J Orthop Sci (2006) 11:459–466
DOI 10.1007/s00776-006-1047-4

JOURNAL OF
ORTHOPAEDIC SCIENCE

Original article

Correction and lengthening for deformities of the forearm in multiple cartilaginous exostoses

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Abstract

Background. Multiple cartilaginous exostoses cause various deformities of the epiphysis. In exostoses of the ulna, the ulna is shortened and the radius acquires varus deformity, which may lead to dislocation of the radial head. In this study, we present the results of exostoses resection, with correction and lengthening with external fixators for functional and cosmetic improvement, and prevention of radial head dislocation.

Methods. We retrospectively reviewed seven forearms of seven patients who had deformities of the forearm associated with multiple cartilaginous exostoses. One patient had dislocation of the radial head. Operative technique was excision of osteochondromas from the distal ulna, correction of the radius, and ulnar lengthening with external fixation up to 5 mm plus variance. We evaluated radiographs and the range of pronation and supination. Furthermore, we conducted a follow-up of ulnar length after the operation.

Results. Dislocation of the radial head of one patient was naturally reduced without any operative intervention. At the most recent follow-up, six of the seven patients showed full improvement in pronation–supination. Ulnar shortening recurred with skeletal growth of four skeletally immature patients; however, it did not recur in one skeletally mature patient. Overlength of 5 mm was negated by the recurrence of ulnar shortening about 1.5 years after the operation.

Conclusions. We treated seven forearms of seven patients by excision of osteochondromas, correction of radii, and gradual lengthening of ulnas with external fixators. The results of the procedure were satisfactory, especially for function of the elbow and wrist. However, we must consider the possible recurrence of ulnar shortening within about 1.5 years during skeletal growth periods in immature patients.

Offprint requests to: H. Tsuchiya
Received: December 7, 2005 / Accepted: June 12, 2006

Introduction

Multiple cartilaginous exostoses are a hereditary condition in which exostoses are formed around the joints and cause various deformities of the epiphysis. Exostoses of the forearm are mostly formed in the distal ulna, which leads to growth disturbance. While growing, the ulna is shortened and the radius acquires varus deformity, and sometimes this leads to dislocation of the radial head. Some cases undergo malignant transformation and become secondary chondrosarcomas.[1]

Although various treatment alternatives have been proposed to manage such deformities and shortening of the forearm, lengthening of the ulna with correction only or with lengthening of the radius is considered to be the most appropriate treatment. However, most reports show only short-term results, and little is known about the postoperative course. In this study, we present the results of resection of exostoses, with correction and lengthening with external fixators for the purpose of functional and cosmetic improvement and prevention of radial head dislocation.

Patients

We retrospectively reviewed seven forearms of seven patients who had deformities of the forearm associated with multiple cartilaginous exostoses (Table 1). The patients were treated in our hospital between 1991 and 2002. There were three boys and four girls, ranging in age from 6.8 to 14 years (average, 10.8 years). The follow-up period ranged from 2.7 to 14.1 years (average, 7.1 years). Deformation was present in three right and four left forearms, dislocation of radial head was found in one patient, and two patients had a restriction of pronation before surgery. An Ilizarov fixator was used alone in one patient, monotube fixation in three patients, both in one patient, and an Orthofix (M-100;

Table 1. Demography of the patients

Case	Sex	Age (years)	Follow-up period (years)	Side	External fixator	Length gain (mm)	Correction angle of radius (°)	EFT (days)	EFI (days/cm)	DI (days/cm)
1	M	8.2	12.0	Left	Orthofix	30	25	139	46.3	22
2	M	14.0	14.1	Left	Orthofix	22	17	141	64.1	13.6
3	F	10.1	9.2	Left	Ilizarov	16	11	80	50	18.1
4	F	6.8	6.0	Left	Ilizarov + Monotube	30	25	128	42.7	13.7
5	F	11.3	2.7	Right	Monotube	36	16	115	31.9	11.9
6	F	11.2	3.7	Right	Monotube	41	19	155	37.8	10
7	M	11.4	3.1	Right	Monotube	15	0	126	97.3	22.7
Average		10.4	7.3			27	16.1	126	52.9	16.0

