

Case Report

Ilizarov reconstruction on forearm deformity due to multiple hereditary exostosesFaisal Miraj,¹Asdi ARB²¹ Department of Orthopaedic & Traumatology, Fatmawati Hospital, Universitas Indonesia²Department of Orthopaedic & Traumatology, Faculty of Medicine, Universitas Indonesia, Cipto Mangunkusumo Hospital, Jakarta, Indonesia**ABSTRACT****ABSTRAK**

Introduction: Multiple hereditary exostoses (MHE) is a rare medical condition that usually located at the most rapidly growing ends of long bones and manifested by multiple lesions and frequently associated with characteristic skeletal deformities. Management of this disease has many problems. Therefore, it is a challenge for orthopaedic surgeons for such cases are rarely found and the complexity they may have. We did an Ilizarov reconstruction for deformity correction while maintaining the length of the forearm with good outcome and no complications.

Methods: We present a series of 3 patients with forearm deformity due to MHE. They were between 5-17 years old. All of them showed a marked ulnar shortening, radial bowing, and radial head dislocation or subluxation. We performed corrections using Ilizarov frame to gradually lengthen the ulna until the most distal part achieved a range of -2 mm from radial styloid for normal ulnar variance. Subsequently, the distal radius ulna was temporary transfixed with K wire and the ulnar lengthening continued along with gradual traction of the radius until the radial head achieved the level of articulation with the humeral capitellum. Open reduction was needed, followed by temporary pin fixation of the radial head to the humeral capitellum. Osteotomy of the radius was performed in patient 3 (with severe bowing deformity) to straighten the radius.

Results: With Ilizarov method, forearm deformities were corrected, ulnar deviation was improved, prominence of the radial head on the lateral elbow was diminished, and gradual reduction of the radial head on humeral capitellum was safely completed, therefore, this method optimally maintained the length of the forearm. All three patients had no complications, especially no radial nerve injury, and no significant pin track infection. During the rehabilitation program, there were no joint stiffness and the elbow and wrist ROMs were improved.

Conclusion: Ilizarov reconstruction is a good choice to treat forearm deformity due to multiple hereditary exostoses. It has less complications, good outcome, maintain the length of the forearm, good ROM, and good radiographic parameters followed before and after the operation.

Level of Evidence: 3

Pendahuluan: Multiple hereditary exostoses (MHE) adalah kelainan yang jarang yang biasanya terletak pada ujung tulang panjang yang paling cepat pertumbuhannya dan tampak sebagai lesi multipel dengan deformitas yang khas. Penatalaksanaan penyakit ini sering bermasalah. Sehingga keadaan ini merupakan tantangan bagi orthopedic karena merupakan kasus yang jarang dan kompleks. Kami melakukan rekonstruksi dengan ilizarov untuk koreksi deformitas sekaligus mempertahankan panjang lengan bawah dengan hasil yang baik dan tanpa komplikasi.

Metode: Kami membawakan serial kasus dari 3 pasien dengan deformitas lengan bawah yang disebabkan oleh MHE. Mereka berumur antara 5-17 tahun. Pada semuanya tampak jelas pemendekan ulna, radius yang melengkung, dan dislokasi atau subluksasi radial head. Kami melakukan koreksi menggunakan ilizarov untuk memanjangkan ulna secara bertahap sampai jarak - 2 mm dari radial styloid untuk mencapai normal ulnar variance. Setelah itu distal radius ulna difiksasi sementara dengan K-wire kemudian pemanjangan ulna dilanjutkan bersamaan dengan tertariknya radial head sampai pada posisi sendi humeral capitellum. Open reduction diperlukan dan diikuti dengan fixasi sementara antara radial head dengan sendi humeral capitellum. Osteotomy dilakukan pada pasien ke-3 dengan deformitas yang berat untuk meluruskan radius.

Hasil: Dengan metode ilizarov, deformitas lengan bawah dapat terkoreksi dengan perbaikan pada ulnar deviasi, hilangnya penonjolan radial head pada sisi lateral siku akibat reduksi bertahap dan aman pada sendi humeral capitellum. Panjang lengan bawah dapat dipertahankan. Seluruh pasien tidak mengalami komplikasi khususnya cedera radial nerve dan infeksi. Sejalan dengan itu, program rehabilitasi dilakukan sehingga tidak ada pasien yang mengalami kekakuan sendi dan perbaikan ROM siku dan pergelangan tangan.

Kesimpulan: Rekonstruksi dengan Ilizarov merupakan pilihan yang baik untuk penatalaksanaan deformitas lengan bawah yang disebabkan oleh MHE. Dengan metode ini tidak didapatkan komplikasi dan hasil yang baik, dapat mempertahankan panjang lengan bawah, fungsi ROM yang baik dan tercapainya parameter radiologi yang baik.

