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# Ulnar lengthening for children with forearm deformity from hereditary multiple exostoses: a retrospective study from a tertiary medical center



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# **Abstract**

**Background** Patients with hereditary multiple exostosis (HME) usually present with forearm deformity with or without radial head dislocation. Ulna lengthening has been proposed to address this condition. Exostosis resection plus ulna lengthening has been adopted in our hospital since 2008, and patients with this condition were retrospectively reviewed. Herein, we aimed to investigate the optimal timing and clinical outcomes of this surgical approach.

**Methods** In all, thirty-five patients (40 forearms), including 22 boys and 13 girls, were enrolled in our study from July 2014 to September 2020. We divided the patients into 4 groups based on the age when they received surgery and the status of the radial head. Pronation and supination of the forearm, flexion and extension of the elbow, wrist ulnar deviation and wrist radial deviation, and radiological parameters including ulnar length (UL), ulnar variance (UV), the percentage of radial bowing (RB/RL), radio articular angle (RAA) and carpal slip (CS), were assessed and recorded.

**Results** The mean UL was significantly improved after surgery in four Groups (P<0.05). In patients with radial head dislocation, we found significant improvement in forearm, wrist function and elbow flexion (p<0.05). For the patients with radial head dislocation, the juniors demonstrated better improvement in % RB and RAA (p<0.05, p=0.003 and 0.031).

**Conclusion** Exostosis resection and ulna lengthening with unilateral external fixation can effectively improve the function and radiological parameters of forearm deformity in HME children. For patients with radial head dislocation, early surgery can achieve better results. For patients not associated with radial head dislocation, we recommend regular follow-up and surgical treatment after 10 years of age.

Keywords Hereditary multiple exostosis, Radial head dislocation, Forearm deformities, Ulnar lengthening

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# **Background**

Hereditary multiple exostosis (HME) is a type of skeletal dysplasia, with approximately 1/50,000 incidence in the population [1, 2]. Exostosis often grow in the metaphysis of the long bone and can cause growth disorders and deformities [1, 3]. Forearm deformities are reported in 30–60% of patients with HME. According to Masada's classification system [4], forearm deformities can be divided into four types, see Fig. 1. Clinically, HME presents with deformities such as wrist ulnar deviation, radial head dislocation, ulnar shortening, and radial bowing [1, 2, 4–6]. Patients with HME usually come to the hospital with local pain, functional limitations and appearance abnormalities.

The purpose of surgical intervention is to correct the skeletal deformity, reduce the dislocation of the radial head and improve the forearm function. Ulnar osteotomy combined with lengthening, simple radial head resection, and distal radial hemiepiphysiodesis has been reported to address this deformity [2, 6, 7]. However, the optimal treatment algorithm for patients with this condition remains controversial [2, 6–9].

In this study, we retrospectively reviewed 36 patients with this condition receiving ulnar osteotomy and lengthening, and patients at different age groups were compared. We aimed to evaluate the effect of ulnar osteotomy lengthening in patients of different ages and provide insights into the optional surgical timing.

# **Methods**

# **Patients**

In all, from July 2014 to September 2020, 52 children with forearm deformities from HME were admitted to our hospital. Finally, 35 patients (40 forearms), including

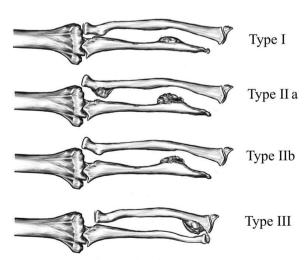


Fig. 1 Masada classification of the forearm osteochondroma

22 boys and 13 girls, were retrospectively reviewed, see Fig. 2. All patient information was obtained from the hospital database. The study was approved by the Ethics Committee of Tongji Medical College, Huazhong University of Science and Technology.

# **Grouping criteria**

The patients were grouped according to the status of radial head dislocation into the dislocation group and the non-dislocation group. Then they were subdivided into 4 groups according to their ages receiving surgery (age  $\leq$  10 years or >10 years): (1) Junior with radial head dislocation (The J-D group), (2) Junior without radial head dislocation (The J-N group), (3) senior with radial head dislocation (The S-D group), and (4) senior without radial head dislocation (The S-N group).

