Acta Orthop. Belg., 2007, **73**, 704-709

ORIGINAL STUDY

Two-stage double-level rotational osteotomy in the treatment of congenital radioulnar synostosis

Wael EL-ADL

From Mansoura Faculty of Medicine, Mansoura University, Mansoura, Egypt

Congenital proximal radioulnar synostosis is a rare congenital anomaly that can be extremely disabling, especially when it occurs bilaterally or if there is severe hyperpronation. Currently, osteotomy to achieve a neutral or slightly pronated position is widely accepted for the management of patients who have severe pronation. The present study evaluates the result of two-stage double-level rotational osteotomy of both the radius and ulna in the treatment of severe congenital radioulnar synostosis. Nine children with severe congenital radioulnar synostosis underwent two-stage double-level rotational osteotomy of both the radius and ulna at Mansoura University Hospital. There were seven boys and two girls with a mean age of 5.6 years who were followed up for a mean of 26 months. The position of the forearm was improved from a mean pronation deformity of 76° (60° to 85°) to 30° of pronation in the affected dominant extremities and 20° of supination in non-dominant extremities in all cases. Bony union was achieved by 5.9 weeks with no loss of correction. The advantages of this technique are that it is easy, safe, with absence of severe postoperative complications and requires a small surgical scar. A drawback of the technique is that the rotation correction depends only on a cast, so that a correction loss might occur if the plaster cast loosens.

Keywords : congenital radioulnar synostosis ; diaphysis of the radius ; rotation osteotomy.

INTRODUCTION

Congenital radioulnar synostosis is a rare anomaly when failure of segmentation of the radius and ulna results in a fixed position of the forearm ranging from neutral rotation to severe pronation (*1,3*). When the deformity is mild, little disability is evident, as the ipsilateral shoulder and wrist can compensate effectively (*1,17*), but with significant pronation the activities of daily living, including dressing, eating, washing and accepting objects in the palm of the hand, can be severely impaired (*1,8,20*).

Two major surgical procedures are available for this disease. One is the mobilisation operation to separate the radioulnar synostosis and restore forearm rotation (*5,9*). The mobilisation operation is thought to be the best method theoretically ; however, it requires a vascularised fat graft after release of the synostosis and it is often difficult to realign the radial head in the proper position in severe cases, in which the radial head shows posterior or anterior dislocation (*5*). More recently, osteotomies distal to the site of the synostosis to fix the forearm in a functional position have been advocated (*16*).

■ Wael Ali Maher Mohammed El-Adl, MD, Lecturer of Orthopaedic Surgery.

Mansoura Faculty of Medicine, Mansoura University, Mansoura, Egypt.

Correspondence : Wael Ali Maher Mohammed El-Adl, 1 Ali Maher Street from Gihan Street, Mansoura, Egypt.

E-mail : waelalimaher@gmail.com

No benefits or funds were received in support of this study

Table I. — Details of patients

Number	Age (years, months)	Sex	Side	Follow-up (months)	Preoperative fixed pronation deformity		Postoperative fixed pronation deformity	
					Right	Left	Right	Left
1	4.7	Boy	Right	38	70°	Normal	20°	Normal
2	6.8	Girl	Bilateral	37	65°	80°	20°	25° of supination
3	7.5	Boy	Right	33	80°	Normal	25°	Normal
4	5.4	Girl	Right	30	80°	Normal	30°	Normal
5	3.9	Boy	Right	27	70°	Normal	25°	Normal
6	8.3	Boy	Bilateral	25	85°	80°	25°	30° of supination
7	4.8	Boy	Left	20	Normal	85°	Normal	25° of supination
8	5.2	Boy	Right	15	75°	Normal	30°	Normal
9	4.2	Boy	Right	13	70°	Normal	30°	Normal

As derotation then takes place over a larger area, this reduces the risk of secondary loss of correction and vascular complications (*6,16,20*).

