Changing Paradigms in the Treatment of Radial Club Hand: Microvascular Joint Transfer for Correction of Radial Deviation and Preservation of Long-term Growth

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Radial longitudinal deficiency (RLD) is a congenital deformity of the upper extremity which can present with a spectrum of upper limb deficiencies. The typical hand and forearm deformity in such cases consists of significant forearm shortening, radial deviation of the wrist and hypoplasia or absence of a thumb. Treatment goals focus on the creation of stable centralization and functionally hand, maintenance of a mobile and stable wrist and preservation of longitudinal forearm growth. Historically centralization procedures have been the most common treatment method for this condition; unfortunately centralization procedures are associated with a high recurrence rate and have the potential for injury to the distal ulnar physis resulting in a further decrease in forearm growth. Here we advocate for the use of a vascularized second metatarsophalangeal joint transfer for stabilization of the carpus and prevention of recurrent radial deformity and subluxation of the wrist. This technique was originally described by the senior author in 1992 and he has subsequently been performed in 24 cases with an average of 11-year follow-up. In this paper we present an overview of the technique and review the expected outcomes for this method of treatment of radial longitudinal deficiency.

Keywords: Radial club hand, Radial longitudinal deficiency, Treatment, Microvascular joint transplantation, MTP-joint transfer
radial deviation and wrist subluxation; unfortunately, this technique has been associated with a high rate of recurrent radial deviation, physeal injury and wrist stiffness.\(^2\)\(^,\)\(^3\) In this paper we present an overview of treatment methods for radial hypoplasia and describe our concepts for the treatment of type III and IV RLD.

**HISTORY OF TREATMENT METHODS**

The surgical treatment of RLD historically has attempted to create a centralized wrist, maintain wrist motion and improve hand function by placing the hand in a more outstretched position.\(^4\)\(^,\)\(^9\) Patients with type I deformities usually need no treatment, while patients with Bayne type II deformities can usually be treated conservatively with wrist splinting or in some cases with radial distraction lengthening. Varying methods of surgical correction have been described during the past century for the treatment of patients with type III and type IV deformities. Soft tissue release with or without ulnar osteotomy and nonvascularized bone grafting have been recommended by some, but these procedures in isolation fail to maintain nonvascularized bone grafting have been recommended from some, but these procedures in isolation fail to maintain long-term wrist alignment.\(^10\) The technique of centralization was originally described by Sayre\(^11\) in 1893, and was later modified by Lidge in 1969.\(^12\) For the past 4 decades, centralization of the carpus on the distal end of the ulna in conjunction with soft tissue release has become the most commonly performed surgical procedure for the treatment of type III and type IV RLD.\(^2\)\(^,\)\(^5\)\(^,\)\(^10\)\(^,\)\(^13\)\(^,\)\(^14\) Studies have demonstrated that centralization can partially correct for radial deviation while preserving some wrist motion.\(^14\)\(^,\)\(^15\) Further modifications to the centralization procedure have been described, including radicalization by Buck-Gramcko\(^16\) in 1985, as an additional means of correcting ongoing muscle imbalance across the carpus.

In cases of severe radial deviation, centralization of the wrist can be difficult at the time of surgery, due to tight and unforgiving soft tissue contractures. Early wrist splinting and manual manipulation has been recommended from first day after birth in order to prevent tight contractures at the time of centralization surgery; however, if contractures still develop or if the child presents to the hand surgeon late after birth, soft tissue release, carpal resection and ulnar osteotomy have been performed to enable centralization of the carpus onto the ulna.\(^8\)\(^,\)\(^9\)\(^,\)\(^13\)\(^,\)\(^15\) In order to avoid this extensive dissection, bony resection and acute stretching of the neurovascular structures, Kessler\(^17\) introduced preoperative soft tissue distraction prior to the centralization procedure using an apparatus originally developed for distraction lengthening of fingers and the forearm. Smith and Greene\(^17\) subsequently presented their small series of successful preliminary soft tissue distraction using an Orthofix external distractor in 4 patients. Pre-centralization soft tissue distraction is now recommended in cases with severe RLD or in neglected late-presenting patients needing significant correction of their wrist deformity.\(^18\)\(^-\)\(^22\)

**FAILURE OF CENTRALIZATION PROCEDURES**

Even though centralization has been the preferred method of treatment for severe RLD, several studies have shown a recurrence of wrist radial deviation with long-term follow-up. Lamb et al.\(^23\) reported a recurrence of radial wrist deviation in 7 out of 15 patients. Shariatzadeh et al.,\(^3\) McCarthy et al.,\(^24\) and Goldfarb et al.\(^25\) reported an average wrist angle of 38 degrees postoperatively that worsened to 71 degrees at follow-up; such recurrence rates have been noted in additional studies.

