

## Congenital Radioulnar Synostosis

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### ARTICLE INFO

Recived article: 13-07-2021

Accepted article: 24-08-2021

Published article: 30-08-2021

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### ABSTRACT

A uncommon finding is to attend for the orthopedic a patient with synostosis cubital radio congenital proximal, appears a few times and it is characterized for her limitation prone - supination, generally bilateral and of congenital origin. the article's purpose is the presentation and this one analysis common bit affection. We presented two patients with lesions, one bilateral and the other at one stroke member, usually they appreciate the few clinical manifestations, being to accomplish impossibility her prono - supination the more showy. Conclusions: proximal radioulnar synostosis is anon frequent affection that brings about difficulties carrying out actions with the hands, Its diagnosis is relatively simple by means of the clinical findings and plain X-rays of the elbow.

**Keywords: congenital radioulnar synostosis, diagnostic radiology.**

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### Introduction

The lack of separation of proximal and of the radio and ulna is a non frequent disorder that causes rigidity in the pronation position of the forearm with variable degrees. It generally occurs due to the longitudinal stop of the segmentation process of the bone in the proximal part of the forearm. This rare congenital defect is detected between the four or five years of age when the functional requests increase. Frequently this disability is compensated by the supination of the shoulder's rotation. <sup>(1)</sup>

The anatomic lesion is represented is the bone fusion between the ulna and the proximal radio, that causes fixed pronation of the limb, the trabecular framework between bones is visible. In other cases, this fusion can be only fibrous, the ulna has a normal aspect, but the radius is arched on its axis; sometimes it has a prominent head, atrophic or it does not exist. In relation to muscle lesions atrophy of pronators and supinators or the is absences be seen. <sup>(2)</sup>

### Case Presentation

**Patient's information (I).** 6 year old child with a family history of a maternal uncle and a mother's uncle who had a history of decreased mobility of the forearm. The child

presented difficulties when brushing his teeth and back handed position when placing bottles and toys. There is no previous history of trauma, pain or swelling.

**Clinical findings.** Child with fixed pronation of both forearms in a prone position between 15- 20 degrees. Moderate hand disability for everyday life, shortening and bending of forearms, hypermobility of the wrist and painless restriction of motion

**Diagnostic evaluation.** Plain radiology offers a safe diagnosis of affection in both elbow. Fusion of bone of forearm in proximal zone and also, the bending of radius's diaphysis.



Figure 1. Note the non existence of radial head in the antero lateral view.



Figure 2. Lateral view. There is fusion of radius and ulna, type III synostosis.

**Therapeutic intervention.** Suggested conservative management according to mild synostosis, ergonomics and activity modification was included.

**Follow up and sequels.** The patient was attended in consultation for more 15 years, keeping a life close to the normality.

#### **Patient's information (II).**

**Clinical findings.** 16- yearsold male student, that come to consultation presenting difficulty to perform certain motion with movements with the left upper limb. His mother said that he makes strange movements with the left upper limb since childhood but has never hindered him to have life.

**Diagnostic evaluation.** On examining it was notice a fixed prone position of the forearm, with a loss of the muscular contour when compared with the right upper limb. No pain.

When suggest to make some movements, it was found ankylosis in discreet pronation of the left-hand forearm. Simple X- rays of both elbows were takes and compared. There was a trabecular bone without articulation between the radius and compared. These was a trabecular bone without articulation between the radius and the proximal ulna, a vestige of the head of radius was appreciated.

**Therapeutic intervention.** Management was only rehabilitative to promote compensatory movement of the shoulder and to compensate for ankylosis in the radioulnar portion of the left elbow.

**Follow up and sequels.** The prognosis is good.



Figure 3. Note of the left elbow proximal radioulnar synostosis, and observe the difference of a normal elbow in other X- rays.