Keywords: Ilizarov reconstruction, forearm deformity, multiple hereditary exostoses, outcome**Corresponding author:** Faisal Miraj, MD. icalorto@yahoo.com

INTRODUCTION

Multiple hereditary exostoses (MHE) is a disorder of enchondral bone growth that usually located at the most rapidly growing ends of long bones and manifested by multiple lesions and frequently associated with the characteristics of skeletal deformities. MHE is a relatively rare autosomal dominant disorder, mainly caused by loss of function as a result of mutations in two genes: exostosin-1 (EXT1) and exostosin-2 (EXT2). [1] Although, MHE is histologically benign lesions, they can cause a variety of clinical problems, including skeletal pain, cosmetic concerns, and limitation to the adjacent joint.[2] In MHE, osteochondromas are almost always located in the proximal humerus, the distal femur, and the proximal tibia. In addition, the incidence is 80% in the distal ulna and lower in the proximal forearm (38% in radius, 37% in ulna). Forearm deformities can be found in 30% to 60% of MHE patients.[2-4] The deformities are a combination of relative shortening of the ulna, bowing of either or both forearm bones, increased ulnar tilt of the distal radial epiphysis, ulnar deviation of the hand, progressive ulnar translocation of the carpus, and dislocation of the head of the radius. Radiologically corresponds to Masada classification. ^{3,5} Sometimes, exostoses can interfere with normal development of the growth plate, giving rise to limb deformities, low stature and scoliosis. Other many neurovascular and associated disorders can lead to surgery. The therapeutic approach to MHE is substantially surgical, whereas the medical one is still at an experimental level.²⁻⁶

The treatment of MHE is a difficult problem because little has been documented. Surgical intervention is indicated for pain or functional deficit due to osteochondroma. Sometimes, surgery is performed to improve the cosmetic appearance.^{3,4} There are several treatment modalities for the forearm deformities. One of them is Ilizarov reconstruction. Based on our literature review, there are only a few case reports on the use of Ilizarov reconstruction in forearm deformity due to multiple hereditary exostoses. The aim of this study is to understand the clinical outcome, including functional status, range of motion of the wrist, forearm, and elbow, and radiological results confirmed to the parameters. (Figure 1)

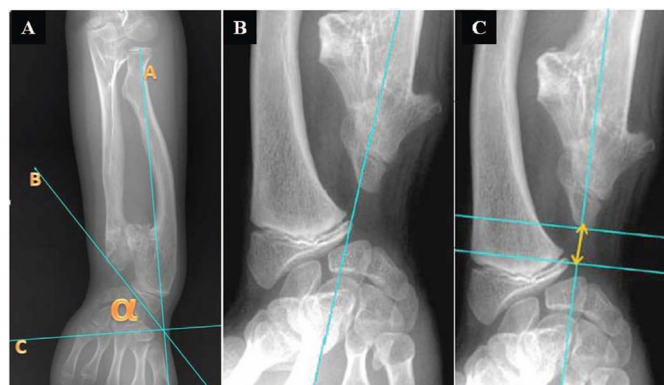


Figure 1. Radiograph parameters as described by Fogel *et al.* (1984) and Burgess and Cates (1993) show (A) RAA (α) was defined as the angle between a line running along the articular surface of the radius (b) and another line (c) that was perpendicular to a line (a) joining the centre of the radial head to the radial border of the distal radial epiphysis (or the radial styloid in skeletally mature patients). (B) CS was measured by determining the percentage of the lunate that is in contact with the radius. A line was drawn from the centre of the olecranon through the ulnar border of the radial epiphysis (or radial articular surface in mature patients). This line normally bisects the lunate. (C) Arrow represents the ulnar variance. We draw a line from the distal ulnar tip to the ulnar border of the distal radial epiphysis. ⁵

CASE ILLUSTRATION

We present a girl, 9 years old, with right elbow deformity, radial head dislocation and short ulna with concurrent bowing of both bones. Her parents also noted reduced elbow and forearm mobility, but she was pain-free with no problem with activities of daily living. On physical examination, both elbows appeared straight with reversal of the normal carrying angle. Bony prominence over the lateral aspect of the right elbow gave an impression of cubitus varus deformity. Flexion and extension of the right elbow were 10-110° while the left side were 0-135°. The right elbow had 50° pronation and 90° supination, while the left elbow had 75° pronation and 90° supination. Bony swelling was palpable over the wrist, ulnar deviation of wrist, no localized tenderness and no significant limitation of wrist motion. Plain radiographs showed right distal ulnar exostoses, ulnar shortening, with anterior-lateral dislocation of radial head corresponding to Masada type IIb. (Figure 2)

Radiographic measurement was conducted. The parameters of the forearm, such as radial articular angle (RAA), carpal slip (CS), and ulnar variances, were measured before the operation (Figure 2). The result for

this patient were 42° for RAA, Carpal slip was abnormal with ulnar displacement of the lunate more than 50% and -35 mm for ulnar variance.

The list of preoperative problems for this first patient consisted of distal ulnar exostoses with shortening of the ulnar bone, radiocapitellar joint dislocation with proximal migration of the radial head, ulnar minus right wrist, ulnar deviation of the wrist joint, slight bowing of both the radius and ulnar bones, limited flexion and pronation of the elbow.

