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CASE REPORT

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Large Exostosis of the Distal Radius in a Pediatric Patient Causing a Dysplastic Ulna and Distal Radioulnar Joint Disruption: A Single-stage Management Approach

Albert Jerome D. Quintos, MD,[1,2] Pasquale Manuel A. Bonavitacola, MD,[2] Candice Elaine C. Lim, MD[2]

[1]*University of the Philippines Manila College of Medicine*
[2]*Department of Orthopedics, The Medical City, Pasig City, Philippines*

ABSTRACT

The distal radioulnar joint (DRUJ) is an important structure that stabilizes the radius and ulna. Any incongruency may result in limited forearm rotation and a weak grip. Exostosis is a benign bony cartilage-capped outgrowth protruding from the surfaces of affected bones. These occur as solitary lesions in 1-2% of the general population. Primary resection at the base is the mainstay of treatment for solitary exostosis lesions. In rare cases, an exostosis of the distal radius can grow particularly large leading to deformities of the adjacent ulna and DRUJ. In this case report, we present a patient with a large distal radius exostosis resulting in a dysplastic ulna with DRUJ involvement. Case reports on this rare phenomenon report good results following osteotomy and gradual lengthening of the ulna with an external fixator – a multi-stage surgical approach. In this case report, treatment involved resection, pinning, bone grafting with mesh, and the use of a fascial sling taken from the radial volar fascia wrapped around the distal ulna, resulting in near full return of function for the forearm and hand – a single-stage management approach.

Keywords. pediatric, exostosis, distal radius, dysplastic ulna, incongruent DRUJ, surgical treatment, single stage, one stage, DRUJ reconstruction

INTRODUCTION

Primary bone tumors arise from bone tissue and range in severity from benign to significantly malignant. An exostosis is a benign growth of bone that typically presents with a characteristic cartilaginous cap toward which the growth is directed. When a cartilaginous cap is present in an exostosis, they are called osteochondromas. These lesions may be solitary and spontaneous or linked to genetics with multiple affected bones. They are more commonly found in long bones such as the radius, humerus, femur, tibia, the pelvis, and shoulders. Prevalence studies show about a 1% rate of occurrence of solitary lesions in the general population.[1]

These can cause dysplasia of adjacent bones and lengthening of adjacent tissues. Deformities of the forearm, when present, are relatively common at a 30–60% incidence rate. These disruptions, particularly around the DRUJ may cause significant impairment in patients' hand function, particularly with forearm pronation, supination, and grip strength. The DRUJ is one of the primary structures that transmit load at the distal forearm and wrist. It acts as a pivot for radioulnar rotation[2] and its disruption may significantly impair function. A large exostosis of the distal radius may lead to a dysplastic ulna in the form of ulnar shortening and excessive localized bowing to accommodate the lesion.

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Corresponding author:
Pasquale Manuel A. Bonavitacola, MD
The Medical City, Ortigas Avenue,
Pasig City, 1800 Metro Manila, Philippines
Tel. No.: +63(2) 86356789
E-mail: pasquale.bonavitacola@obf.ateneo.edu

Figure 1. Pre-operative gross pictures of lateral **(A)** PA **(B)**, with pronation **(C)** and supination **(D)**. X-rays of forearm AP **(E)**, lateral **(F)** and wrist AP **(G)** and lateral **(H)** on presentation to the clinic showing gross limitations in pronation and supination and a large exostosis at the distal radius.

Treatment options for patients who have a large exostosis with DRUJ disruption and dysplastic ulna range from simple removal of exostosis to distraction procedures with external fixation of the ulna.[3] This case report presents a pediatric patient who came in with a large exostosis of the right distal radius causing a dysplastic ulna with disruption of the DRUJ, treated with resection of mass, osteotomy, and pinning of the ulna with bone grafting and mesh reinforcement all in a single-stage approach.

CASE

A 9-year-old right-handed girl with no known comorbidities presented to our service for a mass on her wrist and a limited range of motion of her right forearm. The mass started small four years before consultation without any inciting event or trauma. The mass grew over the years with further limitations in pronation and supination of the right forearm. One year prior the patient began to have difficulty lifting heavier objects with her affected hand. On consult, limited pronation (30 degrees) and supination (25 degrees) were documented. The circumference of the right distal forearm was measured at 15 cm as compared to the left at 12.5 cm. Motor strength for grip was 4/5 for the right in comparison to the left (Figure 1A).