EFT, external fixation time; EFI (external fixation index) = EFT (days)/length gain (cm); DI (distraction index), periods of lengthening procedure (days)/length gain (cm)

Orthofix, Verona, Italy) in two patients. A unilateral fixator was selected when angulation of the radius could be safely corrected in an acute manner, followed by gradual lengthening of the ulna. Monotube (Stryker, Kalamazoo, MI, USA) or Orthofix (M-100) fixators were selected for their fit of the size of the ulna. The Ilizarov external fixator was used in cases that required gradual multifocal or oblique plane corrections or lengthening of the radius.

Surgical indications included radial head subluxation-dislocation, disturbance of activities of daily living because of limited or painful forearm rotation, progressive deformity, and undesirable cosmetic appearance.

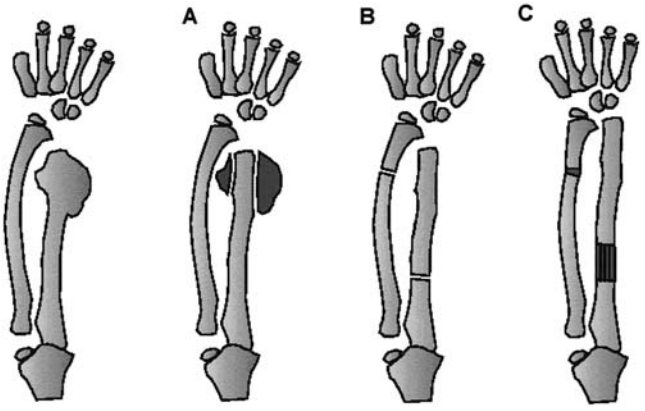

Fig. 1. Operative technique. **A** Excision of osteochondromas of distal ulna. *Blacked area*, osteochondroma. **B** Osteotomy. **C** Correction of the radius, with gradual lengthening of the ulna. *Blacked area*, callus

Operative technique

The operative technique was as follows (Fig. 1): (1) excision of osteochondromas of the distal ulna, (2) corrective osteotomy and internal fixation of the radius (five patients), or osteotomy and gradual correction of the radius with an external fixator (two patients), and (3) gradual lengthening of the ulna using an external fixator up to 5 mm plus variance, with the expectation of subsequent recurrence of ulnar shortening.

Evaluation

Radiographic review compared preoperative and most recent follow-up radiographs. We measured the radial articular angle (RAA; Fig. 2A), bowing of the radius, carpal slip (CS; Fig. 2B), and relative ulnar shortening (Fig. 2C).[2] We also evaluated the amount of ulnar lengthening, correction angle of the radius, external fixation time (EFT), external fixation index (EFI), and the distraction index (DI). EFI was obtained by dividing the total duration of external fixation by the length gained; DI was obtained by dividing the total duration of distraction by the length gained. We evaluated the range of pronation and supination clinically. Furthermore, we conducted a follow-up of ulnar length after the operation.

Results

Before the operation, the ulna was shortened by an average of 12.3 mm (range, 10–15 mm), resulting in a bowing of the radius by an average of 21.9° (range, 18°–27°). RAA before surgery was an average of 38.6°

Table 1. *Continued*

Radial articular angle (°)		Carpal slip (%)		Relative ulnar shortening (mm)		Bowing of radius (°)		
Preoperation	Final follow-up	Preoperation	Final follow-up	Preoperation	Final follow-up	Preoperation	Final follow-up	
47	30	100	50	15	0	19	19	
33	15	66	37.5	12	–1	27	11	
30	15	50	60	10	5	21	10	Relengthening, 13 mm
49	35	30	31	13	8	27	13	
32	24	66	30	15	3	18	0	
41	25	38	42	10	–5	25	0	Radial head subluxation
38	36	47	33	11	2	16	14	
38.6	25.7	56.7	40.5	12.3	1.7	21.9	9.6	

