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Original Article

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Our Experiences in the Surgical Treatment of Bayne Type III and IV Radial Longitudinal Deficiencies

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ABSTRACT

Radial longitudinal deficiencies are characterized by radial deviation of the wrist, a short and bowed forearm and a non-functional or absent thumb. The deformity is caused by varying degrees of underdevelopment and malformation including hypoplasia of the bones, joints, muscles, tendons, ligaments, nerves and vessels. This study describes our results from centralization in 8 limbs with Bayne type III-IV radial longitudinal deficiencies. We reviewed 8 limbs from 6 patients with a centralization performed between 2002 and 2010. The median patient age at the time of the operation was 2.75 years (range 2-14 years). The sex ratio was 4 male and 2 female. All patients underwent centralization. One patient had an additional 6.5 cm elongation of the ulna bone by means of a circular external fixator (CEF). The median follow-up time from centralization was 4,5 years (range 2-10 years). The result was considered to be excellent in 4 cases, good in 3 cases and fair in 1 case. Radial deviations regenerated in all cases. During follow-up, a radial deviation of 15 degrees developed in one patient; this deviation did not require surgical correction. Hand and wrist movements were within acceptable levels. Movement and function of the wrist and forearm can be obtained through centralization of the wrist and ulnar elongation when necessary. A cosmetically pleasant appearance can be ensured.

Key words: *Radial longitudinal deficiency, centralization, distraction.*

Bayne Tip III ve IV Radial Uzunluk Eksikliklerinin Cerrahi Tedavisindeki Deneyimlerimiz

ÖZET

Radial uzunluk eksiklikleri elbileğinin radial eğriliği, kısa ve eğri önkol ve eksik veya fonksiyon görmeyen başparmak ile karakterizedir. Deformite kemikler, eklemler, kaslar, tendonlar, ligamentler sinirler ve damarların hipoplazisini içeren malformasyon ve az gelişmenin çeşitli derecelerine neden olur. Bu çalışma, Bayne tip 3 ve 4 radial uzunluk eksikliği olan 8 ekstremitedeki santralizasyon sonuçlarımızı tanımlamıştır. 2002 ve 2010 arasında santralizasyon yapılan 6 hastanın 8 ekstremitesi gözden geçirildi. Operasyon zamanında ortalama hasta yaşı 2.75 yıldı (2-14 yıl arası). Cinsiyet oranı 4 erkek, 2 kadın idi. Bütün hastalara centralizasyon yapıldı. Bir hastaya ek olarak ulna kemiğine sirkuler eksternal fiksatör (CEF) vasıtasıyla 6,5 cm uzama sağlandı. Santralizasyon için ortalama takip süresi 4,5 yıldı (2-10 yıl arası). Sonuç 4 olguda mükemmel, 3 olguda iyi, 1 olguda orta kabul edildi. Radial eğrilik tüm olgularda düzeldi. Takip sırasında, bir hastada 15 derece radial eğrilik gelişti; bu eğriliğe cerrahi düzeltme gerekmedi. El ve elbileği hareketleri kabul edilebilir düzeyler içindeydi. Elbileği ve önkol hareket ve fonksiyonu elbileğinin santralizasyonu ve gerektiğinde ulnar uzatma yoluyla elde edilebilir. Kozmetik olarak hoş görünüm sağlanabilir.

Anahtar kelimeler: *Radial uzunluk eksikliği, santralizasyon, uzatma*

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INTRODUCTION

Radial hypoplasia is a rare congenital deformity of the upper extremities that presents with clinical radial deviation of the wrist characterized by radial shortness. This deformity presents with a wide spectrum of hand and radial anomalies varying from steady radial hypoplasia to radial aplasia. This condition is also defined as radial longitudinal deficiency (RLD), and the hypoplasia can involve bones, joints, muscles, tendons, ligaments, nerves and veins.

Surgical and non-surgical methods are available for the treatment of radial longitudinal deficiency, and treatment is started as soon as a diagnosis is made. The basic treatment aims are to correct wrist angulation, protect the movement and function of the wrist and hand, improve the regular growth of the radial bone and obtain a cosmetically pleasant appearance (1,2). The aims of this study are to discuss the results of centralization in our cases of radial longitudinal deficiency and to review the literature.

