ECRONICON OPEN ACCESS

EC PAEDIATRICS
Review Article

Overview for Congenital Deformities of Hand in Children

Nguyen Ngoc Hung*

Surgical Department, Vietnam National Hospital for Pediatric, Ha Noi Medical University, Vietnam

***Corresponding Author:** Nguyen Ngoc Hung, Surgical Department, Vietnam National Hospital for Pediatric, Ha Noi Medical University, Vietnam.

Received: March 07, 2025; **Published:** March 26, 2025

Abstract

Congenital hand differences encompass a range of structural abnormalities that impact the function, appearance, and psycho-social domains of affected children. Continued advances in the understanding and treatment of these differences continue to shape management. Over the years, new developments have been made in the areas of molecular genetics, non-invasive treatments, surgical techniques, and outcome measures in some of the most common congenital hand differences. Applying these advances in the understanding and management of congenital hand differences will help surgeons achieve the best possible outcomes for these children.

Keywords: *Congenital Hand Deformities; Trigger Thumb; Bifid Thumb; Radial Club Hand; Syndactyly*

Introduction

Congenital hand defects are abnormalities of the hand that develop before birth and congenital hand defects usually occur between the fourth and eighth weeks of embryonic development [1]. Failure to convert the arm bud cells into the upper limb can result in an abnormal upper limb shape/function that is present at birth. Congenital hand defects are also known as- congenital hand anomalies [2], congenital hand malformations, congenital hand conditions, and congenital hand problems. Some causes are genetic while others are influenced by the environment and the rest are unknown.

Congenital hand or finger abnormalities that are present at birth. Any type of birth defect can become a challenge for a child as he or she grows. Hand defects can be particularly disabling when severe because children learn to interact with the environment through the use of their hands. Types of malformations include extra fingers and thumbs, fused fingers, underdeveloped or overdeveloped hands, fingers and arms, and missing fingers and thumbs or bones in the arms.

Early consultation with a hand surgeon is an important part of treatment. If reconstructive surgery is not possible, there are a variety of prosthetic devices that can be used to increase function. More than half of congenital malformations can be detected in utero. Early surgical correction is crucial to achieving good long-term outcomes with low surgical morbidity and mortality.

Great strides have been made in the past 50 years in the diagnosis and management of congenital malformations, formerly known as congenital anomalies. Formally fatal conditions can now be treated with success rates exceeding 90 percent. However, improvements in

care have been largely confined to high-income countries (HICs), despite the fact that many malformations can be corrected with simple surgeries (Chirdan, Ngiloi, and Elhalaby 2012). If surgery is the neglected stepchild of global health (Farmer and Kim 2008), then pediatric surgery is the unborn child.

Incidence

Two Scandinavian studies provide insight into the epidemiology of congenital upper limb malformations, data on which are often limited in the literature. Ekblom., *et al*. [3] reported an incidence of 21.5/10,000 live births (a total of 562 patients with malformations), with undifferentiated malformations being the most common, followed by duplications and formation malformations. Koskimies., *et al*. [4] reported 419 cases with an incidence of 5.25/10,000 live births in Finland. Radial insufficiency was the most common and 60% had additional congenital anomalies. The difference in incidence is due to the fact that Koskimies., *et al*. considered only reduced defects (formation defects, underdevelopment, and hanging amputations). Ekblom., *et al*. included most of the congenital anomalies classified by the International Federation of Societies for Surgery of the Hand.

Interestingly, if only nonformation, underdevelopment, and stenosis were considered, the incidence of 4.9/10,000 live births was similar to that reported in Finland. Differences between registries, including data recording and evaluation, also play a role in these differences.

Hand anatomy

There are few structures in human anatomy as unique as the hand. The hand must move to position the fingers and thumb. Adequate strength forms the basis for normal hand function. The hand must also be coordinated to perform fine motor tasks accurately. The structures that form and move the hand must be properly aligned and controlled for normal hand function to occur.

The important structures of the hand can be divided into several categories. These include:

- Bones and joints
- Ligaments and tendons
- Muscles
- Nerves
- Blood vessels.

The front or palmar surface of the hand is called the palmar surface. The back of the hand is called the dorsal surface.

Bones and joints

Anatomically, the hand is defined as the area of the upper limb distal to the wrist.

The base of the hand contains 8 bones, each of which is called the carpal bones. The palmar surface of each hand contains 5 metacarpal bones. The fingers contain the phalanges.

The skeleton of the hand consists of 27 bones that can be divided into three groups:

- Carpals: Including 8 wrist bones.
- Metacarpals: Including 5 metacarpals.
- Phalanges: Including 14 phalanges.

Figure 1: *Hand anatomy.*

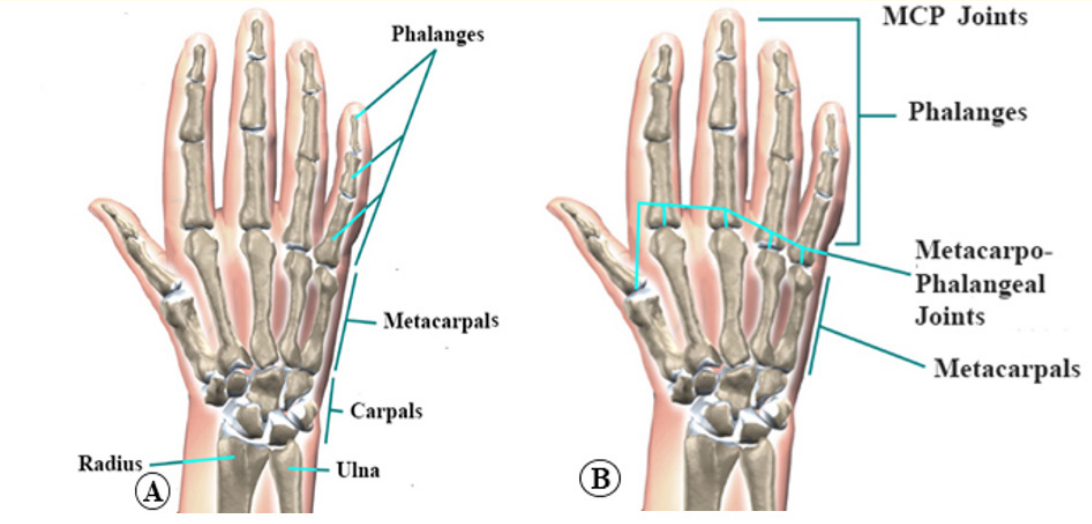


Figure 2A and 2B: *Hand bone.*

There are 27 bones in the wrist and hand. The wrist itself contains eight small bones, called carpals. The carpal bones join with two bones of the forearm, the radius and ulna, to form the wrist joint. Further into the palm, the carpal bones join with the metacarpals. There

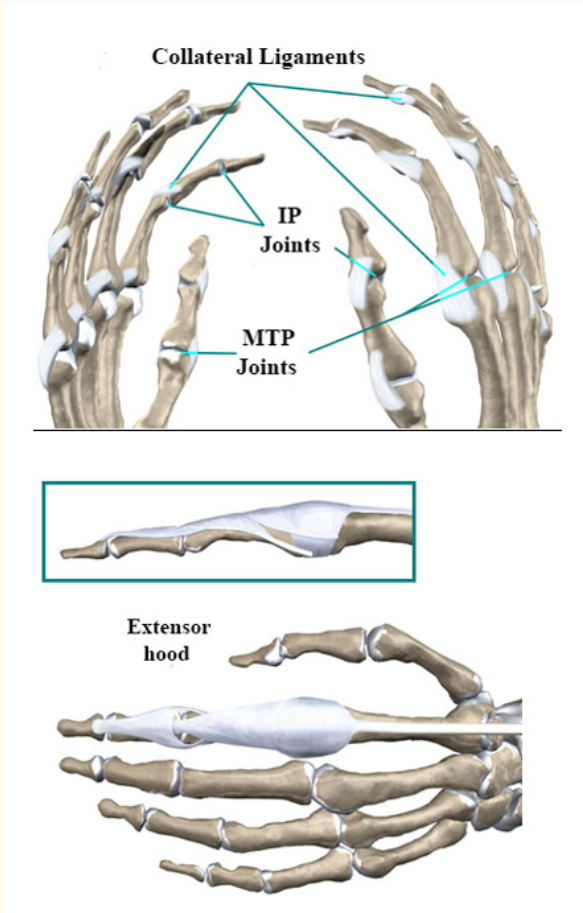
are five bones of the hand that make up the palm. One metacarpal bone joins each finger and thumb. Small bone shafts called phalanges line up to form each finger and thumb.

The main knuckle joints are formed by the connections of the knuckles to the metacarpals. These joints are called the metacarpophalangeal joints (MCP joints). The MCP joints act as a hinge when you bend and straighten your fingers and thumb.

The three knuckles in each finger are separated by two joints, called the interphalangeal joints (IP joints). The joint closest to the MCP joint (the metacarpophalangeal joint) is called the proximal IP joint (PIP joint). The joint near the end of the finger is called the distal IP joint (DIP joint). The thumb has only one IP joint between the two phalanges of the thumb. The IP joints of the fingers also act as hinges when you bend and straighten your fingers and thumb.

The joints of the hand, fingers, and thumb are covered at the ends with articular cartilage. This white, shiny material has a rubbery consistency. The function of articular cartilage is to absorb shock and provide an extremely smooth surface to facilitate movement. There is articular cartilage basically everywhere two bone surfaces move together or articulate.

Ligaments and tendons



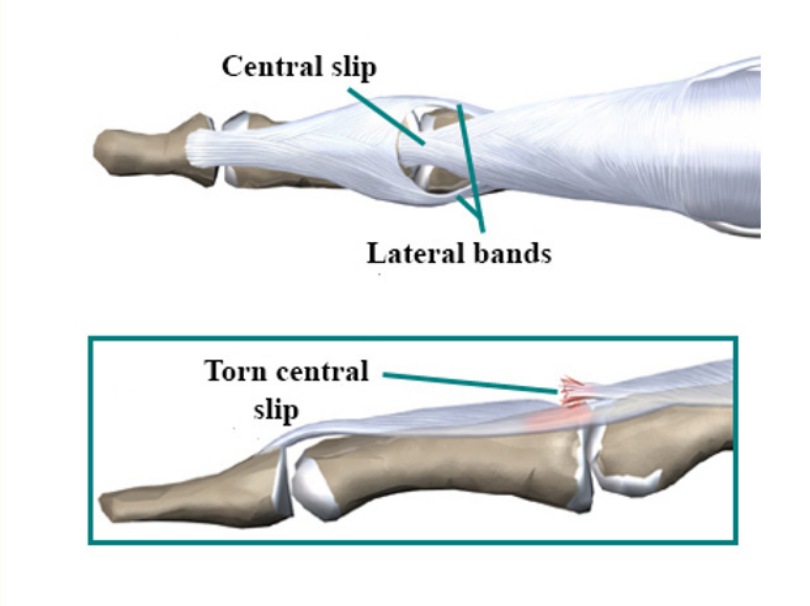


Figure 3: *Ligaments and tendons.*

Ligaments are tough bands of tissue that connect bones to each other. Two important structures, called collateral ligaments, are located on either side of each finger and thumb joint. The function of the collateral ligaments is to prevent abnormal sideways bending of each joint.

At the PIP joint (the middle joint between the distal phalanx and the DIP joint), the strongest ligament is the metacarpal plate. This ligament connects the proximal phalanx to the middle phalanx on the palm side of the joint. The ligament tightens when the joint is straightened and keeps the PIP joint from bending too far backward (hyperextension). Finger deformities can occur when the metacarpal plate becomes loose due to disease or injury.

The tendons that allow each finger joint to straighten are called the extensor tendons. The extensor tendons of the fingers begin as muscles that originate on the back of the forearm bone. These muscles move toward the hand, where they eventually connect with the extensor tendons before crossing over the back of the wrist joint. As they move into the fingers, the extensor tendons become extensor caps. The extensor caps flatten to cover the top of the finger and send out branches on each side that connect to the bones in the middle and end of the finger.

The place where the extensor tendons attach to the middle phalanx is called the central gliding joint. When the extensor muscles contract, they pull on the extensor tendons and straighten the finger. Problems occur when the central gliding joint is damaged, as can happen with a tear.

Muscles

Many of the muscles that control the hand start at the elbow or forearm. They run along the forearm and cross the wrist and hand. Some control only the flexion or extension of the wrist. Others affect the movement of the fingers or thumb. Many of these muscles help position and hold the wrist and hand while the thumb and fingers grasp or perform fine motor actions.

Most of the small muscles that control the thumb and little finger start at the wrist bone. These muscles connect in a way that allows the hand to grasp and hold. Two muscles allow the thumb to move across the palm, an important function known as thumb opposition.

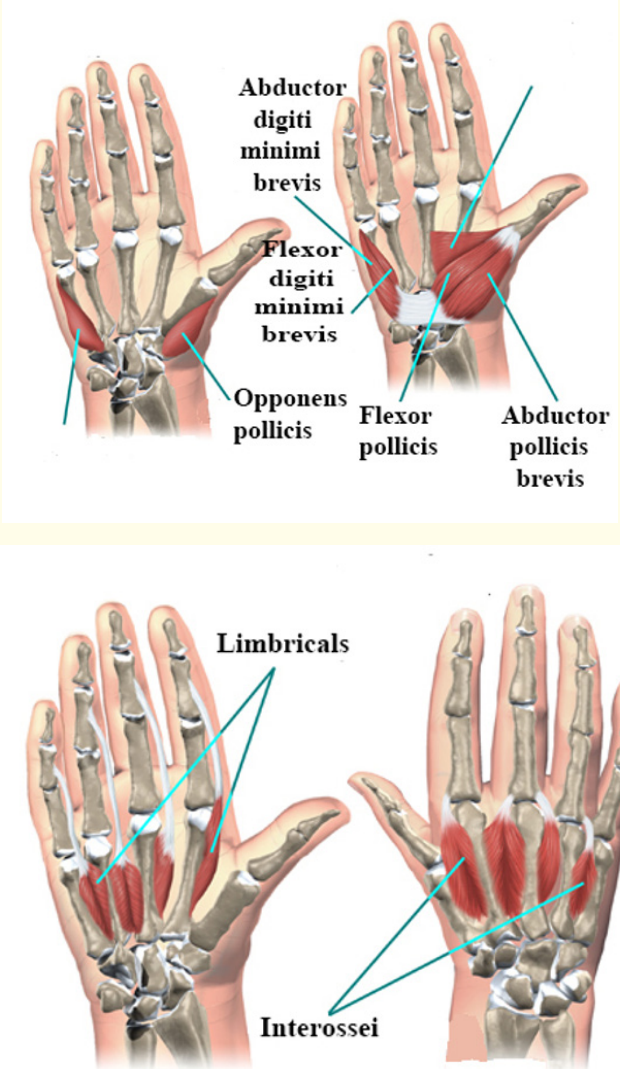


Figure 4: *Musles of hand.*

The smallest muscles originating in the wrist and hand are called intrinsic muscles. Intrinsic muscles guide the fine movements of the fingers by positioning and stabilizing them during hand activities.

