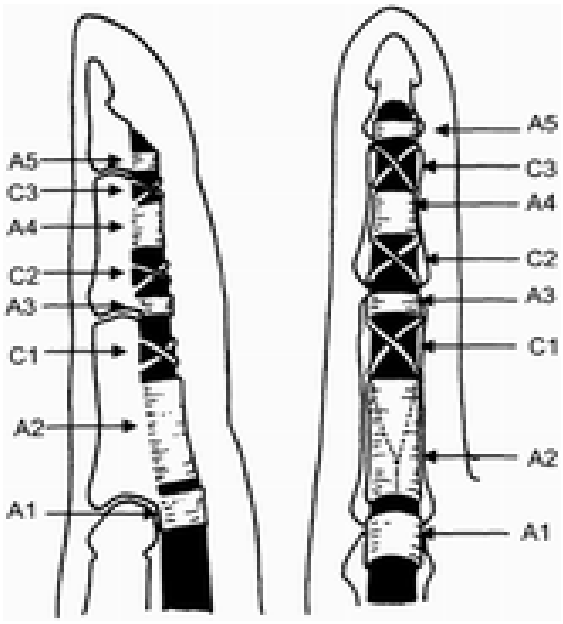


Figure 1: Location of the different pulleys on the finger;
Sagittal (left) and coronal (right) depictions of the pulley system of a typical flexor tendon (black areas) of the finger: fibro-osseous annular pulleys (A2, A4), palmar plate annular pulleys (A1, A3, and A5), and cruciate pulleys (C1, C2, and C3).
From Hauger O et al. (11)

1.2.4 Myology

The muscles of the forearm and hand are divided into a dorsal, palmar and brachioradial muscle group (8).

The dorsal antebrachial muscles form the extensor muscles and are divided into a superficial and a deep group. **Table 3** shows these two groups. The muscles of this group are innervated by the radial nerve.

Superficial group	**Deep group**
Extensor carpi radialis longus	Abductor pollicis longus
Extensor carpi radialis brevis	Extensor pollicis longus
Extensor carpi ulnaris	Extensor pollicis brevis
Extensor digitorum communis	
Extensor digiti minimi	

Table 3: The eight dorsal antebrachial muscles

The palmar antebrachial muscles form the flexor group and are divided into 4 layers.

The muscles of the first layer are:

- Pronator teres
- Flexor carpi radialis
- Palmaris longus
- Flexor carpi ulnaris

The muscle of the second layer:

- Flexor digitorum superficialis

The muscles of the third layer are:

- Flexor digitourim profundus
- Flexor pollicis longus

The muscle of the fourth layer:

- Pronator quadratus

The muscles are innervated by the median nerve, except for the flexor carpi ulnaris which is innervated by the ulnar nerve (8).

The brachioradial muscle group is divided into a superficial and a deep group. The brachioradialis, extensor carpi radialis longus and brevis are parts of the superficial layer. The deep layer consists of the supinator muscle.

The intrinsic muscles of the hand are divided into two groups, the muscles of the thumb on the radial side form the thenar eminence; the muscles of the little finger on the ulnar side form the hypothenar eminence. **Table 4** shows the intrinsic muscles of the hand (8).

Thenar eminence	**Intermediate**	**Hypothenar eminence**
Abductor pollicis brevis	Lumbricales	Palmaris brevis
Flexor pollicis brevis	Interosseous	Abductor digiti minimi
Opponens pollicis brevis		Flexor digiti minimi brevis
Adductor pollicis brevis		Opponens digiti minimi

Table 4: The intrinsic muscles of the hand

The nerval innervation of the muscles of the thenar eminence is assured by the median nerve, except for the medial head of the flexor pollicis brevis and the adductor pollicis brevis which are supplied by the ulnar nerve.

The median nerve supplies the two lateral lumbrical muscles, the two medial lumbrical muscles and all interosseous muscles are supplied by the ulnar nerve, as are all the muscles of the hypothenar eminence (8–10).

1.2.5 Blood supply

The arterial supply is guaranteed by the radial and ulnar artery. The radial artery begins at the bifurcation of the brachial artery just below the elbow; it follows the course of the radius. At the wrist it passes through the tabatière to the dorsum of the hand crossing the first and second metacarpal bones and uniting with the deep palmar branch of the ulnar artery to form the deep palmar arch. On its way to the deep palmar arch the radial artery gives rise to some branches supplying the lower arm, hand and fingers.

The ulnar artery usually is the bigger terminal branch of the brachial artery. It begins below the elbow and passes through the pronator teres muscle. From this point on it accompanies the ulnar nerve on the radial side of the M. flexor carpi ulnaris to the superficial palmar arch.

The venous drain is guaranteed by superficial and deep veins (8–10).

1.3 Anatomy of the foot

1.3.1 Osteology

The skeleton of the foot can be divided into three parts: the tarsal bones, the metatarsal bones and the phalanges.

The tarsus consists of seven bones, the talus and calcaneus are arranged in a proximal row. Four are arranged in a distal row: three cuneiform bones and the cuboid. Only the talus forms the connection to the bones of the lower limb. Between the talus and the cuneiform bones lies the scaphoid (8–10).

The metatarsus is made up of five long bones, numbered beginning from the medial side. Each bone consists of a body, a base and a head.

The phalanges of the foot are similar to those of the hand. 3 for each toe and 2 for the big toe and in some case also the little toe (8–10).

1.3.2 Joints and Ligaments

The ankle-joint connects the lower limb to the foot. The joint is a hinge articulation and allows the following movements: dorsiflexion/palmarflexion and inversion/eversion. The joint is formed by the lower ends of the tibia and fibula and the talus. The joint is strengthened by different ligaments, such as the anterior and posterior ligaments of the capsule, the deltoid ligament, the anterior and posterior talofibular ligaments, the calcaneofibular ligament and the synovial membrane (8,9).

The talus connects to the calcaneus with two different articulations, the anterior part formed by the talocalcaneonavicular articulation and a posterior part formed by the talocalcaneal articulation. The anterior part the talus articulates with the calcaneus and the scaphoid bone. The articulation is strengthened by different

ligaments and the capsule and permitted movements are inversion and eversion of the foot (8,9).

The other bones of the tarsus form strong articulations strengthened by different ligaments. The permitted movements are a gliding and sliding upon each other.
The tarsometatarsal articulations form the Lisfranc-line and are built of the three cuneiforms and the cuboid articulating with the bases of the metatarsal bones I-V. The first cuneiform articulates with the first metatarsal bones, the second and third metatarsal bones articulate with the second and third cuneiform and the cuboid articulates with the fourth and fifth metatarsal bone. The bones are connected by plantar, dorsal and interosseous ligaments (8,9).

The metatarsus is linked to the phalanges by metatarsophalangeal joints. These joints are condyloid articulations and are formed by the heads of the metacarpal bones and the proximal ends of the first phalanges. The dorsal aponeurosis and two collateral ligaments strengthen the joint. Possible movements of these joints are flexion/extension and adduction/abduction (8,9).

The interphalangeal articulations are the joints between the proximal, medial and distal phalanges and are hinge-joints. Each joint has collateral ligaments and the dorsal aponeurosis of the extensor muscles. The possible movements are extension and flexion of the toes (8,9).

The foot has arches to support the weight of the body. A longitudinal and a transverse arch of the foot are differentiated. The longitudinal arch can be divided into a medial and lateral arch. The medial arch consists of the calcaneus, talus, navicular, the three cuneiforms and metatarsal bones I-III. The calcaneus, cuboid and the fourth and fifth metatarsal form the lateral arch. The arch is supported by the tendons of the tibialis posterior and the short muscles of the sole, the plantar ligaments and the plantar aponeurosis. The transverse arch is due to the position of the calcaneus, talus and navicular bone and the cuneiforms and cuboid. It is strengthened by interosseous, plantar and dorsal ligaments, the short muscles of the foot and the tendons of the tibialis posterior and the peroneus longus (8–10).

1.3.3 Myology

The intrinsic muscles of the foot are divided into the short muscles of the sole and the short muscles of the dorsum of the foot (8).

The dorsal muscles of the foot are the extensor digitorum brevis and hallucis brevis. The dorsal muscles are supplied by the deep peroneal nerve.

The muscles of the foot sole are:

- Abductor hallucis
- Flexor hallucis brevis
- Adductor hallucis
- Flexor digitorum brevis
- Quadratus plantae
- Lumbricales, interossei
- Flexor digiti minimi brevis
- Abductor digiti minimi
- Opponens digiti minimi

The medial plantar nerve supplies the flexor digitorum brevis, the flexor hallucis brevis, the abductor hallucis, and the first lumbricalis. The other muscles of the sole of the foot are supplied by the lateral plantar nerve (8–10).

1.3.4 Blood supply

The arterial supply is guaranteed by the arteria dorsalis pedis and the medial and lateral plantar arteries. The arteria dorsalis pedis is a direct branch of the anterior tibial artery and accompanies the tendon of the hallucis longus and the deep peroneal nerve. It divides into two branches and is a part of the deep plantar arch.

The medial plantar artery is smaller than the lateral and a branch of the posterior tibial artery. It passes forward along the medial side of the foot between the abductor hallucis and flexor digitorum brevis (8,9).

The lateral plantar artery passes between the flexor digitorum brevis and the quadratus plantae. It unites with the deep plantar branch of the dorsalis pedis artery, completing the plantar arch.

The venous drain is guaranteed by superficial and deep veins, concomitant, communicating and perforating veins (8).

2 Deficiencies of the hand and foot

All patients treated with pathologies of the hand and/or foot at our Department were assigned to the different entities classified by Van Schoonhoven (12). The most commonly seen deficiencies of the hand and foot during a 11 year period were trigger thumb/finger and polydactyly. Those entities were further analyzed.

2.1 Trigger thumb/finger

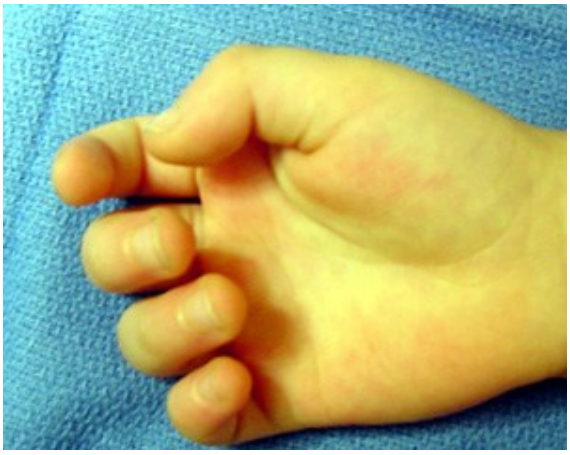


Figure 2: Depiction of a trigger thumb in a child
Data from: Ken MacKenzie Communications (13)

Pollex or digitus flexus is defined as a fixed flexion deformity of the interphalangeal joint due to a stenosing tenosynovitis of the tendons. It is one of the most common anomalies of the hand but relatively uncommon in children (14–18). The diagnosis is obtained clinically and the treatment is either surgically (14,15,19–21) or non-surgically (14,22,23).

