

*Short Communication***Recurrence rate after radial club hand surgery in long term follow up***Hooman Shariatzadeh^a, Dawood Jafari^a, Hamid Taheri^a, Farid Najd Mazhar^{*b}***Abstract**

BACKGROUND: Radial club hand includes wide spectrum of malformations that involve radial side of forearm. Conservative treatments are recommended to the milder types and surgical interventions specially centralization for the severe forms. Recurrence after centralization is one of the challenging problems in this anomaly. In this study, we assessed the recurrence rate of centralization in radial club hand surgery in long term follow up.

METHODS: In this retrospective study, we reviewed the records and radiographies of 9 patients with radial club hand who underwent centralization to correct the deformity in the hand surgery department. Patients' age, type of the thumb anomaly, associated anomalies in other systems, involved side, type of radial club hand, hand-forearm angle preoperative, post operative and in the last follow up visit, and centralized wrists motion range in the final follow up visit were all assessed and reported.

RESULTS: Eleven cases of radial club hand in nine patients underwent centralization. Mean age at the time of the centralization was 17 (6-72) months and mean follow up was 90 (48-170) months. Preoperative hand-forearm angle was 75 (30-110) degrees, immediate postoperative angle was 25 (15-35) degrees and in the last follow up visit it was 52 (40-60) degrees. The amount of correction was 66% and loss of correction in long term was 54%.

CONCLUSION: Centralization still can be a standard procedure in treatment of patients with radial club hand with acceptable results.

KEYWORDS: Radial club hand, centralization, recurrence, surgical treatment, wrist, deformity.

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Radial club hand or radial deficiency includes wide spectrum of malformations that involve radial side of forearm (radius, radial carpus, and thumb), including hypoplasia of the bones and joints, muscles and tendons, ligaments, nerves, and blood vessels.^{1,2} It is the most common type of longitudinal failure of formation.³ Incidence of Radial club has been reported from 1:30,000 to 1:100,000 live births, so it is an uncommon deformity.^{2,4} Radial deficiency is commonly associated with congenital anomalies and involvement of musculoskeletal system. The most common anomalies occur in upper extremities, including humeral hypoplasia, proximal radioulnar synostosis, congenital radial head dislo-

cation and fingers stiffness. Less common findings are metacarpal synostosis and syndactylia.⁵ Bilateral deformities occur in approximately 50% of patients; when the deformity is unilateral, the right side is more commonly affected. Both sexes are equally affected.⁴ Children with bilateral and severe radial deficiency have considerable functional impairment due to thumb dysfunction, shortness of upper extremities and wrist instability.² Different classification systems have been developed for radial club hand, among them Heikel classification and Bayne classification are more acceptable.^{2,4} Nonoperative treatment has been advocated for mild deformities but surgical intervention especially centralization are used for

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severe deformities.² Long-term follow-up in few studies report problems with recurrence and stiffness.^{6,7} Centralization has been used as a surgical procedure for decades to correct the malformation with noticeable achievements. Statistically significant difference between the preoperative, postoperative, and follow-up hand forearm angles has been reported after this procedure.^{6,7} The purpose of this study is to report the concomitant abnormalities, clinical course, results and recurrence rate of this rare malformation after centralization in long term follow up.

Methods

We retrospectively reviewed the medical records and radiographies of patients with radial club hand who underwent centralization to improve function and appearance at our hospital between the years of 1990 and 2002. From these patients 9 cases had the requested criteria to enter the study. The inclusion criteria were:

- 1- The patient must have congenital deficiency of radius or radial club hand
- 2- The patient must have undergone centralization to improve function and appearance
- 3- The records must be complete
- 4- There must be an acceptable quality of radiographies.

The age of patient at the time of operation, the abnormality type of the thumb, abnormalities in other systems, the side of involvement, the type of radial club hand, hand-forearm angle before correction, immediately after surgery and in final follow up visit, type of surgery for abnormal thumb, wrist motion range in patients with unilateral involvement compared to uninvolved side, all were assessed and recorded. Similar to other surgeons, the authors believe that the accepted and most useful classification of congenital radial dysplasias is a modification of that proposed by Heikel, in which four types are described, type one is mildest and type four is the most severe form of the malformation.⁴

For assessment of hand-forearm angle the authors did respect the method which has been proposed by Manske et al.^{8,9} According to this method, hand-forearm angle is the angle between the longitudinal axis of third metacarpal bone and longitudinal axis of the ulna. The longitudinal axis of the ulna is a line which is perpendicular to the distal physis of the ulna. The bowing of the ulna was the angle between the longitudinal axis of proximal and distal ulna (figure1). There were also contraindication for surgical intervention which included:



Figure 1. Method of determining the hand-forearm angle and bowing of ulna.