Nerves

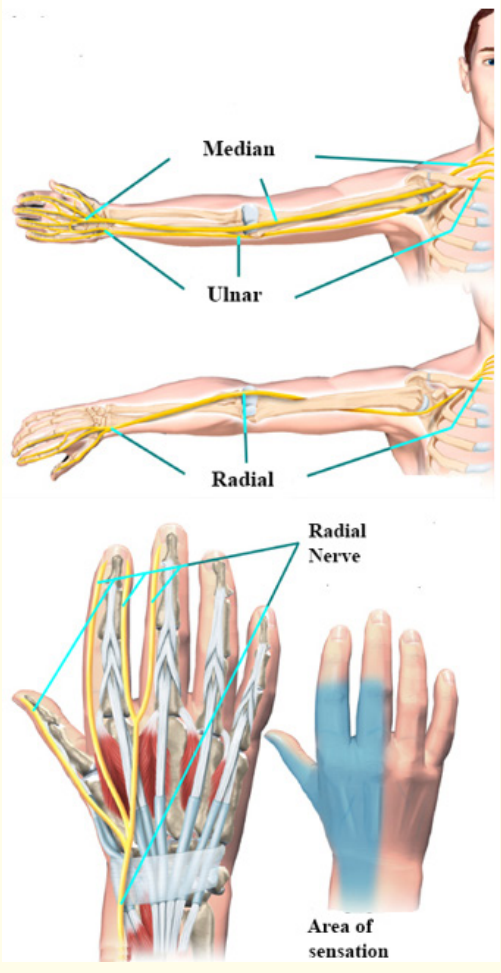
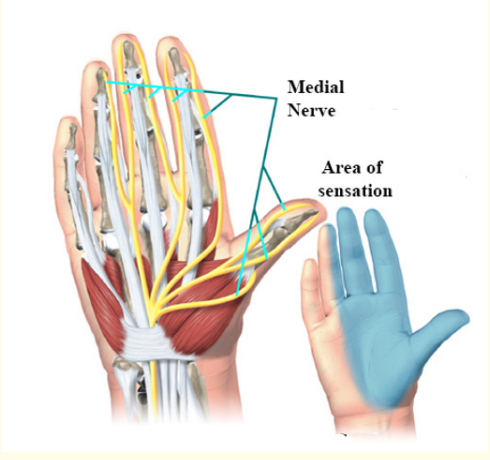


Figure 5: *Nerve of hand.*



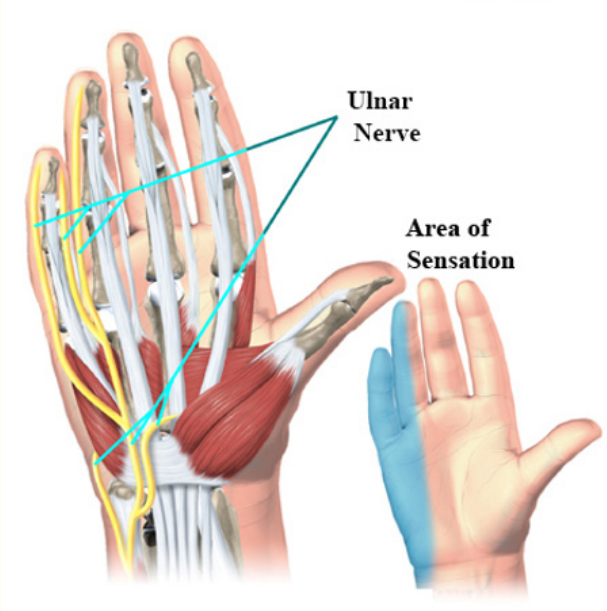


Figure 6: *Nerve movement and sensory of the hand.*

All the nerves that go to the hand and fingers start together at the shoulder: the radial nerve, the median nerve, and the ulnar nerve. These nerves carry signals from the brain to the muscles that move the arm, hand, fingers, and thumb. The nerves also carry signals back to the brain about sensations like touch, pain, and temperature.

The radial nerve runs along the side of the thumb on the forearm. It wraps around the head of the radius bone toward the back of the hand. It supplies sensation to the back of the hand from the thumb to the third finger. It also supplies the back of the hand and just behind the first phalanx of the back of the ring and middle fingers.

The median nerve passes through a tunnel inside the wrist called the carpal tunnel. This nerve supplies sensation to the thumb, index finger, long finger, and half of the ring finger. It also sends a nerve branch to control the thenar muscles of the thumb. The thenar muscles move the thumb and allow you to touch the pad of the thumb to the tip of each finger on the same hand, a movement called opposition.

The ulnar nerve passes through a separate tunnel, called Guyon's canal. This tunnel is formed by the two carpal bones, the piriformis and the hook bone, and the ligaments that connect them. After passing through the tunnel, the ulnar nerve branches out to supply sensation to the little finger and half of the ring finger. Branches of this nerve also supply the small muscles in the palm of the hand and the muscle that pulls the thumb toward the palm.

Blood vessels

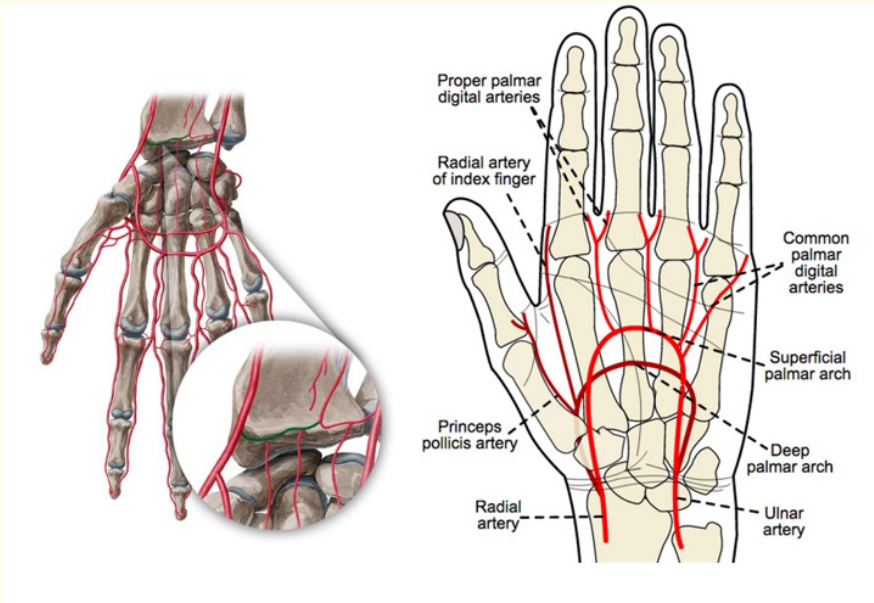


Figure 7: *Artery supply for hand.*

Along with the nerves are large blood vessels that supply blood to the hand. The largest artery, the radial artery, runs across the front of the wrist, closest to the thumb. The radial artery is where the pulse is felt in the wrist. The ulnar artery runs next to the ulnar nerve through Guyon's canal (mentioned earlier). The ulnar and radial arteries curve together in the palm of the hand, supplying blood to the front of the hand, fingers, and thumb. Other arteries run across the back of the wrist to supply blood to the back of the hand, fingers, and thumb.

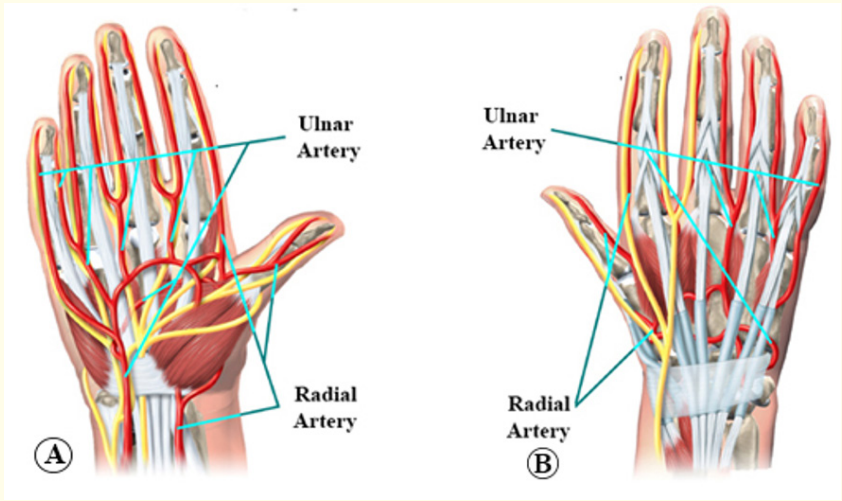


Figure 8A and 8B: *A: Palmar artery; B: Dorsal artery.*

Arteries are multilayered tubes that carry blood from the heart to other parts of the body. There are six arteries that enter the hand. They are:

- **Deep palmar arch:** Named for the shape of an arch, the deep palmar arch is small but important. This blood vessel gives off small branches that supply blood to the thumb and index finger.
- **Superficial palmar arch:** Also named for the shape of an arch, this blood vessel communicates with the deep palmar arch and also gives off important branches that supply blood to the fingers. These are called the common digital arteries.
- **Common digital arteries:** The common digital arteries are small blood vessels that arise from the palmar arch and supply blood to the fingers. They are called "common" because when they split to become the proper digital arteries, most of these blood vessels supply two different fingers.
- **Digital arteries to the thumb:** The thumb receives its blood supply from the digital arteries.
- **Proper digital arteries to the fingers:** The proper digital arteries originate from the common digital arteries, right where they split between the fingers. This split supplies blood to two different fingers. Each finger has two proper digital arteries running on either side. If one of these blood vessels is injured, the remaining blood vessels can usually maintain blood supply to the entire finger.

Congenital deformities of the hand

Duplication of the thumb

Thumb duplication is the most common anomaly of the hand [5]. The shape of the duplication depends on changes in the bone and soft tissue structure, which vary according to the degree of branching and the extent of duplication. An accurate understanding of the anatomical abnormalities is a prerequisite for successful thumb reconstruction in such hands.

Thumb duplication is usually unilateral and sporadic, and is more common in Asians. In China, polydactyly is the most common congenital anomaly of the hand. Duplication at the level of the middle interphalangeal joint (MPJ) (Wassell type IV) is the most common type of thumb duplication.

In the treatment of polydactyly, simple amputation of one finger does not yield satisfactory results, and doing so often results in malalignment or instability of the remaining ligaments of the remaining thumb. Furthermore, the reconstructed thumb often develops complications of growth plate dystrophy and ankylosis later in life. Therefore, modern reconstructive strategies involve combining elements from both thumbs to achieve the desired functional and aesthetic results.

Classification

The Wassel classification is the most commonly used classification (Figure 9) [6]. Depending on the degree of duplication and bone structure, seven types are distinguished: (1) distal phalanx duplication, (2) distal phalanx duplication, (3) proximal phalanx duplication, (4) proximal phalanx duplication, (5) bifid metacarpal, (6) metacarpal duplication, and (7) metacarpal duplication involving the triphalangeal thumb. Horii's Rotterdam classification (Figure 10) and Horii's classification of Wassel type IV polydactyly (Figure 11).

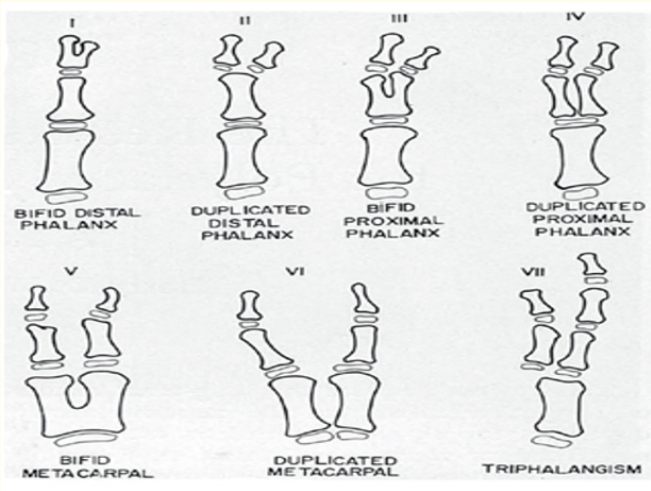


Figure 9: *The Wassel classification for radial polydactyly [6].*

Rotterdam classification (Figure 10)

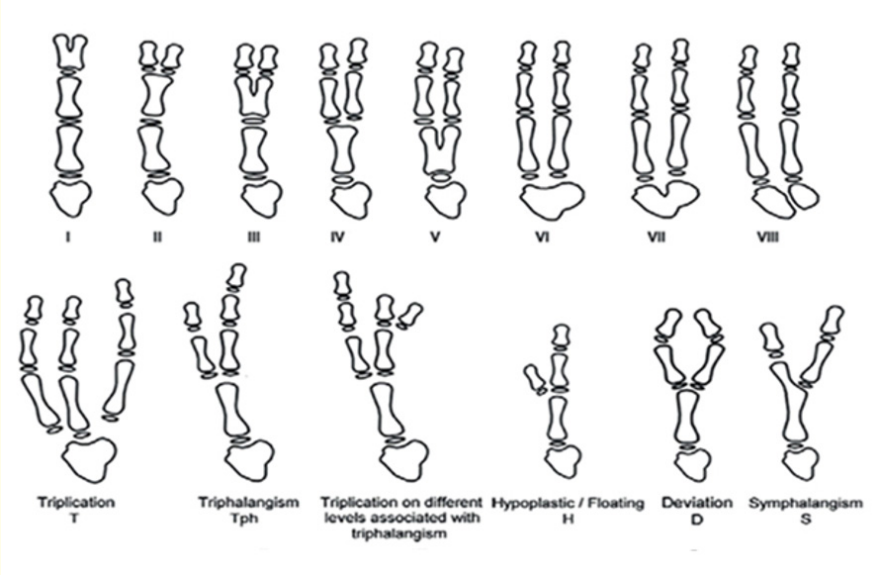


Figure 10: *The Rotterdam classification [7].*

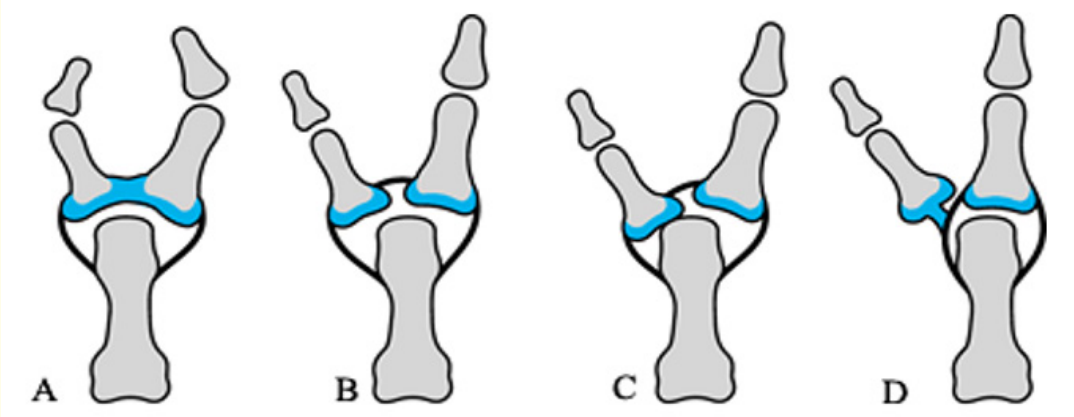


Figure 11: *Horii's subdivisions of Wassel type IV thumb polydactyly into four types [8].*

Treatment-surgery

Principles of treatment

Timing of surgery

While surgeons focus primarily on function, many parents and older children are extremely concerned about the appearance of hand deformities. They may hope that surgery can help the hand appear normal, but normal appearance is only possible in certain types of deformities. Surgery to change appearance is elective surgery. If the outcome is similar regardless of age, surgery should be postponed until the child is older than 2 years, when general anesthesia is safer [9]. Surgery performed before the child starts school may help reduce teasing and avoid school absence. Parents and older children should share the surgeon's goals and understand the risks and possible outcomes of surgery. In most cases, simultaneous surgery for bilateral hand deformities is contraindicated, as postoperative bilateral immobility is uncomfortable for young children and disabling for older children. If the surgery is minor and no casting is required, or if the child is very young and anesthesia is high-risk, the inconvenience of bilateral immobilization may be outweighed by the benefit of bilateral surgery. A long-arm splint (above the elbow) is used to immobilize the hand after surgery in infants and young children, who may have more difficulty with a short-arm splint.

