

CONGENITAL RADIOULNAR SYNOSTOSIS, A REPORT OF 11 CASES AND REVIEW OF LITERATURE

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Abstract- During the last 10 years, we have had 11 cases of radioulnar(RU) synostosis, a very rare congenital anomaly of the upper extremity. Only 3 of them required surgical intervention. So we evaluated these three cases of proximal radioulnar synostosis corrected by proximal derotational osteotomy. The indication for surgery was severe pronation deformity that caused functional problem. Mean age at the time of surgery was 4.5 years (3-6 years) and mean postoperative follow up was 3.5 years (1-5 years). Forearm position after surgery was 15° supination. Anesthetic and functional results in all patients were good. In only one patient it was complicated by impending compartment syndrome that was treated by conservative measures. We recommend derotational osteotomy for correcting RU synostosis in earlier age. *Acta Medica Iranica: 40(2): 126-131; 2002*

Key Words: Synostosis, congenital, radioulnar

INTRODUCTION

Congenital proximal radioulnar synostosis can be severely disabling, especially if it is bilateral or if severe hyperpronation exists. Functionally, patients with severe deformity have trouble in getting a cup to the mouth, using eating utensils, or accepting objects in an open palm (1). Of 11 patients with radioulnar synostosis that we found during the last ten years in Imam Khomeini hospital, only 3 patients met the criteria for operation. One of the patients had both proximal and distal synostosis, a feature that has not been reported in English language literature so far.

The mean age at presentation was 4.6 years for operated patients (3-6 years) and 17.5 years for non-operated patients (2.5-47 years). The most common complaint of patients was limitation of forearm rotation. We had 6 female and 4 male patients. In 5 of our patients, we couldn't find any associated anomaly. In 5 patients, the radioulnar synostosis was as a part of a syndrome, multiple synostosis syndrome in 4 patients and Poland's syndrome in the

other one.

We operated through a posterior approach and after osteotomy of ulna distal to fusion mass, we put an intramedullary pin in the ulna and then derotated the limb to the desired position. Tourniquet was released and the vascular status of limb checked, and if acceptable the second pin was inserted to maintain the corrected position. This pin was placed in to facilitate its removal if there was any compromise in circulation postoperatively. The mean follow up period of operated patients was 3.5 years (1-6 years). All patients were satisfied with their extremity and their functions improved considerably. The mean postoperative position of operated limb was 13.3 degree of supination (10 to 15 degrees).

The results of three operated cases are summarized in table 2.

Of eight patients that were not candidates for operative treatment, four were members of a family with multiple synostosis syndromes. Another patient was an 18-year-old man who worked as a mechanic without any functional problem. We also had a 5-year-old girl and a 2.5-year-old boy with bilateral disease and no functional problem. We had a 4-year-old girl with double synostosis in one forearm, a feature that was not reported in literature so far.

DISCUSSION

Congenital radioulnar synostosis, a deformity characterized by a fixed position of the forearm ranging from neutral rotation to maximum pronation, is a rare congenital anomaly (1-5). Only 350 cases have been reported in literature until now (3). The condition can be disabling especially when it occurs bilaterally or if there is severe hyperpronation (1,2).

Congenital radioulnar synostosis may be presented as an isolated anomaly without any associated features as in 6 of the 11 patients. It may be presented as a component of multiple congenital deformity (6-9). We had a patient with Poland's syndrome and 4 patients with multiple synostosis syndromes.

Wilkie classified the synostosis roentgenographically into tow types: type I, there is lack of proximal of radius, bony fusion for 3-6 cm, and medullary canal of radius and ulna is connected

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together. In type II, there is a normal radius and the synostosis is located just distal to proximal radial

epiphysis, so the radial head is dislocated anteriorly or posteriorly (9).