The incidence of trigger thumb lies between 0.5 and 3 per 1,000 children (1,17). The exact etiology still remains unknown (25,26). However three possible theories have been raised: a congenital (14,21), a trauma resulting (27) or an acquired triggering during the early months of childhood (17,28–30). Recent studies have shown that the trigger finger or thumb is acquired (28).

Clinically an abnormally flexed thumb or finger at the interphalangeal joint can be seen combined with a nodule (Notta´s node (31)) palpable at the A1-pulley (16,32). Sometimes a snapping and triggering of the affected digit is present (32).

In most of the cases treatment of this abnormality is surgically by a release of the A1-pulley. The surgical procedure should not be delayed too long since developing contractures have been described (20,33,34).

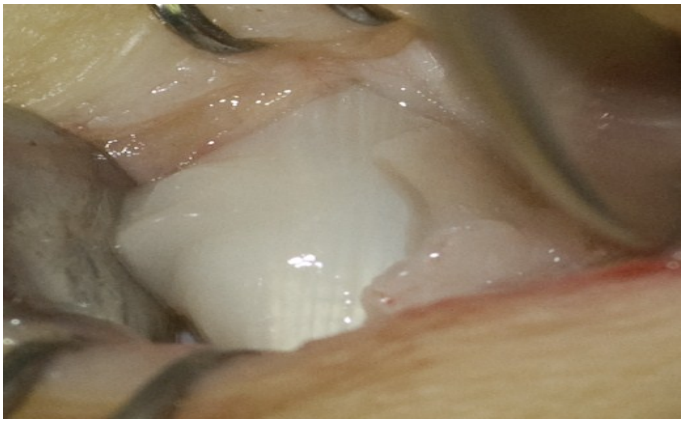


Figure 3: Intraoperative situs of a trigger thumb with a thickened tendon

Nevertheless, some studies have shown a successful conservative therapy by treating the concerned digit with a splint (22,23) or in some cases a spontaneous recovery has been observed (14,16). The goal of the treatment is to regain full range of motion at the interphalangeal joint (19,35–40).

2.2 Polydactyly

Polydactyly is defined as a congenital condition in which too many fingers or toes are present.

The extra digit can range from a small cutaneous nubbin to a fully complete and functional digit. In the hand, the digit can be located either on the ulnar (little finger) or radial (thumb) side or centrally, within the middle three fingers. On the foot the tibial, fibular or central rays can be affected. Radial and ulnar as well as tibial and fibular polydactyly are more common than central polydactyly (41). It can occur unilaterally or bilaterally.

Polydactyly is one of the most common malformations of the hand and foot. Temtamy and McKusick (42) described the polydactyly as one of the most common hand anomalies among African Americans.

The detailed etiology still remains unknown (1).

It can appear isolated, in an autosomal dominant inheritance, or associated with syndromes (1,43). **Table 5** shows a list of syndromes associated with polydactyly.

Syndromes associated with polydactylies
Ellis van Creveld-Syndrome
Apert-Syndrome
Syndactyly
Cleft hand
Trisomy
Poland-Syndrome
Bardet-Biedl-Syndrome

Table 5: Syndromes commonly associated with polydactylies

Temtamy and McKusick (42) classified polydactyly into preaxial, central and postaxial types. A patient suffering from preaxial polydactyly has a duplication of the first digit or ray. Central polydactyly involves the duplication of the three middle fingers or rays and postaxial means a duplication of the fifth digit or ray. This classification is preferably used for polydactylies of the hand.

The diagnosis is obtained clinically. Nevertheless, a radiological evaluation is necessary to determine bony involvement, the extension, the level of abnormality and to assure an adequate preoperative planning.

In all of the cases treatment consists of surgery and depends on the complexity of the deformity. The aim of the surgical treatment is to regain optimal function and appearance (1).

2.2.1 Ulnar Polydactyly

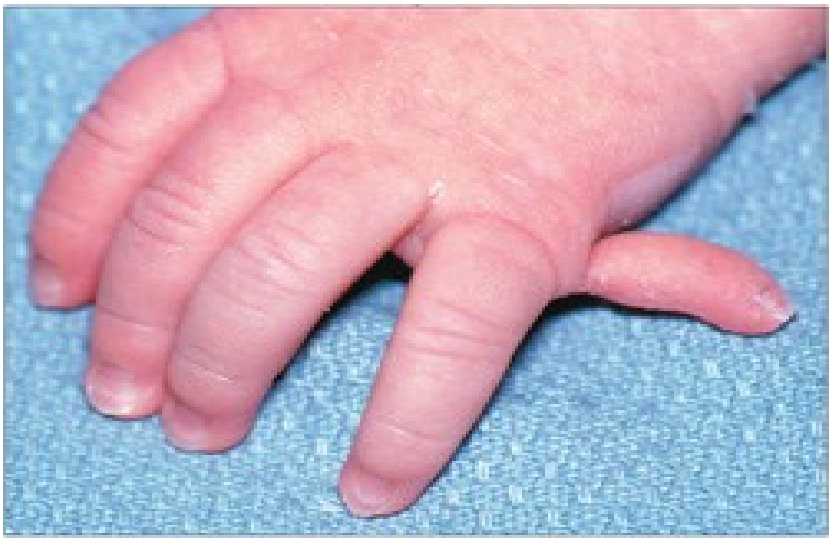


Figure 4: Depiction of ulnar polydactyly
Data from Robert P. Blereau, MD (44)

The incidence of ulnar polydactyly is much higher in African Americans than in Whites (42). Ulnar polydactyly can occur isolated, as part of a syndrome or associated with syndactyly and polydactyly of the feet (41,45).

The Stelling (46) and Turek (47) classification is used to classify ulnar polydactyly. Three types of polydactyly are differentiated: type 1 is a floating cutaneous appendage, type 2 is a partial duplication with an articulation and a shared metacarpal or phalangeal bone and type 3 shows a complete duplication of the digit. Other classifications such as the modified Rayan-Frey classification (45) are also used when referring to ulnar polydactylies.

The treatment of choice is the surgical ablation of the supernumerary digit; normally the hand function is not restricted due to the abnormality, so that the surgical treatment is used to approve the physical appearance rather than the functional (1,39,41). The surgical method depends on the extent and level of abnormality (**Table 6**).

Stelling and Turek Type	Appearance	Surgical treatment
1	Small cutaneous appendage	Surgical excision
2	Partial duplication with a common metacarpal head	Surgical ablation, collateral ligament or intrinsic muscle reconstruction if needed
3	Complete duplication	Surgical ablation, collateral ligament or intrinsic muscle reconstruction if needed

Table 6: Classification of ulnar polydactylies and preferred treatment according to Stelling (46) and Turek (47)

Complications after surgery can be chronic instability of the metacarpophalangeal joint due to an inadequate reconstruction of the collateral ligament, deviation of the little finger to the ulnar or radial side due to a suturing under too much tension. When the metacarpus is broadened and not narrowed during surgery a deformed ulnar border of the hand will remain (1,41,45,48).

2.2.2 Radial Polydactyly

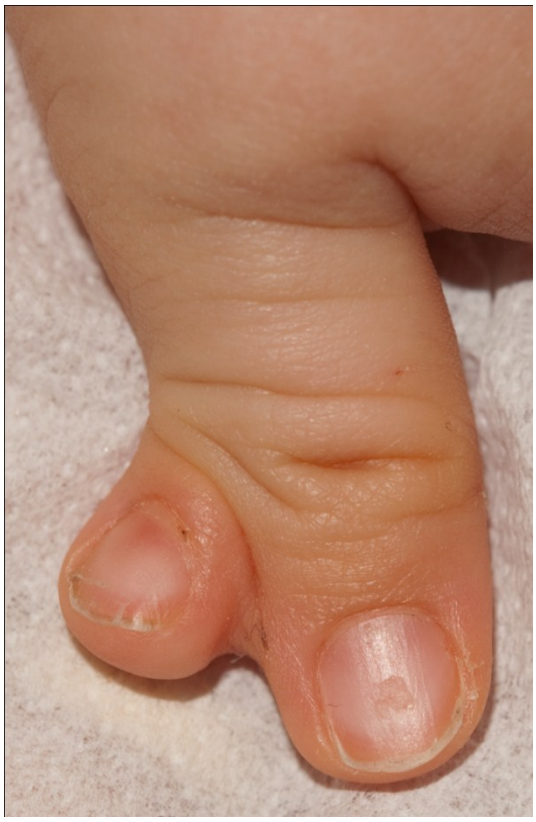


Figure 5: Depiction of radial polydactyly

Radial polydactyly (**Figure 5**) is defined as a duplicated thumb and can appear in a variety of forms ranging from a small nubbin to a fully functional thumb.

Duplication of the thumb is the most common congenital anomaly of the hand, the incidence is approximately 1 in 3000 live births (49). The cause of the duplication remains unknown, some genetic disorders are assumed to be important for the development of this anomaly (1). Triphalangism and an associated polysyndactyly are often associated with an autosomal dominant heredity (1,41). As in all the types of polydactyly several syndromes can be associated (43).

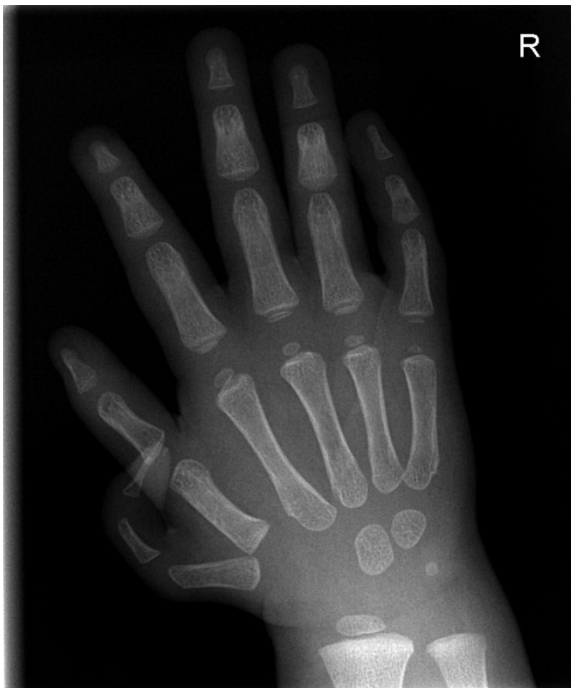


Figure 6: X-ray showing a radial polydactyly Wassel type VI

The Wassel (50) classification is used to describe the anatomy and the involved pathology of the thumb. The classification consists of six subtypes of the radial duplication and a seventh including the triphalangeal duplicated thumb, the rudimentary type of radial polydactyly is not included in this classification (1). **Figure 7** shows the Wassel classification (50) and the appearance of the different types.

