Chapter 29

Congenital Hand Deformities

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Since it is not always easy to find an up to date article on certain congenital hand deformities, several common ones are included here with simple guidelines for treatment. There are many deformities but only a few are seen frequently in Africa. These are syndactyly, duplication, radial dysplasia or radial club hand, macrodactyly or gigantism and constriction bands.

The author realizes that many may not operate on these, but this is summary of the approach to the common anomalies. For most congenital anomalies, one should have loupes for magnification. Pollicization of the thumb will not be discussed since one needs magnification for the dissection.

All congenital deformities occur in the first 8 weeks of gestation and usually between 4 and 8 weeks. Etiology is often genetic but may be sporadic and spontaneous. An increasing number of deformities are being linked to a specific gene abnormality. Children are aware of a difference by age 2 and often seek an explanation by age 4 and by age 5-6 they are self-conscious of the abnormality. Thus, it is important to correct these deformities by school age.

**Syndactyly**

According to the literature, syndactyly is uncommon in Africans but these cases will likely show up at a large referral hospital. Syndactyly is twice as common in males and it may be inherited or spontaneous or associated with a syndrome as Apert’s. Syndactyly of the toes is also seen with congenital syndactyly.

Syndactyly is classified according to the length of the syndactyly—partial to complete and according to the tissue within the syndactyly—simple if only skin is involved, complex if bone is also involved and complicated if there are extra, missing or angulated bones within the syndactyly.

Long/ring syndactyly is most common. If syndactyly involves the border digits as small/ring or thumb/index, these must be divided first at 6 months to prevent angulation of the longer digit. Syndactyly of central digits may be delayed to 18 months when the finger has doubled in size and with less fat.
Surgery is easier at this time with fewer complications. Never release adjacent webs at the same time as there is not enough skin for both releases, but non-adjacent fingers may be divided at the same time. Syndactyly release always requires skin grafts since the circumference of two fingers together is 1/3 less than two fingers separated. Full-thickness skin grafts are more pliable and durable and thus preferred. For multiple skin grafts the groin is preferred since the closure of the incision is in the inguinal crease. The axis of the donor site is from pubic tubercle to the Anterior Superior Iliac Spine (ASIS). See chapter on skin grafts for the technique of harvesting these grafts. The family must be warned that the recipient areas on the fingers will always be dark in a dark skinned individual.

Surgery is performed with a tourniquet and one dose of a cephalosporin is used preoperatively. The author releases the tourniquet after the web space is released and while the skin graft is harvested. Bleeding is controlled by pressure or gauze soaked in dilute adrenalin. The tourniquet is once again inflated before skin grafting.

The key to successful surgery is creating a good web space. Therefore, the length of the dorsal flap must be sufficient to reach the volar margin of the web space and a few mm. more to overcorrect for possible “web creep,” which is a slow loss of web space depth over time. In multiple adjacent webs, it may be difficult to determine the proximal end of the web space. It is approximately half way between the PIPJ crease and the distal palmar crease. Since skin grafting will be necessary, a dorsal flap with an extra mm. or more in length will allow for a good deep web space and will not affect closure of each side of the adjacent fingers. The flaps between the adjacent fingers can be drawn out in a zigzag fashion. The width of each flap should be from midline to midline of the adjacent digits with the apex at the crease or midway along each phalanx. The opposite side should be a mirror image of the other. See Figure 1 below.
Different technique: Note the length of the dorsal flap which easily transfers to the web with a little extra to account for the possible future web creep (Courtesy of Marybeth Ezaki MD)

Sometimes the defect on each side may be nearly approximated and some may defat the flaps to attempt closure of the defect but this could be risky. Closure should be with absorbable 4 to 6-0 suture—chromic or Monocryl. Dressing should be with non-adherent gauze such as Vaseline or Xeroform, moist cotton balls, dry gauze and a bulky dressing covering the fingers completely and a long arm splint with the elbow flexed for elevation. The author inspects the dressing at 7 days but does not remove the entire dressing for 2 weeks. There is little gained by early complete removal of the dressing. There are times when K-wires may be placed down the fingers for immobilization to allow the grafts to take better (or #21 hypodermic needles may be used in young children if K-wires are scarce). Even with a bulky dressing and splinting, fingers in small children may still move around and the grafts may not take well. Adjacent web spaces may be divided after 3-6 months.

