

Congenital Radioulnar Synostosis and Radial Head Dislocation with Elbow Joint Blockage: A Case Report

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Abstract

Background: The proximal radioulnar joint is a syndesmal joint which stabilizes the forearm during pronation and supination, and the elbow during flexion and extension. Radioulnar synostosis is a congenital condition in which this joint is abnormally merged and, sometimes, the radial head is also dislocated. It's mainly diagnosed in males (2:1) and the average age of diagnosis is 6 years old. Genetics is the main cause of this condition.

Case Summary: We present the case of a 16-year-old male patient, diagnosed with radioulnar synostosis with congenital radial head dislocation, who presented with recurrent events of elbow blockage. A surgical resection of the radial head was performed, allowing the patient to regain a more functional elbow range of motion (R.O.M.) and, consequently, a more active lifestyle. The synostosis was not treated since it was not the main complaint, and the patient presented good compensatory movements. He was released with no complications and came for follow up at 2-weeks and 6 months post-operative with no further complications.

Conclusion: Since this is a rare congenital condition, it is difficult to find in literature articles addressing this topic, especially how to approach and treat the disease to provide patients the maximum range of motion possible. This can lead to more case reports and further research and consensus on what the best surgical approach for these patients should be.

Keywords: Elbow Locking; Radio-ulnar; Synostosis; Joint; Radial Head; forearm; congenital.

Introduction

The proximal radioulnar joint is constituted by the circumferential radial head and an osteofibrous ring on the ulna, all joined by the annular ligament. It is a syndesmal joint, hence, its main function is to stabilize the forearm during prone-supination movements and flexo-extension of the elbow.

Radioulnar synostosis is a rare malformation of this joint, resulting from a segmentation failure of the two bones, resulting in a single bony-like structure, instead of the joint with its corresponding annular ligament, and can present along with radial head dislocation. Normal separation of the bones occurs during the 7th week in-utero.

Epidemiology

This malformation is more commonly found in males than females (3:2) and the average age of diagnosis is 6 years old. 60% of cases are bilateral.

Etiology

Genetics

Literature shows a higher incidence of the condition with familial cases, with evidence of autosomal dominant inheritance (20% with positive family history), in some cases associated with chromosomal abnormalities.

Development

Upper extremities develop between the fifth and the eighth week of pregnancy and begin as a whole with a continuum of the humerus, radius and ulna by cartilaginous bridging. During the sixth week separation and differentiation of the cartilage occurs to become 3 separate bones. Radio ulnar synostosis may appear at approximately 7-week gestation due to a lack of differentiation in the cartilage union of these two bones.

There are associated symptoms in 30% of the cases such as acrocephalosyndactyly, acropolysyndactyly, Arthrogyrosis, Mandibulofacial dysostosis, Klinefelter's Syndrome (XXY), hip dysplasia, polydactyly and talipes equinovarus.

Clinical Presentation

Typically, patients present with an inability to pronate and supinate (commonly fixed in pronation at 30°), that, however, does not interfere much with elbow function in daily activities. However, some can present difficulties performing specific tasks such as keyboard use and overall tabletop activities, which are associated with deficient pronation; eating, face washing or catching a ball, which are associated with deficient supination. Elbow flexion is usually preserved, nevertheless sometimes joint blockage may be associated, especially when dislocation of the radial head is present, further limiting flexion and extension.

In patients with congenital radioulnar synostosis prono-supination is limited to an average of 30° when the elbow is at a flexed position, however with elbow extension and compensatory motions of the shoulder (abduction, adduction and internal/external rotations) a greater range of motion in prono-supination can be achieved.

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Table 1: Cleary Classification.

Type I	Radial head reduced, no osseous synostosis
Type II	Radial head reduced, osseous synostosis
Type III	Radial head hypoplastic and posteriorly dislocated, long osseous synostosis
Type IV	Radial head mushroom-shaped and anteriorly dislocated, short osseous synostosis

Case Presentation

In this report, we present the case of a healthy 16-year-old male, with no familial or hereditary history relevant to the case. The patient refers symptoms onset at 2 years of age, with limitation for active and passive prono-supination of the right forearm without pain or limitation of other movements, such as flexion and extension. He was diagnosed at a young age (no precise age known) with congenital right proximal radioulnar synostosis and congenital radial head dislocation by the pediatric orthopedic department with no other abnormalities or findings and remained under observation. Over the last 4 years, he's presented 5 isolated events in which he experienced locking and limitation while extending the elbow from hyperflexion, especially during physical activities.

