Radial Longitudinal Dysplasia: An Alternative Treatment Paradigm

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Abstract

Radial longitudinal dysplasia continues to challenge the pediatric hand and upper limb surgeon. Despite decades of clinical experience and research, surgical strategies to improve wrist alignment and upper limb function remain imperfect, with persistent concerns regarding recurrent deformity, stiffness, and distal ulnar growth disturbance. For these reasons, many authors have proposed alternative treatment paradigms to traditional centralization techniques. The purpose of this review is to explore the arguments for surgical treatment, to identify the potential sequelae of traditional centralization, and to present an "alternative treatment paradigm" for radial longitudinal dysplasia.

Key Words: Radial longitidinal dysplasia, centralization, bilobe flap

Introduction

Radial longitudinal dysplasia (RLD) refers to failure of formation of the radial aspects of the hand and forearm, and is currently classified as an abnormality of formation and differentiation of the antero-posterior axis of the developing upper limb by the Oberg-Manske-Tonkin classification system [1].The incidence is thought to be between 1 in 30,000 and 100,000 live births , and RLD has a slight male and right-sided predisposition. Over half of patients have bilateral upper limb involvement.

There is a broad spectrum of anatomic anomalies involving the skeletal, muscular, and neurovascular structures, and associated syndromes are common, including VACTERL association, Holt-Oram syndrome, Cornelia de Lange syndrome, thrombocytopenia-absent radius, Diamond-Blackfan anemia, Rothmund-Thomson syndrome, and Fanconi's anemia [2,3,4,5]. A host of condition-specific schemes have been proposed, based mostly upon the severity of bony involvement, and to date the modified Bayne and Klug classification continues to be used by most providers [6,7].

Overall, the goals of treatment include maximizing upper limb and hand function, ideally by preserving wrist motion, improving wrist alignment, maintaining limb length and growth potential, and preserving the ability for subsequent hand reconstruction and pollicization. Furthermore, the aesthetic differences –and secondary effects thereof-- imparted by RLD must be recognized and considered in an appropriate fashion.

A host of questions continue to be raised with the growing experience of RLD treatment, particularly with longer term follow-up and patient-derived functional outcome measures. What is the natural history of RLD? Is surgical treatment for deformity correction worthwhile? And if so, what is the best method of treatment to maximize function and minimize complications?

The purpose of this review will be to address these questions using the available information from India and the United States, and in doing so, present an alternative treatment algorithm for patients with RLD.

Natural History versus Surgical Reconstruction

With any congenital difference, the first fundamental task of the pediatric orthopaedic surgeon is to understand the natural history of the condition and the comparative outcomes of natural history versus surgical treatment. Kotwal et al. published an important paper seeking to address this fundamental question as it relates to RLD [8]. One hundred and three patients with RLD treated non-operatively were compared to 239 patients treated with surgical reconstruction. All patients had Bayne type 3 or 4 deficiency and were treated at a single center from 1985 to 2004. Patients were assessed according to standard clinical parameters, radiographic measurement of the hand-forearm angle (HFA), and visual analogue scores (VAS) for pain and aesthetic appearance. Grip strength was evaluated using dynamometry, though pinch strength was not measured due to the heterogeneity of hand/thumb involvement. The Prosthetic Upper Extremity Functional Index (PUFI), originally developed to evaluate function in patients with transverse upper limb deficiencies, was used to assess upper limb function.

Non-operative treatment consisted of early stretching, serial casting, splinting, and.or functional bracing. Centralization consisted of centralization (n=202) or radialization (n=107) via an ulnar incision, excising redundant ulnar skin, notching the carpus, reefing the wrist joint capsule, preserving the distal ulnar physis, and stabilizing with a 1.6mm Kirschner (K-) wire [9,10]. Tendon transfers were performed in selected cases. Radialization was performed in a similar fashion, though tendon transfers were more commonly performed and K-wire fixation

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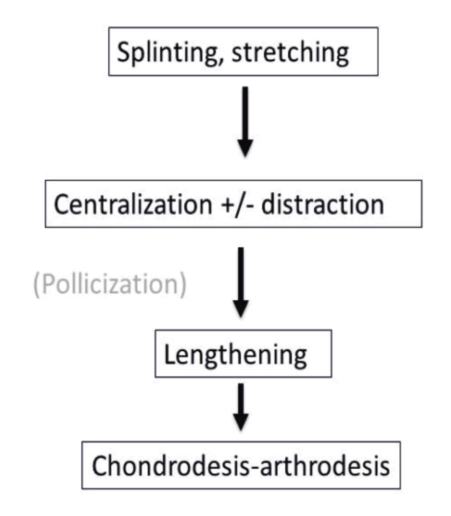

Figure 1: "Traditional" treatment algorithm for radial longitudinal dysplasia, utilizing centralization or radialization.

