Parent/Patient Education Series: Ulnarization: Treatment Strategy for Severe Radial Dysplasia

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Radial dysplasia, also known as radial club hand, can vary in severity. This diagnosis can be an isolated growth anomaly, or it can be related to multiple syndromes with varied coexisting medical issues.

Syndromes Related to Radial Dysplasia:

- Thrombocytopenia-Absent Radius Syndrome (TAR)
- Fanconi Anemia
- Holt-Oram Syndrome
- Okihiro Syndrome
- Lacrimo-Auriculo-Dento-Digital Syndrome (LADD)
- Nager Syndrome
- Baller-Gerold Syndrome
- Richieri-Costa-Pereira Syndrome
- Roberts Syndrome
- VACTERL Syndrome (Vertebral anomalies / Anorectal Malformations / Cardiovascular anomalies / Tracheoesophageal fistula / Esophageal atresia / Renal or Radial anomalies / Limb deficits)

CLASSIFICATION

There are several classification systems to describe the severity of radial dysplasia. The Heikel classification of radial dysplasia in Figure 1 is similar to the Bayne and Klug classification; the differences are not clinically significant. The radius is one of the two large bones of the forearm. It extends from the side of the elbow to what would normally be the thumb-side of the wrist. The ulna is the other large bone of the forearm that runs parallel to the radius. The carpus is a group of bones that form the wrist joint bridging the hand to the forearm.