The patient was previously admitted to E.R. complaining inability to extend his right elbow due to a joint blockage in a flexed position, thus limiting his physical activity; physical examination showed a normotrophic right upper limb, with limitation of forearm rotation and inability to perform hyperextension of the elbow confirming the diagnosis. Pulse, mobility, and sensitivity were normal, with no other physical findings. X-rays (Fig. 1) showed a joint blockage due to a dislocated radial head locking with humeral condyles, which was reduced with a closed maneuver and patient was released.

Surgical treatment was proposed, consisting of excision of the radial head to avoid new joint-blockage episodes. Although many different techniques have been described we had to take another approach to resolve our patient's primary concern. The radial head dislocation and deformity would not allow for anatomical reduction and the radial head held no anatomical normal properties for functionality. Simple synostosis removal was not considered because this would not prevent from another elbow blocking to happen again. And a full radial head removal, synostosis removal would end in radial migration and instability. By only removing the radial head and not the synostosis the radius of this patient will not move and there will be no affectation to the wrist. Going for more complex treatment was limited due to lack of fundings. The patient accepted this procedure and was admitted for surgery.

The physical exam taken immediately before the surgical procedure, showed elbow extension of 180°, elbow flexion of 90°, elbow pronation of 75° and supination of 45°.

He was admitted to the operating room where he was placed in a supine position and administered general anesthesia. Proper antiseptic and aseptic technique were performed on his right upper limb and covered with surgical dressings. An anterior approach was used to expose the radio humeral joint and the radial head.

Proper care was taken to avoid damage to the neurovascular package (Fig. 2).

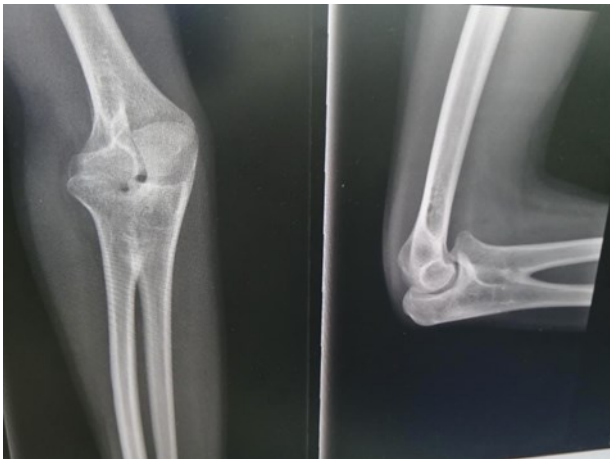


Fig. 1: AP and lateral X-ray of the elbow with synostosis of the radioulnar joint and the dislocation of the radial head which causes the elbow blocking.



Fig. 2: Sharp and blunt dissection retracting and protecting the neurovascular package.

Once the radial head was exposed (Fig. 3), a 1.2mm sagittal bone saw blade was used to excise the radial head (Fig. 4 and Fig. 5) and gel foam and bone wax were used to control bleeding. Fluoroscopic guide imaging was used (Fig. 6 and Fig. 7) to corroborate there was no extra bony edges and that flexion was achieved without blockage. 2-0 and 3-0 Monocryl suture was used for deep wound closure and 3-0 nylon for skin closure. The wound was covered with simple gauze and sterile elastic bandages.

In the immediate post-op the patient was able to achieve a 180° elbow extension and 110° elbow flexion, pronation remained unmodified due to the synostosis not being treated.

At 2-week post-op examination, wound healing was uneventful, and sutures were removed. Flexion remained at 110° with no signs of pain or blocking.

6 months after surgery our patient was asymptomatic, with elbow extension at 180°, flexion at 110°, pronation at 75° and supination at 45° with no elbow blocking. Control X-rays were taken (Fig. 8). The patient reported subjective improvement in quality of life and function. Before surgery our patient was interested in playing football but quit due to causing of elbow blockage; he was able to return after surgery. He was also allowed to go back to weightlifting.