MATERIALS AND METHODS

We reviewed 8 cases of hands in 6 patients with congenital radial longitudinal deficiency deformities between 2002 and 2010 (Table 1). The patients were classified according to Bayne (3), and only patients with Bayne type III-IV deformities with indications for centralization were included in this study. The median patient age at the time of surgery was 2.75 years (range 2-14 years). Two patients were male, and 4 were female. All of the patients underwent centralization. Five patients had type IV deformities, and 3 had type III radial longitudinal deficiency deformities. A nonfunctioning thumb was associated with a radial longitudinal deficiency deformity in all hands. After centralization, we performed pollicization on the index finger. Two cases had Vater syndrome, and one case had brachyphalangia. For one patient with Vater syndrome (Figure 1-2), a circular external fixator (CEF) was placed at the same time as the centralization (after centralization), and an ulnar osteotomy was performed to extend the ulnar bone (Figure 3-4). During the postoperative follow-up visits, a 6.5 cm lengthening of the ulna was achieved with the help of the distractor (Figure 5-6). The range of motion at the shoulder, elbow, wrist and fingers was recorded at the final follow-up. At the final follow-up visit, a questionnaire was given to parents for subjective evaluation of appearance improvements and overall satisfaction. It is a self-made questionnaire (Questionnaires were not administered because some of the children were very young).

***Figure 1**. (a-b) Preoperative view of the patient (3).*

Surgical Technique

Centralization without arthrodesis was performed in all cases to stabilize the hand on the ulna for all patients. All surgeries were performed under general anesthesia. Utilization of tourniquet and loop magnification was standard.

***Table 1**. Patients' clinical data*

Patient	Centralization age	Sex	Bilateral side	Bayne type (R/L)	Associated anomalies	Lengthening (cm)	Follow-up (yr)	Parents satisfaction score
1	2.5	Female	Right	IV	No		9	8/10
2	4	Male	Left	III	Vater Synd		4	8/10
3	14	Male	Left	IV	Vater Synd	6.5	2	6/10
4	2	Male	Bilateral	IV/III	No		4	10/10
5	2	Female	Left	III	Brachyphalangia		5	8/10
6	3	Male	Bilateral	IV/IV	No		10	9/10

Figure 2. Preoperative x-ray of the patient.

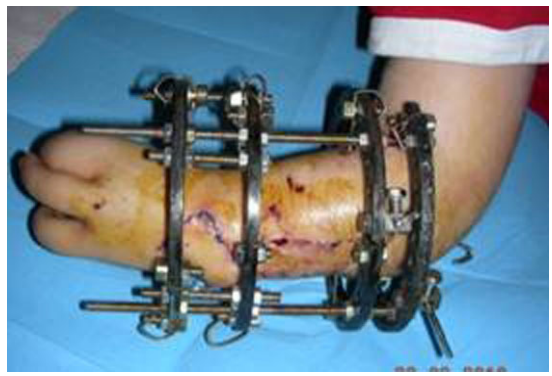

Figure 4. Clinical appearance of centralization and lengthening of forearm.

For centralization of the hand, two skin incisions were performed. The first incision was a Z-plasty on the tight radial side, and the second incision was a transverse wedge incision over the prominent distal ulna. The former incision was for skin lengthening, and the latter incision provided sufficient exposure to the ulnar side of the wrist and allowed removal of excess fibrofatty tissue and skin. A distally based capsular flap containing the ulnar collateral ligament was created when exposing the ulna, and the blood supply to its epiphysis was preserved. The ulnar nerve and its dorsal cutaneous branch and the ulnar artery were preserved. The extensor retinaculum was released. The radial extrinsic extensors and flexors, which share a common muscle mass in these patients, were released from the carpal bones. After palmar capsule release, the hand was sufficiently free to be placed in line with the second or third metacarpal on the end of the ulna. A slight shaving of the cartilage on the end of the ulna and the opposing carpal surface was performed to produce a plane joint. A strong, double-pointed K-wire was placed retrograde through the metacarpal, brought out through its head and then went down proximally through the center of the ulna epiphysis and up into the ulnar shaft. An x-ray was used to control the placement of the K-wire. Stabilization was achieved by reinforcing the supports on the ulnar side of the hand. The released radial flexors and extensors were attached to the ulnar carpus. The extensor carpi ulnaris was placed back in its sheath on the ulna and reattached more distally on the shaft of the fifth metacarpal. If necessary, the flexor carpi ulnaris was also moved. The hypothenar muscle mass was detached from its origin and reattached to the ulna if it was able to reach. The distally based capsule was firmly reattached. A padded cast was used from palm to

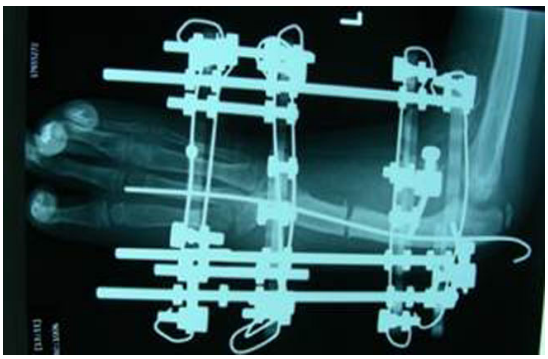

Figure 3. X-ray of centralization and lengthening of forearm. K-wire was removed at 2 mos.

