Derotational osteotomy in the treatment of congenital radioulnar synostosis: A report of 11 cases

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Abstract

Introduction: Congenital proximal radio-ulnar synostosis is a rare congenital anomaly of the upper extremity which is severely disabling as it causes severe pronation deformity. So the aim of this study was to assess the efficacy of proximal radioulnar derotational osteotomy followed by internal fixation for the treatment of congenital radioulnar synostosis.

Materials and Method: Between June 2018 and April 2020, 11 patients with CRUS who underwent derotational osteotomy at the proximal radioulnar synostosis site were evaluated. There were 7 boys and 4 girls. The mean age at the time of surgery was 10.5 years (range, 2 to 13) years. The forearm was derotated to the goal position (20 degrees of supination to 10 degrees of pronation) using plates for internal fixation and plaster splints for external immobilization. Pre- and postoperative positions of the forearm were recorded; forearm function was evaluated based on the classification system proposed by Failla et al.

Results: The mean follow-up duration was 6 months. The mean initial pronation deformity was 65.0 ± 5.2 (55 to 80) degrees. The mean correction achieved was 75.27 ± 5.25 (50 to 90) degrees, resulting in a mean final position of 9.45 ± 3.25 degrees of supination (20 degrees of supination to 10 degrees of pronation). Based on the Failla classification system, 7 forearms were rated as good, 4 were rated as fair preoperatively. At the final follow-up, 9 forearms were rated as excellent and 2 were rated as good. All patients achieved bone union after 10 weeks. Post-operative period was uneventful and no complications were seen.

Conclusion: Proximal radioulnar derotational osteotomy followed by plate fixation is a safe and feasible procedure with a low complication rate. The technique can effectively improve the function of the forearm.

Keywords: Congenital radioulnar synostosis (CRUS), Derotational osteotomy, Failla classification system

Introduction

Synostosis is the union of any adjacent bones and can involve any part of the body. Synostosis between the radius and ulna can be either, congenital or post traumatic. Congenital radioulnar synostosis is a rare upper limb malformation and was first described by Sandifort’s in 1793. The elbow is first identifiable 5 weeks after conception. At this stage, the cartilaginous anlagen of the humerus, radius, and ulna are continuous. Subsequently, longitudinal segmentation produces separation of the distal radius and ulna. However, temporarily, the proximal ends are united and continue to share a common perichondrium. Abnormal genetic or environmental factors operating at this time could interrupt subsequent proximal radioulnar joint morphogenesis. Congenital radioulnar synostosis may be isolated or associated with other abnormalities such as brachydactyly, polydactyly, syndactyly, thumb aplasia, and Madelung’s deformity. It may also be found as a part of well-known syndromes (arthrogryposis, Apert’s syndrome, Carpenter’s syndrome, Williams’ syndrome) and chromosomal abnormalities (Klinefelter’s syndrome). Both sexes are affected with 3: 2 male to female ratio. It is bilateral in 60% of the cases. The main feature of this malformation subtype is that the forearm is fixed at the pronation position. The loss of forearm rotation results in hindrance of actions that require this type of motion. The ipsilateral shoulder and wrist can effectively compensate for mild deformity.
When pronation deformities are severe, daily activities such as eating, washing, dressing, engaging in personal hygiene care, and accepting objects in the palm of the hand can be severely impaired. Different classifications of radioulnar synostosis are present. Cleary and Omer proposed four radiographic types:

1. Type I: fibrous synostosis with a reduced and normal-appearing radial head.
2. Type II: osseous synostosis with normal radius.
3. Type III: osseous synostosis with a hypoplastic and posteriorly dislocated head.
4. Type IV: a short osseous synostosis with an anteriorly dislocated radial.

Currently, derotational osteotomy remains the most commonly performed procedure in patients with CRUS, which alters the position of the forearm from hyperpronation to a more functional position to reduce supination limitations and to allow patients to more easily perform activities of daily life.

**Materials and Methods**

Between June 2018 and April 2020, eleven patients with CRUS were managed with derotational osteotomy and plate fixation. In our study, there were 7 boys and 4 girls among them left elbow was involved in 10 patients and the right elbow was involved in 1 patient. All the patients had unilateral CRUS, the mean age at the time of surgery was 10.5 (range, 2 to 13) years. The inclusion criteria were children presenting with either unilateral or bilateral CRUS, skeletally immature children with open growth plates, forearm pronation ≥ 55 degrees, and a score ≤ 10 points based on the Failla classification system. The exclusion criteria were children with inadequate clinical or radiographic data; other combined ipsilateral upper limb anomalies.

Preoperative assessment was performed for all patients, including both clinical and radiological assessment. The clinical assessment included the ability of performing the activities of daily life, the degree of pronation deformity, and the range of motion of the elbow and wrist. The radiographic assessment was based on the classification of Cleary and Omer.

Preoperative assessment of the patients revealed that all of them had limitation of the activities of daily life. All patients were type II according to Cleary and Omer, with visible osseous synostosis with normal radius.