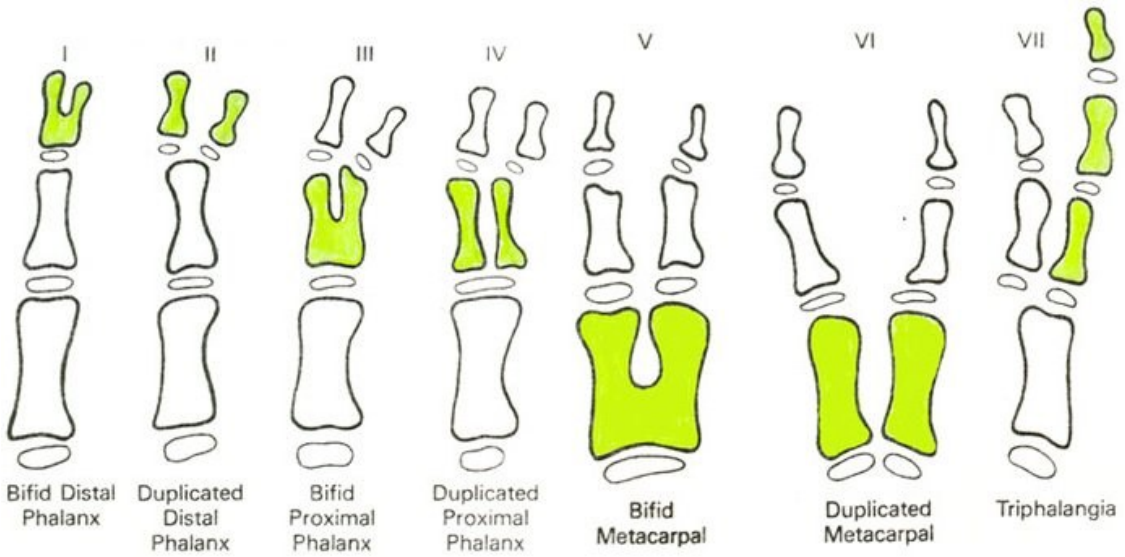


Figure 7: Wassel classification describing the anatomy and the involved pathology of the thumb From Birmingham Radiology Training Scheme *(51)*

Table 7 shows the Wassel classification (50) and the respective surgical treatment.

Type	Surgical treatment
Wassel I	Ablation
Wassel II	Ablation with ligament reconstruction
Wassel III	Resection of the radial thumb, osteotomy, fixation with a Kirschner-wire, mobilizing of the abductor pollicis brevis muscle and reattaching to the extensor pollicis longus tendon Bilhaut Cloquet (52) procedure
Wassel IV	Resection of the radial thumb, osteotomy, fixation with a Kirschner-wire, mobilizing of the abductor pollicis brevis muscle and reattaching to the extensor pollicis longus tendon
Wassel V	Resection of the radial thumb and the radial metacarpal, correction of the ulnar angulation of the ulnar metacarpal, increase of the first web space.
Wassel VI	Resection of the radial thumb and the radial metacarpal, correction of the ulnar angulation of the ulnar metacarpal, increase of the first web space.
Wassel VII	Excision of extra phalanx, arthrodesis of IP joint, pollicization, on-top plasty
Rudimentary	Surgical excision

Table 7: Wassel classification and preferred surgical treatment data adopted from (50)

Complications following surgery are chronic instability due to an ineffective reconstruction of the collateral ligament, deviation of the digit due to a suturing under too much tension, Z-deformity and metacarpal joint instability. When the metacarpus is broadened and not narrowed during surgery a deformation of the radial border will remain. Functional disabilities are rare but can appear, however the cosmetic impact often is the cause for a reoperation (1,53–57).

2.2.3 Central polydactyly of the hands

Central polydactyly means that the index, middle or ring finger is duplicated. The same condition can appear on the feet.

This condition rarely appears isolated and is mostly associated with syndactyly and other anomalies (1,7,42).

Polydactyly of the index finger is very rare; duplication of the middle finger is more commonly seen than that of the index finger. Duplication of the ring finger is most frequently seen (1,58).

This duplication is also treated surgically, but there is scarce information about the procedures used. Due to the commonly associated syndactyly the treatment is complicated and a single procedure is not sufficient in most of the cases. The syndactyly has to be separated, the accessory bone excised, osteotomy performed to correct the axial deviation and transposition of a digital ray and tendon reconstruction are necessary steps to obtain an esthetic and functional acceptable outcome (58,59).

Early onset complications include skin or fingertip necrosis. Late onset complications are scar contractures, deviation, loss of function or motion, instability of the joints and a limited growth in length of the affected finger (58).

2.2.4 Polydactyly of the foot

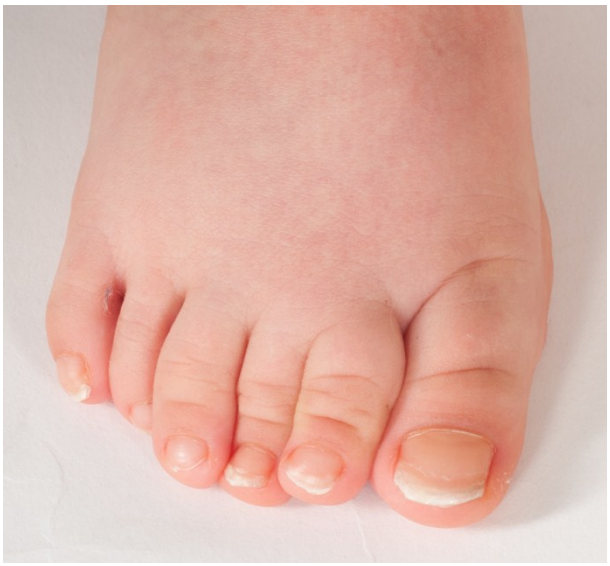


Figure 8: Depiction of fifth ray polydactyly

Polydactyly of the foot is defined as a duplication of the toe or the metatarsal (**Figure 8**).

It is the commonest congenital malformation of the foot, ethnic differences exist and as in all of the forms of polydactyly an autosomal dominant inheritance can be present. The incidence is approximately 1,7:1000 births (60,61).

In most cases the patients present because of the cosmetic impact or an existing shoe difficulty (41,62).

Different classification systems exist to describe the patterns of polydactyly of the foot, the currently used classification is the one described by Blauth and Olason (63). They classify the duplication of the toe along two axes, the longitudinal ranging from the distal phalanx to the tarsal bone; and a transverse one labeling the involved ray (I-V).

Three different types of polydactyly can be distinguished: post-axial (fifth ray), central and preaxial polydactyly (first ray) (60,61). In most of the cases the fifth ray is duplicated representing a fibular polydactyly (61).

Central duplication is rarely seen, the duplication is mostly hypoplastic. A preaxial duplication is the second most commonly seen polydactyly of the foot. Usually the phalanx is duplicated and the metatarsal appears as a block (1,60).

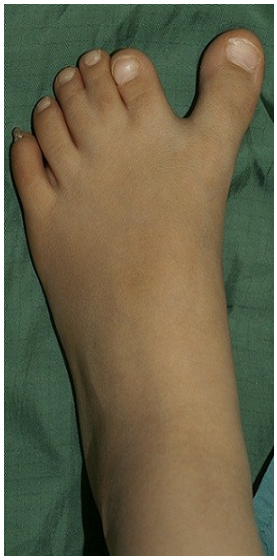


Figure 9: Depiction of first ray polydactyly

The treatment is composed of the excision of the duplicated digit, division of the capsule and reconstruction of the remaining ray; depending on the localization of the affected ray. If the first ray is affected a Kirschner-wire is used to avoid postoperative axial deviations (60–62,64).

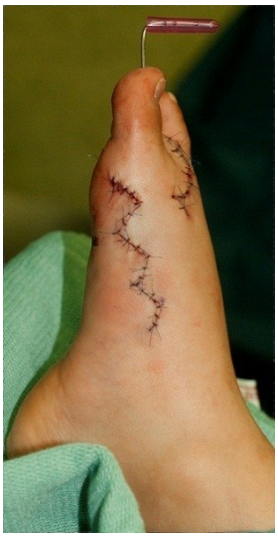


Figure 10: Depiction of a surgically corrected first ray polydactyly

Surgical methods and common complications are shown in **Table 8**.

Affected ray	Surgical method	Complications
First ray	Excision of the toe, reattachment of the abductor and adductor hallucis, trimming of the metatarsal head, if needed	Hallux varus causing pain and a shoe conflict, metatarsophalangeal joint subluxation, deformity or instability
Fifth ray	Excision of the hypoplastic toe, narrowing of the metatarsal head if needed, repairing of the joint capsule	Angular deformity, metatarsalphalangeal joint subluxation, deformity or instability
Central ray	Excision of the duplicated digit, reapproximation of the intermetatarsal ligament	Wider forefoot, metatarsophalangeal joint subluxation, deformity or instability

Table 8: Surgical method according to the affected ray and postoperative complications

3 Patients and Methods

For this study, data were retrospectively collected at the Department of Pediatric and Adolescent Surgery at the Medical University of Graz. All patients treated with pathologies of the hand and/or foot during an 11 years period (2000-2010) were included. The patients were assigned to the different entities classified by Van Schoonhoven (12) (**Table 9**).

Van Schoonhoven Classification	Number of cases
Camptodactyly	15
Syndactyly	6
Klinodactyly	41
Pollex/digitus flexus (trigger thumb)	100
Polydactyly (and thumb duplication)	86
Mirror hand	0
Macrodactyly	8
Brachydactyly	13
Thumb hypoplasia	22
Radial deficiency	no data collected

Table 9: Van Schoonhoven (12) classification and number of cases

Patients with the most common entities (trigger thumb or trigger finger and polydactyly) were further analyzed.

For trigger thumb/fingers the following parameters were collected and analyzed:

- Gender
- Date of birth
- Side and number of involved fingers
- Further diagnoses
- Age at the time of surgery
- Time of surgery
- Number of presentations (including preoperative visits)
- Complications and reoperations
- Time of hospitalization
- Outcome

For polydactyly the following parameters were collected:

- Gender
- Date of birth
- Side
- Degree of polydactyly
- Family history
- Further diagnoses
- Age at the time of surgery
- Time of surgery
- Number of presentations (including preoperative visits)
- Complications and reoperations
- Time of hospitalization
- Post-surgery treatment
- Outcome

Data were retrieved from the medical database (MEDOCS®), and analyzed using Microsoft Office Excel®.

