Simultaneous radio-ulnar distraction by a single ulnar corticotomy in a case of Multiple Hereditary Exostosis

**INTRODUCTION:**
Hunter and Boyer first described case of multiple exostosis and hereditary multiple exostosis (HME) respectively[1]. HME is inherited as autosomal dominant disorder with exostosin 1 (EXT1) and exostosin 2 (EXT2) genes being involved which results in reduction of heparan sulphate chain elongation[2]. The above mutations are noted in 70-80% cases and rest show spontaneous mutation [3]. Radiologically, they present as bony outgrowths with a cartilage cap which is juxaphyseal in origin with continuation of cortex and medullary canal from normal bone into the exostosis[4]. Forearm deformities are the most common in MHE with cubitus varus, asymmetric growth of radial and ulnar physis resulting in bowing of radius, shortening of ulna, increased ulnar tilt of distal radius physis, ulnar translocation of carpals, carpal instability and possible dislocation which eventually lead to restriction of pronation supination motion[5].

**Case presentation:**
A 17-year-old male presented with right forearm deformity for past nine years. On examination, his radial head seemed dislocated with ulnarly deviated wrist with forearm shortening of two centimetres. A mid-diaphyseal ulnar corticotomy was done and both the radial and ulnar pins were connected to each other through a connecting rod over the wrist joint. Gradual distraction of ulna with secondary radial migration of the radial head. This helps in improvement of the dislocated radial head at the elbow. Uniplanar external fixator has advantage of technical ease with low complication rates.

**Background:** The rationale of ulnar lengthening in Multiple Hereditary Exostosis is that it frees the tethering of distal radial physis along with increased support to the ulnar-sided carpals and also improves the radial head dislocation due to simultaneous radial dislocation.

**Case summary:** A 17-year-old male presented with right forearm deformity for past nine years. On examination, his radial head seemed dislocated with ulnarly deviated wrist with forearm shortening of two centimetres. A mid-diaphyseal ulnar corticotomy was done and both the radial and ulnar pins were connected to each other through a connecting rod over the wrist joint. Gradual distraction of ulna with secondary radial migration was done for three weeks followed by consolidation for eight weeks. Improvement in range of motion at elbow as well as wrist joint along with DASH score was noted.

**Conclusion:** This procedure was based on the premise that lengthening of the ulna which is connected to the radius through the connecting bar over the wrist will also help in pulling the radius distally leading to distal migration of the radial head. This helps in improvement of the dislocated radial head at the elbow. Uniplanar external fixator has advantage of technical ease with low complication rates.

**Table 1**

<table>
<thead>
<tr>
<th>Forearm Pronation</th>
<th>Pre-operative</th>
<th>Post-operative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elbow Flexion</td>
<td>0-70</td>
<td>0-150</td>
</tr>
<tr>
<td>Elbow Extension</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Forearm Supination</td>
<td>0-90</td>
<td>0-80</td>
</tr>
<tr>
<td>Forearm Pronation</td>
<td>0-45</td>
<td>0-15</td>
</tr>
</tbody>
</table>
Wrist flexion 0-10 0-30
Wrist extension - 0-10
Ulnar deviation 25 0
Forearm length difference 2 cm 0 cm
DASH Score 25 55

Table 1. Illustration of change in range of motion of elbow, forearm and wrist along with DASH score pre-operatively and post-operatively.

**Figure 1.** Preoperative x-ray of the patient showing multiple exostosis, with radial head displacement and ulnar shortening.