## Inclusion criteria

The patients had a diagnosis of HME and at least one of the following radiographic or functional parameters: (1) the ulna is shorter than the radius by > 5 mm, (2) a radial

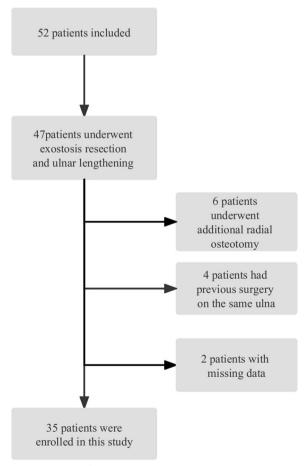
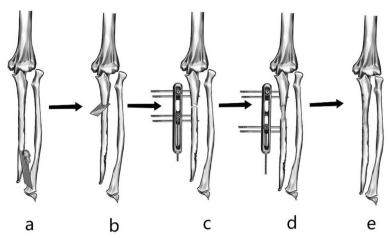


Fig. 2 Flowchart of patient selection

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**Fig. 3** Flowchart of patient selection. Osteochondroma resection(**a**), ulna osteotomy(**b**), ulna extension using a unilateral external fixator (**c**, **d**), remove the external fixator(**e**)



**Fig. 4** Radiological presentations of a 7-year-old boy with HME with a forearm deformity. Preoperative radiographs of the patient showed distal ulnar exostosis, ulnar shortening, and radial head dislocation (Masada IIb type) (a). Postoperative radiography of ulnar osteotomy and lengthening surgery (b). Radiological presentation of one, two and six months after surgery (c, d and e). Radiographic picture of fourteen months after surgery showing correction of the deformity and radial head reduction (f)

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**Fig. 5** Radiographic parameters. (1) RB, radial bowing (the maximum distance from the ulnar border of the radius to the long axis of the forearm was measured and then divided by the radial length). (2) UL, ulnar length (the length of the ulna was measured); UV, ulnar variance (the distal ulnar shortening from the most distal ossified point of the ulna to the plane of the distal radial surface); RAA, radio articular angle (the angle of inclination of the distal articular surface of the radius to the long axis of the forearm). (3) carpal slip (the percentage of the lunate on the ulnar side of a continuation of the linear axis of the forearm)

articular angle of >30 degrees, (3) a carpal slip of >60% (Fig. 5), (4) dislocation of the radial head, (5) limitation of forearm movement that interferes with daily activities. Patients who meet one or more of the 5 inclusion criteria are selected for the first time. Among all patients undergoing surgery, we excluded cases with the following conditions: (1) patients with metabolic diseases, (2) previous surgery on the same limb, (3) patients with incomplete data, (4) additional surgery for this treatment (other than exostosis resection and ulna lengthening) (Fig. 2).

# Surgical methods

Three senior surgeons performed surgeries. The treatment procedure is described in Fig. 3. All procedures were performed under general anaesthesia and tourniquet control (180-200 mmHg for a maximum of 60 min). According to the location of the exostosis, a longitudinal incision was made on the forearm. The preferred location for osteotomy was the proximal one-third to one-fourth of the ulna. The osteotomy was performed using a sharp drill and a chisel. Four Schanz pins (diameter 3 or 4 mm) were placed on the ulna in parallel. The external fixator (YIJIABAO, China) was then connected between these Schanz screws. All patients underwent ulnar or radius exostosis resection and ulnar lengthening using a unilateral external fixator.

One week after surgery, the ulna was extended in 0.25 mm increments 3 to 4 times daily to achieve a traction rate of 0.75 to 1 mm/ day. Follow-up was conducted in the second week, 1 month and every 1 month after. The increase in the ulna axis and degree of callus at the

osteotomy region could be monitored by X-ray. Radiological presentations of a 7-year-old boy with HME with a forearm deformity were showed in Fig. 4. Parents should pay attention to their child's forearm sensation, range of motion, and peripheral circulation. Parents could contact us by phone or visit the hospital whenever the children experience discomfort during the extension process. The distraction was discontinued when the radial head showed reduction or positive ulnar variance reached 5 mm (the

**Table 1** The demographic characteristics

	Number/Mean(±SD)	Percentage (%)
Sex		
Boys	22	62.9
Girls	13	37.1
Side		
Right	11	31.4
Left	19	54.3
Bilateral	5	14.3
The junior group(N/D)	23(12/11)	57.5
The senior group(N/D)	17(8/9)	42.5
Modified classification		
	16	40.0
lla	9	22.5
IIb	15	37.5
Age	9.04(±3.49)	
During of follow-up	21.22(±14.06)	

The junior group( $y \le 10$ ); The senior group( $y \ge 10$ )

N:without radial head dislocation

D:with radial head dislocation

SD Standard deviation

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ulna was longer than the radius). The external fixator was kept in place until the distracted region demonstrated consolidation on the X-ray, or a plate could be inserted to maintain the length until the bone union was achieved.

