Bilateral Uncemented Ceramic-on-Ceramic Total Hip Arthroplasty in a 26-Year-Old Man With Morquio Syndrome

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Morquio syndrome, also known as type IV mucopolysaccharidosis, is an autosomal recessive disorder caused by deficiency of *N*-acetyl-galactosamine-6-sulfate. Morquio,[1] in Uruguay, and Brailsford,[2] in England, simultaneously described this syndrome in 1929. Its incidence is unknown but is estimated to be between 1 in 75,000 (Northern Ireland) and 1 in 200,000 (British Columbia, Canada).[3,4] Morquio syndrome is characterized by a defect in degradation of keratan sulfate and chondroitin-6-sulfate resulting in accumulation of mucopolysaccharides, which interfere with cell function.

A child with Morquio syndrome may appear normal at birth but usually begins showing manifestations of the disease between ages 2 years and 4 years. Major orthopedic problems result from a unique spondyloepiphyseal dysplasia and ligament laxity. There is severe growth retardation with short-trunked dwarfism, genu valgum, lumbar and thoracic kyphosis, abnormal hip development, and malalignment of the lower limbs.[5] The pelvis typically shows both acetabular and femoral head dysplasia with varying degrees of hip subluxation, short and wide femoral necks with valgus deformity, and a wide symphysis pubis. Odontoid hypoplasia may lead to atlantoaxial instability, which can give rise to myelopathy, quadriplegia, and even sudden death.[6]

Other features are aortic valve incompetence, aortic stenosis, hepatomegaly, splenomegaly, inguinal hernia, mixed hearing loss, and ocular complications, particularly clouding of the corneas. Pulmonary complications include a restrictive defect, caused by kyphoscoliosis, which can result in pulmonary hypertension and cor pulmonale.

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Although hip dysplasia leading to degenerative arthritis is one of the most significant orthopedic manifestations in Morquio syndrome, this problem has not received much attention in the literature. In the present article, we report the case of an active 26-year-old man with Morquio syndrome and advanced degenerative hip arthritis managed with bilateral uncemented ceramic-on-ceramic total hip arthroplasty (THA), followed up for 5 years. In this first English-language report on use of bilateral uncemented THA in Morquio syndrome, we highlight the unique aspects of the case. The patient provided written informed consent for print and electronic publication of this case report.

Case Report

A 26-year-old man with Morquio syndrome presented to the outpatient clinic with an 18-month history of bilateral hip pain. The patient, a shopkeeper, weighed 63 kg (139 lb) and was 150 cm (4 ft, 11 in) tall. He described the pain as constant, 9/10 in severity, and said it woke him from sleep. It was more severe on the left side at time of presentation.

Clinical examination of the patient's hips revealed fixed flexion deformities of 10° with flexion to 80° and abduction and adduction of 10°. Rotation examination, which was painful, revealed almost no movement.

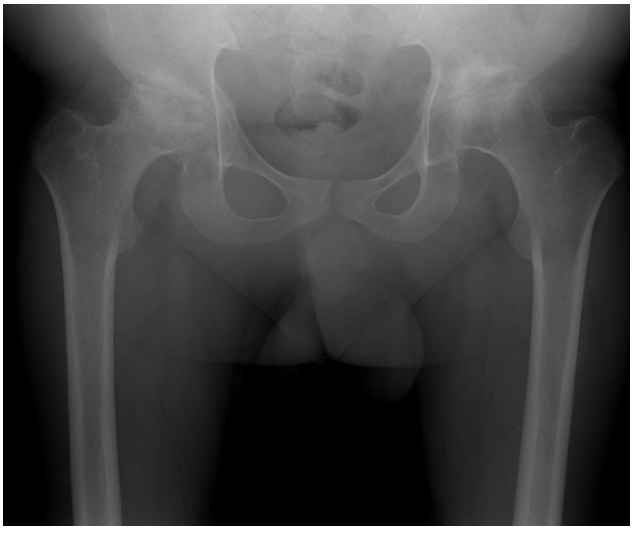

Figure 1. Preoperative anteroposterior radiograph shows advanced arthritic change in both hips, acetabular dysplasia, and small femora.

