

Treatment of neglected ulnar longitudinal deficiency associated with radial head dislocation in a 9-yr old male patient. Case-study and review of the current literature

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ABSTRACT

Ulnar longitudinal deficiency represents a challenging clinical entity in terms of treatment and functional recovery.

Case study

A 9-year old male patient was referred to our outpatient clinic due to severe shortening and ulnar deviation of his right upper limb. Range of motion of the elbow joint was limited including both flexion-extension and pronation-supination. Radiographic examination revealed absence of the distal two thirds of the ulna as well as bowing of the radius and dislocation of the radial head. The patient underwent surgical treatment involving excision of the fibrocartilaginous anlage extending between the proximal ulna and the carpus, excision of the proximal radius and creation of a single-bone forearm in neutral rotational position.

Results

At 6 weeks postoperatively bone healing at the site of the fusion was achieved. In terms of functional results, flexion and extension of the forearm were comparable to the contralateral extremity.

Conclusion

Creation of a single-bone forearm represents a viable treatment option in cases of ulnar longitudinal deficiency associated with radial head dislocation and limited pronation – supination of the forearm. Surgical intervention should be encouraged early so as to prevent deformation of the radius.

KEY WORDS: Ulnar longitudinal deficiency, ulnar clubhand, single-bone forearm.

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Introduction

Ulnar dysplasia or club hand represents a rare clinical entity, much less common than central longitudinal deficiency or radial clubhand. Furthermore, its therapeutic approach remains quite challenging in terms of functional recovery as well as cosmetic appearance. We present a case of a 9-year old male patient with neglected ulnar longitudinal deficiency and associated radial head dislocation treated with single bone forearm creation.

Case study

A 9-year old male patient was referred to the outpatient department of our clinic for further investigation of an increasing deformity of his left forearm. During initial clinical evaluation, a remarkable ulnar deviation of the right forearm was noted accompanied with upper-limb length discrepancy of approximately 7 cm compared to the contralateral side (**fig.1**). Furthermore, a bony prominence was present on the lateral side of the elbow joint suggesting dislocation of the radial head. Range of motion of the elbow joint was also affected. Pronation and supination of the forearm were limited to an extent of less than 10 degrees, elbow flexion was deficient by 40 degrees and extension by approximately 20 degrees resulting in a poorly functional upper limb (**fig.2**). No other musculoskeletal malformation was present. Numerous hyperpigmented lesions representing *café au lait* macules were also present on the patient's chest wall, abdomen and back. Following clinical evaluation, the patient underwent radiologic examination of the right upper extremity. Absence of the distal two thirds of the ulna was noted, as well as severe bowing of the radius accompanied with dislocation of the radial head suggesting a type II ulnar deficiency according to the Bayne classification⁽¹⁾ (**fig.3**). Further laboratory work up consisting of serologic, hormonal and immunologic tests turned out normal. Based on the type of the deficiency as well as the poor functional state of the affected limb, surgical treatment was proposed.

Preoperative planning

In type II deficiency of the ulna associated with dis-



Figure 1. Shortening and ulnar deviation of the right forearm.

location of the radial head and limited forearm rotation, the main surgical goals include excision of the dislocated radial head, excision of the fibrous anlage extending between the distal ulna and the carpus and creation of a single-bone forearm in neutral rotational position so as to provide an optimal functional outcome. Precise measurement of the present proximal ulnar length in the x-rays is mandatory so as to estimate the length of the proximal radius that needs to be removed.

Surgical procedure

Patient lies supine with the shoulder abducted at 90 degrees and the elbow and forearm on a radiolucent table. An anterolateral approach to the elbow extended distally along the forearm is used in order to provide adequate exposure to the proximal third of the radius as well as the hypoplastic ulna. The fibrous anlage, arising from the tip of the deficient ulna, is identified and excised (**fig.4**) Proximal radial osteotomy is then carried out by means of an



Figure 2. Limited flexion and extension of the right forearm.



oscillating saw according to the preoperative length measurements (**fig.5**). Following removal of the proximal third, the distal part of the radius is mobilized, rotated into a neutral position in terms of pronation-supination and fused with the proximal ulna using multiple k-wires. Radiographic examination is carried out (**fig.6**) and a long-arm splint is applied before the patient leaves the operation room.

