

Staged reconstruction of radial longitudinal deficiency and thumb aplasia – case report and a review of the literature

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Purpose: Presentation of surgical treatment of a girl with type IV radial longitudinal deficiency and type IV thumb hypoplasia as well as review of the most relevant literature published in the previous 25 years.

Methods: The treatment consisted of placement of an external fixator and subsequent distraction in the radioulnar and proximodistal axis as the initial procedure followed by radialization according to Buck-Gramcko around the 2nd birthday, 10 weeks after the primary procedure. The last step of the treatment was a pollicization of the index finger performed before the 4th birthday.

Results: The surgical treatment resulted with a very favorable outcome – a clinically stable wrist, radiologically well aligned carpus over the ulna and an index pollicised in such a manner that enables adequate opposition to the other fingers.

Conclusion: A review of literature in the last 25 years shows that radialization of the ulna with a preceding distraction offers benefits over centralization alone. Distraction of the wrist protects the distal ulnar growth plate and thus promotes growth. Soft tissue release with a bilobar flap can also provide a good outcome as well as a microvascular joint transfer from the 2nd toe whereas the latter requires advanced microsurgical skills.

Key words: SURGICAL PROCEDURES, OPERATIVE; THUMB; ABNORMALITIES

INTRODUCTION

Radial longitudinal deficiency (RLD) is a malformation of the upper limb in the radio-ulnar axis (1). Even though the deficiencies of the bones of the forearm, wrist and thumb are most prominent, muscles, nerves, joints and vessels tend to be abnormal as well and play a significant part in this malformation (2). It often accompanies an underlying syndrome or association, such as Fanconi's anaemia, Holt-Oram syndrome and the VACTERL association. The incidence is estimated between 1:30 000 - 1:55 000 live births and in around 50% of patients, it occurs bilaterally. Various etiological factors have been proposed, such as genetic mutations (due to correlation with above-mentioned syndromes) or potential intrauterine insults such as vascular insufficiency, environmental effects, or maternal drug exposure (3).

Modified Bayne and Klug classification recognizes 4 types of RLD based on the radiographic severity of skeletal deficiency, (Figure 1) ranging from mild thumb or distal radial hypoplasia to a complete absence of the radius. It considers factors such as: thumb absence or hypoplasia; carpus hypoplasia, absence or coalition; distal part of the radius hypoplasia; proximal radius hypo/aplasia.

We are presenting a case of a 20 month old girl with a unilateral (right forearm) Bayne and Klug type IV radial longitu-

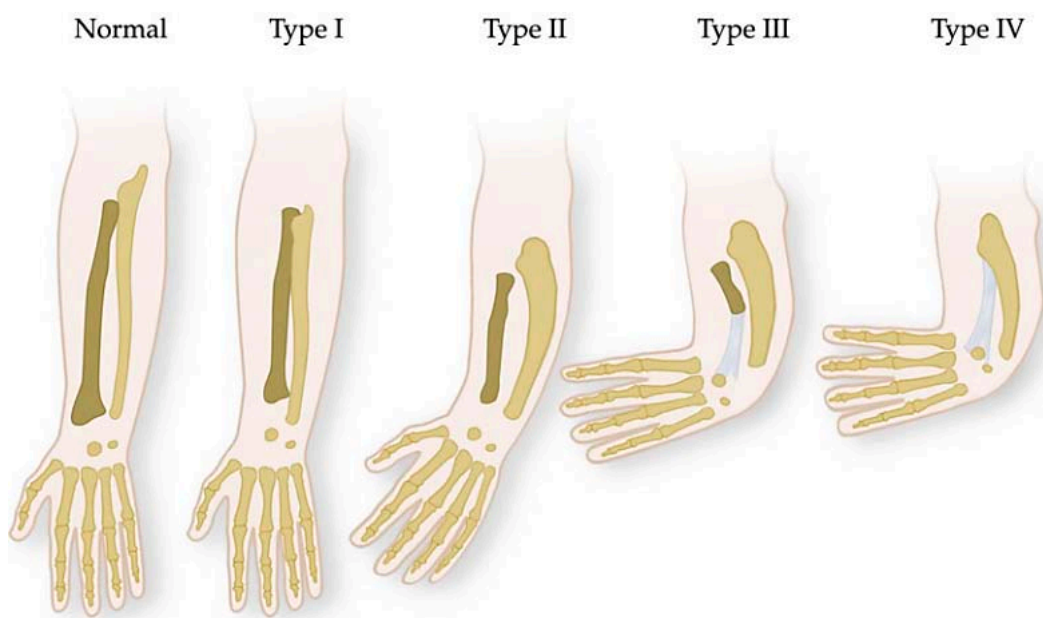
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From: Van Alphen NA, Moran SL. Radial Deficiency. In: Abzug JM, Kozin SH, Zlotolow DA ed. The Pediatric Upper Extremity. New York: Springer;2015:259.