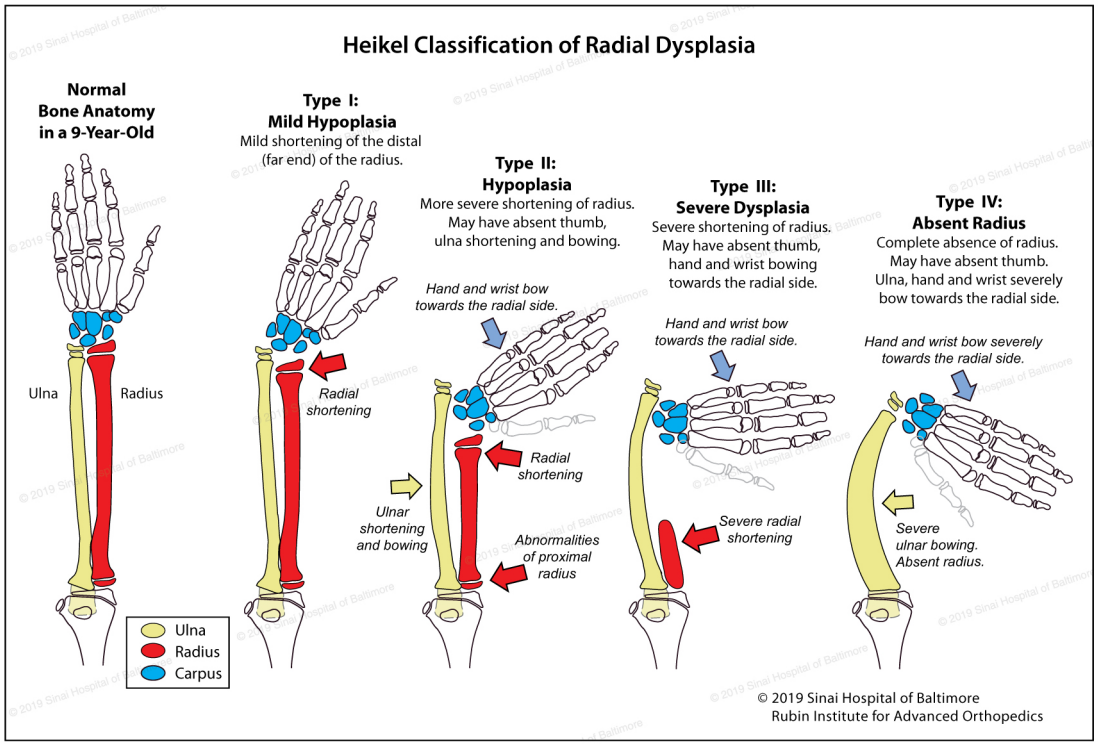


Figure 1

TREATMENT STRATEGIES DEPENDING ON CLASSIFICATION

Type I Radial Dysplasia

I treat this type of radial dysplasia with lengthening of the radius through a bone cut in the distal (wrist end) part of the radius. This strategy grows the radius to support the wrist bones (carpus) and correct the relationship between the radius and ulna. This is accomplished by either a one- or two-stage treatment plan in the mid-childhood ages. The first stage is performed by application of an external fixation device to the hand and wrist for gradual re-positioning of the hand and wrist into a normal position if needed. The second stage consists of adjusting the external fixation to hold the wrist and hand in the corrected position and the initiation of lengthening of the radius through to a distal radial osteotomy (bone cut). This type of strategy usually needs to be repeated towards the end of growth to accomplish permanent correction. The end of long bone growth usually occurs around the age of 14 years for females and 16 years for males.

Type II Radial Dysplasia

This type of radial dysplasia falls in between the above treatment strategy and the procedure called ulnarization. The vast majority of the Type II radial dysplasia patients in my practice have significantly abnormal proximal (upper end) and distal (lower end) growth areas of the radius. This translates to significant growth arrest in the radius with overt shortening during growth. This limits the ability to lengthen the radius enough to make the above strategy viable and successful.

Types III & IV Radial Dysplasia

I have treated both of these severe types of radial dysplasia successfully with ulnarization. This procedure is a modification of Dr. Buck Gramko's radialization procedure by Dr. Dror Paley. There are multiple differences in both surgical strategy and surgical technique when comparing ulnarization to radialization. I have made several positive modifications to both the technique and treatment protocol. The ulnarization procedure I perform will be explained in great detail below.

SURGICAL TREATMENT OPTIONS FOR RADIAL DYSPLASIA

The most common surgical treatment options for radial dysplasia are centralization and radialization. Both of these procedures attempt to place the hand and carpus (wrist bones) on the end of the ulna. Figure 2 demonstrates the positioning of the hand/carpus after centralization, radialization and ulnarization. Both centralization and radialization are limited by high recurrence rates of deformity, growth arrest of the distal ulna physis (growth plate) and loss of wrist motion.

The centralization procedure attempts to balance the hand and wrist on a rounded end of the ulna. The tight tissues create a deforming force that eventually pulls the hand back into a radial club hand position. If the correction is maintained, this usually means the carpus (wrist bones) have fused to the end of the ulna with permanent loss of wrist motion and loss of ulna growth potential.