Table 1. Clinical data of the eleven patients

	Involved side	Age at operation/presentation	Sex of patient	Position of ankylosis	Preoperative elbow ROM/deformity	Shortening of ipsilateral extremity	Wrist hypermobility	Wilkie classification
Case1	L	6	F	120°Pron.	Full	+ive	+ive	I
Case2	R	5	F	110°Pron	10° flexion contracture	+ive	+ive	I
Case3	L	3	M	110°Pron	Full	+ive	+ive	I
Case4	R	5	F	8° Pron	Full	+ive	+ive	I
Case4	L			5° Pron				II
Case5	L	18	M	10 ° Pron	Full	+ive	+ive	I
Case5	R			5° Pron				I
Case 6	R	4	F	Neutral	Full	+ive	-ive	II
Case 7	R	2.5	M	5° Pron	Full	----	+ive	I
Case 7	L			10° Pron			+ive	I
Case8	R	45	M	Neutral	80°-90°	----	----	II
Case8	L			Neutral	30°-100°			II
Case9	R	24	M	Neutral	75°-90°	----	----	II
Case9	L			Neutral	45°-130°			II
Case 10	R	23	F	Neutral	Fixed in 30°	----	----	I
Case 10	L			Neutral	Fixed in 30°			I
Case 11	R	19	F	Neutral	Fixed in 60°	----	----	I
Case 11	L			Neutral	Fixed in 45°			I



Fig. 1. Pre operative X-ray of case 1. A- AP, B- Lat.

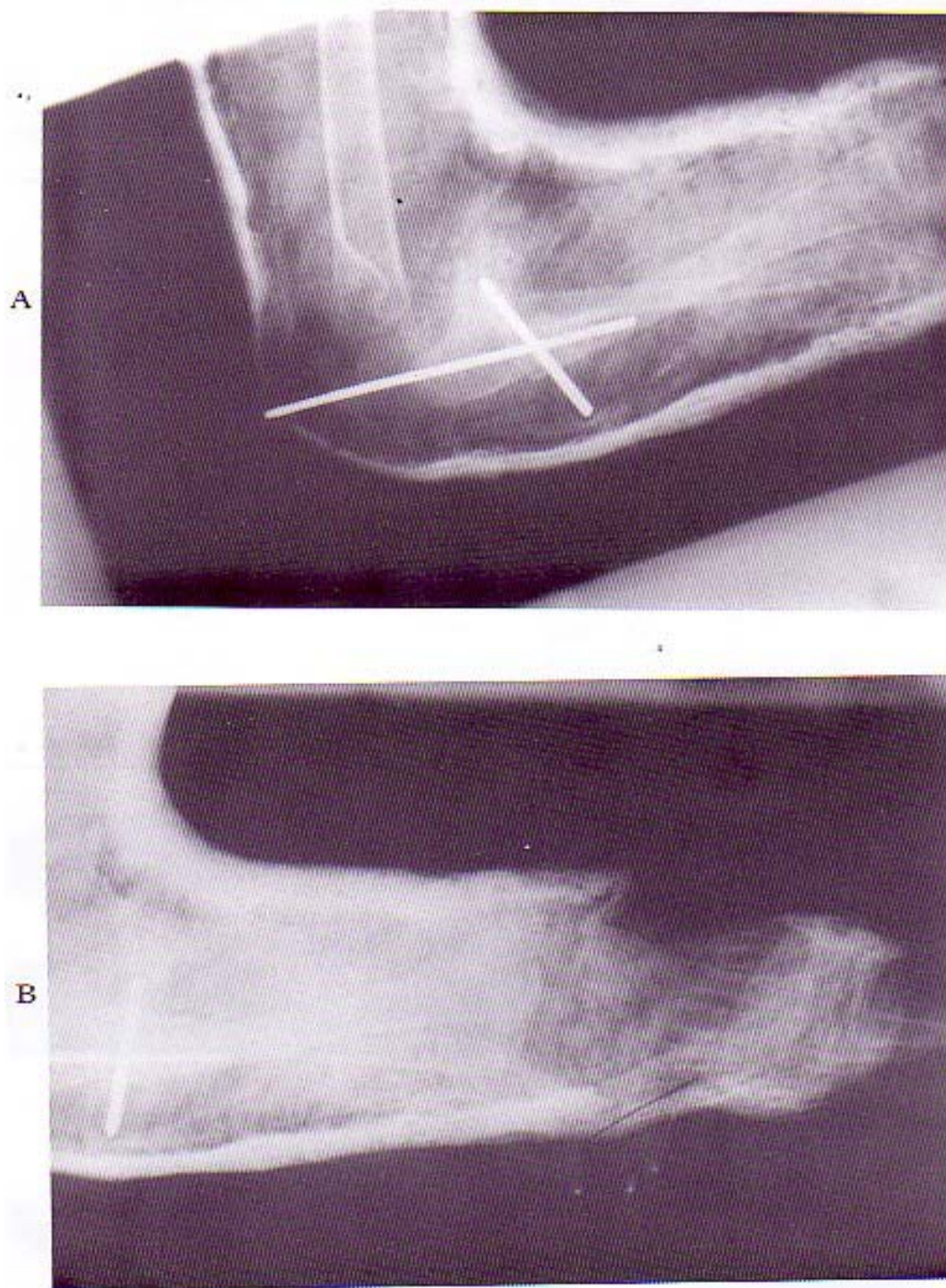


Fig 2- Postoperative X-rays, case 1.