The present study evaluates the result of two-stage double-level rotational osteotomy of both the radius and ulna in the treatment of severe congenital radioulnar synostosis.

PATIENTS AND METHODS

During the period 2003-2006, nine children underwent a prospective study for correction of severe congenital radioulnar synostosis with two-stage, double levels rotational osteotomy of both the radius and ulna at Mansoura University Hospital. There were seven boys and two girls. Their ages at operation ranged from 3 years and 9 months to 8 years and 3 months (mean : 5 years and 6 months). The right forearm was involved in six children and the left in one. There was bilateral involvement in two. All were right handed (table I).

The mean preoperative range of motion of the elbow joint was from 2.6° extension (range : 2 to 4) to 133.8° flexion (range : 130 to 140). The mean preoperative pronation deformity was 76° (range : 60 to 85) (table II). It was measured with the patient's elbow held fixed to the side of the chest, the forearm at 90° and the angle between the longitudinal axis of the humerus and the line of the radial and ulnar styloid processes was measured with a goniometer, as described by Ogino and Hikino (*17*). Nine forearms were classified as type III according to Cleary and Omer (*1*) (table II) with visible osseous synostosis associated with posterior dislocation of a hypoplastic radial head, while two forearms were of type II, with visible osseous synostosis and a normal reduced radial head.

Table II. — Cleary and Omar (*1*) classification for congenital radioulnar synostosis

Types	**Criteria**
Type 1	there is a lack of involvement of the bone, and the radial head is located and normal
Type 2	there is a visible osseous synostosis with a normal radius
Type 3	there is an osseous synostosis with a hypoplastic and posteriorly dislocated radial head
Type 4	there is a short osseous synostosis with an anteriorly dislocated radial head

Operative technique

The child is supine under general anaesthesia, with a well-padded tourniquet. The ulnar osteotomy is carried out distal to the site of the synostosis via a subcutaneous subperiosteal posterior approach. The radial osteotomy is undertaken at the distal diaphyseal-metaphyseal junction through an incision over the dorsolateral ridge of the distal third of the radius. The bone is exposed subperiosteally and the osteotomy site is marked with several fine drill holes that penetrate both cortices. The division of the radius and then the ulna is completed using a sharp osteotome. At this point, no attempt should be made to change the position of the arm. The tourniquet is deflated and haemostasis is achieved. The wounds are irrigated and closed with subcuticular sutures. A long arm plaster cast is placed over sterile dressings.

Fig. 1. — A girl aged 5 years and 4 months with congenital radioulnar synostosis of the right forearm and had fixed pronation deformity of 80°.

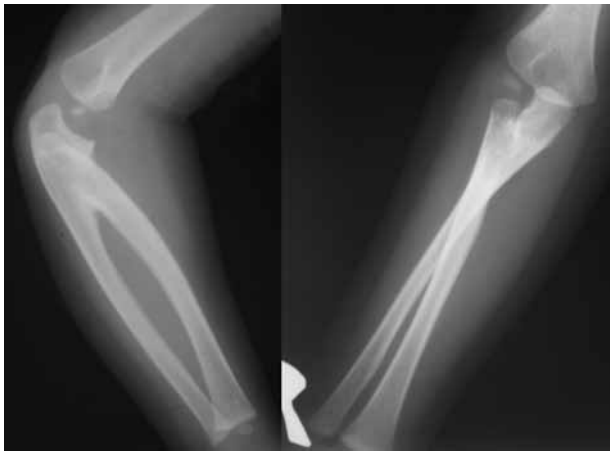

Fig. 2. — Antero-posterior and lateral radiograph of the child showing type 3 congenital radioulnar synostosis with an osseous synostosis and a hypoplastic and posteriorly dislocated radial head.

