



## Recurrence rate of radial deviation following the centralization surgery of radial club hand

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Received: 24 Apr 2017

Published: 8 Mar 2018

### Abstract

**Background:** Congenital radial club hand (RCH), as a rare congenital deformity of the upper extremity, is characterized by a wide spectrum of malformations including radial deviation. Centralization surgery is the standard treatment for severe cases that have been associated with a high rate of recurrence. This study reports the long-term results and recurrence rate of radial deviation following the centralization surgery of RCH.

**Methods:** The medical records of 13 congenital RCH patients (16 hands), who underwent centralization surgery, were reviewed retrospectively. Hand-forearm angle (HFA), hand-forearm position (HFP), and ulnar bow (UB) were used to assess forearm angles.

**Results:** The mean age of the patients was 19.4±8.9 months, and their mean follow-up was 62.1±39.9 months. The mean HFA correction was 29.4°±23.9°, the mean HFA recurrence was 13.3°±13.7°, the mean correction of HFP was 13.4±7.3 mm, and the mean recurrence of HFP was 1.4±2.8 mm. The mean UB showed 7.6°±12.5° correction immediately after surgery and a further 3.6°±7.3° at the last follow-up (overall 11.2°±17.6°). A number of 12 out of 13 parents were completely satisfied with the results.

**Conclusion:** According to our results, an acceptable long-term result is expected after the centralization surgery of RCH. However, the risk of the recurrent radial deviation is high and needs to be optimized in future investigations.

**Keywords:** Radial club hand, Radial deviation, Centralization, Recurrence

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**Cite this article as:** Najd Mazhar F, Shariatzadeh H, Balvardi M, Nakhaei Amroodi M, Mirzaei A. Recurrence rate of radial deviation following the centralization surgery of radial club hand. *Med J Islam Repub Iran.* 2018(8 Mar);32:18. <https://doi.org/10.14196/mjiri.32.18>

### Introduction

Radial club hand (RCH) is a congenital anomaly of the upper extremity, characterized by various degrees of hand and forearm deformities, ranging from mild radial hypoplasia to complete absence of the radius. Since the entire radial column of the forearm is hypoplastic, the affected child often lacks a functional thumb and usually need pollicization later in life (1, 2). It is a relatively rare deformity with an incidence of 5 per 100 000 live births (3).

Treating this anomaly is based on its severity. While conservative treatment is recommended for milder deformities, centralization technique is the choice of treat-

ment for more severe cases, in which the carpus is surgically moved to the central portion of the distal ulna to correct radial deviation and wrist subluxation. This technique has been used and recognized as the standard correction method of RCH malformation for decades and the results have shown significant improvement in postoperative forearm angles following this operation (4, 5). However, this technique has been associated with a high rate of recurrent radial deviation (6, 7).

The present study aimed at reporting the long-term results and recurrence of radial deviation following the cen-

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#### ↑What is “already known” in this topic:

Centralization surgery has been widely used in the treatment of severe cases of congenital radial club hand (RCH), as a rare congenital deformity of the upper extremity. However, it has been associated with a high recurrence rate of radial deviation, and thus further evaluation of this method is of prime importance.

#### →What this article adds:

Our results revealed that in spite of the high recurrence rate of radial deviation following the centralization surgery, the satisfaction rate was high enough to justify the application of this method, at least until a more appropriate method is developed.

tralization surgery of RCH.

**Methods**

This study was approved by institutional review board (IRB) of Iran University of Medical Sciences (code number: 9111242008), and informed consent was obtained from the parents to report the study. The clinical and radiographic records of patients, who underwent centralization surgery for the correction of RCH deformity at our center during 1992 and 2016, were reviewed retrospectively.

Only patients with congenital RCH, who underwent centralization, were included. In addition, those patients with incomplete or inadequate medical records including unacceptable quality of radiographs were excluded from the study. In this respect, 3 patients with incomplete or inadequate medical records were removed from the study. Finally, 13 out of 16 patients with congenital RCH, managed by centralization surgery, were included in this study. Since the deformity was bilateral in 3 cases, 16 centralization surgeries were assessed in the final study.

