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Ulnar hemimelia in deformed left forearm treated with ilizarov fixator

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Abstract

Introduction: Ulnar hemimelia is a congenital ulnar deficiency of the forearm characterized by complete or partial absence of the ulna bone.

History and Examination: 18 years female presented to us with deformed left upper limb. Cosmetically, the forearm looked curved with concavity towards the ulnar side.

Materials and Method: The correction was carried out in 2 stages. In first stage, the wedge shaped bone was osteotomised and correction was then fixed with dynamic compression plate. The ulna was fixed with ilizarov frame. In second stage, the corticotomy of proximal ulna was done and rush nail was passed intra medullary before distraction to prevent translation. The ulna was lengthened until it reached the distal radio ulnar joint.

Result and Conclusion: In our case report, the girl with ulnar hemimelia was not able to do routine activities of daily living before the operation. Hence treatment was necessitated to improve cosmetic and functional outcomes. Management of such cases is highly individualised and mainly involves improvement of function.

Keywords: hemimelia, osteotomy, ilizarov, congenital

Introduction

Ulnar hemimelia is a congenital ulnar deficiency of the forearm characterized by complete or partial absence of the ulna bone. When one side of the distal half of limb is absent or underdeveloped, such cases are termed as hemimelia. Incidence is estimated at 1/ 1,00,000-1,50,000 live births, with male to female ratio of 3:2. Ulnar hemimelia is a rare congenital condition which occurs in about 1 in 1.5 million population. Ulnar hemimelia is unilateral in approximately 70% of cases, tends to be right-sided, and is usually incomplete and non-syndromic. Most patients have some shortening of the forearm. The position of the hand tends to drift to the ulnar –side of the wrist. The extension of the elbow is generally limited to about 90 degrees. In severe cases, the elbow is held at about 160 degrees of flexion. Ulnar hemimelia may present with other skeletal anomalies (absence or hyperplasia of the radial digits, thumb duplication, or syndactyly) or syndrome (Poland anomaly, goltz-gorlin syndrome, schinzel syndrome, klippel –feil syndrome or cornelia de lange syndrome). Ulnar deficiency is also one of the manifestations of the femur-fibula-ulna syndrome. In our case, we report a case of farming girl who presented to us with deformed left upper limb with radial head dislocation. Ulnar hemimelia was first reported in 1683 by Goller and hence is probably the first of the paraxial hemimelias to be identified as such, there being some doubt about the true identity of the case of hemimelia described by Pare in 1573. Although chronological tables of all the early cases of radial, tibial, and fibular hemimelia are available in the literature, no such list other than the bibliography provided by Rabaud and Hovelacque seems to have been prepared for ulnar hemimelia.

History and examination: 18 years female presented to us with deformed left upper limb. Cosmetically, the forearm looked curved with concavity towards the ulnar side.

The pronation and supination at the distal and proximal radioulnar joint and elbow movements were restricted. Distal sensations and pulsations were present.

Preoperative radiographs

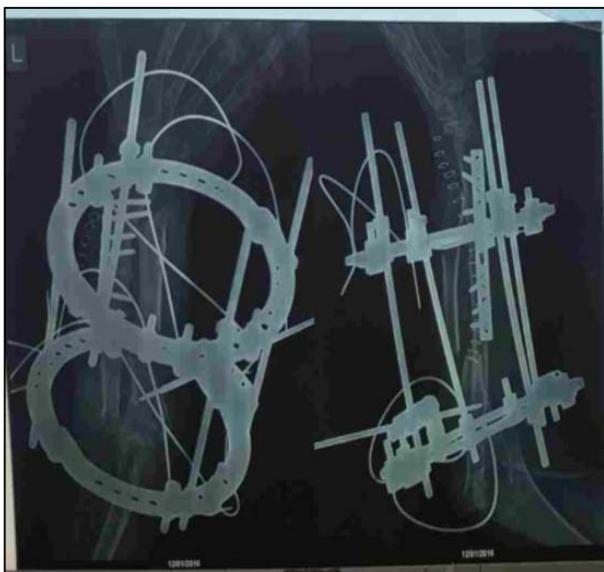


X ray examination of forearm AP and lateral views show radial head dislocation with radial bowing and shortened ulna.

