



Case report

Simultaneous bilateral total hip arthroplasty in Morquio syndrome

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ABSTRACT

A 16-year-old girl who had Morquio syndrome presented with severe bilateral hip pain and limited mobility because of bilateral hip osteoarthritis and fixed flexion deformities. She was wheelchair bound for the previous 6 months. Cervical spine flexion-extension views showed mild subluxation (<3 mm), and there was thoracolumbar spine kyphosis. Magnetic resonance imaging of the cervical and thoracolumbar spine showed hypoplasia of the odontoid and vertebral bodies, but no spinal cord compression. Bilateral cemented total hip arthroplasty was performed through a posterior approach under general anesthesia with fiberoptic intubation. The femoral canals accepted a small-diameter stem, the right femoral head was used as a graft for superior right acetabular deficiency, and low-profile all-polyethylene acetabular cups were implanted. Follow-up at 15 years after surgery showed that the patient was fully ambulatory without pain or supports, and radiographs showed no loosening. In summary, total hip arthroplasty at a young age may be necessary in patients who have Morquio syndrome because of severe arthritis and soft tissue contractures. Extensive preoperative evaluation that includes imaging of the entire spine is mandatory because of the risk of developing spinal cord compression.

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Introduction

Morquio syndrome is a type IV mucopolysaccharidosis, which is an autosomal recessive disorder, first described in 1929, characterized by the deposition of mucopolysaccharides in the cells of multiple organs [1]. This syndrome has several musculoskeletal manifestations, especially hip dysplasia with a shallow and dysplastic acetabulum, femoral head dysplasia and avascular necrosis, hip subluxation, and severe arthritis at an early age. In the severe form, the femoral head hinges around the superior wall of the acetabulum and becomes fixed, with no or limited motion. The femoral neck may be short, wide, and markedly anteverted. Soft tissue contractures around the hip joints cause severe fixed hip flexion deformity [2–5].

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Other skeletal manifestations of Morquio syndrome include short stature, genu valgum, and foot deformities. Cervical spine involvement with odontoid hypoplasia and subluxation may cause cervical cord compression, myelopathy, and paralysis, and this may be a major anesthetic risk because the spinal cord may be vulnerable to injury during endotracheal intubation. Thoracolumbar kyphosis may present with cord or root compression [6–8]. These patients also may have aortic valve stenosis or incompetence, ocular and hearing problems, hepatosplenomegaly, and restrictive pulmonary disease [9]. Unlike patients who have other mucopolysaccharidosis, Morquio syndrome patients have normal intellect [5].

In patients who have Morquio syndrome, total hip arthroplasty may be indicated at a young age because of severe bilateral hip involvement. However, literature search showed only 2 English-language case reports about hip arthroplasty and outcomes in young patients [10–12].

We treated an adolescent girl who had Morquio syndrome with simultaneous bilateral total hip arthroplasty for severe bilateral dysplastic hips and extensive soft tissue contractures, and who returned for long-term follow-up. The patient provided written informed consent for print and electronic publication of this case report and photographs.



Figure 1. Preoperative anteroposterior (AP) radiograph of the pelvis showing severe bilateral hip osteoarthritis.

Case history

A 16-year-old girl who had Morquio syndrome presented to us in 2001 because of a 2-year history of severe, progressively increasing pain in both hips. There had been progressive reduction in her mobility; she was able to stand and take a few steps with great difficulty, but otherwise she was completely wheelchair bound during the 6 months before presentation. Body weight was 45 kg and height was 145 cm. Examination showed cervical spine restricted extension and thoracolumbar spine kyphosis. There was fixed flexion deformity of both hips (40° , with further flexion to 90°), and there was no hip internal rotation, external rotation, abduction, or adduction. The knees had slight genu valgum, and the feet were normal. The upper and lower limbs had normal neurovascular examination.

Radiographs showed severe bilateral hip osteoarthritis (Fig. 1). As both hips were involved there was no preoperative leg length discrepancy. Cervical spine flexion-extension views showed mild subluxation (<3 mm), and there was thoracolumbar spine kyphosis. Magnetic resonance imaging of the cervical and thoracolumbar spine showed hypoplasia of the odontoid and vertebral bodies, but no spinal cord compression. Computed tomography scan of the pelvis confirmed hip dysplasia. The patient was evaluated by a cardiologist and anesthetist and cleared for surgery.