#### **Discussion**

The pathology presented and named as proximal radioulnar synostosis is also known as Sandifort- Lennoire deformity, the first autor recognized the pathology in 1793 and the second understood it as a cause of sickests. <sup>(3)</sup>

Etiology of the forearm begins as a single cartilaginous bud and divides from distal to proximal into the radius and ulna at week seven in the maternal uterus, so failure in differentiation results in synostosis in proximal aspect of the forearm. Frequently there are other syndromes (30%), for example, Apert syndrome (acrocephalosyndactyly), Arthrogryposis and Carpenter's syndrome (acropolysyndactyly). <sup>(4)</sup>

The description of the development that the embryology of the upper limb bud arises from the unsegmented body wall at weeks four. The elbow becomes visible at weeks five, initially, the three cartilaginous buds of the humerus, radius, and ulna are connected before segmentation. Therefore, the radius and ulna share a common perichondrium. Abnormal events at that time may lead to a failure of segmentation. Duration and severity of the insult may determine the degree of subsequent synostosis. According to these pathogenesis explanations, because the detainment of development is contradictory due to the fact that in the initial stage both bones are joined to their cartilaginous out line, but in supination, while fusion can be seen in pronation. <sup>(5)</sup>

Clinically, the elbow goes forward with a variable degree of pronation, totally blocked, in contrast, there is integrity of the flexion and the extension. The differential diagnosis is among acquired, traumatic or infectious synostosis. <sup>(6)</sup>

There is no pain, it is commonly asymptomatic recognized by parents and teachers. The child suffers from difficulties to perform specific tasks, like using a keyboard, failure for supine activities a deficient pronation, when eating, washing his face, catching a ball.

Standard age for clinical examination is at age 6 years , because in smaller children signs and symptoms may be unnoticed until early teenage, especially in unilateral cases. The motion of the elbow is usually reserved, there is fixed pronation of the forearm around 30° commonly; there is compensatory abduction, the motion of the shoulder compensates the loss of pronation with active abduction, and there may be also hypermobility of the wrist. <sup>(7)</sup>

Three kinds of congenital proximal radioulnar synostosis, in type I there is no head of the radius and there is complete and uniform fusion between the radius and ulna; it is the most severe lesion. In type II the higher portion of the radius is present but more or less deformed and there is fission of both bones in the neck of the radius. In the type III, the head of the radius is deformed and subluxed and the fusion occurs in the proximal area of interosseous membrane.

Clery and Omer Classification is based on appearance of the synostosis and radial head reduction, Type I- Lacks of bone involvement, reduced normal appearance of radial head, Type II- Distinct bone synostosis, but otherwise normal findings, Type III- Distinct bone synostosis with hypoplastic and posteriorly dislocated radial head and Type IV- Short osseous

synostosis, anteriorly dislocated radial head, usually with a mushroom shaped deformity.

Wilkie Classification offers 2 types.

- Type I: Lack of proximal portion of radius, bone fusion of 3-6 cm, and radius and ulna are connected at medullary canal.
- Type II: Normal radius, synostosis is located just distal to proximal radial epiphysis and the radial head is dislocated anteriorly or posteriorly.

The treatment is under discussion because the need of practice some motion or procedure, is question need because the lack of supination is compensated minimally with the shoulder's rotary motion.<sup>(8)</sup>

In children with bilateral congenital radioulnar synostosis, surgeons have traditionally recommended the reposition of the forearms in supination. However, the author considers that this position is not advisable nowadays because working with the computer's keyboard stops a bilateral pronation. In general the surgery is not indicated, due to the adaptation that offers the abduction movement of the shoulder.<sup>(9)</sup>

The recommendation of resection the proximal portion of the radius must be accompanied by the resection of the bone membrane along the ulna, but it is also necessary to operate the soft parts. There are other surgical interventions as Galeazzi's operation, another surgical possibilities as Kelikian's intervention or Palagi's intervention.<sup>(10)</sup>

Pasupathy B, Tholgappiyan T, Sureshbabu M, assessed the functional outcome using double rotation osteotomy and osteotomy at synostosis site in congenital radio ulnar synostosis, but the double osteotomy at both radius and ulna should be reserved as a choice for older children with bilateral hyperpronation deformity.<sup>(11)</sup>

Surgical treatment would be indicated if there is a severe deformity in pronation that causes serious functional failures.

## Conclusions

Congenital proximal radioulnar synostosis is a non-frequent affection, that causes restriction for the performance of actions with the hands. It's diagnosis is very simple by means of clinical procedure and plain simple radiology of the elbow. There are several methods of treatment, but in the results are not satisfactory. It is a well tolerated condition therefore, the behavior in both cases was conservative and expectant. The publication's principal contribution is presentation two - cases too little frequent.

**Contribution of the authors:** the authors assisted the cases and wrote the paper and analysis of the documents.

**Conflicts of interest:** there are no conflicts of interest.

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