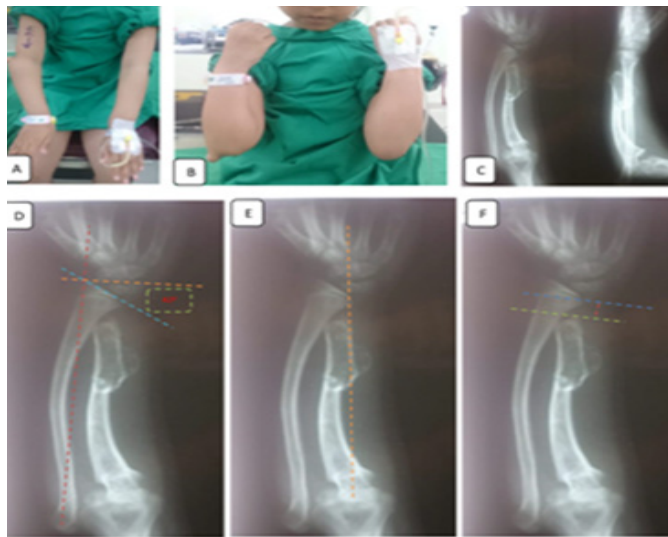


Figure 2. (a) limitation in the pronation of the right elbow (range of motion $0-50^\circ$) with prominence over the lateral aspect of the left elbow and ulnar deviation of the wrist (b) Pre-operative picture showing limited flexion of the right elbow (range of motion $10-110^\circ$) (c) Plain radiograph of radius and ulnar with ulnar minus deformity of the wrist, distal ulnar exostoses and anterolateral radio-capitellar dislocation (Masada type IIB deformity). These radiographs showed the radiographic measurements: (d) The radial articular angle (RAA) was 42° which defined as the angle between a line running along the articular surface of the radius (blue line) and another line (orange line) that was perpendicular to a line (red line) joining the centre of the radial head to the radial border of the distal radial epiphysis (the radial styloid in skeletally mature patients). (e) Carpal slip was abnormal with ulnar displacement of the lunate more than 50%. It was measured by determining the percentage of the lunate that was in contact with the radius. First, a line was drawn from the centre of the olecranon through the ulnar border of the radial epiphysis (radial articular surface in mature patients). This line normally bisects the lunate. (f) Red line represented the ulnar variance. It was measured from the distal ulnar tip to the ulnar border of the distal radial epiphysis. This patient had -35 mm

Second case, a 5 year old boy was referred to our clinic with forearm deformity and dislocation of radial head. He was previously diagnosed with hereditary multiple exostoses. The patient had marked shortening with forearm rotation limitation in pronation and elbow flexion. Clinical pictures showed a bump on her right elbow showing the dislocated head of radius and varus angulation of the elbow. Flexion and extension of the right elbow were $20-100^\circ$ while on the left side were $0-130^\circ$. The right elbow had 40° pronation and 90° supination, while the left elbow had 75° pronation and 90° supination. Bony prominence was on distal ulna, ulnar deviation of wrist and no limitation of wrist motion. Plain radiographs showed distal ulnar exostoses, ulnar shortening and anterior-lateral dislocation of radial head corresponding to Masada Type IIB. (Figure 3)

Radiographic measurement of the second patient was 46° for RAA, abnormal for CS and -20 mm for ulnar variance (Figure 3). The list of pre-operative problems for this patient was similar with the first patient, which was distal ulnar exostoses, shortened ulnar, anterolateral radiocapitellar joint dislocation, ulnar deviation of the wrist joint, slight bowing of both radius and ulnar bones, limited flexion and pronation of the elbow.



Figure 3. (A) Pre-operative picture showing a prominence on the right elbow which is a dislocation of radial head with limitation on pronation (range of motion $0-40^\circ$) (B) Plain radiograph of distal ulnar exostoses, ulnar shortening and anterolateral radio-capitellar dislocation (Masada type IIB deformity) (C). Radiographic measurement of the second patient was 46° for RAA, abnormal CS and -20mm ulnar variance

The third patient was a girl aged 17 years old with a severe radial bowing, varus angulation of elbow, non-dislocated of radial head, short ulna, ulnar deviation of wrist. The flexion-extension of elbow was normal, no pain, no limitation of activities but she had difficulty

to pronation. Plain radiographs showed distal ulnar exostoses, ulnar shortening and no dislocation of radial head corresponding to Masada type I. (Figure 4) Radiographic measurement of the third patient was 50° for RAA, abnormal for CS and -30 mm for ulnar variance. (Figure 4) The list of pre-operative problems for this patient consisted of distal ulnar exostoses, shortening of the ulnar bone, ulnar deviation of the wrist joint, bowing of both the radius and ulnar bones, limited pronation of the elbow.

