Hemiepiphyseal Stapling for Ankle Valgus in Multiple Hereditary Exostoses

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Abstract

If left uncorrected, valgus ankle deformity in multiple hereditary exostoses can cause significant disability in skeletally immature children and in adults. Various management methods have been described, including hemiepiphyseal stapling, transphyseal screw placement, fibular-Achilles tenodesis, distal tibial osteotomy, and ablative epiphyseodesis.

In this article, we report the cases of 3 skeletally immature children who had undergone hemiepiphyseal stapling of the medial distal tibial epiphysis for correction of valgus ankle deformity in multiple hereditary exostoses. Correction of the tibiotalar axis, in relation to chronological and bone age, was evaluated.

Hemiepiphyseal stapling of the medial distal tibial epiphysis provides ipsilateral corrective potential while allowing staple removal for reversal of growth retardation. This procedure is useful in the management of ankle valgus in multiple hereditary exostoses.

The bone tumors most common in children are osteochondromas,1,2 cartilage-capped bony prominences that extend from the edge of the physis and metaphyseal cortex of long bones. These lesions are normally solitary, but some patients have multiple osteochondromas in the condition known as multiple hereditary exostoses. This is an autosomal dominant disorder recently linked to a mutation in the EXT gene family.2,3 Multiple hereditary exostoses are typically discovered in the first decade of life and do not have a sex predilection.1,4 The multiple osteochondromas can lead to deformities in many areas of the body. Approximately 50% of people develop a valgus deformity of the ankle.5,6

In the skeletally immature child, a valgus deformity of the ankle is progressive. As the child grows, the distal tibial epiphysis may develop wedging, and the fibula may shorten, forcing the ankle into valgus.4,5 This is thought to be secondary to a discrepancy in the pressures on the physis.7 Growth is slowed on the lateral side, with higher pressures on the fibular physis and the lateral tibial physis, while lower pressures on the medial tibial physis accelerate growth.7 The valgus alignment causes abnormal loading of the joint itself and may result in compensatory problems with nearby joints of the foot or the knee. If severe enough, the deformity can cause problems with shoe wear, bracing, or skin breakdown and may lead to gait abnormalities. As the child becomes an adult, uncorrected deformities at the ankle lead to degenerative changes and pain.6

Valgus ankle deformity may be a primary condition, as in multiple hereditary exostoses, or may represent compensation for deformities elsewhere in the lower extremity. It has been described in various other conditions, including neurogenic, congenital, and chromosomal abnormalities.1-17 Many surgical techniques have been described to correct the deformity, including hemiepiphyseal stapling, transphyseal screw placement, fibular-Achilles tenodesis, distal tibial osteotomy, placement of Ilizarov frame, and ablative epiphyseodesis.3,8-12,15-17

In this article, we report the cases of 3 patients who had valgus ankle deformity in the setting of multiple hereditary exostoses and describe the outcomes of hemiepiphyseal stapling.

Figure 1. Case 3—preoperative (A) and postoperative (B) standing radiographs of right ankle.

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Materials and Methods

Institutional review board (IRB) approval was obtained to conduct an International Classification of Diseases, Ninth Revision code search for hospital patients with multiple hereditary exostoses. A waiver of informed consent was granted by the IRB committee. Included in the search were cases from private orthopedic offices and from the orthopedic clinic at the hospital. Three patients were identified as having had multiple hereditary exostoses managed with hemiepiphyseal stapling of the medial distal tibial epiphysis. One other patient had had multiple hereditary exostoses and ankle valgus but had been treated with transphyseal screw placement.

All 3 patients underwent medial distal tibial hemiepiphyseal stapling of both ankles after serial radiographic examinations had shown progression of valgus ankle deformity. Timing of surgery was based on clinical judgment involving a combination of chronologic age, bone age, severity of symptoms, and degree of deformity.

Surgery was performed by exposing the medial distal tibial physis, taking care to protect neurovascular structures during the exposure. Keith needles were placed in the physis to display the orientation of the physis; their location was confirmed by fluoroscan. Three Blount staples were then placed around the physis with the middle staple centered in the sagittal plane. Placement was then checked on multiple views using fluoroscan to ensure the staples were not violating the articular surface or the physis. No patient had pain at the site of the osteochondroma, so the osteochondroma was not removed in any of the cases.

The tibiotalar axis was radiographically evaluated with true standing weight-bearing mortise views of the ankle and was measured as the angle formed by a line across the talar dome and a line from the middle of the distal third of the tibia to the midpoint of the mortise. All patients were carefully positioned to avoid internal or external rotation of the hip and were radiographically evaluated by the authors and under our direction. The patients provided written informed consent for print and electronic publication of their respective case reports.

