The Recurrence of Deformity After Surgical Centralization for Radial Clubhand

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Fourteen children representing 19 cases of radial clubhand had centralization of the carpus on the distal ulna during an 18-year period. Age at the time of the initial surgery averaged 3.2 years (range, 0.7–8.1 years) and the follow-up periods averaged 6.5 years (range, 1.5–22.2 years). There were 16 type IV radial and 3 type III clubhands. Preoperative, postoperative, and follow-up x-rays were used to determine the initial deformity, amount of surgical correction, and degree of recurrence. The total angulation (the combination of the radial deviation of the hand and the ulna bow) was measured. The average preoperative angulation measured 83° (range, 55° to 110°). Centralization corrected the angulation an average of 58° (range, 15° to 95°) to an average immediate postoperative total angulation of 25° (range, 5° to 60°). At the final follow-up examination there was a loss of 38° (range, 5° to 105°) and the total angulation increased to an average of 63° (range, 20° to 120°). The difference between the preoperative, postoperative, and follow-up angles was statistically significant. There was a significant correlation between the preoperative angle and the final angle, the preoperative angle and the amount of correction, the amount of correction obtained at surgery and the recurrence of the deformity, and the age at time of initial surgery and the amount of recurrence. (J Hand Surg 2000;25A:745–751. Copyright © 2000 by the American Society for Surgery of the Hand.)

Key words: Radial clubhand, centralization, surgery, recurrence.

Radial clubhand was first described in 1733 in an autopsy of a newborn with bilateral absent radii.[1] 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.[2] Early management recommendations concentrated on manipulation and casting. Initial surgical treatment involved ulnar osteotomy to correct the bow and splitting the distal ulna for insertion of the carpus.[2,3] Reconstruction of the radius by a bone graft to support the carpus was reported in 1928[4] and nonvascularized epiphyseal transfer was reported in 1945.[5] The results of these procedures were disappointing; multiple 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.[2,4]

Centralization of the carpus on the distal ulna has emerged as the preferred surgical technique to correct radial clubhand. 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.[2,6–10] Radialization involves overcorrecting of the carpus on the ulna combined with tendon transfer

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Table 1. Associated Anomalies

Associated Anomalies	*No. of Cases*
Cardiac	
Atrial septic defect	3
Ventricular septic defect	1
Skeletal	
Congenital scoliosis	4
Kleppel-Feil syndrome	1
Spina bifida	1
Fused ribs	1
Metatarsus adductus	1
Clubbed feet	1
Fibular hemimelia	1
Gastrointestinal	
Imperforate anus	1
Tracheoesophageal fistula	1
Neurologic	
Hydrocephalus	1
Mental retardation	1
Hematologic	
Thrombocytopenia	1
Genitourinary	
Absent kidney	1

to rebalance the wrist.[11] The results of centralization have been reported but few studies report the degree of recurrence over time.[8–10,12,13] The purpose of this study was to examine the correction obtained at the time of centralization and subsequent loss during the follow-up period.

Materials and Methods

Fourteen patients had centralization for radial clubhand between 1974 and 1992. Bilateral surgery was performed in 5 patients, for a total of 19 centralization procedures. There were 8 girls and 6 boys. Eleven patients had numerous associated anomalies (Table 1). There were 16 type IV and 3 type III radial clubhands (Table 2). The initial surgery was performed at an average age of 3.2 years (range, 0.7–8.1 years). The average age at the follow-up examination was 9.7 years (range, 4.1–22.8 years). Four patients required revision surgery for recurrence of wrist deformity; x-rays obtained after the revision procedure were used to evaluate outcome.

Surgical Technique

Therapy was initiated immediately after birth with passive placement of the wrist into ulnar deviation while stabilizing the forearm. This maneuver is recommended at each diaper change to facilitate soft tissue stretching. Nighttime splinting was started at approximately 3 months of age when the arm was large enough to don a splint. Serial splinting was used to maintain the correction obtained by passive manipulation. This regimen of stretching and splinting was continued until the time of centralization. Surgery was performed following the extensive preoperative therapy to stretch the taut radial structures.