In total, 4 females and 9 males were included in this study. The mean±SD age of patients was 19.4±8.9 months, ranging from 6 to 36 months; the mean±SD fol-

low-up of patients was 62.1±39.9 months, ranging from 12 to 132 months.

Modified Heikel RCH typing system (8) has been used to classify RCH in this study. Accordingly, 4 RCH types were categorized, with Type 1 the mildest and type 4 the most severe form of deformity.

The method introduced by Manske et al. was used to assess forearm angles (9). Based on this method, hand-forearm angle (HFA) is described as the angle between longitudinal axis of the third metacarpal bone and longitudinal axis of the ulna. In this setting, the longitudinal axis of the ulna would be a line perpendicular to the distal physis of the ulna (Fig.1A). Hand-forearm position (HFP) could be easily calculated, accordingly (Fig.1B). In addition, ulnar bow (UB) would be regarded as the angle between the longitudinal axis of proximal and distal ulna (Fig.1C).

**Surgical technique**

The centralization approach was performed to correct the RCH (Fig.2). The incisions were different, as the surgeries were performed by 3 different hand surgeons. However, the rest of the operations were done in accord-



Fig. 1. Assessment of forearm angles based on Manske method; (A) Hand-forearm angle; (B) Hand-forearm position; (C) Ulnar bow

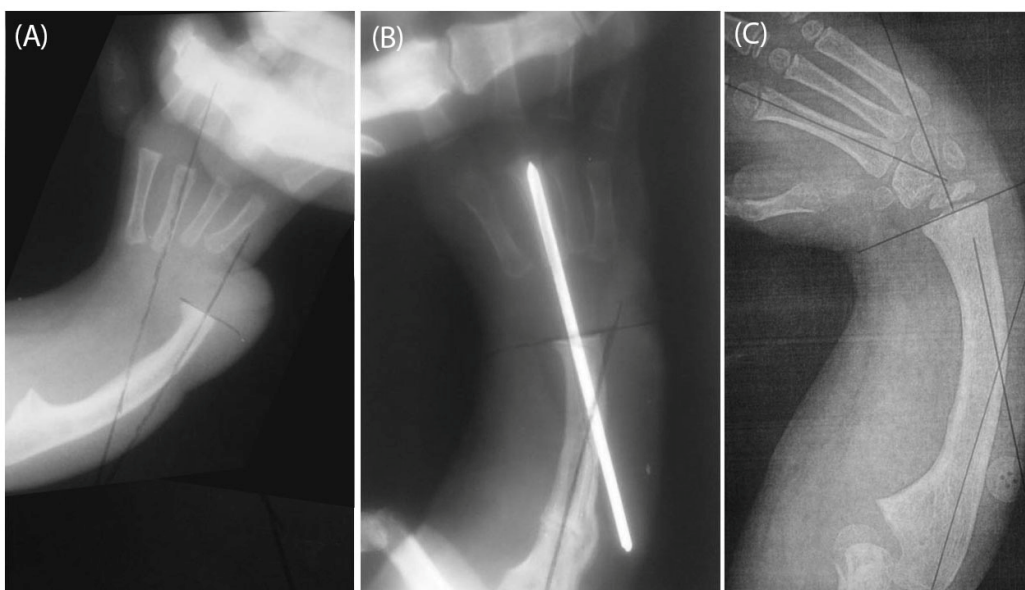


Fig. 2. Radiographs of a type IV RCH; (A) Preoperative radiograph; (B) Radiograph of immediately after centralization surgery; (periosteal reaction of the ulna was due to the sequel of the direct trauma not from ulnar osteotomy); (C) Radiograph of 89 months after the surgery (Polycization was performed for this patient after centralization)(case number 5).

