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Congenital Radio-Ulnar Synostosis (About 7 Cases)

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Abstract: Congenital radio-ulnar synostosis is rare deformity, causing limitation of prono-supination. Often discovered at school-age. The forearm is mostly fixed in position of pronation. The transverse rotational osteotomy through the fusion mass the intervention that we propose. Objectives of our study remind clinical data, describe the techniques used for therapeutic management and compare our results with those of the literature. This is a retrospective analysis of 7 cases of congenital radio-ulnar synostosis followed and treated at the pediatric orthopedic department of the hospital of children of Rabat between January 2006 to February 2017. In the results of our study, we have the average age who is about 32.5 months with predominance of male; the involvement is bilateral in 57.7% of cases and unilateral in 42.8% of cases. According to the Cleary and Omer classification, 57.1% of cases were classified as type III, and 42.8% of cases were classified as type IV. In all cases, the forearms were fixed in position of pronation. Regarding the treatment, 10 forearms were operated consisted on a rotational osteotomy through the fusion mass, without any postoperative complications.
Keywords: Radio-ulnar synostosis - prono-supination – osteotomy.

INTRODUCTION

Forearm synostosis is a rare condition characterized by proximal radio-ulnar fusion, responsible for the disappearance of prono-supination movements, with a forearm blocked mostly in the neutral position or in hyper-pronation.

The etiology of synostosis of the forearm remains unknown, a genetic cause is suggested in front of the existence of family histories or its more frequent association with genetic diseases like the syndrome of Apert, William or Klinefelter [1].

A longitudinal defect of segmentation occurring early at the 7th week of fetal development would be at the origin of the malformation, responsible for the persistence of a fibrous or bone bridge between the radius and the ulna, secondary to an anomaly of the path of the posterior interosseous artery.

Since synostosis definitively fixes the skeleton of the upper forearm in a neutral position relative to the humeral paddle, the difference in growth between the arterial tree and the radius in the subsequent development of the skeleton is responsible for the usual pronation of the known forearm in this anomaly [2].

It associates anomalies of the radial head which can be hypoplastic, dislocated or for some absent authors and which is at the origin of various classifications. However, this does not change the handling of the malposition.

MATERIALS & METHODS

It is a retrospective mono-centric study made through the analysis of 7 cases of congenital radio-ulnar synostosis treated in the orthopedic and reconstructive surgery department "B" of the Rabat Children's Hospital. During a period from January 2006 to February 2017.

In our series, the extremes of age ranged from 5 months to 8 years with an average age of about 32.5 months (2.7 years). No cases occurred before the age of 5 months. 4 patients presented between the age of 5 months and 2 years, about 57.1%. 2 patients consulted between the age of 2 years to 4 years, or 28.5%. Only one case, 14.2%, presented in consultation between the age of 4 years and 8 years.

We were able to identify: 4 male cases, 57.1% and 3 female cases, or 42.8%. There was a slight male predominance of CRUS in our study. The unilateral involvement was found 3 times, ie 42.8%. For all patients with unilateral involvement, the left side was always concerned, ie 100%.

Right-sided involvement was not noted in any patient with unilateral involvement. Bilateral involvement is recorded in 4 cases, 57.1%. 4 patients

had type III CRUS (57.1%) and 3 patients had type IV CRUS (42.8%) according to the Cleary and Omer classification [3]. No patients had a type I or type II SRCC.

All patients (the 7 cases) presented in consultation for a deficiency of the prono-supination, the forearm for all these patients was always fixed in position of pronation according to different degrees (from 60 ° to 90 ° pronation for most patients). In no case was the forearm fixed in supination.

Only one patient had a poly-malformative syndrome, associating agenesis of the 5th finger of both hands, agenesis of the 2nd fingers of both feet, and congenital glaucoma. For the rest of the cases the CRUS was isolated without any associated lesions. No case, in particular, presented a similar case in the family. However, first degree consanguinity was noted in 2 cases.

The reason for consultation in all the patients was a functional discomfort with a limitation of the prono-supination noted by the parents or the teachers, which are at the origin of a handicap in the current life and difficulties with school, besides they report the notion of abnormal gestures made by their children when handling objects.