Surgical procedure

After general anesthesia, the patient is placed supine on the operating table. The upper limb is prepared with a standard antiseptic solution and draped. The other hand is kept available for comparison if needed during the procedure (Figure 12-14). We use magnifying loupes and place pneumatic ligatures around the arms. We have an operating microscope in case we need to separate the thumb nerves of both thumbs.

When the thumb are the same size (Type I and II -13.0%).

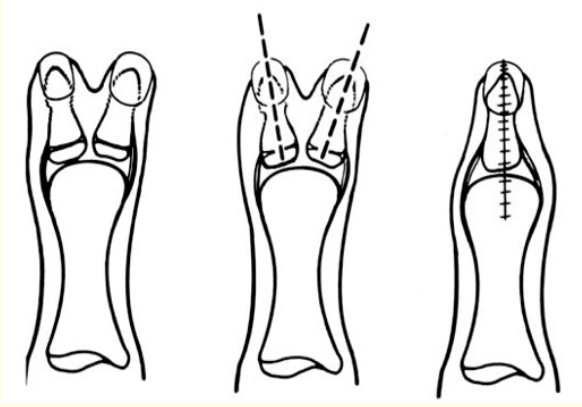


Figure 12: *Bilhaut-Cloquet technique for type II.*

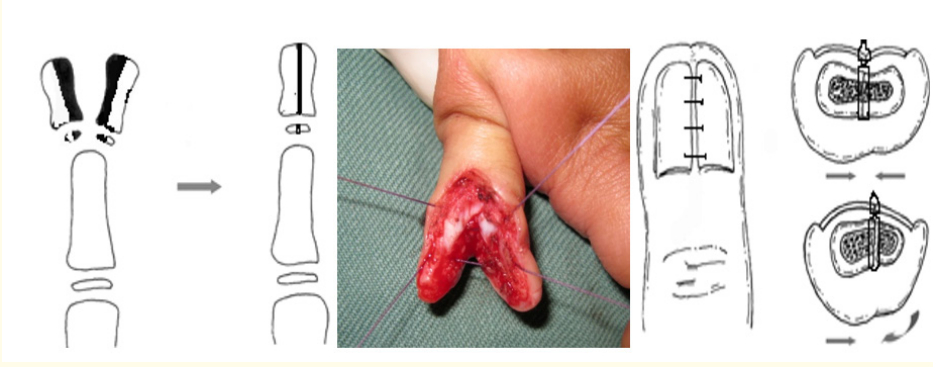


Figure 13: *Preoperative thumb are the same size (Type I and II).*

Axial malformation

Do not wedge osteotomy and axial realigment of the phalanx at the time first operation.

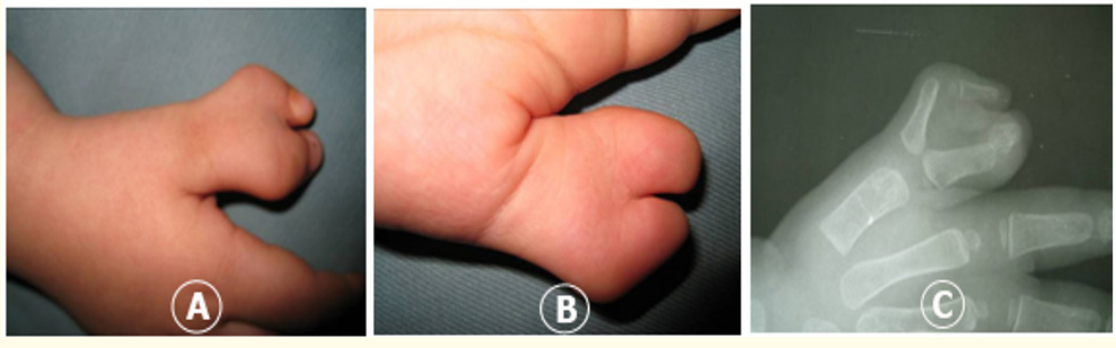


Figure 14A-14C: *Axial malformation.*

Wessel's type IV

Transferring abductor pollicis brevis muscle for Wessel's type IV [10].

Hung NN 2010 [10] reported "Cleft thumb type IV in children: transposition of the proximal phalanx with the abductor pollicis brevis tendon". We planned the incision to avoid incision along the radial contour of the reconstructed thumb. A racket incision was made around the base of the thumb duplicating the radius and the surgeons examined the thumb for tendon abnormalities. We separated the extensor tendon from the lesser thumb. For Wassel IV thumbs, the incision should allow careful exposure of the extensor and flexor surfaces, so that the tendon branch points and neurovascular structures in the digit can be clearly seen. Exposure usually requires several types of racket incisions with proximal and distal extensions (Figure 15 and 16).

Variant 1 (V1)

The racket incision is made on the least developed thumb (usually the most radial). Through the incision, we expose the adductor pollicis brevis tendon as it is introduced into the proximal phalanx of the thumb and carefully preserve this tendon. If the thumb is to be amputated, we identify the adductor pollicis muscle; remove and carefully preserve a segment of the epiphysis, with the insertion of the adductor pollicis brevis; and remove a segment of the epiphysis on the radial aspect of the thumb ulna (Figure 1a and 1b). The thumb is then separated from the wound. We separate the lateral ligament distal to the phalanx to be amputated (Figure 1a and 2). is attached to the radial aspect of the thumb ulna with a 4-0 Dexon suture (Figure 1c and 3). We center the remaining digits on the remaining articular surface and suture the lateral ligament and the medial tendon tightly to the phalanx.

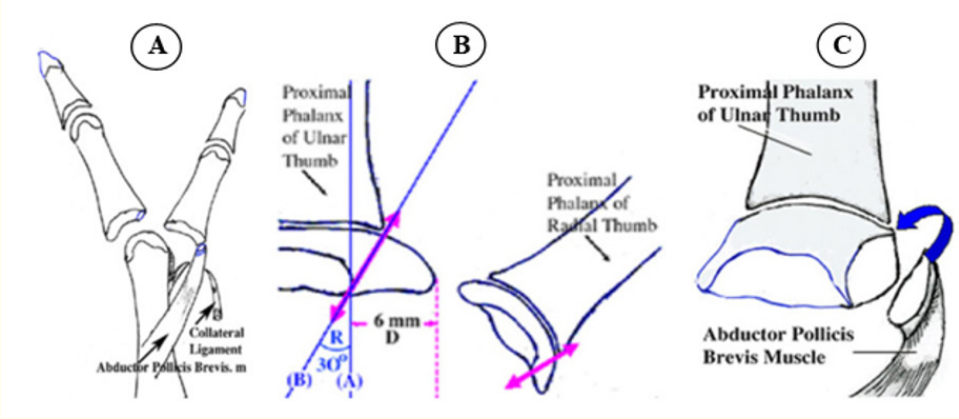


Figure 15A-15C: *A: Cutting the epiphyseal radial and ulnar thumb and creating collateral ligament. B: A phalangeal axis, B line of cutting of the epiphyseal proximal ulnar thumb, R range at the point of intersection of A and B is 30, D distance between the lateral side of the epiphyseal ulnar thumb and the point of intersection of A and B is 6 mm. C: Transferring the segment of the epiphyseal radial thumb with insertion of the abductor pollicis brevis muscle to the radial side of the ulnar thumb.*

Variant 2 (V2)

The metacarpal bone and its two surfaces are removed using the surgical procedure outlined in V1, after which the thumb is separated from the wound and the metacarpal head is identified. The metacarpal head usually has two surfaces, one for the amputated thumb and one for the remaining thumb.

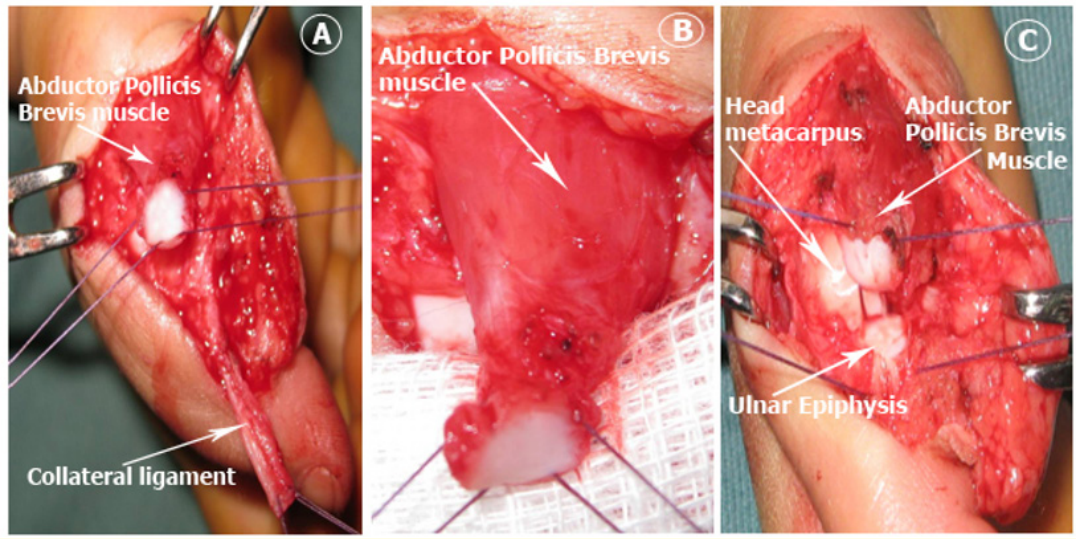


Figure 16A-16C: *Wessel's type IV with transferring abductor brevis muscle.*

Variant 3 (V3)

Cartilaginous connection between the proximal radius and ulna Using the surgical procedure outlined in V1, the cartilaginous connection between the proximal radius and ulna was identified and we then separated the connected cartilage (Figure 4b).

Variant 4 (V4)

This procedure was performed in cases where the MPJ angle deformity was a closed wedge osteotomy was performed to correct the deformity at the metacarpal neck.

Variant 5 (V5)

Surgical treatment of tortuosity deformity In these thumbs, where the radial component was removed, there was ulnar deviation at the MPJ and radial deviation at the IPJ. The aim of the reconstructive surgery was to provide good alignment and stable joints. We performed a closed wedge osteotomy at the metacarpal and proximal phalanges to correct the abnormal bone union (Figure 17).

Complications

The reconstructed thumb is always shorter than the normal thumb. Additionally, angulation, joint instability, limited range of motion, and scar contracture are common problems [8]. Reoperation rates of 20 to 25% have been reported. More complex duplicated thumbs create more postoperative problems. Any compensation in the bony axis, persistent joint instability, or tendon malalignment will lead to problems. Joint instability and/or imbalanced tendon action will cause thumb deviation and a ‘‘Z’’ deformity over time.

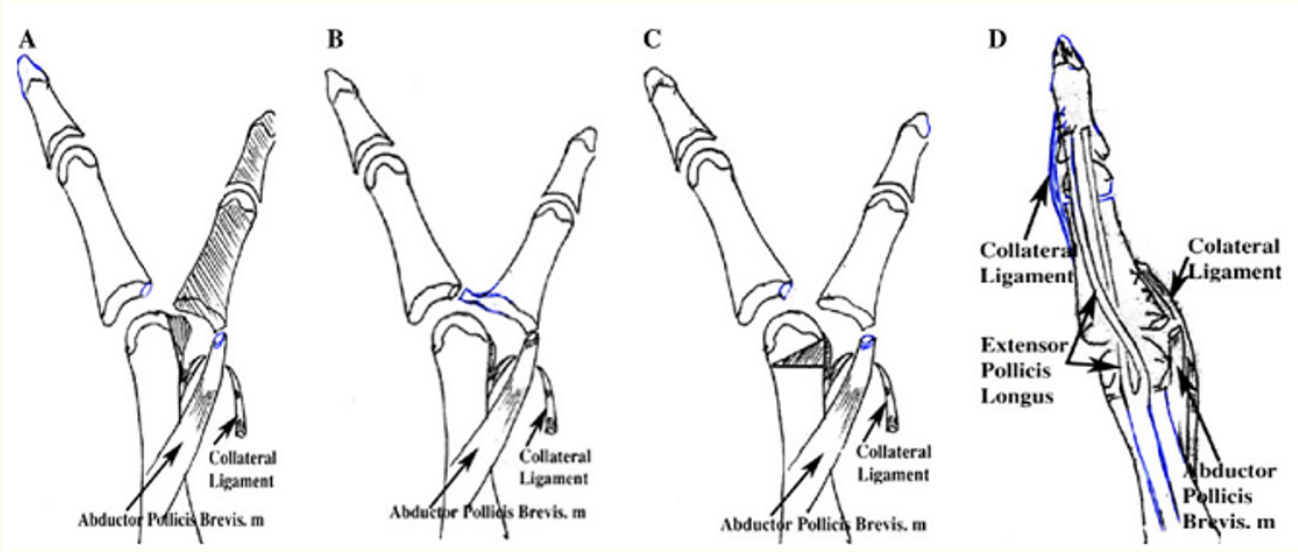


Figure 17: *Surgical procedures: A: Variant 2; B: Variant 3; C: Variant 4; D: Variant 5.*

Congenital constriction ring

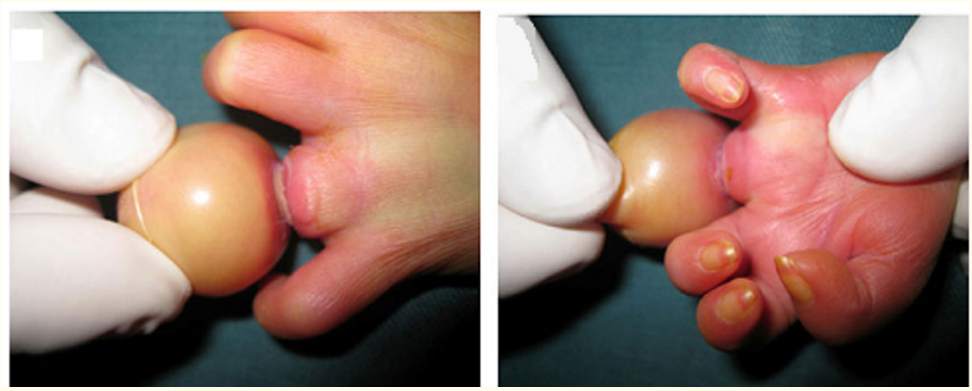


Figure 18: *Congenital constriction ring in the fourth finger.*

Amniotic band syndrome, first described by Montgomery in 1832 (Figure 18) [11], is a term used to describe a variety of related congenital anomalies, including multiple limb constrictions, oligodactyly, missing middle fingers, clubfoot, cleft lip and palate, and hemangiomas. Additional, less common clinical manifestations include complete limb absence, short umbilical cord, craniofacial disorders, neural tube defects, cranial defects, scoliosis, and body wall defects, such as gastroschisis and ectopic heart. Some of these manifestations occur

at birth with very low frequency because they result in spontaneous abortion [12] Amniotic band syndrome (ABS) or constriction band syndrome, is a congenital disorder characterized by circumferential bands around the affected limbs, resulting in an hourglass appearance, deformities, and even amputations depending on the severity. ABS occurs in 1 in 1,200 to 15,000 live births.