ance with the standard centralization technique, which was described by Manske et al. previously (9). Briefly, the distal ulna was located at the middle of carpus, without disturbing the physis of the ulna. Reefing or advancement of extensor carpi ulnaris was part of the procedure, and tendon transfer was performed for 3 patients (case numbers 2, 8, and 10), which transferred the flexor and extensor carpi radial tendons to the reefed extensor carpi ulnaris one. Longitudinal pin was used to maintain correction following the surgery. The limb was immobilized in long-term cast for 6 to 8 weeks. The pin was remained at its place for 8 to 10 weeks. The hand and forearm were placed in appropriate splint for 8 to 12 weeks after the removal of the pin, considering the patient's compliance. Osteotomy of the ulna was performed for only 1 patient (case number 8) and pollicization for 6 patients.

HFA, HFP, and UB were calculated before the operation, immediately after the operation, and at the last follow-up session. Wrist passive range of motion was assessed by a goniometer and a fellowship trained hand surgeon, who was not involved in any of the operations. At the end, we evaluated the level of satisfaction from the surgery by asking the following question from the patient's parents: Are you pleased from the operation and do you recommend it to others? Values of 1, 0.5, and 0 were assigned whenever both, one, or none of the parents were satisfied, respectively.

### Statistical analysis

Central tendency and variability were evaluated using mean and standard deviation (SD), respectively. The Pearson's correlation coefficient test was used to evaluate potential correlations. Statistical analysis was performed using IBM SPSS for windows, Version 16. A p value of less than 0.05 was considered as statistically significant.

### Results

The demographic, clinical, and radiological characteristics of the patients are demonstrated in Table 1.

The mean preoperative, postoperative and last follow-up of HFA was  $49.5^{\circ} \pm 26.2^{\circ}$ ,  $6.8^{\circ} \pm 5.6^{\circ}$ , and  $20.1^{\circ} \pm 13^{\circ}$ , re-

spectively. While the mean HFA showed  $42.7^{\circ} \pm 22.3^{\circ}$  correction immediately after the surgery, this correction value was decreased by  $13.3^{\circ} \pm 13.7^{\circ}$  at the last follow-up. In conclusion, the final correction was  $29.4^{\circ} \pm 23.9^{\circ}$ .

The mean preoperative, postoperative, and last follow-up HFP was  $9.4 \pm 5.4$  mm,  $-5.4 \pm 4.1$  mm, and  $-4 \pm 3.6$ , respectively. In spite of a record of  $14.8 \pm 6.7$  mm mean correction of HFP immediately after the surgery, the mean correction revealed  $1.4 \pm 2.8$  mm recurrence at the last follow-up session. Consequently, the final mean correction of HFP was  $13.4 \pm 7.3$  mm.

The mean preoperative, postoperative and last follow-up UB was  $39.5^{\circ} \pm 16.7^{\circ}$ ,  $31.9^{\circ} \pm 14.3^{\circ}$ , and  $28.3^{\circ} \pm 7.8^{\circ}$ , respectively. The mean UB showed  $7.6^{\circ} \pm 12.5^{\circ}$  correction immediately after surgery and a further  $3.6^{\circ} \pm 7.3^{\circ}$  at the last follow-up. Overall mean correction of UB was  $11.2^{\circ} \pm 17.6$ .

No significant correlation was observed between the recurrence rate of HFA and other clinicopathologic characteristics of the patients such as preoperative HFA ( $p=0.37$ ), age at operation ( $p=0.33$ ), gender (0.2) and type of deformity ( $p=0.7$ ).

No significant correlation was observed between the ROM and preoperative HFA ( $p=0.33$ ) and HFP ( $p=0.67$ ).

The mean satisfaction level, provided by the patient's parent (13 cases), was 12 out of a total value of 13.

### Discussion

Although RCH was first described in 1733 (4), managing this deformity still remains a challenge in reconstructive hand surgery. Treatment focuses on the construction of a stable centralized and functional hand, preservation of a stable and mobile wrist, and maintenance of a longitudinal forearm growth (10). Evaluation of the different aspects of the existing approaches might result in valuable information, leading to the optimization of the available methods. Thus, we evaluated the outcome of patients after centralization surgery of RCH, mainly the recurrence of deformity, as the most important complication of this surgical method.