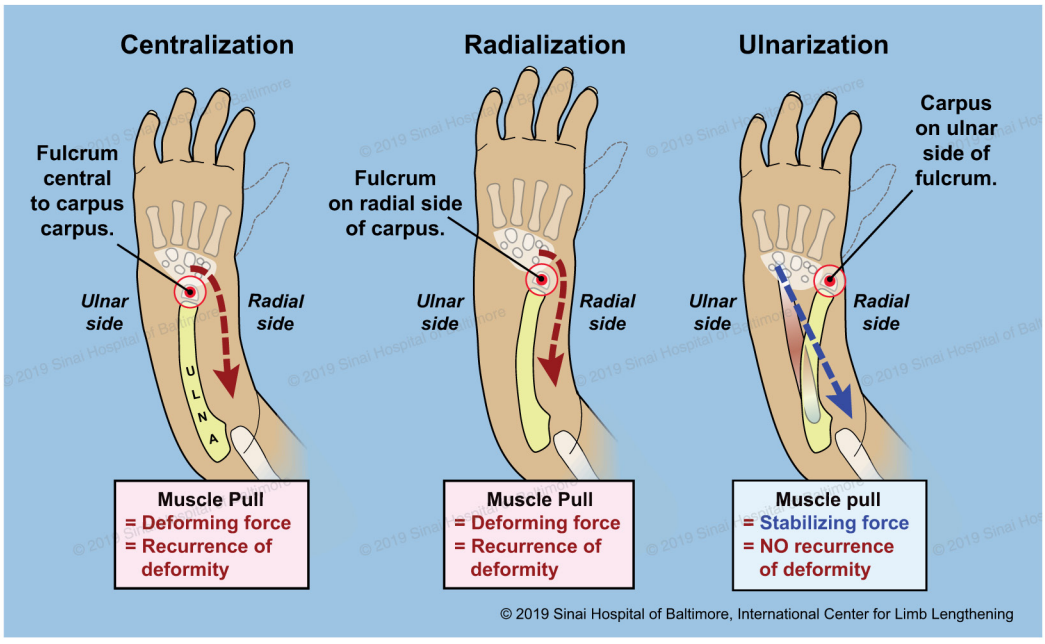


Figure 2

The radialization procedure was developed to attempt to decrease the recurrence rate of the radial club hand deformity after centralization. The radialization includes a hand and carpus position farther to the ulna (little finger) side of the distal ulna and a transfer of the radial based muscles (flexor carpi radialis, extensor radialis longus and extensor radialis brevis) to the ulnar-based muscles (extensor carpi ulnaris). The muscle transfer is an attempt to balance the deforming forces around the hand and wrist. Unfortunately, these muscles that are used in the radialization are rarely present in the severe forms of radial dysplasia. If these muscles are present, they are very small and weak. Similar to the centralization procedure, the radialization procedure has a very high incidence of recurrent club hand deformity, distal ulna growth arrest and stiffness of the wrist.

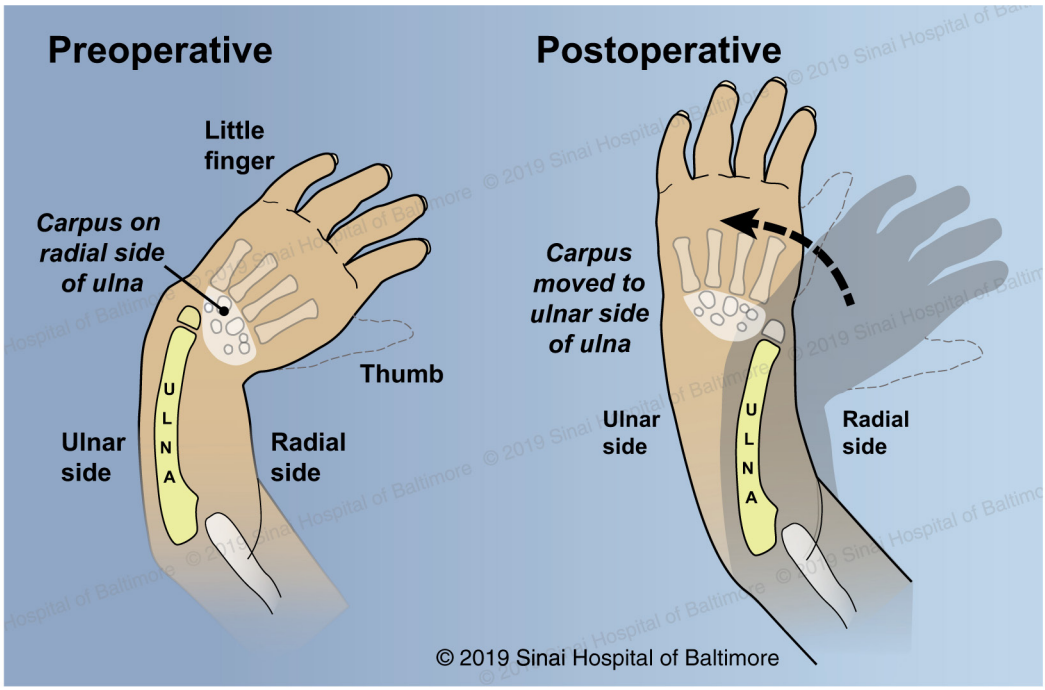
Figure 2 also demonstrates a schematic of the ulnarization procedure where the hand and carpus are moved to the little finger side of the ulna. The deforming forces are now converted to stabilizing forces by using the distal ulna as a fulcrum. The ulnarization procedure has the advantages of maintaining wrist motion and distal ulna growth. Also, I have not experienced a recurrence of radial club hand deformity over the last 14 years of using this procedure.

