Management of Masada type I/IIb forearm deformity by gradual ulnar lengthening in patients with hereditary multiple osteochondromas

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Research article

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Abstract

Background

Gradual ulnar lengthening is the most commonly used procedure in treatment of Masada type I/II deformity in patients with hereditary multiple osteochondromas. However, the treatment remains controversial for the recurrence of deformity in growing children and only slightly handicapped of daily life activities in untreated adult patients. The aim of this study was to evaluate mid-term clinical and radiological outcomes of ulnar gradual lengthening in our clinic.

Methods

We retrospectively reviewed patients who underwent ulnar lengthening by distraction osteogenesis from June 2008 to October 2017. Patients with less than two years of follow-up were excluded. The surgical procedures consisted of ulnar lengthening by external fixator and/or excision of the osteochondroma at the distal ulna. The carrying angle (CA) and range of motion (ROM) of the forearm and elbow were clinically assessed, and the radial articular angle (RAA) and ulnar shortening (US) were radiologically assessed before lengthening, two months after external frame removal, and at the last follow-up.

Results

The current study included 15 patients (17 forearms) with a mean age of 9.4 ± 2.3 years at index surgery. The mean follow-up period was 4.2 ± 2.4 years. There were 9 patients (10 forearms) with Masada type I deformity and 6 patients (7 forearms) with Masada type IIb deformity. The mean amount of ulnar lengthening was 4.2 ± 1.2 cm. The mean RAA improved from 37.3 ± 7.9° to 29.8 ± 6.5° initially (P = 0.003) and relapsed to 34.4 ± 7.6° at the last follow-up (p = 0.234). There was minimal deterioration of US yet significant improvement at the last follow-up compared to pre-op (p < 0.001). The elbow flexion and forearm pronation were improved significantly at last follow-up (p < 0.001 and p = 0.024, respectively). The mean carrying angle improved significantly from −6.3 ± 7.4° preoperatively to 7.4 ± 11.1° at last follow-up (p < 0.001).

Conclusions

Gradual ulnar lengthening significantly reduces cosmetic deformity and improves function; it is recommended for patients with Masada type I/IIb deformity. Therefore, we advocated aggressive individual treatment protocol for patients with Masada type I/IIb deformities.

Background
Hereditary multiple osteochondromas (HMO) is an autosomal dominant condition characterized by multiple benign cartilage-capped tumors, which typically occur at the juxta-epiphyseal region of tubular bones. Forearm deformities resulting from tumor-induced growth disturbances of distal radial and ulna are common, with a prevalence of 40–74% of HMO patients[1]. Masada et al. classified forearm deformities into three types[2]. In type I, the main osteochondroma is located in the distal ulna and results in ulnar shortening and ulnar deviation at the wrist, with a secondary bowing radius often observed. Type II is ulnar shortening with a dislocated radial head; this type can be further divided into two subgroups: type IIa (proximal radial osteochondroma involved) and type IIb (no proximal radial osteochondroma involved). In type III, the main osteochondroma involves the distal part of the radius with relative shortening of the radius. Because of the cross-sectional diameter of the distal ulnar physis being smaller than the radius, the ulna is more vulnerable to growth impairment[2], and therefore type I and type II are more common.

A shorter proportional ulnar length is associated with a diminished range of motion of the forearm in type I, while in type II the deformities result in restriction of both elbow movement and forearm rotation.[1, 2]. Gradual ulnar lengthening has been widely used with successful reported results in managing forearm deformity[3–7]. However, the treatment remains controversial. Stanton and Hansen[8] and Arms et al. [9] suggested less aggressive corrective surgery for forearm deformities because skeletally mature patients have a good tolerance to minimal functional impairment and appearance deformity. Furthermore, some long-term follow-up studies reported a high risk of recurrence of early intervention in skeletally immature patients [10–12]. The current study aim to evaluating our mid-term clinical and radiological outcomes of gradual ulnar lengthening for Masada type I/II deformity, and we hypothesized that simple gradual ulnar lengthening would be effectively improve the cosmetic problems and forearm function for Masada type I/II deformity.