We performed a retrospective study in which two doctors measured and collected all data. Before and after surgery, we evaluated imaging and functional outcomes. Based on the radiographic imaging, the following parameters for each forearm were assessed in Fig. 5: (1) the percentage of radial bowing( radial bowing divided by radial length; RB/RL) (2) ulnar length, ulnar variance, radio articular angle (3) carpal slip; Considering that some of the younger children had incomplete metacarpal development, carpal slip was not included as a reference in the younger groups. Functional results include the pronation and supination of the forearm, the flexion and extension of the elbow, wrist ulnar deviation and wrist radial deviation.

# Statistical analysis

Statistical analysis was performed with SPSS statistical software (version 25.0; SPSS, Chicago, IL, USA). Continuous variables were summarized as the mean and the standard deviation (SD). The parametric Kolmogorov-Smirnov test was used to check for normal distribution of the data. A matched-sample T-test was used for functional and imaging indicators before surgery and at the last follow-up visit. Two independent sample T-tests were used to compare the imaging and functional indicators of the J-N and S-N groups. The J-D and S-D groups used the same method. The Fisher exact test was used for nominal variables. The level of significance was set at a *p*-value < 0.05.

# Results

# Study population

This study included 35 pediatric patients with HME (40 forearms). Twenty-two patients were boys, and 13 were girls. The mean age receiving the surgery was 9.04 (±3.49) years (range, 3.5 to 15.2 y; median= 8.3 y). The junior group included 20 patients (age range,3.5 to 9.8 y; median=7.5 y) and 23 forearms, including 11 forearms with radial head dislocation. The senior group included 15 patients (age range,10.2 to 15.2 y; median=12.5 y) and 17 forearms, including 9 forearms with radial head dislocation. The mean follow-up time was 21.22(±14.06) months. According to the Masada classification system, 16 forearms (40.0%) were classed as Type I; 9 (22.5%), as Type IIA; and 15 (37.5%), as Type IIB. The patient demographics are shown in Table 1.

# The junior groups

In the J-N group, the average UV decreased from  $0.53\pm0.23$ cm to  $-0.04\pm0.45$ cm (p=0.01). Forearm

pronation increased significantly from  $66.0\pm4.34^{\circ}$ to  $79.89\pm4.99^{\circ}(p < 0.001)$ . In the J-D group, the %RB and RAA of preoperative and last follow-up recorded p values of 0.003 and 0.031, respectively, which were statistically significant. Also, the forearm rotation function, Wrist ulnar deviation, and Wrist radial deviation were significantly improved (p<0.05).

# The senior groups

In the S-N group, the average CS decreased from  $54.12\pm22.01\%$  to  $40.24\pm14.12\%$  (p=0.039). Forearm pronation increased significantly from  $64.25^{\circ}\pm11.29^{\circ}$ to  $78.25^{\circ}\pm7.80^{\circ}$ (p=0.014). In the S-D group, the % RB and RAA of preoperative and last follow-up recorded p values of 0.083 and 0.069, respectively, which were statistically non-significant. However, the forearm rotation function, wrist ulnar deviation and wrist radial deviation were significantly improved (p<0.05).

## Compare

The mean UL was significantly improved after surgery in four Groups ( $P \le 0.05$ ). For patients without dislocation,

**Table 2** Radiographic outcomes in the patients without radial head dislocation

	Preoperative <sup>a</sup>	Last Follow-up <sup>a</sup>	Difference <sup>a</sup>	P Value
UL				
The J-N	12.95 <b>(</b> ±2.08 <b>)</b>	15.89 <b>(</b> ±2.31 <b>)</b>	-2.94 <b>(</b> ±2.05)	0.005*
The S-N	16.88 <b>(</b> ±2.83 <b>)</b>	18.96 <b>(</b> ±3.53 <b>)</b>	-2.08 <b>(</b> ±1.09 <b>)</b>	0.032*
P'Value	0.021	0.097	0.459	
UV				
The J-N	0.53 <b>(</b> ±0.23 <b>)</b>	-0.04 <b>(</b> ±0.45 <b>)</b>	0.57 <b>(</b> ±0.53 <b>)</b>	0.019*
The S-N	0.47 <b>(</b> ±025 <b>)</b>	0.13 <b>(</b> ±0.29 <b>)</b>	0.34 (±0.23)	0.062
P'Value	0.683	0.511	0.434	
%RB				
The J-N	9.40 <b>(</b> ±2.47 <b>)</b>	9.24 <b>(</b> ±2.90 <b>)</b>	0.17 <b>(</b> ±1.26 <b>)</b>	0.720
The S-N	9.48 <b>(</b> ±2.53 <b>)</b>	8.31 <b>(</b> ±2.57 <b>)</b>	1.17 <b>(</b> ±1.82 <b>)</b>	0.288
P'Value	0.960	0.602	0.285	
RAA				
The J-N	24.18 (±3.09)	22.46 <b>(</b> ±2.22 <b>)</b>	1.72 <b>(</b> ±3.14 <b>)</b>	0.165
The S-N	29.26 (±4.07)	26.80 <b>(</b> ±4.01 <b>)</b>	2.47 <b>(</b> ±3.52 <b>)</b>	0.256
P'Value	0.036	0.034	0.717	
CS				
The J-N				
The S-N	54.12 <b>(</b> 22.01 <b>)</b>	40.24 <b>(</b> ±14.12 <b>)</b>	15.24 <b>(</b> ±8.02 <b>)</b>	0.039*
P'Value				