- 1- Severe associated anomalies not compatible with long life
- 2- Inadequate elbow flexion.
- 3- Mild deformity with adequate radial support
- 4- Older patients who have accepted the deformities and have adjusted accordingly
- 5- Patients with severe soft tissue contracture including nerves and vessels.¹⁰

Besides the surgical incisions which were different, the surgical method in these patients was in accordance with the recommendations of Manske et al.⁹ According to this method of

centralization, distal ulna without disturbing the physis was located at the middle of carpus. Tendon transfers were not used but reefing or advancement of extensor carpi ulnaris were part of procedure. After surgery the correction was maintained by longitudinal pin and the extremity was immobilized in long arm cast for 6-8 weeks (figure 2-6). Pin was maintained in its place for 8-12 weeks. The forearm and hand were placed in appropriate splint after removing the pin as long as possible in accordance to the patient's compliance.



Figure 2. Type 4 radial club hand



Figure 3. The same patient after 6 months

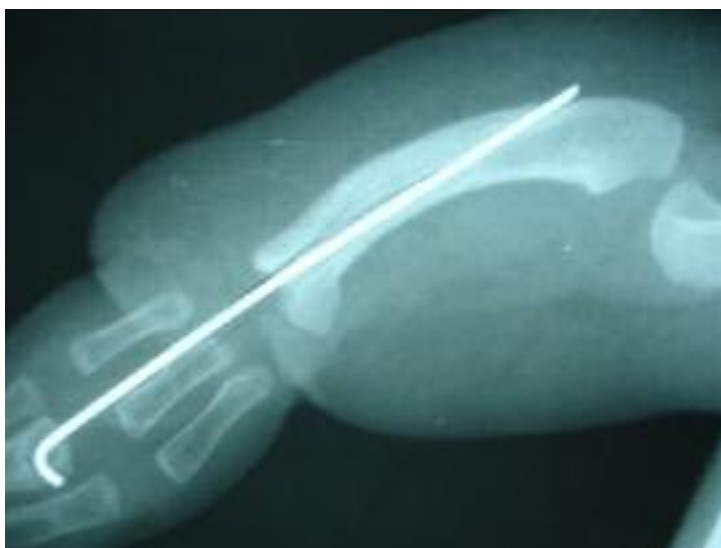


Figure 4. After centralization at the age of 18 months.



Figure 5. Six months post surgery: pin is broken due to early range of motion.



Figure 6. Hand and forearm of the same patient at the age of 7 years.

Results

Eleven cases of centralization were done in nine patients. Four patients had bilateral involvement and centralization was done only in severely involved extremities. We had 4 male and 5 female patients. The patients' profile is summarized in table 1. According to Heikel's classification, two cases had type one, four cases had type 3 and seven cases had type four involvement. Thumb abnormality was present in all cases and it included 2 hypoplastic thumbs, two bilateral floating thumbs and in the remaining the thumb was absent. Associated anomalies were present in 5 cases, one fibular hemimelia, one bladder extrophy, one congenital bilateral dislocation of hip, one unilateral dislocation of hip and one tibia hemimelia. There was no hematopoietic system involvement. Mean age was 17(6-72) months at the time of centralization and 33(24-72) months at the time of pollicization. Mean

follow up period was 90(48-170) months. Mean hand-forearm angle before surgery was 75(30-110) degrees, immediately after correction it was 25(15-35) degrees and in final follow up visit, this angle was 52(40-60) degrees. The amount of correction was 66% and loss of correction in long term was 54%. Six pollicizations were done in 6 patients. The average ulnar bowing was 30 degrees. Ulnar corrective osteotomy was done in three cases which had 35, 40 and 40 degrees of bowing. In unilateral involvement we compared the wrist range of motion in corrected side with the normal one in the last follow up visit. In involved side the average range of motion in flexion extension direction was 49% of the uninvolved side and in ulnar radial direction it was 63% (table 2). The average period for post operative splintage was 9(3-24) months. In 3 cases splintage time was 24 months and in these 3 patients the mean loss of correction was 20 degrees.

Table 1. Patient's profile.