An additional concern with the centralization procedure is the potential injury to the ulnar physis during soft tissue dissection and cartilaginous manipulation. Heikel\(^26\) demonstrated in the 1959 that the ulna in children with RLD may grow between one half to three quarters the length of the unaffected ulna. Centralization procedures, with concomitant ulnar osteotomy and transphyseal pins can all result in potential distal ulnar physeal injury resulting in further shortening of the forearm unit. Sestero et al.\(^27\) verified this finding in 2005 when they found that untreated limbs of patients with RLD grew to 64% of normal ulnar length, while centralized limbs grew to only 48-58% of normal ulnar length. This potential for physeal injury and a high incidence of radial wrist recurrence have dissuaded the authors from using the centralization technique for the treatment of Bayne type III and IV RLD.

**VASCULARIZED SECOND METATARSAL PHALANGEAL JOINT TRANSFER FOR CORRECTION OF RLD**

Currently microsurgical techniques allow for the possibility of transferring vascularized bone grafts which can include a viable physis. Experimental transplantation of metacarpophalangeal joints with microvascular anastomoses in dogs were shown to be possible in 1979 by Hurwitz\(^28\) and several subsequent studies have shown satisfactory growth potential remaining in the epiphysis following transfer and revascularization.\(^28\)\(^,\)\(^29\) Clinical series of vascularized MTP-joint transfer with epiphyseal growth have been reported following the reconstruction of pediatric joint injuries and following toe transfer for reconstruction...
of the adactylous hand.\textsuperscript{30,31}

In 1992 the senior author introduced a novel technique for the treatment of Bayne type III and IV radial deficiency using the second metatarsal phalangeal (MTP) joint as a vascularized graft to create a new radial column within the wrist.\textsuperscript{32} The metatarsal and proximal phalanx of the second toe are transferred to the forearm to create a Y-shaped distal ulna with the potential for growth at both limbs of the “Y”. The first description of a Y constructs for the treatment of radial hypoplasia was by Albee\textsuperscript{33} in 1928 in which a tibial bone graft was used to stabilize the wrist. Starr\textsuperscript{34} modified this procedure in 1945 using nonvascularized fibular grafts; however these constructs lack the capacity for growth if performed in young children.

A detailed description of the surgical technique for vascularized MTP joint transfer for the treatment of RLD was published in 1998 and long-term results were subsequently published in 2008.\textsuperscript{6,35,36} The procedure consists of three distinct stages. The first stage involves soft tissue distraction of the wrist with the use of an external distraction device in order to centralize the wrist and create a space for placement of the MTP joint. The second stage involves MTP transfer using standard microsurgical technique. The final stage is the bony healing phase, during which the distraction device is left in place for an addition 6 to 8 weeks as the metatarsal bone heals to the shaft of the ulna. The section below provides a brief overview of the operative technique.

**Indications and Timing**

This procedure is recommended only in types III and IV radial club hands. Also a case of a failed centralization or radialization is considered mostly very suitable. Contra indications may be a complete inability to flex the elbow and a very poor quality of fingers and forearm muscles with extremely tight wrist radial deviation over 90 degrees.

In contrast to centralization procedures, which are usually performed in the 1st year of life, MTP joint transfer is performed at 2.5 to 4 years of age. The reason for delay is to allow the metacarpals time to reach adequate size to allow for placement of the distraction device. In addition, it is easier for the child to remain in the distraction device for 4 to 5 months once they are ambulating and moving out of the toddler phase of development.

During the years prior to MTP transfer we encourage the parents to stretch the wrist, and night time splinting is encouraged to prevent significant fixed soft tissue contractures. If warranted in children with thumb hypoplasia, pollicization may be performed before or after MTP joint transfer. Our preference is prior to MTP joint transfer, usually during the first to second year of life when the child begins initiating pinching between the index and long finger.

**Soft Tissue Distraction and Optional Surgical Release**

All children will require soft tissue distraction in order to straighten the wrist and provide enough space for placement of the MTP joint. Severe cases may require open soft tissue release at the time of distracter placement, particularly those cases with thrombocytopenia-absent radius (TAR) syndrome where the brachiocarpus muscle can significantly limit correction of wrist position. If soft tissue release is required it should be done through an incision over the radial aspect of the wrist. It is important to protect all superficial veins, particularly the cephalic vein in order to preserve options for recipient vessels during the second stage of the operation. Through the radial incision, all fibrotic and non functional flexors and extensors should be cut and the tightened capsule should be released. It is important to leave those tendons intact that move the fingers.