Women are affected more frequently than men (1.44 to 0.91), and African Americans are affected 1.76 times more frequently than whites. No specific risk factors have been identified, but an abnormal pregnancy event is noted in 60% of patients.

Craniofacial abnormalities, body wall defects, clubfoot, and visceral abnormalities have all been associated with ABS.

The cause of ABS is not fully understood, with several theories of cause. First described by Streeter in 1930, the intrinsic theory states that an abnormality in germline development causes amniotic banding [13].

This theory helps support the association with body wall defects and visceral and craniofacial abnormalities and has been supported by several studies [14]. The external theory, perhaps the most widely accepted theory today, suggests that amniotic sac rupture due to failure of the amniotic sac and chorion to fuse leads to the development of amniotic bands that encircle and constrict body parts; finally, some point to subsequent vascular disruption causing systemic alterations of the embryonic blood supply.

The timing of surgery to repair amniotic bands varies with presentation. In cases of acute limb ischemia or severe distal edema, amniotic band release is performed urgently with nerve and soft tissue reconstruction performed in the second stage. This is rare and most amniotic bands can be treated in a delayed fashion, the timing determined by the depth of the amniotic band and the distal outcome.

Classification

The timing of surgery to repair amniotic bands varies with presentation. In cases of acute limb ischemia or severe distal edema, amniotic band release is performed urgently with nerve and soft tissue reconstruction performed in the second stage. This is rare and most amniotic bands can be treated in a delayed manner, the timing being determined by the depth of the amniotic band and the resulting distal extent.

Stag I	Simple constriction ring	
Stage II	Constriction ring with the deformity of distal part lymphedema may be present	
Stage III	Distal fusion (acrosyndactyly)	
	Type I	Normal web space with distal fingertip fusion
	Type II	Incomplete web space formation with distal fingertip fusion
	Type III	Sinus tracts between digits with distal fingertip fusion
Stage IV	Intrauterine amputation	

Table 1: *Patterson classification for amniotic constriction bands [15].*

Surgical technique

After general anesthesia and tourniquet application, magnification was obtained using a magnifying loupe. First, the joints were placed in a neutral position and the proximal and distal skin edges of the tightening band were pulled together. Then, the incision site was marked (Figure 1a) and a perpendicular incision was made through the skin to the fibrous band and fascia (Figure 1b and 1c). The tightening area had to be carefully and meticulously dissected to avoid damaging the underlying neurovascular system. Fasciotomy was performed after removing all fibrotic tissue, separating the subcutaneous layer from the fascia by 1.5 cm on both sides (Figure 1d). Direct closure was

completed without suturing the fascia (Figure 1d), allowing the adipose tissue to reposition itself under the skin. To close the wound, we used 4-0 Monocryl sutures (Ethicon, Johnson & Johnson, Somerville, NJ) in the deep layer and 5-0 Monocryl sutures in the subcutaneous layer. A two-stage approach is used in all cases, with half the circumference removed in the first surgery and the other half removed 7 days later. This avoids any problems with the distal circulation of the limb, which may have been compromised. Lymphedema, if present, will improve significantly within a few weeks of the first surgery.

Postoperative care After surgery, the limb is cast and placed in a neutral position. Approximately 3 weeks after sinus surgery, the cast is removed and the limb can move freely.

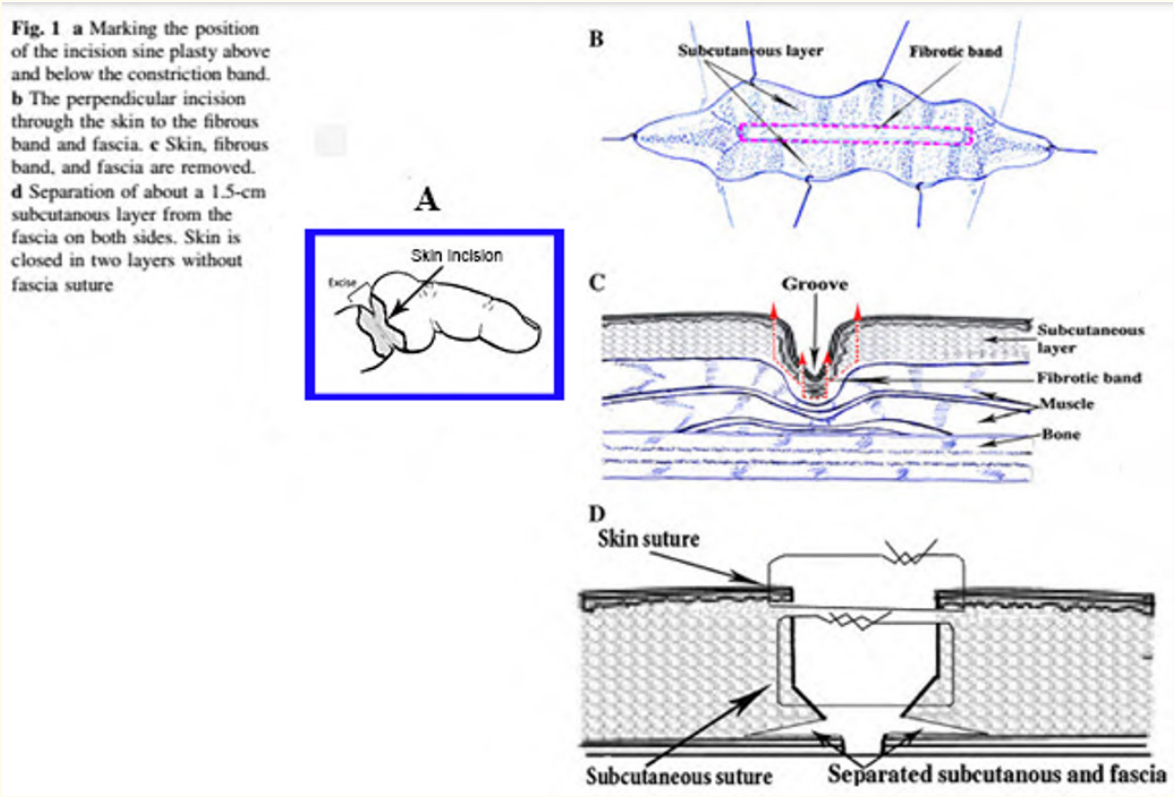


Figure 19: *Schematic diagram of Hung's S plasty technique.*

Complications

Surgical repair of tight bands is associated with complications similar to those associated with other hand surgeries, including infection, gangrene, and graft loss due to inadequate immobilization, combined with flap necrosis and distal digit ischemia due to impaired circulation. Other complications, including sensory loss, contractures, and/or immobility, are difficult to identify because they may also be due to the initial damage caused by the tight band. Although most tightness is superficial, the maximum depth is on the dorsal sur-

face of the hand and wrist [10]. The depth of the groove can vary from a partial defect with mild subcutaneous tissue loss to a deep circumferential depression that disrupts veins, lymphatic channels, tendons, and even nerves [10] in cases of complete tightness with or without distal swelling or lymphedema [10]. The greater the involvement of the palmar surface of the limb, the greater the likelihood of neurovascular and tendon involvement. The lack of involvement of the dorsal surface of the arm or axilla can often result in severe distal neurovascular and lymphatic damage, including paralysis and sensory deficits [10].

In our sinus surgery, the skin incision is perpendicular to the fibrous band and fascia, and the ligated area must be carefully and meticulously dissected to avoid damaging the underlying neurovascular system. Six patients (6.9%) had sensory deficits due to distal ligation after surgery. However, all six patients had fully recovered by the time of the last follow-up examination.

Syndactyly of hand

Syndactyly is the second most common congenital anomaly of the hand and occurs in approximately 1:2500 live births, is more common in males and is most commonly seen in the third space. The condition tends to be strongly familial and is usually bilateral [16]. It can be primary or secondary, primary being due to non-disjunction or separation. Secondary syndactyly is the result of previous events and is caused by fusion abnormalities. Syndactyly can present as the sole anomaly or can be associated with other syndromes such as Poland syndrome, palmar clefting or Apert syndrome.

Clinical

Syndromic syndactyly

Other manifestations of syndactyly are part of a syndrome, often accompanied by other congenital abnormalities. The following is a brief overview of the major genetic regions in the more common syndromes.

Distal syndactyly; there are five types of distal syndactyly, a condition related to syndactyly and early syndactyly.

Type I, synonymous with the term distal syndactyly, is Apert syndrome (MIM101200). This syndrome involves the FGFR2 gene and the 10q26 locus, and includes midfacial dysplasia, syndactyly of the fingers and hands, and a tendency to distal bone fusion [17].

Type II, Carpenter syndrome (MIM201000), involves the RAB23 gene at 6p11, with malformations including syndactyly of the feet and short hands/fingers, and acromegaly [18].

Type III (MIM101400), involves syndactyly of the second and third fingers, as well as third and fourth toes, as well as eyelid malformations and skull abnormalities. This syndrome involves the 7p21.2 and 10q26 loci involving the TWIST 1 and FGFR 2 genes [19].

Type IV, formerly known as Goodman syndrome (MIM2010 20) but now thought to be a variant of type II [20].

Type V, also known as Pfeiffer syndrome (MIM101600) involves the FGFR 1 and 2 genes [21].

Roentgenography

Testing and diagnosis

Sometimes syndactyly can be diagnosed prenatally during an ultrasound, although it is usually discovered after birth.

To determine the best treatment for your child, doctors at Children's Hospital of Philadelphia (CHOP) will perform a complete physical exam and take your family medical history. Imaging tests such as X-rays can be used to look at the structures underneath your child's hand and determine if the bones are fused together.

Classification

Syndactyly is classified as simple when only the soft tissue structures involved are involved and as complex when the bones or nails of adjacent fingers are involved. With complete syndactyly, the entire length of adjacent fingers is involved by the webbing; with incomplete syndactyly, the webbing does not extend the entire length of the finger. Complex syndactyly is the term used to refer to complex cases involving a mixture or set of syndactyly [22].

Syndactyly: Genetic studies and clinical classification issues

Nonsyndromic forms of syndactyly are easiest to analyze in their subgroups, rather than as an entity, and are shown in figure 20-28.