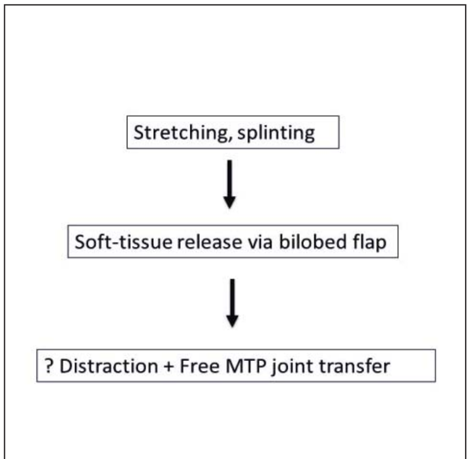

Figure 2: An alternative treatment paradigm for radial longitudinal dysplasia, emphasizing soft-tissue releases via bilobe flap reconstruction, which acknowledges risks of recurrent deformity after centralization and prioritizes preservation of wrist motion and distal ulnar physeal integrity

was achieved through the index metacarpal [11].

Overall, patients with type 3 and 4 RLD treated with surgical reconstruction demonstrated better PUFI scores, had improved HFA, and reported improved aesthetics than non-operatively treated patients. However, surgical patients also had decreased wrist range of motion (ROM), and interestingly functional results as assessed by the PUFI correlated best with digital ROM, regardless of treatment type. Ekblom et al. similarly reported that digital and wrist ROM is more important for activity and participation than HFA in their analysis of 20 patients with RLD [12]. Holtslag et al. similarly found that only ROM of the didgits correlated with function, as assessed by the Sequential Occupational Dexterity Assessment [13]. No significant differences in activity and participation between patients with mild versus severe RLD. Thus, while there is published information suggesting that surgical treatment provides functional, alignment, and aesthetic improvements that are superior to natural history, it is important to remember the importance of wrist and digital ROM in upper extremity and hand function.

Concerns with centralization

Surgical treatment in the form of centralization or radialization is not without concerns. First the longevity of correction and functional improvement continues to be debated. In their longer-term follow-up study of 25 wrists treated with centralization, Goldfarb et al. reported recurrent deformity (HFA of 25 degrees), limited wrist ROM (mean arc 31 degrees) and forearm shortening (ulnar length 54%), perhaps due to iatrogenic distal ulnar physeal disturbance [14].

In efforts to minimize the soft-tissue tension and thus the risks of recurrent deformity, stiffness, and growth arrest, many authors have advocated pre-centralization distraction using a host of external fixation constructs. In theory, pre-centralization distraction reduces soft tissue tension, facilitates centralization, obviates the need for carpal notching or prolonged implant placement which may contribute to distal physeal disturbance. Thatte and Mehta reported on a series of 29 wrists treated with distraction and subsequent radialization at a young age [15]. HFA improved from 74 to 10 degrees, ulnar length was maintained at over 70%, and correction was maintained at mean 6.5 year follow-up. These results are excellent by all accounts, but have not been consistently reproduced in all centers.

Manske et al. compared 13 wrists treated with centralization alone to 13 wrists treated with centralization preceeded by distraction [16]. With centralization alone, HFA improved from 53 degrees pre-operatively to 13 degrees immediately post-operatively to 27 degrees at most recent follow-up. With pre-operative distraction, HFA improved from 53 degrees pre-operatively to 21 degrees immediately post-operatively to 36 degrees at last follow-up. Based upon the similar amount of recurrence, the authors concluded that distraction may facilitate surgical centralization but does not necessarily minimize recurrence. Similar findings were noted by Dana et al. in a series of 8 patients with over two-year follow-up, as well as Lamb et al, McCarthy et al., Shariatzadeh et al, and others [17,18,19,20].