The X-ray shows a 19-year-old female who had a centralization procedure at the age of 18 months. The wrist is fused and the ulna is only 8.5 cm (3 1/2 in) in length.

ULNARIZATION STRATEGY FOR RADIAL DYSPLASIA

As described above, the ulnarization procedure was developed as a modified approach to radial dysplasia by Dr. Dror Paley. The concept was to perform a comprehensive release of the radial club hand deformity through an extensive exposure of the volar (palm side) of the wrist and forearm. The wrist/carpal joint capsule is released and the hand and carpus is transferred to the ulna side of the ulna. As demonstrated in Figure 2, the deforming forces causing the radial club hand deformity are now converted into stabilizing forces using the distal ulna as a fulcrum.

This position is held with an external fixation device postoperatively for three months to allow fibrous healing and stabilization of the new hand position. This essentially creates a syndesmosis or fibrous joint connection between the carpus (wrist bones) and the distal ulna. This is similar to the joint between the distal tibia and fibula just above the ankle joint. The advantages of a syndesmosis type joint formation in the wrist are maintained wrist motion and hand position stability.



The previous radial club hand correction techniques, both centralization and radialization, create either a synchondrosis (cartilage to cartilage connection) or

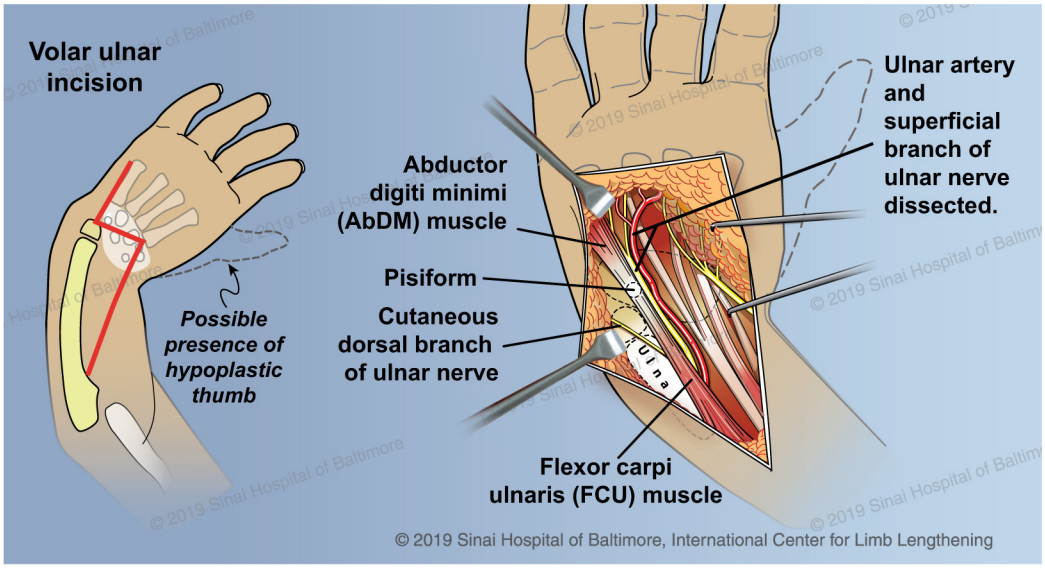
synostosis (bone to bone connection) resulting in the loss of the wrist joint and wrist motion. Another technical difference in the ulnarization technique is the transfer of the Flexor Carpi Ulnaris muscle (FCU) to the back of the hand to increase wrist extension (pulling the hand up) power. The centralization and radialization techniques describe transferring muscles on the radial side of the forearm; however, these muscles are rarely present.

In contrast, the ulnarization technique utilizes the flexor muscle on the ulna side of the forearm that is always present and very strong. This muscle is detached from its connection on the palm side of the wrist, where it acts more as a deforming force, and is transferred or moved to the back of the wrist to improve wrist position and motion. Enhancing wrist extension is important due to the fact that the most functional hand position is neutral position to 15-30 degrees of wrist extension.