Fig. 3: Exposition of the Radial Head (*RH).



Fig. 4: Excision of the Radial Head (*RH).



Fig. 5: Radial head and a section of the annular ligament completely removed.



Fig. 6: Intraoperative AP fluoroscopic imaging.



Fig. 7: Intraoperative Lateral fluoroscopic imaging.



Fig. 8: Immediate Post op lateral X ray image.

Discussion

According to the DASH Score Interpretation (Table 2), our patient presented a severe disability, since the elbow blocking didn't allow him to have an active lifestyle for a boy his age and participate in sports. It was also painful and recurrent, so a definitive treatment to the condition was considered adequate. In other reports of elbow locking with congenital proximal radioulnar synostosis (Nezar, et. Al) the blockage was treated with orthosis in full extension for 3 weeks and, subsequently, the annular ligament was removed. This treatment was not considered, since the orthosis showed no benefits in Nezar's article, neither for the blockage of the joint, nor the pronation and supination caused by the synostosis and had to be surgically intervened despite orthosis (*"Instantly after cast removal, the patient reported a recurrent, painful, locking elbow"*); hence we proposed a surgical approach from the beginning. Nezar et al, report 3 children with radial head excision with significantly good outcomes, and it is why we decided to replicate these techniques, and to skip the conservative treatment. They reported R.O.M in flexion and extension from 0 – 140° and we were able to obtain an elbow R.O.M from a 0° extension to a 110° flexion.

Due to the limitation in bibliography and treatment options, since some authors suggest treating both the synostosis and the dislocation while others recommend treating the dislocation alone, it is still unclear which is the better treatment options for these patients. We opted for treating only what the patient referred to limit his lifestyle and cause pain which was the elbow locking in a flexed position and leave the radio ulnar synostosis as it was. Our patient had grown used to compensate the pronation-supination movement with his shoulder so that could remain as it was. (1-6)

Table 2: The DASH Score (Disabilities of Arm, Shoulder, and Hand) is a self-administered region-specific instrument developed as a measure of and self-rated upper-extremity disability and symptoms.

DASH Score Interpretation	
0% to 20%: minimal disability	Most living activities can be fulfilled. Usually, no treat-
20% to 40%: moderate disability	Patients present some difficulties and pain while lifting, sitting, and standing. Some activities like traveling and social life in general can be limited due to the current condition. Work, personal care and sleeping are not severely affected and can be managed with con-
40% to 60%: severe disability	Pain is the main problem in this group of patients, but
60% to 80%: crippled	Back pain impinges on all aspects of the patient's life.
80% to 100%	Patients are either bed-bound or exaggerating their symptoms.

Conclusion

Since this is a rare congenital condition, there is not much information about which approach, and treatment are best to regain the most range of motion possible for the patients. This can lead to more case reports and further research and consensus on what the best surgical approach for these patients should be and when the best time to treat it.

Conflict of Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

1. Nezar B. Hamdi, Abdulmuhsen N. Alshammari, Abdullah Y. Almarshad, Faisal M. AlFayyadh, Painful Locking Elbow in a Child with Congenital Proximal Radioulnar Synostosis, *Journal of Hand Surgery Global Online*, Volume 1, Issue 2, 2019, Pages 127-130,
2. Congenital radial ulnar synostosis (2021) *Orthobullets*.
3. Congenital radial ulnar synostosis (2021) *Orthobullets*. Available at: <https://www.orthobullets.com/hand/6071/congenital-radial-ulnar-synostosis>
4. Garg, G. and Gupta, S.P. (2015) "Surgical outcome of delayed presentation of congenital proximal Radioulnar synostosis," *SICOT-J*, 1, p. 33.
5. Okotcha, E. et al. (2022) "A rare case of congenital Radioulnar synostosis," *Consultant*, 62(6). Available at: <https://doi.org/10.25270/con.2021.11.00004>.

6. VanHeest, A.E., Lin, T.E. and Bohn, D. (2013) "Treatment of blocked elbow flexion in congenital radioulnar synostosis with radial head excision," *Journal of Pediatric Orthopaedics*, 33(5), pp. 540–543.

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