



Correction of forearm deformities in children with multiple osteochondroma, by corrective radial osteotomy and ulnar lengthening by distraction osteogenesis

Fikri Erkal BILEN, Levent ERALP, Halil I. BALCI, Mehmet KOCAOGLU, Harzem ÖZGER

From Istanbul University Medical School, Istanbul, Turkey

We retrospectively evaluated the results after ulnar lengthening and radial deformity correction using an external fixator for forearm deformities caused by osteochondromas. Eight forearms were treated surgically in seven patients with multiple hereditary osteochondroma. The mean follow-up time was 40 months (range, 20 to 60 months). The average radial articular angle improved from 43° to 35.5° (range, 28 to 56°) and the carpal slip improved from 69.5% to 55% (range, 40 to 60%) postoperatively. The average shortening of the ulna was reduced from 2.06 cm to 0.44 cm (range 0 to 1cm) after the treatment. There were no serious complications associated with the surgery ; two minor pin track infections were successfully treated by local wound care and antibiotics. Although technically demanding, ulnar osteotomy and gradual lengthening by an external fixator provided promising results in the treatment of forearm deformities in children with multiple osteochondroma.

Keywords : multiple hereditary osteochondroma ; multiple exostosis ; forearm deformity ; surgical correction.

INTRODUCTION

Many patients with multiple hereditary osteochondroma present a variety of forearm deformities which result in cosmetic and functional problems (3). Several surgical treatment options have been reported in the literature, with no consensus about the optimal treatment. Masada *et al* proposed

a classification system for forearm deformities in order to help in planning of the treatment (3). In this retrospective study, we present our results in the treatment of forearm deformities caused by multiple osteochondroma.

PATIENTS AND METHODS

Between 2003 and 2007, eight forearms in seven patients with multiple hereditary osteochondroma were treated surgically in our institution. There were four female and three male patients with an average age of 10 years (range : 5 to 14 years). No patients were lost to follow-up. The left side was involved in five forearms, the right side in three (one patient had bilateral forearm deformities). Prior to reconstruction, anteroposterior and lateral roentgenograms of both forearms were obtained for all patients (fig 1A & 1B). Ulnar shortening, the radial articular angle (RAA) and carpal slip (CS) were

-
- Fikri Erkal Bilen, MD, FEBOT.
Acibadem University Medical Faculty, Department of Orthopaedics and Traumatology, Istanbul, Turkey.
 - Levent Eralp, MD, Associate Professor.
 - Halil I. Balci, MD, Resident.
 - Mehmet Kocaoglu, MD, Professor.
 - Harzem Özger, MD, Professor.
Istanbul University Istanbul Medical School, Department of Orthopaedics and Traumatology, Istanbul, Turkey.
- Correspondence : Fikri Erkal Bilen, MD, FEBOT, Okul Yolu sk Arkheon sitesi B2 Blok D6 Ortakoy Istanbul 34347, Turkey. E-mail : dr@erkalbilen.com
-
- © 2009, Acta Orthopædica Belgica.



Fig. 1. — Preoperative radiological assessment. 1A : anteroposterior view ; 1B : lateral view of a patient with forearm deformity ; 1C : measurement of radial articular angle (RAA) ; 1D : measurement of the carpal slip ratio (CS).

measured and recorded (fig 1C & 1D). RAA was measured between two lines on AP films : one along the articular surface of the distal radius and the other perpendicular to a line from the centre of the radial head to the radial edge of the distal radial epiphysis (1). The normal value of RAA is 15 to 30°. CS was measured as the percentage of the lunate proximal surface in contact with the radius, as limited by a line drawn from the centre of the olecranon through the ulnar edge of the radial epiphysis (1). This line normally bisects the lunate, and the CS is abnormal when ulnar displacement of the lunate is more than 50%. Preoperatively, the average RAA was 43° (range : 32 to 75°), and the average CS was 69.5% (range : 43 to 100%). The mean shortening of the ulna prior to surgery, measured with a perpendicular drawn from the distal end of the ulna to the linear axis of the forearm, was 2.06 cm. We used Masada's classification (3) for guidance of the surgical procedure. In type I forearm deformities, the main osteochondroma formation is in the distal ulna. There is a relative shortening of the ulna and bowing of the radius. In type II forearm deformities there is radial head dislocation, either associated with an osteochondroma at the proximal radius (type IIA) or not associated with an osteochondroma at the proximal radius (type IIB). In type III deformities there is an osteochondroma formation at the distal radius which results in relative radial shortening. According to Masada's classification, five forearm deformities were of type I and the remaining three forearm deformities were of type IIB in our series (3).