ULNARIZATION TECHNIQUE

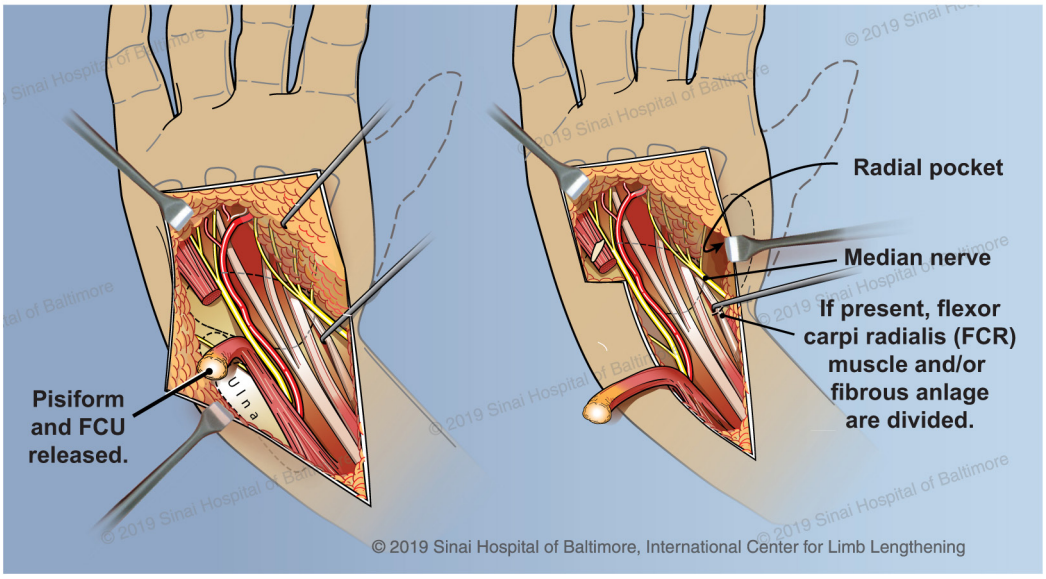
The following is a schematic explanation of the ulnarization procedure:

1. An incision is performed on the palm (volar) side of the hand and forearm in a zig-zag fashion. This style of incision helps to increase exposure and to avoid crossing the wrist flexion creases with a perpendicular incision. Crossing skin creases in a perpendicular incision would increase the risk of scar contractures of the adjacent joint, so this is avoided.
2. After the skin flaps are carefully elevated, all vital structures are identified. The ulna nerve and ulna artery are isolated and protected. The Flexor Carpi Ulnaris muscle (FCU) is identified and traced to its connection to the pissiform bone (part of the carpus).



3. A portion of the pissiform bone along with the FCU muscle is released from the remainder of the carpus and elevated in preparation for transfer later in the procedure.

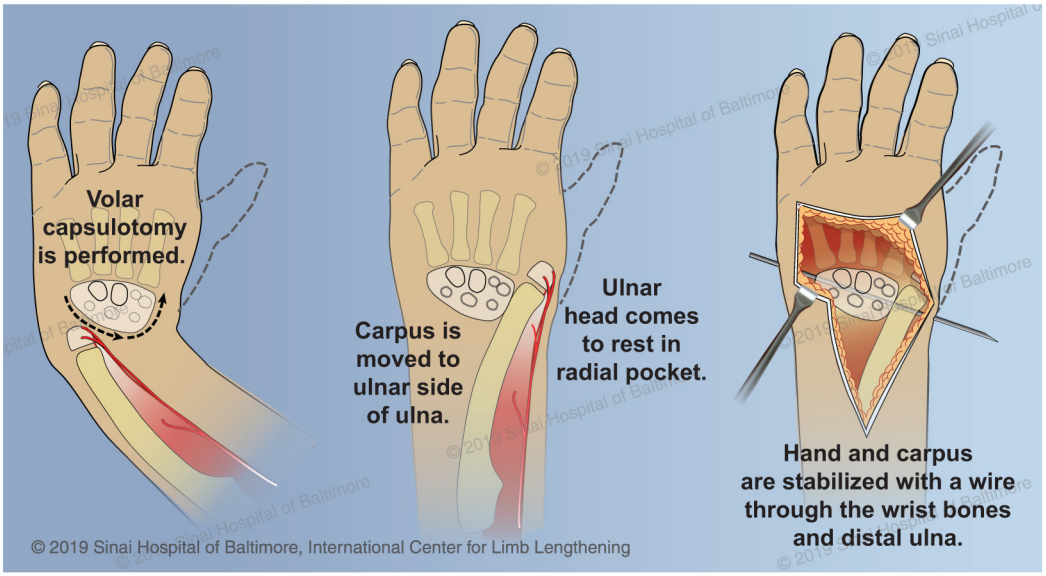
4. The central and radial zones of the wrist are explored to identify all flexor tendons, the median nerve and any fibrous radial remnant. The "radial pocket" is created to allow room for the eventual transfer of the distal ulna.