The diagram below will show the method the author uses to deal with synostosis of the distal phalanx where division of the bone may leave some exposed bone along the edges. At times a small amount of bone, nail and nail bed along the side of the synostosis will need to be removed in order for distant the finger-tip flaps shown below to cover the bone.
Complications include the rare case of infection and loss of the grafts. If grafts are lost completely, then repeat grafting is necessary.

Complex syndactyly where just the distal phalanx tufts are involved will require division of the synostosis as described above. Complex and even complicated syndactyly where several fingers are involved as in Apert’s Syndrome requires careful planning and some innovation in carrying out the release. When all or several fingers are involved in a complicated syndactyly, release of one web space at a time is recommended. Because of the abnormal bones it may not be possible to leave the child with a normal number of digits. In Apert’s, symphalangism with fusion of the PIP joints and stiff fingers is also present and the PIPJ crease as well as the normal web space crease are often missing. The goal is to give the child as normal appearing hand as possible but most often it will only be a “helping hand” with no motion at the PIPJ.
Congenital syndactyly is not the same as burn syndactyly. In burn syndactyly, the fingers and web spaces are scarred and the skin is not pliable and does not transfer easily as in congenital syndactyly. For burned syndactyly one should use the hour-glass flap as described in the burn reconstruction chapter.

**Camptodactyly**

Camptodactyly means “bent finger.” This can occur sporadically or as part of a syndrome. It usually involves the small and ring fingers at the PIPJ. When camptodactyly is part of a syndrome all the fingers may be affected. When it occurs in childhood, the cases are equally divided between male and female. At puberty it may occur in girls without a previous history. If diagnosed in early childhood, splinting may be attempted but must be continuous over a long period of time. This condition is mentioned here only to recommend that **operative correction not be attempted.** Surgery will be difficult and is often without significantly improvement especially in older children with a deformed, chisel-shaped head of the proximal phalanx. This flexion deformity is usually just a cosmetic problem and the patient will function fairly well even with the deformity. If one has considerable experience in congenital hand surgery, techniques may be found in Green’s *Operative Hand Surgery.*

![Fig 9](image1.jpg) ![Fig 10](image2.jpg)

**Camptodactyly**

On left is typical 90° flexion deformity of PIPJ in young child

On right is chisel-shaped head deformity of proximal phalanx and this is indicative of a poor result with surgery

**Duplication**

Post axial or small finger duplication is quite common and it often consists of a small tag along the ulnar border of the hand. This is often excised or tied off soon after birth. In the black population this is not associated with systemic
findings whereas in Caucasians it is associated with congenital systemic conditions.

Preaxial or **thumb duplication** is sporadically seen and is usually unilateral. It is classified according to the Wassel Classification from I to VII:

I. Bifid distal phalanx

II. Duplicated distal phalanx

III. Bifid proximal phalanx

IV. Duplicated proximal phalanx—most common

V. Bifid metacarpal

VI. Duplicated metacarpal

VII. Triphalangism with duplication

(Triphalangism without duplication is inherited as autosomal dominant and associated with many anomalies)

**Type IV:** Since this is most common it will be discussed here. The aim of surgery is to give the child one good, normal appearing thumb with good opposition, alignment and a normal first web space without scarring. Most often the ulnar duplication is largest or the same size as the radial duplication and the radial duplication can be removed, sparing the first web space. The nail bed of the retained thumb is preserved. Surgery is performed between one and two years of age.

Operative approach requires salvage of the radial MPJ collateral ligament and insertion of abductor pollicis brevis into the radial base of the proximal phalanx which will be discarded. The periosteum on the radial side of the proximal phalanx is stripped off with the ligament and muscle insertion. The duplication may have a normal appearing ulnar thumb or both parts may be
angulated away from each other. In such cases, it is important to leave behind a straight thumb. This requires:

♦ Re-inserting the radial collateral ligament and abductor pollicis brevis into the base of the remaining proximal phalanx and also realigning the tendons and bones.
♦ Occasionally the tendons to the discarded part are large and need to be saved and reinserted into the tendons of the preserved thumb.
♦ The metacarpal head may need an osteotomy on the radial side to remove the articular surface for the discarded proximal phalanx and to trim down the large metacarpal head to allow easy insertion of the ligament and APB into the base of retained proximal phalanx. This will also help straighten the angulation. See Fig 7 below. Black arrow points to site of osteotomy.