The surgical procedures consisted of excision of the osteochondroma and lengthening osteotomy of the ulna by an external fixator. Circular external fixators (Ilizarov) were used in two forearms and unilateral exter-



Fig. 2. — The circular external fixator is assembled in step-cut fashion in order to allow elbow flexion.

nal fixators were used in six forearms (LRS, Orthofix, Bussolengo, Italy). The circular external fixator frame was assembled as two step cut half rings so that the proximal part permitted elbow flexion (fig 2). Two patients (# 4 and 5) required correction osteotomy and plate fixation of the radius due to radial bowing. After application of the external fixator, either circular or monolateral, immediate range-of-motion exercises were allowed (fig 3A & 3B). In patient # 2, distal radioulnar fixation was performed, and the ulna was lengthened until the radial head was relocated. The distal radioulnar fixation was then removed, and lengthening was continued. In the same session, an open lunar ligament reconstruction was carried out with triceps fascia strips. External fixators were removed upon completion of the consolidation phase. In two patients, intramedullary Rush pins were used to guide the lengthened ulna. Radiographs of a patient demonstrate the steps of the surgical procedure (figs 4A, 4B & 4C).

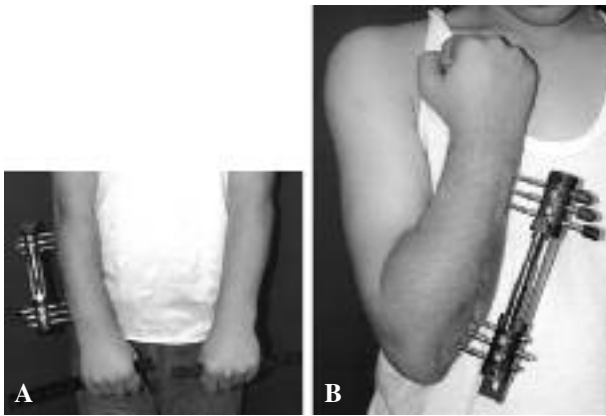


Fig. 3A and 3B. — Early function allowed with the external fixator.

RESULTS

The mean follow-up time was 40 months (range : 20 to 60 months). The average RAA improved from 43° to 35.5° (range : 28 to 56°) and the postoperative CS improved from 69.5% to 55% (range : 40 to 60%) (table I). The average postoperative shortening of the ulna was reduced from 2.06 cm to 0.44 cm (range : 0 to 1 cm). The radial bowings in two patients (no. 4 and no. 5) who underwent osteotomy and plate fixation were satisfactorily corrected and united. There was no limitation of the forearm and elbow range of motion postoperatively. In three patients with type IIB deformity the radial heads were relocated at a mean period of six weeks (patients # 2, 3 & 6). At the latest follow-up all radial heads remained relocated.

We encountered no recurrence of ulnar shortening. There were no nonunions and fractures of the regenerate bone. There were no post-operative neurological impairments. Two minor pin track infections were successfully treated by local wound care and oral antibiotics.

DISCUSSION

Multiple hereditary osteochondroma, also known as multiple hereditary exostosis, is an inherited skeletal dysplasia with an autosomal dominant pattern (5). About 40% of the cases are sporadic. Clinically, these patients have skeletal deformities

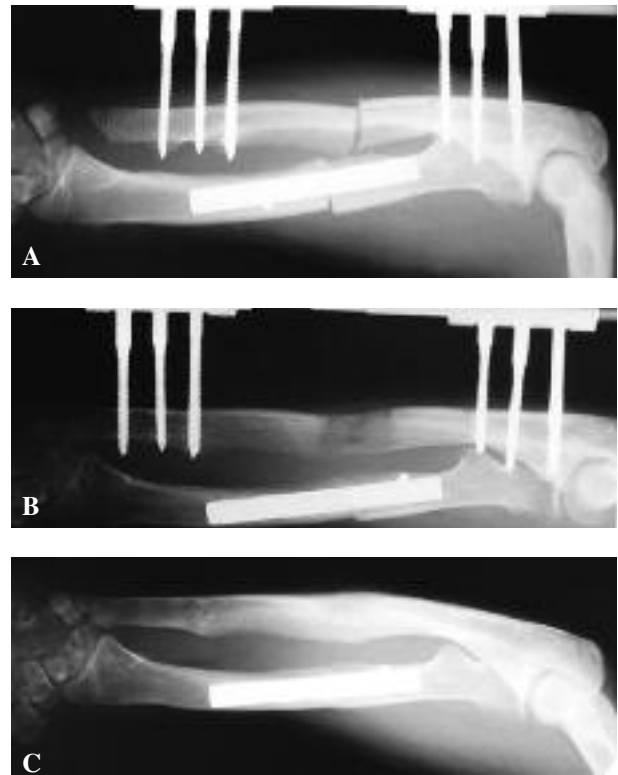


Fig. 4. — Radiographs showing the ulnar lengthening and radial correction osteotomies. A The ulna is osteotomized for lengthening, the radius is osteotomized, and stabilized by plate fixation following acute correction of the bowing ; B : New bone formation during lengthening ; C : Consolidation of the regenerate bone at the lengthening site. Note the overlengthening of the ulna at the distal radioulnar joint.

and short stature. The lesions are radiologically and histologically similar to solitary osteochondromas. In comparison to solitary osteochondromas, these patients have higher risk for malignant transformation (5 to 10% versus $< 1\%$). Deformities occur due to disorganized endochondral ossification in the growth plate and may require surgical correction, especially in the paired bones such as tibia/ fibula and radius/ulna (5).