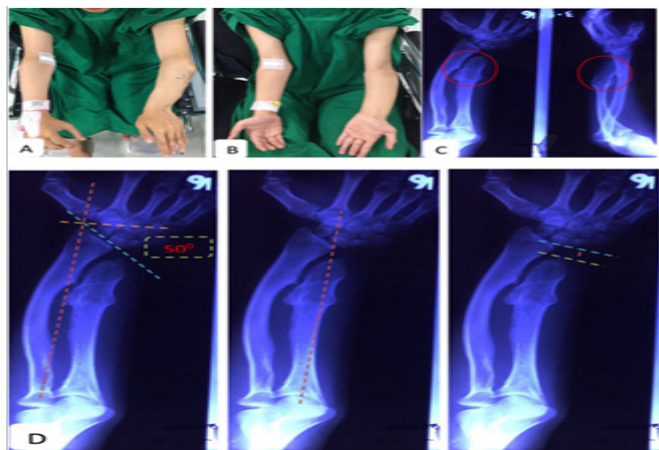


Figure 4. (A and B) Clinical picture showing limitation of pronation (range of motion 0-50°) and normal limit for supination (C) Plain radiograph of distal ulnar exostoses, ulnar shortening and without dislocation of left radial head (Masada type I deformity) (D) Radiographic measurement showing 50° for RAA, abnormal CS and -30 mm ulnar variance

METHODS

We performed the operation into two methods, based on the type of the disease (Masada Type 2B and 1). For Masada Type 2B (patient 1 and 2), we excised the ulna exostoses that just prominence to the skin and followed by osteotomy on metadiaphysis of the ulna for gradual lengthening using ilizarov frame. The frame consisted of two full rings with 3 rods, each on proximal and distal side. One or two 4.5 mm diameter half pins and one tensioned wire 1.6 mm put on proximal side of the ulna and two or three 4.5 mm half pin on distal. (Figure 5)

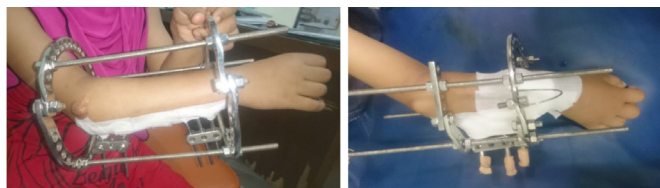


Figure 5. Clinical photo of the right forearm in patient 1 and 2 with ilizarov frame

1 mm per day gradual distraction started 7 days after operation until the tip of distal ulna achieved range -2 mm from radial styloid (normal value of ulnar variance). Temporary transfixing K-wire 2.0 on distal ulna and radius was put subsequently and ulnar lengthening continued followed by gradual traction of the radial head spontaneously to the level of articulation with humeral capitellum. Open reduction of radial head with temporary pin fixation needed in order to facilitate good reduction and soft tissue reconstruction. In patient 1, Ilizarov frame was kept until the distraction callus consolidated in 3 months. (Figure 6A) In patient 2, the frame was converted into internal fixation as soon as ulnar lengthening completed along the reduction of radial head was achieved. Internal fixation shortened the ilizarov frame period and could minimize ulnar distraction callus angulation that occurred in patient 1. (Figure 6B)

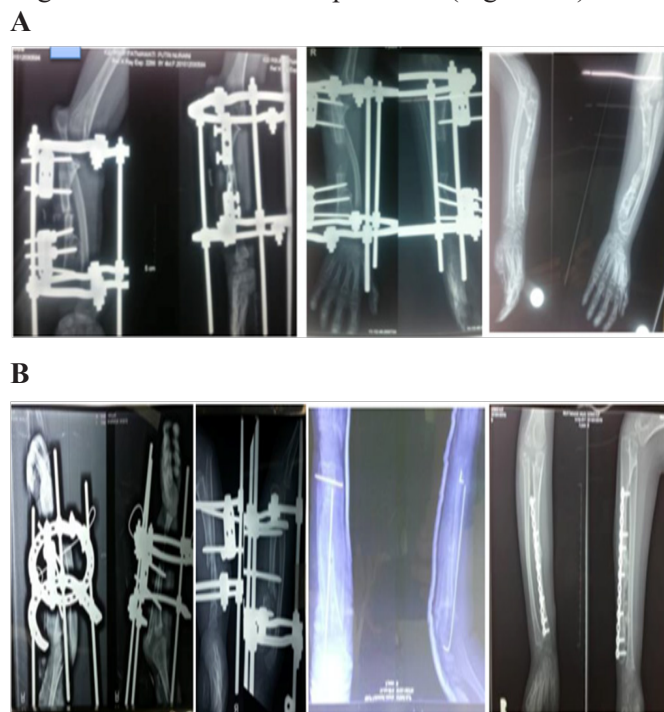


Figure 6. A Right forearm radiograph of patient 1 shows the sequences of correction until complete healing of the osteotomy sites with reduced radial head B. Radiographs of patient 2 shows the sequences of correction, ulnar lengthening with reduced radial head facilitated by transfixing wire on distal radius ulna, followed by conversion of ilizarov frame into internal fixation.