Table 2. Radial Clubhand Patient Data

Patient ID No.	*Involvement*	*Surgical Side*	*Type*	*Age at Time of Initial Surgery (yr)*	*Age at Final Follow-up (yr)*	*Duration of Follow-up (yr)*	*Medial Reefing*	*Tendon Advance*
10845L	Left	Left	IV	0.7	22.8	22.2	Yes	No
11657L	Left	Left	IV	3.1	7.9	4.8	Yes	No
11935R	Bilateral	Right	IV	6.6	9.5	2.9	Yes	No
11935L	Bilateral	Left	IV	6.9	9.5	2.6	Yes	No
12308R	Bilateral	Right	III	1.7	19.5	17.8	Yes	No
12308L	Bilateral	Right	III	1.7	19.5	17.8	Yes	No
12651R	Right	Right	IV	8.1	12.8	4.7	Yes	No
14469R	Right	Right	IV	4.1	15.5	11.4	Yes	No
22787R	Bilateral	Right	IV	1.9	8.5	6.6	Yes	No
22787L	Bilateral	Left	IV	2.8	8.5	5.7	Yes	No
23186L	Bilateral	Left	IV	3.5	5.0	1.5	Yes	No
23186R	Bilateral	Right	IV	4.1	5.0	0.9	Yes	No
25984R	Bilateral	Right	IV	1.7	5.3	3.6	Yes	Yes
26683R	Right	Right	III	3.8	7.7	3.9	Yes	No
26726L	Left	Left	IV	1.8	4.3	2.4	Yes	No
27471R	Bilateral	Right	IV	0.8	4.1	3.3	Yes	Yes
27471L	Bilateral	Left	IV	0.9	4.1	3.2	Yes	Yes
27834R	Bilateral	Right	IV	3.7	7.0	3.3	Yes	No
27834L	Bilateral	Left	IV	3.8	7.0	3.3	Yes	No

The surgical technique followed previously established principles for centralization.[6,7,9,10,13] Two incisions were used for adequate exposure of the wrist. A zigzag incision centered along the radial aspect of the wrist was made through the skin and subcutaneous tissue. This incision allowed Z-plasty skin lengthening after centralization. The enlarged median nerve and its anomalous dorsal branch were identified in the skin fold at the wrist. Aberrant preaxial musculotendinous units and anomalous fibrous bands were released to allow passive correction of the carpus to a neutral position.

A second incision was made beginning at the middorsum of the wrist and extended ulnarly in a transverse and elliptical fashion to the volar midline. This approach provided exposure of the carpus and excision of redundant skin and subcutaneous tissue. The flexor carpi ulnaris and ulnar neurovascular structures were identified and protected. The carpus was exposed by a transverse arthrotomy in the dorsal capsule and fibrous tissue excised from the ulnocarpal joint. The carpus was then reduced onto the distal ulna and secured with a K-wire. Failure to achieve reduction requires repeat examination of the radial structures for any remaining contracted or fibrotic tissue. In severe cases adequate reduction could not be obtained; thus, alternative measures were necessary, such as carpectomy, limited shaving of the distal ulna epiphysis while avoiding injury to the growth plate, or closed wedge osteotomy of the ulnar to correct the ulnar bow (Table 2).[9,10]

Postoperative cast immobilization was used for 6 to 8 weeks followed by pin removal and splint fabrication. Motion was encouraged and the splint was weaned to nighttime usage, which was recommended until skeletal maturity.

Measurement of Deformity

X-ray measurements were made to quantitate the initial deformity, amount of surgical correction, and degree of recurrence. The amount of deformity was calculated by the angle formed by 2 intersecting lines, the first being drawn along the longitudinal axis of the third metacarpal and the second along the proximal axis of the ulna. This angle represents the total angulation of the clubhand and is the combination of the radial deviation of the wrist and the ulna bow.

Statistical Analysis

The SPSS for Windows statistical program (release 6.1.4; SPSS Inc, Chicago, IL) was used to analyze the data. All tests were performed in the null hypothesis and statistical significance was established at $p = .05$. The initial analysis consisted of a

				Angulation		
Carpectomy	*Distal Ulnar Excision*	*Diaphyseal Osteotomy*	*Revision*	*Preoperative (°)*	*Postoperative (°)*	*Final (°)*
Yes	Yes	No	Yes	70	15	20
Yes	No	Yes	Yes	64	22	55
Yes	No	No	No	90	15	105
Yes	No	No	No	90	25	62
No	No	No	No	95	25	90
No	No	No	No	75	60	85
Yes	No	No	No	75	20	80
Yes	Yes	No	No	90	30	120
Yes	Yes	Yes	No	110	30	68
Yes	Yes	No	No	95	60	78
Yes	Yes	No	No	80	5	35
Yes	No	No	No	95	25	35
No	No	Yes	Yes	70	12	65
Yes	No	No	No	55	5	24
No	Yes	No	No	100	35	40
No	Yes	No	No	80	15	47
No	No	No	Yes	65	15	37
Yes	Yes	No	No	95	30	60
Yes	No	Yes	No	90	30	57

