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Congenital Radioulnar Synostosis, a Report of 7 Cases and Review of Literature

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Abstract **Original Research Article**

Background: Congenital radio-ulnar synostosis is rare condition, characterized by limited pronation and supination. It is typically diagnosed in school-aged children. The forearm is mostly fixed in a hyperpronated position. Transverse derotational osteotomy at the site of synostosis is the intervention we propose. ***Patients and methods*:** This is a retrospective study including 7 cases of congenital radio-ulnar synostosis followed and treated at the pediatric orthopedic department of the children's Hospital of Rabat between January 2006 to February 2017. ***Results:*** The average age is about 32.5 months with a male predominance. Bilateral involvement presents 57.7% of cases. According to the Cleary and Omer classification, 42.8% of cases are classified as type IV, and 57.1% of cases are classified as type III. The average degree of preoperative fixed rotation of the forearm is 66° (from 0° to 85°). Only one patient presents a poly-malformative syndrome. Ten forearms underwent surgery, with transverse derotational osteotomy at the site of synostosis, without any postoperative complications.

Keywords: Congenital radioulnar synostosis. Forearm. - Pronation-supination.

INTRODUCTION

Congenital radiocubital synostosis (CRS) is a rare bone disorder characterized by proximal fusion of the two bones of the forearm. As a result, the forearm is usually fixed in a neutral or hyperpronation position, limiting pronation-supination movements. Although the origin of this condition remains unknown, genetic factors are being considered due to the condition's association, at times, with family history and genetic diseases such as Apert, William or Klinefelter syndrome [1]. A defect in longitudinal segmentation secondary to an anomaly in the trajectory of the posterior interosseous artery, occurring early, generally around the 7th week of fetal development, is thought to be at the origin of this malformation. This defect leads to the persistence of a fibrous or bony bridge between the radius and ulna. This bony fusion immobilizes the upper forearm in a neutral position in relation to the humeral pallet. Associated anomalies, such as hypoplasia, dislocation or even absence of the radial head, may be present, giving rise to different classifications. CRS are usually treated surgically, although conservative treatment may be recommended for well-tolerated forms. Numerous surgical techniques have been described, divided into two categories: procedures aimed at restoring pronosupination of the forearm by releasing the synostosis, and procedures aimed simply at restoring a more functional position of the forearm.

OBJECTIVES

The aims of this work are to describe and evaluate the techniques used to correct CRS, to identify the indications and the most appropriate surgical technique, and to compare our results with those in the literature.

MATERIALS AND METHODS

STUDY MATERIALS

This is a retrospective study carried out in the traumatology-orthopedics department of Rabat Children's Hospital, during a period from January 2006 to February 2017. Seven files of children with congenital proximal radioulnar synostosis are included in our study. After obtaining approval from the hospital's ethics committee.

STUDY METHODS

The data were collected from patient files available in the archives. We analyzed the files using an evaluation sheet based on the following items: Age, sex, side affected, history, radiological classification, clinical

presentation intraoperatively, clinical presentation postoperatively, functional evolution and finally the follow-up period (Table 1).

Table 1: Clinical data of the seven patients

Case N°	1	2	3	4	5	6	7
Sex	F	M	M	F	M	M	F
Age	1year 6 months	2years 6months	8years	4years	7months	5months	2years
Involved side	Left	Bilateral	Left	Bilateral	Bilateral	Bilateral	Left
Dominant side	Left	Right	Right	Right	Right	Right	Right
Medical History	No	No	No	No	No	PMS*	No
Type (Cleary et Omer)	Type IV	Type III	Type III	Type IV	Type IV	Type III	Type III
Preoperative position	45°	DT : 85° G : 80°	90°	DT : 65° G : 80°	DT : 0° G : 70°	DT : 65° G : 70°	70°
Final position of fore arm	0°	**R** :25° Pronation **L** : 30° supination	20° pronation	**R** :20° Pronation **L** : 25° supination	**R** : - **L** : 0°	**R** :20° Pronation **L** : 20° supination	0
Correction degree	45°	**R** :60° **L** : 50°	70°	**R** :45° **L** : 55°	**R** : - **L** : 70°	**R** : 45° **L** : 50°	70°
follow-up period	5 years	4 years	1 year	9 years	9 years	12 years	3years
Functional improvement	Good	Good	Good	Good	Good	Good	Good

