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**Amit Narang**

Senior Resident, Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

**Anand Gupta**

Post Graduate Student, Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

**Ayanjit Chattopadhyay**

Post Graduate Student, Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

**Amit Sharma**

Assistant Professor, Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

## Surgical outcome of both bone de-rotation osteotomy in congenital radioulnar synostosis: A case report and literature review

**Amit Narang, Anand Gupta, Ayanjit Chattopadhyay and Amit Sharma**

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### Abstract

Radioulnar synostosis is found as a congenital deformity in children or as a post-traumatic deformity in adolescents and adults. Congenital variety is more commonly located in the proximal part of the radius and ulna and restricts the range of motion of the forearm leading to some functional disabilities in the child such as using daily objects and carrying out basic hand to mouth activities like eating. The pronation deformity occurring as a result of this entity can be effectively dealt with single stage both bone forearm de-rotation osteotomy done at different levels in radius and ulna. We report our experience using this procedure in a 4-year old child with unilateral proximal congenital radioulnar synostosis in the non-dominant side. This procedure is relatively simple and we were able to achieve a good functional outcome at one year of follow-up with no associated complications.

**Keywords:** Radioulnar synostosis, de-rotation, osteotomy

### 1. Introduction

Congenital radio-ulnar synostosis is a rare phenomenon leading to loss of forearm range of motion and a fixed pronation deformity in children. The possible cause behind this phenomenon is the failure of in utero differentiation of radius and ulna. The most common location is at the proximal level where sometimes the radial head is completely fused with the proximal ulna or it may be completely absent or dislocated <sup>[1]</sup>. It has been reported that children with more than 60 degrees of pronation deformity have considerable difficulty in carrying out daily activities <sup>[2]</sup>. We report our experience of single stage both bone forearm de-rotation osteotomy in a 4 year old child with proximal congenital radioulnar synostosis.

### 2. Case report

A 4 years old child presented to us in the out patient department with complaints of inability to supinate the right forearm and an inappropriate position of the forearm while doing hand to mouth activities like eating. On clinical examination, the child had fixed pronation deformity of 80 degrees and could only take eatables to his mouth with excessive dorsiflexion of the wrist.



**Fig 1:** Clinical photograph of the patient taking a cookie from hand to mouth demonstrating excessive dorsiflexion at the wrist

**Corresponding Author:**

**Amit Narang**

Senior Resident, Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

Radiographic findings confirmed the diagnosis of proximal radioulnar synostosis with complete bony fusion of the radius and ulna at the proximal one third level.



**Fig 2:** Antero-posterior and lateral radiograph of the patient showing proximal radio-ulnar synostosis

Family history was negative for such type of deformity in any other relative. After a complete evaluation of the patient to rule out any syndromic association, surgical correction of the deformity with both bone forearm diaphyseal de-rotation osteotomy was planned.

**2.1 Surgical procedure:** With the patient in supine position, the forearm was carefully draped after complete aseptic preparation of the surgical site. We didn't use the tourniquet due to the fear of increased chances of compartment syndrome during reperfusion after fixation of de-rotation osteotomy. Also, it is mandatory to check the radial pulse once the forearm it rotated at the osteotomy site and applying a tourniquet can mask a possible vascular compromise intra-operatively. A volar incision was used for the radius and a transverse osteotomy was performed at the mid radius level. For ulna, dorsal incision was used and osteotomy was performed at a level proximal then the radial osteotomy. A careful de-rotation of the forearm was done till the mid-prone position as this was the non-dominant hand. Radial osteotomy was fixed using a plate and the ulnar osteotomy was fixed using an intramedullary 2mm K wire (Figure 3a, 3b & 3c).



**Fig 3a:** Intraoperative photograph showing radial side osteotomy fixed with a plate



**Fig 3b:** Intraoperative photograph of ulnar side showing the percutaneous K wire going through the ulnar osteotomy



**Fig 3c:** Post-operative radiograph showing the both bone osteotomies fixed with plate and K wire

Post-operatively the patient was kept in above elbow cast for a period of 6 weeks till the signs of radiological union were seen, at which time the K wire from the ulna was removed and the patient was allowed gentle range of motion at elbow.