**Methods**

This study received institutional review board approval. We retrospectively reviewed the medical records of all patients with HMO who underwent gradual ulnar lengthening in our hospital from June 2008 to October 2017. Patients with full radiographs pre- and postoperatively were included in this study and patients with less than 2 years of follow-up were excluded. The forearm deformity was classified according to Masada et al.’s classification[2]. Demographic data and chief complaints of patients prior to operation were recorded. Clinical results included the carrying angle (CA) and range of motion (ROM) of the forearm and elbow, compared between preoperative and last follow-up. The radiological assessment included the radial articular angle (RAA)[13], as measured on the anteroposterior radiograph, and ulnar shortening (US), as measured on the lateral radiograph (Fig. 1); these data were recorded before lengthening, 2 months after the external frame removal, and at last follow-up.

The operative procedure included gradual ulnar lengthening with an external fixator and/or excision of the distal ulnar osteochondroma. The lengthening began 10 days after surgery with distraction at a rate of 0.75 mm/day. The anticipated lengthening should meet the two following criteria. Proximally, the
radial head should pull the trochlear notch of the ulna, while distally, the positive ulnar variance should be obtained by overlengthening at 5–10 mm. The obtained lengthening was measured and recorded on film on the last day of lengthening.

The parametric Kolmogorov-Smirnov test was used to check for normal distribution of the data. A one-way analysis of variance was used to compare continuous variables. Post hoc tests were used to compare the radiologic values between pre-op and different follow-up stages, Levene’s test was used to compare variance between patients with Masada type I and type IIb deformity. A \( p \) value < 0.05 was considered statistically significant. All statistical analyses were conducted using the Statistical Package for the Social Sciences (SPSS Inc., Chicago, IL) version 17.0.

**Results**

This study included fifteen patients (17 forearms) with a mean age of 9.4 ± 2.3 years at index surgery. The mean follow-up period for all patients was 4.2 ± 2.4 years. The main complaints were cosmetic forearm deformities in all patients, followed with larger bone bump near the wrist joint in 10 forearms. All patients were free of pain preoperatively and at last follow-up. The mean ulnar lengthening was 4.2 ± 1.2 cm. Ten forearms were excised of osteochondromas concurrently at the distal ulna. There were 9 patients (10 forearms) who were Masada type I, with a mean age of 8.2 ± 1.5 years, and 6 patients (7 forearms) were type IIb, with a mean age of 11.0 ± 2.2 years. The ulnar was significantly shorter for patients with Masada type II deformity (\( p = 0.021 \)), demonstrating less pronation and flexion than those with type I deformity (\( p = 0.015 \) and 0002, respectively) (Table 1)
Table 1

Characteristics of 17 forearms before operation (Mean ± SD)

|                                  | Patient with Masada type I deformity | Patient with Masada type IIb | P value |
|----------------------------------|--------------------------------------|-----------------------------|---------|
| Age (Mean yrs ± SD)              | 8.2 ± 1.5                            | 11.0 ± 2.2                  | 0.006   |
| Length of follow-up              | 3.6 ± 1.4                            | 5.0 ± 3.4                   | 0.333   |
| RAA (°)                          | 34.1 ± 4.1                           | 41.9 ± 10.6                 | 0.106   |
| US (cm)                          | −2.3 ± 0.7                           | −4.0 ± 1.4                  | 0.021   |
| CA (°)                           | −8.9 ± 5.6                           | −2.6 ± 8.4                  | 0.082   |
| Pronation (°)                    | 56.0 ± 15.8                          | 32.9 ± 19.1                 | 0.015   |
| Supination (°)                   | 93.0 ± 9.5                           | 85.0 ± 22.3                 | 0.400   |
| Arc of forearm rotation (°)      | 149.0 ± 19.6                         | 117.9 ± 33.4                | 0.028   |
| Flexion (°)                      | 140.5 ± 6.4                          | 128.6 ± 6.9                 | 0.002   |
| Lack of extension (°)            | 0.5 ± 2.8                            | 2.8 ± 5.7                   | 0.273   |
| Arc of elbow motion (°)          | 140.0 ± 5.8                          | 125.7 ± 6.7                 | < 0.001 |

RAA, radial articular angle; US, ulnar shortening; CA, carrying angle

The preoperative data was compared between patients with Masada type I and Masada type IIb. The preoperative, postoperative, and last follow-up RAA and US values are shown in Table 2. Initially, the mean RAA was improved significantly after external fixator removal (p = 0.003); relapse occurred at the last follow-up. The mean change in US was −3.0 ± 1.3 cm preoperatively and was reduced significantly to 0.8 ± 0.5 cm overlength of the ulna after fixator removal (p < 0.001). There was some regression with a mean −0.8 ± 0.9 cm of ulnar shortening at the last follow-up examination, but overall there was a significant improvement compared to pre-op (p < 0.001).