**Treatment:**
Two pins of 4mm each were applied to proximal and distal ulna. Distal radius was also held by two 4mm pins and both the medial and lateral systems were connected to each other by a connecting rod centred over the wrist joint. A mid-diaphyseal corticotomy of the ulna approached through the subcutaneous approach between extensor carpi ulnaris and flexor carpi ulnaris was done (Figure 3). Both the ulnar and radial pins were connected to two uniplanar external fixators. Elbow was left free to allow for mobilisation. Distraction was started at the ulnar corticotomy site from post-operative day 7. A total distraction of 1mm per day was carried out every day in quarterly intervals. The distraction was continued for 3 weeks. Weekly radiographs were obtained to evaluate the regenerate formation. At 3 weeks, the distraction was stopped and regenerate was allowed to consolidate for a period of 8 weeks. The external fixators were removed after 6 weeks. Radiographs showed distal migration of the radial head with improved ulnar length and also reduction of the ulnar angulation of the carpals. Range of motion exercises of elbow and wrist joint were being done in the time period. Weight bearing was gradually allowed and full weight bearing and resuming of normal activities was done by post-operative week 14. At 24 months follow up, the range of motion has improved and DASH score was calculated to be 25 (Table 1) (Figure 4, 5). The diagrammatic representation of the change in ulnar length is shown (Figure 6).

**Figure 2.** Immediate post-operative x-ray after ulnar osteotomy and pin placement (A) and at 24 months follow up showing distal placement of radial head compared to preoperative imaging along with improved ulnar length and ulnar carpal support (B).

**DISCUSSION:**
MHE usually inherits in autosomal dominant pattern with around 40% of cases being sporadic[6]. Patients typically present with multiple swellings resembling solitary enchondromatosis with deformities of long bone, especially paired bones of forearm and leg, which occur due to defective endochondral ossification of the physis. The forearm deformities occur due to the difference between cross sectional area of both distal physes and the growth associated with it[5]. The distal ulnar physis being smaller in cross section gets affected more severely. Secondly, it contributes 10% more to the longitudinal growth of forearm compared to distal radial physis. Masada proposed the classification of forearm deformities in MHE to simplify the surgical correction procedure[8]. He recommended ulnar acute lengthening of type I deformities and gradual lengthening in type IIb deformities. Radial osteotomy was needed in most of their cases. There are contrasting opinions regarding timing of surgery, with some advocating late intervention after skeletal maturity to prevent recurrence and some advocating early intervention to have more potential in remodelling[7]. Matsubara et al reported that age is not a factor to predict re-surgery[5]. Since growth potential of any tethered physis cannot be predicted, deformities which will recur also cannot be predicted. But the onset of radial head subluxation or its progression warrants early surgery. The decision regarding acute or gradual lengthening of ulna depends on the amount of lengthening. Up to 2.5cms or 25% of ulnar length lengthening can be done acutely[5].

The patient in this study with forearm deformity was mainly concerned with the cosmetic and functional deformity of the wrist joint. He presented well beyond his skeletal maturity. Gradual lengthening was done since a deficit of about 2cms was needed to be corrected. Both wrist and elbow joints were left free to be mobilized during the distraction and consolidation phase. Post-operatively patient had improvement of elbow flexion and supination due to distal migration of the radial head and improvement of wrist flexion due to increased ulnar length and ulnar carpal support leading to increased joint stability. DASH score showed an increase from 25 to 55.

The earliest literature showing surgical outcomes in 18
Surgical excision of exostosis in most series. Most of the external fixators (Table 2). This was accompanied by acute or gradual by either uniplanar or multiplanar in the recent literature has been ulnar lengthening, either showed improvement of function as well as forearm ulnar lengthening or radial hemiepiphyseal stapling but only reduction of pain whereas patients undergoing simple excision didn’t show any functional improvement as the surgical procedures. The patients undergoing lengthening and radial hemiepiphyseal stapling was done for osteochondromas, excision with gradual ulnar Table2
disease control osteotomy.