The study was reviewed and approved by the ethics committee of the Medical University of Graz (EK23-276 ex 10/11).

4 Results

4.1 General

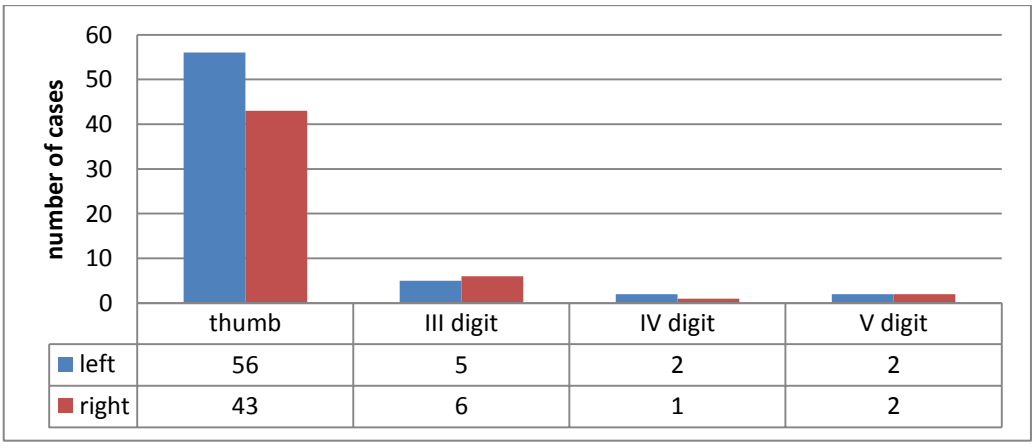
Between 2000 and 2010, a total of 186 patients were suffering from either trigger thumb or finger or polydactyly. Of those, 100 patients were treated because of trigger thumb/finger and 86 with polydactyly.

4.1.1 Trigger thumb/finger

In total, 100 patients with a total of 117 trigger thumbs and fingers were included. Out of the 100 children 51% were girls (n=51) and 49% were boys (n=49). Mean age of the patients was 3.14 years ranging from 7 months to 13 years.

There were 10 patients with bilateral thumb involvement, 75 with unilateral trigger thumb. Out of the 12 patients diagnosed with trigger finger, 4 showed a bilateral involvement. 3 patients showed finger and thumb involvement.

The involved digits included 99 thumbs, 11 middle fingers, 3 ring fingers and 4 little fingers **(Figure 11)**. Interestingly, none of the patients showed an involvement of the index finger.



	thumb	III digit	IV digit	V digit
left	56	5	2	2
right	43	6	1	2

Figure 11: Distribution of the 117 trigger fingers according to the affected fingers and concerned side

The mean age of the treated patients with trigger thumb was 3.19 years (range 7 months to 13 years), while the mean age of the patients with trigger finger was 3.6 years (range 1 to 8 years).

Table 10 shows the number and distribution of trigger fingers of all 100 patients including the children with multiple affected fingers.

	Left hand					Right hand				
Patients	**I**	**II**	**III**	**IV**	**V**	**I**	**II**	**III**	**IV**	**V**
44						X				
31	X									
10	X					X				
4			X							
3								X		
1			X					X		
1				X						
1					X					
1										X
1					X				X	X
1						X		X		
1	X					X			X	
1	X							X		

Table 10: Number and distribution of trigger fingers of 100 patients

Out of 51 girls 84% had a trigger thumb, 12% a trigger finger and 4% a trigger finger and thumb. 86% of the boys had a trigger thumb, 12% a trigger finger and 2% a trigger finger and thumb. **Figure 12** shows the gender distribution split into affected fingers.

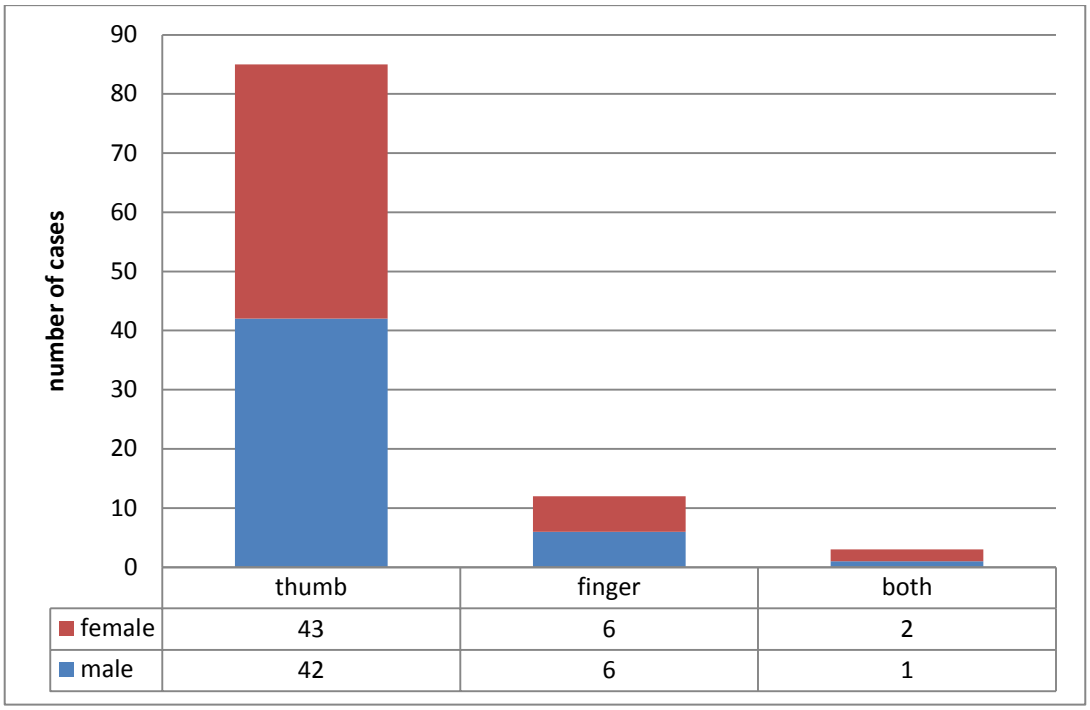


Figure 12: Gender distribution split into affected fingers

None of the treated patients showed a family history of trigger thumb or finger. Additionally, no associated abnormalities were diagnosed. 5 out of the 100 patients have been born preterm. One of those was born before the 30th week of gestation.

83% of the patients were treated surgically by splitting of the A1-pulley, 4% were treated conservatively by either splinting (n=3) and massaging (n=1). The remaining 13 patients were treated by watchful waiting (**Figure 13**).

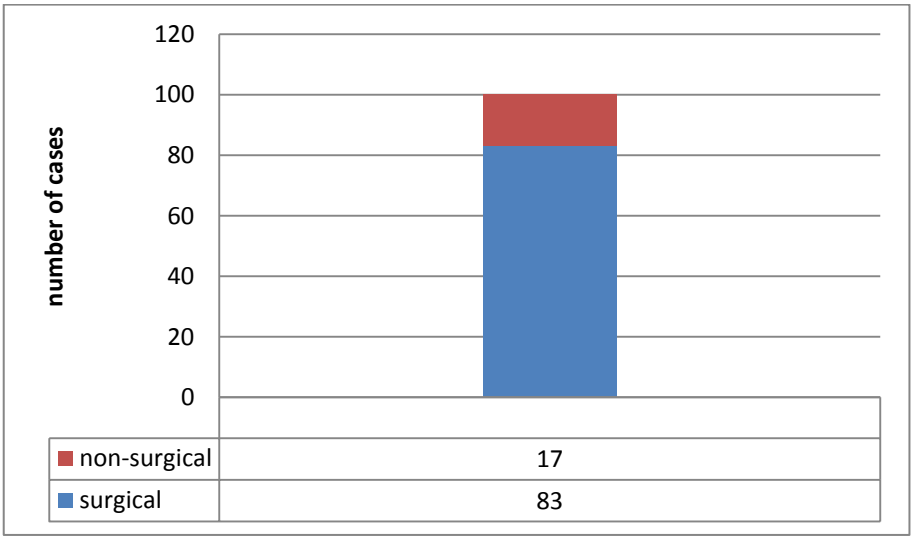


Figure 13: Treatment of 100 patients with 117 trigger thumbs/fingers.

Of the 85 patients with a single trigger thumb 88% (n=75) were treated surgically. Out of the 12 patients with single or multiple finger involvement, 42% were operated (n=5). In all of the patients with both thumb and finger involvement (n=3) splitting of the A1 pulley was performed.

Figure 14 shows the treatment according to the affected fingers.

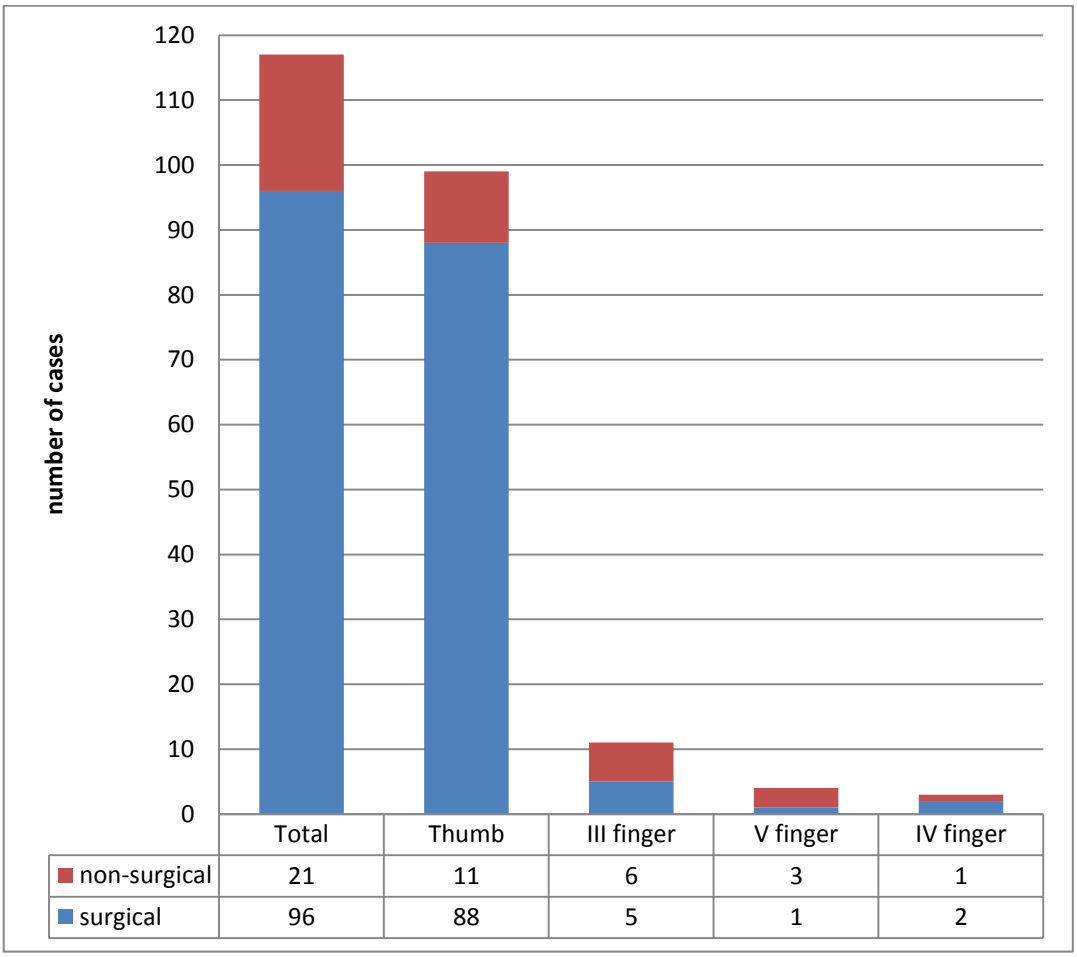


Figure 14: Treatment according to the affected fingers

The mean age at surgery was 3.1 years ranging between 7 months and 13 years.