Results

Wound healing proceeded without complications. The splint was removed at 6 weeks postoperatively following radiographic confirmation of bone healing at the site of the fusion and the patient was encouraged to undertake range of motion exercises. At 8 weeks postoperatively flexion and extension of the elbow were comparable to the contralateral side providing a satisfactory functional outcome (**fig.7**). K-wires were removed 6 months postoperatively. Follow-up clinical and radiographic examinations were carried out at 12 and 24 months; no complications were noted.



Figure 3. Lateral x-ray of the affected forearm manifesting distal ulnar absence, severe bowing of the radius in association with radial head dislocation.

Discussion

Ulnar longitudinal deficiency represents a rare clinical entity affecting 1 sibling in 100.000 live births⁽²⁾. More than 70% of the cases are unilateral with a boy-to-girl ratio of 3:2⁽³⁾. Anomalies of the capitate, lunate, hamate, triquetrum, pisiform as well as absence or deformity of the 4th and 5th metacarpals and digits usually accompany the deficiency⁽⁴⁾. Most cases are regarded as sporadic in occurrence.



Figure 4. Excision of the fibrous anlage extending between the proximal ulna and the carpus.



Figure 5. Osteotomy of the proximal radius.



Figure 6. Creation of a single-bone forearm; radioulnar fusion with multiple k-wires.

However, a series of hereditary conditions and syndromes have been associated with ulnar longitudinal anomalies including Klippel-Feil syndrome⁽⁵⁾ and Goltz-Gorlin syndrome⁽⁶⁾. Ulnar longitudinal deficiency has been classified by Baine in four distinct types⁽¹⁾, reflecting the severity of the deformity. Type I is characterized by distal shortening of

the ulna with both epiphyses present and, occasionally, slight bowing of the radius. In type II ulnar clubhand, there is distal ulnar aplasia along with the presence of a fibrocartilaginous anlage attaching the ulnar stump to the carpus. The latter acts as a tether, progressively bowing the radius. Total ulnar aplasia represents the cardinal manifestation




Figure 7. Functional outcome at 8 weeks postoperatively. Flexion – extension of the forearm is comparable to the contralateral side.

of type III ulnar clubhand, accompanied by elbow instability. Lastly, type IV is characterized by radiohumeral synostosis along with complete absence of the ulna.

Numerous treatment options have been proposed over the last decades; accurate identification of both the anatomical and functional deficits is of paramount importance in determining the most suitable treatment for each case. Hence, we believe that preservation and/or restoration of elbow stability, correction of the bowing deformity of the radius and prevention of its recurrence represent the primary endpoints of surgical intervention. Excision of the fibrocartilaginous anlage along with the creation of a single bone forearm as initially described by Straub⁽⁷⁾ and modified by Lloyd-Roberts⁽⁸⁾ accomplishes all of the aforementioned goals. Even though rotation of the forearm is sacrificed, function is astonishingly improved partly due to the stabilization of the elbow as well as the rotatory compensation provided by the shoulder joint.

Other treatment options include excision of the

anlage along with radial osteotomies in order to correct the bowing deformity and application of distraction osteogenesis of the ulna by use of an external fixator. The former, though well documented in literature^(9,10), would not suffice in our case since elbow stability would not be restored. The latter technique, published recently by Chen⁽¹¹⁾, represents a viable alternative that could be used in order to restore appropriate ulnar length and at the same time reduce and stabilize the elbow joint. However, it requires a much longer treatment period, as compared to the time needed for the radio-ulnar fusion to heal and is associated with high complication rates.

Overall, creation of a single bone forearm represents an acceptable option for patients with type II ulnar longitudinal deficiency, preventing further deformation of the forearm and at the same time improving function. 

Conflict of interest statement

The authors declare that there is no conflict of interest regarding the publication of this article.

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