*polymalformative syndrome

SURGICAL TECHNIQUE

The indications for surgical treatment are dominated by the degree of functional discomfort reported by the patient, affecting daily activities. The surgery consists in performing a transverse derotation osteotomy at the site of synostosis of the ante-brachial skeleton, under general anaesthesia. A 3 cm posterior longitudinal incision is made on the outer edge of the olecranon to locate the synostosis (Fig 1a). A Kirschner wire is inserted laterally, distally to the olecranon growth plate, and pushed into the medullary canal. The periosteum is loosened with a rugine to expose the synostosis. The Kirschner wire is retracted to create a sub-periosteal osteotomy with an oscillating saw, horizontal in the proximal half of the synostosis. We then rotate the limb as required. The derotation position of the ante-brachial skeleton is stabilized by a second, obliquely directed Kirschner wire (Fig 1b). The lack of

vascular disturbance observed in the patient's new position allowed for the application of a brachio-antebrachio-palmar cast, which will be worn for six weeks (refer to Fig 2). The patient undergoes continuous monitoring for 48 hours to detect any potential vascular or nerve complications.

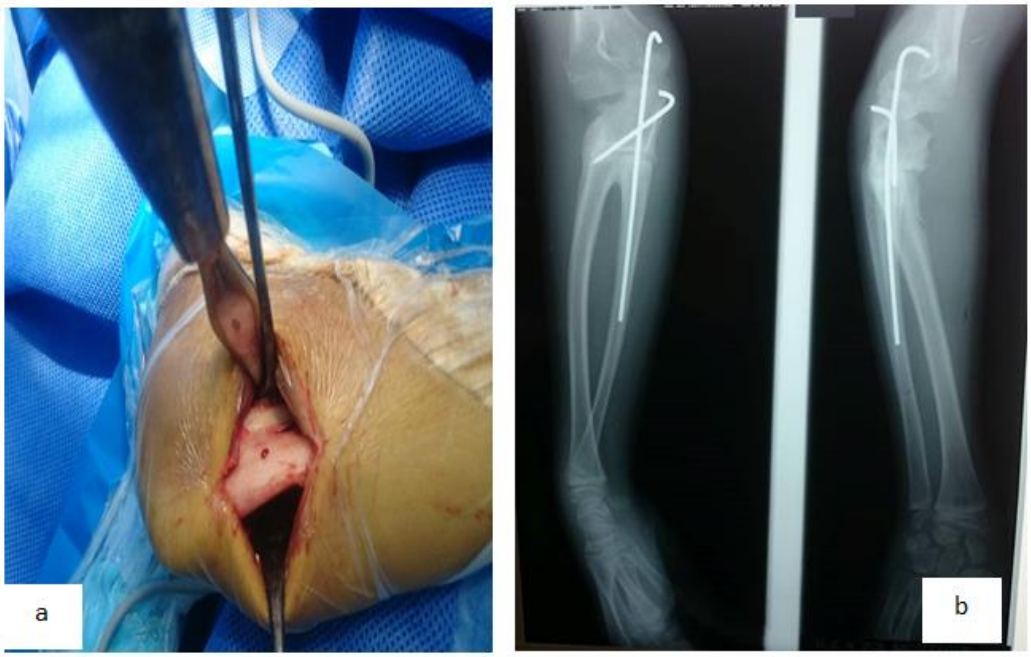

Fig 1: (a): Exposed site of synostosis (b): postoperative radiological aspect after transverse derotation osteotomy

