CONGENITAL RADIAL CLUB HAND: RESULTS OF CENTRALIZATION IN 10 CASES

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Abstract- Congenital radial club hand is an intercalary or terminal deficiency characterized by radial deviation of the hand, marked shortening of the forearm, and generalized underdevelopment of the extremity. We performed centralization of the ulna in 12 forearms of 10 patients with severe congenital radial club hands. The mean follow up was 42 months after operation (24 to 68 months). The mean age at the operation was 16.8 months (7 to 42 months). The mean angular deformity before operation was 100 degrees (75 to 135). The angular deformity was corrected in all patients initially, but in the latest follow up the mean residual angular deformity was 19.58 degrees (5 to 45). The mean correction of angular deformity was 79.57 degrees (55 to 120). Range of motion in the wrist increased significantly after operation in both flexion-extension range and radial-ulnar deviation range (P <0.05). Stability of the wrist was acceptable in the latest follow up in all patients. We recommend that correction of congenital radial club hand by centralization should be performed in three stages. The first is preoperative splinting and corrective casting, the second centralization of the ulna and correction of angular deformity, and the third which may be longer than previous stages is postoperative splinting and bracing. This stage of treatment should be continued till skeletal maturity.

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INTRODUCTION

Radial club hand was first described in 1733 in an autopsy of a newborn with bilateral absent radii. The etiology was discussed in the 19th century as either a congenital defect of the primary radial ray anlage or an acquired lesion secondary to syphilis (1). Early management recommendations concentrated on manipulation and casting. Initial surgical management involved ulnar osteotomy to correct the bow and splitting the distal ulna for insertion of the carpus (1, 2). Reconstruction of the radius by a bone graft to support the carpus and non-vascularized epiphyseal transfer was reported in 1928 (3). The results of these procedures, however, were disappointing; causes of failure included disruption of the ulnar growth plate and subsequent increase in limb length discrepancy, inadvertent ankylosis or arthrodesis of the wrist and loss of motion, and failure of the transplanted bone to grow and with loss of the radial support (1, 3).

Centralization of the carpus on distal ulna has emerged as the preferred surgical technique to correct radial club hand. The basis for this procedure has been developed by pioneers in congenital hand surgery, who have described numerous modifications to obtain or maintain correction of the wrist on the ulna (1,4,5). Radialization involves overcorrection of the carpus on the ulna combined with tendon transfer to rebalance the wrist (6). The results of centralization have been reported in several studies but we could not find any study reporting the results of this operation in Iran. The purpose of this study was to examine the correction obtained at the time of centralization and the subsequent outcomes in the latest follow-up.

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MATERIALS AND METHODS

We treated 12 congenital radial club hands in 10 patients by centralization from 1990 to 2000. We obtained informed consent from parents of all patients.

Surgical Technique

Therapy was initiated immediately after birth with passive placement of the wrist in ulnar deviation while stabilizing the forearm. This stretching maneuver is recommended at each diaper change to facilitate soft tissue stretching. Serial casting was started at 6 weeks when the arm was large enough to do casting. Serial casting was used to maintain the correction obtained by passive manipulation. This regimen of stretching and serial casting was continued to stretch taut radial structure until the time of centralization. The surgical technique followed the established principles of centralization (4,7,8). Two incisions were used for adequate exposure of the wrist. A zigzag incision centered along the radial aspect of the wrist that allowed Z-plasty skin lengthening after centralization. Aberrant pre-axial musculo-tendinous contracted structures were released to allow passive correction of the carpus to a neutral position. A second incision was made beginning at the mid-dorsum of the wrist and extended ulnarly in a transverse and elliptical fashion to the volar midline. The flexor carpi ulnaris and ulnar neurovascular bundle were identified and protected. The carpus was exposed through a transverse carpectomy in the dorsal capsule and fibrous tissue excised from the ulno-carpal joint. The carpus was then reduced onto the distal ulna and fixed with a K-wire. Failure to achieve reduction requires repeat examination to the radial structures for any remnants of contracted tissue. In severe cases adequate reduction could not be obtained; thus alternative measures such as carpectomy, limited shaving of the distal ulna epiphysis while avoiding injury to the physis, or closed wedge osteotomy of the ulna to correct the ulnar bow may be necessary. None of our cases required carpectomy or distal ulnar excision. Postoperative cast immobilization was used for 6 to 8 weeks followed by pin removal and splinting. Motion was encouraged and the splint was weaned to night time usage, which must be continued until skeletal maturity. X-ray measurements were used to quantitate the initial deformity, amount of surgical correction, stability and degree of recurrence. Stability was classified according to X-ray. If third metacarpal, lunate and ulna were aligned in neutral, flexed and extended positions, the wrist would have grade 3 stability. If they were aligned only in neutral position and their alignment was deranged with flexion and extension, it would have grade 2 stability. If they had no alignment in neutral position, it would have grade 1 stability.