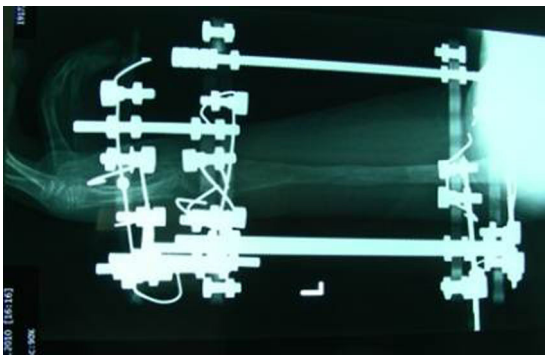

Figure 5. Postoperative follow-up 5.5 months after the operation.

***Figure 6**. After removal of the circular external fixator. Forearm was lengthened by 6.5 cm.*

axilla with the elbow flexed. The K-wire was removed 6 to 9 weeks after surgery, and the patients had the short arm plastic splint that allowed finger movement for 4- 6 weeks.

RESULTS

The median follow-up time was 4.5 years (range 2-10 years). The results were considered to be excellent in 4 cases, good in 3 cases and fair in 1 case (We were asked to describe the average the functional and aesthetic patient results from the doctors and parents on a 1-10 number scale. The results were evaluated on average of two scale. Above the 8 number was considered to be excellent, between 6-8 was considered to be good, and below 6 was considered to be fair). Radial deviation recurred in all limbs. During follow-up, a radial deviation of 15 degrees developed in one patient; this deviation did not require surgical correction. Patient noncompliance and inefficient adherence of the Kirshner wire were determined to be recurrence factors. Better results were obtained on the ulnar side when plication was performed on the capsule. Hand and wrist movements were within acceptable levels. The aim of this operation is to obtain basic hand function (to allow the patient to perform basic needs) (Figure 7). No significant change in the extent of shoulder and elbow motion was found at the final follow-up. Centralization had no effect on the range of motion of the finger joints. Parental satisfaction scores are given in Table 1. There were no cases of neurovascular injury or wound infection.

DISCUSSION

Radial longitudinal deficiency is a birth anomaly that was defined by Petit in 1733 as a radial longitudinal deficiency occurring in 30,000 to 100,000 live births. RLD frequently involves a loss of function or the existence of a thumb in children (1,4-8). The etiology of this anomaly is indeterminate, and the facts are sporadic. Rarely, RLD patients have life-threatening systemic problems. Renal USG, EKO and blood tests must be given to patients. RCH is generally seen in the following diseases: Fanconi anemia, Holt – Oram syndrome (RCH, ventricular septal defect), VATER syndrome (vertebral anomaly, anal atresia, tracheoesophageal fistula, esophageal atresia, renal agenesis), VACTERL syndrome (vertebra anomaly, anal atresia, cardiac anomaly, tracheoesophageal fistula, renal agenesis,

***Figure 7**. Long-term functional and cosmetic result of centralization and index finger pollicization.*

limb defect) and TAR syndrome (thrombocytopenia, radius deficiency) (2,4,6,8,9). Vater syndrome was found in two cases presented here.

Deformities were divided into 4 groups by Bayne and Klug according to the existence of the radius: 1) A moderate level of defect is present in the distal radial epiphyseal, minor radial deviations are seen, and thumb hypoplasia may be obvious. 2) The radius is characterized by limited growth, both distally and proximally. The radius is miniaturized, and moderate radial deviation is present in the wrist. 3) Two-thirds of the radius is nonexistent, and severe distal radial deviation is found in the wrist. 4) This type is extensive and very severe; the radius is nonexistent (1,2). James and colleagues modified this classification. They classified cases with normal distal radius longitudes as N and O according to isolated anomalies of the thumb carpal bone (type N: Normal radius longitude and normal wrist with thumb hypoplasia; type O: Normal radius longitude, radial side carpal anomalies, radial deviation). However, James et al. did not offer any classification change for the other 4 groups (1,10). In the group of patients presented here, two-thirds had a nonexistent radius in three hands, whereas the radius was completely nonexistent in 5 hands.