Patient no.	Sex	Involved side	Type of anomaly	Involvement	Age (month)	Follow up (month)
1	F	R	III	Unilateral	6	60
2	M	R	IV	Unilateral	10	48
3	M	L	IV	Unilateral	6	120
4	M	R	III	Unilateral	24	60
5	F	R-L	I- IV	Bilateral	24	70
6	F	R-L	III- IV	Bilateral	7-13	72
7	F	L	IV	Unilateral	72	170
8	M	R-L	I- IV	Bilateral	18	52
9	F	R-L	III-IV	Bilateral	24-32	160

F Female

M Male

R Right

L Left

Table 2. Motion range of centralized wrists in comparison to uninvolved side in unilateral cases at final follow up visit.

Patient no.	Flexion – Extension Degrees	Ulnar - Radial Degrees
	(% of normal wrist)	(% of normal wrist)
1	85 (56%)	35 (75%)
2	75 (50%)	30 (70%)
3	70 (43%)	30 (60%)
4	80 (53%)	35 (75%)
7	65 (43%)	25 (62%)

Discussion

After 250 years of first description, radial club hand still remains one of the most controversial and complex management challenges in reconstructive hand surgery.^{10,11} Radial side of upper extremity is hypoplastic in this anomaly in different grades. The obvious deformity of a short forearm and radially deviated hand is almost invariably present at birth. The forearm is between 50% and 75% of the length of the contralateral forearm, a ratio that usually remains the same throughout periods of growth.^{2,10} Radial club hand can be associated with severe anomalies in other systems including hematopoietic, cardiovascular, urinary and gastrointestinal systems which can be life threatening.^{10,12,13} Type one treatment usually is confined to the thumb reconstruction but in severe forms and in the absence of contraindications, surgical treatment for centralization can be proposed in most situations and for years.¹⁰ Also it is said that centralization can improve the appearance of the extremity but has not been proven to enhance function.² But some reports emphasized that centralization has been shown to improve function, particularly in bilateral involvement.⁴ After centralization the surgeon may encounter early or late complications. The most important late complication is recurrence of deformity due to failure of surgical treatment and progressive bowing ulna.^{10,14} There are few reports in literature which did pay attention to this kind of complication.¹⁴⁻¹⁶ Kozin et al in their report followed up 19 cases of centralization in 14 patients.¹⁵ In their study, preoperative, postoperative and follow-up x-rays were used to determine the initial deformity, amount of surgical correction and degree of recurrence. The average preoperative angulation measured 83 degrees (55-110). Centralization corrected the angulation an average of 58 degrees (15-95) to an average immediate postoperative angulation of 25 degrees (5-60). At the final follow-up examination there was a loss of 38 degrees (5-105) and the total angulation increased to an average of 63 degrees (20-120). Age at the time of the initial surgery averaged 3.2 years (0.7-8.1) and the

follow-up periods averaged 6.5 years (1.5-22.2).¹⁵ They also found a significant correlation between the preoperative angle and the final angle.¹⁵ Comparison of the results of their study with our study confirms that the results of centralization are similar.

Some clinicians believe that, because of stiffness after surgery, centralization is not recommended in radial club hand especially in bilateral involvements.¹⁷ But according to Bayne, although recurrence and complications are common after centralization, patients and their parents are pleased with the procedure.¹⁰ In an attempt to reduce the complications and recurrence after centralization, some authors recommend preoperative traction and external fixator application.^{18,19} Recently in an effort to reduce the recurrence rate Buck-Gramcko proposed the method of "Radialization" with emphasizing to soft tissue reconstruction and more ulnar deviation or the wrist.²⁰ He believes that the procedure is successful and recommends it at the age of 6-12 months. But Lourie et al did not recommend the radialization procedure in patients who will have pollicization in future.¹⁰ Also, Geck's study found no significance difference between the two procedures of radialization and centralization, despite more soft tissue dissection in radialization.^{7,10} In our study, in spite of 54% loss of correction, patients have acceptable range of motion in the corrected wrists comparing to the uninvolved wrists in patients with unilateral radial club hand. And we believe that this is in accordance with James et al that "the wrists tend to be either flexible and deviated or stiff and straight".²

The authors do believe that current study has following limitations:

- 1- Different surgeons did the procedures
- 2- We did not compare centralization with radialization.
- 3- Follow up period in our study is shorter than some other studies.
- 4- We did not evaluate the functional abilities of centralized wrists by standard tests such as Jebsen-Taylor test and DASH questionnaire.