Ten days later, children are re-admitted, the cast is removed with the patient under general anaesthesia and the forearm is supinated or pronated into the desired position. Anteroposterior and lateral roentgenograms confirm bony apposition and alignment. Generally, affected dominant extremities should be placed in 20 to 30° of pronation and nondominant extremities in 20° of supination. Pulses are checked carefully after manipulation and the extremity is closely monitored to detect signs of compartment syndrome. An above-elbow plaster cast is applied with the elbow flexed to 90°, and children are reviewed every three weeks until radiographs

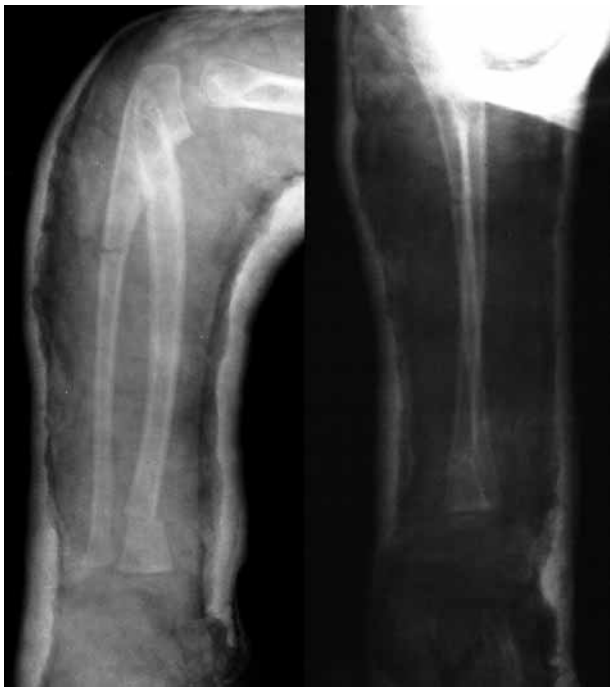

Fig. 3. — Immediate postoperative radiograph with double-level osteotomy of both the radius and ulna.

show bony union, then the cast is removed. If healing is delayed, the cast is retained until union is achieved. The patients are followed up at six-monthly intervals, and the position of rotation of the forearm is recorded.

RESULTS

The average duration of follow-up was 26.4 months (range : 13 to 38). The mean correction achieved at surgery was 58.6° (range : 35 to 110) to a final position of 20 to 30° of pronation in the affected dominant extremities and 20° of supination in the nondominant extremities. Bone union was achieved in all patients, with average duration of 5.9 weeks (range : 5 to 7).

The time for complete removal of the cast ranged from 6 to 9 weeks (mean : 8 weeks). In three children, with pronation deformities in the affected nondominant side, marked dorsal angulation of the radial osteotomy was seen after derotation to the supinated position. However, remodelling to nearly normal had occurred by an average of 18 months.

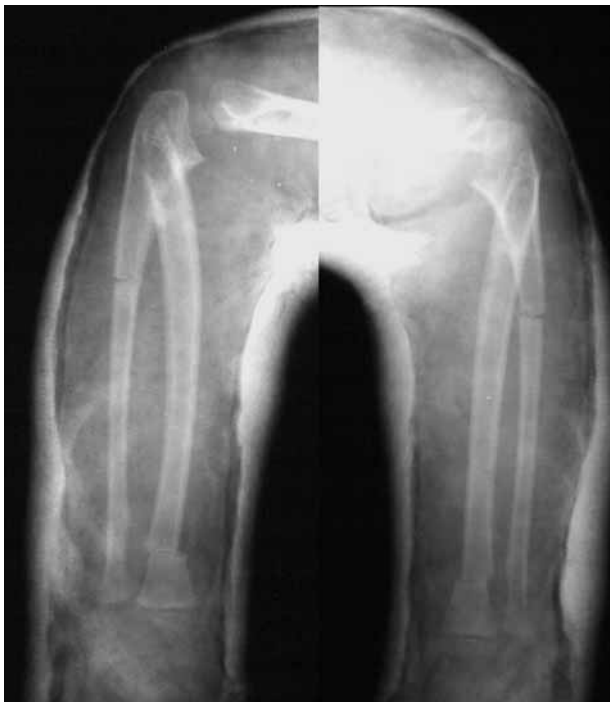

Fig. 4. — 10 days postoperative radiograph after derotation of the osteotomy was done.

