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Unusual presentation of congenital radioulnar synostosis with osteoporosis, fragility fracture and nonunion: A case report and review of literature

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Abstract

BACKGROUND

Congenital radioulnar synostosis (CRUS) is a rare deformity of the upper extremity. It is characterized by loss of rotation of the involved forearm and functional limitations in daily activities. No studies on CRUS with osteoporosis have been reported to date, and osteoporosis is usually recognized as an important dimension of genetic disorder in children. We discuss the possible relationship among this disorder, osteoporosis and fracture nonunion, investigate the strict surgical indications and recommended treatments.

CASE SUMMARY

A 14-year-old male patient with bilateral CRUS with osteoporosis, fragility fracture and nonunion of fractures in ulna and radius presented our institution for further treatment, complaining of limitation in rotation. The bone mineral density of the hip and lumbar spine was 0.687 g/cm² and 0.705 g/cm², respectively, and the Z-score for both was -2.1, which revealed osteoporosis and a high risk of fracture. Two serum bone turnover markers indicated an imbalance of bone metabolism. Reoperation for ulna fracture with autogenous bone grafting and a postoperative physiotherapy program were adopted rather than the separation of pathological synostosis. Radiological examination, observational posture assessment and limb function scale were evaluated before and 1 year after surgery. At 1 year, the fracture nonunion had almost recovered, forearm movement function on the fracture side was restored, and function on the healthy side was significantly improved compared with that before rehabilitation.

CONCLUSION

Surgical indications for CRUS vary from person to person. Surgery should not be the first choice of treatment, and physiotherapy is not inferior to surgical treatment.

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Core tip: Congenital radioulnar synostosis is a rare deformity of the upper extremity, which is characterized by loss of rotation and a variety of functional limitations. We present a rare case of bilateral congenital proximal radioulnar synostosis with osteoporosis, fragility fracture and nonunion. This case suggests that internal fixation plus bone grafting or stimulus of osteogenesis may result in a much better outcome than expected. In addition, this case highlights that surgery should not be the first choice except for those with severe daily activity limitations, and physiotherapy is not inferior to surgical treatment.

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INTRODUCTION

Congenital radioulnar synostosis (CRUS), which is caused by fusion between the proximal end of the radius and ulna with the forearm fixed in some degrees of pronation, is a rare deformity of the upper extremity. It is common, however, in congenital elbow anomalies, and approximately 60%-80% of cases are bilateral^[1-3]. It is characterized by loss of rotation of the involved forearm and a variety of functional limitations in daily activities in severe cases, presumably as a result of an insult *in utero* at the time when the proximal ends of the radius and ulna were connected by a common perichondrium^[4,5]. The etiology has been reported to be genetic and multifactorial^[1]. Pain or restriction of elbow flexion-extension movements is absent in most patients with CRUS; their forearm is usually fixed in the pronation, and the function of supination is lost to varying degrees^[1,6]. In most cases, the evaluation of the malformation is based on the classification by Cleary and Omer^[7,8]. It is classified into four radiographic types, as shown in Table 1^[9].

No studies on CRUS with osteoporosis have been reported to date, but osteoporosis is usually recognized as an important dimension of genetic disorder in children. Childhood osteoporosis is typically divided into primary and secondary causes, with osteogenesis imperfecta (OI) representing the prototypical primary osteoporosis of childhood, predisposing to bone fragility and fracture^[10-13]. Low-trauma non-vertebral fractures in children with osteoporosis are observed most frequently at the femur, tibia, forearm, humerus, feet and ankles^[14-16].

We here report a 14-year-old male patient with bilateral CRUS with osteoporosis, fragility fracture and nonunion of fracture in ulna and radius. Informed consent was obtained from the patient and his guardians for using the data of this case to be submitted for publication.

CASE PRESENTATION

Chief complaints

A 14-year-old right-handed boy with CRUS of the bilateral forearm and a fragility fracture of both bones of the forearm in a fall presented to the Department of Orthopedics of our hospital.

History of present illness

In another hospital, open reduction and internal fixation were performed, while the ulna was fixed with a Kirschner wire, maintaining neutral forearm position in a plaster cast for 8 wk before rehabilitation was initiated. At 3 mo after surgery, since the location of the reduced ulna was lost and the fracture was nonunion, the patient came to our hospital and asked for further treatment.