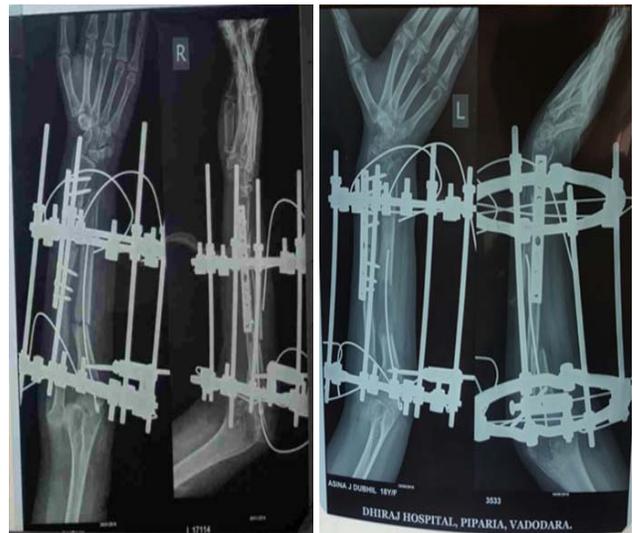
Operative procedure

The correction was carried out in 2 stages. In first stage, the wedge shaped bone was osteotomised and correction was then fixed with dynamic compression plate. The ulna was fixed with ilizarov frame. In second stage, the corticotomy of proximal ulna was done and rush nail was passed intra medullary before distraction to prevent translation. The ulna was lengthened until it reached the distal radio ulnar joint. The radial head automatically relocated with functional improvement in pronosupination and elbow movements.

Post operative photographs



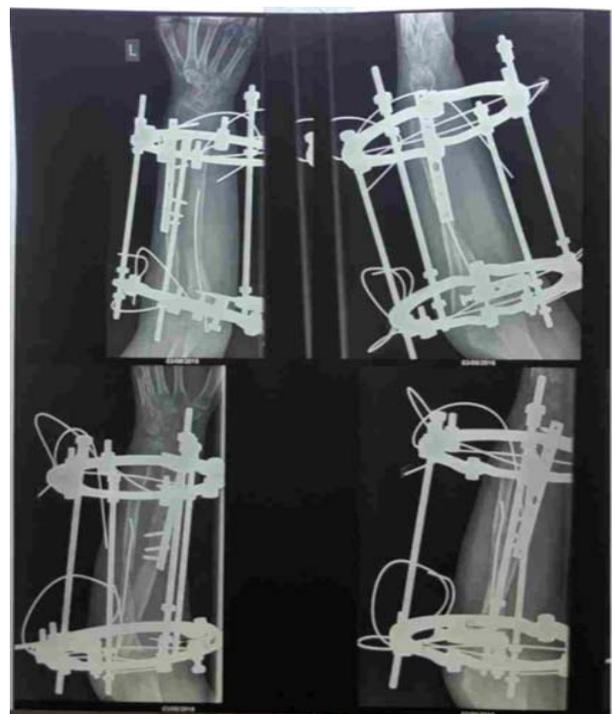
Immediate post operative x rays show AP and lateral views of the forearm with radius deformity correction by diaphyseal osteotomy and plate fixation along with ilizarov fixation for ulna



Post operative x rays after corticotomy of ulna was carried out after insertion of intra medullary rush nail to guide neo-osteogenesis during the process of distraction

Discussion: Ulnar hemimelia is characterised by partial or complete absence of ulna, radial bowing, fixed or mobile elbow with abnormal digits. Most of the cases are males and are unilateral mainly involving the right side. The most critical period for the development of the limb anomalies is from 24-36 days of the embryonic life. Hence early diagnosis in antenatal period reduces the chances of occurrence. Management of such cases is highly individualised and mainly involves improvement of function. The steps involved in our case were as follows:

- STEP I Corrective osteotomy: radius with compression plate
- STEP II Isolated ulnar Ilizarov frame fixation
- STEP III Upper metaphyseal ulnar corticotomy
- STEP IV Ulnar lengthening
- STEP V After enough lengthening, lower end of ulna reaches the wrist
- STEPVI Removal of frame after consolidation of lengthening followed by plaster application and physiotherapy
- STEPVII Rehabilitation





Radiological and clinical photographs showing the lengthening of the affected forearm during the distraction process

Conclusion: In our case report, the girl with ulnar hemimelia was not able to do routine activities of daily living. Hence treatment was necessitated to improve cosmetic and functional outcomes.



X rays showing 6 months follow up with new bone formation after corticotomy and distraction of ulna. Ilizarov frame fixator was removed. The forearm was kept in plaster for 2 months for proper consolidation of the new bone. Thereafter, radial head excision was done to improve pronation and supination at the forearm.



Final clinical photographs of patient after removal of ilizarov frame and plaster after achieving functional length of left forearm.

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