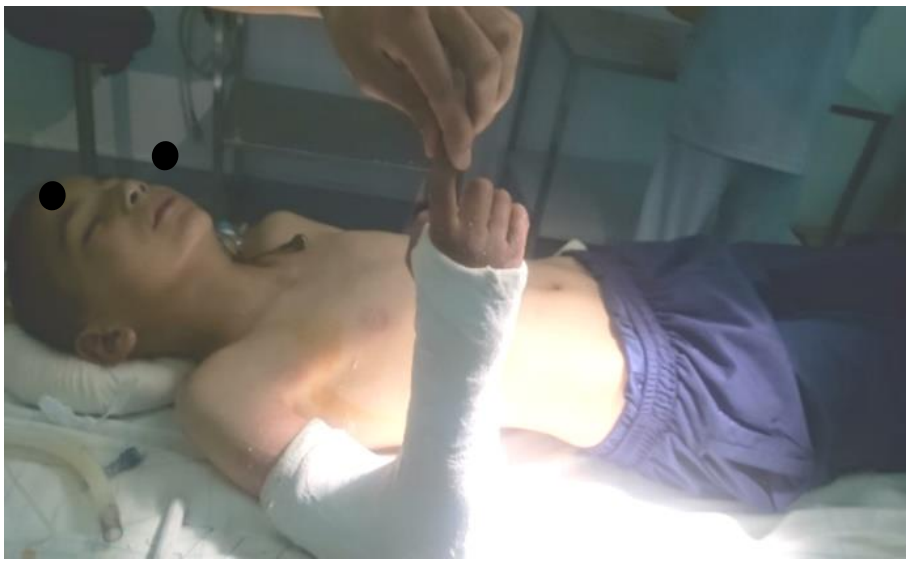

Fig 2: Brachial-antebrachial-palmar cast

RESULTS

This series included seven patients, with a slight male predominance, 4 boys and 3 girls, with a mean age of 34.2 months (from 5 months to 8 years). Unilateral involvement was found in 3 patients (42.8%) and bilateral involvement in 4 cases (57.1%). For all patients with unilateral involvement, the left side was always involved, i.e. 100%. 6 patients were right-handed and only 1 left-handed. The average degree of preoperative fixed forearm rotation was 66° (range 0° to 85°). The main complaints presented at the consultation were: difficulty eating (inability to hold a spoon or glass properly), difficulty washing the face and inability to groom, as well as difficulties at school for those children attending school. In terms of radiological classification, six forearms were classified as Cleary and Omer type III, with visible bony synostosis associated with posterior dislocation of a hypoplastic radial head, and five forearms were type IV, with short bony synostosis associated with anterior dislocation of the radial head (Fig 3). In the present series, only one patient presented with a poly-malformative syndrome, associating agenesis of the 5th finger of both hands, agenesis of the 2nd finger of both feet and congenital glaucoma. In the remaining cases, the CRS was isolated, with no

associated lesions. We performed 10 surgeries on 7 patients (10 forebar), with an average age on surgery of 6 years and 7 months, with extremes of 18 months and 8 years. Post-treatment forearm position ranged from 0° (neutral position) to 20° pronation for patients with unilateral involvement (3 cases) and from 20° to 25° pronation on the dominant side and from 0° (neutral position) to 30° supination on the non-dominant side for patients with bilateral CRS (4 cases) (Table 2). In our series, no post-operative complications were noted, and the improvement in forearm position was satisfactory in both functional and aesthetic terms for all patients.

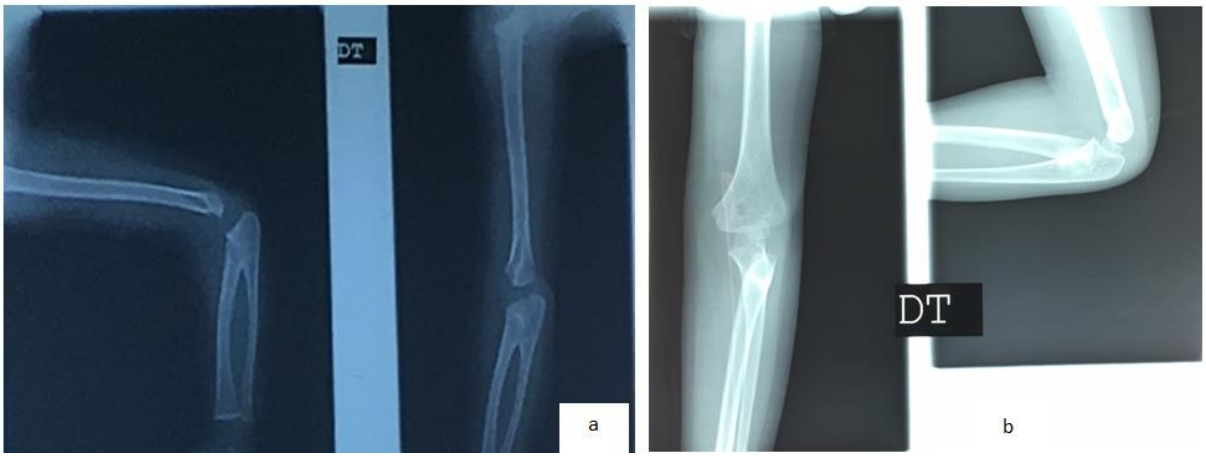

Fig 3: (a) Rx of right elbow showing CRS type III in case 2; (b) Rx of right elbow showing CRS type IV in case 4