In Masada type 1 (patient 1), there were severe bowing on radius with subluxation of radial head and short ulna. Open reduction of radial head performed with subsequent correction of radial bowing through osteotomy on the

apex, followed by intramedullary fixation straightened the radius without shortened it to preserve the radial length. Osteotomy on metadiaphysis ulna was performed for further lengthening using ilizarov frame. The construct consisted of three 5/8 ring with three rods, each on proximal, middle and distal side, One 4.5 mm diameter half pins and one tensioned wire 1.6 mm put on proximal ulna, two 4.5 mm half pin on distal ulna and one 4.5 mm half pin on distal radius. 1 mm per day gradual ulnar lengthening started 7 days after surgery by pulling downward the middle ring until the tip of distal ulna achieved range of -2 mm from the radial styloid (normal ulnar variance). (Figure 7) Conversion into internal fixation was performed as soon as ulnar lengthening completed along the reduction of radial head was achieved in order to shortened the ilizarov frame period and minimized ulnar distraction callus angulation. (Figure 8,9, 10)

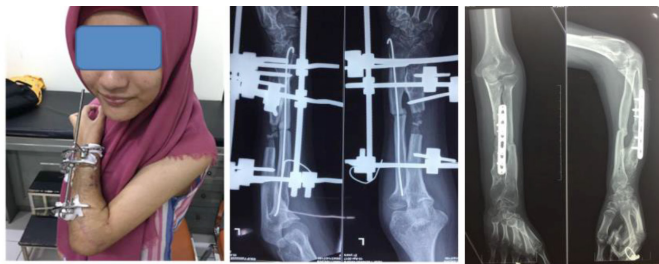


Figure 7. Clinical and radiographs of the left forearm in patient 3, sequences of ulnar lengthening, with osteotomy for correction of radial bowing and conversion of ilizarov frame into internal fixation

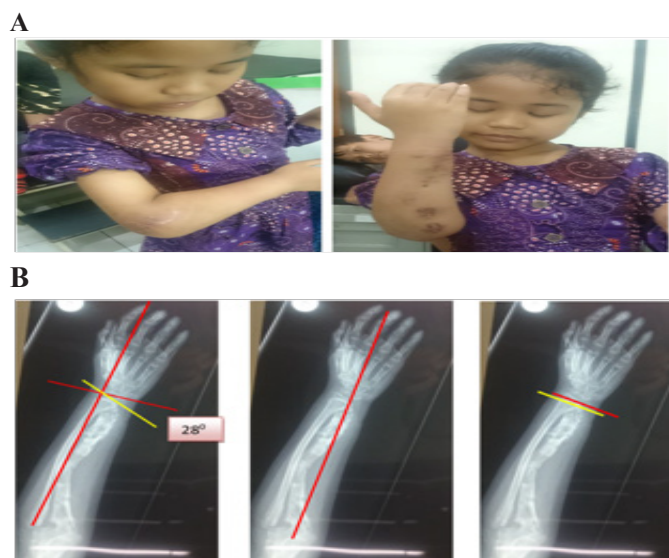


Figure 8. Clinical and radiographic photo of the right forearm in patient 1 A.full range of pronation and flexion elbow at a week after the operation. B. Radiographs after the operation with all of the parameter was normal limit

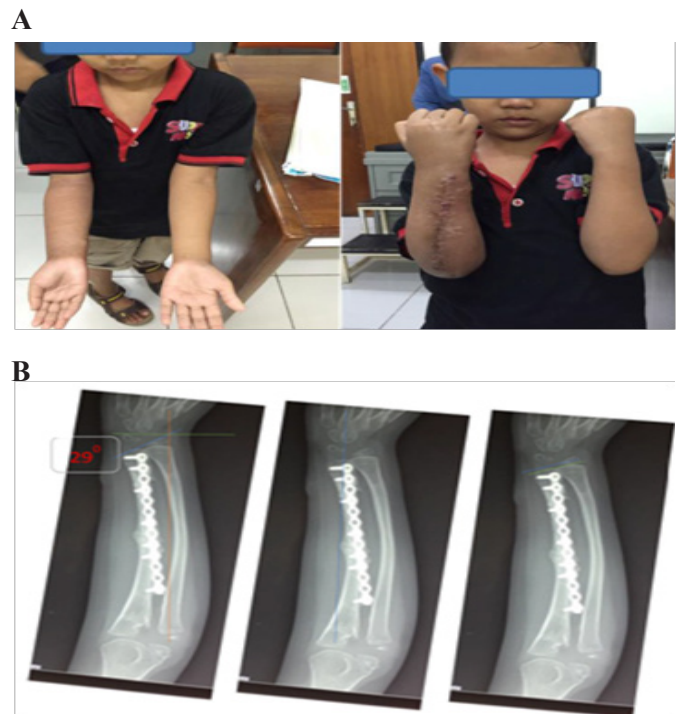


Figure 9. Clinical and radiographic photo of the right forearm in patient 2 A. achieved full range of flexion elbow after the operation. B. Radiographs after the operation with all of parameter was normal limit