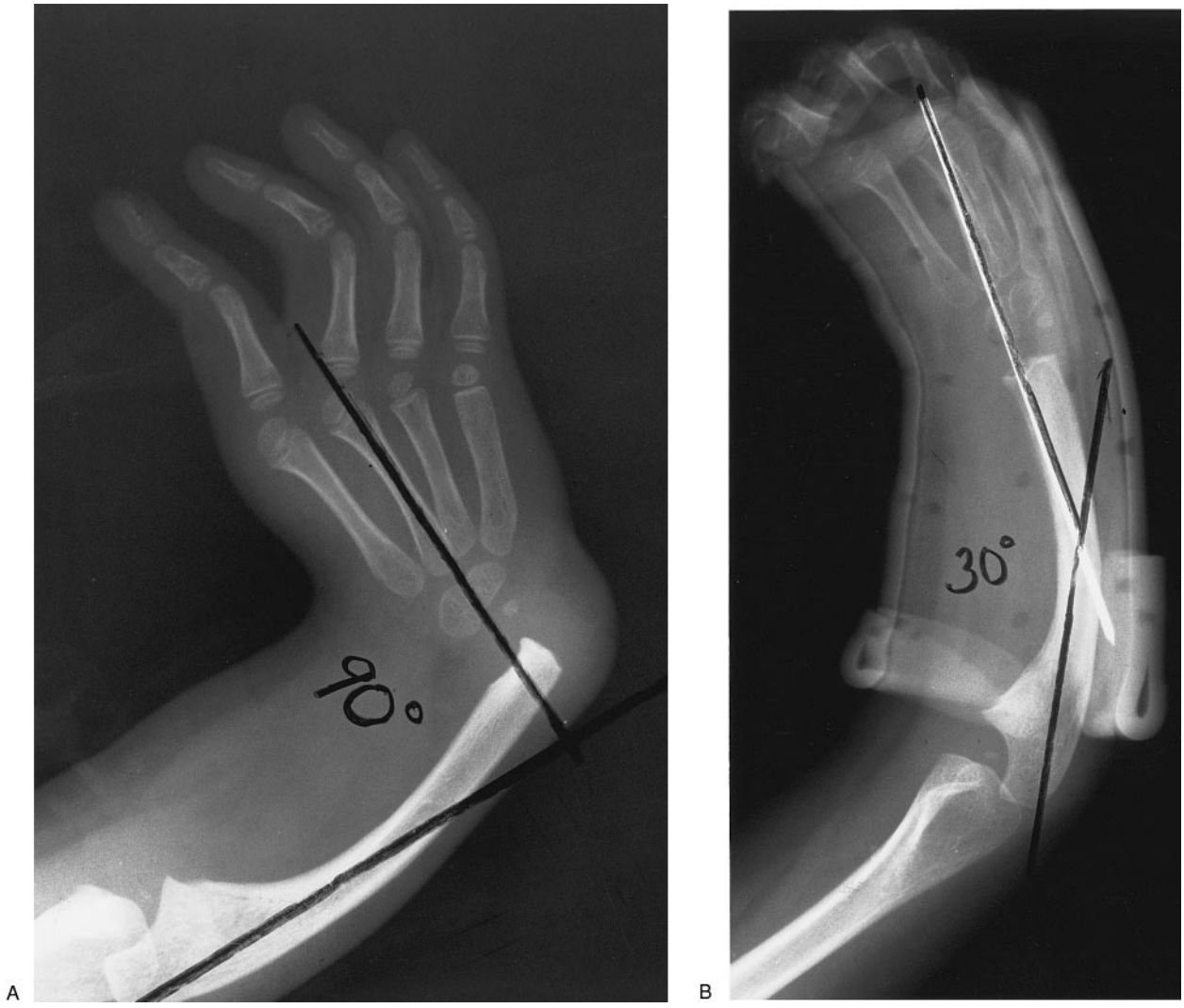
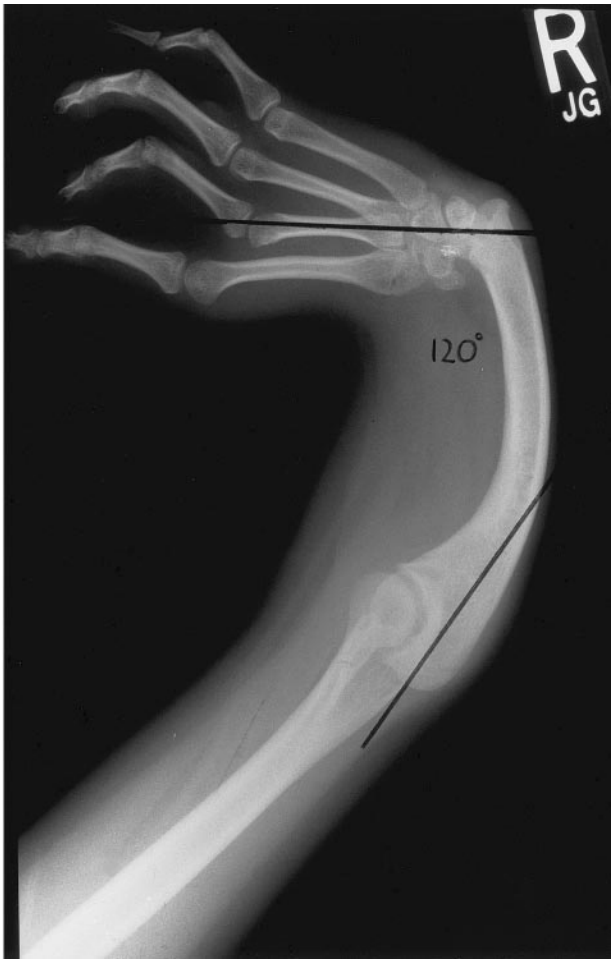


