Patients with skeletal dysplasia are prone to developing advanced osteoarthrosis of the knee requiring total knee replacement (TKR) at a younger age than the general population. TKR in this unique group of patients is a technically demanding procedure owing to the deformity, flexion contracture, generalised hypotonia and ligamentous laxity. We retrospectively reviewed the outcome of 11 TKRs performed in eight patients with skeletal dysplasia at our institution using the Stanmore Modular Individualised Lower Extremity System (SMILES) custom-made rotating-hinge TKR. There were three men and five women with mean age of 57 years (41 to 79). Patients were followed clinically and radiologically for a mean of seven years (3 to 11.5). The mean Knee Society clinical and function scores improved from 24 (14 to 36) and 20 (5 to 40) pre-operatively, respectively, to 68 (28 to 80) and 50 (22 to 74), respectively, at final follow-up. Four complications were recorded, including a patellar fracture following a fall, a tibial peri-prosthetic fracture, persistent anterior knee pain, and aseptic loosening of a femoral component requiring revision. Our results demonstrate that custom primary rotating-hinge TKR in patients with skeletal dysplasia is effective at relieving pain, with a satisfactory range of movement and improved function. It compensates for bony deformity and ligament deficiency and reduces the likelihood of corrective osteotomy. Patellofemoral joint complications are frequent and functional outcome is worse than with primary TKR in the general population.

Skeletal dysplasia encompasses a number of genetic disorders characterised by abnormal development and remodelling of bone and cartilage.\(^1\) The presence of multiple joint pathology, abnormal alignment, generalised hypotonia and ligament laxity predisposes to knee pathology at a young age.\(^2\) Early joint salvage options include soft tissue releases and corrective osteotomy to improve congruity, alignment and stability. However, when painful osteoarthritis (OA) develops, total knee replacement (TKR) is indicated. In these patients TKR is often complicated by articular and metaphyseal angular deformity, bone loss, narrowness of the femoral and tibial canals, flexion contracture and ligament laxity.\(^3\)\(^-\)\(^7\) Furthermore, abnormal lower limb rotational profiles often predispose to a chronically subluxed or dislocated patella.\(^5\)\(^-\)\(^7\)

Total hip replacement (THR) reduces pain and improves function in patients with skeletal dysplasia,\(^8\) but there is little information on the results of TKR in such patients: one study reported six complications (43\%) using a variety of modular prosthetic designs in 14 patients followed up for a mean of 3.4 years.\(^9\) Two patients required intra-operative distal femoral osteotomy because of metaphyseal angular deformity, both of which failed to unite and required further surgery. Given the limited follow-up of that study, the mid- to long-term results of TKR in this situation remain unknown. We therefore evaluated the mid-term functional outcome, radiological findings, survival and complications after implantation of a computer-aided design and computer-aided manufactured (CAD-CAM) rotating-hinge TKR system in patients with skeletal dysplasia.

**Patients and Methods**

We identified and retrospectively reviewed eight patients who had 11 primary custom-made rotating-hinge TKRs for advanced OA secondary to skeletal dysplasia between 2000 and 2008. The indications for using this device were painful end-stage OA of the knee with associated substantial femoral or tibial bone loss and angular deformity combined with ligamentous instability due to deficient collateral ligaments (Fig. 1). Clinical data were obtained from the case notes, outpatient reviews, imaging studies and functional knee questionnaires. There were three men and five women, with a
mean age of 57 years (41 to 79) at the time of surgery, a mean height of 138 cm (122 to 155) and a mean weight of 56 kg (40 to 102); seven different dysplastic conditions were involved (Table I). Three patients had bilateral procedures. No patient was lost to follow-up, which left 11 knees available for review. Patients were followed clinically and radiologically for a mean of seven years (3 to 11.5). Functional scoring was performed during telephone interviews and clinic reviews, and the radiographs were examined.

The prosthesis. The Stanmore Modular Individualised Lower Extremity System SMILES prosthesis (Stanmore Implants Worldwide Ltd, Stanmore, United Kingdom) is a second-generation rotating-hinge design manufactured using CAD-CAM technologies (Fig. 2). The femoral component is made of cobalt–chromium–molybdenum (Co-Cr-Mo) alloy with a titanium stem. It consists of a Co-Cr-Mo axle, a pair of ultra-high-molecular-weight polyethylene (UHMWPE) bushes and a titanium circlip. The tibial component is also made of Co-Cr-Mo alloy and consists of a UHMWPE bearing surface and a titanium stem and casing. Rotation occurs between the tibial component and the casing. The length, diameter, curvature and angulation of the femoral and tibial stems are customised based on pre-operative biplanar radiographs to optimise fixation and compensate for skeletal deformities and bone loss. The bearing surfaces are bevelled to constrain rotational movement beyond 5°, and there is a hyperextension bumper pad. These design features compensate for ligamentous instability, which is a common feature in patients with skeletal dysplasia (Fig. 3). Customised plateau plates and wedges are also available. The SMILES prosthesis incorporates many of the design features of second-generation rotating-hinge devices to improve the biomechanics of the joint and provide better clinical outcomes.\textsuperscript{10}