The procedure was carried out under general anaesthesia with the patient in supine position under tourniquet control. A posterior thompson’s approach with longitudinal incision of 3 cm on the outer edge of the olecranon first allowed the synostosis to be exposed. A Kirschner wire was inserted laterally, distal to the olecranon growth cartilage and pushed into the medullary canal. The periosteum was detached to expose the synostosis. The Kirschner wire then retreated enough to perform a horizontal subperiosteal osteotomy, in the middle 1/2 of radius and ulna. and the forearm was gradually derotated to the target position (20 degrees of supination to 10 degrees of pronation) with the elbow bent at 90°. Both forearm bones i.e radius and ulna were fixed with 3.5mm locking plate and screw system. Full flexion and extension of the elbow joint were confirmed to exclude any impingement of the joint. Plaster was applied in neutral position for the duration of 6 weeks, with a follow-up of 6 months. Our osteotomy easily consolidated within ten weeks and no complications were found.

![Fig 1: Patient with Congenital radio-ulnar synostosis of left elbow exhibiting restriction of supination and pronation at left elbow.](image)

![Fig 2: Pre-operative X-ray](image)

![Fig 3: Intra-operative X-ray (Transverse osteotomy)](image)
Bone union of the osteotomy site was evaluated postoperatively by X-ray, and the incidence of complications was calculated. Evaluation of the surgical effect was first performed by preoperatively and postoperatively measuring the axial position of the forearm to determine improvements of forearm function. The pronation deformity was measured with the patient’s elbow held fixed to the side of the chest and the forearm at 90 degrees of flexion, 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. The forearm function of the children was also assessed via the classification system established by Failla et al. \[12\] for 15 tasks described by Morrey et al. \[13\] (Table 2).

Table 2: The functional ranges of rotation of the forearm classification system used by Failla et al.

| Quantity | Daily activities | Complete:1 point/ cannot complete:0 point |
|----------|------------------|------------------------------------------|
| 1        | Touch hand to the vertex (head) | 1/0                                      |
| 2        | Touch hand to the occiput | 1/0                                      |
| 3        | Touch hand to the neck | 1/0                                      |
| 4        | Touch hand to the chest | 1/0                                      |
| 5        | Touch hand to the waist | 1/0                                      |
| 6        | Touch hand to the sacrum | 1/0                                      |
| 7        | Touch hand to the shoe | 1/0                                      |
| 8        | Pour from a pitcher | 1/0                                      |
| 9        | Put glass to the mouth | 1/0                                      |
| 10       | Cut with a knife | 1/0                                      |
| 11       | Put fork to the mouth | 1/0                                      |
| 12       | Use a telephone | 1/0                                      |
| 13       | Read a newspaper | 1/0                                      |
| 14       | Rise from a chair | 1/0                                      |
| 15       | Open a door | 1/0                                      |

Total score: 0–15
Excellent, 15 points; good, 10–14 points; fair, 6–9 points; and poor, < 6 points

Results
The mean follow-up duration was 6 months. The mean initial pronation deformity was 65.0 ± 5.2 (55 to 80) degrees. The mean correction achieved was 75.27 ± 5.25 (50 to 90) degrees, resulting in a mean final position of 9.45 ± 3.25 degrees of supination (20 degrees of supination to 10 degrees of pronation). Based on the Failla classification system, 7 forearms were rated as good, 4 were rated as fair preoperatively. At the final follow-up, 9 forearms were rated as excellent and 2 were rated as good. All patients achieved bone union after 10 weeks. There were no cases of transient nerve palsy, compartment syndrome, correction loss, infection, or broken plate. Post-operative period was uneventful and no complications were seen.

Table 3: Comparison of results of before and after surgery in the operated forearms

| Pronation deformity (mean ± SD; degrees) |
|----------------------------------------|
| Preoperative | 65.0 ± 5.2 |
| Postoperative | 9.45 ± 3.25 |

Table 4: The forearm function by the Failla classification system

| Excellent | Preoperative | Postoperative |
|-----------|-------------|---------------|
|           | 0           | 9             |
| Good      | 7           | 2             |
| Fair      | 4           | 0             |
| Poor      | 0           | 0             |

Discussion
CRUS is a rare deformity thought to be caused by a failure of prenatal longitudinal segmentation with persistence of the cartilaginous anlage between the radius and the ulna during the fifth to seventh week of embryogenesis \[14\]. The resultant
bridge may be fibrous or bony [15]. The main feature of this malformation subtype is that the forearm is fixed at the pronation position. When pronation deformities are severe, daily activities such as eating, washing, dressing, engaging in personal hygiene care, coo ming and accepting objects in the palm of the hand like accepting prasad am can be severely impaired. Over time, the specific indications for operative intervention have been debated. Cleary and Omer [5] believe that operative intervention is rarely indicated and their indications for surgery are based more on functional deficits than absolute forearm position. However, Simmons et al. [6] stated that a pronation over 60 degrees was an absolute indication for surgery. Ogino and Hikino [6] suggested that a fixed pronation of 15 to 60 degrees was a relative surgical indication based on the associated functional deficit. Thus, we selected forearm pronation ≥ 55 degrees and a Failla classification system score ≤ 10 points as the surgical indications in this study.