![Fig 13](image1)

Release of radial collateral ligament and insertion of AbPB from radial duplication, the ablated thumb, and reattaching these to the base of proximal phalanx of ulnar thumb (From Green’s Operative Hand Surgery, 5th edition, courtesy of Elsevier)

♦ Additional metacarpal osteotomies may be needed if there is a severe ulnar angulation of the distal joint.

![Fig 14](image2)

An additional metacarpal osteotomy may be required to straightened the ulnar thumb. These osteotomies are usually done at the neck of the MC but
occasionally at the neck of the proximal phalanx. This prevents an angulated zigzag deformity (From Green's Operative Hand Surgery, 5th edition, courtesy of Elsevier)

♦ In some cases imbrication of the ulnar collateral ligament at the IPJ is all that is needed.
♦ Usually a K-wire is inserted into the thumb to maintain alignment. It is left in 4 weeks. A thumb spica splint or cast is applied.
♦ At four weeks the splint and pin are removed and range of motion is allowed.
Complications include malalignment with a zigzag deformity secondary to bone, tendon or ligament imbalance, instability and stiffness.

One can refer to basic hand surgery texts for detailed description or can contact the author and he will give you further details.

**Triphalangeal Thumb without duplication** is mentioned as it is relatively common and it is often associated with other anomalies, including cardiac anomalies, and thenar muscle deficiency.

**Radial dysplasia or radial club hand:**

Radial dysplasia (radial deficiency or radial club hand) is also relatively common in Africa. It is often associated with other syndromes with a variety of findings including cardiac and blood abnormalities. The entire radial side, radial corridor, of the upper extremity may be deficient with angulation toward the radial side. This includes the bones and soft tissue. The most common finding is an absence of the entire radius with bowing of the ulna toward the missing radius—Type IV. The thumb and radial side carpal bones may or may not be present. This condition is bilateral 50% of the time. The elbows may be extended and stiff. Children with radial dysplasia should be carefully checked for possible cardiac valvular disease and hematologic abnormalities.

The goals of surgery have been to correct the radial deviation of the wrist, balance the carpus on the forearm, maintain motion, allow for growth and
promote function. In the past, recommended treatment was centralization of the ulna on the carpus with or without osteotomy to straighten the ulna. Later radialization of the ulna was recommended whereby the ulna is aligned with the scaphoid or trapezium. Though good results can be obtained, the recurrence rate with subluxation/dislocation of the ulna, relapse of wrist flexion, stiffness and poor function is high. With these techniques it is difficult to preserve the ulnar epiphysis. Therefore the present treatment that is recommended can be done at the district hospital: early stretching and splinting in the newborn

Then one can refer the patient to a major center for further surgery at 18-24 months which should include:

- Release all tight structures on the radial side including skin, tendons, capsule, radial anlage, etc.
- Ulnar osteotomy with preservation of ulnar epiphysis—do not try to centralize since this often damages the ulnar epiphysis
- Use ulnar-sided flap to reconstruct skin loss on radial side of wrist—see below
- Possible ECRL/ECRB to ECU or imbricated ECU for tendon balance and splint for three weeks

In adolescence perform a wrist fusion. Waiting until adolescence allows full growth without disturbing ulnar epiphysis

Contraindications for surgery anytime include:

- life threatening systemic conditions, as cardiac anomalies
- bilateral stiff elbows: straightening the hand removes it away from face and mouth
- older adapted children—have learned to use their deformed hands

Summary: Extensive surgery with centralization or radialization is usually NOT recommended and especially not recommended in the district hospital setting. The complications of aggressive surgery must be explained to the parents so they will not be discouraged when the hand is not straightened in early childhood.

Thumb Hypoplasia

A small or absent thumb is often associated with radial dysplasia but maybe seen as an isolated deformity. Thumb hypoplasia may be minimal, Type I, or the thumb may be absent, Type V. Small but functional thumbs, Type I to Type IIIA, can be augmented with tendon transfers and a Z-plasty to deepen the first web space. Types IIIB to V require pollicization of the index finger—a
demanding procedure. The difference between IIIA and IIIB is the presence of a CMC joint. IIIA has a CMC joint. In IIIB the CMC joint is absent.

![Fig 18](image1.png) ![Fig 19](image2.png) ![Fig 20](image3.png)

Fig 18: shows normal thumb; Fig 19: hypoplastic thumb; Fig 20: X-ray showing absent CMC joint in Type IIIB hypoplastic thumb. IIIB thumb hypoplasia requires a pollicization

**Macrodactyly or Gigantism:**

![Fig 21](image4.png) ![Fig 22](image5.png)

Macrodactyly/Gigantism secondary to lipofibromatosis

Gigantism in early childhood is commonly seen and is usually associated with lipofibromatosis. It is also seen with neurofibromatosis, vascular anomalies, and other bony conditions but usually not until adolescence or later in life.