The surgical treatment took place on a day-surgical basis in 89% of the cases, 4 patients stayed for one night (5%) and one patient (1%) stayed for 2 nights after the surgical treatment (**Figure 15**). The patients staying for more than one night had preexisting conditions or other operations performed during the same anesthesia.

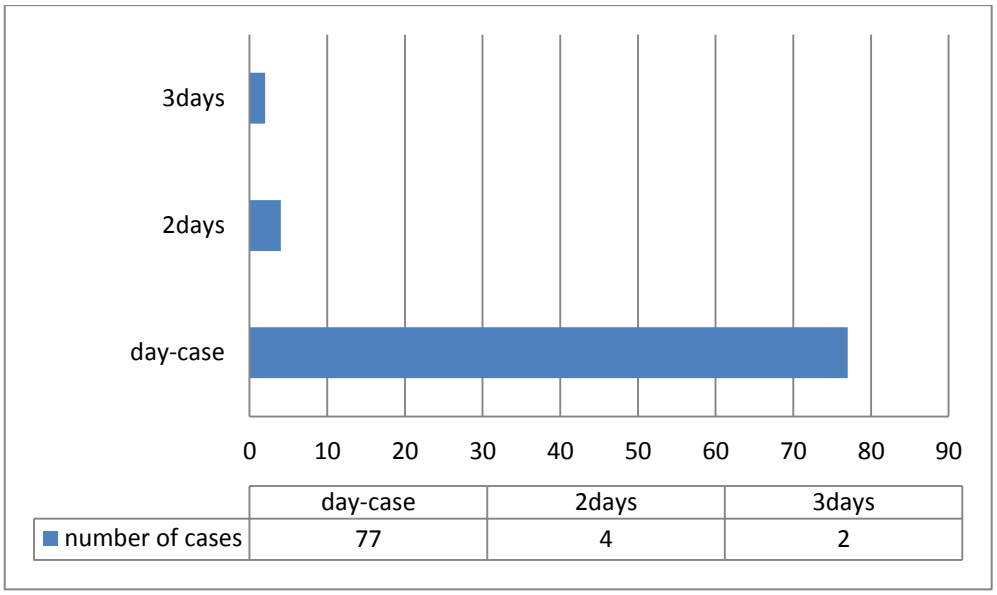


Figure 15: Time of hospitalization after the surgical treatment

The time of surgery was divided into three periods:

- < 20 min
- 20-30 min
- > 30 min

	<20 min	20-30 min	>30 min
Number of patients [n]	7	23	34
Percentage [%]	11	36	53

Table 11: Time of surgery for 64 patients suffering from a single trigger finger
(For 8 patients data were not available)

In 11 cases more than one trigger finger was treated surgically during the same anesthesia and the time of the surgical intervention took more than 30 minutes. Each surgically treated patient presented himself in average three times to our outpatient department (range 1 to 12).

Three patients showed minor complications (superficial wound infection and delayed wound healing) necessitating treatment with oral antibiotics, no additional surgery was needed for these patients. Three patients needed revision surgery due to an inadequate release of the A1-pulley. Three patients had a 15° reduced interphalangeal joint motion and one patient showed a hypertrophic scar.

Complications	Number of patients	Percentage
Superficial wound infection	3	3.6%
Reoperation	3	3.6%
Reduced interphalangeal joint motion	3	3.6%
Hypertrophic scar	1	1.2%

Table 12: Complications following treatment of trigger thumbs and fingers

4.1.2 Polydactyly

A total of 86 patients were diagnosed with polydactyly of the upper and/or lower limbs.

Our study included the analysis of:

- 53 ulnar polydactylies in 32 patients
- 29 radial polydactylies in 26 patients
- 53 polydactylies of the foot in 39 patients.

The distribution of the different forms of polydactyly is shown in **Table 13**.

Ulnar Polydactyly	Radial Polydactyly	Foot	Combined
22	24	29	11

Table 13: Distribution of the different forms of polydactyly

4.1.2.1 Ulnar Polydactyly

53 ulnar polydactylies in 32 patients were treated during the last eleven years. In 22 cases only the hands were involved and in 10 cases another polydactyly affecting the limb was present (**Table 14**). Of the 32 patients 20 were male (63%) and 12 female (37%).

	Left			Right		
Patients	**Ulnar**	**Radial**	**Foot**	**Ulnar**	**Radial**	**Foot**
14	X			X		
7	X					
5	X		X	X		X
1	X		X	X		
1	X	X	X	X	X	X
1				X		
1	X		X			X
1			X	X		
1			X	X		X

Table 14: Number and distribution of ulnar polydactylism in 32 patients

In most of the cases the ulnar polydactyly occurred bilaterally (66%). In 11 cases of unilateral occurrence (44%) it was more frequent on the left (73%) than on the right (27%) side.

The 53 hands with ulnar polydactyly were classified according to the Stelling (46) and Turek (47) classification **(Table 15)**.

Type	Appearance	Number of polydactylies	Percentage
1	Floating cutaneous appendage	41	77.4%
2	Partial duplication	8	15.1%
3	Complete duplication	4	7.5%

Table 15: Classification according to Stelling (46) and Turek (47)

A family history of polydactyly was present in 6 patients and absent or unknown for the remaining patients.

Five patients presented with a syndrome; two with Ellis Creveld, two with a dysmorphic syndrome and one with Jeune Syndrome.

Other anomalies were documented in 15 patients (47%). These are shown in **Table 16**. Most of the musculoskeletal anomalies were additional hand or foot deficiencies such as polydactylism or syndactylism.

Associated anomaly	Number of patients
Musculosceletal	9
Cardiac	3
Preterm babies	2
Gastrointestinal	1

Table 16: Associated anomalies of 32 patients with ulnar polydactyly

27 patients were treated with surgical ablation under general anesthesia, 4 in local anesthesia and one patient by ligation.

The mean age at the time of operation was 0.72 (range, 0-10) years.

The surgical treatment took place on a day-surgical basis in 8 (25%) cases, 17 (53.1%) patients stayed 2 to 4 days in the hospital; 3 (9.4%) patient stayed for 5 to 6 days and 4 (12.5%) patients for more than a week following the surgical treatment. In average the patients stayed for 3.8 days.

Each surgically treated patient presented himself 5.9 times mean to our department (including preoperative, operative and postoperative visits).

The outcome of the patients is shown in **Table 17**. In total, 9% of all patients needed a reoperation due to a hypertrophic scaring. Reversible wound dehiscence, swelling of the soft tissue and a hypoplastic left little finger were seldom seen.

Outcome/Complications	Number of patients	Percentage	Classification
Hypertrophic scaring	3	9%	I
Swelling of soft tissue	1	3%	III
Deviation of the axis	1	3%	I
Hypoplastic left little finger with restriction of motion	1	3%	II
Wound dehiscence	1	3%	I

Table 17: Complications and outcome of treated patients with ulnar polydactyly

4.1.2.2 Radial Polydactyly

In total 29 radial polydactylies in 26 patients were treated during the last eleven years. In 92% of the cases the patients were diagnosed with radial polydactyly alone and in 8% of the cases another polydactyly was present.

Out of the 26 children 54% were boys (n=14) and 46% were girls (n=12).

Three patients had bilateral involvement, unilateral involvement was more often seen on the right (n= 16) than on the left (n=7) hand **(Table 18)**.

	Left			Right		
Patients	**Ulnar**	**Radial**	**Foot**	**Ulnar**	**Radial**	**Foot**
16					X	
7		X				
1		X			X	
1	X	X	X	X	X	X
1		X	X		X	X

Table 18: Number and distribution of radial polydactylism in 26 patients

The Wassel classification (50) was used to classify the different types of radial polydactyly. Results are shown in **Table 19**.

Type	Appearance	Number of hands	%
Wassel I	Bifid distal phalanx	None	None
Wassel II	Duplicated distal phalanx	11	38
Wassel III	Bifid proximal phalanx	2	7
Wassel IV	Duplicated proximal phalanx	7	24
Wassel V	Bifid metacarpal	2	7
Wassel VI	Duplicated metacarpal	3	10
Wassel VII	Thumb duplication with triphalangism	1	3
Rudimentary	Small appendage with a narrow pedicle	3	10

Table 19: Wassel classification (50) I-VII; rudimentary types are not included in the Wassel classification

One of the treated patients showed a family history of radial polydactylism in a first-degree relative.

One patient was diagnosed to have an associated dysmorphic syndrome, 6 had associated deficiencies of the hand and foot and 4 had associated renal, gastrointestinal and cardiac abnormalities.

Out of 26 patients 25 underwent surgery. One patient was lost for follow-up.

The mean age at the time of operation was 1.16 (range 0-7) years.

The surgical treatment took place on a day-surgical basis in 4% of the cases, 15 patients stayed 2 to 4 days in the hospital; 9 patient stayed for 5 to 6 days after the surgical treatment. In average the patients stayed for 3.9 days **(Figure 16**).

