**CASE REPORT**

Total knee replacements in a patient with the Morquio syndrome

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The Morquio syndrome is a rare disorder which presents with a number of musculoskeletal problems. The literature describing total knee replacement in these patients is sparse. We describe the management of a patient with bilateral instability and pain in the knees using bilateral constrained knee replacements, and followed up for five years with pre- and post-operative knee scores. We highlight the difficulties encountered and discuss the end results.

The Morquio syndrome is a rare condition with an incidence of 3:1 000 000. It is a mucopolysaccharidosis type IV and is inherited as an autosomal recessive. The main defect is in the metabolism of glycosaminoglycans, especially keratan sulphate, due to enzyme defects in lysosomes resulting in abnormal deposition in the connective tissues. The condition does not affect the development of the brain, but has characteristic clinical features.

The musculoskeletal presentation is typically short-trunked dwarfism with genu valgum. Patients almost invariably have atlantoaxial instability, prominent lax joints, kyphoscoliosis and pectus carinatum.

The extraskeletal manifestations are medically significant and include aortic stenosis and hypoplastic lungs. There may be associated hepatomegaly and splenomegaly, which accounts for the abdominal distension seen in infancy.

Management of arthropathy of the knee joint is a challenging biomechanical undertaking because of the short stature and limb morphology with deformity in three planes, and is further complicated by potential medical problems. This paper describes our experience of bilateral total knee replacement (TKR) in a patient with the Morquio syndrome and highlights the challenges encountered in management.

**Case report**

A 22-year-old man with the Morquio syndrome presented with a two-year history of progressive instability and pain in the knees. Although his pain was bilateral, his left knee was worse at the time of presentation. Functionally, he had limited mobility and was largely dependent on his wheelchair. He was able to walk short distances, but only indoors.

He had undergone an occipitocervical fusion as a child, and MRI of the cervical spine did not reveal any evidence of spinal stenosis or myelopathy.

Examination revealed that he had a marked spinal kyphosis and was of short stature. His knees were valgus in alignment, with accompanying planovalgus feet and marked joint laxity (Fig. 1). Radiographs (Fig. 2) showed bilateral severe arthropathy of the knee joints. He scored 22 on the Oxford Knee Score in both his knees.

In view of his excessive joint laxity and short stature a full constrained custom-made prosthesis was ordered (Zynergy Orthopaedics, Rotherham, United Kingdom). Full-length radiographs of his legs and CT images were reformatted to produce three dimensional (3D) reconstruction (Fig. 3), following which a virtual computer model was developed and the implants were designed from these images.

Pre-operative investigations revealed no cardiac or renal problems, but he was noted to have reduced lung volume on spirometry. Surgical technique. A standard midline approach was used and his right knee was replaced first. This procedure was carried out without a tourniquet because of the small size of the leg. The femoral component had to be shortened with a diamond saw in order to seat it appropriately in the femur. A generous soft-tissue release was performed. The iliotibial track had to be divided to allow full extension of the knee. The operation was complicated by severe blood loss, necessitating admission to the intensive care unit. Post-operatively he lost terminal extension of the knee, which improved only marginally with physiotherapy.
Four months following the first operation he underwent a left TKR. A standard midline approach was used, but this time a paediatric tourniquet was employed. The anterior flange on the tibial component was removed and the femoral component was shortened to allow adequate insertion. Unfortunately, he developed a post-operative peroneal nerve palsy, which recovered completely by 12 months. Radiographs (Fig. 4) and alignment (Fig. 5) were satisfactory. The Oxford knee score was 39 in both knees. At five years there was a good range of movement of both knees with flexion to 90° and loss of the terminal 10° extension on the right. Both knees were stable and without pain.

Discussion
The literature on the management of problems in the knee in the Morquio syndrome is sparse. Osteotomies have been used to correct limb alignment, but the joint laxity requires the subsequent use of a brace, with poor functional results. Neither of these procedures slow down the destruction of the joint or allow improved function.

Two previous papers have described the use of a joint replacement to manage arthropathy of the knee in the Morquio syndrome. DeWaal Malefijt et al used the smallest available posteriorly stabilised prosthesis and needed extensive soft-tissue releases to achieve ligament balancing. A custom-made prosthesis was used by Heisel and Hesselschwerdt. In our case, custom-made prostheses were used. The difficulty was further compounded by the shape of the patient’s femur, which had multiplanar deformities, and despite having tailored implants, difficulty was still encountered in inserting the stemmed femoral components. Both of the stems had to be shortened intraoperatively. This problem occurred because of overestimation of the dimensions of bony anatomy on the 3D CT reconstructions.

The first operation was complicated by severe bleeding because the tissues were generally very vascular, and this may have contributed to an inadequate release of the posterior capsule, leading to the post-operative loss of extension. The patient also developed an acute hepatic transaminitis (isolated rise in the transaminases), which led to a coagulopathy. No direct cause was identified, but it was thought to be due to a combination of tissue hypoperfusion and hypoxia on a background of regular use of paracetamol. However, he made a full recovery. For the left knee replacement, a paediatric tourniquet was used to good effect, allowing adequate soft-tissue release.
At the five-year follow-up he was satisfied with the outcome. The knees were stable, and he was no longer in pain and had improved knee scores.

Total knee replacement in patients with the Morquio syndrome is a challenging procedure. These patients require a full and thorough anaesthetic assessment, because they often have clinically subtle but significant malfunction in their lungs or heart. As a consequence, intensive-care facilities must be available to manage any complications. They may also experience unpredictable post-operative problems, such as a coagulopathy, or hepatic malfunction. In view of the small size of the patient and the ligamentous laxity, custom-made fully constrained implants are required, and facilities for modification of the implant should be available intra-operatively.

We recommend the use of a tourniquet in view of potential bleeding secondary to the increased vascularity of the soft tissues. Paediatric tourniquets are the most appropriate for this purpose.

Although the Morquio syndrome is a rare condition, bilateral constrained custom knee replacement provides an excellent restoration of function with the elimination of pain and instability.

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References