To the authors’ knowledge, few reports have been published in the English literature of using total knee arthroplasty and total hip arthroplasty for the treatment of hereditary multiple exostoses. This article describes 2 patients with hereditary multiple exostoses, 1 treated with total hip arthroplasty and 1 treated with total knee arthroplasty. Bony deformities make arthroplasty uniquely challenging in patients undergoing total hip or knee arthroplasty. An expanded metaphysis of the proximal femur, coxa valga deformity, and the presence of hardware from previous reconstructive surgeries can make total hip arthroplasty technically difficult. Substantial bony deformity of the distal femur, valgus deformity of the knee, and sizing issues that necessitate custom implants can make total knee arthroplasty difficult. The most common bony deformities in hereditary multiple exostoses are short stature, limb-length discrepancy, valgus deformity at the knee and ankle, and asymmetry of the pectoral and pelvic girdles. Most reported surgical treatments for patients with hereditary multiple exostoses focus on the pediatric population or the management of malignant transformation of exostoses. Studies that specifically address the conditions associated with knee deformities focus on extra-articular deformity correction rather than arthroplasty. When arthroplasty is necessary in this patient population, an understanding of the commonly occurring deformities can help with preoperative planning and surgical management. All painful lesions must be evaluated for malignant transformation. Bone scans can be useful during workup. All specimens should be sent for pathologic evaluation. Such patients are challenging because of the distorted hip anatomy and valgus knee deformity. The current 2 cases illustrate specific challenges that can be anticipated and underscore key principles for arthroplasty in the management of hereditary multiple exostoses.

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Hereditary multiple exostoses is an autosomal-dominant condition with full penetrance that results in multiple osteochondromas. These benign bone tumors are found at the metaphyses of rapidly growing long bones. The prevalence of hereditary multiple exostoses is 0.9 to 1.4 per 100,000 in Caucasians and 100 to 1310 per 100,000 in the Chamorros of Guam and the Ojibway Indians of Canada.

The malignant transformation of osteochondromas to low-grade chondrosarcoma is a well-recognized feature of hereditary multiple exostoses. The prevalence of chondrosarcoma transformation is estimated to be 0.5% to 25%. A cartilage cap greater than 2 cm is associated with an increased risk of malignancy. Hereditary multiple exostoses can cause deformities in the upper and lower extremities, as well as short stature. Because osteochondromas can develop adjacent to faster-growing growth plates, such as those around the knee, normal bone growth is disrupted, and angular deformities occur at the joints. Loss of range of motion eventually occurs. The mesomelic or distal segments of the limb (ie, forearm and leg) are preferentially affected by the exostoses, further contributing to the angular deformities of the involved joints. Cosmesis is also a common concern.

In the lower extremities, the proximal femur, knee, and ankle are involved. The metaphyseal bone region typically increases in size because of osteochondroma formation adjacent to the growth plate. Distal femur and proximal tibial involvement can contribute to valgus knee deformity. The shortened fibula often seen in hereditary multiple exostoses can also instigate valgus knee joint angulation. The proximal femur is characterized by coxa valga and acetabular dysplasia.

In the surgical management of hereditary multiple exostoses, preoperative planning with special attention paid to unique bony deformities is crucial. This article describes 2 clinical cases to highlight key technical considerations when using arthroplasty to treat this condition.

**Case Reports**

**Patient 1**

A 52-year-old man with hereditary multiple exostoses presented with left knee pain. His medical history included a chondrosarcoma of the diaphyseal region of the right femur treated via intercalary resection and reconstruction with allograft and exostoses removal from the left proximal femur. At physical examination, left knee range of motion was 15° to 95°. The knee ligaments had good stability to varus and valgus testing. Knee radiographs showed severe tricompartmental arthritis with valgus deformity (Figure 1A). The patient underwent total knee arthroplasty (TKA).

The patient initially underwent a left cemented posterior-stabilized TKA (LPS; Zimmer, Warsaw, Indiana). He was fitted with the largest off-the-shelf femoral component available. Intraoperatively, several exostoses interfered with the placement of the cutting blocks and were removed. To balance the valgus deformity, the posterolateral capsule and popliteus tendon were released and the iliotibial band was piecrusted.

His postoperative hospital course was uneventful, and he was discharged to a subacute rehabilitation facility. There, fecal impaction precipitated the development of a painful rectal fissure, which limited his participation in physical therapy. At the 6-week follow-up visit, knee range of motion was 5° to 80°. Manipulation under anesthesia was required, and 95° of flexion was obtained. At the 39-month follow-up visit, his left knee was pain free. Radiographs showed excellent alignment of all components (Figure 1B).

**Patient 2**

A 61-year-old man with hereditary multiple exostoses presented with severe right hip pain. On physical examination, he achieved 85° of hip flexion, 45° of external rotation, and 0° of internal rotation. Hip radiographs showed severe joint-space narrowing and coxa valga (Figure 2A). Pelvic radiographs revealed a large, irregularly shaped osteochondroma in the left proximal femur. A triple-phase bone scan ensured that the lesion was not active. No suggestion of metabolic activity was detected.

He subsequently underwent a right total hip arthroplasty (THA). The capsule was densely adherent to the underlying bone. An extensive metaphyseal flare existed. The acetabular portion of the procedure was routine. On the femoral side, because of the widened metaphysis, a fully coated stem (VerSys Beaded Fullcoat; Zimmer) that engaged the diaphysis was used. Radiographs at the 27-month follow-up showed that the THA components were well aligned (Figure 2B). His pain had significantly improved.