Treatment of radial club hand is determined based on the degree of anomaly (1,2). Radial club hand treatment involves current surgical approaches, centralization without carpal bone resection, stabilization with tendon transfer and improvement of thumb deformation using pollicization. The optimal age for surgery is 6-9 months (8,11). To protect against the development of contractures during centralization, soft tissue deregulation, carpal resection and ulnar osteotomy were used on the ulna to activate wrist centralization (1). Inefficient soft tissue deregulation before centralization causes unequal ulnar growth, distal ulnar bowing, intercarpal fission and recurrence (7,12). Some authors described soft tissue distraction in radial longitudinal deficiency before centralization. Extreme dissection was improved to prevent bone resection and tension of acute neurovascular structures, whereas Kessler improved preoperative soft tissue distraction for the thumb and radius. 1). Nanchahal used a Kessler device and a monolateral external fixator. Preoperative distraction with the Kessler device permits hand realignment without skeletal resection or excessive tension on radial neurovascular structures (7,13). With a Smith and Greene Orthofix monolateral fixator, soft tissue was successfully supplied in 2 patients (1,7,14). A Vilkki monolateral external fixator was used to improve angular deformities and to form a flap area in 9 patients in microvascular 2nd toe transfer of the foot (7,15,16). Distraction angulation using the Ilizarov technique improves translation and also results in a more functional position of the wrist by improving the deformity (7). R. K. Kanojia and colleagues used a bilateral fixator before surgery for soft tissue distraction in patients with type 3 and type 4 deformities. Unilateral external fixators do not improve combined radial deviation and flexion deformity. Unilateral fixators can only provide radial improvement (12). Sanjeev Sabharwal and colleagues used the Ilizarov method for soft tissue distraction before centralization in type 4 deformity cases. The treatment was started with the use of a splint to hold the wrist at the maximum ulnar deviation for 6 weeks together with stretching exercises. Then, the Ilizarov method was used for soft tissue distraction. Those authors completed centralization using a dorsal bilobed skin flap (17), as described by Evans (7). We preferred to improve both soft tissue and bone shortness by bone distraction instead of soft tissue deregulation or distraction. Matsuno and colleagues described an improvement in wrist radial deviation using radial extension in Bayne and Klug type 2-3 cases. Radius extension provides wrist and radial movements in type 2-3 patients; because of continuous growth in children, other extensions may be required to avoid recurrences (18). Suneel B. Bhat and colleagues used the multi-axial correction (MAC) method for ulnar extension in Bayne type 4 cases. The MAC system is a monolateral external fixator system. This approach is especially beneficial in early adolescents for improving angularity and length and in cases with previous inefficient soft tissue procedure. The MAC system has some advantages compared to circular distracters, such as ease of use, easy acceptance by patients, short treatment times, low risk of complications, three-dimensional control and efficient extension (4). Kawabata H and colleagues made ulnar extensions using the Ilizarov method to improve recurrent wrist deformity with ulnar shortness and bowing in 7 patients with severe radial longitudinal deficiency who had previous wrist surgery. They described a multi-step treatment for radial longitudinal deficiency. They first performed wrist centralization and stabilization, then extended the ulna using the Ilizarov method and improved angular deformity (19). Pickford MA and colleagues made ulnar extensions in radial longitudinal deficiency using the Ilizarov method (20). In our study, ulnar extension was preferred in addition to cen-

tralization using a CEF in a 14-year-old Vater syndrome case with unilateral type 4 deformity because of serious ulnar bone and radial shortness. In the same session, centralization was performed first, and then a CEF was placed. In postoperative follow-ups, an extension of 6.5 cm. was obtained in the ulnar bone by distraction. By improving soft tissue and bone shortness in the radius, the functional and esthetic results of centralization were improved. The number of surgeries was reduced by performing two procedures in the same session, and the recovery time after surgery and the adaptation period was shortened.

The aims of type 3 and 4 radial longitudinal deficiency treatment are to improve and protect wrist deformities, to provide movement, to retain the growing capacity of the ulna and to improve appearance. In young patients, centralization, radialization and microvascular epiphyseal transfer are performed, whereas in older cases and recurrent deformities, distraction of the ulna is used (7). In one step carpal resection surgeries, ulnar shortening may be required. In these types of situations, some complications arise including plaques growing over trauma of the distal ulnar, traumas due to tension in radial neurovascular structures, extreme edema in the hand and wound site problems (7,12). Centralization is still a standard surgical treatment for radial longitudinal deficiency deformities. More positive results can be observed by using stretching exercises, splints, familial education and early surgery, especially if treatment is initiated as soon as the patients are diagnosed. We believe that better results may be achieved by performing ulnar extension with distraction in cases of advanced age, where ulnar shortness predominates.

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