Table 2

Radial parameters and clinical functional movement of patients (Mean ± SD)

|                   | Preoperative | Postoperative | Last follow-up | P₁ value | P₂ value |
|-------------------|--------------|---------------|----------------|----------|----------|
| RAA (°)           | 37.3 ± 7.9   | 29.8 ± 6.5    | 34.4 ± 7.6     | 0.003    | 0.234    |
| US (cm)           | −3.0 ± 1.3   | 0.8 ± 0.5     | −0.8 ± 0.9     | < 0.001  | < 0.001  |

P₁, data between the pre-op and post-op; P₂, data between the pre-op and last follow-up
At the last follow-up, the carrying angle and the pronation improved significantly \((p < 0.001)\). Although supination deteriorated slightly \((p = 0.155)\), significant improvement of the total arc of forearm rotation was observed \((p < 0.001)\). The flexion of elbow improved significantly \((p = 0.024)\), while the mean arc of flexion and extension of elbow did not reach statistical significance \((p = 0.084, Table 3)\).

One patient who was originally diagnosed as Masada type I deformity progressed to type II deformity at the last follow-up, experiencing a fracture at the callus site after external frame removal. A similar fracture also occurred in one patient with Masada type II deformity. Although the radial heads have been pulled to the level of trochlear of the ulna, no patient with type II deformity had a concentric reduction of the radiocapitellar joint after external frame removal (Fig. 1). Three patients had severe lateral posterior radial head dislocation at the last follow-up, but there was no obvious deformity of forearm appearance with a mean carrying angle of 11°.

### Table 3

| Range of movement and carrying angle based on follow-up stage (Mean ± SD) |
|------------------------------------------------|
| Preoperative | Last follow-up | P value |
| CA (°)        | −6.3 ± 7.4     | 7.4 ± 11.1 | < 0.001 |
| Pronation (°) | 46.5 ± 20.4    | 73.2 ± 15.8 | < 0.001 |
| Supination (°) | 89.7 ± 16.0    | 84.4 ± 10.9 | 0.155 |
| Arc of forearm rotation (°) | 136.2 ± 29.7 | 157.7 ± 23.1 | < 0.001 |
| Flexion (°)   | 135.6 ± 8.8    | 140.6 ± 8.1 | 0.024 |
| Lack of extension (°) | 1.5 ± 4.2 | 2.9 ± 5.3 | 0.351 |
| Arc of elbow motion (°) | 134.2 ± 9.4 | 137.6 ± 9.2 | 0.084 |

### Discussion

The osteochondroma-induced growth disturbance of the distal ulnar physis usually leads to unbalanced growth between the radius and ulna, resulting in functional limitation of the forearm and elbow, as well as an unappealing cosmetic appearance of the arm. Ulnar lengthening by distraction osteogenesis with or without an associated procedure has been the most prevalent procedures in treatment of Masada type I/II deformity\[14\]. However, the necessity of operation for such deformities is still controversial, and appropriate treatment is challenging, especially for children. The core of these problems is the concern of recurrence after surgery in skeletally immature patients\[2, 10, 15\], and less restriction of daily life activities in adult patients children\[8, 9\]. Noonan et al. found that only 13% of untreated adult patients, often with varying degrees of functional deficit, had substantial pain or limitations related to their job performance\[16\], and patients were found adaptable and with excellent Quick DASH questionnaire.
responses in previous study[10]. Unlike adult patients, self-assessed functional impairment and patient satisfaction can't be evaluated in children. The indications for surgery in the present study was cosmetic forearm deformity, cubitus varus and/or the protrusion of the radial head at the elbow and the large bump (osteochondroma) at the distal ulna were the initial problems noted by their parents, while functional limitations were observed through examination by a doctor rather than by the parents.