X-rays revealed an exostosis of the distal radius abutting against the ulna resulting in a dysplastic ulna. The widened radio-ulnar interval and negative ulnar height suggested DRUJ laxity. The figures below are the pre-op x-rays taken one month before surgery (Figure 1B).

The goals of treatment were to remove the exostosis, prevent recurrence, stabilize the DRUJ, and restore the normal function of the forearm and wrist. The surgeon performed a resection of the exostosis, pining of the distal ulna, and autologous bone grafting from the radius with mesh application.

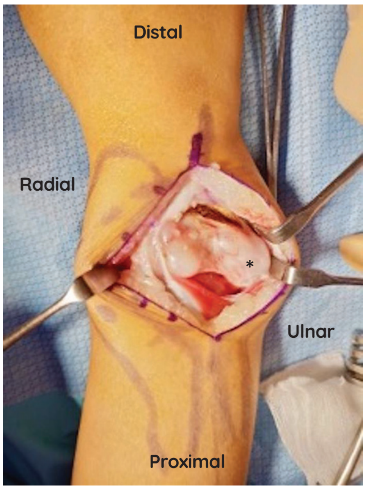

Figure 2. Gross photo of exostosis intraoperatively. *Area abutting the ulna.

A dorsal approach between the 4th and 5th dorsal compartments was used to expose the exostosis and dysplastic ulna (Figure 2). The exostosis was removed using an osteotome after stripping the surrounding periosteum ensuring that the entire cartilaginous cap was resected along with the stalk. As shown in Figure 3A, after resection of the exostosis, a dysplastic ulna was noted resulting from the abnormal growth of the resected mass. Corrective osteotomy was performed to align and restore the height of the ulna. The distal fragment was fixed to the intramedullary canal of the proximal ulna using a 0.062" K-wire, leaving a gap (Figure 3B). A bone graft obtained from the periphery of the exostosis resection site was placed over the gap at the ulnar osteotomy site and secured with a resorbable mesh (Figure 3C-D). Stripped periosteum and surrounding fascia from the area of resection (Figure 3E) was then passed underneath the extensor muscles toward the ulna, looped around the ulna osteotomy site, and secured onto itself with sutures acting as a soft tissue stabilizer recreating some function of the DRUJ (Figure 3F). An ulnar gutter splint was applied post-operatively for protection.

Post-op x-rays showed complete resection of the mass with the bone graft located at the ulnar gap (Figure 4). The patient followed up at two weeks for wound inspection then at two months with repeat x-rays showing consolidation of bone graft and incorporation into the distal ulna with callus formation (Figure 5).

Histological studies revealed an osteochondroma with no malignant potential or transformation (Figure 6). The intramedullary pin was removed at 10 weeks post-operatively (Figure 7).

There was a gradual improvement in the pronation and supination of the right forearm (Figure 8, Table 1). Serial piano key tests also showed near-symmetric stability of the DRUJ throughout the range of the forearm. On serial X-rays after removal of the pin, ulnar remodeling occurred (Figure 9). The callus bridging of the gap was achieved and the patient regained 50 degrees of pronation.

At four months post-pin removal, the patient had good supination reaching up to 80 degrees and pronation of 70