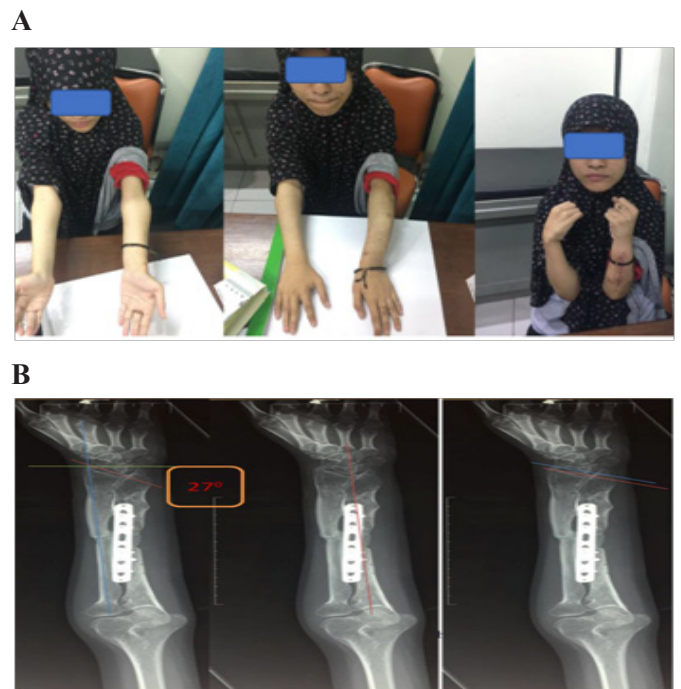


Figure 10. Clinical and radiographic photo of the right forearm in patient 3 A. achieved full range of flexion elbow and pronation after the operation with no complications.. B. Radiographs after the operation with all of the parameter was normal limit

RESULTS

| No. | Parameters | Patient 1 | | Patient 2 | | Patient 3 | |
|-----|--------------------------------|-----------|--------|-----------|--------|-----------|--------|
| | | Pre- | Post- | Pre- | Pro- | Pre- | Post- |
| 1 | RAA | 42° | 28° | 46 | 29° | 50° | 27° |
| 2 | CS | 74% | 26% | 72% | 26% | 71% | 27% |
| 3 | Ulnar variance | -35mm | -2mm | -20 mm | -2 mm | -30 mm | +1 mm |
| 4 | Pronation | 0-50° | 0-75° | 0-40° | 0-75° | 0-50° | 0-75° |
| 5 | Flexion elbow | 10-110° | 0-135° | 20-100° | 0-135° | 0-135° | 0-135° |
| 6 | Radial head dislocation | + | - | + | - | - | - |
| 7 | Radial nerve palsy | - | - | - | - | - | - |
| 8 | Joint stiffness | - | - | - | - | - | - |
| 9 | Bowing of the bone | + | - | + | - | + | - |
| 10 | Ulnar deviation of the wrist | + | - | + | - | + | - |
| 11 | Prominences of the distal ulna | + | - | + | - | + | - |

Table 1. The result of our technique by comparing the pre- and post-operative procedure

Deformity and clinical outcome. All patients showed improvement after the course of treatment. Normal ulnar variance was achieved. Bowing of both bones were satisfactorily corrected while successfully maintained the length since no shortening procedure of the radius was performed. (Table 1).

Range of movement. Post-operative improvements of full range of motion on elbow, forearm and wrist at the time of the final follow-up evaluation were regained. The mean forearm pronation was 75°. The mean elbow flexion was 135° (figure 8,9,10). The mean range of motion was improved compared with the pre-operative range (0-46° on forearm pronation and 15-105° on elbow flexion). Patient 1 and 2 had severe radial head dislocation at the time of surgery and underwent progressive ulnar gradual lengthening in order to distract radial head until it reduced or achieved the level of humeral capitellum, which then followed by open reduction. In evaluation after surgery, no complication of limited range of motion occurred. (Table 1).

Radiological parameters. The average pre-operative radial articular angle, carpal slip and ulnar variance were 46°, 73%, -28 mm. There were improvements at the time of the final evaluation, the measured parameters averaged

28°, 26%, -2mm respectively. (Table 1).

Outcome of Excision of Exostoses. Excision of exostoses on distal part of ulna was performed to all patients. Simple excision of exostoses in the part that prominence to the skin improved the range of pronation with good cosmetic result since there was no more prominence on distal ulna (Table 1).

Complications. There were no complication of radio-ulnar synostosis, radial nerve palsy, joint stiffness and non-union. No pin tract infection occurred, and no breakage of wires or pins. All the resected tumors were pathologically confirmed as exostoses, and none was malignant. No tumor recurrence was found during the follow-up period. (Table 1).

Functional score. The range of pronation/supination and elbow flexion/extension were pre-operatively evaluated. Each patient was rated according to the Functional Assessment Criteria. (Table 2) The presenting symptoms were no pain, loss of range of motion of the forearm and cosmetic problems in all patients (score 4). After surgery, all patients were given score 5, they were satisfied with the result, no limitation in all activities, good cosmetic and no complication.

| Score | Please check the one box that most closely describes the current condition of your hand and wrist |
|-------|--------------------------------------------------------------------------------------------------------------------------------------|
| 5 | I have no limitations of my activities and no pain |
| 4 | I have no pain. I have some limitation of my activities but have not had to change my lift (Sports activities or Job because of it . |
| 3 | I have no pain. I have had to change or limit my job or give up certain sports activities because of the condition of my hand. |
| 2 | I have pain in my hand, wrist, or elbow, but i have no limitations because of it. |
| 1 | I have pain in my hand, wrist, or elbow, which limits my activities. |
| 0 | I have pain for which I take medications. |