The S-N Group consists of patients older than 10 years without radial head dislocation (n=8)

The J-N Group consists of patients aged 10 years or less without radial head dislocation (n=12)

 $\it UL$  Ulnar lengthening,  $\it UV$  Ulnar variance,  $\it RB$  Radial bowing,  $\it RAA$  Radial articular angle,  $\it CS$  Carpal slip, P data between the pre-op and the last follow-up; P' data between the junior group and the senior group

<sup>&</sup>lt;sup>a</sup> The values are given as the mean and the standard deviation

<sup>\*</sup> denotes *P* < 0.05

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there was no significant difference in imaging and functional improvement between the younger and older groups. For patients with radial head dislocation, the radial head was reduced at the last follow-up. The difference in radiographic indexes was mainly %RB and RAA. In the J-D group, the %RB and RAA of preoperative and last follow-up recorded p values of 0.003 and 0.031, respectively, which were statistically significant. The S-D group recorded P values of 0.083 and 0.069, respectively, which are statistically non-significant. In patients with radial head dislocation, we found significant improvement in forearm, wrist function and elbow flexion. However, there was no difference in the improvement of functional parameters before and after operation between the younger group and the older group. Table 2 and Fig. 6 compare the radiographic parameters between groups J-N and S-N. The radiographic results of patients in

groups J-D and S-D are shown in Table 3 and Fig. 7. The functional results in patients are shown in Tables 4 and 5.

Radial head reduction was not achieved in 4 patients at the end of the lengthening in the S-D group, and additional surgery of radial head reduction or resection was performed. Of all the patients, only one in the S-D group underwent a radial head resection. At the end of ulna lengthening, the head of the radius was excised, considering that the patient was older than 10 years and had severe deformation of the head that affected reduction. At the last telephone follow-up, none of the patients had further dislocation of the radial head. We found three patients in the J-D group who had relapsed. The main manifestations of recurrence were tumor growth and limited functional activities. One case of ulnar nonunion was also found. No postoperative impingement symptoms were observed in any of the wrists.

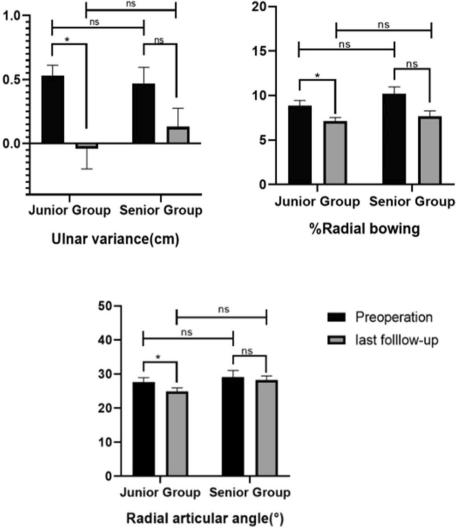


Fig. 6 Comparisons between preoperative and follow-up radiographic measurements in patients without dislocation of radial head. \* p < 0.05

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**Table 3** Radiographic outcomes in the patients with radial head dislocation