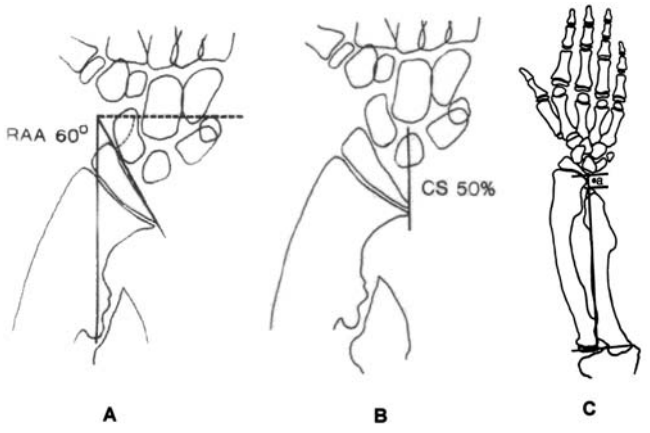

Fig. 2. Evaluation on radiographs. **A** Radial articular angle (*RAA*) is the angle between two constructed lines: one along the articular surface of the radius and the other perpendicular to a line that bisects the head of the radius and passes through the radial edge of the distal radial epiphysis (dashed line). **B** Carpal slip (*CS*) is measured as the percentage of contact of the lunate with the radius, determined by an axial line drawn from the center of the olecranon through the ulnar edge of the radius. **C** Ulnar shortening (*a*) is measured with a perpendicular drawn from the distal end of the ulna to the linear axis of the forearm

(range, 30°–49°), and CS before surgery was an average of 56.7% (range, 29%–100%).

At the most recent follow-up, RAA was an average of 25.7° (range, 15°–36°), improved compared with the normal side. The ulna was gradually lengthened by an average of 27 mm (range, 16–41 mm), resulting in 1.7-mm ulnar length minus variance at the most recent follow-up. However, in four patients shortening of the ulna recurred, and in one patient a 13-mm relengthening of the ulna was performed. CS was improved by about 16%. Correction angle of the radius was an average of 16° (range, 0°–25°) (Table 1).

EFT was an average of 126 days, EFI was an average of 52.9 days/cm, and DI was an average of 16.0 days/cm. Dislocation of the radial head of one patient was naturally reduced without any operative intervention. Before surgery two patients had a restriction of pronation; however, five patients had a restriction of pronation–supination just after completing the lengthening, which occurs because of the ulnocarpal impaction syndrome. At the most recent follow-up, six patients showed full improvement in pronation–supination; the remaining patient had a slight restriction of supination. All the tumors resected were pathologically confirmed as exostoses, and none was malignant. No tumor recurrence was found during the follow-up period. Superficial infection occurred in one patient and was treated with antibiotics, but there was no deep infection or breakage of wires or pins.

Ulnar shortening recurred with growth of four immature patients; however, it did not recur in one skeletally mature patient. The overlength of 5 mm was negated by the recurrence of ulnar shortening within about 1.5 years after the operation (Fig. 3).

Case presentations

Case 2 (Fig. 4)

A 14-year-old boy complained of deformity of the left forearm and limitation of pronation because of a giant