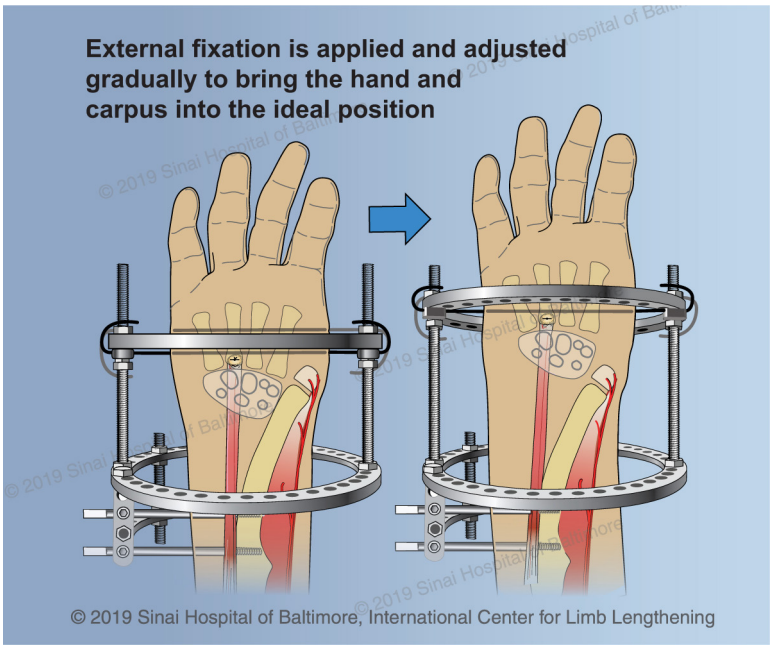
5. After all the important structures are identified and protected, the fibrous capsule around the wrist bones (carpus) is released. This is termed the volar capsulotomy.

6. Once the carpus is mobile and freed from the distal ulna, the hand and carpus are moved or transposed to the opposite side of the ulna. The hand and carpus are now located on the "ulna" side of the ulna, thus the procedure's name, ulnarization.