Table 1 Cleary and Omer classification of congenital proximal radioulnar synostosis

Type (proportion)	Description
I (19%)	Fibrous synostosis with a reduced normal-appearing radial head
II (8%)	Visible bony synostosis with a reduced radial head
III (56%)	Visible bony synostosis with a hypoplastic and posteriorly dislocated radial head
IV (17%)	Short bony synostosis with an anteriorly dislocated mushroom-shaped radial head

History of past illness

The boy had two previous fractures, including one distal radius fracture and one tibiofibular fracture.

Personal and family history

There was a possible hereditary history in his family, where his mother and younger brother had a similar elbow deformity.

Physical examination upon admission

On physical examination, a hard but painless lump was found in the posterolateral region of the bilateral elbow, and it could not be reduced in flexion-extension or in pronation-supination. A deficiency in active motion was present at the left forearm (Figure 1). Right forearm function was slightly limited for the compensation of the ipsilateral shoulder, and the patient had no flexion and extension dysfunction on both sides of the forearm.

Laboratory examinations

Bone mineral density (BMD) was measured by Dual Energy X-ray Absorptiometry. BMD of the hip and lumbar spine was 0.687 g/cm² and 0.705 g/cm², respectively, and the Z-score for both was -2.1. The 2013 International Society for Clinical Densitometry recommended a BMD Z-score threshold (-2.0 or worse) in children to denote childhood osteoporosis, provided such children also have a clinically significant fracture history, defined as ≥ 2 long bone fractures by age 10 and ≥ 3 long bone fractures by age 18^[17]. According to the recommendation, the patient was identified as childhood osteoporosis with an increased fracture risk. Serum collagen type I cross-linked C-telopeptide (a marker of bone resorption) and serum procollagen I N-terminal propeptide (a marker of bone formation) of the patient were 1832 ng/mL and 347 ng/mL, respectively. These two bone turnover markers are recommended by the International Osteoporosis Foundation and the International Federation of Clinical Chemistry and Laboratory Medicine for the assessment of bone health^[18].

Imaging examinations

Plain radiography showed a fracture of both bones of the left forearm and pathologic fusions between the proximal diaphysis of radius and ulna, with a small bone diameter (Figure 2).

FINAL DIAGNOSIS

Bilateral CRUS with osteoporosis, fragility fracture and nonunion of fractures in ulna and radius.

TREATMENT

Bilateral synostoses of this case were both classified as type II according to Cleary and Omer. Considering that the child's forearm function of the dominant side was not significantly affected with the compensation of the ipsilateral limb and could ensure necessary living requirements, reoperation for ulna fracture with autogenous bone grafting and a postoperative physiotherapy program were adopted rather than the separation of pathological synostosis. Physiotherapy was performed by a professional rehabilitation physician, and all exercises were practiced bilaterally. These exercises consisted of actively stretching the muscle to the point of limitation of supination, followed by a passive supination movement, and then practicing compensatory movement of adjacent joints.

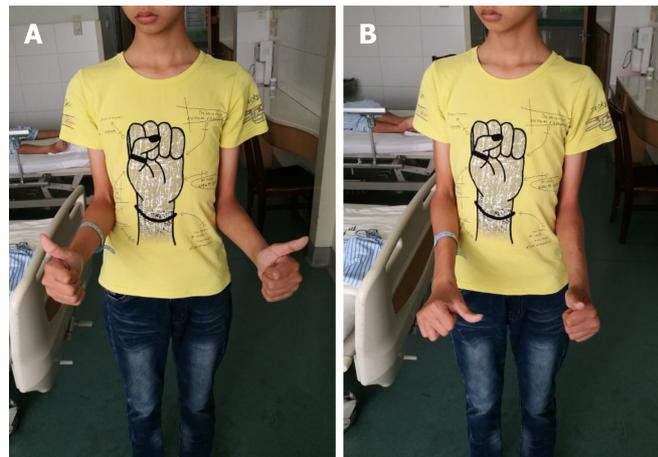


Figure 1 Active motion of the left forearm was limited. In order to reduce the rotation error caused by the compensation of elbow motion, the elbow joint was tightly attached to the waist. A: Supination; B: Pronation.