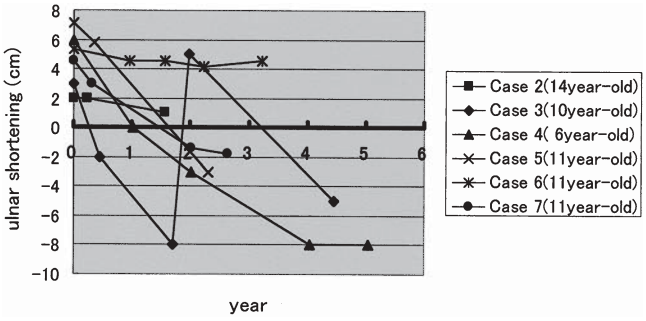

Fig. 3. Sequence of ulnar shortening

exotosis. Range of pronation was 30°. On X-ray, RAA was 33°, CS was 66%, and ulnar shortening was 12 mm (Fig. 4A,B). During the operation, the range of pronation improved to its full extent after resecting an exostosis of the distal ulna. The radius was corrected acutely and fixed with a plate, an Orthofix (M-100) external fixator was applied to the ulna, and osteotomy was performed (Fig. 4C). The ulna was lengthened 22 mm after a waiting period of 10 days (Fig. 4D). External fixation time was 141 days (Fig. 4E). At the most recent follow-up, he had no pain, unlimited range of pronation, and no recurrence of ulna shortening (Fig. 4F,G).

Case 6 (Fig. 5)

An 11-year-old girl complained of deformity of the right forearm and limitation of pronation. Range of pronation was 30°. X-ray radiography demonstrated that RAA was 33°, CS was 66%, ulnar shortening was 12 mm, and lateral dislocation of the radial head had occurred (Fig. 5A–C). The radius was corrected acutely and fixed with a plate, a monotube external fixator applied to the ulna, and osteotomy performed (Fig. 5D). The ulna was lengthened 45 mm after a waiting period of 7 days. The radial head was reduced without any open reduction for the dislocated radial head (Fig. 5E). External fixation time was 155 days (Fig. 5F). At 3 years 8 months after the operation, she had no pain and unlimited range of pronation. There was no hindrance to her activities of daily living. Furthermore, on X-ray radiographs, no recurrence of ulnar shortening or dislocation of the radial head could be seen (Fig. 5G,H).

Discussion

Multiple cartilaginous exostoses are a benign autosomal dominant skeletal dysplasia affecting bones formed by endochondral ossification. About 40% of the patients are sporadic. Estimations of its prevalence have ranged from between 0.9 and 1.4 in 100,000[3] in two European populations. There has been a wide variation in the reported rates of malignant degeneration of a benign exostosis to chondrosarcoma or to some other sarcoma in patients who have hereditary multiple cartilaginous exostoses. Reports have documented the risk to be 2%–5%.[4] Multiple cartilaginous exostoses often cause an obvious deformity of the forearm. The incidence of forearm lesion in multiple exostosis patients was reported to be between 39% and 59%.[3,5]

Prichett reported that the characteristic relative ulnar shortening is related to three factors.[1]

1. The ulnar physis had a cross-sectional area that is only one-quarter of that of the distal radius.
2. The distal ulna is more commonly involved in the condition than the distal radius.
3. The distal ulnar physis contributes more to total ulnar length than the distal radial physis contributes to radial length.

Osteochondroma in the distal ulna can result in a relative shortening of the ulna called candy-stick deformity, increased inclination of the distal radial articular surface with ulnar migration of the carpus, and sometimes proximal radial head subluxation or dislocation.[6]

For these deformities, several surgical interventions have been reported.[7–11] Fogel et al. reported three different procedures[7]: excision of osteochondromas only, ulnar lengthening with excision of osteochondroma, and ulnar lengthening with radial hemiepiphyseal stapling and excision of osteochondromas. Excision of distal osteochondromas did not improve the deformity, but it did reduce the preoperative rate of progression of ulnar shortening. Ulnar lengthening with excision of osteochondroma did not result in a significant improvement in forearm rotation, radial articular angle, or carpal slip. On the other hand, acute ulnar lengthening (0.3–1.6 cm) with radial hemiepiphyseal stapling and excision of osteochondromas led to good results. However, this procedure made the final length discrepancy unpredictable because the final length of the forearm could not be predicted.

