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

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ABSTRACT

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 distraction.

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 translation 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.

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 juxtaphyseal in origin with continuation of cortex and medullary canal from normal bone into the exostosis [2]. 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[4]. Ulna shortening is more pronounced because distal ulnar physis contributes more to total ulnar length compared to what distal radial physis contributes to total radius length and also owing to less cross-sectional area of ulna leading to its more severe involvement[1]. Masada et al. classified the deformities of forearm[5]. Treatment proponents have suggested operative intervention at one spectrum for functional impairment to others who suggest that these operative interventions do not produce any significant results. Surgery in form of excision of exostosis or lengthening of ulna or lengthening of both radius and ulna or stapling of distal radius physis or excision of radial head or open reduction of radial head or reconstruction of distal ulnar physis by proximal fibular physis or Suave Kapandji procedure may be contemplated[5]. The rationale of ulnar lengthening 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[5].

We present a case report of a 17-year old male of MHE presenting forearm deformities which was managed by ulnar osteotomy and distraction by limb reconstruction system (LRS).

CASE PRESENTATION:

A 17-year old male presented with history of nine years of right forearm deformity with occasional pain in the elbow joint on routine daily activities for the past two years. Trauma and infection as the etiological factors were ruled out. A family history of multiple bony prominences was noted in the mother of the patient but which didn't create any functional deficit in the person. Multiple bony prominences were present in the patient in proximal humerus and proximal tibia bilaterally. The right elbow was internally rotated attitude compared to the opposite side with an abnormal prominence over the lateral aspect of the joint. The abnormal prominence seemed to arise from the proximal radius which moved along with movements of the radius. The olecranon and distal humerus were normal on examination. At the wrist joint, the hand was ulnar deviated. The ulnar styloid along with distal ulna were palpable around 1.5 cms proximal to the radial styloid. Bony prominences were also arising from the distal radius. The measurements of elbow and wrist motion are tabulated (Table 1). A true shortening of two centimetres of the right forearm was noted. Radiographs showed radial head dislocation with bowing of both radius and ulna along with shortening of ulna with carpal displacement ulnarly (Figure 2). Multiple exostoses were also present in the distal forearm (Masada type IIb). DASH score was calculated to be 55.

Table1

	Pre-operative	Post-operative
Elbow Flexion	40-120	12-135
Elbow Extension	-	-
Forearm Supination	0-70	0-90
Forearm Pronation	0-45	0-15

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).

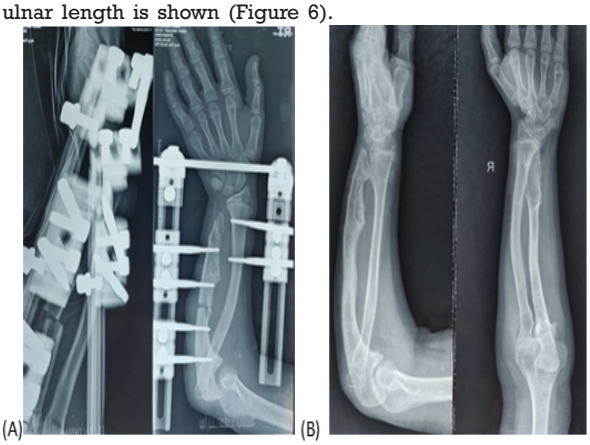

Figure 2. Immediate post-operative x-ray after ulnar osteotomy and pin placement (A) and at 24 months follow up showing distal placement of radial head compared to preoperative imaging along with improved ulnar length and ulnar carpal support (B).

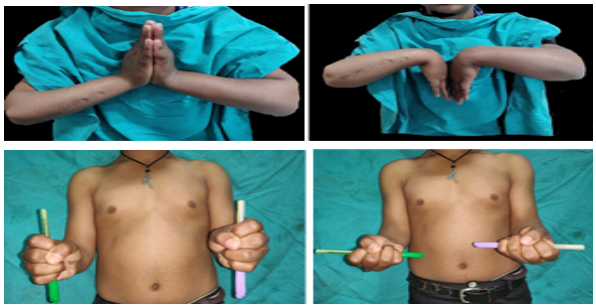

Figure 3. Clinical and functional outcome of the patient at 24 months follow up.

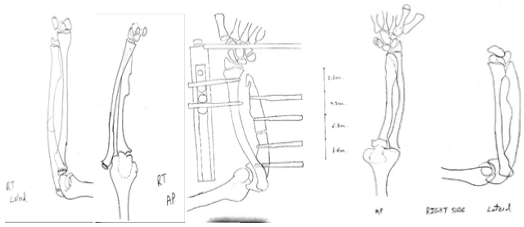

Figure 4. Line diagram showing change in the forearm relationship of radius and ulna over the follow-up period.

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 in 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[5]. 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[8]. 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[9].

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

forearms was reported in 1984[10]. Simple excision of osteochondromas, excision with gradual ulnar lengthening and radial hemiepiphyseal stapling was done as the surgical procedures. The patients undergoing simple excision didn't show any functional improvement but only reduction of pain whereas patients undergoing ulnar lengthening or radial hemiepiphyseal stapling showed improvement of function as well as forearm rotation. Gradually, the most common procedure reported in the recent literature has been ulnar lengthening, either acute or gradual by either uniplanar or multiplanar external fixators (Table 2). This was accompanied by surgical excision of exostosis in most series. Most of the series reported improvement in radiographic measures whereas patient reported outcomes weren't either validated or lacked in depth. Evidence suggests that ulnar lengthening alone or in conjunction with other procedure improves radiological parameters with maintenance of radiocapitellar congruency with reduced pain. Elbow carrying angle as well as forearm rotation improvement has been noted. Wrist symptoms didn't improve significantly in comparison to elbow symptoms in the literatures reported, probably because of the fact that mid-diaphyseal osteotomy of ulna treated radius and ulna as one unit due to strong interosseous membrane and also probably because there is distal physeal tethering of ulna which doesn't improve significantly on lengthening.