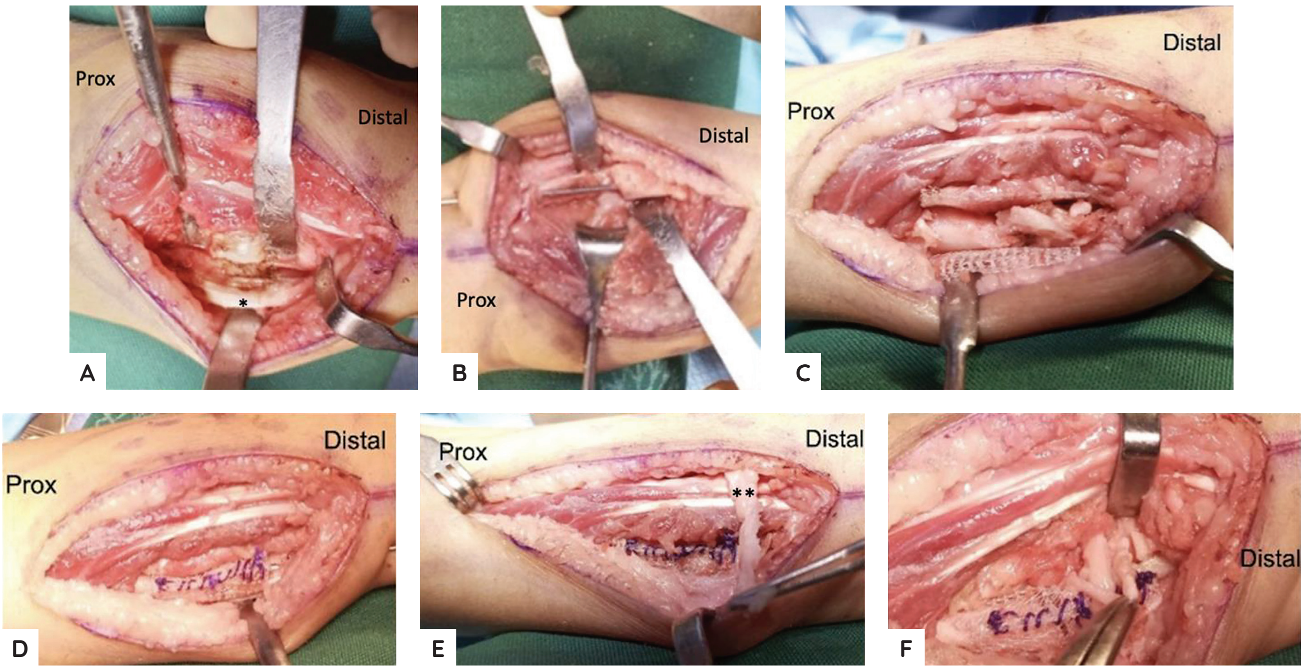

Figure 3. Intraoperative images taken. After resection of mass noting the *dysplastic flattened ulna **(A)**; pinning of the ulna after corrective osteotomy was performed to secure fragments **(B)**; application of bone graft and mesh done to fill the defect between proximal and ulnar fragments **(C)**; closure of mesh with absorbable sutures – securing the graft in place **(D)**; ** sling taken from loose periosteum and surrounding fascia from radius looped around ulna and sutured onto itself acting as a soft tissue stabilizer **(E)**; final inspection prior to closure **(F)**.