	Preoperative <sup>a</sup>	Last Follow-up <sup>a</sup>	Difference	P Value
UL				
The J-D	10.96 <b>(</b> ±1.72 <b>)</b>	14.30 <b>(</b> ±2.11 <b>)</b>	-3.34 <b>(</b> ±1.51)	0.000*
The S-D	14.45 <b>(</b> ±2.09 <b>)</b>	17.34 <b>(</b> ±3.57 <b>)</b>	-2.89 <b>(</b> ±1.94 <b>)</b>	0.002*
P'Value	0.001*	0.029*	0.573	
UV				
The J-D	0.91 <b>(</b> ±0.39 <b>)</b>	0.08 (±0.43)	0.82 <b>(</b> ±0.51 <b>)</b>	0.000*
The S-D	0.46 <b>(</b> ±0.51 <b>)</b>	-0.08 <b>(</b> ±0.29 <b>)</b>	0.54 <b>(</b> ±0.70 <b>)</b>	0.049*
P'Value	0.039*	0.340	0.310	
%RB				
The J-D	8.84 <b>(</b> ±1.99 <b>)</b>	7.12 <b>(</b> ±1.36 <b>)</b>	1.72 <b>(</b> ±1.45 <b>)</b>	0.003*
The S-D	10.21 <b>(</b> ±2.31 <b>)</b>	7.65 <b>(</b> ±1.88 <b>)</b>	2.56 (±3.86)	0.083*
P'Value	0.173	0.473	0.516	
RAA				
The J-D	27.64 <b>(</b> ±4.24 <b>)</b>	24.86 (±3.57)	2.78 <b>(</b> ±3.69 <b>)</b>	0.031*
The S-D	29.11 <b>(</b> ±5.87 <b>)</b>	28.23 (±3.58)	0.88 <b>(</b> ±5.43 <b>)</b>	0.639
P'Value	0.522	0.050	0.366	
CS				
The J-D				
The S-D	57.15 <b>(</b> ±18.12 <b>)</b>	40.42 (±18.07)	17.67 <b>(</b> ±12.10 <b>)</b>	0.003*
P'Value				

The J-D Group consists of patients aged 10 years or less with radial head dislocation (n=11)

The S-D Group consists of patients older than 10 years with radial head dislocation (n=9)

# **Discussion**

Gradual ulna lengthening can effectively improve the function and radiographic results of forearm deformity in HME patients. For patients with radial head dislocation, the improvement in function is more significant. Radiographic parameters showed that the improvement of radial deformities was more significant in younger patients with dislocation. Although no radial deformity correction was performed, % RB was improved during ulna lengthening, and the improvement of % RB and RAA was more pronounced in the J-D group (P = 0.003; P = 0.031), which is consistent with some studies [10-14]. Ulna lengthening facilitates the correction of radial deformities due to spontaneous bone remodeling in pediatric patients [10, 11]. The traction force in the ulnar against the radius through the distal radioulnar joint and interosseous membrane reduces the radial head and contributes to the improvement in the RB [12-14]. Hsu et al., in their study, observed similar results in patients ( $\leq 10$  years of age) who did not undergo corrective radial osteotomy [15].

An essential goal of surgical intervention for forearm deformities is to disrupt the progression of deformities and dysfunction, especially radial head dislocation [12].

Some scholars believe that resection of osteochondroma alone can effectively improve the functional activities of the forearm and delay the progression of the deformity [16, 17]. However, resection of osteochondroma alone did not improve the radiographic parameters [17, 18]. In our study, all patients underwent ulna and radius osteochondroma resection and gradual ulnar lengthening, and the patients achieved good radiological and functional results after surgery. Many studies have reported good radiographical and functional results after osteochondroma resection and gradual ulnar lengthening [10, 15, 19]. In addition, the recurrence of ulnar shortening and radial head dislocation can be reduced by the resection of osteochondroma before the correction of ulnar shortening and deformity [20, 21].

In recent years, gradual ulnar lengthening has been considered by most scholars as a crucial surgical step to correct forearm deformities [11, 22, 23]. However, the exact procedure and timing of surgical intervention remains controversial. First of all, the choice of osteotomy point is significant. The proximal third of the ulna for the osteotomy does not damage critical anatomical structures such as the interosseous membrane and the proximal oblique cord [24]. The tension of the interosseous membrane is conducive to reducing the radius head during the elongation process [20]. Subsequently, the ulna was gradually lengthened using an external fixator, which allows controlled lengthening and possible reduction of the radial head without significant soft tissue damage [25].

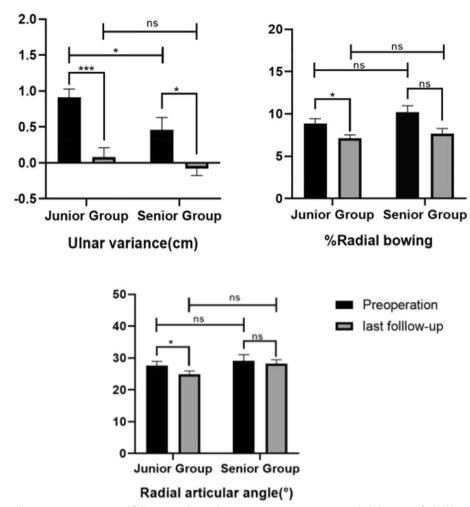
Regarding the selection of external fixators, many studies have applied unilateral external fixators to treat forearm deformities caused by HME and achieved good results [10, 11, 25, 26]. In this study, we used a unilateral external fixator to extend the ulna. During the follow-up, we found that due to the advantages of small size and easy postoperative care, most patients said the impact on their daily lives was acceptable. Also, when compared with other lengthening devices, the complication rates appear to be lower with unilateral fixators [27].