Wood et al. reported other procedures: closing wedge osteotomy of the radius and fixing with a compression plate and screws, excision of the osteochondroma of the distal ulna, and ulnar lengthening using a long step-cut osteotomy and securing the bone in its lengthened position. Although appearance was markedly improved using this method, functional improvement was only minimal.[10]

Prichett reported that lengthening of the ulna and correction of the radius, particularly with the use of an external fixator, has given predictable results and is a useful method of treating forearm deformity in patients with hereditary multiple exostoses.[1]

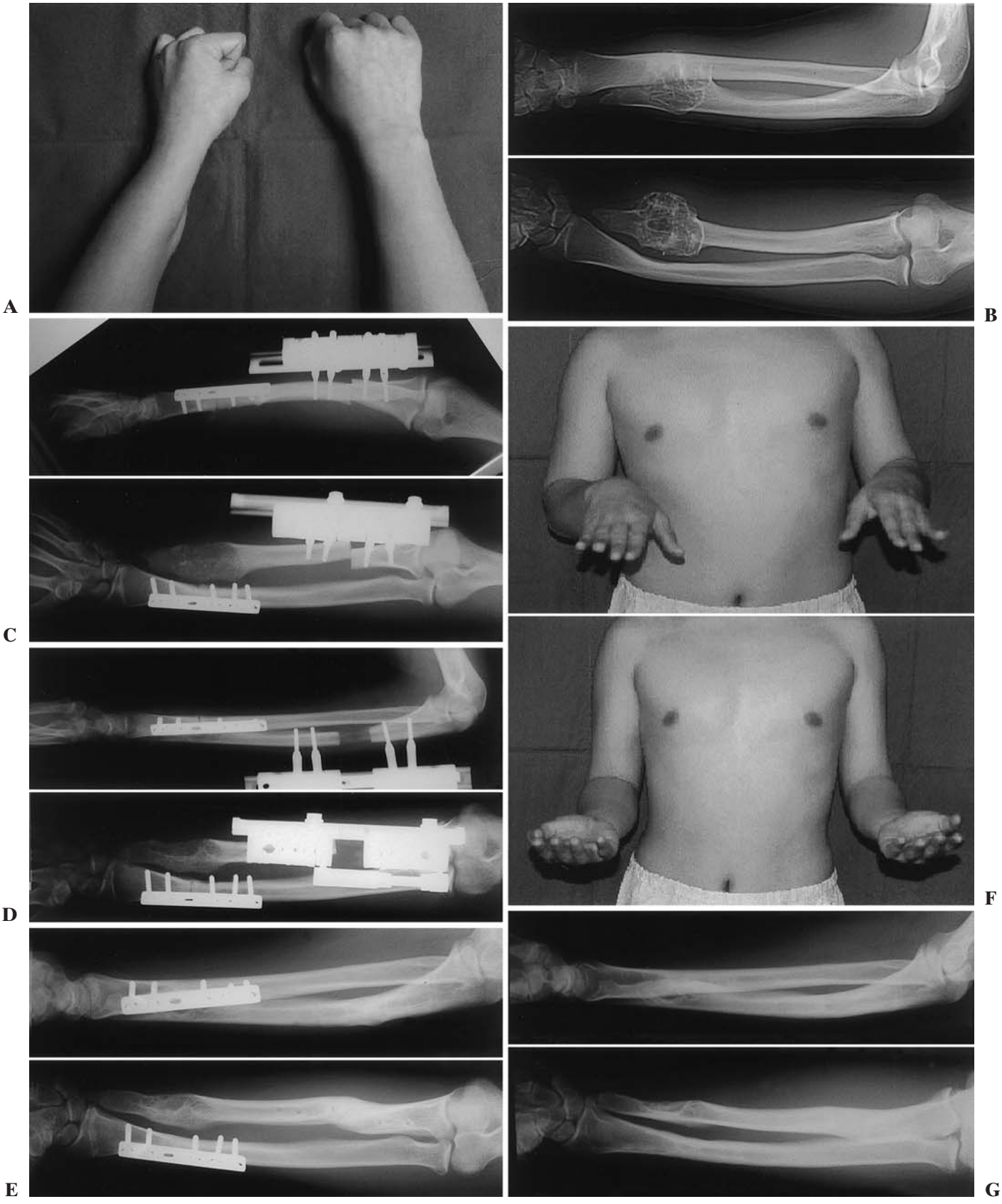

Fig. 4. Case 2: 14-year-old boy. **A** Restriction of pronation before the operation. **B** Radiographs before the operation (front and lateral view). **C** Radiographs after the operation (front and lateral view). **D** Radiographs after ulnar lengthening (front and lateral view). **E** Radiographs after the removal of external fixator (front and lateral view). **F** Full range of pronation and supination at 1.8 years after the operation. **G** Radiographs at 1.8 years after the operation (front and lateral view)