Table 2: Position of the operated forearms after derotation osteotomy

Unilateral involvement	**Bilateral involvement**	
Neutral position (0°) at 20° pronation.	**Dominant side**	**Non-Dominant side**
	20° to 25° pronation.	Neutral position (0°) to 30° supination.

DISCUSSION

Wilkie's research [2] dates the initial documentation of congenital radioulnar synostosis back to 1793 by Sandifort at the Museum Anatomicus. This condition is a rare anomaly of the upper limb, with approximately 350 cases recorded in specialized literature [3]. Despite its rarity, it stands as the most prevalent congenital abnormality affecting the elbow joint. While genetics account for 25% of cases, congenital radioulnar synostosis often occurs in isolation [4, 8, 9]. Various associated skeletal anomalies have been documented, including clubfoot, congenital equinus varus, carpal synostoses, Madelung's disease, arthrogryposis, thumb agenesis, multiple synostoses disease, and hip dislocation [4]. In our observed series, there was no familial history of CRS among patients, with only one case displaying a polymalformative syndrome.

While congenital radioulnar synostosis is present at birth, it often remains undetected until later in a child's school years [13]. The functional limitations vary depending on the severity of the deformity, its bilateral or unilateral nature, and whether the affected side is dominant. In milder cases, compensatory movements from adjacent joints (shoulder and wrist) can mask limited pronation and supination, allowing for decent functionality initially. However, as children grow and engage in more demanding activities, limitations in arm movements become noticeable to parents and educators [14]. This limitation significantly impacts daily life and school performance, manifesting in difficulties such as holding objects like pencils, knives, and glasses properly. Tasks like buttoning clothes or eating can become challenging due to the pronation deformity. Additionally, basic grooming tasks like placing the hand at the perineum can be problematic. Participation in sports that require effective use of the upper limbs, such as catching or throwing a ball, or swinging a tennis racket, can also be hindered [15]. Based on our series, the average age of onset is around 2.7 years, consistent with findings reported by Tsuyoshi Murase [16] and Simmons *et al.*, [4].

In line with literature findings, CRS is reported as bilateral in 60-80% of cases [6, 7]. In our series, bilateral involvement was observed in 57.7% of cases, consistent with this range.

Cleary and Omer [5] categorized CRS into four radiological types (Table 3), with type III being the most prevalent, a trend also observed by other researchers [10, 11]. However, it's noteworthy that functional disparities among these types are minimal, and the radiological appearance can evolve with age. Consequently, the classification holds limited clinical significance [5, 12].

Table 3: Cleary & omer classification [5]

Type I	Fibrous ankylosis with normal radial head
Type II	Osseous synostosis with normal radial head
Type III	Osseous synostosis with posteriorly dislocated and hypoplastic radial head
Type IV	Pseudo-synostosis and anteriorly dislocated, mushroom-shaped radial head

The decision to pursue surgical intervention for CRS is primarily driven by the level of functional discomfort experienced by the patient, especially considering the relatively high complication rate, which can be as high as 36% [17]. The main risks associated with surgery include vascular or nerve damage due to overstretching during derotation, as well as the potential for compartment syndrome. Simmons *et al*., [4] proposed criteria for surgical intervention based on the degree of pronation deformity. They suggested that surgery is warranted when the forearm is fixed in pronation at 60° or more, as significant functional deficits are typically present in such cases. For pronation deformities ranging between 15° and 60°, the decision for surgery should be made based on factors such as the unilateral or bilateral nature of the condition and the results of the physical examination. If forearm function is severely limited, surgical intervention may be warranted. Conversely, cases with fixed pronation of less than 15° generally do not require surgery, as forearm function is typically satisfactory. Furthermore, Simmons *et al*., recommend prioritizing surgical intervention on the dominant side in cases of bilateral CRS. This approach aims to address the most functionally limiting condition first.