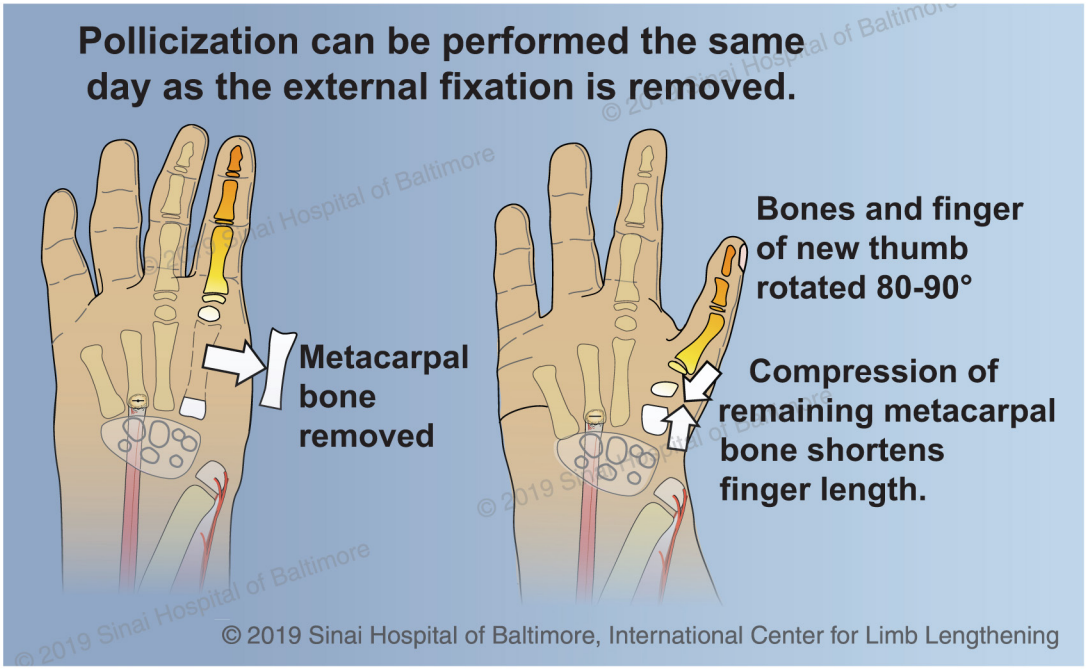
7. The hand and carpus are stabilized with a wire through the wrist bones and distal ulna. If the desired position of the carpus in relation to the distal ulna is achieved at the time of surgery, the wire is left in place and an external fixation is placed to hold the reconstruction stable. This type of external fixation device will not need adjustments or turns.



8. The correct position of the wrist bones or carpus is next to the distal portion of the ulna. The most distal part of the ulna is still cartilage in most toddlers and does not appear on X-rays. It is not uncommon for the hand and wrist to "settle" more distal to the correct position at the time of surgery. If this occurs during surgery, then the initial stabilizing wire that is inserted between the carpus and distal ulna is removed after the external fixation device is applied. The external fixation is adjusted gradually during the first two weeks after surgery to slowly distract or pull the hand and carpus into the ideal position. After the ideal hand and carpus position is achieved and confirmed with X-rays, the external fixation device is maintained for a total of three months.



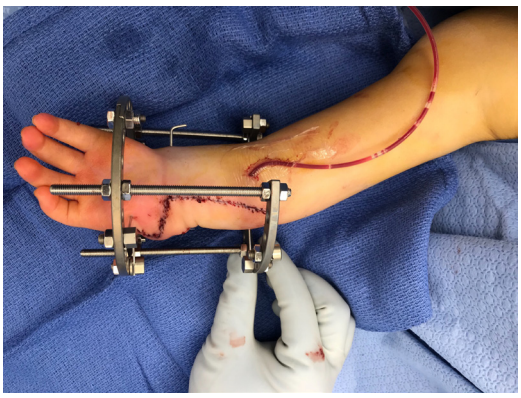
9. If a pollicization (creating a functional thumb by transferring another finger to the thumb position) is needed, this can usually be done at the time of the removal of the fixator.



A case example of intraoperative photos before and after ulnarization prior to application of the external fixation device is below.

The following is a case example of intraoperative photos before ulnarization, after ulnarization and external fixation, after external fixation removal at three months, and after pollicization was performed the same day as the external fixation removal.

Before Ulnarization



After Ulnarization and External Fixation

External Fixation Removal at 3 months

Pollicization performed same day as external fixation removal.

CONCLUSION

Ulnarization treatment provides many advantages over the more commonly performed centralization and radialization procedures. There has been no recurrence of the radial club hand deformity in our fourteen years of performing ulnarization surgery. There is no growth arrest of the ulna, so it can continue to grow for many years as the child grows. The resulting hand is in a much more functional position with improved grip strength and wrist mobility.

International Center for Limb Lengthening patients benefit from our team-centered approach with world-renowned pediatric orthopedic surgeons, anesthesiologists with extensive experience working with pediatric patients, and specialized physician assistants, nurses and physical therapists. We also have a large group of patient families who have children undergoing radial club hand treatment that offer support through our International Center for Limb Lengthening (ICLL) Facebook Group, ICLL Families. We help patients with radial club hand achieve their best possible result.