Table2

Author	year	Number of cases	Intervention	outcome
Mandar Agashe[11]	2018	1	Removal of diseased ulna and radius with nailing in the first step fusing left out parts of ulna and radius with revision plating	Better flexion and increased grip strength with better cosmesis
Amin Abdel[12]	2018	12	Illizarov lengthening of ulna	Increase in: Flexion- 117 to 145 Supination- 46 to 73 Pronation- 38 to 70
John Ham[13]	2016	14	Radial head resection, hemi-interposition using a local tissue flap, and LUCL ligament graft reconstruction after previous distal forearms procedures including excision of osteochondroma(s), ulnar lengthening and/or radial correction osteotomy	Satisfactory results in the postoperative pain score, range of motion, elbow stability, patient satisfaction, and quality of life.
KousoukeIba[14]	2017	3	Excision of the osteochondroma of the distal ulna and gradual lengthening of the ulna up to 5mm plus variance using an external fixator	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
Stephen Refsland[15]	2016	17	Gradual mid diaphyseal ulnar lengthening with uniplanar external fixator(all 17 cases) With osteochondroma excision (14 cases) With radial osteotomy (5 cases)	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
Riccardo D'Ambrosi[16]	2015	15	Excision of ulnar osteochondromas with radial head dislocation reduction and ulnar lengthening by Ilizarov external fixator	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)
Marco Massobrio[17]	2015	1	Asymmetric lengthening of both the radius and the ulna with two different and separated monoaxial external fixators	Limitation in elbow extension (-15°), and wrist ulnarization (25°), radialization (10°), and flexion-extension (50°–130°). Post op 12 months, elbow extension limitation was -5° and wrist range of motion was completely restored.
Jason P Kelly[18]	2015	16(18 forearms)	Hemiepiphyseal stapling of the radial side of the distal radius	The radial articular angle, carpal slip, ulnar tilt, lunate subsidence, and metaphyseal epiphyseal angle were measured on preoperative and final postoperative radiographs and compared. Improvement in 4 out of 5 radiological measurements (lunate subsidence did not show much improvement)
Yong Jin Cho[19]	2014	4	Excision of ulnar osteochondromas with ulnar lengthening by Ilizarov external fixator	Improvement of radial length, radial bowing angle, and radial articular angle
B G Beutel[20]	2014	1	Gradual ulnar lengthening with multiplanar external fixator	Complete restoration of elbow ROM and resolution of pain. Forearm was unaffected preoperatively
Yun-fa Yang[21]	2013	2	Distal ulnar osteochondroma resection and reconstruction with vascularized proximal fibular graft	Improvement in the ROM, pain and function of the wrist

S H Song[22]	2012	16 (23 forearms)	(1) corrective osteotomy (monofocal or bifocal) and gradual lengthening of the ulna, (2) corrective osteotomy (monofocal or bifocal) of the radius, and (3) excision of exostoses	Most patients were satis ed with forearm appearance, had no pain on strenuous activities & could manage daily life activities easily.
Zhong-wen Tang[23]	2012	14	Osteochondroma excision (ulna), gradual distal ulnar osteotomy and lengthening uniplanar external xator	Percentage of grip power, ROM, pain and activity of hand. All but 2 cases had no limitation of daily activities,4cases reported mild pain. All ROM parameters in forearm and biplane wrist motion improved except for one case.
Bilal Demir[24]	2011	6	Circular external fixator in 4 Single rail road fixator in 2 Proximal ulnar osteotomy in 3 Mid diaphyseal osteotomy in 2 Distal ulnar osteotomy in 1	Radial articular angle, carpal slip, ulnar variance, radial bowing. MRI, CT scanning and bone scintigraphy were used and patients showed improvement in all indices
Bjoern Vogt[25]	2011	12	Gradual diaphyseal ulnar lengthening, uniplanar ortho fix external fixator (12),+/- osteochondroma excision(5ulnas),+/- proximal radioulnar synostosis resection(1) +/-radial osteotomy(4)	Forearm:41% improved,25% deteriorated,4% unchanged. Elbow & wrist fiexion/extension: 84%unchanged,1case improved and another deteriorated. Wrist adduction/abduction: 59%improved and 41% unchanged.
Hidenori Matsubara[8]	2006	7	Excision of osteochondromas from the distal ulna, correction of the radius, and ulnar lengthening with external fixation up to 5mm plus variance.	ROM & pain were used as tools for assessment and showed significant improvement. Before the operation, the ulna was shortened by an average of 12.3mm resulting in a bowing of the radius by an average of 21.9°.
G R Fogel (9)	1984	18	excision of the osteochondromas (10 patients), ulnar lengthening with excision of the osteochondromas (3 patients), and ulnar lengthening with radial hemiepiphyseal stapling (7 forearms of 5 patients)	Isolated excision: no improvement in neither rotation nor ulnar shortening but significant improvement in pain, no osteochondroma recurrence. Ulnar lengthening & excision – improved function and rotation

Table 2. Review of literature of the last 25 years showing various surgical procedures performed and its outcome.

Gradual ulnar lengthening either by uniplanar or multiplanar external fixator, results in improved functional forearm rotation with improvement of elbow carrying angle and wrist motion. LRS has advantage of technical ease with low complication rates as compared to multiplanar or circular external fixator. This procedure should be considered as treatment option in patients of MHE with impending radial head dislocation or complete radial head dislocation.

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