Figure 20: *Syndactyly I: Zagodactyly.*

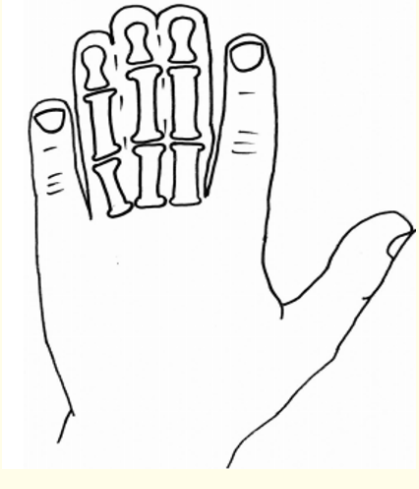


Figure 21: *Syndactyly II: Synpolydactyly.*

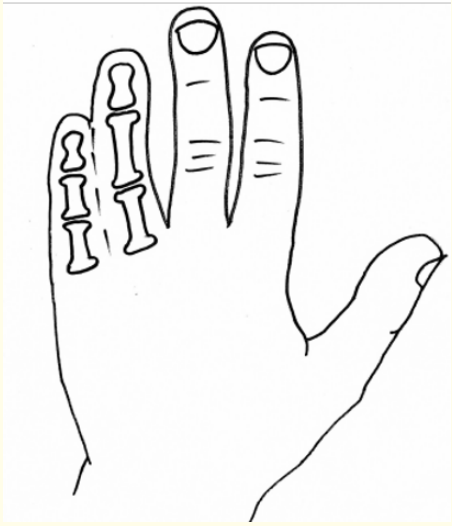


Figure 22: *Syndactyly III: Spectrum of oculodentodigital dysplasia.*

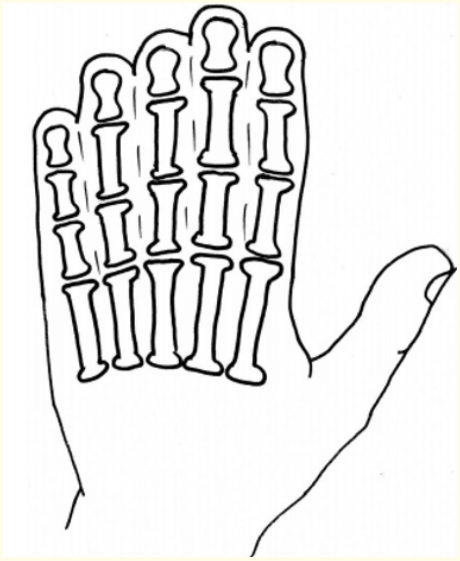


Figure 23: *Syndactyly IV: Haas type.*

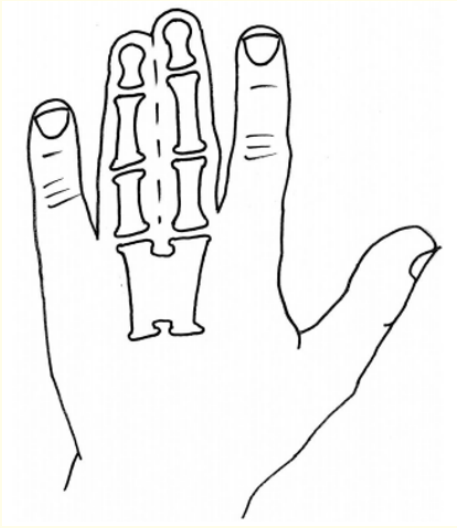


Figure 24: *Syndactyly V.*

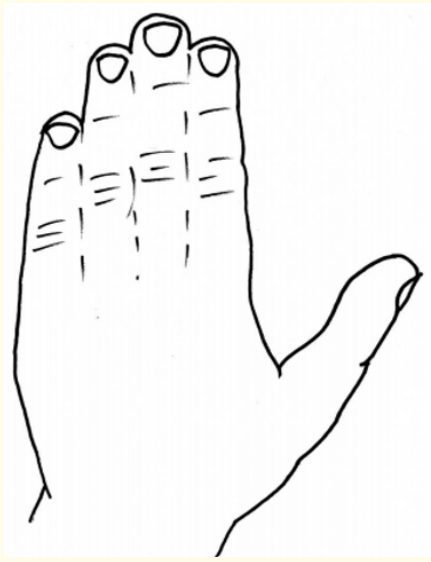


Figure 25: *Syndactyly VI: Mitten hand.*

Figure 26: *Syndactyly VII: Cenani lenz.*

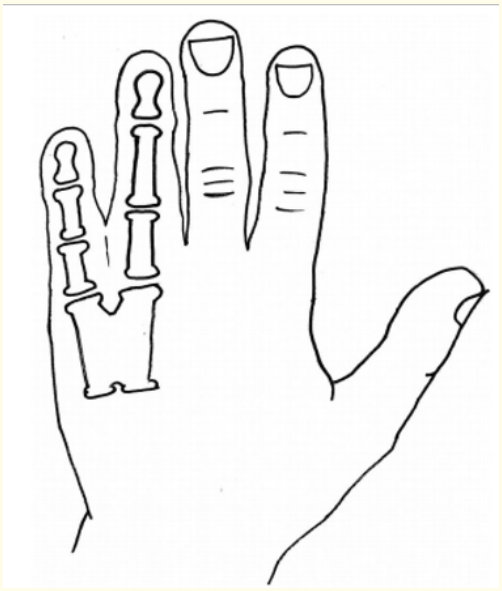


Figure 27: *Syndactyly VIII.*

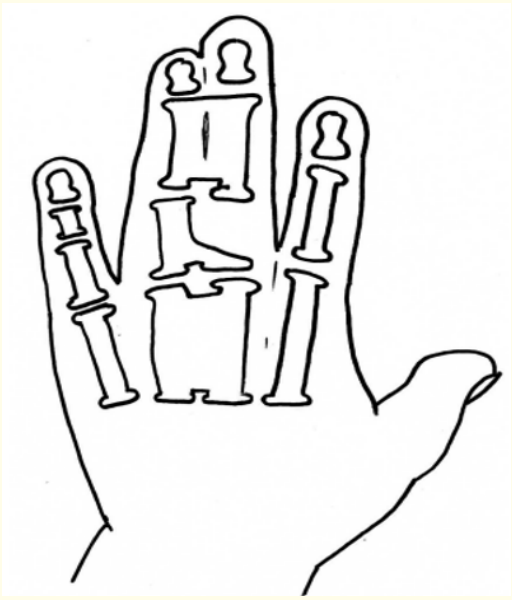


Figure 28: *Syndactyly IX: Mesoaxial synostotic.*

Figure 20-28: *Syndactyly from tType 1-8.*

Syndactyly type I (SD1; MIM 185900): Also known as zygodactyly, SD1 is characterized by involvement of the interdigital space between the third and fourth fingers and/or the interdigital space between the second and third toes. This is the more common non-syndromic manifestation of syndactyly and has also been described with involvement of other fingers and underlying bones.

Syndactyly type II (SD2; MIM 185900): Synpolydactyly (SPD), both genetically and clinically, is one of the most heterogeneous types of non-syndromic syndactyly. It also appears to be non-persistent in families with SPD.

Syndactyly type III (SD3; MIM 186100): In type III syndactyly, the normal phenotype and first described involved complete bilateral syndactyly between the fourth and fifth fingers. This is usually a soft tissue syndactyly but occasionally the distal phalanges are found to be fused. The fifth finger is usually short with an absent or rudimentary middle phalanx. The feet are usually spared. Johnston and Kirby (1955) [23] presented one of the largest fully described pedigrees, consisting of 7 affected males and 7 affected females over 5 generations in an autosomal dominant pattern of inheritance.

Syndactyly type IV (SD4; MIM 186200): Haas was the first to describe the condition and it is often referred to by his name (Haas polysyndactyly). Syndactyly is described as complete, affecting the fingers of both hands, with polydactyly usually involving the 6 metacarpals and 6 digits.

Syndactyly type V (SD5; MIM 186300): This rare form of syndactyly is typically characterized by fusion of the metacarpals and metatarsals. The most common fused metacarpals and metatarsals are the 4th and 5th metacarpals or the 3rd and 4th metacarpals.

Syndactyly type VI (SD6; MIM undetermined): SD6, also known as glove syndactyly, involves unilateral syndactyly of digits 2 through 5 [24]. A family has been described with this anomaly, which is likely to be autosomal dominant, but with variable expression and incomplete penetrance.

Syndactyly type VII (SD7; MIM 212780): In 1967, Cenani and Lenz [25] described two brothers with a form of syndactyly resembling Apert syndrome. However, they noted that there were additional features including severe shortening of the radius and ulna at fusion, fusion of the metacarpals, and 'disorganization' of phalangeal development.

Syndactyly type VIII (SD8; MIM not allocated): Fusion of the fourth and fifth metacarpals is also an uncommon manifestation of syndactyly. First described by Orel in 1928 [26], it was initially thought to be an X-linked recessive trait, supported by families described in later papers.

Syndactyly type IX (SD9; MIM609432): Type IX, mesoaxial syndactyly (MSSD), has only been described in two families. Originally found in a family known to have SD1, the phenotype of axial syndactyly is characterized by complete syndactyly and osteophytes of the third and fourth digits with severe osteolysis of the proximal phalanges, hypoplasia of the thumb and index finger, aplasia/hypoplasia of the middle phalanges of the second and fifth digits, and complete or partial soft tissue syndactyly of the toes.

Classification

The classification of syndactyly is shown in figure [27]. The term simple syndactyly is used if only the soft tissues are affected and complex syndactyly if the bones of adjacent digits are fused (synostosis). The term incomplete syndactyly is used to describe proximal syndactyly involving the distal phalanx and complete syndactyly if the syndactyly extends to the distal phalanx. The term complex syndactyly describes syndactyly with bony formation between the two finger rays (polydactyly occult) and multiple syndactyly describes syndactyly involving the distal phalanges, but with a close spacing.

Classification and causes

Hand development begins on day 27 of intrauterine life [28]. The fingers are usually separated by days 44-46 (Figure 29). Most syndactyly occurs during this period and may be due to a defect in the programmed cell death (apoptosis) pathway that normally twists the embryonic interdigital tissue and allows the formation of individual digits (Figure 30) [29,30]. This tissue is often twisted from distal to proximal, and the explanation for syndactyly is not entirely straightforward.

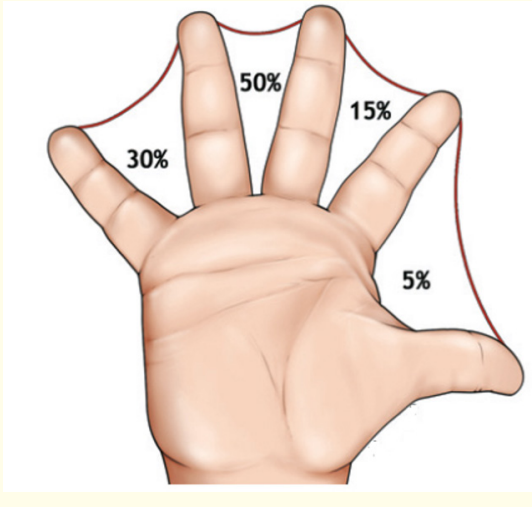


Figure 29: *Schematic representation of involved web spaces [31].*

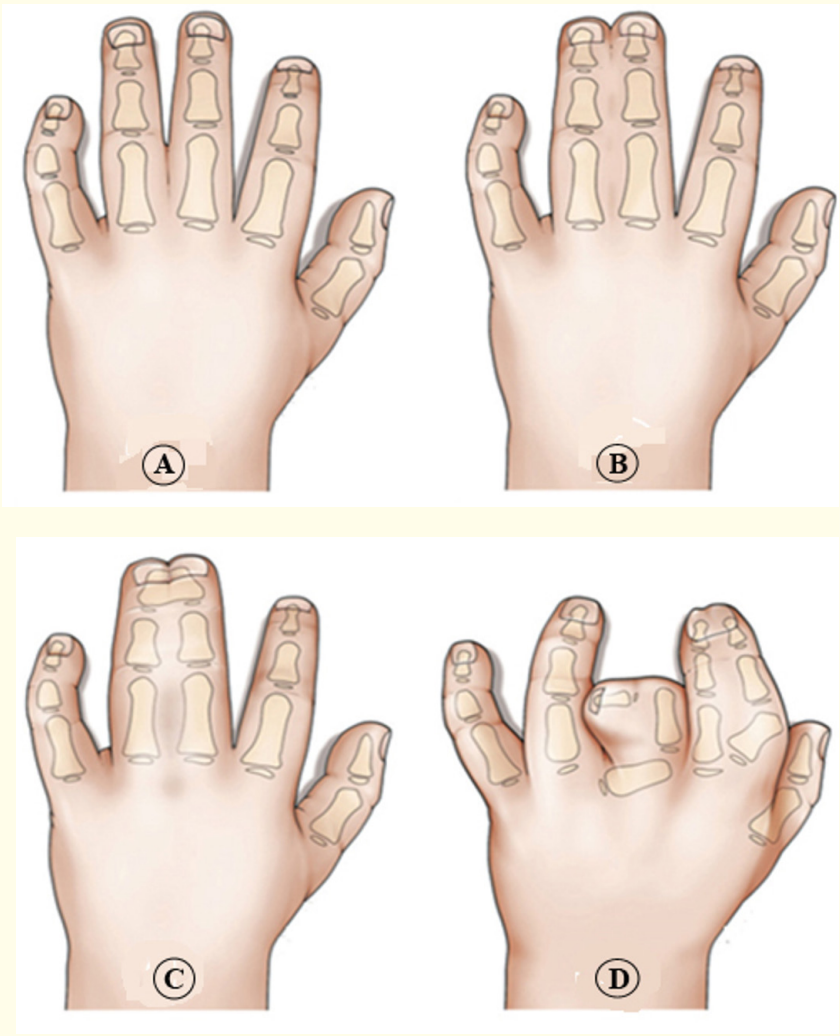


Figure 30A-30D: *Classification of syndactyly. Syndactyly between long and ring finger. A) Simple incomplete syndactyly. B) Simple complete syndactyly. C) Complex complete syndactyly. D) Complicated syndactyly.*

The exception to the rule is acrosyndactyly, where the most common form is associated with amniotic band syndrome, which causes fingers to fuse after the separation process is completed.

Treatment - Surgery

Surgical treatment

The general goal of syndactyly surgery is to produce a hand with as many independent and functional digits as possible with the least number of surgeries and complications. Many techniques have been described to achieve this goal; Upton [26] reported 46 different methods used over the past two centuries.

Timing of surgery

The timing of surgery has been well studied. We typically perform surgery when the child is about 12 months old. Surgery should be performed early enough to allow normal development, but late enough to avoid postoperative complications. Surgery before 12 months of age is associated with a higher rate of scar contractures.

Indications for surgery

The main indication for syndactyly surgery is a soft tissue bridge, with or without a bony joint, connecting two or more digits and affecting the functional use of the hand. A contraindication to surgery for syndactyly is webbing associated with the so-called extra digit. Two types of syndactyly have been described: type I involves two metacarpals supporting a single oversized digit; type II involves a single metacarpal supporting two or more distal digits.

Goals of surgery

Normal function of the fingers requires proper placement and depth of the webbed space. Therefore, the goals of surgery are to create a normal webbed space and improve the shape of the affected digits. There are many ways to separate the digits and reconstruct the webbed space and will be described in more detail later.

Technical considerations

Surgical techniques for treating syndactyly have not changed significantly over the past century. Separating webbed digits by separating them with a pair of scissors in the nursery, as was done in the early 1800s, is not recommended because of the inevitable risk of flexion contractures due to longitudinal scarring. Full-thickness skin grafting (FTSG) is often recommended to cover areas of the finger not covered by a local flap.

Overview of surgical technique

The most common reconstructive design for the joint, and that used at our institution, involves a dorsal rectangular flap originating proximally. Many geometric designs have been used for this flap, including rectangular, triangular, omega, and multilobular. Flap design is highly dependent on surgeon training and preference, and can be easily modified to attempt to recreate a more anatomic web space.

Surgical technique

The surgery should be performed under general anesthesia with a ligature in the affected limb; the contralateral groin is prepared for removal of the FTSG. The joint is designed dorsally to have a medial and lateral root border in the longitudinal midline of the adjacent digits, starting at the metacarpophalangeal joint (Figure 31).

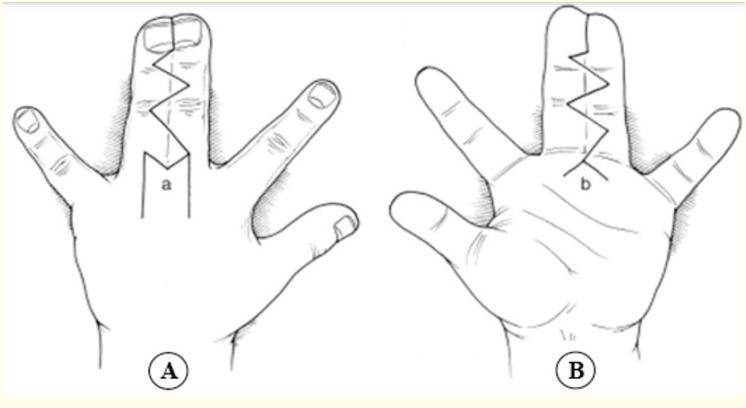


Figure 31A and 31B: *Dorsal (A) and palmar (B) views. The dorsal flap (a), the V-shaped palmar flap (b), and the zigzag flaps for the syndactylized digits.*

Starting at the distal end of the finger, the dorsal incision is extended in a zigzag pattern from the midline of one finger to the adjacent finger. If there are two separate fingernails, the dorsal incision is extended distally, bisecting the two fingertip tips. When there is a complex nail, as in complex syndactyly, the nail bed and nail bed should be divided along the distal phalanx line.