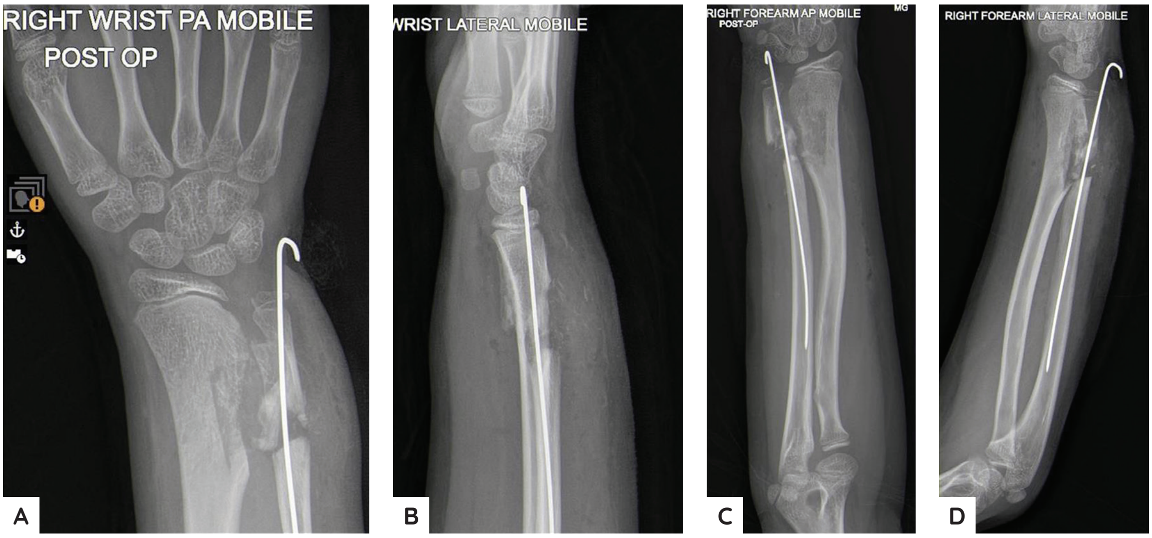

Figure 4. Post-op X-rays. Showing resected distal radius with an intramedullary pin of ulna. Bone graft shown at defect of the ulna in wrist AP **(A)**, lateral **(B)**, and forearm AP **(C)**, and lateral **(D)** x-ray views.

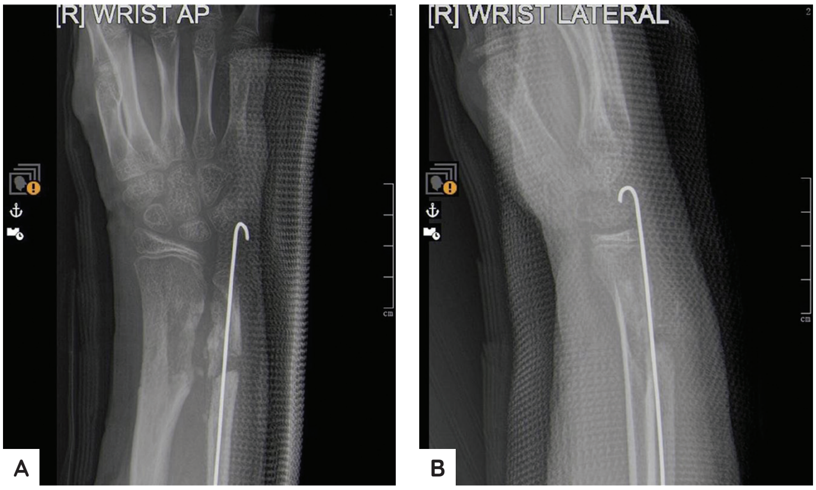

Figure 5. X-rays of wrist AP **(A)** and lateral **(B)** at 2 months follow-up showing intact pin and beginning consolidation of ulna with callus formation.

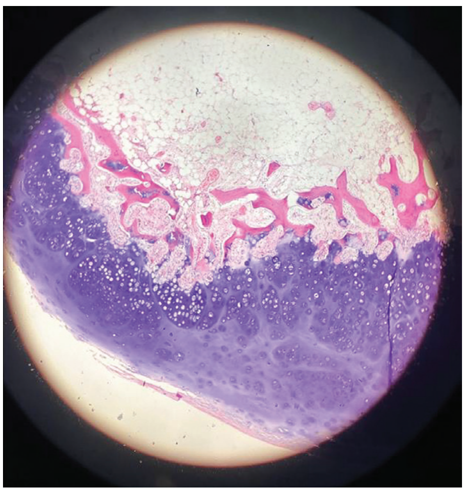

Figure 6. Histological studies revealed an osteochondroma with no malignant potential or transformation. Intramedullary pin was removed with note of some residual gapping and beginning bridging callus over area of defect (10 weeks post-op op).

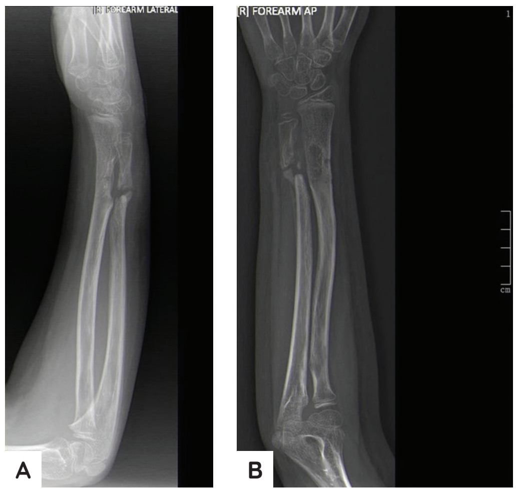

Figure 7. Forearm AP **(B)** and lateral **(A)** X-ray views. Intramedullary pin was removed with note of some residual gapping and beginning bridging callus over area of defect (10 weeks post-op).