OUTCOME AND FOLLOW-UP

The patient was evaluated before and 1 year after surgery with radiological examination, observational posture assessment, the Activities of Daily Living Scale, the Liverpool Elbow Score, the Disabilities of the Arm, Shoulder, and Hand score and the Failla classification system. The Activities of Daily Living Scale was used to evaluate subjectively the degree of improvement in activities of daily living, including 11 that related to hygiene and self-care, eight that related to feeding, and seven that assessed environmental interaction on a 5-point scale. The Liverpool Elbow Score was first introduced in 2004 as an elbow-specific outcome score to be completed by both the clinicians and patients, combining a nine-item patient-answered questionnaire and a six-item clinical assessment score^[19]. The Disabilities of the Arm, Shoulder, and Hand score is a 30-item questionnaire that quantifies physical function and symptoms in persons with any or multiple musculoskeletal disorders of the upper limb^[20]. The Failla classification system is a grading system used by Failla *et al*^[21] to evaluate the functional ranges of rotation of the forearm. The evaluation results are given in [Table 2](#). One year later, the fracture had almost recovered ([Figure 3](#)), and the range of motion was significantly improved ([Figure 4](#)).

DISCUSSION

The forearm complex functions to rotate the hand in space and allow for a redistribution of forces during functional tasks. Disruptions of any one of the components can potentially lead to limitations in forearm rotation and function^[22]. CRUS as a rare malformation is thought to be an anomaly of longitudinal segmentation that develops in the early fetal life. The elbow begins to appear at day 34-35 of fetal life, while the humerus, radial bone and ulna appear at day 37^[7]. The humerus, radius and ulna are continuous with each other and are joined by a common perichondrium within the first 60 d of embryologic development. The cartilaginous anlage between these bones is separated by condensation, while the forearm is in a neutral position. Deformity of the radial head may be closely related to complete proximal union caused by early interference with joint formation^[23] or unequal growth of the radius head caused by limited distal epiphyseal fusion^[24].

CRUS is usually bilateral and subject to autosomal dominant inheritance^[25]. Moreover, CRUS is one of many components of malformation syndromes in children with chromosomal aberrations^[8,26]. A number of authors have claimed that there is a genetic basis for radioulnar synostosis. Evidence for this etiology, like in this case, includes familial occurrence and the close relation with other congenital syndromes such as acrocephalosyndactyly, microcephaly, Apert's syndrome, Carpenter's syndrome, arthrogryposis, Klinefelter syndrome, multiple exostoses, Holt-Oram syndrome, fetal alcohol syndrome, Ayme-Gripp syndrome, Saethre-Chotzen syndrome, mandibulofacial dysostosis with microcephaly and William's syndrome^[2,27-30].

Because children can easily compensate for rotational limitations caused by joint deformities, the disease is usually not diagnosed early^[31]. Although the pronation and



Figure 2 Plain radiographs of the bilateral forearm at 3 mo after surgery. A, B: Left side with loss of reduced location; C, D: Right side with the same deformity of radioulnar synostosis.

supination movements of forearm are limited, elbow flexion and extension exercises are generally unaffected. Most patients with CRUS can manage daily activities through the overuse and compensation of shoulder and wrist^[6,32]. Shoulder abduction compensates for inadequate pronation, and shoulder adduction compensates for inadequate supination^[33].

Our patient had type II bilateral synostoses, according to the Cleary and Omer classification, and had an accessory bone in the proximal part of his bilateral forearm wedged between the radius and ulna. In addition, there was a fracture with slender long bones and thin cortex after a low-energy injury. Laboratory examination also revealed the presence of osteoporosis as compared with peers. The only available method to determine the pathogenesis with certainty is to measure directly bone formation and resorption on trabecular surfaces *via* transiliac bone biopsy. However, the patient refused this procedure because of the potential risks. We hypothesize that patients with CRUS may have OI or calcification disorder due to genetic defects. Therefore, for such patients with fractures, in addition to strong internal fixation, bone grafting or stimulation of osteogenesis may have more benefits than expected.