Fig. 5. Case 6: 11-year-old girl. **A** Restriction of pronation before the operation. **B** Deformity and shortening of her right forearm. **C** Radiographs before the operation (front and lateral view). *Arrow*, dislocation of radial head. **D** Radiograph during ulnar lengthening: radial head was not reduced (arrow). **E** Radiograph after ulnar lengthening: radial head was reduced naturally (arrow). **F** Radiographs after the removal of the external fixator (front and lateral view): radial head was reduced (*arrow*). **G** Full range of pronation and supination at 3.7 years after the operation. **H** Radiographs at 3.7 years after the operation (front and lateral view)

It appears that we have reached a consensus on the need for lengthening and normalization of the relationship of the radius and ulna, but controversy in the literature now exists regarding whether forearm lengthening is best achieved immediately or gradually. Some authors have reported that immediate lengthening of the ulna should be limited to 20 mm because greater lengthening can cause neurovascular problems.[6,12] On the other hand, Waters et al. reported that it was safe to lengthen acutely up to 25 mm or 20% of total length.[13]

To prevent possible recurrence, however, it would be better to overlengthen the shorter ulna. Most patients required lengthening from about 20 to 40 mm, considering the recurrence.[14] By lengthening gradually, patients can avoid neurovascular system problems and the shorter radius can be lengthened as well. Stapling or hemiepiphysiodesis of the distal radial physis, or shortening of the radius to prevent recurrence, was reported by Siffert and Levy[15]; this procedure is not acceptable because the affected limbs will be shorter. Furthermore, a dislocation of the radial head can be reduced using gradual lengthening with an external fixator.[1] Therefore, we believe that our method is a good way to treat forearm deformity caused by osteochondroma.

The timing of the surgery is extremely important, and there are many reports regarding this issue. Some recommend early intervention, because earlier intervention has more potential for remodeling and leads to better surgical results.[1,8,9,16,17] Others recommend intervention at a later age, because a recurrent operation can be avoided by postponing the procedure and good function can be acquired despite significant deformity after skeletal maturity.[14,18] We perform the operation if the patients have the indication without considering their age, because some patients lose function that cannot be regained by postponing the operation, especially with a dislocation of the radial head.

There are some reports of follow-up of lower leg lengthening,[19–21] and there is a smaller body of literature regarding middle- to long-term follow-up of ulnar lengthening. In this study, our results showed that an overlength of 5 mm was negated by the recurrence of ulnar shortening about 1.5 years after the operation. Dalmonte et al. reported that the tendency for recurrence of ulnar shortening was inversely proportional to the age at operation; it varied from 5.3 mm per year in patients aged 5 years to 2.7 mm per year in patients aged 11 years.[6] This finding is similar to ours and provides surgeons with useful information to explain the operation to the patients and their parents. However, our results demonstrated no relationship between the amount of recurrent ulnar shortening and patient age at operation. We hypothesize that the amount of recurrent ulnar shortening depends on the extent of damage of the distal ulnar growth plate caused by the osteochondroma.

Conclusions

We treated seven forearms of seven patients by excision of osteochondromas, correction of the radius, and gradual lengthening of the ulna with an external fixator. The results of the procedure were satisfactory, especially for the function of the elbow and wrist. However, we must consider the possible recurrence of ulnar shortening within about 1.5 years during skeletal growth periods in immature patients.

The patients and their families were informed that data from their cases would be submitted for publication and gave their consent.

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