Distraction is performed with a unilaterial or a multiaxial distraction device (Swemac Orthopedics AB, Linkoping, Sweden or Biomet Microfixation Inc., Jacksonville, FL, USA) placed on the ulnar dorsal aspect of the hand (Fig. 1). The distraction device should lie parallel to the ulnar. If significant radial deviation remains after manual correction and/or surgical release in the operating room an angular distraction device should be placed so that the distraction device can remain in line with the wrist.
ulnar aspect of the wrist. The distraction device is stabilized with 2 to 3 bicortical pins placed proximally and 2 to three pins distally. Distal pins are placed through the small and ring finger metacarpal bases and if possible through the long finger base as well to provide additional stability. Small finger metacarpal purchase alone will be inadequate for distraction. The frame remains in place until bony consolidation is achieved 6 to 8 weeks following MTP transfer, so the surgeon must be satisfied with the position of the distraction device at the initial operation. Spatial frames or true Ilizarov distraction devices are not used for this procedure as the radial aspect of the hand must be free of hardware so the surgeon can perform the toe transfer during the second stage of the procedure.

Approximately 25 mm of distraction is necessary following distraction device assembly. Distraction is performed at 0.5 mm a day to avoid any injury to the physis. This means that distraction can take up to 50 days prior to second stage reconstruction. Previous publications have found that physeal injury and muscle ischemia can occur with rapid distraction speeds.\textsuperscript{37,38} The goal of distraction is to centralize the wrist and to create a space between the base of the second metacarpal and the visible distal ulna. The MTP joint construct transferred with need to be 45 to 50 mm in total length between the base of the second metacarpal and the metaphyseal flair of the ulna at three years of age (Fig. 2).\textsuperscript{39}

**Microvascular Joint Transfer**

A two team approach is used, with one surgeon preparing the wrist while the other surgeon prepares the second toe. The ipsilateral second toe is harvested using standard technique. Multiple arterial sources should be taken including the 1st and 2nd dorsal metatarsal artery and 2nd and 3rd plantar metatarsal artery. Long arterial length is recommended as many children with radial RLD may lack a radial artery and persistent median artery. In such cases a more proximal end to side anastomosis to the ulnar artery may be necessary. Flexor and extensor tendons to the toe are preserved and anchored to the remaining proximal phalanx with suture. Dorsal nerves are taken to serve the skin paddle and also to provide some proprioception to the joint and flexor and extensor tendons. The distal and middle phalanges are removed. The skin must be carefully filleted from the fibular aspect of the graft and brought to the tibial side to provide a skin paddle that will cover the radial aspect of the wrist. Care must to taken to protect the peristeal vessels going to the physis of both the proximal phalanx or metatarsal during this portion of the procedure (Fig. 3). The toe is allowed to re-perfuse for thirty minutes prior to vessel division. During the reconstruction of the wrist the toe donor site is closed using standard technique. Special attention is given to the repair of the intrametatar-
sal ligament to prevent spreading of the toes later in life (Fig. 4).

As the toe is being prepared the wrist is opened over the radial aspect. Careful dissection of the cephalic vein is performed (if present) or 2 large dorsal veins. Occasionally a radial artery may be identified, but more commonly a persistent median artery is present which can be utilized for anastomosis in an end to end fashion. All intact finger flexors and extensors are preserved while remnants of tight muscles, brachioradialis, flexor carpi radialis, and extensor carpi radialis are divided and separated.

The MTP construct will be placed into a space extending from the metaphyseal flair of the ulna to the base of the second metacarpal (or remaining radiocarpal cartilage). The bony preparation for MTP joint transfer first involves the identification of the distal ulnar physis. The position of the distal ulnar physis and ulnocarpal joint are marked with 25-gauge needles and protected. The base of the second metacarpal is also identified and a small notch is created to accept the distal end of the proximal phalanx. In cases of TAR syndrome or in patients with intact radial cartilage, the proximal phalanx of the MTP joint construct may be inset into the remaining radial carpal remnant instead of the second metacarpal base. Just proximal to the metaphyseal flair of the distal ulna, approximately 10 mm proximal to the physeal plate, an osteotome is used to create a cortical strut based on a proximal hinge. A 20-mm bone flap is pulled proximal and radial to form a platform upon which the toe will be transferred.