FIGURE 1. The original classification of radial longitudinal deficiency according to Bayne (Copyright Mayo Foundation).



FIGURE 2. AP view of the forearm preoperatively.

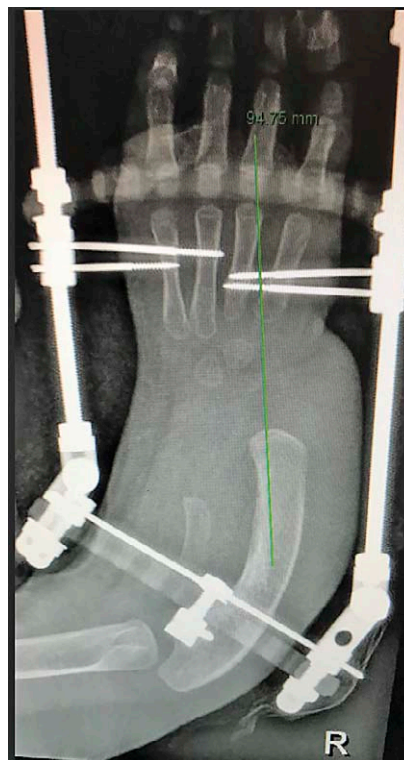


FIGURE 3. A profile view of the forearm preoperatively.

dinal deficiency (Figure 2.) as well as a Blauth type IV hypoplastic thumb as an isolated anomaly. In addition to the presentation of our case, we have also performed a review of literature pertaining to RLD in the Pubmed database for the period of last 25 years.



FIGURE 4. An AP radiograph of the forearm after 4 weeks of distraction.

CASE REPORT

The patient initially presented to our Clinic at 20 months of age. After consultation with the parents, we opted for a three-step reconstruction. The first procedure, which was undertaken when the child was 23 months old, was the application of an external fixator (Litos GMBH Ahrensburg, Germany). The fixator was placed distally in the 2nd and the 4th metacarpal bone and proximally in the proximal ulna. In the following four weeks, distraction was performed in a radioulnar as well as in a proximodistal direction which enabled a satisfying position of the carpus on the ulna prior to radialization (Figure 3.). After additional 6 weeks during which external fixator remained on the forearm but no distraction took place, we have removed the external fixator and performed a radialization procedure according to Buck-Gramcko. This procedure included severing of the ECRB and ECRL tendons at their insertions, opening and mobilizing the wrist joint in an ulnar direction after which the position of the wrist was ultimately fixed with a K wire which extended from the ulna into the 2nd metacarpal bone. Due to the breakage of a pin which was placed in the proximal ulna, we have had to leave a part of the pin in the bone. Postoperatively, an upper arm splint was placed with the forearm in the middle position for the period of three weeks followed by a placement of an orthosis which was initially worn permanently. The K wire which held the position of the wrist joint was removed after 3 months in the outpatient clinic. The third step of the reconstruction, the pollicization procedure was performed when the child was a



FIGURE 5. An AP radiograph of the affected forearm after the radialization procedure (Notice that a part of the pin from the external fixator got broken during the removal. We made no attempt to remove it from the bone).



FIGURE 6. The outcome after the pollicization of the index finger.

month shy of 4 years of age. Pollicization was done in a standard fashion with skin flaps raised according to Carter-Ezaki, ligation of the radial neurovascular bundle to the index finger, shortening of the 2nd metacarpal bone at the level of the epiphysis proximally and at the base distally and fixation of the new thumb in a hyperextended and abducted position (Figure 4). During the follow-up period,

the patient experienced a postponed healing of a smaller skin area which resolved without functionally impairing scarring.

At the latest follow up, 6 months after the final step of the treatment, the patient presented with a clinically stable joint and good radiological alignment of the wrist over the distal ulna (Figure 5). The pollicized index was in a position which enabled a good opposition and she displayed satisfying pinch strength (Figure 6).