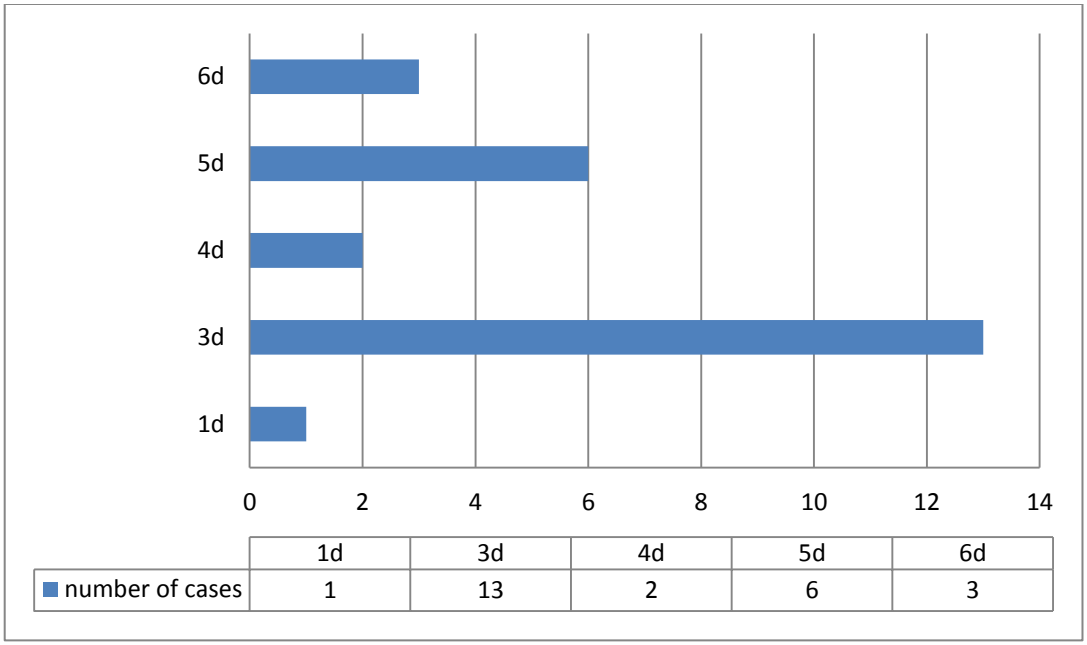


Figure 16: Time of hospitalization after the surgical treatment

15 patients were treated with a splint after surgery (60%).

	Wassel II	Wassel III	Wassel IV	Wassel V	Wassel VI	Rudimentary
Splint	4	2	4	2	1	1

Table 20: Splint treatment according to the Wassel classification (50)

The time of surgery was divided into four periods:

- < 60 min
- 60-120 min
- 120-180 min
- > 180 min

	<60 min	60-120 min	120-180 min	>180 min
Number of patients [n]	2	3	6	7
Percentage [%]	11	17	33	39

Table 21: Time of surgery for 18 patients suffering from radial polydactyly
(For 7 patients data were not available)

Each surgically treated patient presented himself in average 8 times to the hospital (including preoperative, operative and postoperative visits).

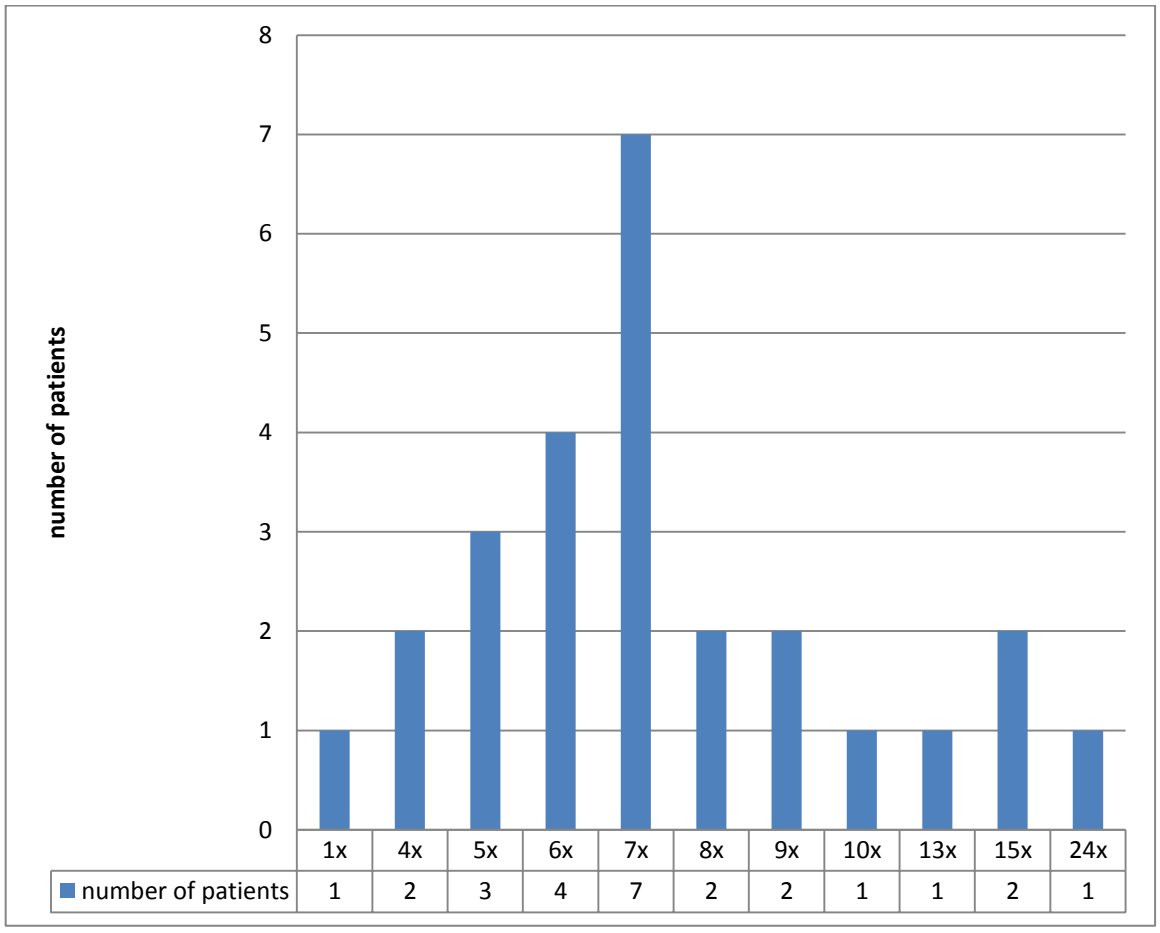


Figure 17: Number of presentations at our Department

One patient developed a joint instability, in one patient the pincer grip was not possible, in another the pulp to side grip was not possible, one patient showed a loss of strength in the surgically treated finger, another patient showed a restriction of motion and 7 patients showed a mild deviation, but none needed a second surgical treatment **(Table 22)**.

Only one patient needed a reoperation due to a deviation of the distal phalanx.

Complications/outcome	Number of patients	Percentage	Wassel classification(50)
Mild radial deviation	5	20%	II; IV; V
Mild ulnar deviation	2	8%	II, V
Pincer grip n.p.	1	4%	II
Pulp to side grip n.p.	1	4%	V
Reoperation	1	4%	
Decreased strength	1	4%	III
Joint instability	1	4%	VI
Restriction of motion	1	4%	II

Table 22: Complications following treatment of polydactylism

4.1.2.3 Polydactyly of the foot

In total 39 patients with 53 polydactylies of the foot (tibial, fibular and/or central) were included. Only the feet were involved in 29 patients (74%); in 10 patients (26%) the hands and feet were involved. Out of the 39 patients, 22 were female and 17 male.

Mean age by the time of the first presentation was 1.35 years ranging from 0 to 9 years.

In 25 patients the polydactyly occurred unilaterally (65%). Bilateral involvement was seen in 14 patients (35%). Unilateral involvement was more frequent on the left (18 cases) than on the right (7 cases) side. **Table 23** shows the exact distribution of the polydactylies of the foot.

	Left				Right			
Patients	**First ray**	**Fifth ray**	**Central**	**Hand**	**First ray**	**Fifth ray**	**Central**	**Hand**
3	X							
2	X				X			
2	X			X	X			X
1					X			
12		X						
5						X		
4		X				X		
3		X		X		X		X
2		X		X		X		
1		X						X
1		X		X		X	X	X
1			X					
1							X	
1			X	X				X

Table 23: Distribution of polydactylies of the foot

The polydactylies were classified according to Blauth and Olason (63).

The most common level of duplication was the metatarsal (60%), followed by proximal phalanx duplication (22%). We did not observe any patient with a tarsal duplication. The four patients that were not classified presented with rudimentary skin or nail appendages, which are not listed in the Blauth and Olason´s(63) classification (**Table 24)**.

Patterns of polydactyly	Number of patients	Percentage
Distal phalanx	5	9%
Middle phalanx	3	6%
Proximal phalanx	20	38%
Metatarsal	21	40%
Tarsal	None	None
Other	4	8%

Table 24: Classification according to Blauth and Olason (63)

The fifth ray was affected in 70% (n=28), followed by the first ray in 20 % (n=8), the second ray in 5 % and the fourth ray in 5%.

Out of the 28 cases with involvement of the fifth ray, 7 patients presented with an additional polydactyly. In 6 of the 8 patients with involvement of the first ray the polydactyly was the only present deficiency. Half of the cases with central ray affection were isolated and in the remaining half the deficiency was associated with another polydactyly (**Table 25**).

Affected ray	Isolated	Associated polydactyly	Total
First ray	6	2	8
Fifth ray	21	7	28
Central	2	2	4

Table 25: Distribution of 39 patients, one patient showed a first-ray-central-hand associated anomaly

More than half of the treated patients showed other associated features as shown in **Table 26**.

Associated anomaly	Cases
Polydactyly of the hand	10
Syndactyly	6
Clinodactyly	2
Bardet-Biedl syndrome	1
Ellis-Creveld Syndrome	1

Table 26: Associated anomalies of 39 patients with polydactyly of the foot

7 patients showed a family history of polydactylism of the foot or hand.

35 of the 39 patients underwent surgery. One patient moved back to France, one patient didn´t show up for the surgical appointment, for another no data was available and one patient with a fifth ray polydactyly with a small duplicated nubbin, did not show a shoe conflict, so surgical treatment was not necessary.

Polydactyly	Total	Surgery	Mean age in years
All polydactylies	39	35	1.5
Tibial polydactyly	8	7	2.25
Fibular polydactyly	28	26	1
Central polydactyly	4	2	2

Table 27: Treatment and age at operation of 39 patients with polydactyly of the foot according to the affected ray

The mean age of the patients at the time of surgery was 1.5 years (range; 0-9).

All the surgically treated patients except one received a cast or splint after surgery, for at least 2 weeks.

The surgical treatment took place in a day-surgical environment in 2 case, 17 patients stayed 2 to 4 days in hospital; 15 patient stayed for 5 to 6 days in hospital after the surgical treatment and 2 patients stayed one week in hospital after their surgical treatment. On average the patients stayed in hospital for 4.5 days (**Figure 18**).