Many studies have examined different surgical methods for CRUS, such as derotational osteotomy of radial shaft, radial head excision and graft applications (see Table 3 for details). As CRUS is rare, it is difficult to conduct randomized-controlled trials, and there are generally only case reports or case series in the literature. Surgical techniques for treating radioulnar synostosis can be categorized into two groups: (1) Operations that improve pro-supination through synostosis resection, with or without interposition of biological or synthetic materials; and (2) Operations that improve the forearm's fixed position^[34]. Surgery is performed with the intention of restoring normal anatomical alignment, thus preserving normal forearm biomechanics. However, some studies have shown a high incidence of postoperative complications^[35-37]. It is almost impossible to restore the complete rotation of the

Table 2 The evaluation results before and after surgery

Outcome assessment	Before surgery		At 1 yr after surgery	
	Nondominant, fractured side	Dominant, non-fractured side	Nondominant, fractured side	Dominant, non-fractured side
ROM of elbow E/F (°)	0/140	0/140	0/140	0/140
ROM of forearm P/S (°)	0/5	-30/0	-45/45	-45/30
ADL ¹				
Hygiene and self-care items	3.5	4.0	4.2	4.5
Feeding-related items	3.3	4.6	4.5	4.9
Environmental interaction	3.3	4.3	4.3	4.7
Average	3.4	4.3	4.3	4.7
LES ²				
Clinical assessment	12	15	17	17
Patient-answered questions	11	30	31	34
Final scores	4.4	8.2	8.7	9.2
DASH ³				
Function	48		32	
Symptoms	14		7	
DASH value	26.7		7.5	
Failla classification system	Fair	Good	Good	Excellent

¹Scores range from 1 to 5. The lower scores represent greater limitation on activities of daily living.

²Scores range from 0 to 10. The lower scores represent greater symptom and functional severity.

³Value ranges from 1 to 100. Greater scores indicate more extremely limited upper limb function. ROM: Range of motion; E/F: Extension/flexion; P/S: Pronation/supination; ADL: Assessment of activities of daily living; LES: Liverpool Elbow Score; DASH: Disabilities of the arm, shoulder and hand.

affected forearm, and although the initial results might be satisfactory, the range of motion would return to the preoperative status over time^[37,38]. Satisfactory results were usually achieved in patients with very severe preoperative disability^[31,39-42].

Although an indication for operation in a patient with more than 60 degrees of fixed pronation was proposed, it must be individualized in patients with pronation between 15 and 60 degrees^[43]. The patients with > 60 degrees of pronation usually have restrictions in daily activities^[44]. Generally, the surgical protocol is dependent more on functional deficits than absolute forearm position. Studies have shown that physiotherapy is effective in the functional recovery of the upper extremities in patients with CRUS^[45].

In the present case, forearm rotation function was compensated well through the adjusted movements of the adjacent joints, wrists and shoulders, and there was no obvious limitation in daily life. Therefore, we suggest that the choice of procedure should be individualized as not all the patients are eligible for surgery.

CONCLUSION

In summary, based on a review of all similar reported cases, we believe that patients with CRUS complicated with osteoporosis and fracture, due to the suspicious gene-induced OI or calcification disorder, may benefit not only from indispensable firm internal fixation but also bone grafting or stimulus of osteogenesis. In addition, surgical indications in such patients vary from person to person. Surgery should not be the first choice except for those with severe limitations in daily activities, and physiotherapy may be a new method of functional rehabilitation. However, these are just conjectures obtained from a common case, and more research is needed to confirm these predictions in the future.

Table 3 Different surgical methods used in the treatment of congenital radioulnar synostosis