Fig 3. Double synostosis

Table 2. Postoperative data

	Duration of F/U	Elbow ROM	Final position of forearm	Aesthetic improvement	Functional improvement
Case 1	5 years	Full 10° flexion	10° supination	Good	Good
Case 2	4 years	contracture	15° supination	Good	Good
Case 3	1 year	Full	15° supination	Good	Good

Cleary and Omer described four types: in type 1, there is lack of involvement of the bone and the radial head is located; in type 2, there is a visible synostosis with normal radius; in type 3, there is an osseous synostosis with hypoplastic and posteriorly dislocated radial head; and in type 4, there is a short osseous synostosis with an anteriorly dislocated radial head (2,10). The main indications for surgery are hyperpronation and bilateral synostosis, in both of them disability must be very severe (1,2,4,5,11-13). The severity of hyperpronation that requires surgery is controversial but in our opinion it must be determined on individual basis and the major concern is the function of the extremity. In authors' opinion, patients with congenital radioulnar synostosis who haven't severe deformity and functional limitation need no operative treatment. Recently, some authors present new procedures for mobilization of the forearm, but long-term results have not been presented.

Several operative procedures have been suggested for congenital radioulnar synostosis, that can be categorized in 2 major groups: 1) operations that were designed to restore the rotational motion of forearm (i.e., supination and pronation) in addition to removal of synostosis, 2) operations that improve the fixed position of forearm in a more functional position.

Several authors have reported separation of the synostosis and interposition of fat or muscle or silicone, but their results have not been satisfactory and recurrence of the ankylosis has been the major complication (2,3,14,15). Hansen and Andersen performed a partial resection of the left radius in a sixteen-year-old girl. Eighteen month postoperatively, osseous contact was noted roentgenographically (14). Miura et al. operated eight upper extremities in seven patients. They placed anconeus between the separated radius and ulna, but synostosis recurred in all the patient (15). Kelikian and Doumanian reported good results with use of a swivel prosthesis in patients who had post-traumatic

proximal radioulnar synostosis; however, Tachdjian noted disappointing results with the swivel prosthesis in patients who had congenital synostosis, with recurrence of the ankylosis at eighteen-month follow-up examination (16,17). Kanaya reported that separation of a congenital radioulnar synostosis with a vascularized fascio-fat graft and osteotomy of the radius could achieve pronation and supination of the forearm (2).

Currently, osteotomy to achieve more functional position is accepted for the management of patients who have severe pronation (1,2,5,11,12,18-20). The ideal position depends on the type of involvement (bilateral or unilateral, dominant or nondominant site), the social and cultural environment of the patient, and projected future activities (19). Because it is not possible to predict patient's future activities, it is impossible to determine the optimum position of the forearm (2). Simmon's and Green's recommendations for optimum position are summarized in table 3. All of our operated cases were unilateral and we preferred to bring their forearm to 15-degree supination after osteotomy.

The preferred site for derotational osteotomy is distal of fusion mass, osteotomies distal to fusion mass tend to have greater soft tissue restrictions (1, 14-15,18-19). Gradual correction using an Ilizarov frame decreases the risk of neurovascular compromise and allows the patient to select the most functional position (21). Using a small external fixation device allows precise rotational correction and affords adequate stabilization yet avoids cast immobilization (4). We agree with Green and Mital that a longitudinal K-wire in ulna and a transverse K-wire through the distal fragment that is incorporated in the cast is good for one stage operation (18).

Some authors believe that operation is best to carry out between the ages of 4 and 10 years (5), but we recommend the operation between the ages 5 and 7 years. The radial head should have been appeared and the earlier age has less neurovascular complications.

Table 3. Recommended positions after osteotomy

Author	Unilateral	Bilateral	
		Dominant	Nondominant
Simmons	Neutral to 20° pron.	10-20°pron.	Neutral
Green	10-20° supination	30-45°pron.	20-35°sup.

Complications of derotational osteotomy are vascular compromise, compartment syndrome, loss of derotation, decreased elbow ROM, which are very rare. We have only one case of impending compartment that got relieved with conservative treatment.

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