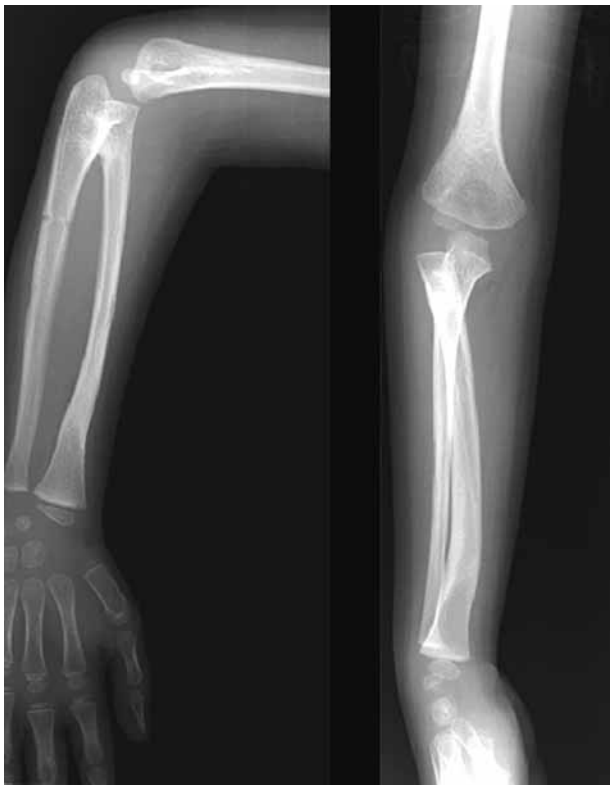

Fig. 5. — Follow-up radiograph after 30 months with complete union and remodelling of the osteotomy.

At latest follow-up there was no loss of correction in any child. There were no complications and no circulatory disturbances, neuropathies, or hypertrophic scars on the forearm. Flexion and extension of the elbow and movements of the wrist were unaffected by the operation. All children showed marked functional improvement and they and their families were satisfied with the results.

DISCUSSION

Congenital proximal radioulnar synostosis, a deformity characterised by a fixed position of the forearm ranging from neutral rotation to maximum pronation, is a rare congenital anomaly (*9*). According to Wilkie (*22*), the first case of congenital radioulnar synostosis was described in 1793 by Sandifort in Museum Anatomicum. It is thought to be caused by a failure of longitudinal segmentation with persistence of the cartilaginous anlage between the radius and ulna during the seventh week of development. The resultant bridge may be fibrous or bony (*1,14*). The condition can be extremely disabling, especially when it occurs bilaterally or if there is severe hyperpronation, as occurs in 50% to 80% of cases. Children who have a severe deformity have trouble bringing a cup to the mouth or accepting objects into an open palm (*20*).

The aetiology is unknown but a genetic basis has been suggested through the association with a positive family history and disorders such as the Apert and Klinefelter syndromes (*3,20*). Skeletal anomalies associated with congenital radioulnar synostosis include talipes equinovarus, an absent thumb, coalescence of the carpal bones, symphalangism and bowing of the radial shaft (*20*). However, in this study, there were no cases with associated skeletal anomalies.

Many authors consider the synostosis as either type 1, a true bony fusion in which the radius and ulna are smoothly joined proximally for a variable distance, or type 2 in which there is congenital dislocation of the radial head with the synostosis just

Fig. 6. — Clinical follow-up after 30 months with fixed pronation deformity of 30° ; the child is able to use her hand.

distal to the proximal radial epiphysis (*8,14,22*). Cleary and Omer (*1*) described four radiographic types : in type 1, there is a lack of involvement of the bone and the radial head is located and normal ; in type 2, there is a visible osseous synostosis with a normal radius ; in type 3, there is an osseous synostosis with a hypoplastic and posteriorly dislocated radial head ; and in type 4, there is a short osseous synostosis with an anteriorly dislocated radial head. According to the classification system of Cleary and Omer, nine of our children had a type-3 and two, a type-2 synostosis. However, because there is little functional difference between the radiographic types (*1*) and the classification changes with time, these systems may have little clinical value (*15*).