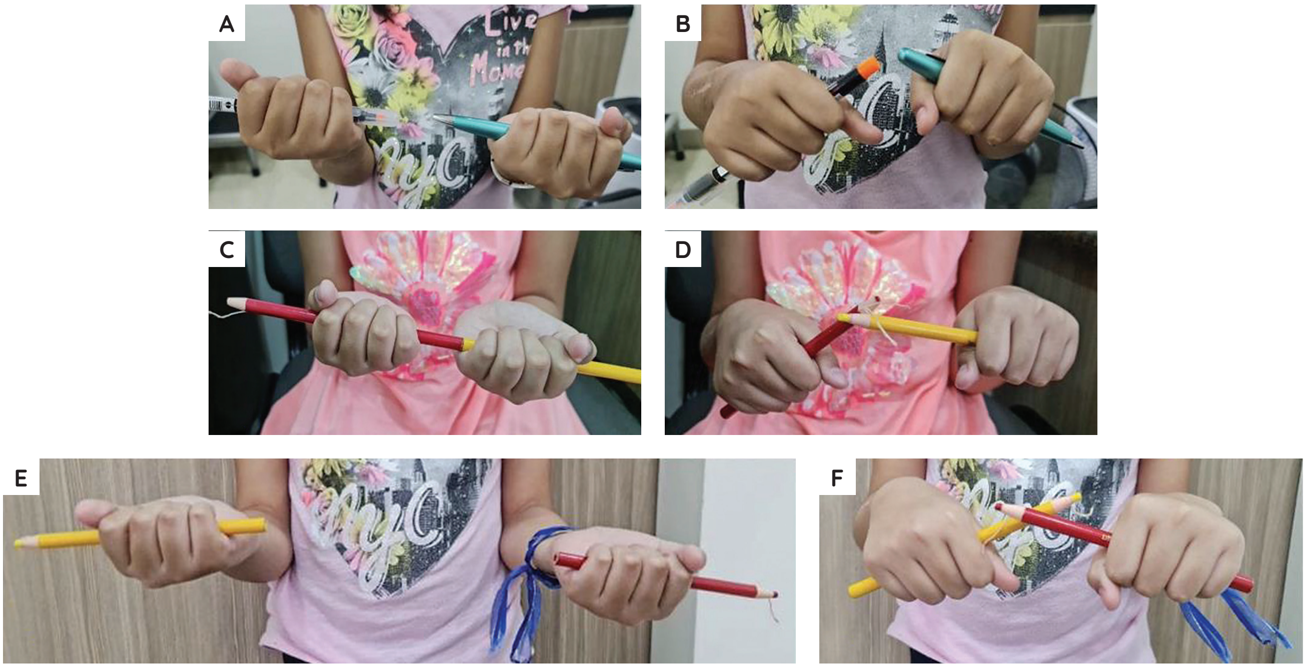

Figure 8. Gross pictures show improving pronation and supination of both forearms. Follow up pronation and supination at 2 months **(A,B)**, 4 months **(C,D)**, and at 7 months **(E,F)**.

Table 1. Range of motion tests after removal of pin measured in degrees from neutral position

Date	Range of Motion (deg)	
	Pronation	Supination
Pre-op	0-30	0-25
1 month	0-50	0-80
2 months	0-50	0-80
4 months	0-70	0-80
7 months	0-70	0-90