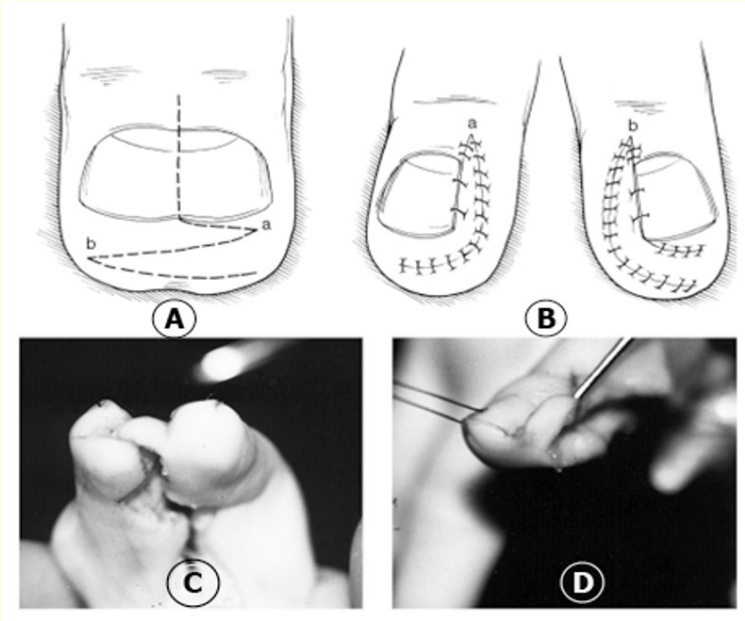


Figure 32A-32D: *Creating a nail fold using skin from adjacent fingertips. A: Dorsal flap, B: Palmar flap. C: Intraoperative photograph of double flap being raised. D: The flap is swung proximally before it is secured with sutures.*

The palmar curve is then drawn. Proximally, this line meets the center of the previously drawn V-shaped palmar anchor line, approximately midpoint between the proximal and middle thirds of the proximal phalanx (Figure 32).

Special considerations

Although the techniques described are appropriate for most patients, other conditions, such as milder sleep, incomplete ring finger, complex, saddle hand, multiple middle fingers, and no ring, require alternative techniques.

Data grafting is required. A, Obtaining an elevated dorsal arch. B and D, Rotating and inserting the dorsal and palmar arches. C, Design of the palmar triangular flap. a = dorsal arch, b and c = palmar triangular flap.

For incomplete thumb-index digits or other mild digits, a four-flap Z-plasty or a medial V-Y-plasty with lateral Z-plasty can be used to moderately deepen the interdigital space between the ring fingers. These techniques avoid the need for skin grafting on the lateral wall, which is the working surface of the thumb. For non-retractable first digits, proximal displacement can create a functional working surface (Figure 33).

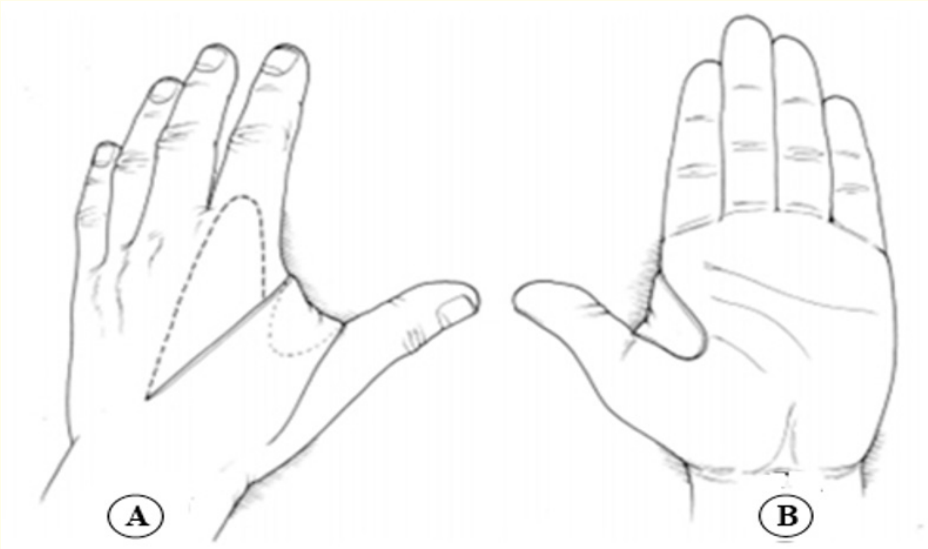


Figure 33A and 33B: *Dorsal A. and palmar B. views demonstrating correct placement of a dorsal transpositional flap for first web-space release.*

Complex syndactyly

Syndactyly involving bony connections between adjacent digits requires a plan that prioritizes early establishment of normal or near-normal bone structure, after which definitive periosteal release can be performed. Syndactyly can be separated by releasing the syndactyly with a scalpel or osteotome. The metacarpal angle is corrected by closed wedge osteotomy with a Kirschner wire through the midmedullary and advanced retrogradely.

Complications

As with any surgical procedure, infection can occur. Although infection is a relatively rare complication of syndactyly surgery, it can be devastating if not promptly recognized and treated with surgical debridement or antibiotics. Recent data suggest a postoperative infection rate of 1.6%. Surgical complications are more likely to occur in patients with syndactyly and multiple preoperative medical diagnoses. Other potential complications include scarring and contractures [32]. This may result from the use of split-thickness skin grafts or from surrounding bone growth. The use of full-thickness grafts is less associated with contractures and is preferred when possible for this reason. Another common complication is web creep, which is defined as the web space moving distally as the patient ages. This can lead to progressive deformity of the hand. The patient's age at the time of surgery may be an important factor in web creep. Patients should be monitored periodically after surgery to detect this complication early. Finally, devascularization of the digit or nerve damage may occur. Only one digit should be operated on at a time to ensure that the digit retains a functioning digital artery [33]. Devascularization can result in prolonged healing and recovery times or, in severe cases, amputation. In cases of nerve injury, several techniques have been described to address finger nerve injury, including primary nerve restoration, nerve conduit restoration, and nerve grafting. Finger nerve injury, even after restoration, can result in loss or decreased sensation in the fingers.

If nonsurgical treatment is chosen, this can result in reduced dexterity and function in the hand. Nonsurgical treatment or observation of syndactyly between fingers of different lengths can lead to increased deformity and growth disturbances. The shorter finger may have the effect of forcing the longer finger into a flexion deformity toward the shorter finger and into a flexed state [32].

Radial club hand

Congenital clubfoot or agenesis of the radius is a relatively rare congenital abnormality of the upper limb, usually occurring in association with radial atrophy or agenesis of the radius and also absence of a functional thumb in most cases (Figure 34).

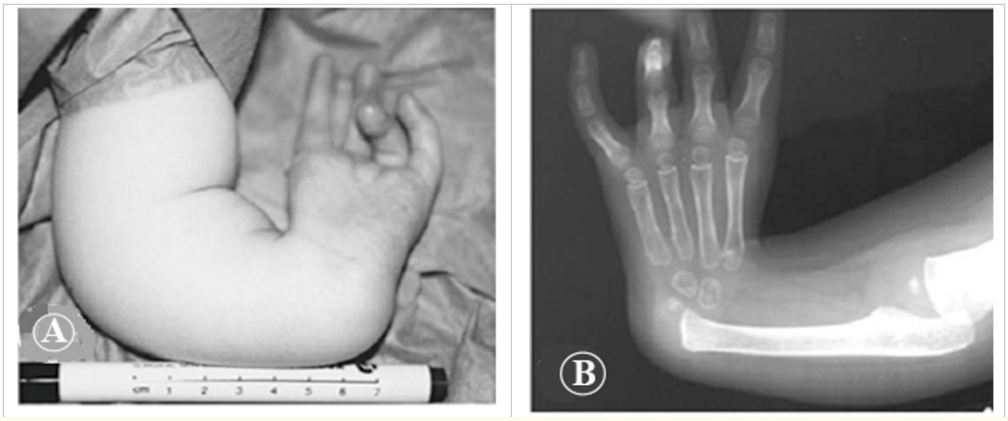


Figure 34A and 34B: *A: Preoperative view of the hand of a girl at the age of 4 years 3 months. B: Preoperative radiograph of the same patient before operation.*

The traditional treatment for radial clubfoot is a variation of centralization [35] and has recently been combined with preoperative soft tissue decompression to enhance surgical procedures. Some of these reconstructive efforts have been accomplished using a vascularized fibula to replace the missing radius. With the aid of advanced microsurgical techniques, developed in conjunction with vascularized metatarsal-toe-toe fusion (MTP-II) for congenital digit defects, the authors introduced metatarsal-toe-toe-toe fusion (MTP-II) in 1992 as a treatment to stabilize the radial-displaced hand.

Clinical

Congenital radial clubfoot or vertical plane radial defect is characterized by radial deviation of the hand, marked forearm shortening, and general underdevelopment of the limb [36]. The thumb is often absent or underdeveloped, and the ulna is usually 60% of its normal length.

Radial clubhand may be an isolated anomaly or may be associated with one of several complex syndromes. Unilateral clubhand is usually an isolated anomaly, while bilateral clubhand is often associated with syndromes. Several non-syndromic skeletal anomalies may be seen in children with unilateral clubhand, including radioulnar syndactyly, syndactyly, congenitally elevated scapulae, scoliosis, clubfoot, and developmental dysplasia of the hip.

Impaired hand function

Effective grasping and gripping are both impaired in children with unilateral clubhand, and the impairment may be due to failure of the thumb to develop normally, abnormal fingers, and altered muscle function.

Thumb: In some cases, the thumb is completely absent, while in others, the thumb may be present but completely nonfunctional. In these cases, the grip function is lost. When the thumb is present, the range of motion of the thumb can be significantly reduced and a true pinch is often impossible.

Fingers: The index finger is often stiff; generally, the little finger is the most mobile finger. When the thumb is absent, the index or little finger takes the place of the thumb. The range of motion of the interphalangeal joints is reduced and this is a factor in poor grip strength.

Muscles: Abnormally oriented muscles pull the hand outward and bend it inward. The finger flexors are ineffective in generating sufficient force to bend the fingers.

Wrist instability

Wrist instability contributes significantly to poor hand function. Grip strength is significantly reduced because the wrist cannot maintain dorsiflexion when the child attempts to grasp an object.

Short forearm

A short forearm itself may not cause any functional limitations but it does add to the unsightly appearance of the limb. However, the short radius in children with clubfoot type II contributes to instability at the wrist.

Stiff elbow

- Improving overall hand function. Although improving hand function is desirable, in some cases this may not be possible. It is essential to clearly identify cases where hand function cannot be improved because any attempt at surgical intervention may actually further impair function.
- Improving limb form radial and flexion deviation at the wrist, ulnar bowing, short forearm, and associated thumb deficiency may all need to be addressed to improve limb form.

Classification

Although radial deficiency has a wide range of phenotypes, ranging from thumb hypoplasia to complete absence of the radius and first ray, Bayne., *et al*. [37] classified radial deficiency into four types based on the radiographic severity of the radius deficiency (Figure 35):

- Type I, short distal radius (radius slightly shorter than ulna, distal growth plate present);
- Type II, underdeveloped radius (radius smaller/thinner than ulna, absent growth plate);
- Type III, partial radial absence (only a small proximal radius segment present); and
- Type IV, complete absence (complete absence of radius, ulna may be curved).

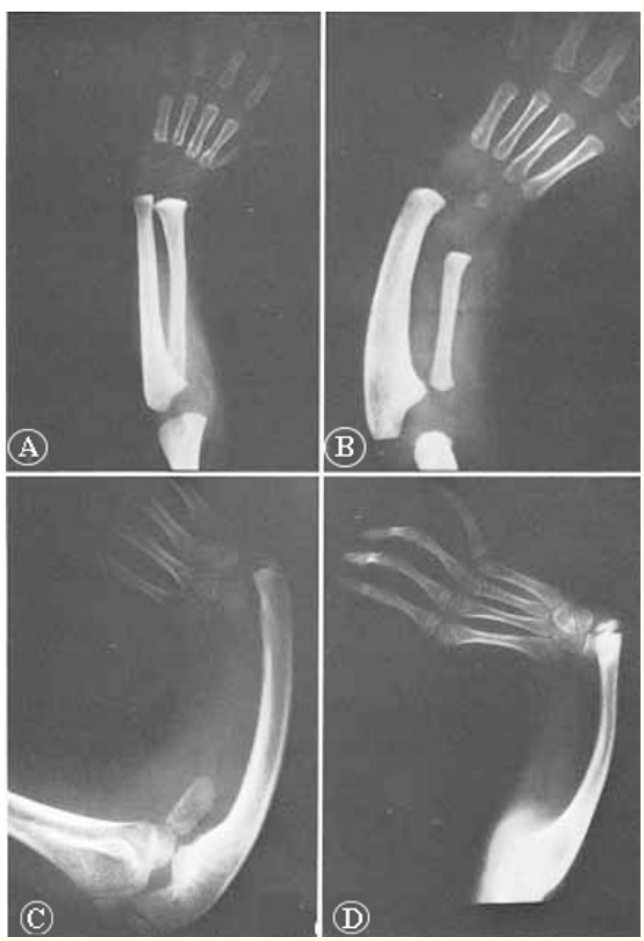


Figure 35A-35D: *Radiographs of varying severity of radial club hand. In the mildest form or A: type I radial club hand the radius is slightly short, B: while in type II the radius is clearly hypoplastic. C: In type III there is partial absence of the radius; D: In type IV the entire radius is absent [37].*

Heikel classification of radial dysplasia [38]

Clinical examination, preoperatively and at follow-up, assessed shoulder and elbow range of motion, wrist deformity, and finger chain range of motion. Six patients with stiff extensor elbows underwent preliminary rehabilitation and flexion orthosis to achieve 90° preoperative flexion. Comprehensive evaluation, performed by geneticists, screened for associated abnormalities (Table 2).