No cases were diagnosed at birth during routine clinical examination performed at the delivery room. On the other hand, the functional discomfort felt by the patients was judged according to the GRIFFET classification [4]: important in two cases (discomfort with writing and feeding), average in three cases (discomfort with food), 2 cases could not be classified because they are two infants, no patient had a discomfort considered minimal.

The other functional tests were not used in our study. Clinically, in 6 patients in our series the forearm was fixed in pronation position, forearm position varied from 45 ° to 90 ° (mean of 70 °) pronation, noted only one case which had a forearm fixed in 0 ° neutral position.

The elbow flexion-extension was altered in 2 cases: in one case, the flexion was reduced by 15 °; in the other, the extension has been reduced by 20 °. The rotational hyper-mobility of the wrist (compensatory pronoun-supination) was present in all cases, with a mean mobility of 40 °. The flexion-extension of the wrist was normal.

In our series, 5 children had compensatory shoulder abduction and reversed hand position to compensate for pronounced supination deficiency. In infants, shoulder abduction was absent age. None of the patients complained of pain.

RESULTS

We performed 10 procedures in 7 patients (10 forearms), the average age at surgery was 4 years and 7 months with extremes of 18 months and 8 years. The intervention consists of performing a derotation osteotomy of the anti-brachial skeleton. A posterior longitudinal incision of 3 cm on the outer edge of the olecranon, first allows to identify the synostosis (Figure 1). A Kirschner wire is inserted laterally, distally to the olecranon growth cartilage and pushed into the medullary canal. The periosteum is detached on the rugin to expose the synostosis.

The Kirschner wire is retracted to perform a subperiosteal osteotomy with oscillating saw, horizontal in the proximal half of the synostosis. Then we realize the desired rotation of the member. Our derotation position of the anti-brachial skeleton is stabilized by a second obliquely directed Kirschner pin (Figure 2). The absence of vascular disturbance noted in this new position, allows then the making of a plaster BABP, for a period of 6 weeks (Figure 3). Monitoring for 48 hours made it possible to look for possible neuro-nervous disorders.

The position of the forearm after the derotation osteotomy, for patients with unilateral involvement (3 cases) is between 0° (neutral position) in 2 cases at 20° of pronation in one case. For patients with bilateral CRUS (4 cases): the position of the forearm after the derotation osteotomy is 20° (in 2 cases) to 25 ° (in 1 case) of pronation on the side dominant, in 1 case the dominant forearm was not operated because it was fixed in neutral position.

For the non-dominant side the position of the forearm was placed from 0 ° (neutral position) to 30° supination (0° in 1 case, 20° in 1 case, 25° in 1 case and finally 30° in 1 case). No postoperative complication was noted with good satisfaction of all patients both functionally and aesthetically.

