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PII: S1877-0568(22)00300-0

DOI: https://doi.org/doi:10.1016/j.otsr.2022.103445

Reference: OTSR 103445

To appear in: *Orthopaedics & Traumatology: Surgery & Research*

Received Date: 2 May 2021

Accepted Date: 28 September 2021

Please cite this article as: Bom F, Barbier O, Libouton X, Docquier P-Louis, Forearm deformity in multiple hereditary exostosis. Radiologic predictors of radial head dislocation, *Orthopaedics and Traumatology: Surgery and Research* (2022), doi: https://doi.org/10.1016/j.otsr.2022.103445

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Original article

Forearm deformity in multiple hereditary exostosis. Radiologic predictors of radial head dislocation.

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Abstract
Introduction: Radial head dislocation in patients with multiple hereditary exostosis (MHE) can lead to functional deficit. We investigated whether the location of the exostosis and certain radiological criteria predict risk of radial head dislocation/subluxation.
Hypothesis: We hypothesized that the radiological criteria differentiate between patients who need closer follow-up of the forearm and others for whom multiple radiographs are superfluous.
Patients and Methods: We retrospectively reviewed the demographics of patients with MHE in our hospital, and radiographic measurements were made on forearm radiographs: radial length, ulnar length, ulnar variance, radial articular angle, and radial bowing.
Results: 49 forearms were analyzed in 30 patients. Mean age was 9.5 years at first evaluation and 11.8 years at last evaluation. Radial head dislocation or subluxation was found in 6 forearms (12%). Risk factors comprised isolated exostosis in the distal portion of the ulna or exostosis in the distal part of both the ulna and radius, radial or ulnar shortening > 4.6 cm, radial bowing > 8.1%, radial articular angle > 35°, and ≥ 3 exostoses in the forearm.
Discussion: In patients with MHE with risk factors for radial head dislocation, close follow-up with regular radiography is indicated and early surgery should be performed before the radial head dislocates.

Level of Evidence: IV; retrospective study
Keywords: multiple hereditary exostosis, osteochondroma, forearm deformity, radial head dislocation

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Introduction

Multiple hereditary exostosis (MHE) generally involves autosomal dominant transmission [1], although de-novo mutations may also occur. Most cases implicate EXT1 or EXT2 gene mutation [2]. Prevalence is 1/50,000 births [3,4]. Forearm involvement is seen in 30-74% of cases [5-17]. Location is typically in long-bone metaphyses and, in the upper limb, in the proximal humerus, distal radius, distal ulna or the hand [18]. Clinical manifestations occur in the first decade of life in 80% of cases [11,18]. In the forearm, functional deficits may be induced by ulnar or radial shortening or bowing, ulnar deviation of the wrist or radial head dislocation [7-10,15,19-21].

As the natural history of the pathology is poorly known, treatment options and timing are hard to define [8,10,15,22,23]. Good esthetic results are reported with surgery, although functional results are controversial [11,14,15,21-30]. It has, however, been shown that, once the radial head has dislocated, attempted reduction tends to be ineffective [6,11,15,17,28]. Intervention ahead of dislocation could avoid esthetic and functional complications.

We investigated whether exostosis location and certain radiological factors might predict radial head dislocation. The study hypothesis was that the radiological criteria can differentiate between patients who need closer follow-up of the forearm and others for whom multiple radiographs are superfluous.

Patients and methods

Review board approval

Institutional review board approval was obtained under n° B403201523492.

Patients

Sixty consecutive patients managed in our hospital for MHE between 2007 and 2021 were recruited. Patients without documented involvement or complete forearm radiography were excluded. Forearms without exostosis were also excluded (Figure 1). The following data were collected retrospectively: age at radiography, gender, and duration of follow-up.

Methods – Radiographic analysis

AP and lateral forearm radiographs were analyzed retrospectively by a single independent observer. Deformity was assessed following Burgess [18] (Figure 2): radial length (RL), ulnar length (UL), ulnar variance (UV), radial articular angle (RAA), and radial bowing (RB). Radial and ulnar shortening was determined by subtracting measured length from predicted length according to Burgess [18]. Proportional UL was determined as:

$$proportional\ ulnar\ length\ = \frac{ulnar\ length\ (cm)}{radial\ length\ (cm)}$$

Carpal slip (CS) was assessed as the percentage of the lunatum on the ulnar side of a projection of the forearm linear axis (LA) (Figure 3).

All forearm exostoses were located and any radial head dislocation was noted. Forearm deformities were classified following Jo [19] on initial X-rays.

Radial and ulnar z scores were calculated following Gindhart [31]: negative if measured length shorter than predicted length [5,31].