This will be the subject of our studies in the future.

In conclusion, centralization still can be a standard procedure in surgical treatment of radial club hand in our country. The authors

recommend further studies on new areas such as preoperative traction with external fixator application and also functional assessment of the operated wrist.

Conflict of Interest

Authors have no conflicts of interest.

Authors' contributions

HSh carried out the design and coordinated the study, participated in most of the experiments and prepared the manuscript. DJ provided assistance in the design of the study, carried out all the experiments and participated in manuscript preparation. HT provided assistance in the design of the study, carried out all the experiments and participated in manuscript preparation. FNM provided assistance in the design of the study, coordinated and carried out all the experiments and participated in manuscript preparation. All authors have read and approved the content of the manuscript. They participated in editing and revision processing.

References

1. Skerik SK, Flatt AE. The anatomy of congenital radial dysplasia. Its surgical and functional implications. *Clin Orthop Relat Res* 1969; 66:125-43.
2. Green D, Hotchkiss R, Pederson W, Wolfe S. *Green's operative hand surgery*. 5th ed. Philadelphia: Churchill Livingstone; 2005.
3. Giele H, Giele C, Bower C, Allison M. The incidence and epidemiology of congenital upper limb anomalies: a total population study. *J Hand Surg [Am]* 2001; 26(4):628-34.
4. Jobe MT, Wright PE. Congenital anomalies of the hand. In: Canale ST, Beaty JH, editors. *Campbell's operative orthopaedics*. St Louis: Mosby; 2007. p. 4375-90.
5. James MA, McCarroll HR, Jr, Manske PR. Characteristics of patients with hypoplastic thumbs. *J Hand Surg [Am]* 1996; 21(1):104-13.
6. Damore E, Kozin SH, Thoder JJ, Porter S. The recurrence of deformity after surgical centralization for radial club-hand. *J Hand Surg [Am]* 2000; 25(4):745-51.
7. Geck MJ, Dorey F, Lawrence JF, Johnson MK. Congenital radius deficiency: radiographic outcome and survivorship analysis. *J Hand Surg [Am]* 1999; 24(6):1132-44.
8. Buck-Gramcko D. *Congenital malformations of the hand and forearm (The hand & upper limb)*. Philadelphia: Churchill Livingstone; 1998.
9. Manske PR, McCarroll HR, Jr, Swanson K. Centralization of the radial club hand: an ulnar surgical approach. *J Hand Surg [Am]* 1981; 6(5):423-33.
10. Lourie GM, Lins RE. Radial longitudinal deficiency. A review and update. *Hand Clin* 1998; 14(1):85-99.
11. Gupta A, Kay SPJ, Scheker LR. *The growing hand: diagnosis and management of the upper extremity in children*. Philadelphia: Mosby Ltd; 1999.
12. Beals RK, Rolfe B. VATER association. A unifying concept of multiple anomalies. *J Bone Joint Surg Am* 1989; 71(6):948-50.
13. Dell PC, Sheppard JE. Thrombocytopenia, absent radius syndrome: report of two siblings and a review of the hematologic and genetic features. *Clin Orthop Relat Res* 1982; 162:129-34.
14. Bayne LG, Klug MS. Long-term review of the surgical treatment of radial deficiencies. *J Hand Surg [Am]* 1987; 12A(2): 169-79.
15. Damore E, Kozin SH, Thoder JJ, Porter S. The recurrence of deformity after surgical centralization for radial club-hand. *J Hand Surg [Am]* 2000; 25(4):745-51.
16. Geck MJ, Dorey F, Lawrence JF, Johnson MK. Congenital radius deficiency: radiographic outcome and survivorship analysis. *J Hand Surg [Am]* 1999; 24(6):1132-44.

17. Frankel ME, Goldner JL, Stelling FH. Radial club hand: is centralization necessary? A rational surgical approach. J Bone Joint Surg Am 1971; 53:1026.
18. Nanchahal J, Tonkin MA. Pre-operative distraction lengthening for radial longitudinal deficiency. J Hand Surg [Br] 1996; 21(1):103-7.
19. Smith AA, Greene TL. Preliminary soft tissue distraction in congenital forearm deficiency. J Hand Surg [Am] 1995; 20(3):420-4.
20. Buck-Gramcko D. Radialization as a new treatment for radial club hand. J Hand Surg [Am] 1985; 10(6 Pt 2):964-8.

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