Six months post-operatively, at the time of implant removal from the radial side, the patient as well as the parents were satisfied with the outcome and he could eat without any difficulty and excessive use of wrist dorsiflexion (Figure 4a&4b).





**Fig 4a:** Radiograph at the time of implant removal from radius (6 months post-operative)



**Fig 4b:** Clinical photograph at 1 year of follow-up showing improved hand to mouth function

At present the boy is 5 years old and the forearm is maintained in a mid-prone position with good hand to mouth function at one year follow-up period.

### 3. Discussion

Congenital radioulnar synostosis is an uncommon entity affecting males more than females. Familial cases are predominantly of autosomal dominant inheritance. Syndromic association has also been found with syndromes like klinefelter's syndrome, arthrogryposis, carpenter syndrome etc [3]. Cleary classified radioulnar synostosis into four types. Type 1 had a non-osseous synostosis with reduced radial head, Type 2 had osseous synostosis with reduced radial head, Type 3 had long osseous synostosis with hypoplastic posteriorly dislocated radial head and Type

4 had a short osseous synostosis with mushroom shaped radial head dislocated anteriorly [4]. As per the cleary classification, our patient had type 2 radioulnar synostosis.

The usual concern in patients with lesser degrees of pronation deformity (up to 20-30 degrees of fixed pronation) is cosmetic as they are able to all the activities without much difficulty. But with higher degrees of fixed pronation, the patients have difficulty in using day to day items like spoons, forks and in other hand to mouth activities. These are cases that need to be treated surgically for the correction of deformity. Moreover, this condition is found bilaterally in 60 percent of the cases, where the functional limitation of child is even more and warrants surgical correction [5]. In our case, the deformity was unilateral and the affected limb was non-dominant.

Different surgical options mentioned previously include excision of the synostosis and interposition of vascularized fat graft, osteotomy through the synostosis and excision of radial fragment and both bone forearm de-rotation osteotomy. Synostosis excision and interposition fat grafting is difficult to execute surgically. Also, the amount of dissection and the morbidity of the procedure is much more with possible complications of neurovascular compromise and chances of recurrence. Osteotomy done through the synostosis and excision of the radial fragment can be done in late presenting cases, but it should be avoided in younger children due to the risk of DRUJ (distal radioulnar joint) instability and also there is increased risk of vascular compromise as the soft tissues are tight and the surgical field is complex. De-rotation osteotomy done in the diaphysis of radius and ulna is a relatively simple procedure to provide functional mid-prone position of the forearm. The recommended age at which this procedure is done is 4-10 years as it is easy to perform the osteotomy and adequate remodeling can occur in this age group. A delay in executing this procedure till later age has increased chances of complication as the deformity becomes long standing and soft tissues tend to tighten with time [6]. Shingade VU *et al.* reported a successful outcome of single stage de-rotation osteotomy of proximal ulna and distal radius in their study on congenital radioulnar synostosis [7].

There are different reports in literature regarding the optimum position of forearm to fix the osteotomy. Some suggest to fix the osteotomy in 0-30 degrees of pronation in dominant side and 0-20 degrees on supination in non - dominant side while others fix it in 0-10 degrees of supination irrespective of hand dominance arguing that compensation at the level of shoulder is enough to adapt for writing and using keyboard with the dominant hand [8, 9]. We chose to fix the forearm in a mid-prone position considering non-dominant role of the affected side in our patient.

Cases such as these need to be brought to attention for an improved management of such children at an appropriate age and to provide better alignment of forearm with respect to hand before they enroll in school.

### 4. Conclusion

Radioulnar synostosis with greater degrees of pronation deformity limiting the day to day activities of the child should be considered for surgical correction. Single stage both bone forearm osteotomy is useful and simple procedure for correction of deformity resulting from congenital radioulnar synostosis in children. It improves the function by realigning the forearm in the appropriate position with

respect to the wrist and thus improves the quality of life of the patient.

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**7. Statement of informed consent:** We ensure that a valid written consent was taken from the parents of the patient (as the patient is a minor) for the purpose of treatment as well for the purpose of publication of the data and photographs.

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