SCIENTIFIC ARTICLE

An Evaluation of Forearm Deformities in Hereditary Multiple Exostoses: Factors Associated With Radial Head Dislocation and Comprehensive Classification

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Purpose This study attempted to evaluate a series of patients with hereditary multiple exostoses (HME) who could not be categorized according to the widely accepted Masada classification and to identify radiographic variables such as radial bowing, ulnar shortening, ulnar variance, radial articular angle, and carpal slip predictive of deformity.

Methods We retrospectively reviewed data on 102 upper limbs of 53 pediatric patients with HME. Demographics, site of forearm involvement, and radiographic parameters were documented. Patients with exostoses of the forearms were categorized into 6 groups based on location of the exostoses and presence or absence of a dislocated radial head. Proportional ulnar shortening was calculated as the ratio of ulnar length to radial length.

Results According to the Masada classification, 4 limbs were normal, 10 were type I, 2 were type II, and 24 were type III. Sixty-six limbs were unclassifiable. We classified those 66 limbs using a modification of the Masada classification. Of the 106 limbs, 11 (10.3%) had a dislocated radial head. Based on the radiographic analysis, patients with proportional ulnar shortening of less than 0.9 had a higher risk of radial head dislocation than did those with proportional ulnar shortening of 0.9 or greater. Patients with radial bowing greater than 8.1% showed a higher frequency of radial head dislocation than did those with radial bowing of 8.1% or less. Exostoses of both the distal radius and ulna tended to increase the rate of radial head dislocation. A greater amount of negative ulnar variance caused more radial bowing and a greater radioarticular angle.

Conclusions We propose a new comprehensive forearm classification for patients with HME. Proportional ulnar shortening less than 0.9 and radial bowing 8.1% or greater can be used to predict the risk of radial head dislocation. *(J Hand Surg Am. 2017;■(■):1.e1-e8. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Type of study/level of evidence Prognostic IV.

Key words Forearm deformity, hereditary multiple exostosis, radial head dislocation, classification.

Online Exclusive

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Received for publication May 29, 2015; accepted in revised form January 10, 2017.

No benefits in any form have been received or will be received related directly or indirectly to the subject of this article.

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0363-5023/17/■ ■-0001$36.00/0
http://dx.doi.org/10.1016/j.jhsa.2017.01.010

Hereditary multiple exostoses (HME) is an autosomal dominant skeletal disorder that begins to be clinically apparent during childhood. The prevalence is estimated at 1 in 50,000 births and it occurs as a result of a *de novo* mutation in 20% to 30% of patients.[1,2] This disorder can cause a multitude of problems in the upper extremities. Altered growth results in shortening of the involved bones, angular deformity, decreased range of motion, and functional impairment, and it may lead to pain as a result of local muscle, tendon, or nerve irritation.

The focus of the current study was on lesions involving the forearm. The prevalence of these deformities among patients with HME is as high as 40% to 74%[3–6]; they include relative shortening of the ulna; bowing of the radius; ulnar deviation of the distal aspect of the radius, wrist, and hand; and dislocation or subluxation of the radial head.[3–6] Deformity may progress to carpal and elbow instability.[3,5,6] A dislocated radial head is associated with reduced forearm rotation and functional impairment.[7] Peterson[8] suggested that preventing and mitigating progression of deformity, particularly radial head dislocation, is the most important goal of early treatment. Understanding the natural history and risk factors associated with impaired forearm rotation and radial head dislocation would identify at-risk patients who may benefit from early surgery. However, the natural history of the condition is ill-defined; as a result, appropriate timing of forearm surgical treatment remains controversial.[4,6–9]

Masada et al[10] classified forearm deformities into 3 groups according to the location of the tumor and the morphology of the deformity. Type I involves osteochondromata of the distal ulna. Type II is characterized by dislocation of the radial head, either with osteochondromata of the proximal radius (type IIA) or resulting from more distal involvement. Type III includes cases with osteochondromata of the distal radius. However, it is difficult to categorize many cases using the Masada classification. The purposes of this study were to modify the Masada scheme to allow a more comprehensive HME classification, to identify clinical predictors of radial head dislocation, and to evaluate associations of the classifications with radiographic parameters.

MATERIALS AND METHODS

After we received institutional review board approval, we retrospectively reviewed the charts of all patients diagnosed with HME at our institution from January 2004 to December 2014. Patients with anteroposterior and lateral forearm radiographs were included. Patients in whom forearm involvement was not documented on radiographs were excluded, as were patients who had joint degeneration on radiographs. We recorded patient demographic data, including age at the time of radiographic evaluation, sex, location of the exostoses, and the presence or absence of radial head dislocation. This was a cross-sectional study in which all data were collected at a single time point.

The time point used for data collection was either when the patient reached skeletal maturity or when radiographs were taken at the last visit. If a patient underwent surgery to treat symptomatic forearm masses, we evaluated the radiographs taken just before surgery. Radiographs were used to determine the location of all forearm exostoses and the presence of radial head dislocation.

Patients were classified into 4 categories based on the location of the forearm exostoses on radiographs taken at the time of the patient's presurgical visit.

Our categorization is based on the Masada system, with modifications. Type I, II, and III deformities were classified as per the Masada system, and we added type IV, unclassifiable in the Masada system and defined as exostoses arising from both the distal radius and the ulna. In addition, we subdivided type IV into 2 groups depending on the existence of radial head dislocation: the radial head is in the correct anatomic location in type IVA patients, whereas it is dislocated in type IVB patients.

Forearm deformities were evaluated based on radiographic measurements, as described by Burgess and Cates.[11] The length of the bone varies widely in skeletally immature children but the size of the 2 bones in the forearm is strongly correlated; therefore, rather than use absolute measures of forearm length, we measured the proportional length of the radius relative to the ulna by calculating the ratio between the 2. We assessed the following radiographic parameters for each subject using anteroposterior and lateral forearm radiographs (Fig. 1).

Proportional ulnar length

The length of the ulna relative to the length of the radius was determined. Ulnar length and radial length (from the center of the proximal physis to the center of the distal physis) were measured (Fig. 1A) Proportional ulnar length (PUL) = ulnar length (cm) / radial length (cm).

Ulnar variance

Ulnar variance UV (in millimeters) was determined by drawing a perpendicular line from the long axis of the distal radial articular surface to the most distal ossified portion of the ulna. A negative UV value indicates a