The second concern with centralization techniques has been the risk of iatrogenic ulnar growth disturbance, resulting in upper limb length discrepancy and shortening of an already dysplastic forearm. Potential factors contributing to the risk of ulnar physeal arrest include devascularization from soft tissue dissection around the distal ulna, carpal notching and increased pressure imparted on the distal ulna during centralization, as well as the placement and retention of an intramedullary implant crossing the ulnar physis. Sestero et al. evaluated ulnar growth patterns in 124 limbs of 90 patients and found decreasing ulnar growth in patients treated with non-notching and notching centralization techniques [21]. Indeed, non-operatively treated limbs retained 64% of "normal" length, whereas those patients who underwent non-notching and notching techniques of centralization had ulnar lengths of 58% and 48%, respectively.

To counteract and address both recurrent radial deviation deformity as well as ulnar shortening, several authors have proposed late reconstruction consistent of ulnar lengthening with or without subsequent ulnocarpal arthrodesis. Farr et al. reported on 8 forearm lengthenings performed in 6 patients at mean age of 10 years [22]. At almost 5 year followup, a mean of 7cm of lengthening was achieved. HFA improved from 25 degrees pre-operatively to 11 degrees post-operatively, though recurrent deformity was again observed, with a final follow-up HFA of 23 degrees.

Arthrodesis –or chondrodesis in the skeletally immature—allows for correction of wrist deformity at the expense of ulnocarpal motion. Pike et al. reported on 12 patients with recurrent radial deviation of greater than 45 degrees who underwent ulnocarpal chondrodesis with Kirschner (K-) wire fixation [23]. Patients had a mean post-oepative DASH score of 24.5, with visual analog scores (VAS) of 7 and 8 for appearance and function, respectively.

Based on these and many other reports, the "classic" or traditional treatment algorithm in the United States and around the world has been (Fig. 1) early splinting, stretching

Figure 3: Clinical photographs depicting elements of soft-tissue release via a volar bilobed flap. (a) Pre-operative resting position, depicting radial deviation of the wrist and a hypoplastic thumb. (b) After pre-operative stretching and splinting, the wrist may be passively stretched to neutral position. (c, d, e) Intra-operative photographs depicting the (c) radial, (d) volar, and (e) ulnar aspects of the incision. A volar approach was chosen given the aesthetic advantages of leaving the dorsal wrist free of scars. (f) Intra-operative photograph after flap elevation, allowing for access to the musculotendinous units for release and/or transfer. Note the soft-tissues about the distal ulnar physis are carefully preserved. (g) Intra-operative depiction of flap rotation, providing tissue to the previously taught radial wrist. (h, i) Clinical photographs after wound closure, demonstrating improved alignment of the wrist. (j) At 3 months post-operatively, incisions are well healed, flaps remain viable, and the wrist is supple with improved resting alignment. (All images courtesy of Children's Orthopaedic Surgery Foundation, copyright 2016)

and/or casting, followed by centralization with or without pre-operatively distraction. For those with recurrent deformity and/or excessive limb length discrepancy, ulnar lengthening and ulnocarpal chondrodesis or arthrodesis may be considered.

Is there another way?