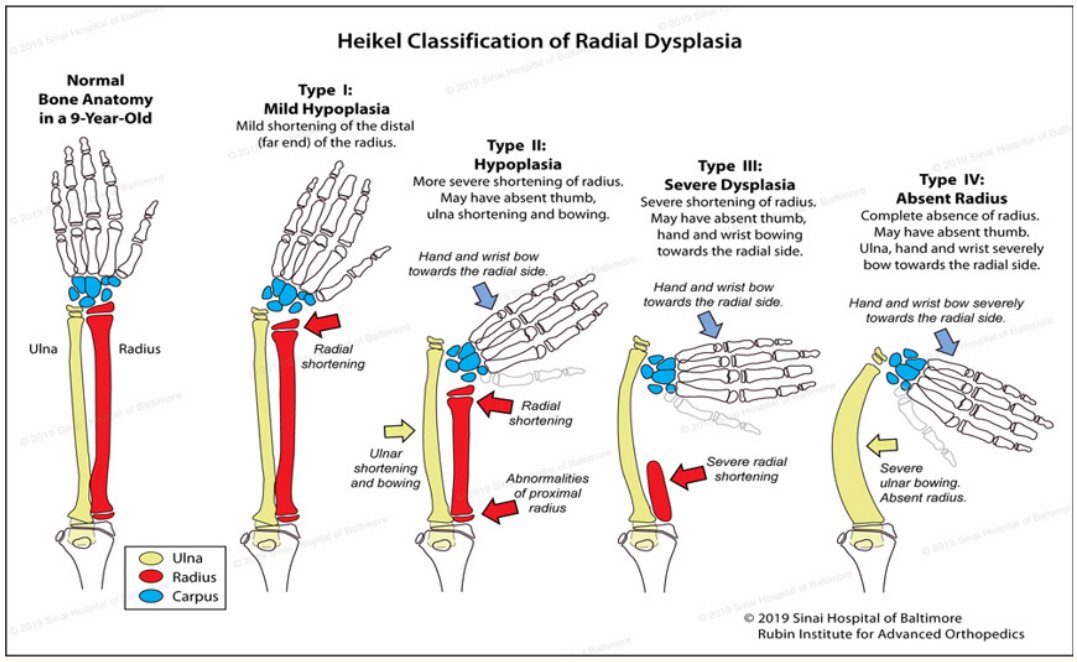


Figure 36: *Heikel classification of radial club hand:*

- *Type I: Short distal radius.*
- *Type II: Hypoplastic radius.*
- *Type III: Patial absence of radius.*
- *Type IV: Complete absence of radius.*

Patient		Range of passive elbow flexion (º)	Thumb
1	Cardiac (VSD), hearing loss, contralateral thumb hypoplasia	110	Index Pollicization
2	Cardiac (VSD), persistent arterial canal), psychomotor retraction	90	
3	Hypoplasias, large vessel malpositioning, thumb hypoplasia	70	Index Pollicization
4	Isolated	120	Index Pollicization
5	Spine (binucleate T 10 vertebra)	110	Index Pollicization
6	Testicular ectopia	70	
7	VACTERL	100	

7	Dextrocardia, cervical spine deformities	N/A	Index Pollicization
8	Spine, Cardiac	60	Index Pollicization
9	Cardiac (VSD), renal (incomplete duplicity, mega ureter, low ears	90	Index Pollicization
10	Isolate	80	
11	TRA, G6PD, deficit	N/A	
12	TRA, contralateral thumb hypoplasia	120	

Table 2: *Assessment, performed by geneticists, screened for associated abnormalities.*

VACTERL: Association of at least 3 of the following: vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or esophageal atresia, renal anomalies and limb defects; VSD: Ventricular Septal Defect; TRA: Thrombocytopenia + Radial Aplasia; N/A: Information Not Available.

Roentgenography

Patients were evaluated radiographically by analyzing preoperative and postoperative hand and wrist radiographs as described by Manske., *et al*. [39]. Two measurements were taken. The hand-forearm angle (HFA) is the angle between the long axis of the long metacarpal and the distal ulna as seen on anteroposterior radiograph of the hand and forearm. A ‘‘negative’’ HFA refers to ulnar deviation of the long metacarpal and long metacarpal bones relative to the distal ulna, the more desirable position. The hand-forearm position (HFP) refers to the distance in millimeters between the base of the little metacarpal and the distal ulna. If the proximal pole of the little metacarpal is ulnar relative to the distal ulna, the (HFP) position is recorded as a positive millimeter; a change in ulnar orientation is considered an improvement in hand position. The goal is to achieve mild correction of the ulnar deviation of the hand and ulnar displacement of the hand and wrist by the end of the treatment regimen.

Treatment - Surgical

Indications and timing

For infants with radial clubfoot, careful splinting is always the mainstay of treatment and should be started on the first day of life and continued to improve hand alignment and wrist mobility. In addition, parents should start treatment early combined with regular gentle manipulation. Radial deviation will quickly become more difficult to treat if these measures are not started early or splinting is discontinued. Current treatment for radial clubfoot is considered appropriate for grades 3 and 4 Bayne defects. In grades 2, when the radius is only hypoplastic, the deformity is usually not progressive. Typically, such a shortened radius can be treated with conservative monitoring and careful splinting. In severe cases, correction of the radial wrist deviation by lengthening the hypoplastic radius may be necessary. In types 3 and 4, the radius is not supported and the hand will be severely deformed and deviated in the radial direction and in most cases, surgical treatment is considered necessary. The method presented is also useful after attempting to concentrate with moderate or severe recurrence.

Treatment - Geberal principle

Management of radial insufficiency should begin as soon as possible after birth. Treatment should include manipulation and splinting, which will stretch the stretched soft tissues and radial structures and allow passive correction of the deformity by aligning the hand and wrist with the ulna. Adequate preoperative soft tissue stretching is a prerequisite for successful surgery and should be strictly followed up to the time of surgery.

Soft tissue stretch and optional release

When wrist alignment can be achieved relatively easily, release is not required in the early stages. Preoperative radiographs with a test of wrist alignment by pulling from the fingers will provide guidance as to its need. In addition, clinical stiffness may clearly dictate its need. However, it is rarely needed in well-treated conservative cases. Unnecessary scarring from the incision on the radial aspect of the wrist may also add to the difficulty of subsequent major surgery. If release is required, the surgery will be performed through a transverse incision that cuts all remaining extensor and flexor muscles of the radial wrist along with release of the tightness of the joint capsule. All tendons that move the fingers must be left intact. This should be a delicate and non-traumatic procedure, taking into account the blood vessels and nerves needed for the actual joint grafting later (Figure 37).

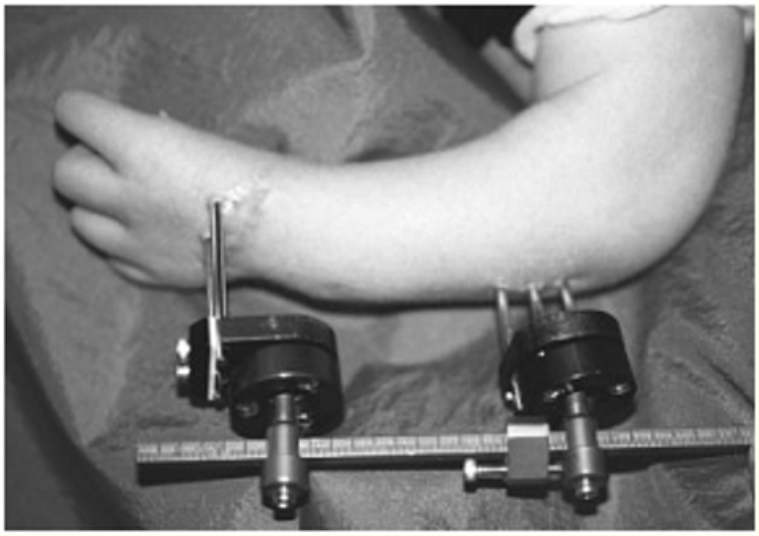


Figure 37: *Soft tissue distraction in progress. The Hammer distraction device is lightweight and it weighs 120g with 6 pins. The ball joints allow easy assembling.*

The average amount of soft tissue stretch required is 25 mm after the initial manual adjustment during the external fixation procedure. This means that 45 to 50 days of stretch are required before the second part of the reconstruction can be performed. Soft tissue stretch is always a time-consuming process. Stretching should not exceed 0.5 mm per day. There is clear evidence that articular cartilage detachment and forearm muscle ischemia will occur at higher rates of stretch.

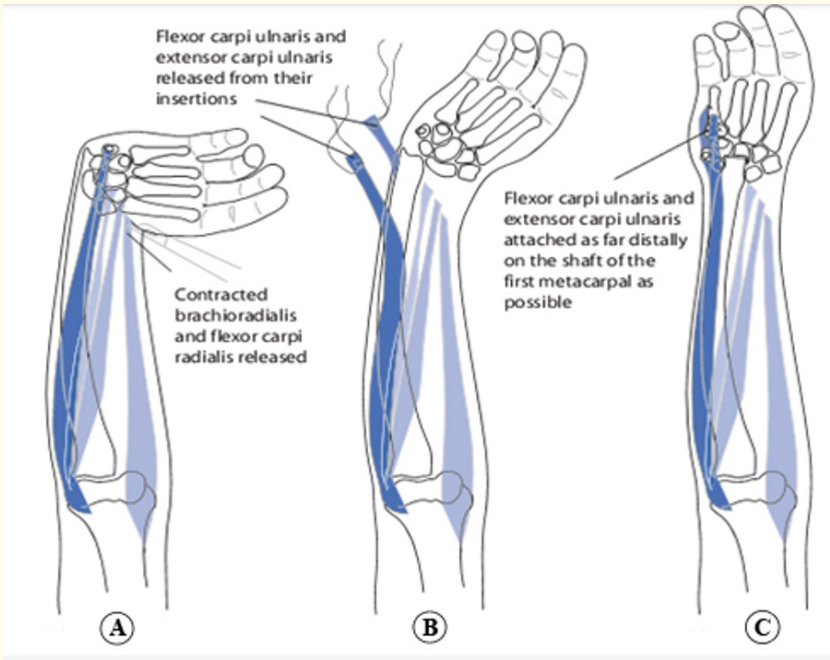
Treatment history

Previous surgical treatments for RLD have attempted to create a centralized wrist, maintain wrist motion, and improve hand function by placing the hand in a more extended position (Figure 37). Patients with type I deformities usually do not require treatment, whereas patients with type II Bayne deformities can often be treated conservatively with a wrist splint or, in some cases, radial extension. Various surgical approaches have been described over the past century to treat patients with type III and type IV deformities. Some have proposed soft tissue release with or without ulnar osteotomy and nonvascular bone grafting, but these procedures alone do not maintain long-term wrist alignment. The initial centralization technique was described by Sayre [40] in 1893 and was later modified by Lidge in 1969 [41].

Radical advancement

Traditionally, the treatment of radial clubfoot (RCH) has been to centralize the carpus to the ulnar head. Partial or complete recurrence of the radial clubfoot deformity and growth arrest of the distal ulna are common sequelae after centralization. In 1985, Buck-Gramcko [35] described an alternative to centralization, which he called radical advancement. Heuseda makes a dorsal incision to separate the hand from the carpus and then transfers the hand to the ulnar side of the ulnar head.

The Buck-Gramcko radialization is performed through a dorsal incision, without visualization of the neurovascular structures, overdissecting the ulna, and without visualization of the radial vascular pedicles of the distal ulnar head and the ulna. Furthermore, the flexor and extensor carpi radialis tendons are often absent and cannot be transferred. The English name "radialization" is misleading (Figure 38). It can be mistaken for moving the hand in a radial direction. Radialization moves the hand in an ulnar direction.



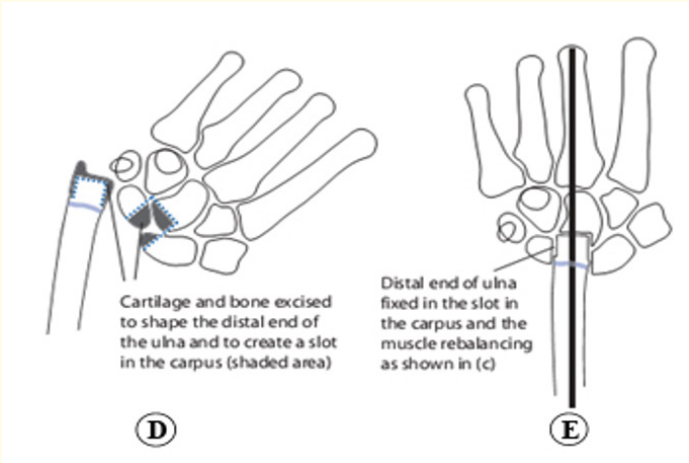


Figure 38A-38F: *A-C: Muscles transfer; D: Pre-Operative LRS; E: Centralization; F: Radicalization.*

We believe that the Ilizarov device offers certain advantages over previously described external fixators for soft tissue reduction before centralization in children with Bayne type IV congenital radius deficiency. By having the option of placing multiple bone anchors in different planes and applying movable hinges corresponding to the apex of the deformity and the ability to suspend dynamic slings at the distal end of the fixator, a more complete and physiologically sound reduction can be achieved.

Centralization (Phase one)

Centralization was performed. The goal of centralization was to stabilize the carpal bones and correct the angle. An S-shaped incision on the dorsum of the hand was made from the dorsum of the hand to the distal third of the forearm. This allowed access to the radial muscle, the ulnar shaft, and the carpal bone itself. Except for the skin incision, the actual centralization technique was based on the technique described by Bayne and Klug [37].

The wrist was fixed with a 1.8 mm Kirschner wire. The skin was closed easily by rotating the flap. A long arm cast was applied for 4 weeks. A short arm splint was then applied to protect the wire and prevent recurrence of the deformity.

Centralization by tendon transfer

Centralization by tendon transfer is a common method for maintaining the corrected position of the radial hand [37]. The first centralization of the carpus to the distal ulna, an operation that still exists today in a modified form, was performed by Sayre in 1894 [39] (Figure 39). On the other hand, various methods of correction have been proposed, such as rotation [35].

Figure 39A-39C: *A: Preoperative appearance at one week of life. B: The hand is radially deviated by 90°. C: Preoperative X-ray film of the radial club hand at one year.*

In the present review, we explain our centralization procedure with tendon transfer for Bayne Type III or Type IV and our radius lengthening with temporary external fixation of the wrist [37]. for Bayne Type I or Type II (Figure 40).

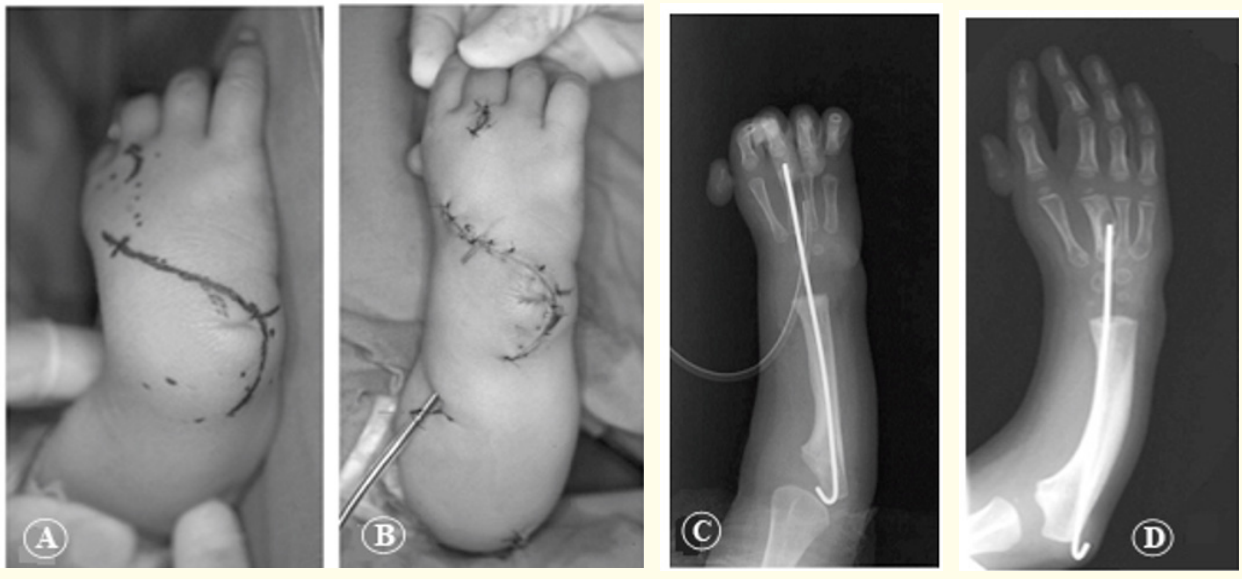


Figure 40A-40D: *Centralization and postoperative X-ray film. A-C: Centralization at the age of 22 months. A: Dorsal S-shaped skin incision. B: Appearance immediately after operation. C: X-ray film immediately after operation. D: X-ray film shows cyst formation, possibly due to mechanical stress caused by the Kirschner wire, at 9 months after centralization (at 31-months-old).*

Stage two (Index fiber pollicization)

A thumb transfer was performed, i.e., the transposition of the thumb to create a new thumb. In this case, the actual thumb transfer technique was based on the description of Carroll [43]. The skin incision for the thumb transfer was designed to extend from the incision used for centering (Figure 41).