In our series, the decision for surgical intervention primarily hinges on the degree of functional discomfort experienced by the patient. Pronation fixed at over 60° is a clear indication for surgery, especially if it affects the dominant side. When pronation falls between 20 and 60°, the decision for surgery is relative and must be evaluated on a case-by-case basis. In instances where pronation is less than 15-20°, functional discomfort tends to be minimal, and surgical correction isn't typically necessary, as compensation can be easily achieved through shoulder movement. In total, we performed 10 interventions:

- Eight cases involved a forearm fixed in a pronated position greater than 60°, all deemed appropriate for surgical correction.
- In one instance, the forearm was fixed in a pronated position of 45° on the dominant side, necessitating surgical intervention due to the extent of functional discomfort.
- Another case presented with a neutral position at 0° for the right side and 70° for the left side. Surgical intervention was recommended for the left side due to the significant pronation deformity, while abstaining from surgery on the right side where functional discomfort was less pronounced. This individualized approach ensures that surgical decisions are tailored to each patient's specific needs and level of functional impairment.

The initial surgical techniques devised for CRS focused on restoring pronosupination through radical resection of the synostosis, often with interposition of various materials such as fascia, fat, muscle, or external implants. Unfortunately, outcomes from these procedures were disappointing, often resulting in subsequent restriction of movement [19, 12]. An alternative approach introduced by Kelikiian and Doumanian [20] involved the insertion of an intramedullary metal prosthesis of the radial head to re-establish supination and pronation. However, this technique also yielded unsatisfactory results. Contemporary surgical techniques, as advocated by many authors [4, 18, 21], diverge from attempts to restore pronosupination. Instead, the focus is on positioning the forearm in a more functional orientation, irrespective of pronosupination restoration. This shift in approach reflects a more pragmatic perspective aimed at improving overall functionality and quality of life for patients with CRS.

The transverse derotation osteotomy in synostosis was first described by Green and Mital, MA [7] based on a transverse osteotomy in synostosis. Murase *et al*., [10] performed osteotomies in the distal third of the radius and proximal third of the ulna in 4 patients with forearms fixed at more than 70° pronation. They achieved an average correction of 65°, the only complication being a 20° loss of correction during plaster cast immobilization. The Ilizarov method is based on progressive correction of the deformity after a derotation osteotomy, using an Ilizarov fixator at a rate of 1mm/day. Bolano [22] reported progressive derotation of the forearm fixed in pronation at 150° in 1 patient, after the use of an IIizarov fixator, an osteotomy through the synostosis was performed, the author made an immediate acute correction of 60°, followed by progressive derotation of 4° / d over 1 month, a neutral position s was achieved, and infection of the plugs was the only complication. Another study by Rubin [23] demonstrated the possibility of progressively correcting deformities greater than 90° using an Ilizarov external fixator. In our series, we advocate for the transverse derotation osteotomy at the synostosis site as the preferred surgical technique. This method involves the use of two Kirschner wires for osteosynthesis, followed by plaster cast immobilization for a period of 45 days to two months. The simplicity of this technique is a significant advantage, requiring only a single incision and exerting less compression on the surrounding soft tissues. Moreover, it is associated with a lower incidence of severe post-operative complications. Despite its simplicity, certain precautions must be taken when employing this technique. During the posterior approach,

it is crucial to perform aponeurotomies as thoroughly as possible. Additionally, shortening the skeleton in the osteotomy zone is imperative to relax the soft tissues. Correction can be easily achieved and should be secured with two pins—one radial and one ulnar. Post-operatively, close attention must be paid to ensure proper vascularization of the limb and normal finger movement. Monitoring these factors is essential for ensuring optimal post-operative outcomes and patient recovery.

The optimal position of the forearm after surgical correction of the synchronous radio-ulnar capitellar coalition remains a subject of debate. Several factors influence this decision, including whether the disease affects one or both forearms, the predominance of the affected side, and the patient's social and cultural environment. For bilateral cases, Green and Mital [7] proposed a position of pronation of 30° to 45° for the dominant forearm and 20° to 35° of supination for the non-dominant side. In unilateral cases, the ideal position is recommended between 10° and 20° of supination. In our practice, we offer a correction range of 0° to 20° of pronation for unilaterally affected patients. For bilateral cases, we recommend a correction of 20° to 25° of pronation for the dominant side and 0° to 30° of supination for the non-dominant side. Results were deemed satisfactory for patients and their families in both functional and aesthetic terms (Fig 4).