DISCUSSION

Radial longitudinal deficit comprises a spectrum of abnormalities – from type 0 in which the deviation of the distal radius is minimal (4) to type IV where the radial part of the hand rests on the radial forearm (5). The most established therapeutic modalities for radial longitudinal deficiency types III and IV according to Bayne are as follows: centralization, radialization, soft tissue release with a bilobed skin flap and a microvascular 2nd MTP joint transfer. The classical therapeutic approach is the centralization procedure described over a century ago (6). This technique could either be done with the creation of a notch in the carpus for the ulnar head or as a release of all structures that produce tethering effect such as the radial wrist capsule, radial wrist extensors together with pinning of the carpus in a neutral position with the ulna being aligned with the middle finger (7). In various reports, this technique resulted in the formation of a shortened forearm, which was more pronounced in the notched centralizations (8) as well as a high recurrence rate of angular deformity (9-12). Buck-Gramcko has modified this method by aligning the ulna with the index finger metacarpal in a procedure known as radialization (13). Both centralization and radialization may or may not be preceded by a soft tissue distraction procedure. Several authors have confirmed that preoperative distraction lengthening enables a tension-free alignment of the carpus over the ulna which prevents premature closure of the ulnar epiphyseal growth plate and consequently a longer forearm (14, 15). However *Manske et al.* have noted that preoperative distraction results in higher recurrence of the angular deformity in the long term follow-up (11). In the article by *Dana et al.*, radialization with soft tissue distraction resulted in a mean correction of angulation deformity of 33°(16). *Bhat et al.* have shown in a long term follow-up study that radialization (without previous soft tissue distraction) results in an aesthetic and functional improvement even though loss of correction can be expected in the long-term (17). *Mittal et al.* have shown in a randomized controlled trial that radialization without previous distraction resulted in a better correction, lower recurrence rate and a longer forearm length than a notched centralization (18).

Reconstruction with radial soft tissue untethering and a bilobed skin flap is an option in the treatment of severe radial longitudinal deficiency popularised by the group from Texas Scottish Rite Hospital in Dallas (19). The purpose of the skin flap is to substitute the lack of skin at the point of maximum skin deficiency. This procedure comprises a release of tight fascial bands and transfers or releases of wrist flexors to prevent progressive angulation. In cases in which the ulnar angulation is greater than 30°, a mid/dyaphyseal osteotomy is warranted. Despite not achieving a straight wrist with this technique, patients express a high rate of satisfaction for both appearance and function.

A microvascular joint transfer for the reconstruction of the radial longitudinal deficiency is another procedure which enables a stable wrist with a preserved forearm length and a low recurrence rate (20). This technique is, however, technically demanding, requires advanced microsurgical skills and entails procurement of the 2nd ray of the foot as the donor site. *Morsy et al.* recommend this technique as a salvage procedure after failed centralization(21).

Luangjarmekorn et al. have published an impressive case series in which they showed a single stage reconstruction of radial longitudinal deficiency with thumb hypoplasia (22). Even though this approach enables complete treatment during a single general anaesthesia it however entails a higher risk of arterial insufficiency of the pollicized thumb and postoperative venous congestion.

Radial lengthening was attempted as a therapeutical modality for Bayne and Klug type II and III radial longitudinal deficiency (23). Even though some clinical benefits have been noted, this method requires 3 or 4 lengthening procedures at 3 year intervals until the cessation of ulnar growth.

In our patient, we have opted for the radialization technique with a distraction that preceded the procedure and a pollicization of the index finger which completed it. Main reason for our approach is that co-author (D.M.W) has brought the experience needed for this procedure. To our knowledge, this is the first time that a reconstruction of radial longitudinal deficiency was done in this manner in Croatia. Ultimately, the achieved outcome was highly satisfactory for both the parents and us as the treating surgeons.

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SAŽETAK

Etapna rekonstrukcija radijalnog longitudinalnog deficita sa aplazijom palca - prikaz slučaja i pregled literature

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Svrha: Prikaz kirurškog liječenja djevojčice sa radijalnom longitudinalnom deficijencijom tipa IV te hipoplazijom palca tipa IV kao i pregled najrelevantnije literature objavljene na Pubmed-u na ovu temu u proteklih 25 godina.

Osnovni postupci: Liječenje se sastojalo od postavljanja vanjskog fiksatora te posljedične distrakcije u proksimodistalnoj i radioulnarnoj ravnini nakon čega je slijedila radijalizacija prema Buck-Gramcku oko druge godine života djeteta, 10 tjedana nakon prvog zahvata. Posljednji korak je bila policizacija kažiprsta u dobi od 4 godine.

Rezultati: Kirurško liječenje je rezultiralo vrlo dobrim ishodom – klinički stabilnim ručnim zglobovima, radiološki dobrim odnosom karpusa i ulne te policiziranim kažiprstom koji omogućava adekvatnu opoziciju prema preostalim prstima.

Zaključak: Pregled literature u proteklih 25 godina pokazuje kako radijalizacija ulne uz prethodnu distrakciju ima prednost u odnosu na centralizaciju. Distrakcija ručnog zgloba čuva distalnu ploču rasta ulne i tako omogućava rast u duljinu. Oslobođanje mekih tkiva s bilobarnim režnjem može pružiti dobar ishod kao i mikrovaskularni prijenos zgloba s 2 prsta stopala pri čemu potonja tehnika zahtijeva napredne mikrokirurške vještine.

Ključne riječi: OPERATIVNI POSTUPCI; PALAC; ABNORMALNOSTI