Table 2

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Number of cases</th>
<th>Intervention</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mandar Agashe[17]</td>
<td>2018</td>
<td>1</td>
<td>Removal of diseased ulna and radius with nailing in the first step fusing left out parts of ulna and radius with revision plating</td>
<td>Better flexion and increased grip strength with better cosmesis</td>
</tr>
<tr>
<td>Amin Abdel[14]</td>
<td>2018</td>
<td>12</td>
<td>Ilizarov lengthening of ulna</td>
<td>Increase in: Flexion- 117 to 145 Supination- 46 to 73 Pronation- 38 to 20</td>
</tr>
<tr>
<td>John Ham[19]</td>
<td>2016</td>
<td>14</td>
<td>Radial head resection, hemi-interposition using a local tissue flap, and LUCL ligament graft reconstruction after previous distal forearm procedures including excision of osteochondroma(s), ulnar lengthening and/or radial correction osteotomy</td>
<td>Satisfactory results in the postoperative pain score, range of motion, elbow stability, patient satisfaction, and quality of life.</td>
</tr>
<tr>
<td>Kousoukelba[24]</td>
<td>2017</td>
<td>3</td>
<td>Excision of the osteochondroma of the distal ulna and gradual lengthening of the ulna up to 5mm plus variance using an external fixator</td>
<td>With respect to forearm pronation, supination, and ROM, the postoperative values were improved in comparison with the preoperative values. However, forearm pronation, elbow flexion, and extension were not significantly altered from the preoperative value</td>
</tr>
<tr>
<td>Stephen Refsland[5]</td>
<td>2016</td>
<td>17</td>
<td>Gradual mid diaphyseal ulnar lengthening with uniplanar external fixator (all 17 cases) With osteochondroma excision (14 cases) With radial osteotomy (5 cases)</td>
<td>Radial articular angle, carpal slip, radius of curvature, ulnar variance, angle of the radial and ulnar physis, elbow carrying angle, amount of radial head coverage. Non-significant improvement in all the parameters with no pain postoperatively in all cases</td>
</tr>
<tr>
<td>Riccardo D’Ambrosi[14]</td>
<td>2015</td>
<td>15</td>
<td>Excision of ulnar osteochondromas with radial head dislocation and ulnar lengthening by Ilizarov external fixator</td>
<td>MAYO Elbow score improvement- 34 to 93 VAS score- 8.2 to 2.3 Pronation- 35 to 70 Supination- 51 to 80 Functional assessment criteria preop; 1.6 (0–2) Postop; 4.4 (3–5)</td>
</tr>
<tr>
<td>Marco Massobrio[17]</td>
<td>2015</td>
<td>1</td>
<td>Asymmetric lengthening of both the radius and the ulna with two different and separated monoaxial external fixators</td>
<td>Limitation in elbow extension (-15°), and wrist ulnarization (28°), radialization (10°), and flexion-extension (50°–130°). Post op 12 months, elbow extension limitation was -5° and wrist range of motion was completely restored.</td>
</tr>
<tr>
<td>Jason P Kelly[18]</td>
<td>2015</td>
<td>18</td>
<td>Hemiepiphyseal stapling of the radial side of the distal radius</td>
<td>The radial articular angle, carpal slip, ulnar tilt, lunate subsidence, and metaphyseal epiphysial angle were measured on preoperative and final postoperative radiographs and compared. Improvement in 4 out of 6 radiological measurements (lunate subsidence did not show much improvement)</td>
</tr>
<tr>
<td>Yong Jin Cho[19]</td>
<td>2014</td>
<td>4</td>
<td>Excision of ulnar osteochondromas with ulnar lengthening by Ilizarov external fixator</td>
<td>Improvement of radial length, radial bowing angle, and radial articular angle</td>
</tr>
<tr>
<td>B G Beutel[55]</td>
<td>2014</td>
<td>1</td>
<td>Gradual ulnar lengthening with multiplanar external fixator</td>
<td>Complete restoration of elbow ROM and resolution of pain. Forearm was unaffected preoperatively</td>
</tr>
<tr>
<td>Yun-fa Yang[25]</td>
<td>2013</td>
<td>2</td>
<td>Distal ulnar osteochondroma resection and reconstruction with vascularized proximal fibular graft</td>
<td>Improvement in the ROM, pain and function of the wrist</td>
</tr>
</tbody>
</table>


11. Agahaei M, Shah A, Parikh SN. A Rare Presentation and Management of Forearm Deformity in a Patient with Hereditary Multiple Exostoses. JBJS Case Connector. 2018; 8: e63 [PMID: 24405189 DOI: 10.1099/bij.0.002151]


23. Song SH, Lee H, Yoonsal H, Oh SM, Park JH, Song HR. Modified Ilizarov