SURGICAL TREATMENT OF CONGENITAL RADIOULNAR SYNOSTOSIS IN CHILDREN: A CASE REPORT

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ABSTRACT
Congenital radioulnar synostosis is a rare condition, only 400 cases have been reported in the world literature. Often bilateral, it can be recognised from birth by a clinical examination if it is attentive (examination of the pronou-supination of the two elbows). It is in fact often discovered later, especially in unilateral forms, in children of school age. The proximal congenital radioulnar synostosis often results in functional, cosmetic limitations of the upper limb, especially in the bilateral forms. Rotational osteotomy through the synostosis site is the usual procedure. The techniques and sites envisaged for this surgery are numerous, with multiple difficulties, risks and possible complications. We propose the observation of an eight-year-old boy with an asymptomatic form of proximal radioulnar synostosis. The surgical treatment consisted of a resection of the synostosis with rotational transverse osteotomy of the two bones of the forearm completed by a plaster in a neutral position. Clinical results, evaluated at an average of twelve months postoperatively, were found to be satisfactorily on aesthetic and functional levels. The aim of our work is to highlight our straightforward and reliable surgical procedure of the osteotomy of the synostosis maintained by spindles, allowing the functional improvement of the movements of the forearm of our child and thus a healthy life.

KEYWORDS: Radioulnar synostosis, Osteotomy, forearm

Introduction
Synostosis, or bone union, of two adjacent bones, may involve any part of the upper end. In 1793, Sandifort provided the initial description of congenital radioulnar synostosis. This condition results from the failure of the radius and the ulna to separate [1, 2]. The synostosis between the radius and the ulna can be either congenital or post-traumatic.

Synostosis of the forearm is a rare disorder characterised by proximal radioulnar fusion, responsible for a severely limited forearm rotation, usually blocked in a neutral position or pronation. The aetiology of forearm synostosis remains unknown, a genetic cause is suggested in the presence of family histories or its more frequent association with genetic diseases such as Apert’s syndrome, William’s syndrome or Klinefelter syndrome [3].

This genetic cause is found in 25% of cases. However, there are sporadic cases [4, 5, 6]. A failure of longitudinal segmentation occurring at around the seventh week of fetal development is the cause of the deformity, which is responsible for the persis-
We performed a rotational osteotomy of the forearm skeleton. With this condition, including transverse rotation osteotomy and reed osteotomy [8], we stabilised our rotation position of the forearm skeleton with a posterior longitudinal incision of 3 cm on the outer edge of the olecranon first allowed the synostosis to be exposed (figure 3). A Kirschner spindle was inserted laterally, distally of the olecranon growth cartilage and pushed into the medullary canal. The periosteum was detached to expose the synostosis. The Kirschner spindle then retreated enough to perform a horizontal subperiosteal osteotomy of the forearm skeleton. Then, we rotated the forearm at 90° to have it in the neutral position with the elbow bent at 90°. A second obliquely directed Kirschner spindle (figure 4) stabilised our rotation position of the forearm skeleton. Plaster was made for the duration of 6 weeks. The patient recovered and discharged two days after the surgery with no vascular nor nervous complications. With a follow-up of one year, the postoperative angle of pronation was on average 45°. The improvement of the preoperative condition was on average 45° (the average of modification satisfactory is between 35° to 70°). Our osteotomy easily consolidated within ten weeks and no complications were found, which allowed removing the pins.

**Discussion**

The radioulnar synostosis fixes the skeleton of the upper part of the forearm in a neutral position. The difference in growth between the arterial tree and the radius in the later development of the skeleton is responsible for the natural pronation of the forearm known in this deformity [9]. It is associated with abnormalities of the radial head, which may be hypoplastic, dislocated, or for some authors absent which is at the origin of various classifications and whose mechanism is poorly elucidated [3]. The fixed position of pronation observed in most cases was explained by Wilkie’s studies [10], which showed that the forearm of the young embryo is usually in an intermediate pronation position and that the failure of segmentation makes it remain permanently in that position. In patients with unilateral synostosis such as the observation of our patient, shortening is observed in 32% of cases [11], which is very high compared to the other series [4, 9, 12]. Functional disorders are not always correlated with the degree of fixed position of the forearm, as compensatory motion is observed with age. This adaptation is weaker when the forearm is blocked in hyper-pronation (angle of prono-supination> 90°) or the case of a bilateral synostosis. There was no correlation between the fixed position of the forearm and radiological signs, as pointed out by Cleary and Omer [9]. Functional tolerance is excellent when the hand is blocked in an intermediate position, which probably explains why many of these cases of congenital radioulnar synostosis remain undiagnosed [7]. Surgical treatment is indicated in two circumstances: A) When the forearm is fixated in pronation beyond 90° (PS>90°), the treatment is designed to restore a better position of the hand. B) When the deformity is bilateral, the treatment will improve daily functional activities. In both cases, the disability must be severe before considering surgery. For some authors, surgery is justified whenever the forearm is blocked at an angle of 60° of pronation. We will consider this attitude to be rather abusive, especially considering the remarkable adaptive capacities and functional tolerance of these patients. The optimal age for surgery is hard to define. In the Yammine series [11], children were operated on at the age of 4 years, five years when...
Intraoperative image is objectifying proximal radio-cubital synostosis. After performing the desired rotation, a second oblique pin was positioned bridging the osteotomy site to fix the rotation permanently. We chose the use of pins to stabilise our osteotomy rather than the screwed plate, because of the ease of insertion and removal, and according to the choice of the majority of recent series. This therapeutic method proved its efficacy with no observed complications compared to the results of the other series of the literature using other methods sometimes more invasive with vertical or horizontal trans-synostotic osteotomy. Simmons et al. [5] reported a rate of 36% of complications such as a lodge syndrome, paralysis of the ulnar nerve or lesions of the median nerve. Among the other therapeutic possibilities, the osteotomy of the two diaphyses, called “reed” for radius and transversal for ulna, as in the series of Yammine et al. [11], imposes wide approach, with a risk of hematoma and postoperative edema [7], and remains difficult to perform on such small sized diaphysis. In the case of conservative management, there was an increase in the risk of fracture of the forearm, the anterior brachial frame being vulnerable to torsional trauma especially in sports. [17]

Conclusion

Congenital radioulnar synostosis is a rare malformation, the management of which is either conservative or surgical depending on the degree of functional limitations. The therapeutic indications commonly adopted by the authors are any forearm fixated beyond 90° of pronation and the bilateral forms. Our technique, the rotational transverse osteotomy through the synostosis site has proved its effectiveness with an average follow-up of more than one year, regarding complications, relapse rate and loss of derotation.

Authors’ Statements

Competing Interests

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

There were no financial support or relationships between the authors and any organization or professional bodies that could pose any conflict of interests.

Authors’ contributions

All authors contributed to the writing of this manuscript, all read and approved the final version.
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Written informed consent of the patient and her guardian was taken for publication of this case report.

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