We advocated aggressive intervention for such deformities for several reasons as follow. Our data confirms that forearm deformity progresses with age, and functional impairment in Masada type IIb was severer (Table 1). Clement and Porter reported that radial head dislocation and a shorter ulna are independent risk factors associated with forearm rotation[1]. Early intervention can maintain a normal radiocapitellar joint with less functional impairment. Furthermore, a reduction of a dislocated radial head might be more difficult than prevention of radial head subluxation. In this study, good congruency of the radiocapitellar joint was observed in all patients with type I deformity except one, who progressed to radial head dislocation at last follow-up, and the patient suffered a callus fracture after external fixator removal and refused to have another ulnar lengthening during the follow-up. By contrast, no concentric reduction of the radiocapitellar joint was obtained in patients with type II deformity, we attribute this phenomenon to the adaptive lateral angulation of the proximal radius and other pathological changes of the soft structure of the elbow due to the long-standing dislocation of the radial head (Fig. 1); simple ulnar lengthening and other ligament reconstruction surgeries are rarely successful in such conditions[11]. Similar results were reported by Hsu et al, who treated 14 patients by ulnar lengthening and suggested ulnar lengthening along in treating patients younger than 10 years, while additional radial corrective osteotomy to correct the radial bowing deformity for older than 10 years patients[17]. Meanwhile, Iba and colleagues reported good results by using simple axis correction and ulnar osteogenesis distraction without corrective osteotomy of the radius to treat radial head dislocation within one year in HMO patients[18], Ahmed showed similar results in a series of 12 Masada type IIb forearmed patients, showing that a spontaneous reduction was achieved by ulnar lengthening in all patients[19], and demonstrating that the short interval (12 months) between occurrence of radial head dislocation and surgery is the key to a successful procedure. All these studies showing that earlier intervention with simple procedure has better outcomes. In addition, we found that in patients with severe dislocation, the pulling force of the interosseous membrane is limited and could no longer pull the radius moving distally after a large amount of lengthening, continuous traction only resulted in a positive ulnar variance without any displacement of radial head (Fig. 1).

However, in a study with longer-term follow-up, Akita et al reported recurrence in children who underwent surgery too early[11], similar results were reported by Litzelmann[10]. Therefore, Abe et al. proposed postponing ulnar lengthening until after 10 years old to avoid recurrence of deformities[20]. Ham et al. also suggested delaying the lengthening until after the age of 13–15 years[12]. Other studies reported satisfactory results after ulnar lengthening, but the mean follow-up times of their patients were short (< 2.5 years)[3, 4] or the age at intervention was older than 10 years[5]. With a mean follow-up time of 4.2 years, the analysis of the radiographic parameters in this study shows initial improvement and then deterioration at last follow-up, however, the ulnar shortening and forearm pronation improved
significantly by ulnar lengthening, and the cosmetic appearance improved significantly with the change of carrying angle (Fig. 2). As described previously in literature, mild ulnar shortening was acceptable in patients [2, 21], repeated lengthening is not recommended unless a progressive radial head dislocation is detected. We performed 0.5–1.0 cm overlengthening of the ulna in this study, but the limited overlengthening of the ulna seems not useful in the prevention of recurrence in skeletally immature patients. As the natural course of the deformities is unpredictable, the exact length discrepancy between the radial and ulna at skeletal maturity is difficult to predict. Furthermore, the forearm is comprised of two bones, the ulna and radius, which articulate with each other at the proximal and distal radioulnar joints. Thus, unlike one bone lengthening, the ulna cannot be lengthened to the anticipated length at one stage without considering the balance between the radius and ulna. In addition, some authors proposed that ulnar overlengthening could result in ulnocarpal impaction and should be not recommended even in patients with higher risk for recurrence [22, 23]. In contrast, Hill et al., who mentioned that overcorrection was partially successful at preventing subluxation or dislocation of the radial head [24]. We share the same experience, we have no patient complained about the pain of wrist joint, and even by overlengthening, slight ulnar shortening still exists at the last follow-up. We consider minimal overlengthening of ulnar could partially compensate for the growth imbalance between the radius and ulna (Fig. 1) and could thus delay the second lengthening of ulna.