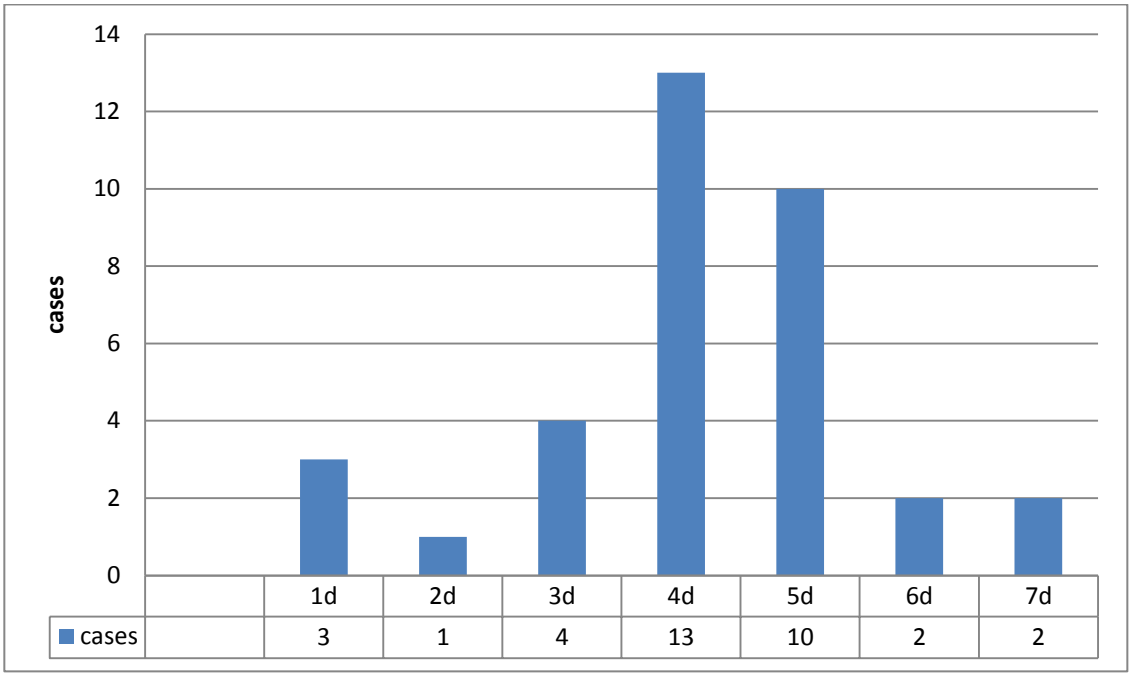


Figure 18: Time of hospitalization after the surgical treatment

Each reviewed patient presented in average 8.3 times to the hospital for a clinical examination.

Complications were rare (**Table 28**). Three patients required subsequent surgery due to a deviation of the axis. One patient developed fever as a sign of infection and was treated with oral antibiotics. All patients showed a good cosmetical result. No major functional or shoe conflict was documented.

Complications/Outcome	Number of patients	Affected ray
Rotated toe	2	First ray
Reoperation	3	Fifth ray
Varus/Valgus-position	2	First ray
Small nubbin	1	Fifth ray
Mild divergence	2	First ray
Instability of the ligament	1	First ray
Hyperextended position of the small toe	1	Fifth ray
Hypoplastic middle and endphalanx	1	Fifth ray

Table 28: Complications and outcome after surgical treatment

5 Discussion

Deficiencies of the hand and foot are rare conditions and are nowadays corrected surgically during the first years of life. Trigger thumb and polydactyly rank among the most commonly seen deficiencies of the hand and foot in children. These deficiencies imply restrictions for the concerned child; the appearance of the hand or foot is altered, which attracts psychological issues, due to a not normal locking hand or foot. For this reason the deficiencies should be corrected before school time, to prevent negative psychosocial effects on the child.

Therefore the aim of this study was to analyze our experiences with the most commonly seen deficiencies of the hand and foot in children.

5.1 Trigger thumb/finger

Notta (31) was the first who described the condition of a trigger finger. Nevertheless, trigger thumb is a relatively rare condition seen in infancy and childhood and the etiology still remains uncertain even if different theories are proposed by different authors (14,28,29,65): a congenital, a trauma and an acquired triggering during early months of childhood.

100 patients with 117 trigger fingers have been treated at our Department; most of the patients were treated by splitting the A1-pulley. The surgical treatment at our Department shows excellent functional results, some minor complications such as superficial wound infection, reduced interphalangeal motion and hypertrophic scar were seen, but no long term treatment was needed.

Our data show that the trigger finger/thumb is one of the most commonly seen anomalies of the hand in children. Our female to male ratio (51:49) was almost equal as it is described in most of the published reports (15,25,29,30).

The distribution according to the concerned digit showed that the thumb was affected in the majority of the cases (85%). Other published data such as Ryzewicz et al.(35), Weilby (65), De Smet et al. (66) and Moon et al. (30) found a similar predominance of the thumb.

Trigger finger or thumb usually occur unilaterally (15,25,29,65). In our series a bilateral involvement was found in 14% (thumbs and fingers). The right side alone was affected in 48 patients (including thumb and fingers) compared to 38 affected patients for the left side. This finding is confirmed by most of the published reports (29,40,65,67).

Trigger thumb was identified in one child at birth. However, the real age of onset is nearly impossible to determine because symptoms often remain uncertain or undetected due to a minor deformity. In some cases the thumb is held in undetected flexion for a long time while injuries may draw attention to the condition (29). However, the time of onset can be set approximately during infancy or early

childhood (18,67).

The treatment of trigger digit is discussed by several authors. Some authors prefer a conservative treatment by splinting (22,23), others recommend a surgical release and some others wait until the age of three for a spontaneous recovery (14). In 83% of our patients surgical release of the A1-pulley was performed; the mean age at the time of surgery was 3.1 years (ranging from 7 months to 13 years). Dinham et al. (14) reported that surgery should not be delayed beyond the age of 4 years due to an increase in developing contractures. In our study and also in other published reports (25,68) no developing contracture was observed at a surgical treatment after the age of 4 years.

Surgery was performed under general anesthesia in all of the cases. In 3 patients minor complications occurred and 3 patients needed a revision due to an incomplete A1-pulley splitting. No digital nerve or artery was injured and no other major complications occurred. Our results of the surgical release are concordant to those of other published studies.

In conclusion, the results of the present study confirm that the results of surgical release of trigger thumb and/or finger in children are excellent. Nevertheless, conservative treatment can be considered in young children and in children presenting with a short duration of symptoms.

5.2 Polydactyly

Polydactyly is a congenital deficiency occurring on the hand or foot. Among the 86 treated patients at our Department, 25% were diagnosed with ulnar polydactyly, 28% with radial polydactyly, in 34% the foot was affected (tibial, central, fibular, or mixed) and in 13% a mixed polydactyly was seen. Radial polydactyly is more frequently seen than ulnar polydactyly at our Department. This finding is concordant with Bates et al.(7), who published a dominance of the radial over the ulnar type of polydactyly.

Racial differences can be observed (1). While radial polydactyly is more common in the caucasian or oriental population, ulnar polydactyly more frequently occurs in the black population (1).

Interestingly, a cutaneous appendage or a duplicated distal phalanx was the most common finding of paediatric hands, while the metatarsal was duplicated in most of the cases of polydactylies of the foot.

Nearly all our patients were treated surgically, dependent on the affected finger; extent of the bony and tissue involvement and the level of the abnormality. In general, treatment had an excellent functional as well as aesthetic outcome, the complication rate was low, and only minor complications occurred (1,7).

5.2.1 Ulnar Polydactyly

There are not many reports evaluating the outcome of ulnar polydactyly. Most of the papers analyse a uniform population (Middle Eastern, Afro-Americans, and Caucasians). Therefore, our data cannot be compared to the published data in terms of racial differences since in our report a mixed population was included.

Ulnar polydactyly of the hands was present in a total of 32 patients with 53 hands; consequential more common than radial polydactyly. We reported a 1.7:1 male predominance in our study, accordant with Mellin (69), Rayan and Frey (45), Stelling (46) and Turek (47). In most of the cases no familial heritance existed, with 19% of our patients showing a positive family history. Assuming that most of our patients are of Caucasian origin, this finding can be compared with those of Rayan and Frey (45), Simmons (70), Watson et al.(71) who describe only sporadic familial associations of ulnar polydactyly.

The polydactyly was limited to the hands in 22 patients. Both hands and/or feet were affected in 10 patients. In cases of a unilateral occurrence the left side was affected more common (73%) than the right (34%). Rayan and Frey (45), Simmons (70) and Watson et al. (71) also noted a predominance of the left hand. We reported a bilateral occurrence in 66% of the affected hands, in the African form of polydactyly a similar bilateral occurrence is reported, only the Caucasian form is different, in this form the majority of the polydactylies is unilateral (48).

We used the Stelling (46) classification to describe the different morphological appearances of ulnar polydactyly, unfortunately this classification is rarely used in the literature. Therefore, a comparison is difficult to make. In our study population a duplication of soft tissue was most frequently observed, followed by a duplication of the phalanges. This results are similar to those reported by Al-Qattan et al. (48) and Rayan and Frey (45).

Al-Qattan et al. (48) published data about ulnar polydactyly and their clinical features in a native Saudi population. They analyzed a uniform population of 84 Saudis, and documented features like a positive family history, associated

syndromes, sex, unilateral/bilateral, right/left hand and the type of polydactyly, based on clinical data and radiographic features. Out of 84 patients 53 were females, in 11% a positive family history was documented, 6% showed associated syndromes. In 50 cases the polydactyly occurred unilateral, with a dominance of the left hand (35 cases). They used the modified Rayan-Frey classification (45) to evaluate the different types of polydactyly. This classification is slightly different from the Stelling classification (46) used in our series. The Rayan-Frey classification (45) is divided into 5 types, ranging from a small soft tissue nubbin without bone, duplication with a separate sixth metacarpal to other cases, including polysyndactyly and a triplication of the little finger. The commonest type was type II (pedunculated digit), followed by type III, I, V and IV.

Rayan and Frey (45) reviewed 148 patients with ulnar polydactyly. The population was slightly mixed (African Americans, Caucasians and Native Americans) with a dominance of African Americans (103 patients). The following features were collected: types of polydactyly, patterns of involvement, associated anomalies, treatment and outcome. In 20 patients hand and feet were involved, 5 showed a mixed radial and ulnar polydactyly. Ulnar polydactyly was more frequent among males. Among African Americans bilateral occurrence of polydactyly was more common. If unilateral the polydactyly was more frequent on the left side. The most common pattern of involvement was type II (pedunculated digit), followed in order of frequency by type III, V, I and IV. 5 patients showed associated syndromes. In 71%, the treatment was ligation in the nursery and complications such as tender or unacceptable nubbins and infection appeared in 23.5%.