The mean age of our patients was 19.4 months at the

Table 1. The demographic, Clinical, and radiological characteristics of the congenital RCH patients

ID	Age* (month)	Gender	Type	Limb	Follow-up (month)	HFA		HFP			UB			ROM	
						pre	post	pre	post	FU	pre	post	FU		
1	18	male	IV	left	64	50°	12°	24°	12 mm	-2 mm	0 mm	60°	60°	40°	30°
2	25	male	IV	right	52	66°	8°	22°	10 mm	-5 mm	-10 mm	34°	34°	24°	35°
3	29	male	IV	right	68	50°	0°	50°	4 mm	-15 mm	0 mm	48°	48°	28°	30°
4	9	male	IV	right	98	17°	10°	16°	5 mm	-1 mm	-3 mm	14°	14°	20°	25°
5	28	male	IV	Left	89	17°	0°	45°	3 mm	-7 mm	-3 mm	24°	24°	32°	30°
6	20	female	III	left	93	20°	0°	14°	6 mm	0 mm	-2 mm	38°	38°	30°	50°
7	16	male	IV	right	29	45°	10°	10°	14 mm	-2 mm	-5 mm	37°	37°	28°	55°
8	36	female	IV	right	12	75°	10°	21°	18 mm	-6 mm	-4 mm	50°	20°	18°	15°
9	9	male	III	right	20	60°	5°	30°	16 mm	-7 mm	0 mm	33°	33°	22°	25°
10	29	male	IV	right	102	78°	12°	16°	19 mm	-10 mm	-10 mm	10°	10°	20°	60°
11	25	male	III	left	108	60°	8°	14°	12 mm	-2 mm	-1 mm	24°	24°	22°	50°
12	12	female	IV	right	19	15°	0°	5°	3 mm	-10 mm	-7 mm	45°	45°	30°	45
13	19	female	IV	left	12	22°	0°	5°	3 mm	-5 mm	-3 mm	28°	28°	38°	30
14	6	female	IV	left	80	100°	18°	5°	10 mm	-3 mm	-6 mm	45°	45°	25°	60
15	30	male	III	right	16	40°	5°	25°	10 mm	-10 mm	-10 mm	40°	40°	46°	50
16	10	male	IV	left	132	78°	12°	20°	6 mm	-2 mm	0 mm	10°	10°	30°	45

HFA = Hand-forearm angle; HFP= Hand-forearm position; UB = Ulnar bow; Pre= Preoperative; Post = postoperative; FU = Last follow-up; ROM = Range of motion; \* Age at the time of centralization

time of surgery, and the mean follow-up of the patients was 62.1 months. According to our results, the centralization surgery corrected the mean HFA 42.7° immediately after the surgery. However, a mean recurrence of 13.3° (26.8%) was observed at the last follow-up examination. In other words, the mean preoperative HFA of 49.5° turned into the mean value of 20.1° at the last follow-up, which equals a 59.4% correction.

The outcome of centralization surgery has been evaluated in other investigations as well. Damore et al. reported the results of 19 centralization surgeries in 14 patients, with a mean follow-up of 78 months. The mean age of their patients was 3.2 years. Based on their results, the mean preoperative HFA of 83° decreased to a mean HFA of 25° immediately after the surgery, while it recurred 38° (45.7%) at the last follow-up. Accordingly, the final HFA correction of their study was 20° (24%). Damore's study also revealed a significant correlation between the age of the patient at the first surgery and recurrence rate (4). Consequently, the higher recurrence rate of their study could be correlated to the higher age of their patients. Our study did not indicate a significant correlation between the age and recurrence rate of the patients.

The higher recurrence rate of their study could also be attributed to the longer follow-up period of their study. Since the recurrence of radial deviation starts immediately after the operation, it could be concluded that higher follow-up period of patients might result in higher recurrence rate. However, this assumption needs to be examined in future investigations.