Reconstruction

The toe is transferred to the forearm and the metatarsal base is placed on the ulnar platform while the distal component of the proximal phalanx is placed against the base of the second metacarpal or against the scaphoid if present. The joint is positioned in 15-20 degrees of flexion which makes for the greatest stability. The tendons which were saved with the toe are re-attached to the remaining radial flexor and extensors tendon muscle bellies to help provide stability to the MTP joint. Small K-wires are then used to secure the construct at the distal and proximal docking sites. The wires are driven through to the ulnar side of the forearm where they are brought out through the skin and capped. One of the authors (SLM) prefers to bend one wire and bury it beneath the skin with the bent wire embedded into the bone, as it has been noted that small caliber K-wires left externally often become loose within 4 weeks. These bent K-wires are left in place and have not been found to result in any problems but may be removed at a later date if necessary (Fig. 5).

Once the bones are secured the toe is revascularized.

Fig. 4. Final appearance of the foot 2 years after toe harvest.

Fig. 5. Bony fixation is initially created with K-wires. Bent K-wires may be buried beneath the skin and used to further stabilize the bony construct. (A) An anterior posterior (AP) radiograph showing final construct. (B) An AP radiograph of the reconstruction following distraction device removal at 8 weeks with consolidation of metatarsal to ulna shaft. (C) Metatarsal phalangeal (MTP) joint longitudinal growth is followed to ensure the MTP joint maintains equivalent growth with distal ulnar physis.
The dominant vessel to the MTP joint is anastomosed end to end into the radial artery or end to end into a persistent median artery. If necessary, an additional anastomosis of the MTP joint plantar arterial system can be performed end to side into the ulnar artery or end to end into the median artery. If only an ulnar artery is identified within the forearm then an end to side anastomosis is necessary. Once the toe is revascularized the skin paddle is inset and the skin is closed. The distraction device is locked into a fixed position.

**Postoperative Management**

Patients are monitored overnight in a microsurgical unit using the skin paddle to monitor the status of the underlying flap, an implantable Doppler probe is also helpful for monitoring the patency of the small vessels (Fig. 6). A bulky dressing is applied over the distraction device for the first 2 weeks. The foot is casted for four weeks. External K-wires are removed at 4 to 6 weeks. The distraction device is removed after bony consolidation which usually occurs at 8 to 9 weeks. The arm is then casted for an additional month. Prophylactic antibiotics and daily pin care are recommended to prevent pin tract infections. Post-distraction casting is important as the new joint is getting extra load at the time of distractor removal and this may be the reason for secondary fractures.

**Complications**

The most common complication is pin tract infection. If such an infection occurs during the consolidation phase antibiotics should be started and distracter device removal may be required. Vascular complications are rare and in the senior authors experience of 24 cases there were 3 arterial failures which resulted in problems with long-term growth in the transferred bones. If growth of the MTP joint is not adequate to keep up with the ulna, secondary bone lengthening procedures can be done at the time of adolescence to correct small recurrence in radial deviation.

**RESULTS OF SECOND MTP-JOINT TRANSFER**

In 1998 Vilkki reported the results of the first nine patients treated with a vascularized MTP joint transfer for Bayne type IV RLD. The age of the patients at the time of surgery ranged from 1 to 14 years (mean, 3.9 years) and the average follow-up time was 6 years. The correction of radial deviation was successful in all cases. One patient developed a traumatic fracture of the transplanted joint. Consequently the treatment protocol was adjusted to using the distraction device for at least nine weeks following MTP transfer.

In 2008 Vilkki presented a long-term follow-up study of 19 wrists in 18 patients, with a mean follow-up of 11 years (range, 4 to 19.5 years). All patients had been treated for either Bayne type III and IV RLD. Average age of the patients at the time of follow-up was 14.4 years.

**Fig. 6.** Flap is monitored by examination of the skin paddle, temperature evaluation of the skin paddle and (if possible) the placement of an implantable Doppler probe around one of the veins to the toe joint. Fig. 6 is an image of the arm at the time of the first dressing change at 5 days. The blue wire at right aspect of image represents the implantable Doppler probe wire.