Fig-1: Intraoperative image objectifying proximal radio-ulnar synostosis

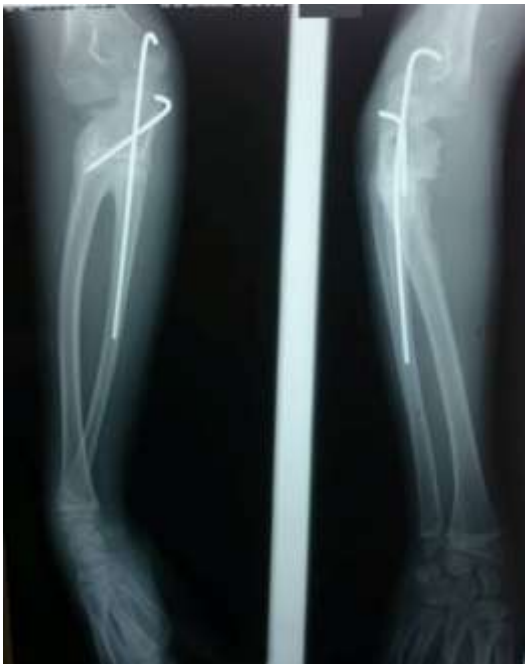

Fig-2: Postoperative radiological aspect after transverse derotation osteotomy

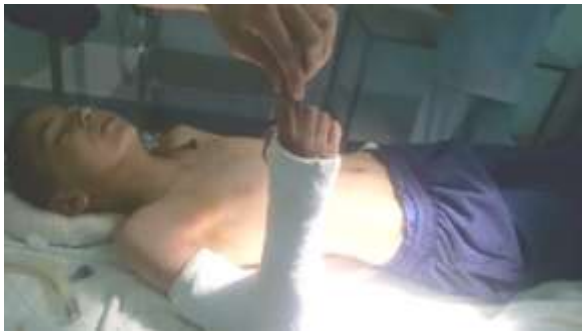

Fig-3: Brachio-antebrachio-palmar cast in correction position

DISCUSSION

According to the literature, CRUS is bilateral in 60 to 80%, with male predominance. However, it is often sporadic and isolated without associated lesions [3-7]. The circumstances of discovery are variable: It can be recognized very early, from birth, by a systematic clinical examination of the prono-supination of the two elbows [8]. During a checkup done in a young patient carrying a polymalformation [9].

The diagnosis may be fortuitous during a radiological assessment following a trauma [10]. In the case of an infant, it is often the mother who notices that her child has a certain difficulty in turning his hand or that he is incapable of this movement [11].

Although this malformation is present at birth, it is often not discovered until late in school age children [11]. Functional deficits associated with CRUS depend on the severity of the deformity, its bilateral character or not and also depending on whether the affected side is dominant or not, in the less severe forms the function of the forearm can be satisfactory due to compensation by the joints above and below (shoulder and wrist), where a good tolerance, it is often in adolescence for aesthetic or functional reasons that the patients come to consult. Children with bilateral involvement and more pronounced pronation tend to consult earlier [12].

Clinically, the main symptom of CRUS is the limitation of prono-supination [13]. The position of the forearm under synostosis is variable. it is rarely and even exceptionally fixed in supination, most often the forearm is blocked in a neutral positron or in a hyper-pronation [8].

Imaging is essential to establish the diagnosis and draw up an assessment of the lesions with a view to proper management of CRUS. Imaging should focus on establishing the diagnosis of synostosis and its topography, studying the features of synostotic bridges, evaluating the impact on adjacent structures, investigating associated abnormalities and ensuring post-therapeutic follow-up.

Synostosis imaging is essentially based on standard radiography and 3D CT [14]. Occasionally, MRI of the proximal radioulnar joint may reveal a cartilaginous synostosis that has not yet ossified or a fibrotic attachment that restricts movement, in a patient with limited forearm rotation and normal radiographs [15].

Forearm synostosis is often well tolerated functionally because it is compensated for by the mobility of the underlying and underlying joints. Its repercussions depend on the uni- or bilateral character of the malformation, the attack on the dominant side or not, the degree of deformity, and the effectiveness of compensatory movements of the shoulder and wrist [16].

Even in cases of bilateral involvement, both hands can be perfectly functional if they are fixed in a position of complementarity with each other [17]. It is the importance of functional discomfort that must guide the therapeutic indication, especially as the complication rate of this surgery is high, sometimes reaching up to 25 to 30% of cases. It is essentially a risk of vascular or nervous pain due to excessive stretching during derotation and a high risk of lodge syndrome [18].

However, indications of surgical treatment of CRUS are controversial and have changed in different studies: for some authors it depends on functional occupations, for other authors it depends on functional limitations, the uni or bilateral character, whether is on the dominant side or not, of the social and cultural environment of the patient and future planned activities [12].

In our series, it was the importance of functional impairment that conditioned the therapeutic indication. Starting from a neutral prono-supination (0°), a pronation fixed at more than 60° is a surgical indication, especially if it is the non-dominant side. Between 20 and 60° of pronation, the indication is relative and to be evaluated case by case. In the case of impairment of less than 15-20° pronation, functional discomfort is minimal and does not require surgical correction, since compensation is easy at the shoulder.

Most authors agree for early surgery at the age of 3-4, because at this age the osteotomy is easier so you can get a sufficient remodeling of the radius and ulna. Functional adaptation is also better in a young child and postoperative complications were higher in the older age group [7]. In our series, the average age at surgery on 10 procedures was 4 years and 7 months consistent with literature data.

The agreement is unanimous on the futility of surgical interventions to release synostosis in the hope of rendering a prono-supination active. The interosseous membrane, muscle contractions, atrophy of the pronator and supinator muscles, the radial head and the abnormal lower radioulnar joint are opposed [19].