The patient had simultaneous bilateral total hip arthroplasty through a posterior approach under general anesthesia with fiberoptic endotracheal intubation. The femoral neck was osteotomized with reference to the lesser trochanter on both sides. Soft tissue releases included bilateral iliopsoas tendon release and adductor tenotomy, which were necessary to balance the hip and performed often in similar cases [13,14]. The acetabula were reamed and the femoral canals were prepared carefully. Bilateral cemented metal-on-polyethylene hip prostheses were used. The femoral canals accepted a small-diameter stem (DDH stem, size 5, Protek, Sulzer Medica, Winterthur, Switzerland), and an intraoperative right calcar fracture was treated with cerclage wires. The right femoral head was used as a graft for right superior acetabular deficiency, and a 40-mm right low-profile all-polyethylene cemented acetabular cup was implanted; the left acetabulum was fitted with a 36-mm cemented acetabular cup (Protek, Sulzer, Winterthur, Switzerland; Fig. 2). After the implants were placed, the lengths increased symmetrically on both sides with no postoperative leg length discrepancy. The patient was mobilized full



Figure 2. Postoperative AP radiograph of the pelvis after 15 years showing bilateral total hip arthroplasty. The right femoral head was used as a graft for right superior acetabular deficiency.

weight bearing from the first day. The intraoperative fracture was secured with a circle wire and the hip was fully cemented; therefore, no restriction to weight bearing was applied. Postoperative care included physiotherapy with hip precautions and venous thromboembolism prophylaxis (heparin, 5000 units twice daily) for 28 days along with pneumatic compression devices and early mobilization. She was discharged from the hospital on the fifth postoperative day after mobilizing to full weight-bearing.

The patient was able to continue unrestricted activities of daily living including unlimited walking. She, however, did not take part in any sporting activities, as she herself had no wish to take part in any sports. If she elects to participate in any sport in the future, we would advise against competitive sports. She had no deformities in her lower extremities and had had no surgeries for feet ankles or the knees. The patient had regular postoperative follow-up annually for 15 years. The patient remained fully ambulatory without pain or supports. At the 15-year follow-up, both hips had excellent range of motion with no leg length discrepancy, and radiographs showed no evidence of loosening; the hip dysfunction and osteoarthritis outcome score was 74.4 [13].

Discussion

The patient had typical features of hip involvement with Morquio syndrome including severe hip dysplasia, osteoarthritis, flexion contractures, pain, and restricted mobility [2-5]. Literature search showed no previous case report of simultaneous bilateral total hip arthroplasty in a very young patient with Morquio syndrome, which was indicated in this patient because of extensive bilateral hip contractures and osteoarthritis that caused extreme immobility and disability. Extensive preoperative evaluation that included imaging of the entire spine was mandatory because of the risk of developing spinal cord compression in patients who have Morquio syndrome [14].

The long-term outcome after total joint arthroplasty in Morquio syndrome is unknown because there are only 2 cases of staged bilateral total hip arthroplasty reported in the English literature [15,16]. In one report, a 26-year-old patient who had Morquio syndrome had noncemented hip arthroplasty because the femoral canals were large enough to accommodate a larger stem (size 3, ABG II, Howmedica, Mahwah, NJ), and a right calcar fracture was treated with cerclage wires as in the present patient; 50-mm acetabular shells were used, and follow-up at 5 years showed good function and no loosening [14]. The other reported patient

was a 25-year-old woman who had cemented femoral stems (Elite CDH, DePuy, Warsaw, IN), but the authors did not comment on the size of the femoral stems or cups; follow-up was 7 years [15]. There were 2 other case reports identified in the non-English literature that reported good to excellent results after total hip arthroplasty in Morquio syndrome [16,17]. In this patient group, the femoral canals are very narrow with mismatch between the metaphysis and diaphysis along with possible osteopenia related to nonweight bearing, which could lead to increased risk of fractures as seen in our case and previous reported case. A preoperative bone densitometry and the preoperative therapy with vitamin D supplements may help restore bone stock. Also, the use of custom made implants with accurate stem sizes could prevent fractures. We would have preferred to use noncemented implants, but cemented components were necessary because the small femurs enabled use of only the smallest available stem in the arthroplasty system, and the shallow acetabula necessitated the use of cemented cups.

Summary

Total hip arthroplasty at a young age may be necessary in patients who have Morquio syndrome because of severe arthritis and soft tissue contractures. The present patient had excellent long-term hip function and mobility. Extensive preoperative evaluation that included imaging of the entire spine was mandatory because of the risk of developing spinal cord compression in patients who have Morquio syndrome.

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