Ref.	Post-op follow-up in mo	Surgical methods	Preoperative fixed forearm position (°) ¹	Final forearm position (°) ⁴	Complications
Simcock <i>et al.</i> ^[31] , 2015	46 (range, 6-148)	Derotational osteotomy	85 (range, 60-100)	-8 (range, -30-0)	12%
VanHeest <i>et al.</i> ^[35] , 2013	33 (range, 24-42) ²	Radial head excision ³	27.5 (range, 0-60)	Mainly improve elbow extension movement	25% (one with transient radial nerve neuropraxia)
Shingade <i>et al.</i> ^[46] , 2014	54 (range, 36-84)	Radial osteotomy	56.3 (range, 30-86)	27.2 (range, 20-30)	Not available
Hung <i>et al.</i> ^[39] , 2008	64 (range, 30-129)	Derotational osteotomy	78 (range, 65-85)	8 (range, 0-30)	0%
Bishay <i>et al.</i> ^[40] , 2016	30.4 (range, 24-36)	single-session double-level rotational osteotomy	70.7 (range, 60-85)	-15.5 (range, -30-20)	0%
Garg <i>et al.</i> ^[36] , 2015	32.1 (range, 16-65)	Radial head excision with a tensor fascia lata graft	51.6 (range, 30-70)	15 (range, 5-32)	50% (superficial infection, posterior interosseous nerve palsy and a persistent extension lag)
Horii <i>et al.</i> ^[41] , 2014	60 (range, 12-156)	Derotational osteotomy	72 (range, 40-100)	0	0%
Pei <i>et al.</i> ^[42] , 2019	55.19 (range, 24-123)	Derotational osteotomy	62.92 (55-80)	7.94 (range, -10-20)	9.7% (transient nerve palsies and compartment syndrome)
Satake <i>et al.</i> ^[47] , 2018	163.2 (range, 120-228)	Derotational osteotomy	51.3 (range, 30-90)	4 (range, -20-30)	0%
Hwang <i>et al.</i> ^[38] , 2015	33 (range, 12-72)	Derotational osteotomy	47 (range, 30-65)	27 (range, 25-30)	0%
Kanaya <i>et al.</i> ^[37] , 2016	120 (range, 96-144)	Radial osteotomy with a free vascularized adipofascial graft	14.2 (range, 0-25)	6.7 (range, 0-25)	66.7% (flap congestion, transient radial nerve palsy, synovial fold and posterior or anterior radial head dislocation)

¹Degree of pronation.

²The fourth case without specific follow-up time.

³A method of only treating the anteriorly dislocated radial head in type IV radioulnar synostoses with radial head impingement.

⁴Degrees of supination: -8, 8° of pronation; -10, 10° of pronation; -15.5, 15.5° of pronation; -20, 20° of pronation; -30, 30° of pronation.

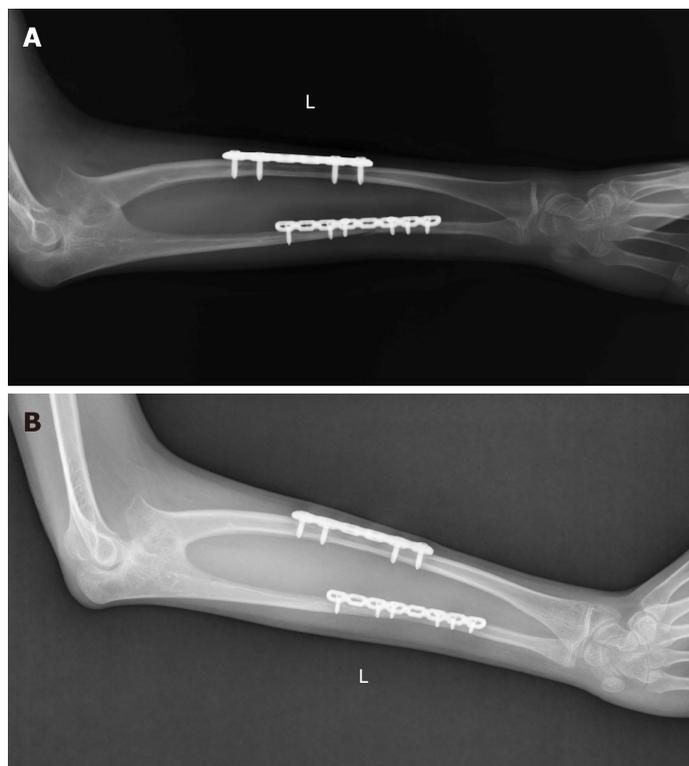


Figure 3 Plain radiographs showing the formation of callus, the disappearance of fracture line and the healing of fracture. A: At 2 mo after surgery; B: At 1 yr after surgery.



Figure 4 The range of motion of the left forearm was restored to the pre-injury state and was significantly improved after operation. A: Supination; B: Pronation.

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