Several operating procedures have been suggested, they can be divided into two main groups:

- Interventions that improve the fixed position of the forearm in a more functional position (derotation osteotomies): there are several but the best known and most used being the transverse derotation osteotomy of the synostosis which, at At present, it seems to have many advantages over other techniques, it aims to put the hand in a better functional position. This procedure is easier than trying to change osteotomy rotation to other levels and preserves good coaptation of the divided extremities and rapid consolidation of the osteotomy site.
- This technique was first described by judet [20] in France and then was repeated by the American authors: Green.WT and Mital.MA [21] who performed a transverse osteotomy in synostosis in thirteen patients.

The result obtained by this method was satisfactory with a much lower complication rate compared to other techniques. They concluded that this method is safe, easy and effective for the treatment of CRUS. This technique was adopted by several authors:

Simmons.BP [5], Castello.JR [7], Griffet.J [4], Hankin.FM [22] and Ogino.T, Hikino. K [23].

Interventions aimed at restoring the prothrombinating motion of the forearm by releasing synostosis: This technique is based on restoration of motion by resection of synostosis and interposition of a fat or muscular. However, this procedure led to unsatisfactory results with recurrence in all patients operated by the authors who described it. [24]

The ideal position to place the forearm after surgical correction remains controversial. The ideal position depends on whether the deformity is unilateral or bilateral, whether the affected side is dominant or non-dominant, the social and cultural environment of the patient, and the projected future activities.

Historically, a high incidence of postoperative complications has been reported [3,5], reaching 36% of cases in some series, these complications include: loss of rotation after surgery; neurovascular compression, which may sometimes require fasciotomy or reduction of derotation; Volkman's syndrome; caliciousness; shortening and angulation of the forearm, and temporary radial paralysis (posterior interosseous nerve).

Careful monitoring of pain, tenderness and finger motility should be performed postoperatively in order to early detect a complication, particularly a log syndrome, which would require urgent surgical revision, prophylactic fasciotomy or resection a segment of the synostotic bone reduces the incidence of this complication [18]. The appearance of vasculo-nervous disorders postoperatively requires decreasing the derotation, it can be increased again gradually thanks to the external fixator or a surgical recovery a week later.

In our series, none of these complications were met, no patient had any significant complication. Probably because we did not in any case perform a derotation osteotomy of more than 90 ° in a single time, the skeleton of the forearm was each time shortened in the osteotomy area to relax the soft tissues and we carefully checked the limb's vascular state and the sensibility and motility of the fingers postoperatively in order to early detect any complications.

The average duration of follow-up in our patients being 1 year. During the last postoperative examination, we achieved a satisfactory functional position of the forearm in all operated patients:

- forearms were in 20° pronation, and only one forearm was in 25° pronation.
- forearms were between 20° to 30° supination.
- forearms were in neutral position (0°).

The results were rated as satisfactory for all patients and their families on both functional and aesthetic levels. Patients no longer had disabilities in daily and school activities and the position of the forearm was considered ideal; There was no loss of correction, we observed no degeneration in the elbow or wrist. Bone healing was achieved after 8 weeks in all 7 cases.

Conflict of interest

The authors declare that they have no conflict of interest.

CONCLUSION

Congenital radio-ulnar synostosis is a rare, often bilateral malformation with predominance in the male sex. It is caused by segmentation failure between the radius and ulna caused by an abnormality in the path of the posterior interosseous artery.

The etiology of CRUS remains unknown, a genetic cause is suggested in the presence of familial histories and the presence of this anomaly, sometimes in the context of certain genetic syndromes, but sporadic cases remain the most frequent.

The clinical expression of CRUS is the blocking of prono-supination, the forearm is often fixed in pronation position, functional adaptation is possible thanks to compensatory movements in the shoulder and wrist. The surgical indications are based on the degree of deformation and the bilaterality of the attack but especially on the importance of the functional discomfort.

Management is either conservative or surgical depending on the degree of functional tolerance of patients. Therapeutic indications commonly adopted by the authors are the case of hyper-pronation forearm blockage greater than 90 ° and the bilateral nature of synostosis.

Based on our study, we conclude that cross-brachial skeletal derotation osteotomy is a safe, easy and effective technique for the treatment of CRUS with less risk of complications, relapse rate and loss of derotation.

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