Generally, there are two surgical options to treat congenital radioulnar synostosis. The first option is to separate the radioulnar synostosis to restore forearm rotation, but the reports of other authors are disappointing with high rate of recurrent synostosis [5-7]. The second surgical option is derotational osteotomy to realign the forearm in a suitable position.

Some reports have been published on the separation of synostosis and interposition of fat or muscle (or some other material), but recurrence of the ankylosis has still been noted [16, 17]. Kanaya et al. [18] reported bone resection of synostosis with free vascularized tissue interposition in seven cases, and all patients had no fusion recurrence and gained forearm rotation at an average of 3.67 years of follow-up. However, the technique of separation and reconstruction surgery has not gained wide acceptance, and derotational osteotomy currently remains the most commonly performed procedure in patients with CRUS.

In this study, the osteotomy was performed through the synostosis in all cases and also transverse derotational osteotomy of radius and ulna. All osteotomies were fixed by plate and screws to ensure adequate degree of intraoperative correction and to avoid postoperative loss of this correction. None of our patients showed loss of correction during postoperative follow-up. There were no increases in the incidence of complications by using plate fixation in our study. Also our study showed that in 0 (neutral) to 20 degrees of supination, both the dominant and nondominant forearms, had the best function, i.e., the highest score of 15 points based on the Failla classification system. Patients with forearms in this position could perform movements that require forearm pronation, such as using a computer mouse and keyboard and touching the back of the head, and they could also complete movements that require forearm supination, such as holding a bowl in the palm, receiving prasad am, washing, touching the wrist with the palm of the hand, and engaging in personal hygiene care.

Conclusion

Proximal radioulnar derotational osteotomy followed by plate fixation is a safe and feasible technique in patients with CRUS. The technique has a low complication rate and can effectively improve the function of the forearm and the quality of the patient’s life.

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References

1. Hansen OH, Anderson NO. Congenital radioulnar synostosis. Report of 37 cases. Acta Orthop Scand 1970;41(3):225-30.
2. Elliott AM, Kibria L, Reed MH. The developmental spectrum of proximal radioulnar synostosis. Skeletal Radiol 2010;39:49-54.
3. Castello JR, Garro L, Campo M. Congenital radioulnar synostosis: surgical correction by derotational osteotomy. Ann Chir Main 1996;15:11-17.
4. Simmons BP, Southmayd WW, Riseborough EJ. Congenital radioulnarsynostosis. J Hand Surg Am 1983;8:829-838.
5. Cleary JE, Omer GE Jr. Congenital proximal radio-ulnar synostosis. Natural history and functional assessment. J Bone Joint Surg Am 1985;67:539-45.
6. Ogino T, Hikino K. Congenital radio-ulnar synostosis: compensatory rotation around the wrist and rotation osteotomy. J Hand Surg Br 1987;12:173-8.
7. Hwang JH, Kim HW, Lee DH, Chung JH, Park H. One-stage rotational osteotomy for congenital radioulnar synostosis. J Hand Surg Eur Vol 2015;40:855-61.
8. Green WT, Mital MA. Congenital radio-ulnar synostosis: surgical treatment. J Bone Joint Surg Am 1979;61:738-43.
9. Shingade VU, Shingade RV, Ughade SN. Results of single-staged rotational osteotomy in a child with congenital proximal radioulnar synostosis: subjective and objective evaluation. J Pediatr Orthop 2014;34:63-9.
10. Fuji moto M, Kato H, Minami A. Rotational osteotomy at the diaphysis of the radius in the treatment of congenital radioulnar synostosis. J Pediatr Orthop 2005;25:676-9.
11. Simcock X, Shah AS, Waters PM, Bae DS. Safety and efficacy of derotational osteotomy for congenital radioulnar synostosis. J Pediatr Orthop 2015;35:838-43.
12. Failla JM, Amadio PC, Morrey BF. Post-traumatic proximal radioulnar synostosis. Results of surgical treatment. J Bone Joint Surg 1989;71-A:1208-1213.
13. Morrey BF, Askew LJ, Chao EY. A biomechanical study of normal functional elbow motion. J Bone Joint Surg Am 1981;63:872-7.
14. Bhatt CR, Mehta CD. Case report: congenital radioulnar synostosis and its embryological correlation and functional assessment. J Anat SocIndia 2011;60:236-8.
15. Bauer M, Jonsson K. Congenital radioulnar synostosis. Radiological characteristics and hand function: case reports. Scand J Plast Reconstr SurgHand Surg. 1988;22:251-5.
16. Dawson H. A congenital deformity of the forearm and its operative treatment. Br J Med. 1912;2:833-5.
17. Miura T, Nakamura R, Suzuki M, Kanie J. Congenital radio-ulnar synostosis. JHand Surg (Br). 1984;9:153-5.
18. Kanaya F, Ibaraki K. Mobilization of a congenital proximal radioulnar synostosis with use of a free vascularized fascio-fat graft. J Bone Joint Surg Am. 1998;80:1186-92.