Many syndromes associated with ulnar polydactyly are described in the literature (1,70). However, in our study only 5 polydactylies were seen as a part of a syndrome. Other anomalies associated with ulnar polydactyly (70) like musculoskeletal deficiencies were diagnosed in our study with syndactyly being the most common one (48).

5.2.2 Radial Polydactyly

At our Department 29 patients were treated with radial polydactyly during the last 10 years. In about a tenth of the cases another polydactyly affecting the feet or the hand was present. The sex ratio showed a female:male ratio of 1.16:1. Bilateral radial polydactyly was rarely seen; in most of the unilateral cases the right hand was predominant (1,50,56).

We used the Wassel (50) classification to rank the different types of radial polydactyly. The published literature reported Wassel type IV to be the most common type (1,56,72,73); however Wassel type II was the most frequent type in our series (38%) followed by Wassel type IV (24%), the least common was Wassel I (no patient), this is concordant with Al-Qattan (72). In the British series published by Naasan and Page (73) Wassel type II was most common, followed by type IV and III.

Yen et al. (56) published data of 34 patients with 36 thumbs, Wassel type II (12 cases) was most common followed by type IV (11 cases), type VI (1 case) was least common. The cases compromised 10 female and 24 male patients; a dominance of the right hand was noticed.

Al-Qattan (72) analysed the distribution of polydactyly of the thumb in a middle eastern population. In total 196 patients, with 228 hands were reviewed. The most common type was Wassel type IV (33.8%) and the least common Wassel type I (0.4%). 26% did not fit into the Wassel classification, including 18 cases of rudimentary duplication, 2 cases of triplication and 5 with symphalangism. The left hand was affected in 84 cases, versus the right in 80 cases. 32 bilateral cases were available; a positive family history was present in 18 cases.

Naasan and Page (73) analyzed 43 cases of radial polydactyly. Wassel Type II was the most common, followed by IV and II, the least common was Wassel type I. No cases of rudimentary duplication, triplication or symphalangism were documented.

All patients except one were treated with surgery, in our study the mean age at the time of operation was 1.13 years and the average time of hospitalization was 3.9 days. Yen et al. (56) published a mean age of the patients at the time of operation of 2.8 years.

In general surgical reconstruction of this deficiency should be completed before the age of 5 (34,41) due to social consequences and to prevent negative psychosocial effects to the child and parents (34,74,75).

Cetik et al. (76) published data of 10 male adults with radial polydactyly. All 10 patients were admitted with social consequences of the cosmetic problems resulting from the polydactyly, patients were reporting social isolation and the need to hide their hands. Nevertheless the functional improvement is also a reason for early reconstruction of the anomaly. Some authors (34,74) suggested that angular deformities may develop and treatment may be difficult and results worse, but Cetik et al. (76) demonstrated that the reconstruction of polydactyly in adults has functionally satisfactory results.

60% needed a splint therapy after the surgical treatment to prevent a deviation of the axis, and furthermore a reoperation due to a deviation.

Mostly, minor complications occurred, nevertheless one patient needed a second surgical treatment due to a deviation of the distal phalanx.

Our results after the surgical treatment of polydactyly closely resemble previous reports in order of the occurrence of complications and the outcome (56,77).

5.2.3 Polydactyly of the feet

In total 39 patients diagnosed with polydactyly of the foot were treated in most of the cases surgically (35 cases). The duplication was classified and analyzed according to the affected toe. Our results closely resembled previous reports (60–62), presenting a dominance of fifth ray polydactylies (70%), followed by the first ray. The occurrence was mostly unilateral, with a higher frequency of the left foot (18 cases).

The Blauth and Olason (63) classification was used to classify the different entities. The most common level of duplication was the metatarsal (60%) followed by proximal phalanx (22%) duplication. Our studies showed a difference according the level of occurrence compared with Morely et al.(61), who described the duplication of the proximal phalanx as the most common followed by metatarsal duplication. On the other hand Bader et al. (60) showed a metatarsal duplication at the most common in fifth ray polydactyly, followed by a partial duplication and metatarsal duplication as most common in first ray polydactyly followd by proximal phalanx duplication.

More than half of our patients showed associated anomalies, with polydactyly of the hand being the most common one. Similar results are reported by several other authors (61,62,78).

Compared to other studies the cases of a positive family history of polydactylies of the foot in 18% of our patients are lower compared to values of 32% reported by Morely et al.(61), and higher compared to values of 10% reported by Watanbe and coworkers (62).

The vast majority (90%) underwent surgery with an average age of 18 months. Morley et al. (61) reported in his study of 25 patients an average age of 29 months at time of operation. We noted a difference in the age at the time of operation according to the different forms of polydactylies; first ray polydactyly 2.25 years, fifth ray 1 year and central polydactylism 2 years. The operation should be done at this age due to the resulting shoe difficulties, pressure on the foot, difficulties in

finding and fitting into shoes, all these resulting difficulties may play a role in the walking process of the child (60). The cosmetical impact should not be neglected.

In our series complications were rarely seen, 3 patients with polydactyly of the fifth ray needed a revision due to a deviation of the axis. Our results resembled closely those of Morley et al.(61), showing no difficulties in shoe fitting and walking. Bader et al. (60) described a worse outcome in patients affected with first ray polydactyly, in our series this could not be confirmed.

First ray polydactyly requires a surgical intervention due to cosmetical and functional problems and the operation should not be underestimated. All in all the results are excellent. Nevertheless, a surgical intervention should be well considered in fifth ray polydactyly if no major shoe difficulty and no abnormal cosmetic exists according to Bader et al. (60).

6 Conclusion

Congenital deficiencies of the hand and foot a rare conditions. In most of the cases the diagnosis can be obtained clinically.

100 patients were diagnosed clinically with trigger thumb, 83% underwent surgery with excellent results. 3 patient needed a reoperation due to an incomplete A1-pulley splitting, minor complications were rare and a conservative management sufficient. Exact knowledge of the anatomy can prevent reoperations. Nevertheless, conservative treatment can be considered in young children and in children presenting with a short duration of symptoms.

86 patients with polydactyly were diagnosed clinically. Radiological evaluation was needed to plan the surgical procedures and to evaluate the involvement of bone and soft tissue. A careful evaluation of skin, nail, bone, ligament and musculoskeletal elements is mandatory for planning the optimal treatment.
Surgical reconstruction leads too satisfactory cosmetic and functional results. Nevertheless minor complications can occur.

Detailed knowledge of the different morphological, clinical and radiological appearances and classifications systems is essential for the treatment of the patients.

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8 Appendix Vote of the ethics committee

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VOTUM

gültig bis 25.03.2012

EK-Nummer:	23-276 ex 10/11
Studientitel:	Long term outcome of congenital deficiencies of the wrist and hand or foot
Prüfer:	Dr. Georg Singer Univ.Klinik für Kinderchirurgie
Sponsor: *)	MedUniv. Graz
Ansprechpartner:	Stephanie Thein, im Hause,
CRO:	-

*) Antragsteller

Die o.a. Studie wurde von der Ethikkommission erstmals im 'expedited Review' am 18.03.2011 behandelt. Die Ethikkommission ist zu folgendem Schluss gekommen:

Es besteht kein Einwand gegen die Durchführung der Studie in der vorliegenden Form.

Kommissionsmitglieder, die für diesen Tagesordnungspunkt als befangen anzusehen waren und daher gemäß Geschäftsordnung an der Entscheidungsfindung und Abstimmung nicht teilgenommen haben: keine

Zur Beurteilung vorliegende Dokumente:

Dokumente eingegangen am 08.03.2011, begutachtet im 'expedited Review' am 18.03.2011	
Antragsformular	20.02.2011
Originalprotokoll 1.1	07.03.2011
Informed Consent Form 8-14 Jahre 1.1	07.03.2011
Informed Consent Form Elterninformationsblatt 1.1	07.03.2011
Dokumente eingegangen am 20.03.2011 (in der nächsten Begutachtung mitbegutachtet)	
✓ Antragsformular	20.03.2011
✓ Originalprotokoll 1.2	18.03.2011
Informed Consent Form Elterninformation 1.2	18.03.2011
Informed Consent Form 8-14 Jahre 1.2	18.03.2011
Dokumente eingegangen am 24.03.2011, begutachtet im 'expedited Review' am 25.03.2011	
✓ Informed Consent Form 8-14 Jahre 1.3	24.03.2011
✓ Informed Consent Form Elterninformation 1.3	24.03.2011

Die Ethikkommission geht – rechtlich unverbindlich – davon aus, dass es sich weder um eine klinische Prüfung nach AMG noch nach MPG handelt.

Das Votum der Ethikkommission berührt in keiner Weise die alleinige Verantwortung der Prüferin / des Prüfers / der Prüfer für die ordnungsgemäße Durchführung der Studie unter Einhaltung aller einschlägiger gesetzlicher Bestimmungen und Richtlinien.

Weiters machen wir darauf aufmerksam, dass der Kommission unverzüglich zu melden sind:

- Abweichungen vom Protokoll aus Sicherheitsgründen oder Protokolländerungen

- Änderungen, die das Risiko der Teilnehmer/-innen erhöhen oder die Durchführung der Studie wesentlich beeinflussen

- Mutmaßliche unerwartete schwerwiegende Nebenwirkungen - SUSARs (AMG-Studien ab 1.5.2004) oder schwerwiegende unerwünschte Ereignisse - SAEs (andere Studien)

- Jegliche Information über sonstige Umstände, die die Sicherheit der Teilnehmer/-innen oder die Durchführung der Studie beeinträchtigen können

Dieses Votum gilt für ein Jahr ab dem Datum der Ausstellung. Bei längerer Studiendauer ist rechtzeitig vor Ablauf der Gültigkeit des Votums ein Zwischenbericht vorzulegen (Berichtsformular), um eine etwaige Verlängerung zu erlangen.

Graz, 25. März 2011

Univ.Prof.DI Dr.Peter H. Rehak
Vorsitzender

Univ.Prof.DDr.Hans-Peter Kapfhammer
Stv. Vorsitzender

Achtung: Bitte bei allen das Projekt betreffende Schreiben oder telefonischen Anfragen die EK-Nummer angeben!

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Rechtsform: Juristische Person öffentlichen Rechts gem. Universitätsgesetz 2002. Information: Mitteilungsblatt der Universität und www.medunigraz.at. DVR-Nr. 210 9494.
UID: ATU 575 111 79. Bankverbindung: Bank Austria Creditanstalt BLZ 12000 Konto-Nr. 500 948 400 04, Raiffeisen Landesbank Steiermark BLZ 38000 Konto-Nr. 49510.