Rotational osteotomies for congenital radioulnar synostosis

M. Ramachandran,
K. Lau,
D. H. A. Jones

*From Great Ormond
Street Hospital for
Children, London,
England*

In five children, six forearms with a fixed pronation deformity secondary to congenital radioulnar synostosis were treated by a derotation osteotomy of the distal radius and the midshaft of the ulna.

There were three boys and two girls with a mean age of 4.9 years (3.5 to 8.25) who were followed up for a mean of 29 months (18 to 43). The position of the forearm was improved from a mean pronation deformity of 68° (40° to 80°) to a pre-planned position of 10° of supination in all cases. Bony union was achieved by 6.3 weeks with no loss of correction. There was one major complication involving a distal radial osteotomy which required exploration for a possible compartment syndrome.

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.^{1,2} When the deformity is mild, little disability is evident, as the ipsilateral shoulder and wrist can compensate effectively,^{2,3} 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.^{2,4,5}

Attempts at resection of the synostosis in order to restore rotation of the forearm have, on the whole, been unsatisfactory, due to loss of correction and vascular compromise following extensive soft-tissue release and derotation.^{4,6,7} More recently, osteotomies distal to the site of the synostosis which fix the forearm in a functional position have been advocated.⁸ As derotation then takes place over a larger area, this procedure theoretically reduces the risk of secondary loss of correction and vascular complications.^{5,8,9}

We describe the surgical technique, results and complications of rotational osteotomies of the forearm with wire stabilisation of the ulna.

Patients and Methods

Five children with congenital proximal radioulnar synostosis were treated by the senior author (DHAJ) using rotational osteotomies of the radius and ulna with stabilisation of the ulna by an Ilizarov wire. The clinical details of the patients are shown in Table I.

There were three boys and two girls with a mean age of 4.9 years (3.5 to 8.5). They were followed up for a mean of 29 months (18 to 43). All were right handed. The mean pre-operative pronation deformity was 68° (40° to 80°). It was measured with the patient's elbow held fixed to the side of the chest, the forearm at 90° and a line through the styloid processes of the radius and ulna used in conjunction with a goniometer, as described by Ogino and Hikino.³ The left forearm was involved in two patients and the right in one. There was bilateral involvement in two.

One child (case 2 aged 4 years at operation) had staged bilateral procedures due to severe functional limitation secondary to bilateral fixed pronation of 80°. The patient with bilateral involvement (case 1) was aged 8 years 3 months at operation. The fixed pronation was less severe (40° bilaterally) and functional limitation was only present in his non-dominant forearm.

Five forearms were classified as type III according to Cleary and Omer,² with visible osseous synostosis associated with posterior dislocation of a hypoplastic radial head. One was of type II, with visible osseous synostosis and a normal reduced radial head. Case 1 had an associated duplication of chromosome 14. Case 3 had associated ipsilateral bowing of the radial shaft.

Operative technique. The patient is supine under general anaesthesia, with a well-padded tourniquet. The ulnar osteotomy is carried out in the midshaft using an oscillating saw via a

■ M. Ramachandran, FRCS(Orth), Specialist Registrar
■ K. Lau, PhD, Senior House Officer
■ D. H. A. Jones, FRCS, FRCS Ed(Orth), Consultant Paediatric Orthopaedic Surgeon
Orthopaedic Department
Great Ormond Street
Hospital for Children,
London WC1N 3JH, UK.

Correspondence should be sent to Mr M. Ramachandran at 5A Wallace Road, Canonbury, Islington, London N1 2PG, UK; e-mail: manoj@krigroup.com

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Table I. Details of patients