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$$radial\ z\ score = \frac{radial\ shortening\ (mm)}{standard\ deviation\ (\sigma)}$$

$$ulnar\ z\ score\ = \frac{ulnar\ shortening\ (mm)}{standard\ deviation\ \ (\sigma)}$$

Statistics
Statistical analyses used SPSS software, version 27 (IBM). The significance threshold was set at $p<0.05$, with Bonferroni correction in case of multiple tests (0.05/number of tests). Radiological factors were compared between groups with and without radial head dislocation on multivariate analysis (MANOVA). Jo classification groups were compared on chi² test. Normal distribution was checked on Kolmogorov-Smirnov test. Mean values were compared between Jo classification groups on 1-factor ANOVA.

Results

Series
There were 16 male and 14 female patients, for 24 left and 25 right forearms. Mean age at initial radiography was 9.5 years (range, 1.6-16.8), and 11.8 years (range, 4.8-30.6) at lastassessment. Mean follow-up (first to last consultation) was 50 months (range, 7-183).

Exostosis location
Table 1 shows exostosis location according to Jo (Figure 4). Radial head dislocation was found only in case of isolated distal ulna (n=2) or associated distal ulna and distal radius location (n=4) and never in isolated distal radius location ($p<0.0001$).

Radiography (Table 2)
Radial head dislocation was found in 6 of the 49 forearms (12%). The dislocation group showed significantly greater radial and ulnar shortening, with lower radial and ulnar z scores and greater RB (Table 3).
Exostosis was more frequent in the group with radial head dislocation (mean, n=3) than in the group without (mean, n=2) ($p=0.039$).
In types IIA, IIB and IVB (with radial head dislocation), there was significantly greater radial and ulnar shortening ($p<0.001$), lower radial ($p<0.001$) and ulnar z scores ($p<0.001$), greater RAA ($p<0.028$) and greater RB ($p<0.0001$). The same pattern was seen in type IVA, but was less pronounced. There was a trend toward negative UV in types I, IIA, IIB and III. There were no significant differences in CS or proportional UL.

Radiologic factors for radial head dislocation (odds ratio)
Radial shortening >22%, ulnar shortening >20%, RB >8.1% and RAA >35° were predictive of radial head dislocation (Table 4).

Discussion

The study hypothesis was confirmed: there were radiologic criteria differentiating patients at risk of radial head dislocation, requiring closer follow-up, versus those for whom iterative X-ray is superfluous.

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Most publications highlight only 1 or perhaps 2 predictive factors. Matsumoto et al. showed that the EXT1 mutation was associated with greater carpal slip and ulnar deviation than the EXT2 mutation [20]. Gottschalk et al. showed that forearms with isolated distal ulna exostosis more frequently developed radial head dislocation [5]. Radial head dislocation correlated significantly with negative UV [6,15]. Jo et al. showed that <0.9 proportional ulna shortening and ≥8.1% RB predicted radial head dislocation [19]. The present study sought to review all reported radiologic predictors of radial head dislocation and assess relevance in our series, and further to put forward new ones: ulnar and radial shortening and ≥3 forearm exostoses.

The impact of exostosis location on presence and severity of deformity confirmed previous findings [5,6,11,18,19]: isolated distal ulnar or associated distal ulnar and distal radial exostosis correlated with radial head dislocation. Forearms with >2 exostoses also showed elevated risk.

Radiologic predictive factors for radial head dislocation comprised radial and ulnar shortening, RB and RAA. According to Jo et al., RB >8.1% increases dislocation risk 5-fold. In the present series, RB >8.1% increased the risk 16.5-fold and RB >11.1% increased the risk 66.7-fold. RAA >35° increased the risk 16.5-fold. Increased RAA was identified according to soft tissue tether theory as preceding radial head dislocation [5,11-13,21]: distal ulnar growth defect followed by distal radial growth defect induced radial bowing.

In our study, neither CS, proportional UL nor UV proved to be independent factors for radial head dislocation. Meza et al. demonstrated that UV is greater in the early stages of the natural historyof the disease, with lower forearm UV progressing toward subluxation. Perhaps we did not haveenough early-stage MHE radiographs to be able to confirm this. MHE is a progressive disease,requiring monitoring: even if not present at a given moment, a predictive factor for radial headdislocation may appear at any point in the child's growth. Early surgery may be proposed when several of these predictive factors appear or show aggravation over time.

One limitation of the present study was the lack of systematic X-rays of both forearms at the first consultation if forearm symptoms were absent. This prevented tracking UV progression over follow-up. Also, no intra- or inter-observer reproducibility assessment was made for the radiographic measurements.