Case Reports

Case 1
A girl had ankle valgus secondary to multiple hereditary exostoses between the tibia and the fibula, first noted at age 6 years and 7 months. She underwent bilateral hemiepiphyseal stapling at age 10 years and 4 months (bone age 10 years and 6 months). At that time, the tibiotalar angles were 22° (right) and 16° (left). The staples in the right ankle were not removed. The staples in the left ankle were removed 8 months after surgery, at bone age 11 years, with the tibiotalar angle corrected to 0°. Unfortunately, the osteochondroma continued growing, and the left ankle deformity recurred. Radiographs at the time of skeletal maturity (bone age >13 years and 6 months) showed a right tibiotalar angle of 10° and a left tibiotalar angle of 12°. At skeletal maturity, clinical examination revealed improved ankle stability (Table).

Case 2
A boy had ankle valgus secondary to multiple hereditary exostoses between the tibia and the fibula, first noted at age 11 years and 3 months. He underwent bilateral hemiepiphyseal stapling at age 13 years and 2 months (bone age 14 years). At that time, both ankles had a tibiotalar angle of 21°. Radiographs at the time of skeletal
maturity (bone age >15 years and 6 months) showed a right tibiotalar angle of 10° in both ankles. At skeletal maturity, clinical examination revealed improved ankle stability (Table).

Case 3
A boy had ankle valgus secondary to multiple hereditary exostoses between the tibia and the fibula, first noted at age 8 years and 11 months. He underwent right ankle hemiepiphyseal stapling at age 12 years and 6 months (bone age 12 years) when radiographs showed a right tibiotalar angle of 13° (Figures 1A, 1B). He underwent left ankle stapling at age 13 years (bone age 12 years 6 months) when radiographs showed a left tibiotalar angle of 11° (Figures 2A, 2B). At physeal closure, at age 15 years, radiographs showed a right tibiotalar angle of 4° and a left tibiotalar angle of 3° (Figures 3, 4). At skeletal maturity, clinical examination revealed improved ankle stability (Table).

DISCUSSION
To our knowledge, this article is the first to specifically report the outcomes of using only hemiepiphyseal stapling to manage ankle valgus in a uniform population of patients, those with multiple hereditary exostoses. Hemiepiphyseal stapling has long been used to retard growth at the physis in multiple hereditary exostoses. Use of staples at the medial distal tibial physis was described as early as the 1960s, but only in 1989 did Snearley and Peterson report using staples in multiple hereditary exostoses. Since 1989, other surgical approaches, including corrective osteotomies, screw epiphysiodesis, placement of Ilizarov frame, and ablative epiphysiodesis have been advocated for correction of valgus ankle deformities in this population.

The disadvantages of hemiepiphyseal stapling include its invasiveness and the difficulty in timing surgery. Difficulty in timing was evident in our cases, particularly case 1, when premature correction prompted us to remove the staples before physeal closure, which ultimately led to recurrence of the deformity.

Compared with other surgical options, however, hemiepiphyseal stapling has many advantages. It is less invasive, controls many planes, does not necessarily require hardware removal, does not require casting, and allows patients to walk as soon as wounds are healed. It is also reversible; for example, in case 1, growth continued after staple removal. In contrast to hemiepiphyseal stapling, corrective osteotomy is technically demanding, requires immobilization, creates stress shielding, and likely requires hardware removal. In addition, the osteotomy is across the exostoses, which can put the anterior compartment neurovascular bundle at risk from stretching or intraoperative injury. Ablative epiphysiodesis is somewhat more invasive than hemiepiphyseal stapling, but its main disadvantage is that it is irreversible. Many would argue that screw epiphysiodesis offers similar advantages to hemiepiphyseal stapling in that it is also reversible, but this procedure requires attention to detail in screw placement, penetrates the physis, and does not control growth in more than a single plane, which potentially leads to unintended deformity in other planes.

One newer hemiepiphyseodesis technique that could be a possible alternative to staples in this population is the 8-Plate Guided Growth System (Orthofix, Verona, Italy), which uses a nonlocking extraperiosteal plate and screws to guide growth. The efficacy of this system has not been demonstrated in any ankle valgus studies yet, but results are promising in early studies involving other lower extremity deformities in children with open physes.

We have described only a small number of patients, but these cases should remind us of the advantages and disadvantages of hemiepiphyseal stapling as part of the armamentarium for managing ankle valgus, particularly in the setting of multiple hereditary exostoses. In all 3 of our patients, treated ankles showed radiographic improvement in the tibiotalar axis. These radiographic changes were consistent with increased ankle stability and other improvements noted during clinical examination.

CONCLUSION
Valgus deformity of the ankle is a common problem in children with multiple hereditary exostoses. In these children, the differential growth between the medial and lateral aspects of the ankle causes the deformity. However, skeletally immature children still have excellent

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Table. Duration and Amount of Correction of Tibiotalar Axis

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<tr>
<th>Case</th>
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*Staples were removed after improvement to 0°; deformity recurred after removal.
growth potential in all areas of the physis. Many severe deformities must be surgically corrected to improve ankle function and avoid degenerative problems in adulthood. Hemiepiphyseal stapling is a surgical option that should be considered in this setting, as the technique takes advantage of a child’s growth potential to correct the deformity and is reversible.

**AUTHORS’ DISCLOSURE STATEMENT**
The authors report no actual or potential conflict of interest in relation to this article.

**REFERENCES**