Table 2. Functional Assessment Criteria by the Patient

DISCUSSION

Surgical procedure for forearm deformities in MHE have various techniques that have been reported, but the procedure still remains controversial.¹ Masada introduced the classification system for forearm deformities in MHE, he presented a simplification of clinical picture, so that surgeons could aim for acceptable correction.² Akita *et al.* reported that excision of exostoses alone did not correct forearm deformities, but simple excision achieved significant improvement of pronation and might also slow down the progression of deformities.^{5,20} Prichett *et al.* reported that the lengthening of the ulna and correction of the radius, particularly with the use of an external fixator, gave predictable results and was a useful method for treating forearm deformity in MHE. Matsubara *et al.* reported that the results of treatment by excision of exostoses, correction of radius, and gradual lengthening of ulna with external fixators was satisfactory, especially for elbow and wrist functions.⁵ This study reports the results of three operations. Two types of operation procedure performed in patients 1 and 2 with MHE of the forearm corresponding to Masada type IIb and one operation performed in patient 3 corresponding to Masada type I. Open reduction of the head radius performed with subsequent correction of radial bowing with osteotomy on the apex of deformity and intramedullary fixation to straighten the radius without shortening procedure. Gradual lengthening of ulna was performed with Ilizarov frame until normal

ulnar variance was achieved, so that the three procedures preserved the radial length.

The indications of the surgery should be carefully validated since the functional benefits seem to be minimal.¹ Noonan *et al.* reported that seventeen of thirty-nine patients found the arms to be cosmetically unappealing because of shortening, angulation, or bumps. Wood *et al.* reported ten patients who had undergone various operations and concluded that function only showed minimal improvement, the appearance of the limbs was markedly improved.²⁰ In this study, we did surgery to all patients because of limited motion and cosmetic. We achieved significant improvement for pronation and elbow flexion and good cosmetic results with no complication.

Dahl stated that he did not perform direct reduction of a chronically dislocated radial head because stiffness and pain could occur.²⁰ Demir *et al.* reported a successful outcome of the treatment of complete radial head dislocation with gradual ulnar lengthening using an external fixator in Masada type IIa or IIb deformity. They stated that the radial head is gradually transferred to the reduction localization along with ulnar lengthening, and during this procedure stabilization of the head is achieved within a fibrous bed as demonstrated in their study using magnetic resonance imaging. Akita *et al.* reported dislocation of the head of the radius in five extremities/eight forearms. These treatments were not effective in either case. Excision of the dislocated radial head was performed in two patients after their skeletal maturity was reached.² In our study, two patients had a complete radial head dislocation with Masada type IIb deformity. Downward gradual ulnar lengthening was performed with Ilizarov frame until normal ulnar variance were achieved and followed by subsequent transfixing wire to fix the distal radial ulna, so that the ulnar lengthening continued in order to distract the radial head to the level of the capitellum. Subsequently, an easier open reduction performed to facilitate good reduction and soft tissue reconstruction. We believed this procedure could be a good surgical option for treating severe radial dislocation in these cases because it provided safe reduction with minimal risk of nerve injury and preserved the radial length without the need of resection or shortening.

The timing of the surgery is extremely important. Most exostoses become apparent toward the end of the first decade of life or later. Young children show obvious

rapid deforming forearm, therefore, operation should be performed at a younger age. If the child is functionally and cosmetically doing well, the operation may be delayed.^{2-5,19} However, Matsubara *et al.* did not find any correlation between recurrence rate and the patient's age; recurrence rate may depend on the extent of damage to the distal ulnar physis.¹⁶ We conducted this technique to 3 patients with age 9, 5, 17 years old and all results were clinically, functionally, and radiographically good. At the time of our evaluation, there were no recurrence. Limitation in the follow-up period and the number of samples were our weakness to conclude about recurrence.

No complications were observed in this case series. But, other investigators had reported some complications, for example fracture, non-union or delayed bone consolidation, pin track infection and neurovascular damage due to fixator-controlled ulnar lengthening.¹ The reported complication rate for lengthening of the forearm vary widely, from 0% to 100%. Drawbacks of this technique are longer time is needed, the frame is bulky, and it is technically demanding, risk of pin site infection, pin loosening, dermatitis, premature consolidation, and delayed consolidation.²⁰

The technique offers the following advantages: minimal invasive technique with minimal dissection, decreased neurovascular risk, decreased soft-tissue injury, infection or soft-tissue impairment. We summarize our concept of surgical treatment in: Masada type I with excision of exostoses, osteotomy of the radius and intramedullary fixation to straighten the radius without shortening it and immediate ulnar lengthening with Ilizarov frame until normal ulnar variance. Masada type IIb with excision of exostoses, downward gradual ulnar lengthening with Ilizarov frame until normal ulnar variance and followed by subsequent transfixing wire to fix the distal radial ulna, so that the ulnar lengthening continued in order to distract the radial head to the level of the capitellum. Open reduction was needed to facilitate good reduction and soft tissue reconstruction. Conversion into internal fixation was performed as soon as the ulnar lengthening completed and the reduction of radial head was achieved in order to shortened the period of use of Ilizarov frame and minimize the ulnar distraction callus angulation.