There is debate as to whether the outcome of early corrective surgery is superior to that of untreated patients in all patients [11, 18]. In this study, for type I patients, both age groups obtained good postoperative radiographical results. There was no significant difference between the two groups. For patients with radial head dislocation, both groups showed significant improvement in motor function except elbow flexion. However,

<sup>&</sup>lt;sup>a</sup> The values are given as the mean and the standard deviation

<sup>\*</sup> denotes *P* < 0.05

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significant improvement in radial malformations was found in the younger group on imaging (The % RB and RAA of preoperative and last follow-up in the J-D group recorded p values of 0.003 and 0.031). Hsu et al. reported that postoperative radial deformities were also significantly improved in the younger group of patients who underwent only gradual ulnar lengthening (The average RAA decreased from  $35.8^{\circ} \pm 5.2^{\circ} \text{to} 22.9^{\circ} \pm 4.7^{\circ} (p = 0.028)$ [15]. Some studies suggest early and aggressive treatment of forearm deformity to avoid deformity progression and dysfunction [12, 28-30]. Ahmed et al. suggested that early intervention is vital to achieve natural reduction of the radial head [13]. According to Peterson, preventing progressive deformities and dysfunction should be the primary goal [28]. Our findings support early surgery for patients with radial head dislocation, and surgery before the age of 10 can better correct the forearm deformity, probably due to robust bone remodeling.

Some studies have shown a high risk of re-dislocation of the radial head in younger patients [12, 16, 31]. Recurrence of ulnar shortening was also found in younger patients with MHE [29]. Some authors recommend delaying surgery to avoid recurrence [8, 31, 32]. Akita et al. considered that forearm deformities are unrelated to function and no longer recommend corrective procedures to prevent functional impairment. In their study, long-term follow-up (13 years) showed that children who had early surgery continued to relapse [18]. Some authors recommend overcorrection of the ulna by 5 to 10 mm in patients with skeletal immaturity to prevent recurrence. Pritchett reported the results of ulna lengthening in 10 forearms. He recommended excessive

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**Table 4** Functional assessment of forearms without dislocation of radial head

	Preoperative*	Last Follow-up*	Difference*	P Value
Forearm pr	onation			
The J-N	66.00(±4.34)	79.89(±4.99)	-13.87(±6.45)	0.000*
The S-N	64.25(±11.29)	78.25(±7.80)	-14.00(±5.48)	0.014*
P'Value	0.699	0.667	0.974	
Forearm su	pination			
The J-N	77.50(±5.88)	82.50(±4.31)	-5.00(±6.39)	0.063
The S-N	78.50(±3.11)	83.00(±4.97)	-4.50(±3.41)	0.078
P'Value	0.708	0.860	0.888	
Elbow flexion	on			
The J-N	130.50(±3.96)	134.00(±5.68)	-3.50(±3.46)	0.064
The S-N	131.33(±2.45)	138.75(±6.65)	-7.43(±7.08)	0.127
P'Value	0.714	0.224	0.215	
Elbow exte	nsion			
The J-N	0.13(±8.69)	0.63(±8.23)	-0.50(±3.51)	0.699
The S-N	-3.75(±8.18)	0.25(±4.11)	-4.00(±4.55)	0.177
P'Value	0.476	0.934	0.168	
Wrist ulnar	deviation			
The J-N	50.88(±18.15)	34.88(±6.33)	16.00(±13.05)	0.010*
The S-N	50.00(±14.05)	35.75(±9.78)	14.25(±4.99)	0.011*
P'Value	0.935	0.853	0.805	
Wrist radial	deviation			
The J-N	2.25(±3.65)	7.38(±4.56)	-5.13(±2.53)	0.001*
The S-N	0(±4.08)	8.75(±2.50)	-8.75(±4.79)	0.035*
P'Value	0.355	0.592	0.110	

<sup>\*</sup> denotes *P* < 0.05

correction in younger patients who underwent surgery before bone maturity because of the higher tendency for recurrence in younger patients [33]. However, complications of excessive prolongation, such as wrist impingement and ulnocarpal impaction syndrome, have also been observed [12].

In this study, the distraction was stopped when the radial head showed reduction or positive ulnar variance reached 5 mm (the ulna was longer than the radius). At the last telephone follow-up, we found three patients with type IIb who had relapsed and had limited forearm functional activity and were all younger than 10 years old at the time of their first surgery. The relapse may be caused by the insufficient lengthening of the ulna [27]. One patient in our study had functional activity limitations due to poor exercise after surgery. Therefore, it is also important to encourage patients to perform proper functional exercises after removing the external fixator.