**Fig. 7.** Thirteen-year follow-up of the left wrist reconstruction in a 16-year-old woman following vascularized metatarsal phalangeal joint transfer. Note minimal radial deviation of the wrist and ability to generate significant force through a centralized and stable wrist joint. Wrist range of motion is 10 degrees of wrist extension and 90 degrees of wrist flexion. Grip strength is 10 kg compared to 30 kg in the contralateral hand (Images in Fig 7. from courtesy of S.K. Vilkki, MD).
(range, 7.5 to 30 years). Average final hand-forearm angle was 28 degrees of radial deviation. Total active wrist motion averaged 83 degrees (range, 30 to 115 degrees). Overall ulnar growth averaged 15.4 cm and overall relative ulnar length was 67% of the contralateral side (range, 51% to 78%) (Fig. 7). Of the original 9 patients, reported in 1998 series, 7 were noted to have an increase in radial deviation of their wrist during the follow-up period; however this change averaged only 12 degrees over a follow-up of 15.2 years.6

Late and early complications were seen 13 of 24 patients and included distracter device complications in 2 patients, fracture of the MTP joint in 2 patients, delayed bony union of the metatarsal ulna interface in 2 patients and pseudoarthrosis at the MTP joint in 2 patients.6 Arterial failure was seen in 3 patients. Two children required late osteotomies for persistently bowed ulnas. Three children required late joint transport to lengthen MTP constructs that failed to grow adequately. Donor site morbidity was noted in only 4 patients with hallux valgus noted in one patient, prominent scarring in 2 and occasional pain noted in one other.6

**DISCUSSION**

The conventional treatment for RLD with centralization or radicalization has been shown to result in unreliable wrist stability, detrimental effects on longitudinal ulnar growth, and limitations in wrist motion.10,15,25,40 Heikell6 suggested that the only reliable means of maintaining wrist position was to fuse the wrist of these patients. The repetitive failure and dissatisfaction with centralization has led us to seek an alternative means of treatment for this problem.

MTP joint transfer allows for the possibility of preservation of ulnar length, wrist mobility and prevention of significant recurrence of radial deviation. Long-term follow-up from Vilkki’s 2008 study found that ulnar length was 67% of the contralateral side.6 These findings compare favorably with previous reports of Sestero et al.7 who found that untreated limbs of patients with RLD grew to 64% of normal ulnar length, while surgically centralized limbs within their study grew to only 48-58% of normal ulnar length.

In their long-term outcome study, Goldfarb et al.25 noted total ulnar growth to average 12 cm in comparison to 15.4 cm in Vilkki’s series.8,24,25 While 3 cm may seem clinically insignificant, this must be taken into the context of a longer ulna in addition to a wrist that is in a more balanced position when compared to standard centralization procedures. McCarthy et al.24 noted that initial wrist angles following centralization reach only 38 degrees, while average wrist position after long-term values in Vilkki’s study averaged only 28 degrees. In addition, McCarthy et al. noted long-term relapse with centralization resulting in a final wrist position averaging 71 degrees of radial deviation; such recurrences have been noted in multiple studies.24,25

Hand function, despite stable wrist position, still seems to be influenced primarily by the pre-existing stiffness noted within the fingers prior to surgery. Children with severely affected digits may still have limited hand function following successful wrist stabilization procedures. Many children with RLD lack normal intrinsic muscles which limit potential for fine motor movements; parents need to be made aware of this prior to considering surgery. In such cases they may elect for alternative treatment options. However it is our belief that the stiff hand in RLD will be improved with balancing the wrist and preserving wrist motion. In Vilkki’s 2008 study he noted 4 patients with bilateral deformities who underwent treatment of only one side.6 In these 4 patients reach was improved by 6-20 cm over the untreated hand (mean, 11 cm) which again points to the possible advantages of this procedure over centralization.

Major downsides to the procedure include the sacrifice of the second toe, however donor site morbidity was low and cosmesis is very acceptable. In addition, the procedure is lengthy, necessitating a distraction device be in place for up to 6 months; for successful outcomes the parents need to be fully educated with regards to the length of the distraction time and the need for long-term distraction device wear. While complications with this initial group were not uncommon final wrist position, even in cases of arterial failure, were still acceptable. It is also the hope that with the development of new distraction devices and ongoing familiarity with the procedure, complication rates and the time necessary for distraction device wear will decrease over time.

**CONCLUSIONS**

MTP joint transfer is an alternative to centralization for the treatment of patients with severe RLD. The procedure may prevent recurrent wrist deviation, as typically seen in centralization procedures, and help preserve all remaining growth within the distal ulnar physis. The procedure is technically demanding and requires the use of a free vascularized toe transfer; however long-term results have been encouraging with restoration of wrist function, maintenance of wrist position and adequate long-term
growth. Further comparative studies to centralization are warranted with regard to longitudinal limb length, requirement for secondary procedures, donor site morbidity and overall hand function.

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