Statistical Analysis

The SPSS for Windows statistical program was used to analyze the data. All tests were performed and statistical significance was established at $P = 0.05$.

RESULTS

Patients' preoperative, operative and postoperative data are presented in tables 1 and 2. Associated anomalies were found in 6 cases as shown in table 3.

The mean follow up was 42 months after operation (24 to 68 months). The average total preoperative angulation measured 100 degrees (range, 75 to 135 with 16.51 SD). Centralization corrected the angulation an average of 79.57 degrees (range, 55 to 120) to achieve an average total angulation at the latest follow-up visit of 19.58 (range, 5 to 45). The difference between these two angles was statistically significant (P=0.002). The average preoperative total range of motion in flexion extension plane was 64.17 (range, 30 to 85) and it increased to 78.33 (range, 55 to 90) in latest follow up visit. The average preoperative range of motion in radial ulnar deviation plan was 28 (range, 20 to 40) that increased to 35.38 (range, 30-45) in the last follow-up. The differences between preoperative range of motion and postoperative range of motion in both planes were statistically significant (P=0.006). There were grade 1 stability in all 12 forearms preoperatively; all of them had grade 3 stability in the latest follow-up (P=0.048).

There were no major complications. Skin flap necrosis seen in 1 patient was treated with surgical excision of necrotic skin and the resulting defect was covered with excessive ulnar side skin.

Table 1. Patients' preoperative and postoperative data

Patient ID	Side	type	Age at time of initial surgery	Age at final follow- up	Duration of follow-up	Medial reefing	Ulnar osteotomy	Tendon transfer
I	R	4	36	88	52	+	+	-
	L	4	42	88	46	+	+	-
II	R	4	7	75	68	+	-	-
III	R	4	27	87	60	+	-	-
IV	R	4	12	60	48	+	-	-
	L	1	No operation					
V	R	4	7	38	31	+	-	-
	L	4	12	38	26	+	-	+
VI	R	1	9	43	34	+	-	+
	L	4	No operation					
VII	R	3	30	65	35	+	-	-
VIII	R	4	9	71	62	+	-	-
IX	R	4	24	48	24	+	-	+
	L	4	No operation					
X	R	3	14	40	26	+	-	+

Table 2. Preoperative and final range of motion, stability and angulation

Patient ID	Range of motion				Stability		Angulation		
	Flexion-extension		Radial-ulnar dev.						
	Preoperative	Final	Preoperative	Final	Preoperative	Final	Preoperative	Postoperative	Final
I	65	75	30	25	1	3	90	0	25
II	60	75	40	20	1	3	100	0	45
III	80	85	20	30	1	3	90	5	15
IV	60	90	20	30	1	3	100	0	20
V	60	75	25	35	1	3	100	3	40
VI	40	65	30	45	1	3	110	3	4
VII	30	55	40	35	1	3	135	3	15
	70	90	25	40	1	3	90	5	15
IX	85	75	20	45	1	3	75	0	20
X	70	90	30	40	1	3	90	3	15
	80	85	30	45	1	3	125	5	5
XII	70	70	25	40	1	3	95	0	25

Table 3. Associated anomalies

Associated anomalies	No. of cases
Hypertelorism	2
Absent kidney	2
Imperforated anus	1
Scoliosis	1
Inguinal hernia	1
Undescended testis	1

DISCUSSION

In radial club hand there are abnormalities of entire limb, from the shoulder to the digits, which involve bones, joints, muscles, tendons, and nerves. (1, 4, 9). Forearm is the most severely affected part with a hypoplastic or absent radius and a deficient ulna results in a shortened segment. The ulna is approximately 60% of normal length at birth; this discrepancy persists throughout growth (1, 10, 11). The wrist is positioned in radial deviation and will eventually develop a perpendicular relationship with the forearm if untreated.