**Discussion**

The pathophysiology of hereditary multiple exostoses is attributed to mutations in the exostosin (EXT1, EXT2, and EXT3 genes). Exostosin 1 and EXT2 code for transmembrane glycoproteins that help...
regulate cell surface heparin sulfate proteoglycans, which are key components of cartilage. Exostin 1 is found on chromosome 8, and EXT2 is found on chromosome 11. Exostin gene mutations can lead to a disruption of the negative feedback central to normal chondrocyte differentiation. This disruption results in abnormal skeletal development. In 80% of patients, hereditary multiple exostoses is diagnosed in the first decade of life. Coxa valga and increased femoral anteversion can be found in the proximal femur. In the upper extremity, ulnar shortening, subluxation or dislocation of the radial head, disruption of the distal radioulnar joint, ulnar deviation, and ulnar translocation of the carpal bones are seen.

Surgical treatment for hereditary multiple exostoses can involve removal of the exostoses, correction of the various joint deformities, and correction of limb-length discrepancies; in growing children, epiphysiodesis can be used for the management of limb-length discrepancies. Complete removal of symptomatic exostoses is advocated to avoid recurrence and malignancy.

To the current authors’ knowledge, 5 cases of TKA and 5 cases of THA in patients with hereditary multiple exostoses have been reported. The most common bony deformities in hereditary multiple exostoses are short stature, limb-length discrepancy, valgus deformity at the knee, and ankle, and asymmetry of the pectoral and pelvic girdles. Most reported surgical treatments for patients with hereditary multiple exostoses focus on the pediatric population or the management of malignant transformation of exostoses. Studies that specifically address the condition’s associated knee deformities focus on extra-articular deformity correction rather than arthroplasty.

When arthroplasty is necessary in this patient population, an understanding of the commonly occurring deformities can help with preoperative planning and surgical management. All painful lesions must be evaluated for malignant transformation. Bone scans can be useful during this workup. All specimens should be sent for pathologic evaluation.

### Hip Deformities

The proximal femur is involved in 90% of patients with hereditary multiple exostoses. Shapiro et al. reported that 8 of 32 patients (64 hips) younger than 20 years with this disease had coxa valga, as did 1 of the current 2 patients. Increased femoral anteversion has also been reported. Acetabular dysplasia can result from large osteochondromas on the medial femoral head, resulting in uncovering of the femoral head. A case was reported of a pediatric patient who required proximal femoral osteotomy and Steele pelvic osteotomy to address hereditary multiple exostoses–induced acetabular dysplasia.

### Knee Deformities

In the knee, exostoses of the distal femur or proximal tibia can lead to valgus deformity. The distal femur and proximal tibia are involved in 70% to 98% of patients with hereditary multiple exostoses. Valgus deformity is also often the result of a shortened fibula that results in valgus angulation of the tibia. Opening-wedge osteotomy and blade plate fixation of the distal femur have been used to correct valgus deformities of the femur. In the proximal tibia, hemiepiphyseodesis or high tibial osteotomies have been used for valgus deformity correction. The valgus deformity seen in the tibia has been attributed to shortening of the fibula secondary to exostoses. Arthroscopic removal of symptomatic intra-articular osteochondromas of the knee has also been reported. The case report described...
an osteochondroma in the anterolateral aspect of the distal femur in the patellofemoral joint.19 A few studies have reported the role of navigation-assisted TKA in the presence of tibial or femoral deformity.20-22 Although these studies do not represent large series, their results indicate that navigation provides a high accuracy of alignment and can prevent implant misalignment. Navigation may be a useful option for surgeons performing arthroplasty for deformities from hereditary multiple exostoses.20-22

**Total Knee Arthroplasty**

To the current authors’ knowledge, TKA for patients with hereditary multiple exostoses was recently described for the first time.8 For the preoperative planning of patients with hereditary multiple exostoses, it is critical to template appropriately and to plan for a custom implant or ensure that various implant sizes are in stock. The current authors did not use a computer-based program to template the radiographs, but such a program may be a useful adjunct for preoperative planning.23 Despite the current patient’s short stature, the largest femoral implant was necessary. Because valgus deformity is commonly found in such patients, the surgical plan should be tailored to this possibility. The current patient required extensive soft tissue releases to balance his ligaments. The authors also aggressively resected the patient’s exostoses. In the series reported by Kim et al,8 all 5 cases were in valgus (range, 11°-45°), and the authors used lateral epicondyile osteotomies to correct the severe valgus deformities. Of the 5 knees, 2 had a peroneal nerve palsy postoperatively.8 Because a 0.5% to 25% risk exists of chondroosarcoma transformation in hereditary multiple exostoses, it is critical to send the resected exostoses to pathology to determine whether a malignancy exists.1 In the current patient, the pathology specimen was negative for malignancy. Postoperatively, no signs of peroneal nerve palsy were noted. At least follow-up 3 years postoperatively, he was independent with ambulation and all activities of daily living, had a pain-free knee, and was fully compliant with his outpatient physical therapy regimen.

**CONCLUSION**

Experience with arthroplasty in patients with hereditary multiple exostoses is limited. These patients are challenging secondary to the distorted anatomy of the hip and valgus deformity of the knee. Total knee and hip arthroplasty can be performed successfully despite the technical difficulties. Surgeries in patients with hereditary multiple exostoses should be performed at large-volume arthroplasty centers with surgeons who are familiar with techniques of performing arthroplasty in patients with deformities. Future studies are needed to address long-term follow-up and anatomic, clinical, and functional outcomes of arthroplasty in a larger cohort of patients with hereditary multiple exostoses.

**REFERENCES**