Patient number	Gender	Associated syndromes	Associated skeletal anomalies	Age at operation	Affected side	Dominance	Pre-operative position	Indication for operation (functional limitation as reported by patient and/or parents)
1	M	Duplication anomaly chromosome 14	None	8 years 3 months	Bilateral (left forearm operated on)	Right-handed	Fixed 40° pronation both forearms	Left forearm – absent supination, unable to catch ball, accept coins and small objects; unable to clean perineal area for hygiene
2	F	None	None	4 years	Bilateral (staged procedure)	Right-handed	Fixed 80° pronation both forearms	Both forearms – unable to hold plates and cups, play with toys, inability to accept small objects, difficulty dressing, difficult perineal hygiene
3	M	None	Bowing ipsilateral radial shaft	4 years 9 months	Left	Right-handed	Fixed 80° pronation	Left forearm – inability to accept small objects and coins, unable to hold plates and cups
4	M	None	None	3 years 6 months	Right	Right-handed	Fixed 60° pronation	Right forearm – inability to hold cups and plates, difficulty dressing
5	M	None	None	3 years 11 months	Left	Right-handed	Fixed 70° pronation	Left forearm – difficulty accepting coins, difficulty dressing

subcutaneous subperiosteal posterior approach. A 1.8-mm Ilizarov wire is passed retrogradely from the osteotomy to exit through the olecranon, and then antegradely across the osteotomy into the distal ulna. The radial osteotomy is undertaken at the distal diaphyseal-metaphyseal junction through a volar approach using an oscillating saw. The tourniquet is released and the forearm rotated to a position of 10° of supination. In all cases, adequate rotation was achieved without difficulty and satisfactory circulation maintained. The deep fascia of the forearm is incised proximally and distally at the osteotomy sites to allow for expansion of the muscle bellies. The wounds are closed in layers, the Ilizarov wire is bent and left proud of the skin. An above-elbow plaster cast is applied with the elbow flexed to 90° and the forearm in the corrected position. The limb is elevated post-operatively and the patient closely observed for neurovascular complications overnight. The cast is reinforced the next day and the patient discharged.

All patients were admitted three weeks after operation for removal of the wire under general anaesthesia and conversion to a below-elbow cast. A plain radiograph confirmed callus at the osteotomy sites and the patients were allowed to mobilise the elbow. They were reviewed after a further three weeks and if the radiographs showed bony union, the cast was removed. If healing was delayed, the cast was retained until union was achieved. The patients were followed up at six-monthly intervals, and the position of rotation of the forearm recorded.

Results

The mean correction achieved at surgery was 78° (50° to 90°) to a final position of 10° supination in all cases. Radiographic evidence of bony union was seen in five forearms at six weeks and in one at eight weeks. This delay in union was noted in the dominant right forearm in staged bilateral

operations. The dominant right forearm was corrected first and the attempted use of the dominant limb for activities of daily living may have accounted for the increased time to union.

At latest follow-up there was no loss of correction in any patient. In one child (case 4), marked dorsal angulation of the radial osteotomy seen at six weeks remodelled to nearly normal by 18 months (Fig. 1). Flexion and extension of the elbow and movements of the wrist were unaffected by the operation. All patients showed marked functional improvement and they and their families were satisfied with the results.

There was one major complication requiring re-operation. In case 3, a haematoma at the site of the radial osteotomy caused symptoms of a compartment syndrome. This presented as failure to achieve control of pain on the first post-operative day and required prompt surgical exploration. The haematoma extended proximally deep to the flexors, compressing them forward against the deep fascia. Flexor digitorum superficialis was indented by the edge of the fascia. The deep fascia was incised, relieving the constriction. The muscle bellies of the flexors were healthy. After 48 hours the fasciotomy wound was granulating satisfactorily and was left to heal by secondary intention. During follow-up, no neurovascular effects were noted and the scar was cosmetically acceptable.

Discussion

According to Wilkie,⁶ the first case of congenital radio-ulnar synostosis was described in 1793 by Sandifort in *Museum Anatomicus*. It is a rare anomaly, with approximately 350 cases recorded in the literature.¹⁰ 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

Fig. 1a

Fig. 1b

Fig. 1c

Fig. 1d

Serial radiographs of case 4 a) prior to operation, b) immediately post-operatively, c) at six weeks showing marked dorsal angulation and d) showing remodelling to nearly normal by 18 months.

resultant bridge may be fibrous or bony.^{2,11} The condition can lead to significant disability, especially if there is hyperpronation or when it is bilateral, as occurs in 50% to 80% of cases.^{4,9} In this series, the mean fixed pronation was 68° and two patients had bilateral involvement.