Table I. Primary diagnoses of patients

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Achondroplasia</td>
<td>2</td>
</tr>
<tr>
<td>Spondyloepiphyseal dysplasia</td>
<td>1</td>
</tr>
<tr>
<td>Pseudoachondroplasia</td>
<td>1</td>
</tr>
<tr>
<td>Multiple epiphyseal dysplasia</td>
<td>1</td>
</tr>
<tr>
<td>Morquio syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Diastrophic dwarfism</td>
<td>1</td>
</tr>
<tr>
<td>Larson's syndrome</td>
<td>1</td>
</tr>
</tbody>
</table>

Fig. 1a
Radiographs of the features of a female patient aged 42 years at operation with pseudo-achondroplasia in whom a custom rotating-hinge total knee replacement was indicated, in a) anteroposterior and b) lateral radiographs of the knee showing deformity of the metaphyseal bone and a subluxed patella, and c) long-leg radiographs showing valgus malalignment. A custom-made replacement for arthritis has already been performed in the right hip.

Fig. 1b
Fig. 1c

Fig. 2
Pre-operative diagram of a computer-aided design and computer-aided manufactured (CAD-CAM) Stanmore Modular Individualised Lower Extremity System (SMILES) total knee replacement for the patient in Figure 1, demonstrating the sites for bone resection. The tibial and femoral stems may be customised to take into account skeletal deformity.
Surgical technique. All operations were performed at one institution by senior surgeons (JM, RCP, RWJC, JAS, SRC, TWRB). Flexion contractures were present in ten knees, varus malalignment in three and valgus malalignment in eight. Subluxation or dislocation of the patella was present in six knees, and all 11 had mediolateral or anteroposterior instability or both. All knees requiring this procedure had articular incongruity, metaphyseal angular deformity and ligamentous laxity. All procedures were performed with the patient in the supine position with a tourniquet, foot bolster and side support for the operated knee. One dose of prophylactic intravenous antibiotics was given at induction on the basis of the prosthesis and the custom-designed anatomical bow of the femoral stem. Patella infera is present, which is a frequent observation in these patients.

Immediate post-operative anteroposterior (a) and lateral (b) radiographs of the patient in Figure 1, demonstrating the satisfactory position of the prosthesis and the custom-designed anatomical bow of the femoral stem. Patella infera is present, which is a frequent observation in these patients.

Results

Functional outcome. The mean Knee Society score improved from 24 points (14 to 36) pre-operatively to 68 (28 to 80) at final follow-up. The mean function score improved from 20 points (5 to 40) pre-operatively to 50 points (22 to 74) at final follow-up. Knee pain and function improved in all patients. All patients with scores < 50 points had multiple joint pathology that compromised function. All eight patients required walking aids pre-operatively, and four required them post-operatively due to multiple joint involvement. One patient was wheelchair-bound as a result of a stroke four years after the primary operation. The mean range of flexion was 73° (40° to 105°) pre-operatively and 91° (70° to 110°) at final follow-up. Flexion contractures decreased from a mean of 17° (10° to 40°) pre-operatively to 3° (0° to 5°) at final follow-up.

Radiological outcome. All components were found to be stable at final follow-up. A non-progressive RLL (< 2 mm wide) was identified underneath the tibial tray in two
Complications and survival.

Four complications were recorded. One patient sustained a (non-resurfaced) patellar fracture following a fall at one month and required open reduction and internal fixation. The fracture failed to unite and the patient required revision of fixation with bone grafting. The fracture subsequently united and the patient now walks with mild pain in the knee and a 10° extensor lag. One patient sustained a fracture of the tibia below a well-fixed tibial component after a fall. The fracture united following open reduction and internal fixation, and the patient currently has a pain-free knee. One patient, described above, required revision of the femoral component for aseptic loosening at five years. The bushings and circlip were removed, the prosthesis dismantled, and the original femoral component was re-cemented after exchange of the bushings and circlip. The tibial component was found to be well fixed and was not adjusted. This patient currently walks with a pain-free knee and a range of movement of 0° to 90°. One patient with a non-resurfaced patella developed clicking and anterior knee pain post-operatively and underwent patellectomy at 37 months. In this patient an intra-operative lateral release had been performed, but patella infera was present and radiographs showed the patella impinging on the tibial component during flexion (Fig. 4). Pain was improved following patellectomy but a 30° extensor lag remained. There was no requirement for intra-operative osteotomy and there were no intra-operative fractures, implant breakages or infections.

Discussion

Total joint replacement in patients with skeletal dysplasia is technically challenging because of the abnormal bone morphology, soft tissue contractures, ligament deficiency and generalised hypotonia. TKR in particular is often complicated by significant articular and metaphyseal angular deformity, bone loss, flexion contracture and collateral laxity, and the abnormal lower limb rotational profiles often predispose to a chronically subluxed or dislocated patella.