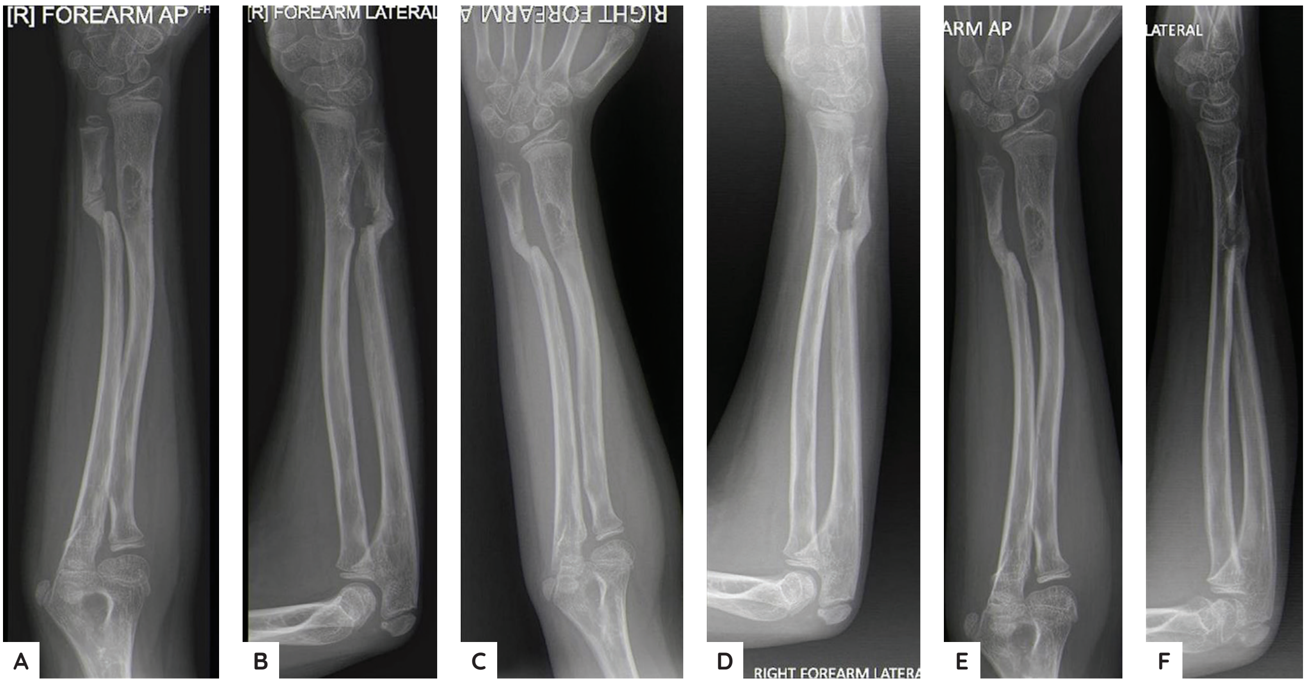

Figure 9. Follow-up x-rays at 1 month **(A,B)**, 2 months **(C,D)** and 4 months **(E,F)** after removal of pin showing bridging callus and progressive remodelling.

degrees. Grip strength also significantly improved as described by the patient and as clinically assessed with the finger squeeze test. A more objective tool such as a dynamometer would have been ideal to document this improvement. Follow-up X-rays showed union of the proximal and distal ends of the ulna, the graft had been incorporated into the defect. No local recurrence of exostosis was noted.

At seven months post-op, full supination was achieved at 90 degrees along with 70 degrees of pronation. X-rays showed further remodeling of the distal ulna now more vertical in orientation with less dorsal attitude.

Of significant note is the presence of some shortening of the ulna on long-term follow-up. Some consequences of this finding may be future instability and ulnar impingement syndrome. These would present with pain at the DRUJ, particularly on supination and pronation. Patients should be followed up regularly to monitor for this pathology. The patient reports being greatly thankful that the surgery was performed. She reported that she can better use her right hand for daily activities and is no longer bothered by gross deformity.

DISCUSSION

The distal radioulnar joint is an important structure that stabilizes the radius and ulna close to the wrist. It is responsible for the transmission of load between the bones of the forearm and the wrist. It is a diarthrodial, synovial articulation that serves as a pivot for pronation and supination between the radius and ulna. any incongruency may result in limitations of forearm rotation and weakness in grip. The DRUJ is also synchronous with the proximal radioulnar joint and is integrated with the ulnocarpal joint. Any of these adjacent joints may be dysfunctional if the DRUJ is not intact.