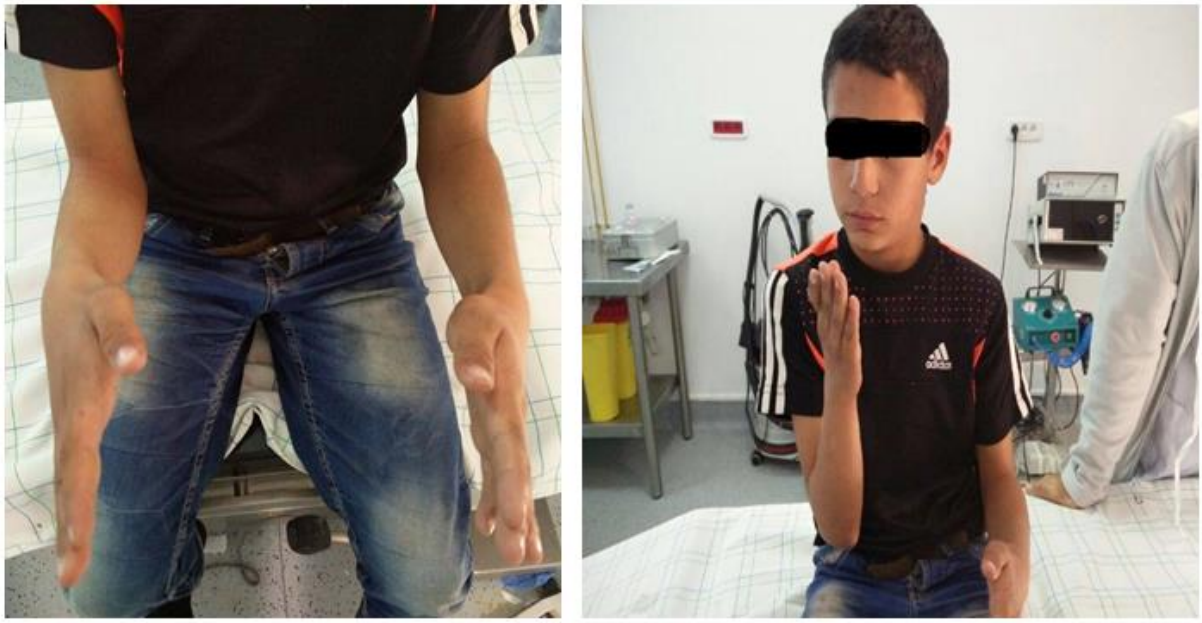

Fig 4: One year post-surgery, Case 3's appearance demonstrates a satisfactory correction angle of 45° in pronation

Table 4: Position of the forearm after surgical correction according to the authors

Authors	**Unilateral**	**Bilateral**	
		Dominant	**No dominnat**
Simmons [4]	15° pronation	10°-20° pronation	Neutral position (0°)
Castello [18]	0° to 15° of pronation for all cases		
Ramachandran [24]	10° of supination for all cases		
Our Sries	Neutral position (0°) - 20° pronation	20°-25° pronation	Neutral position (0°) -30°supination

CONCLUSION

In conclusion, our study advocates for derotation osteotomy at the synostosis site as the preferred surgical approach due to its predictable outcomes and lower complication rates, even though it may not fully restore forearm pronosupination. Surgical intervention is recommended for any forearm fixed at more than 60° of pronation, ideally before the child begins school. Conversely, surgery is less warranted in cases with less than 20° of pronation. For cases falling between 20 and 60° of pronation, the decision for surgery should be made on an individual basis. To mitigate the risk of neurovascular complications, it is prudent not to rotate the forearm more than 90° at any one time during surgery. Furthermore, post-operative vigilance is crucial to ensure adequate limb vascularization and early detection of any complications. These recommendations aim to optimize surgical outcomes and enhance the overall well-being of patients with synchronous radioulnar capitellar coalition.

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