EDITOR'S CHOICE

Ulnar Distraction Osteogenesis in the Treatment of Forearm Deformities in Children With Multiple Hereditary Exostoses

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Purpose To report on the outcomes of using ulnar lengthening combined with acute angular correction for the treatment of forearm deformities in patients affected by multiple hereditary exostoses (MHE). Our hypothesis was that this procedure would improve both radiographic measurements and clinical outcomes with minimal complications.

Methods A retrospective chart review was performed on patients who had a diagnosis of MHE and had undergone ulnar lengthening via a uniplanar external fixator over a 12-year period. Clinical outcomes such as range of motion, pain, and surgical complications were assessed. Radiographic changes were measured using interval radiographs.

Results The series included 17 patients. Median age at surgery was 7 years (range, 3−14 years). Median follow up was 55 months (range, 5−125 months). Improvements occurred in radial and ulnar radii of curvature, carpal slip, ulnar variance, and carrying angle at the elbow. There was 1 major pin track infection. There were 2 failures of the external fixator requiring exchange. Premature consolidation occurred in 1 case. Elbow, forearm, and wrist motion was not affected. Radiocapitellar joint congruency did not change. No patient reported pain at final follow-up.

Conclusions Our approach of using distraction osteogenesis of the ulna with angular correction in the radius and ulna as needed is able to correct carpal slip as well as to improve forearm bowing and elbow carrying angle. All of the patients maintained congruency of the radiocapitellar joint with no postoperative dislocations. Because of the low complication rate, the resolution of pain in patients who presented with pain, and the improvement of forearm bowing, this approach should be considered as a treatment option for children with MHE who are at risk for radiocapitellar dislocation. *(J Hand Surg Am. 2016;41(9):888−895.*

Type of study/level of evidence Therapeutic IV.

Key words Multiple hereditary exostoses, forearm deformity, distraction osteogenesis, ulnar lengthening, multiple osteochondromas.

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MULTIPLE HEREDITARY EXOSTOSES (MHE) is an autosomal dominant genetic disorder of enchondral growth with a prevalence of 1 in 50,000.[1] The disease results from mutations in the tumor suppressor genes EXT1 and EXT2.[2−5] Patients typically present in the first decade of life, with a median presenting age of 3 years.[1] Forearm deformities are found in 40% to 70% of patients with MHE.[1,6] The typical forearm deformity is cubitus varus, often the result of shortening of the ulna with a

compensatory increased radial bow and dislocation of the radial head. A number of factors have been identified as being important contributors to the amount of ulnar shortening. These include the total load of osteochondromas, gender, and lesions of a sessile type.[4,7–9] However, a study that compared loss of longitudinal growth with volume of tumor found no significant association, suggesting that osteochondromas are not "stealing" growth from the physis as previously postulated.[10]

Forearm dysfunction in MHE may be caused by shortening of the ulna, increased radial bowing, impingement of an osteochondroma on the interosseous membrane, radial head subluxation/dislocation, and abnormalities in the distal radioulnar joint (DRUJ).[11] Radial head dislocation has been associated with worse outcomes and higher pain scores.[12–14] However, the natural history of a radial head dislocation is not known. There is evidence to suggest that adults with marked forearm deformity can function well[11,15,16]; however, evidence also points to long-term disability in adults with forearm deformity.[17] With such scant and contradictory evidence, the treatment of forearm deformity in MHE remains controversial.

Variable results have been reported with the use of ulnar lengthening to treat a radial head that is already dislocated.[14,18] Prevention of radial head dislocation may be a better option than relocation, but the success of this strategy has not been established. Because tethering of the radius in a growing forearm can progress to a radiocapitellar dislocation, we consider any new onset of subluxation or any worsening of radiocapitellar subluxation grade in a growing forearm to be indicative of an impending dislocation. Our approach has been to treat forearm deformity in children with MHE when they have any of the following findings: painful range of motion of the forearm, a positive carpal slip (a measure of ulnar translation of the carpus), and/or an impending radiocapitellar dislocation. The purpose of this study was to assess the outcomes of our approach to correct forearm deformity in MHE, in which a gradual ulnar lengthening is combined with osteochondroma excision and corrective osteotomies as needed.

MATERIALS AND METHODS

After internal review board approval, a retrospective chart review was conducted to identify MHE patients treated with an ulnar osteotomy and external fixator placement. This was done by searching appropriate Current Procedural Terminology (CPT) and *International Classification of Diseases—Ninth Revision* (ICD-9) codes over the time period between 2002 and 2013. Nineteen patients were identified; only 17 patients were included owing to incomplete records. Basic demographic data and the details of surgery on all patients were collected.

Range of motion was obtained from the chart and was measured by the treating physicians (S.H.K. and D.A.Z.). Postoperative range of motion was the final value recorded in the chart. The type of range of motion recorded was forearm pronation and supination, elbow flexion and extension, and wrist flexion and extension.

Two fellowship-trained hand surgeons (S.H.K. and D.A.Z.) who specialize in pediatric cases treated all of the patients. All patients who had at least 1 of the indications previously stated were offered surgery. Surgery included excision of all or part of the ulnar osteochondroma, creation of an osteotomy in the middle one-third of the ulna, and application of a uniplanar external fixator (Minirail; Orthofix Ltd, Maidenhead, United Kingdom). Occasionally, a closing wedge osteotomy of the ulna and/or radius was used for acute correction of the cubitus varus. The surgeons made a subjective assessment of the amount of radial bowing to decide on the need for a corrective osteotomy. If they felt that the radial bow would not remodel and would limit motion, a radial closing wedge osteotomy was also performed. After surgery, lengthening was begun 7 to 10 days after application of the external fixator. It was continued until either premature consolidation or achievement of an ulnar-neutral to slightly ulnar-positive wrist. Lengthening was stopped short of neutral in cases in which distraction began to occur through the radiocarpal joint. Lengthening was performed at a prescribed rate of 1 mm/d, although the actual rate may have varied because it depended on patient compliance. The timing of removal of the external fixator depended on consolidation of the lengthened bone, as defined by the appearance of 3 cortices out of 4 on 2 orthogonal radiographic views, and took place in the operating room with the patient under general anesthesia.

The operative record was reviewed for details about the procedure. The postoperative records were reviewed for complications. We looked specifically for premature consolidation, nonunion, and pin-site complications, which were graded as minor or major based on whether or not they were managed as an outpatient or an inpatient.

The start and termination of lengthening were determined from the chart. Length of callotasis was measured on the last day of lengthening. The total length was recorded, as was the number of days of lengthening. The rate of lengthening was calculated from these 2 values. Whether or not the patient had pain