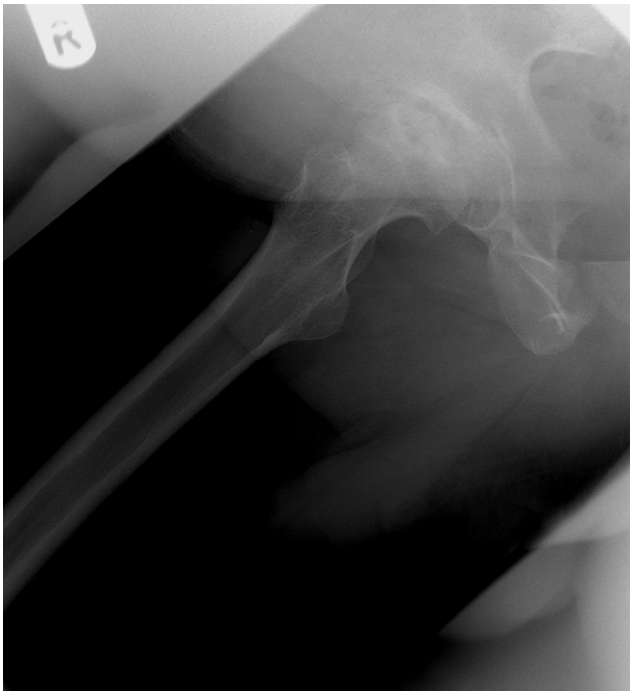

Figure 2. Iliac oblique radiograph of right acetabulum shows significant acetabular dysplasia.

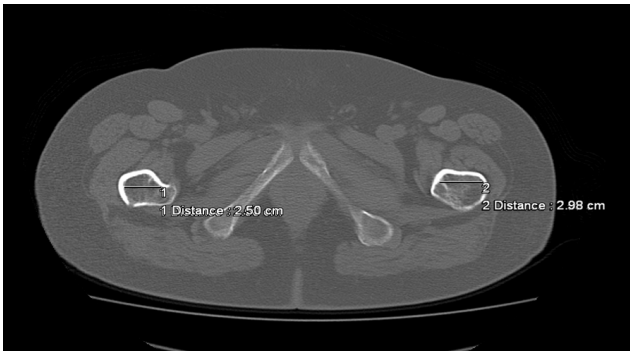

Figure 3. Axial computed tomography shows one of many measurements used to determine size of proximal femur and compatibility with available implants.

An anteroposterior radiograph showed bilateral hip arthritis with acetabular dysplasia (Figure 1). Judet radiographs confirmed advanced acetabular dysplasia, particularly on the right side (Figure 2). Full-length radiographs of the lower limbs were obtained to rule out bony deformities distal to the proximal femur. Given the patient's young age and activity level, staged bilateral uncemented THAs were planned, starting with the left, more symptomatic side. Computed tomography (CT) of the pelvis and the proximal femur was obtained, bony dimensions measured at multiple levels were found to be compatible with available "off-the-shelf" components (Figure 3), and a full preoperative medical assessment was performed.

Preoperative cervical spine radiographs showed odontoid hypoplasia, and flexion-extension views revealed an anterior atlantodental interval of 3 mm, the upper limit of normal. Magnetic resonance imaging (MRI) of the cervical spine showed an intermedullary syrinx extending from the second cervical vertebra down to the thoracic spine—a finding that ruled out regional spinal anesthesia. Neurologic examination was normal. After reviewing these results and consulting the anesthesia team and the patient, we decided to use awake fiberoptic intubation for both THAs.

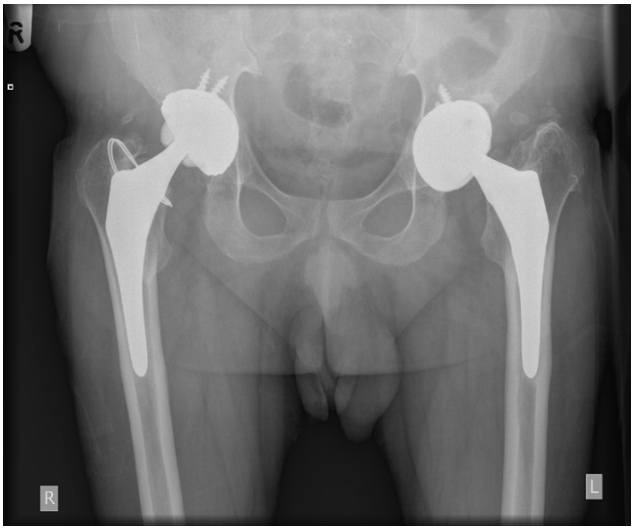

Figure 4. Bilateral total hip arthroplasties 5 years after surgery.