CONCLUSION

We present a summary of treatment protocols and the basis for visualizing, planning and treating forearm

deformities in MHE. We describe our current method of surgical correction of Masada type IIb and the levelling procedure in Masada type I MHE forearm deformities. The advantage that we can take of this modified procedure is that all three patients had no complications, especially no radial nerve injury and no significant pin track infection. Along with the rehabilitation program, there were no joint stiffness, with improvements on elbow and wrist ROMs. Although the initial results are encouraging, more researches are needed for prognostic variables that might influence the outcome and patients satisfaction in surgery for forearm abnormalities caused by multiple heredity osteochondromas. We also conclude that Ilizarov reconstruction is a good choice with no complications, provide good outcome, maintain the length of the forearm, good function of ROM and radiographic parameters.

REFERENCES

1. Vogt B, Tretow HL, Daniilidis K, Wacker S, Buller TC, Henrichs M-P, et al. Reconstruction of forearm deformity by distraction osteogenesis in children with relative shortening of the ulna due to multiple cartilaginous exostosis. *J Pediatr Orthop.* 2011;31(4):393–401.
2. Cho YJ, Jung ST. Gradual lengthening of the ulna in patients with multiple hereditary exostoses with a dislocated radial head. *Yonsei Med J.* 2014;55(1):178–84.
3. Masada K, Tsuyuguchi Y, Kawai H, Kawabata H, Noguchi K, Ono K. Operations for forearm deformity caused by multiple osteochondromas. *J Bone Joint Surg Br.* 1989;71:24–9.
4. Shin EK, Jones NF, Lawrence JF. Treatment of multiple hereditary osteochondromas of the forearm in children: a study of surgical procedures. *J Bone Joint Surg Br.* 2006;88(2):255–60.
5. Song SH, Lee H, Youssef H, Oh SM, Park JH, Song HR. Modified Ilizarov technique for the treatment of forearm deformities in multiple cartilaginous exostoses: case series and literature review. *J Hand Surg Eur Vol.* 2013;38(3):288–96.
6. Le Merrer M, et al. A gene for hereditary multiple exostoses maps to chromosome 19p. *Hum Mol Genet.* 1994;3:717–22.
7. Wuyts W, Schmale GA, Chansky HA, et al. Hereditary Multiple Osteochondromas. *GeneReviews.* Seattle (WA): University of Washington, 2000:1993-2017.
8. Legeai-Mallet L, Munnich A, Maroteaux P, Le Merrer M. Incomplete penetrance and expressivity skewing in hereditary multiple exostoses. *Clin Genet.* 1997;52:12–6.

9. Porter DE, Lonie L, Fraser M, Dobson-Stone C, Porter JR, Monaco AP, Simpson AH. Severity of disease and risk of malignant change in hereditary multiple exostoses. A genotype-phenotype study. *J Bone Joint Surg Br.* 2004;86:1041–6.
10. Malagón V. Development of hip dysplasia in hereditary multiple exostosis. *J Pediatr Orthop.* 2001;21:205–11.
11. Viala P, Vanel D, Larbi A, Cyteval C, Laredo JD. Bilateral ischiofemoral impingement in a patient with hereditary multiple exostoses. *Skeletal Radiol.* 2012;41:1637.
12. Hattori H, Asagai Y, Yamamoto K. Sudden onset of saphenous neuropathy associated with hereditary multiple exostoses. *J Orthop Sci.* 2006;11:405–8.
13. Aldea S, Bonneville F, Poirier J, Chiras J, George B, Carpentier A. Acute spinal cord compression in hereditary multiple exostoses. *Acta Neurochir (Wien)* 2006;148:195–8.
14. Abdullah F, Kanard R, Femino D, Ford H, Stein J. Osteochondroma causing diaphragmatic rupture and bowel obstruction in a 14-year-old boy. *Pediatr Surg Int.* 2006;22:401–3.
15. Bowen ME, et al. Loss-of-function mutations in PTPN11 cause metachondromatosis, but not Ollier disease or Maffucci syndrome. *PLoS Genet.* 2011;7:e1002050.
16. Matsubara H, Tsuchiya H, Sakurakichi K, et al. Correction and lengthening for deformities of the forearm in multiple cartilaginous exostoses. *J Orthop Sci.* 2006;11:459–466.
17. Chin KR, Kharrazi FD, Miller BS, Mankin HJ, Gebhardt MC. Osteochondromas of the distal aspect of the tibia or fibula. Natural history and treatment. *J Bone Joint Surg Am.* 2000;82:1269–78.
18. Cho YJ, Jung ST. Gradual lengthening of the ulna in patients with multiple hereditary exostoses with a dislocated radial head. *Yonsei Med J.* 2014;55(1):178–84.
19. El-Gafary K, El-adly W. Forearm lengthening using Ilizarov external fixator. *Eur Orthop Traumatol.* 2013;4(4):217–24.
20. Akita S, Murase T, Yunenobu K, Shimada K, Masada K, Yoshikawa H. Long term result of surgery for forearm deformities in patients with multiple cartilaginous exostoses. *J Bone Joint Surg Am.* 2007;89:1993–9.