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.^{1,5} To our knowledge there have been no previous reports of congenital radioulnar synostosis and duplication

anomalies of chromosome 14 as in case 1. 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,⁵ which was the only associated skeletal anomaly seen in our series (case 3).

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 distal to the proximal radial epiphysis.^{4,6,11} Cleary and Omer² described four radiographic types: I, fibrous union with a normally-located radial head; II, osseous synostosis with a normal radius; III, osseous synostosis with posterior dislocation of a hypoplastic radial head; IV, a short osseous synostosis with an anterior dislocation of the radial head. They found, as we did, that type III was the most common and this has also been noted by others.^{8,12} Since there is little functional difference among the types described and the appearances may change with time, these classifications may have little clinical value.^{2,13}

The need for operation depends on the severity of the deformity and disability. Simmons et al⁵ 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 Hikino³ 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 our series, all patients had a significant disability, with a mean pronation deformity of 68°.

Various techniques have been used to achieve rotation of the forearm.⁹ The early results of operation at the site of the synostosis were disappointing. Even after radical resection of the synostosis with interposition of fascia, fat, muscle, or extraneous materials such as plexiglass, marked restriction of movement usually recurs.^{10,13} As derotation takes place over a narrow segment, tightness of the soft tissue may cause loss of correction, circulatory compromise, compartment syndrome and posterior interosseous nerve palsy.^{5,14} The early results using free vascularised fasciofat grafts after mobilisation of the synostosis with corrective osteotomy of the radius, appear more promising, with a reported increase in the arc of rotation to between 40° and 83° of pronation and supination, and no associated neurovascular complications.¹²

Kelikian and Doumanian¹⁵ described good results in four cases of post-traumatic radioulnar synostosis, using a cylindrical steel prosthesis in two of their four cases. However, Tachdjian¹⁶ noted disappointing results with the same prosthesis in patients with a congenital synostosis. Other methods of achieving derotation while minimising the risk of neurovascular complications include the Ilizarov technique,¹⁷ correction involving initial osteotomies followed by derotation ten days later^{18,19} and shortening of the forearm by resecting bone from the synostosis.³

Osteotomies to position the forearm in a more functional position are an alternative to resection of the synostosis. Murase et al⁸ described osteotomies in the distal third of the radius and the proximal third of the ulna, held by intramedullary K-wires, in four patients with a mean age of 3.9 years, all with forearms fixed in over 70° of pronation. They achieved a mean of 65° of correction, with the only complication being a 20° loss of correction during immobilisation in a cast. Bony union was seen at a mean of 7.5 weeks, when the wires were removed in clinic. No neurovascular complications were seen.

Our technique is similar in that the osteotomies are performed via simple surgical approaches. We used a 1.8-mm Ilizarov wire in the ulna. The displaced distal radial osteotomy does not require fixation and any deformity remodels well with time (case 4).

There was one major complication in our series when a combination of haematoma, bony displacement and inadequate release of the deep fascia mimicked a compartment syndrome (case 3). This patient also had ipsilateral bowing of the radius, which may have exacerbated the soft-tissue compromise following derotation even if adequate fascial release had been performed.

The best position of the forearm after derotation has not been established. Green and Mital⁹ 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.^{3,8,19} We 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. All our patients were pleased with the final position of their forearms. Although hypermobility of the wrist was subjectively noted in all our patients, it was not formally measured and so may have confounded the outcome. Objective measurements of rotation however were taken at the distal radius and ulna and therefore the hypermobility should not have affected our results.

This study was limited in being retrospective with data retrieved from a review of medical notes. We were unable to formally test pre- and post-operative function in our patients by objective measures, such as the hand-function test of Jebsen et al.²⁰

The principal advantages of this method are the ease of the surgical approach, no loss of rotational correction during follow-up and the need for stabilisation of the ulna only.

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