Left THA was performed through an anterolateral approach with the patient in the lateral decubitus position. In-line reaming of the acetabulum to 50 mm was followed by insertion of an uncemented hydroxyapatite-coated (HA-coated) acetabular shell plus 2 supplementary screws combined with a 32-mm ceramic liner (Trident PSL, Howmedica/Osteonics, Mahwah, New Jersey). On the femoral side, a proximally HA-coated stem, size 3 (ABG II, Howmedica), was combined with a 4-mm head offset 32-mm diameter ceramic femoral head (V40 Alumina, Howmedica/Osteonics). Recovery was uneventful, and the patient was admitted for right THA 7 months later. The same setup, approach, and component dimensions were used. During canal preparation, the procedure was complicated by a medial calcar fracture, which was managed with double cerclage wiring and restricted weight-bearing with crutches for 6 weeks.

At 5-year follow-up, the patient was back at work, reported no hip pain, and did not require any analgesia. Radiographs showed satisfactory component positioning and bony incorporation (Figure 4).

Discussion

Altered joint biomechanics and abnormal articular cartilage are thought to contribute to the onset and progression of degenerative arthritis in patients with skeletal dysplasia.[7,8]

Morquio syndrome, a rare mucopolysaccharide storage disease, results in a unique spondyloepiphy-

seal dysplasia, with hip dysplasia one of the well-recognized sequelae.[9] In the past, many patients with Morquio syndrome died during their young-adult years. Advancements in cardiopulmonary, neurologic, and anesthetic care, however, have dramatically increased the life expectancy of these patients, with many now surviving into their sixth decade.[10] As a result, total joint arthroplasty and prosthesis longevity are important considerations in this patient population.

In a comprehensive review of the current literature on bilateral hip arthroplasty in Morquio syndrome, only 2 case reports were found,[10,11] and only 1, by Lewis and Gibson,[11] was in English. They described the case of a 25-year-old woman, who with spinal anesthesia, underwent bilateral cemented metal-on-polyethylene THAs using Elite CDH femoral stems (DePuy, Warsaw, Indiana). Except for mentioning that radiographs of the cervical spine were obtained, the authors did not comment on the preoperative workup. Although they reported a satisfactory outcome at 7 years, they indicated the patient will need revision arthroplasty at some point.

Managing degenerative hip disease in Morquio syndrome requires extensive preoperative medical assessment and a preoperative plan. The orthopedic surgeon in consultation with the anesthesia service should pay particular attention to the cervical spine. A full cervical spine radiographic series, including flexion and extension views, should be obtained in all cases. We also recommend preoperative spinal MRI, as not only does it give more detailed information on the upper cervical spine, but it may also reveal other relevant anomalies, as in the case presented here. Use of spinal anesthesia in the presence of a syrinx may result in a deleterious increase in craniospinal pressure with the potential for enlargement of the defect and neurologic deterioration.[12]

A thorough preoperative plan is needed to address the articular and periarticular deformities and small bone common in osteochondrodysplasia. Chiavetta and colleagues[13] reviewed 62 THAs performed for osteoarthritis secondary to osteochondrodysplasia. A mix of cemented, uncemented, and hybrid component fixation was used. In 6 cases, customized femoral prostheses were used to accommodate deformities. The authors reported high complication rates, including a 5% incidence of periprosthetic fracture and a 30% revision rate, at a mean follow-up of 11.5 years. Most revisions were performed in patients with cemented components. The authors hypothesized that their patients' young ages and activity levels contributed to these high revision rates.

With extensive preoperative radiologic evaluation of the hip joint and the proximal femur, the surgeon can address many complex technical issues. On the acetabular side, level of true acetabulum, acetabular version, and bone loss require evaluation. Multiple joint involvement, common in patients with osteochondrodysplasia, may alter the gait and place higher loads on the prosthetic joints.[13] Restoration of hip center to true acetabulum confers both anatomical and biomechanical advantages, whereas high hip center and cup lateralization have both been implicated in early failure.[14] Given these results, we believe every effort should be made to restore true center of rotation in these patients. Management of bone loss and version in hip dysplasia is beyond the scope of this case report. Challenges on the femoral side include small bone, altered femoral version, and increased diaphyseal bowing. The combination of full-length lower limb plain radiographs for assessment of femoral bowing and CT measurement of femoral version and bony diameters in the proximal femur should provide the information needed for accurate preoperative planning. Discrepancies among radiologic measurements, patient anatomy, and prosthesis size mandate a backup plan as well. In our patient's case, we made sure we had fracture fixation equipment and a range of small cemented implants readily available. However, we used uncemented components because our patient's anatomy allowed them and because hard-on-hard bearing surfaces provide the best chance of prolonged implant survival in active young men.

This is the only reported case of an active young man with Morquio syndrome and disabling degenerative hip disease managed with bilateral uncemented THAs. For such cases, we recommend a thorough preoperative workup with particular attention paid to the cervical spine and the bony deformity of the hip and proximal femur.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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