There were certain limitations in our study. It was a retrospective case review, and the sample size was relatively small. A control group of non-operative patients is not

**Table 5** Functional assessment of forearms with dislocation of radial head

	Preoperative*	Last Follow-up*	Difference*	P Value
Forearm pro	onation			
The J-D	40.22(±7.72)	68.86(±7.03)	-28.65(±7.33)	0.000*
The S-D	37.56(±6.27)	62.44(±4.30)	-24.88(±4.57)	0.000*
P'Value	0.416	0.028*	0.198	
Forearm su	pination			
The J-D	65.00(±8.06)	78.96(±5.78)	-13.95(±5.36)	0.000*
The S-D	66.22(±6.70)	76.89(±5.01)	-10.67(±4.66)	0.000*
P'Value	0.721	0.410	0.166	
Elbow flexion	on			
The J-D	120.00(±10.79)	134.18(±6.57)	-14.18(±11.27)	0.002*
The S-D	114.56(±3.97)	131.33(±4.74)	-16.78(±4.15)	0.000*
P'Value	0.170	0.291	0.522	
Elbow exte	nsion			
The J-D	-3.55(±6.33)	2.18(±6.35)	-5.73(±9.02)	0.062
The S-D	-4.89(±9.57)	-0.33(±4.95)	-4.56(±6.73)	0.077*
P'Value	0.711	0.345	0.535	
Wrist ulnar	deviation			
The J-D	47.82(±13.95)	33.36(±6.19)	14.46(±9.98)	0.001*
The S-D	47.56(±10.88)	33.67(±5.27)	13.89(±7.25)	0.000*
P'Value	0.964	0.909	0.889	
Wrist radial	deviation			
The J-D	-2.45(±6.41)	5.64(±3.93)	-8.09(±4.81)	0.000*
The S-D	-3.00(±3.43)	2.00(±3.00)	-5.00(±5.03)	0.017*
P'Value	0.821	0.035*	0.178	

denotes *P* < 0.05

included in this study. In addition, follow-up until skeletal maturity is not available.

# **Conclusions**

In conclusion, gradual ulna lengthening can effectively improve the function and radiographic results of forearm deformity in HME patients. For patients with radial head dislocation, early surgery can achieve better results. For patients not associated with radial head dislocation, we recommend regular follow-up and surgical treatment after 10 years of age.

# Abbreviations

HME Hereditary multiple exostosis

UL Ulnar length UV Ulnar variance

RB/RL The percentage of radial bowing

RAA Radio articular angle

CS Carpal slip

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Not applicable.

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#### Authors' contributions

S W, P H and J L designed the review; S W and B H performed the literature searches; S W, P H, J L and ML acquired the data; S W performed the data analysis; SW, B H and YHW prepared and finished the manuscript; P H and J L reviewed the manuscript; S W, P H, J L, B H and BM supervised manuscript drafting and determined the final draft. PH and YHW are in charge of the language polishing and the grammar revision. All authors reviewed and approved the final version of the manuscript.

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## Availability of data and materials

The datasets used and/or analyzed during the current study available from the corresponding author on reasonable request.

#### **Declarations**

## Ethics approval and consent to participate

The studies involving human participants were reviewed and approved by the Ethics Committee of Tongji Medical College, Huazhong University of Science and Technology (ORG No: IORG0003571). Written informed consent was obtained from individual or guardian participants.

## Consent for publication

Not applicable.

# **Competing interests**

The authors declare no competing interests.

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## References

- Schmale GA, Conrad EU 3rd, Raskind WH. The natural history of hereditary multiple exostoses. J Bone Joint Surg Am. 1994;76(7):986–92.
- Zheng C, Han H, Cao Y. Older age and multi-joint external fixator are two risk factors of complications in ulnar lengthening in children with hereditary multiple exostosis. J Orthop Surg Res. 2020;15(1):555.
- Burgess RC, Cates H. Deformities of the forearm in patients who have multiple cartilaginous exostosis. J Bone Joint Surg Am. 1993;75(1):13–8.
- Masada K, et al. Operations for forearm deformity caused by multiple osteochondromas. J Bone Joint Surg Br. 1989;71(1):24–9.
- Canizares MF, Wall LB, Van Heest A, et al. Reliability of the Masada Classification for Forearm Involvement in Patients With Hereditary Multiple Osteochondromas (HMO). J Pediatr Orthop. 2023;43(1):e60–6.
- Huang P, Zhu L, Ning B. Forearm deformity and radial head dislocation in pediatric patients with hereditary multiple exostoses: a prospective study using proportional ulnar length as a scale to lengthen the shortened Ulna. J Bone Joint Surg Am. 2020;102(12):1066–74.
- Li Y, et al. Identification of risk factors affecting bone formation in gradual ulnar lengthening in children with hereditary multiple exostoses: a retrospective study. Med (Baltim). 2019;98(5):e14280.
- Beutel BG, Klifto CS, Chu A. Timing of forearm deformity correction in a child with multiple hereditary exostosis. Am J Orthop (Belle Mead NJ). 2014;43(9):422–5.
- Noonan KJ, et al. Evaluation of the forearm in untreated adult subjects with multiple hereditary osteochondromatosis. J Bone Joint Surg Am. 2002;84(3):397–403.