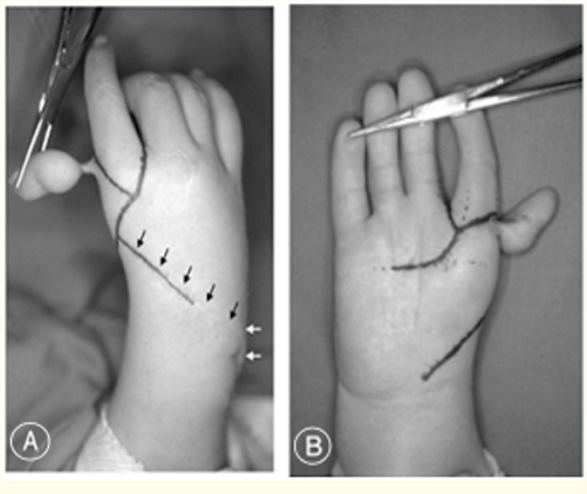


Figure 41A and 41B: *Pollicization at 31-months-old. A: The skin incision for pollicization (arrows) was designed to extend from that for centralization. B: Skin incision line on the palmar side.*

Thumb reconstruction was performed without removing the Kirschner wire that had been inserted during the initial surgery. Instead, the wire was removed when it finally ruptured at 23 months post-anesthesia (14 months post-anesthesia). During the boy's fall, the Kirschner wire ruptured at the ulnar joint. Significant cosmetic improvement without radial displacement and a stable wrist with functional grip were achieved at 27 months post-anesthesia (36 months post-anesthesia).

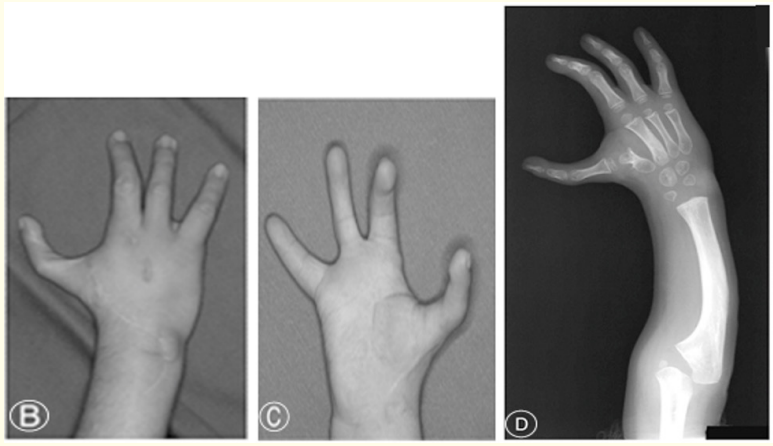


Figure 42B-42D: *The patient at 58-months-old. At 36 months after centralization (27 months after pollicization), an X-ray film demonstrates a change in the distal contour of the ulna, with widening and cupping to support the carpus, so that it resembles a normal radius. Good functional and cosmetic results were obtained.*

Traditionally, the idea of stabilizing a radial malaligned hand with bone grafting and Y-shaped distal ulnar osteotomy was proposed by Albee [45] in 1928 using a simple tibial bone graft, and in 1945, Starr [45] used preoperative skeletal traction for 6 weeks to correct the hand alignment, followed by a nonvascularized fibula graft (Figure 42).

Figure 43A and 43B: *A: Clinical view of the overcorrected radial club hand before pollicization at age 4 years. B: A radiologic view of the same hand 11 months after joint transfer.*

Conclusion of outcome and surgical for radial club hand are include below:

Surgical correction of carpal deformities and wrist stability

Long-arm radial oscillation: In type I and mild forms of type II radial oscillation, wrist instability may be largely due to inadequate radial support due to a short radius. Restoring the bone to normal length so that the distal end of the ulna is extended may improve wrist stability (Figure 43).

Figure 44: *Radiograph of a Y-formed distal ulna in hand. The situation 8 years postoperatively shows a good balance with "radial Y-fork limb" or metatarsal length of 43 mm and distal ulna length of 41 mm.*

Centralization: For over 30 years, centralization has been a common surgical procedure for severe grades II, III, and IV of varus. It involves placing the distal end of the ulna into a slot in the carpal bone aligned with the third metacarpal after releasing contractures on the radial side of the wrist (Figure 43).

Wrist immobilization: Wrist immobilization can achieve stability and alone can improve hand function. However, when performed in skeletally immature children, damage to the distal ulnar growth plate can further shorten an already shortened forearm.

Summary

The radial clubfoot is a complex congenital anomaly at the radius-anterior junction of the upper limb. Centralization by tendon transfer is a common method for maintaining the correct position of the radial clubfoot. In mild cases, lengthening of the radius is accompanied by stretching or releasing the soft tissue at the carpometacarpal joint, while maintaining wrist and forearm motion without causing damage to the growth plate.

Conclusion

There are many congenital anomalies of the upper extremities. Because of their prevalence, plastic surgeons need to be familiar with the different presentations and recognize which anomalies require further investigation due to their association with systemic conditions. In particular, thumb atrophy may be associated with underlying syndromes or associations, while ABS often presents with multiple affected limbs and body parts. Accurate classification of hand anomalies helps determine both the reconstructive technique and the timing of surgery. Timing of surgery minimizes the risk of anesthesia to the developing brain while maximizing hand function and aesthetics.

Bibliography

1. Oberg KC. "Classification of congenital upper limb anomalies: towards improved communication, diagnosis, and discovery". *Journal of Hand Surgery (European Volume)* 44.1 (2019): 4-14.
2. Sullivan MA and Adkinson JM. "Congenital hand differences". *Plastic Surgical Nursing* 36.2 (2016): 84-89.
3. Ekblom AG., *et al*. "Epidemiology of congenital upper limb anomalies in 562 children born in 1997 to 2007: a total population study from Stockholm, Sweden". *Journal of Hand Surgery (American Volume)* 35.11 (2010): 1742-1754.
4. Koskimies E., *et al*. "Congenital upper limb deficiencies and associated malformations in Finland: a population-based study". *Journal of Hand Surgery (American Volume)* 36.6 (2011): 1058-1065.
5. Castilla E., *et al*. "Polydactyly: a genetic study in South America". *American Journal of Human Genetics* 25.4 (1973): 405-412.
6. Wassel HD. "The results of surgery for polydactyly of the thumb". *Clinical Orthopaedics and Related Research* 64 (1969): 175-193.
7. Zuidam JM., *et al*. "A classification system of radial polydactyly: inclusion of triphalangeal thumb and triplication". *Journal of Hand Surgery (American Volume)* 33A.3 (2008): 373-737.
8. Horii E., *et al*. "Duplicated thumb bifurcation at the metacarpophalangeal joint level: factors affecting surgical outcome". *Journal of Hand Surgery (American Volume)* 22A.4 (1997): 671-679.
9. Harris MF. "Milestones in pediatric anesthesia risk". In: Carter PR, ed. Reconstruction of the Child's Hand. Philadelphia: Lea & Febiger (1991): 3.
10. Flatt AE. "Growth, Size and Function". In: Flatt AE, ed. The Care of Congenital Hand Anomalies. St. Louis, MO: Quality Medical Publishing, Inc., i (1994): 25.
11. Hung NN. "Bifid thumb type IV in children: transferring an epiphyseal segment of the proximal phalanx with insertion of the abductor pollicis brevis tendon". *Journal of Children's Orthopaedics* 4.6 (2010): 525-537.
12. Montgomery WF. "Observation on the spontaneous amputation of the limbs of the fetus in utero, with an attempt to explain the occasional cause of its production". *Dublin Journal of Medical and Chemical Science* 1 (1832): 140-144.
13. Gorlin RJ., *et al*. "Syndromes of the head and neck, 4th edition". Oxford University Press, New York: 10-13.
14. Upton J. "Constriction ring syndrome". In: Mathes SJ, Hentz UR (eds) Plastic surgery. Elsevier, Philadelphia (2006): 185-213.
15. Streeter GL. "Focal deficiencies in fetal tissues and their relation to intrauterine amputation". *Contributions to Embryology* 22.126 (1930).

16. Bamforth JS. "Amniotic band sequence: Streeter's hypothesis reexamined". *American Journal of Human Genetics* 44.3 (1992): 280-287.

17. Gogola G. "The pediatric upper extremity". 1st edition (Abzug J, Kozin S, Zlotolow D, eds.). New York, NY: Springer (2015).

18. Percival NJ and Sykes PJ. "Syndactyly: a review of the factors which influence surgical treatment". *Journal of Hand Surgery (British Volume)* 14B.2 (1989): 196-200.

19. Wilkie AOM., *et al*. "Apert syndrome results from localized mutations of FGFR2 and is allelic with Crouzon syndrome". *Nature Genetics* 9.2 (1995): 165-172.

20. Jenkins D., *et al*. "RAB23 mutations in Carpenter syndrome imply an unexpected role for hedgehog signaling in cranial-suture development and obesity". *American Journal of Human Genetics* 80.6 (2007): 1162-1170.

21. Howard TD., *et al*. "Mutations in TWIST, a basic helix-loop-helix transcription factor in Saethre-Chotzen syndrome". *Nature Genetics* 15.1 (1997): 36-41.

22. Cohen DM., *et al*. "Acrocephalopolysyndactyly type II--Carpenter syndrome: clinical spectrum and an attempt at unification with Goodman and Summitt syndromes". *American Journal of Human Genetics* 28.2 (1987): 311-324.

23. Muenke M., *et al*. "A common mutation in the fibroblast growth factor receptor 1 gene in Pfeiffer syndrome". *Nature Genetics* 8.3 (1994): 269-274.

24. Dobyns JH. "Syndactyly". In Green DP, Hotchkiss RN (eds): Operative Hand Surgery, ed 3. New York, NY: Churchill Livingstone, volume 1 (1993): 346-363.

25. Johnston O and Kirby VV. "Syndactyly of the ring and little finger". *American Journal of Human Genetics* 7.1 (1955): 80-82.

26. Temtamy SA and McKusick VA. "The genetics of hand malformations". *Birth Defects: Original Article Series* 14.3 (1978): 1-619.

27. Kelikian H. "Syndactyly, in Congenital Deformities of the Hand and Forearm". Philadelphia, PA: WB Saunders (1974): 331-407.

28. Orel H. "Kleine Beitrage zur Vererbungswissenschaft. Synostosis metacarpi quarti et quinti". *Z-Anatomy* 14 (1928): 244-252.

29. Upton J. "Congenital anomalies of the hand and forearm". I: McCarthy JG, red. Plastic surgery. New York, NY: WB Saunders, 8 (1990): 5218-5398.

30. Tickle C. "Embryology". I: Gupta A, Kay SPJ, Scheker LR, red. The growing hand. London: Mosby, 5 (2000): 25-32.

31. Flatt AE. "Webbed fingers". I: Flatt AE, red. The care of congenital hand anomalies. St. Louis, MO: Quality Medical Publishing (1994): 228-275.

32. Waters PM and Bae DS. "Embryology and development". I: Waters PM, Bae DS, red. Pediatric hand and upper limb surgery. A practical guide. Philadelphia, PA: Lippincott Williams & Wilkins 1 (2012): 1-11.

33. Ikegawa M., *et al*. "Syndactyly and preaxial synpolydactyly in the single Sfrp2 deleted mutant mice". *Developmental Dynamics* 237.9 (2008): 2506-2517.

34. Dao KD., *et al*. "Surgical treatment of congenital syndactyly of the hand". *Journal of the American Academy of Orthopaedic Surgeons* 12.1 (2004): 39-48.

35. Tonkin MA. "Failure of differentiation part I: Syndactyly". *Hand Clinics* 25.2 (2009): 171-193.

36. Goodman FR., *et al*. "Synpolydactyly phenotypes correlate with size of expansions in HOXD13 polyalanine tract". *Proceedings of the National Academy of Sciences of the United States of America* 94.14 (1997): 7458-7463.

37. Buck-Gramcko D. "Radialization as a new treatment for radial club hand". *Journal of Hand Surgery (American Volume)* 10A.6.2 (1985): 964-968.

38. D'Arcangelo M., *et al*. "Radial club hand". In: Gupta A, Kay SPJ, Scheker LR, eds. The Growing Hand: Diagnosis and Management of the Upper Extremity in Children. London: Harcourt Publishers Ltd (2000): 147-170.

39. Bayne LG and Klug MS. "Long term review of the surgical treatment of radial deficiencies". *Journal of Hand Surgery (American Volume)* 12.2 (1987): 169-179.

40. Heikel HV. "Aplasia and hypoplasia of the radius: studies on 64 cases and on epiphyseal transplantation in rabbits with the imitated defect". *Acta orthopaedica Scandinavica, Supplementum* 39 (1959): 1-155.

41. Manske PR., *et al*. "Centralization of the radial club hand: an ulnar surgical approach". *Journal of Hand Surgery* 6.5 (1981): 423-433.

42. Sayre RH. "A contribution to the study of club-hand". *Transactions of the American Orthopedic Association* 6 (1893): 208-216.

43. Lister G. "Reconstruction of the hypoplastic thumb". *Clinical Orthopaedics and Related Research* 66 (1969): 109-112.

44. Takagi T., *et al*. "Bone lengthening of the radius with temporary external fixation of the wrist for mild radial club hand". *Journal of Plastic, Reconstructive and Aesthetic Surgery* 67.12 (2014): 1688-1693.

45. Carroll RE. "Pollicization". In Operative Hand Surgery(ed.) Green DP, 2nd edition (New York: Churchill Livingstone) (1988): 2263-2280.

46. Albee FH. "Formation of radius congenitally absent. Conditions seven years after implantation of bone graft". *Annals of Surgery* 87.1 (1928): 105-110.

47. Starr DE. "Congenital absence of the radius. A method of surgical correction". *The Journal of Bone and Joint Surgery. American Volume* 27A.4 (1945): 572-577.

Volume 14 Issue 4 April 2025
©All rights reserved by Nguyen Ngoc Hung.