A third limitation was the lack of uniform standardized clinical assessment to evaluate functional results, and the lack of genetic data for some patients. The fourth limitation was the small number of patients included; however, MHE is a rare disorder.

Conclusion

We advocate assessing the following radiologic predictors of radial head dislocation in patients with multiple hereditary exostosis:
- isolated distal ulnar exostosis or distal radial and distal ulnar exostosis;
- >22% radial or >20% ulnar shortening;
- >8.1% radial bowing;
- radial articular angle >35°;
- ≥3 forearm exostoses.

In case of multiple predictive factors for radial head dislocation, the possibility of early surgery should be discussed with the patient and parents, as surgery needs to be performed before dislocation occurs [32].

Funding
The study received no outside funding.

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Disclosure of interest
The authors have no conflicts of interest to disclose.

Author contributions:
FB: data acquisition, article writing
OB: critical reviewing
XL: critical reviewing
P-LD: statistical analysis and interpretation, article writing

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Tables

Table 1: Jo forearm classification

Jo type (Figure 3)	**Number of forearms (%)**
Type I	10 (20%)
Type IIA	1 (2%)
Type IIB	1 (2%)
Type III	14 (29%)
Type IVA	19 (39%)
Type IVB	4 (8%)

Type I: distal ulnar exostosis; Type IIA: distal ulnar and proximal radial exostosis with radial head dislocation; Type IIB: distal ulnar exostosis with radial head dislocation; Type III: distal radial exostosis; Type IV: distal radial and ulnar exostosis, Type IVA: without radial head dislocation; Type IVB: with radial head dislocation.

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Table 2: Mean radiographic values

	Mean ± standard deviation	Minimum	Maximum
Radial shortening (cm)	2±2.3	-1.1	9.2
Radial shortening (%)	9.5±10.0	-8	35.9
Radial z-score	-1.8±1.9	-7.5	1.4
Ulnar shortening (cm)	2.2±2.5	-1.2	9.8
Ulnar shortening (%)	9.5±10.0	-8	35.9
Ulnar z-score	-1.9±2.1	-8.0	1.6
U/R ratio	1.1±0.1	0.9	1.2
Ulnar variance (cm)	-0.1±0.4	-1.5	1.2
Ulnar variance (%)	-0.4±2.9	-11.6	9.4
Carpal slip (%)	41±26	0	100
Radial articular angle (°)	32±9.0	3	51
Radial bowing (%)	7.7±2.7	2.8	15.6

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Table 3: Multivariate analysis (MANOVA) of predictive factors for radial head dislocation (p=0.03)

	No dislocation (N=43)	Dislocation (N=6)	p-value (multivariate analysis)
Radial shortening (%)	7.5	24	p<0.0001
Radial z-score	-1.4	-4.4	p<0.0001
Ulnar shortening (%)	7.5	24	p<0.0001
Ulnar z-score	-1.5	-4.9	p<0.0001
U/R ratio	1.1	1.1	NS
Ulnar variance (cm)	-0.06	-0.14	NS
Ulnar variance (%)	-0.29	-0.82	NS
Carpal slip (%)	40.6	41.2	NS
Radial articular angle (°)	31	41	p=0.009
Radial bowing (%)	7.2	11.4	p<0.0001
Number of exostoses	2	3	p=0.039

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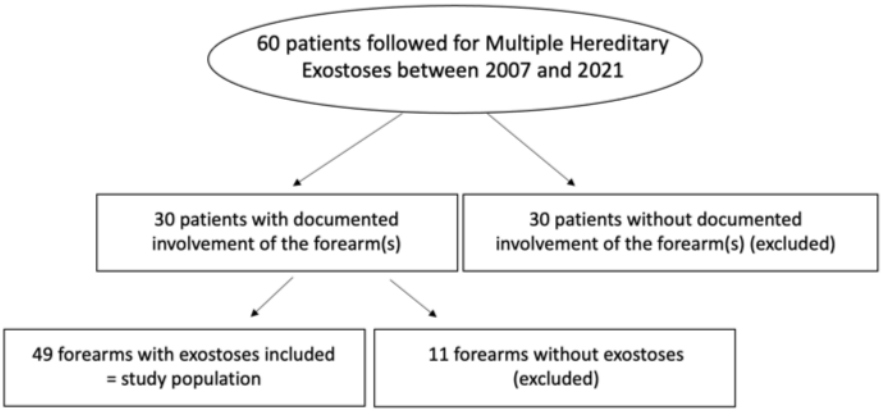
Table 4: Estimated radial head dislocation risk