Unlike reported previously in the literature, the age that ulna lengthening should be performed is not fixed and is dependent on the severity of the deformities in present study. Matsubara et al proposed that the recurrence of ulnar shortening might depend more on the extent of damage to the distal ulnar physis [22]. We recommend an individualized treatment plan for Masada type I deformity. In our study, surgery is not usually arranged at the patient’s first clinical assessment, unless a tendency of dislocation of radial head is observed (Fig. 3). For Masada type IIb deformity with new-onset radial head dislocation, the ulnar lengthening can be postponed no more than 1 year. And for a long-standing dislocation radial head, a concentric reduction might not be obtained by simply ulnar lengthening without radial osteotomy.

There are a few limitations of this study that warrant discussion. First, this is a retrospective study with a small number of patients. Second, we did not measure the ROM of the wrist joint and functional performance, which are important in evaluating stability of the wrist. In addition, most patients in this study were skeletally immature at last follow-up. Therefore, the recurrence risk could not be precisely evaluated. A longer follow-up duration and more detailed data of functional assessment are required to comprehensively assess the results of ulnar lengthening surgery. Furthermore, there is a lack of comparative studies in the literature with regard to functional outcomes for untreated, skeletally immature patients with HMO. Future randomized control studies are needed to better understand these issues. As reported by previous work [11, 13, 25], the pronation improved may be partially due to the excision of the osteochondroma, these results warrant further control studies to specifically evaluate the effect of ulnar lengthening on forearm rotation.

**Conclusion**
Even with a risk of recurrence, gradual ulnar lengthening is worthwhile to maintain function and improve forearm appearance for patients with Masada type I/IIb deformity. The long-standing dislocation of the radial head could not be reduced concentrically by simple gradual ulnar lengthening; thus, we recommended a scheduled follow-up for patients with Masada type I deformity, early intervention should be considered if there is a tendency or new onset of dislocation of the radial head.

**Abbreviations**

HMO: Hereditary multiple osteochondromas; CA: carrying angle; ROM: range of motion; RAA: radial articular angle; US: ulnar shortening; DASH: The Disabilities of the Arm, Shoulder and Hand Score

**Declarations**

**Ethics approval and consent to participate**

Ethical approval was obtained from the Ethics Committee of Shanghai Children's Medical Center. The written informed consent was obtained from all patients' parents and guardians.

**Consent for publication**

Consent for publication was obtained the from parent or guardian.

**Availability of data and material (data transparency)**

The datasets generated and/or analyzed during the current study are not publicly available because they contain patients' personal information, but are available from the corresponding author on reasonable request.

**Competing interests**

The authors declare that they have no conflict of interest.

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**Authors' contributions**

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by Haoqi Cai. Writing - original draft preparation: Yuchan Li. Writing - review and editing: Yuchan Li, Zhigang Wang. Supervision: Mu Chen, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Figures
(a) Type II deformity in a patient with HMO, adaptive lateral angulation of the proximal radius due to the long-standing dislocation of the radial head; (b) In lateral view, radial length (RL) is the distance from the center of the proximal radial physis to the center of the distal radial physis. Ulnar length (UL) is the distance between the trochlear and the ulnar styloid on the longitude axis of the ulna; ulnar shortening was determined by the RL minus the UL; (c and d) The ulna was overlengthened for 1.5 cm, and the radial head was not reduced by gradual ulnar distraction; (e and f) The patient came back 3.1 years after surgery, a computed tomography (CT) was performed, demonstrating the posterolateral displacement of the radial head, but the distal radioulnar joint was remolding well.

![Figure 2](image)

**Figure 2**

(a–c) A 12-year-old girl with type IIb deformity. Her parents complained about an obvious cubitus varus and protrusion of the radial head at the elbow but did not notice functional limitation of the forearm; (d–f) Both appearance and rotation improved at 2.5 years follow-up.
Figure 3

(a and b) Forearm deformity progressed with age, radial head proximal migrant was found 3 years later after first clinical assessment, ulnar gradual lengthening with osteochondroma resection was performed at 10-year-old; (d and e) Lateral radiograph of the same patient; (c and f) 2.1 years after the external fixator removed, both the proximal radioulnar joint and radiocapitella joint were well maintained.