Lipofibromatosis is seen in childhood with uniform enlargement of fingers, toes and sometimes the entire extremity. All anatomic structures enlarge including the bones which enlarge in width and length and the bone age is advanced. There is marked increase in subcutaneous fat and digital nerves.

**The author only recommends partial or complete amputations** in the district hospital. Other treatment consists of debulking procedures and though described below is NOT recommended. (When seen early debulking can be attempted in two or more stages—debulking one side of a finger at a time. Debulking is combined with epiphyseal arrest at each joint once the finger
reaches adult length which is often the time the patient is first seen. Unfortunately, it is difficult to control the transverse axis of growth.) Debunking has limited success, dependent on the age that surgery is begun. Normally there is still continued growth and amputation is often the final result anyway.

Often the parents refuse early ray amputation even though this is the best and most definitive procedure. Ray amputation including the metacarpals of one or more abnormal fingers will often control this condition. The thumb is different and when the thumb is involved, conservative debunking with epiphysiodesis is usually initially recommended, but amputation with later pollicization of a normal index or long finger will often be required. With large “banana fingers” that are several times the length and breadth of a normal adult finger, ray amputation will be necessary. If only one finger is amputated, it is likely no one but a parent will recognize the missing finger.

**Gigantism is commonly seen in the toes.** When gigantism of a large toe is seen early, debunking and epiphyseal arrest can be attempted. With other toe involvement, a ray amputation of one or more toes will be necessary. This amputation is carried back the tarsal/metatarsal joint.

**Constriction Bands:**

The cause for constriction bands is controversial. It has long been thought to be secondary to amniotic bands. Some have linked constriction bands to a Band Syndrome which includes cleft lip and other facial clefts, clubfeet and constriction bands. It is also associated with a type of syndactyly of the fingers, acrosyndactyly. The extremity is normal proximal to the band but with severe bands, the extremity distal to the band may have significant swelling or even amputations as seen in acrosyndactyly.
Surgery should be performed early when there is severe swelling of the distal extremity. If the swelling is mild, then surgery can be delayed until the child is older. Release is best done with multiple Z-plasties—4-6 according to the size of the extremity or finger. A rim of skin and soft tissue is removed on both sides of the band and the adjacent tissue undermined in a subcutaneous plane to allow tension free closure. The tissue on either side of the band must be completely removed nearly down to the bone and one is often concerned about sufficient distal blood supply. There is always ample blood supply so it is important to remove the depths of the band down to normal soft tissue/bone. It may take some time for the distal swelling to resolve. During this time a compression bandage may be used, wrapping from distal to proximal.

**Congenital” Trigger Thumb**

This refers to a flexed IP joint of the thumb which is usually not noticed until several months after birth. At one time it was thought to be congenital but a number of studies have shown this to be neither congenital or trigger. This
condition has not been diagnosed at birth and it does not trigger. It is a fixed flexion deformity. Conditions such as this are rarely noticed in a young child, hence the delay in diagnosis. In the past it was felt that if one waited, the condition would resolve. Though this may occasionally happen, it rarely resolves without surgical intervention.

On exam, the IPJ of the thumb is in fixed flexion and a nodule may be palpated at the palmar digital crease of the thumb. This is Notta’s node and is a swelling on the flexor pollicis longus. It is not possible to extend the thumb.

Treatment consists of division A-1 pulley. The surgery is usually delayed for the child to get a little older for anesthesia and for the hand structures to grow. At 6 months the A-1 pulley can be released under general anesthesia and with a forearm tourniquet. Great care is required to carefully identify and retract the digital nerves, especially the radial digital nerve. The nodule is easily felt and the FPL tendon is identified and the A-1 pulley identified and divided at the palmar digital crease. After division the thumb is extended. Some would splint the thumb in extension for a few days.

The differential diagnosis is flexion of the MPJ with hypoplasia of the extensor tendon, EPB, without extension at the MPJ. This condition usually resolves with the MPJ splinted in extension for several months. The IPJ is not flexed and Notta’s node is not present at the IPJ. If the MPJ flexion is persistent, then an EIP tendon transfer may be done.

One may contact the author of this chapter for specific and detailed information about these conditions: llcartermd@gmail.com