Figure 1. (A) Preoperative x-ray of type IV radial clubhand with 90° of total angulation. (B) Postoperative x-ray obtained after centralization with correction to 30°. *(Figure continues)*

1 × 3 ANOVA with angle and x-ray interval (preoperative, postoperative, and follow-up) treated as within-subject independent variables. Because a significant difference was found, post hoc analysis was performed to determine where statistical significance existed. Bonferoni's *t*-procedure was used to control for type I error in the post hoc analysis. Spearman's correlation coefficient was performed between age at time of initial surgery, age at final follow-up examination, length of follow-up period, surgical variables (capsular reefing, extensor carpi ulnaris advancement, carpectomy, shaving the distal ulna, and ulna osteotomy), preoperative angle, postoperative angle, final angle, amount of correction, and amount of recurrence.

Results

The average preoperative total angulation measured 83° (range, 55° to 110°). Centralization corrected the angulation an average of 58° (range, 15° to 95°) to achieve an average immediate postoperative total angulation of 25° (range, 5° to 60°). At the final follow-up visit there was an average loss of 38° (range, 5° to 105°) from the postoperative angle and the total angulation increased to an average of 63° (range, 20° to 120°). The difference between these 3 angles was statistically significant. For example, Figure 1A shows a preoperative x-ray of a 4-year-old girl with type IV radial clubhand that measured 90° of total angulation. Following centralization, the



C

Figure 1. (C) *(Continued)* Follow-up x-ray showing considerable loss of correction and total angulation measuring 120°.

postoperative angulation measured 30° (Fig. 1B). Follow-up x-rays obtained at 16 years of age showed significant recurrence (90°), with the total angulation measuring 120° (Fig. 1C).

There was a statistically significant correlation found between preoperative angle and final angle ($r = .54$, $p = .02$), preoperative angle and amount of correction ($r = .64$, $p = .01$), amount of correction that was obtained at surgery and amount of angular recurrence ($r = .57$, $p = .01$), and age at time of initial surgery and recurrence of deformity ($r = .51$, $p = .02$).

Discussion

In radial clubhand there are abnormalities of the entire limb, from the shoulder to the digits, that involve the bones, joints, muscles, tendons, and nerves.[2,6,14] The forearm is most affected with a hypoplastic or absent radius and a deficient ulna resulting in a shortened segment. The ulna is approximately 60% of normal length at birth; this discrepancy persists throughout growth.[2,7,8] The wrist is positioned in radial deviation and will eventually develop a perpendicular relationship with the forearm if untreated.[12]

Surgical reconstruction of the radial clubhand is a formidable task secondary to the degree of deformity, constellation of abnormalities, and compensatory patterns that have developed.[2,6,8] The principles of treatment are to correct the radial deviation of the wrist, balance the wrist on the forearm, maintain some wrist and full finger motion, and allow growth of the forearm. Centralization of the wrist on the distal ulna is the standard procedure for radial clubhand correction. Centralization increases the functional length of the forearm by placing the wrist on the ulna and improves the alignment of the digital tendons. Tendon transfer is recommended to restore balance to the wrist and augment function.[10,11]