While commonly utilized, the drawbacks of recurrent deformity, additional limb length discrepancy, and loss of wrist motion despite multiple surgeries have motivated many providers to consider alternative treatment options. Ideally, surgical treatment should preserve wrist motion and forearm length, and in doing so maximize hand and upper limb function. To this end, several authors from the United States and around the world have proposed an alternative treatment algorithm consisting of two stages (Fig. 2). The first surgical step in this algorithm is addressing radial deviation of the wrist through soft tissue releases, capsulorraphy, and tendon transfers via bilobe flap reconstruction [24,25]. Philosophically, this represents a departure from traditional tenets of RLD surgery; the focus of releases is to improve wrist alignment while judiciously preserving the distal ulnar physis. Wrist motion is prioritized over alignment, given the evidence that function is most directly correlated to wrist and digital motion and that typical surgical interventions result in recurrence. Technically, a bilobed flap may be performed either dorsally or volarly, depending upon surgeon preference and awareness of the aesthetic implications of dorsal scars and/or color mismatch between the dorsal and volar skin (Fig. 3). A proximally based flap is designed longitudinally which will be rotated or transposed to provide skin and soft tissue to the tight, often deficient radial wrist. A second lobe is based proximally and incorporating the excessive ulnar soft tissues, which will be rotated to cover the dorsal donor area. Careful flap elevation is performed to maintain vascularity to these random patterned flaps, with care to preserve subcutaneous nerve and vein, when possible. After the flaps have been elevated and mobilized, near circumferential exposure of the deeper structures is possible. Tight radial-sided structures are release, the ulnar capsule may be incised and imbricated, and tendon transfers (particular flexors to ulnar extensors) may be performed to achieve dynamic rebalancing across the wrist. Great care is made to preserve all the soft tissues around the distal ulna in efforts to preserve its vascularity and integrity of the distal ulnar physis. Temporary K-wire stabilization may be performed from the ulnar metaphysis to the carpus, avoiding wire passage across the distal ulnar physis. Splinting, therapy, and ROM exercises are begun at 4-6 weeks post-operatively.

Vuillermin et al. published their series of 18 wrists in 16 patients treated with soft tissue release and bilobe flap reconstruction [24]. A t mean 9 year follow-up, HFA improved modestly from 88 degrees pre-operatively to 64 degrees at most recent follow-up. However, patients maintained a mean of 73 degree wrist flexion-extension and had Disabilities of the Arm, Shoulder, and Hand (DASH) scores and global Pediatric Outcomes Data Collection Instrument (PODCI) scores of 27 and 88, respectively. VAS scores for overall satisfaction were quite high (mean 1.2 on a 10 point scale). These patient-derived measures of outcome compare favorably to other published information regarding the results of centralization. Importantly, there were no physeal growth disturbances, and no patients went on to secondary ulnocarpal chondrodeses or other salvage procedures.

After soft-tissue releases via bilobe flap reconstruction, subsequent soft-tissue distraction and free vascularized metatarsophalangeal (MTP) joint transfer may be performed in a secondary staged fashion [26,27]. Pioneered by Dr. Vilkki, free MTP joint transfer provide an attractive, albeit technically demanding, option to provide more durable carpal support and maintain more normal HFA.

As originally described, distraction lengthening is performed across the ulnar aspect of the wrist, with half pin or smooth wire fixation into both the proximal ulna and ulnar metacarpals [27]. Once adequate distraction has been achieved and more normal wrist alignment restored, free second MTP joint transfer is performed. Arterial inflow is typically achieved from the radial artery to the plantar metatarsal artery. Local veins are used for venous outflow. More extensive radial soft tissues are released, with the native FCR split and reattached to the donor toe flexor and extensor apparatus. K-wire fixation is typically utilized to fix the proximal ulna to the donor metatarsal as well as the radial metacarpals to the donor proximal phalanx; the previously applied fixator is retained for additional support during the early postoperative period. The transferred joint forms a Y-shaped "strut" to support the radial wrist and hand, thereby maintaining HFA while still allowing for wrist motion.

Vilkki previously reported on 19 wrists in 18 patients treated with distraction and MTP joint transfer. At mean 11 year follow-up, HFA was 28 degrees and wrist flexion-extension arc averaged 83 degrees [27,28]. While there was increased radial deviation over time, this change averaged 12 degrees over 10 to 15 years follow-up. Ulnar length is typically preserved, with length approximating two-thirds of normal.

While the author has no personal experience with free vascularized joint transfer, it should be noted that this procedure is generally delayed until later in childhood, given the technical challenges of the procedure. Interestingly, multiple authors have reported that patients often decline secondary MTP joint transfers, given their satisfaction and function following soft-tissue releases via bilobed flap reconstructions [25].

While early reports highlight the theoretical advantages in preserving motion and growth, the merits of this "alternative algorithm" bear further investigation. At present, there is little published information to compare the relative efficacy, durability, and long-term patient-reported functional outcomes of traditional centralization versus soft-tissue releases with or without free MTP joint transfer. In the United States, a multicenter prospective longitudinal cohort study is underway to address this, and other, important congenital questions [29].

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