Surgical reconstruction of radial club hand is a formidable task because of degree of deformity, constellation of abnormalities and compensatory patterns that have developed (12). The goal of treatment of radial club hand is a functional and cosmetically acceptable upper limb. A stable wrist motion and sufficient forearm length are important prerequisites for achieving this (10). The principles of treatment are to correct the radial deviation of the wrist, balance the wrist on the forearm and maintain some wrist and full finger range of motion, and allow growth of forearm. The length of ulna is important, although in most reported series the final length of the ulna was 50% of normal (10).

Centralization of the wrist on the distal ulna is the standard procedure for radial club hand correction. Centralization increases the functional length of forearm by placing the wrist on the ulna and improves the alignment of the digital tendons. Tendon transfer is recommended to restore balance of the wrist and augment function (5, 6). Restoring muscle imbalance is very important for prevention of recurrence of angular deformity (13). Corrective osteotomy of the ulna is recommended if the angular deformity is less than 30 degrees. Lengthening of the ulna is not necessary for improving function, but it may be helpful for correction of cosmetic handicap. It is recommended that lengthening be performed in two stages at 6 and 12 years after centralization procedure that has been performed before the age of one year (13).

Comparison of our results with previous reports is difficult because of the myriad of surgical modifications, variations in technique to determine the deformity, and potential inconsistencies in measurement (1, 10). Lamb monitored 31 centralizations for an average of 5 years and measured a preoperative radial deviation of 78 degree and a follow-up angle of 22 degrees (11). Manske *et al.* reported 21 cases of radial club hand; the hand-forearm angle was 58 degrees before surgery and 26 degrees at an average of 34 months after surgery (5). Watson *et al*. observed 12 centralization for 10 years and reported a recurrence of an average of 30 degree (8). Bayne and Klug monitored 53 patients for an average of 8.6 years and reported 81% good or satisfactory results, defined as a hand-forearm angle of less than 30 degrees (7).

Geck *et al.* recently reported the results of modified centralization in 15 extremities and radialization in 14 limbs with a mean follow-up period of 50 months. The preoperative total angulation measured 33 degrees, with no significant differences between centralization and radialization cases (14).

Goldfarb *et al.* studied functional outcome after centralization for radius dysplasia in 25 wrists at an average of 20 years after surgery (l5). Damore *et al*. reported 14 patients with an average follow-up period of 7 years. They showed initial significant improvement from a preoperative total angulation of 83 degree to an immediate postoperative angle of 25 degree. At the latest follow-up examination the total angulation increased to 63 degrees with a loss of 38 degrees of correction (l2).

Our results of 12 centralizations in 10 patients with an average follow-up of 24 months showed significant improvement from a preoperative total angulation of 100 degrees to 19.58 degrees at the latest follow-up examination. Our follow-up was not long enough to evaluate loss of correction. We will

report our final results after adequate follow-up.

Radial club hand produces a tremendous functional handicap with far greater impairment in bilateral cases (1,11). Therapy should be started at birth with stretching and corrective casting or splinting. The second stage consists of operative procedures which include centralization, and appropriate tendon transfers. We performed centralization at approximately 1 year and attempted to perform radialization of the carpus with tendon transfer. In the third stage, physical therapy to encourage hand-to-mouth function and longterm splinting are emphasized.

Recurrence after surgical treatment of radial club hand is the most common problem following centralization (12).

The reasons for recurrence are multiple and include surgical and non-surgical factors. Surgical factors are the inability to obtain complete correction at surgery, inadequate radial soft tissue release, premature K-wire removal and failure to balance the deforming radial force (7, 14). We removed K-wire 12 weeks after surgery, which is more than previous studies (6-10). Lamb recommended K-wire retention for several months to decrease recurrence (11). Postoperative factors include poor splint use and the natural tendency for the shortened club hand to radial deviate for hand-to-mouth function and to position the more functional ulnar border of the hand toward the objects to be manipulated.

Low recurrence rate in our patients could be due to several reasons. First, our follow up was not long enough. Second we did preoperative preparation of our patients exactly and carefully. Third, we tried our best to relieve the above mentioned surgical factors during operation. And finally postoperatively we followed patients meticulously and planned physiotherapy and appropriate splinting in all patients.

In conclusion, good preoperative preparation (stretching and splinting), on time and correct operation, and finally a comprehensive postoperative management could lead to satisfactory results in patients with radial club hand.

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