Farzan et al. assessed the outcome of centralization surgery in 12 forearms of 10 patients with severe congenital club hand. The mean age of their patients was 16.8 months, and the mean follow-up of patients was 48 months. The mean preoperative angular deviation of their patients was 100°, while it was 19.58° at the latest follow-up (80.4 %). They did not record the angular deviation immediately after the operation, and consequently the recurrence of deviation was not reported in their study (11). The lower recurrence rate of their study could be associated with the lower age of the patients or the lower follow-up period.

Shariatzadeh et al. evaluated the recurrence rate of radial deviation in 11 forearms of 9 patients with RCH deformity managed by centralization surgery. The mean age of their patients was 17 months, and the mean follow-up of their study was 90 months. Preoperative HFA of their study was 75°, which turned into 25° immediately after the operation, and 52° at the last follow-up. Accordingly, the recurrence rate of their study was 36%. Again, the high recurrence rate of their study could be attributed to their longer follow-up period (7).

Several other studies have also evaluated the outcome of centralization surgery of RCH and most of them reported acceptable results (12-15). However, considering the different characteristics of the patients of each study including different age, gender, and type of deformity the outcome of different studies might be incomparable. However, the high rate of the recurrence of radial deviation is noticeable in nearly all studies. In this respect, many ef-

forts have been made to optimize the results of centralization surgery of RCH or develop new techniques.

Buck-Gramcko proposed the radicalization method as a new technique with an emphasis on soft-tissue reconstruction and more ulnar deviation (16). In this technique, fibrotic tissues will be excised, the hand and radial carpal bones will be placed over the distal end of the ulna, and the hand will be fixed with a Kirschner wire in a position of moderate ulnar deviation. However, later investigations did not confirm the priority of radicalization over centralization (5, 15).

To avoid the extensive dissection and acute stretching of the neurovascular structures, Kessler in 1989 suggested preoperative soft-tissue distraction prior to the centralization surgery (17). Subsequently, Smith and Greene reported the successful results of a small series of preliminary soft-tissue distraction using an Orthofix external distracter in 4 patients (18). Precentralization soft tissue distraction is now advised to be performed in neglected late-presenting patients needing significant correction of wrist deformity or in patients with severe RCH (10).

Microvascular joint transplantation was also presented as a new concept for the correction of the congenitally deformed wrists. Vilkki reported the result of a series of 24 radial club hands corrected with this method. According to this study, the procedure can provide a period of nearly 10 years for the affected children with no need for surgical intervention. Besides, usually no splinting is needed during that period. However, an additional corrective osteotomy might be required at early adolescence considering the growth properties of the joint graft and original ulna. The whole procedure is quite demanding and it is recommended to be performed in dedicated microsurgical departments with adequate experience of pediatric tissue transfer (19).

The high recurrence rate of deformity following the centralization and radicalization surgery, led to the development of a new technique called 'ulnarization', which is performed through a volar approach in a vascular and physal sparing fashion. This method dorsally transfers the flexor carpi ulnaris from a deforming to a corrective force, which in turn balances the muscle forces on the wrist biomechanically. Ulnarization is the first treatment of RCH, which has demonstrated no recurrence or growth arrest. Although this method has its drawbacks such as overgrowth of the distal ulna relative to the carpus and excessive ulnar deviation, the promising results of this method might suggest substitution of this method with the traditional techniques in the near future (20).

Our study had some limitations. The small number of patients could be regarded as the main limitation of this study, which prohibited further statistical analysis with higher statistical power. In addition, the surgeries were done by different surgeons and the results of the study could have been affected by intersurgeon variability.

## Conclusion

Our study revealed an acceptable long-term result following the centralization surgery of RCH. However, the high risk of recurrence of radial deviation needs a resolu-

tion in future investigations. This could be obtained by developing new techniques of RCH surgery or modifying the available techniques.

### Conflict of Interests

The authors declare that they have no competing interests.

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