The sigmoid notch of the radius contributes significantly to the stability of the DRUJ. This is the area where the radius rotates about the ulna during pronation and supination. Ulnar variance describes the relation of the height of the ulna to the articulating surface of the radius. In adults the ulnar variance averages at -0.9 mm. The tip of the ulna houses the ligamentous complex that provides stability to the ulnar side of the wrist, the triangular fibrocartilaginous complex (TFCC). Two ligaments in the TFCC provide the most stability to the DRUJ and these are the dorsal and palmar radioulnar ligaments.

Not much literature is described for reconstruction of the DRUJ in patients with large exostosis of the distal radius primarily due to the low incidence of these types of lesions with the aforementioned sequelae in the general population. Several techniques have been described to address and treat these types of deformities. Treatment options range from simple resection of mass to combined procedures such as osteotomy distraction with progressive lengthening and adjustments to address the shortening of the dysplastic ulna.

The goal of treatment is to restore the stability and strength of the DRUJ – an important structure in transmitting load between the forearm and hand and in the articulation of the radius and ulna via pronation and supination. Any disruption or incongruence can lead to limitations in the range of motion of the forearm and wrist. Further enlargement of masses at the wrist may cause weakening of grip or compression of nerves along with contractures of flexor and/or extensor tendons.[2,3] Our patient presented with limitations primarily in forearm supination and pronation, only achieving 30 degrees of pronation and 25 degrees of supination pre-operatively. No objective assessment for grip strength was done but the patient had a good grip and was able to hold several objects with the affected hand.

The Masada classification system was developed to communicate the location of the mass and adjacent structures. It is typically used for patients with multiple osteochondromatosis but may have applications for use in solitary lesions as in our patient. Based on the diagram, our patient may be exhibiting a type III lesion.

Multistage lengthening is described in several papers to address the ulnar dysplasia, however, for some patients, funds issues may limit their options. Aerts et. al.[4] Described a similar case wherein an external fixator was fixed onto the ulna length and progressively adjusted to achieve the desired. In literature, most patients who undergo this type of distractive procedure are pediatric patients since their potential for bone healing and remodeling is greater. In these cases, patient outcomes were good, and return to function was achieved at six months. The decision to proceed with the technique utilized in this paper was to address both the exostosis and the DRUJ reconstruction with lengthening of the ulna in one sitting instead of a multistage approach. The idea was to create a gap and temporarily fix the distal ulna with a k-wire pin in distraction, filling the gap with autologous bone graft from the radius. In this way, no further implantation and no further manipulation of the ulna was needed on follow-ups. The bone graft would simply incorporate into the defect resulting in lengthening of the previously dysplastic ulna.

In terms of the DRUJ, the standard technique for stabilizing the joint is a trans-radio-ulnar pin in a position of greatest stability (more commonly neutral or supination). Other techniques have also been described such as utilization of a tendon graft sling looped around the radio-ulnar junction.[5] However, in cases of dysplasia of the ulna where lengthening is indicated, the option to pin through both bones distally could have aided in further stabilizing the distal fragment and maintaining the ulnar length. Intraoperatively, a fascial sling was determined sufficient to provide tension at the DRUJ. The technique described proved a viable option to achieve DRUJ stability through a single-stage approach with a dysplastic ulna. One possible complication would be the shortening of the ulna. Possible interventions to prevent this complication could be the use of a transverse pin through the distal fragment and radius to maintain ulnar length.

CONCLUSION

The described single-stage procedure may be a viable treatment option for patients with a large exostosis, dysplastic ulna, and DRUJ disruption when multi-stage procedures may not be viable. It was shown that pinning in distraction, bone grafting, and the use of a fascial sling resulted in a stable DRUJ after resection of the exostosis with the return of good forearm supination and pronation.

RECOMMENDATIONS

This paper recommends further documentation of pre- and post-op hand functional scores such as DASH Scoring, grip strength quantification, and long-term follow-up of DRUJ function with close monitoring of potential sequelae of ulnar variance.

ETHICAL CONSIDERATION

Patient consent forms were obtained before manuscript submission.

STATEMENT OF AUTHORSHIP

All authors certified fulfillment of ICMJE authorship criteria.

AUTHORS DISCLOSURE

The authors declared no conflict of interest.

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