Factor	Odds ratio	95% confidence interval	p-value
Radial shortening >22%	26.7	3.4 – 209.9	p<0.0001
Ulnar shortening >20%	66.7	5.8 - 770	p<0.0001
Radial bowing >11.1%	66.7	5.8 - 770	p<0.0001
Radial bowing >8.1%	16.5	1.7 – 158.2	p=0.003
Radial articular angle >35°	16.5	1.7 – 158.2	P=0.003

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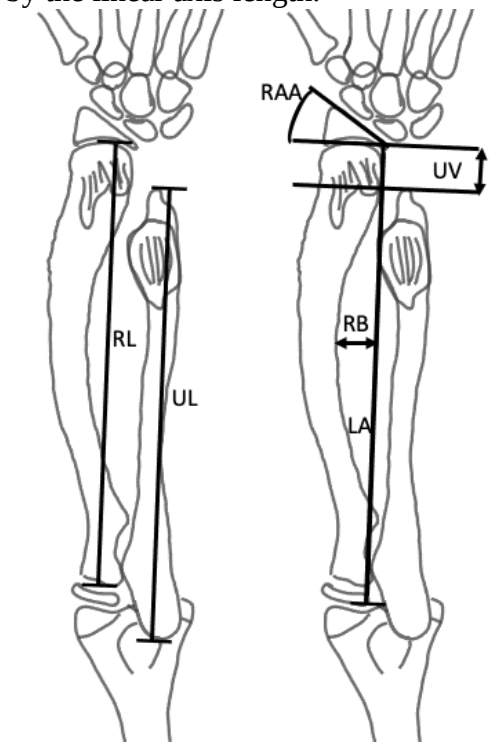
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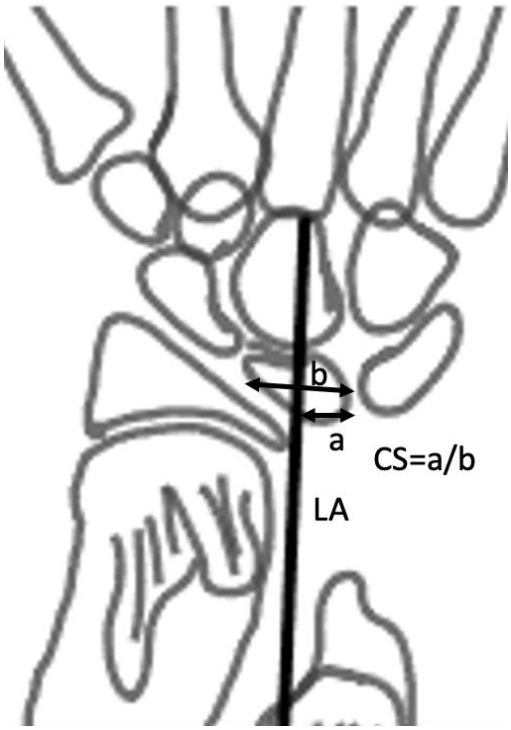


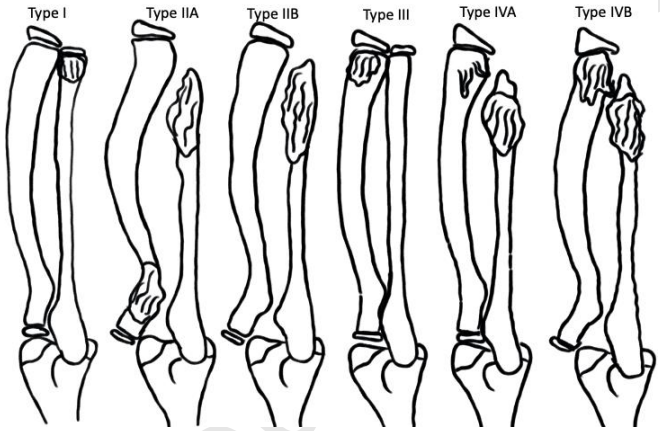
-Figure 1: Study flowchart.

-Figure 2: The linear axis (LA) of the forearm is a line drawn from the ulnar borders of the distal and proximal epiphysis of the radius. 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 from the tip of the olecranon to the ulnar styloid. Ulnar variance (UV) is the distance between 2 horizontal lines perpendicular to the linear axis of the forearm: the first at the level of the growth plate of the radius and the second at the level of the ulnar head. Radial bowing (RB) (%) is calculated as the maximal distance that the radius deviates from the LA, divided by the linear axis length.



-Figure 3: Carpal slip (CS) was measured as the percentage of the lunate situated ulnarly to a projection of the LA (CS=a/b).





-Figure 4: Classification of forearms according to Jo et al.

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