The need for operation depends on the severity of the deformity and disability. Simmons *et al* (*20*) found that pronation of 60° was a definite indication for osteotomy, while pronation of 15° to 60° was a relative indication based on the needs of the individual. Ogino and Hikino (*17*) considered that the mean pronation of patients who complained of disability was 60° and of patients without complaints 20°. Surgery is usually tailored to individual needs. In this study, all children had a significant disability, with a mean pronation deformity of 76°.

Various techniques have been used to achieve rotation of the forearm (*6*). Several authors have reported separation of the synostosis and interposition of fat or muscle (or some other material), but recurrence of the ankylosis has still been noted (*2,8,15,19*). Hansen and Andersen (*8*) performed a partial resection of the left radius in a sixteen-year-old girl. Eighteen months postoperatively, osseous contact was noted roentgenographically. Miura *et al* (*15*) operated on eight upper extremities in seven patients. They placed the anconeus between the separated radius and ulna, but the synostosis recurred in every patient. Kelikian and Doumanian (*11*) reported good results with use of a swivel prosthesis in patients who had a post-traumatic proximal radioulnar synostosis ; however, Tachdjian (*21*) noted disappointing results with the swivel prosthesis in patients who had a congenital synostosis, with recurrence of the ankylosis at the eighteen-month follow-up examination. Simmons *et al* (*20*) reported eight complications in association with twenty-two osteotomies ; the complications included a wound infection (one), loss of correction (three), and circulatory problems (four). Green and Mital (*6*) reported one ischaemic contracture in thirteen patients.

Osteotomies to position the forearm in a more functional position are an alternative to resection of the synostosis. Three types of osteotomy procedures have been used to correct forearm rotation : osteotomy at the synostosis (*12*), osteotomy at two sites in the diaphysis of the radius and the ulna (*10,13,16*), and osteotomy at one site in the distal diaphysis of the radius (*4*). The rotational osteotomy at the synostosis involves operative complexity, and some postoperative complications have been reported, including vascular compromise, such as Volkmann's ischaemia, shortening and angulation of the forearm, and posterior interosseous nerve palsy (*7,17,20*). In the osteotomy at two sites, the procedure is easier and there are fewer complications, although internal fixation is necessary, requiring a second surgery to remove the implant (*4*). In this study, all cases had a double level osteotomy followed by derotation with no need for internal fixation or another surgery for implant removal.

The best position of the forearm after derotation has not been established. Green and Mital (*6*) suggested that in bilateral cases the best position was in 30° to 45° of pronation in the dominant forearm

and in 20° to 35° of supination in the non-dominant. In unilateral cases, the ideal position was 10° to 20° of supination. Other authors have advocated 0° to 20° of supination in the non-dominant forearm and 0° to 20° of pronation in the dominant forearm (*13,16,17*). Ramachandran *et al* (*18*) prefer a position of 10° supination in all cases as compensatory movements at the shoulder and wrist allow the forearm to be located ideally for most daily activities. However, they found that hypermobility of the wrist was subjectively noted in all their patients.

The results of this study of congenital radioulnar synostosis treated by two-stage double level rotational osteotomy of both the radius and ulna were satisfactory. In all cases of this study, no patients reported disabilities in using forearms and hands in writing, eating, washing the face, and all children were pleased with the final position of their forearms. The advantages of this technique are that it is easy, safe, with absence of severe postoperative complications and a small surgical area and scar. A drawback of our technique is that the rotation correction depends only on a cast, so that a correction loss might occur if the plaster cast loosens.

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