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.[2,7,9,10,15,16] Heikel[2] stressed the difficulties with measurements of angles in radial clubhand, including inexact determination of lines and the difficulties in obtaining a standardized x-ray view. Manske et al[10] and Bayne and Klug[9] recommended the hand–forearm angle, while Bora et al[7] used the ulnar bow to assess deformity and recurrence. We preferred total angulation over hand–forearm angle and ulnar bow to assess the deformity because "the clinically apparent radial angulation deformity could be, in part, secondary to bowing of the forearm,"[10] which is accounted for in the calculation of total angulation. Total angulation was initially discussed by Heikel[2] and has been used for the preoperative determination of radial clubhand before Ilizarov application.[15]

Lamb[8] monitored 31 centralizations for an average of 5 years and measured a preoperative radial deviation of 78° and a follow-up angle of 22°. Bora et al[12] reported 14 extremities at an average of 14.6 years after centralization and showed a gradual increase in the hand–forearm angle to an average of 25°. Manske et al[10] reported 21 cases of radial clubhand; the hand–forearm angle was 58° before surgery and 26° at an average of 34 months after surgery. Watson et al[13] observed 12 centralizations for 10 years and reported a recurrence to an average of

30°. Bayne and Klug[9] monitored 53 patients for an average of 8.6 years and reported 81% good or satisfactory results, which was defined as a hand–forearm angle less than 30°. Geck et al[16] recently reported 29 cases in 23 patients with a mean follow-up period of 50 months. A modified centralization procedure was performed in 15 extremities and 14 limbs had radialization. The preoperative total angulation measured 92°, which improved to 21° after surgery. Eight limbs required revision surgery. Final total angulation measured 33°, with no significant differences between centralization and radialization cases.

Our results of 14 patients with an average follow-up period of 7 years showed initial significant improvement from a preoperative total angulation of 83° to an immediate postoperative angle of 25°. At the follow-up examination, the total angulation had increased to 63° with a 38° loss of correction. This recurrence is greater than noted in previous reports, but includes both hand–forearm angle and ulnar bow. This angulation is also less in untreated children, who develop a hand–forearm angle of approximately 100° that remains constant over time.[12]

Recurrence after surgical treatment of radial clubhand is the most common problem following centralization.[2,9] The reasons for recurrence are multiple and include surgical and nonsurgical 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.[9,16] We removed the K-wire 6 to 8 weeks after surgery, which is similar to previous studies.[7,9,11,13,14] However, K-wire retention for several months also has been recommended, presumably to decrease recurrence.[8] Postoperative components are poor splint use and the natural tendency for the shortened clubhand to radial deviate for hand to mouth use and to position the more functional ulnar border of the hand toward objects to be manipulated.[8] These surgical and nonsurgical factors certainly contributed to the considerable recurrence in our patient population.

We obtained the greatest correction in the patients with a high preoperative angle, but these patients had the most recurrence. We were often unable to completely correct the preoperative deformity despite distal ulnar shaving, carpectomy, and ulnar osteotomy. Failure to completely correct the hand–forearm angle has been shown to increase the revision rate.[16] In such cases, recurrence may be related to residual imbalance of the carpus on the distal ulna. Compliance with postoperative splint wear is difficult to monitor and failure to protect the alignment obtained is another potential source of recurrence.

These retrospective results have altered our current approach to radial clubhand. In children with radial clubhand that is supple and easily correctable, we proceed with centralization at approximately 1 year of age and attempt to perform radialization of the carpus combined with tendon transfer. Therapy to encourage hand to mouth function without associated radial deviation and long-term splinting are emphasized. In children with a rigid deformity, preliminary soft tissue distraction to lengthen and reposition the carpus before formal centralization also has increased our armamentarium to treat radial clubhand and is used to lengthen the shortened ulna after centralization. These procedures are offered once the child is old enough to comprehend the concept and to participate in the postoperative care.

Radial clubhand creates a tremendous functional handicap with far greater impairment in bilateral cases.[2,8] However, the degree of initial angulation or subsequent recurrence is difficult to correlate with functional outcome for numerous reasons. First, most children undergo surgery during infancy, which precludes comparison of function before and after surgery. Second, associated systemic (eg, hematopoietic, cardiac, or vertebral) and local (eg, thumb hypoplasia) anomalies directly influence use of the extremity.[2,17] Therefore, we could not determine a reliable method to assess functional outcome that would specifically reflect the degree of radial clubhand deformity.

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