We recognise the limitations of our study, with only a small number of patients and the lack of a control group or published randomised control data against which to compare our results with modular semi-constrained and/or condylar TKR designs. A further limitation is the confounding effect of multiple joint pathology and medical comorbidities. This makes interpretation of functional outcome difficult, as spinal and hip pathologies affect lower limb function, and one patient had had a stroke that rendered him wheelchair bound. Selection, measurement and interviewer bias may have also affected our functional assessment. Nevertheless, this study represents the first report in the literature on the use of custom-made rotating-hinge TKRs in this rare group of patients, and shows encouraging mid-term results.

Early rotating-hinge designs used in revision knee surgery were abandoned because of poor functional results and survival rates. High force transmission across the bone–cement interface resulted in early loosening and bone loss. The second generation of rotating hinges incorporated several design features to improve survival and functional outcomes. The hinge joint became more congruent to reduce stress across the articulation, and a mobile bearing was added allowing the implant to rotate. This design theoretically provided a more congruent articulation, reducing contact stresses and subsequent force transmission across the interfaces. Modular fluted stems were incorporated to improve alignment and press-fit fixation, and modular segments were added for the filling of bone defects.

These features have made these prostheses particularly attractive in the revision setting when global instability and bone loss are present, but they should be used with caution. Pour et al reported a rate of survival at one year of 79.6% in 43 patients with salvage revision knee reconstructions using a modern rotating-hinge TKR, with seven revisions and 15 re-operations at a mean follow-up of 4.2 years.

Rotating-hinge designs are indicated for patients with bone loss and ligament deficiency, which make them suitable as a primary device for patients with skeletal dysplasia. A minimally constrained conventional prosthesis can be used as primary TKR in patients with skeletal dysplasia when these features are not present. Patellofemoral com-
lications are frequent with rotating-hinge designs (6% to 22%)\(^{10,16,20}\) and in patients with skeletal dysplasia (10%),\(^9\) and this was the main complication we encountered in the patients in this series, with an incidence of 18%. In one patient anterior knee pain and clicking necessitated patellectomy owing to impingement of the patella on the tibial component of the prosthesis during flexion. The addition of a deeper patellofemoral groove on the prosthesis may reduce this complication in the future.

Only one patient in the series required revision for isolated aseptic loosening of the femoral component at five years, which was managed with attention to the femoral component and the bearing alone.

The functional outcomes we observed were worse than those reported for condylar designs used for patients with primary OA or rheumatoid arthritis,\(^21\) and marginally worse than for the rotating-hinge designs used in salvage revision knee surgery.\(^{10-19}\) Patients with skeletal dysplasia share many common features, including short stature, spinal deformity, atlantoaxial instability, lower limb malalignment and foot deformity, ligamentous laxity, and generalised hypotonia that predisposes them to premature degenerative joint disease.\(^1\) Multiple joint pathology is one explanation for the low functional scores that we observed relative to the general population. The Harris hip scores\(^22\) following total hip replacement in these patients have also been found to be lower than in the general population.\(^8\)

The only study in the literature on the use of TKR in patients with skeletal dysplasia reported on condylar, semi-constrained and non-custom rotating-hinge designs.\(^9\) In 14 patients with diastrophic dysplasia followed for a minimum of two years (mean 3.4 years), six complications were reported. Dislocation of the patella occurred in two knees, there were two fractures of the distal femur and proximal tibia, and two distal femoral corrective osteotomies were required for metaphyseal angulation. Both osteotomies failed to unite, requiring re-operation, and the two dislocated patellae were managed non-operatively. The investigators reported difficulty inserting the modular components owing to bony deformity and narrowness of the intramedullary canals. The custom rotating-hinge prosthesis used in our study allows for anatomically designed femoral and tibial components. The curvature and diameter of the intramedullary alignment rods and the implant stems are designed to match the abnormal anatomy, avoiding intra-operative fractures and the need for osteotomy and its risk on mal- or nonunion. No patients in our study required intra-operative osteotomy or sustained an intra-operative fracture. Improvements in the range of flexion and correction of flexion contractures were also superior in our study. This probably reflects the greater soft-tissue release achievable with a rotating-hinge design than that possible with a condylar or semi-constrained design before stability is compromised. None of the patients in our series had a weak or non-functioning extensor mechanism jeopardising the outcome of surgery, and we did not encounter any post-operative stability problems. Notwithstanding this, there are occasions when a SMILES fixed-hinge system would be more appropriate, such as in patients with a defective extensor mechanism and significant knee hyperextension pre-operatively.

Thus, good results with satisfactory movement and improved function are obtained following the use of a custom-made rotating-hinge TKR in patients with OA due to skeletal dysplasia.

The authors acknowledge the contribution made by Stanmore Implants Worldwide Ltd and particularly Dr P. Unwin. We also thank Professor G. Blunn, from the Institute of Orthopaedics and Musculoskeletal Science, University College London, for his input.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References