Forearm deformities that occur in multiple hereditary osteochondroma are thought to be related to the difference in cross-sectional areas of the two distal physes and to the growth differential (3). Since the diameter of the distal ulna is significantly smaller than that of the distal radius, the ulna is more vulnerable to growth impairment. Additionally the contribution to the longitudinal growth of

Table I. — Patient demographics and outcome data (RAA : Radial Articular Angle, CS : Carpal Slip)

Patient #	Age, Gender	Forearm	Radial Head Dislocation	Masada Classification	Preop RAA (Degrees)	Postop RAA (Degrees)	Pre op CS (%)	Postop CS (%)	Preop Ulnar Shortening (cm)	Postop Ulnar Shortening (cm)	Complication
1	10, F	1	No	I	40	30	60	55	2.5	0	None
		2	No	I	32	28	75	60	2	0	Pin track infection
2	14, M	3	Yes	IIB	35	30	75	60	2	1	None
3	8, F	4	Yes	IIB	36	31	43	60	2	1	None
4	14, M	5	No	I	42	40	50	50	2.7	0.3	None
5	13, F	6	No	I	75	56	90	60	1.5	0.3	None
6	5, M	7	Yes	IIB	35	30	63	40	1.3	0.5	Pin track infection
7	9, F	8	No	I	50	39	100	55	2.5	0.4	None

the distal ulnar physis is about 10% more than that of the radius, which further increases the asymmetric growth in the affected forearms (3,6).

Masada *et al* proposed a classification for the forearm deformities caused by multiple hereditary exostoses in order to provide guidance for planning of the surgical treatment (3). They recommended excision of the osteochondroma in all cases, immediate ulnar lengthening for type I and gradual ulnar lengthening for type IIB forearm deformities. They also osteotomized the radius in most of the type I and type IIB deformities. In our series we performed excision of the osteochondromas, gradual ulnar lengthening in all cases, and osteotomy and plate fixation of the radius only if there was radial bowing.

Several authors recommend that the surgical intervention be delayed until after puberty to avoid recurrence of the deformities, which may occur when surgery is performed in a growing child (3). However this is usually demanding because of the disturbance caused to the parents by the cosmetic appearance, especially when the radial head is dislocated. Others recommend early intervention, because it has more potential for remodeling and leads to better surgical results (2,3,5,6,7).

In their series with forearms deformities caused by multiple cartilaginous exostoses, Matsubara *et al* overlengthened the ulna up to 0.5 cm plus variance (4). In patient # 6, we overlengthened the ulna

by 0.7 cm to prevent the recurrence of the length discrepancy between the ulna and the radius. At the latest follow-up we encountered no recurrence of the deformity in our patients.

There has been debate as for gradual versus acute lengthening of the ulna. Some authors reported neurovascular problems when acute ulnar lengthening exceeded 2 cm (4). On the other hand, Waters *et al* reported that it was safe to lengthen the ulna acutely up to 2.5 cm or 20% of its total length (8). Since we needed more than 2 cm ulnar lengthening in three patients, and 2 cm lengthening in another three patients, gradual lengthening was chosen.

All patients were satisfied with the cosmetic appearance as well as with the pain relief and improved function obtained postoperatively.

Although technically demanding, ulnar osteotomy and gradual lengthening by an external fixator provides promising results in the treatment of the forearm deformities caused by multiple osteochondromas. When planning the surgery, the expectations of the parents and the patients, as well as the risk for recurrence of the deformity especially in the younger child should be kept in mind.

REFERENCES

1. Fogel GRT, McElfresh EC, Peterson HA, Wicklund PT. Management of deformities of the forearm in multiple hereditary osteochondromas. *J Bone Joint Surg* 1984 ; 66-A : 670-680.

2. **Ip D, Li YH, Chow W, Leong JCY.** Reconstruction of forearm deformities in multiple cartilaginous exostoses. *J Pediatr Orthop B* 2003 ; 12 : 17-21.
3. **Masada K, Tsuyuguchi Y, Kawai H et al.** Operations of forearm deformity caused by multiple osteochondromas. *J Bone Joint Surg* 1989 ; 71-B : 24-29.
4. **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.
5. **Peterson HA.** Multiple hereditary osteochondromata. *Clin Orthop Relat Res* 1989 ; 239 : 222-230.
6. **Prichett JW.** Lengthening of the ulna in patients with hereditary multiple exostoses. *J Bone Joint Surg* 1986 ; 68-B : 561-565.
7. **Stanton RP, Hansen MO.** Function of the upper extremities in hereditary multiple exostoses. *J Bone Joint Surg* 1996 ; 78-A : 568-573.
8. **Waters PM, VanHeest AE, Emans J.** Acute forearm lengthening. *J Pediatr Orthop* 1997 ; 17 : 444-449.