- Demir B, et al. Single-stage treatment of complete dislocation of radial head and forearm deformity using distraction osteogenesis in paediatric patients having multiple cartilaginous exostosis. Arch Orthop Trauma Surg. 2011;131(9):1195–201.
- D'Ambrosi R, et al. Gradual ulnar lengthening in children with multiple exostoses and radial head dislocation: results at skeletal maturity. J Child Orthop. 2016;10(2):127–33.
- 12. Vogt B, 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.
- Ahmed A. Gradual ulnar lengthening by an Ilizarov ring fixator for correction of Masada Ilb forearm deformity without tumor excision in hereditary multiple exostosis: preliminary results. J Pediatr Orthop B. 2019;28(1):67–72.
- 14. Ip D, et al. Reconstruction of forearm deformities in multiple cartilaginous exostoses. J Pediatr Orthopaedics-Part B. 2003;12(1):17–21.
- Hsu PJ, et al. Less is more: ulnar lengthening alone without radial corrective osteotomy in forearm deformity secondary to hereditary multiple exostoses. J Clin Med. 2019;8(11):1765.
- 16. Arms DM, et al. Management of forearm deformity in multiple hereditary osteochondromatosis. J Pediatr Orthop. 1997;17(4):450–4.
- 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.
- Akita S, et al. Long-term results of surgery for forearm deformities in patients with multiple cartilaginous exostoses. J Bone Joint Surg Am. 2007;89(9):1993–9.
- Song SH, et al. 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.
- Lu Y, et al. Distraction osteogenesis at the proximal third of the ulna for the treatment of Masada type I/IIb deformities in children with hereditary multiple exostoses: a retrospective review of twenty cases. Int Orthop. 2022;46(12):2877–85.
- 21. Ishikawa J, et al. Tumor location affects the results of simple excision for multiple osteochondromas in the forearm. J Bone Joint Surg Am. 2007;89(6):1238–47.
- 22. Hill RA, et al. Forearm lengthening by distraction osteogenesis in children: a report of 22 cases. J Bone Joint Surg Br. 2011;93(11):1550–5.
- El-Sobky TA, et al. Current paediatric orthopaedic practice in hereditary multiple osteochondromas of the forearm: a systematic review. Sicot J. 2018;4:10.
- Noda K, et al. Interosseous membrane of the forearm: an anatomical study of ligament attachment locations. J Hand Surg Am. 2009;34(3):415–22.
- Zhang R, et al. Hinge positioning method of Ilizarov apparatus in correcting radial head luxation caused by multiple hereditary exostoses. Jt Dis Relat Surg. 2022;33(1):40–50.
- Li Y, et al. Gradual ulnar lengthening in Masada type I/Ilb deformity in
  patients with hereditary multiple osteochondromas: a retrospective study
  with a mean follow-up of 4.2 years. J Orthop Surg Res. 2020;15(1):594.
- 27. Malot R, et al. Role of hybrid monolateral fixators in managing humeral length and deformity correction. Acta Orthop. 2013;84(3):280–5.
- Peterson HA. Deformities and problems of the forearm in children with multiple hereditary osteochondromata. J Pediatr Orthop. 1994;14(1):92–100.
- Fogel GR, et al. Management of deformities of the forearm in multiple hereditary osteochondromas. J Bone Joint Surg Am. 1984;66(5):670–80.
- Scheider P, Ganger R, Farr S. Age-related outcomes and complications of osteodistraction in the pediatric upper extremity: a large retrospective single-center study of 61 cases. J Pediatr Orthop. 2022;42(2):e181–7.
- 31. Abe M, et al. Lengthening of the forearm by callus distraction. J Hand Surg (Edinb Scotl). 1996;21(2):151–63.
- Ham J, et al. Multiple osteochondromas (MO) in the forearm: a 12-year single-centre experience. Strategies Trauma Limb Reconstr. 2016;11(3):169–75.
- 33. Pritchett JW. Lengthening the ulna in patients with hereditary multiple exostoses. J Bone Joint Surg Br. 1986;68(4):561–5.

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