We present a 25-year-old patient with juvenile rheumatoid arthritis and ankylosis of both hips and both knees treated by staged bilateral hip and knee arthroplasty. She was followed up for 18 months. We discuss the pre-operative planning, surgical details and post-operative rehabilitation.

Ankylosis of a joint describes total loss of movement. This can occur either spontaneously, as a result of an inflammatory or infective condition, or deliberately after operation. Primary joint fusion is now rarely undertaken except for uncontrolled joint sepsis or for patients considered too young for primary joint replacement. In the long-term, problems in the adjacent ipsilateral and contralateral joints are common. After 20 or more years, 35% to 75% of patients with ankylosis of the hip develop back pain, 17% to 28% have been reported to have contralateral hip pain, and 24% to 57% ipsilateral knee pain.

Conversion of an ankylosed joint to a total joint replacement can be technically demanding and requires careful pre-operative planning. Isolated conversion of an ankylosed hip or knee to a total joint replacement has been reported, but to our knowledge, the conversion of bilateral hip and knee ankylosis to a joint replacement in the same patient has not been described previously. We present the results 18 months after surgery of a patient with bilateral fibrous ankylosis of the hips and bony ankylosis of the knees converted to total joint replacement.

Case report
A 25-year-old female presented with a history of juvenile rheumatoid arthritis (RA) since the age of 16 years. She had developed progressive joint deformity in both the upper and lower limbs. She had reasonable function of her hands despite involvement of her shoulders and elbows. However, bilateral ankylosis of her hips and knees severely limited function. She was only able to move with a pulpit type rollator frame and her walking distance was less than 50 metres because of pain in both legs. Sitting was difficult and painful, as the position of her hips and knees dictated that her feet had to be supported on a high foot-rest. Standing from the sitting position was only possible with the help of an assistant. Lying flat required support under the lower limbs. There was a family history of RA.

On physical examination, the right hip had a fixed deformity of 40º flexion, 30º adduction and 35º external rotation. The left hip had a fixed deformity of 40º flexion, 10º adduction and 20º internal rotation. Both knees were ankylosed in 10º of fixed flexion, but because of the rotational deformities at the hip, they appeared to have a valgus angulation. Standard radiographs of both hips and knees showed that all four joints were ankylosed (Fig. 1). The laboratory investigations were normal.

Although she presented initially because of her knees, it was clear that the ankylosis of her hips should be treated first. She was therefore offered staged bilateral total hip replacement (THR) followed by staged bilateral total knee replacement (TKR). The aim of the surgery was to provide movement at the hips and knees, to improve her pelvic and spinal posture and help her ability to stand, transfer and balance.

She underwent right THR followed by left THR 17 days later. The procedures were performed through a posterior approach with the patient positioned laterally, and particular attention was paid to padding and protection of the skin in view of her poor nutrition. A cemented Ogee socket (Depuy, Leeds, United Kingdom) was used. Reaming of the femur was difficult because the fused knee restricted full internal rotation of the hip. A cemented C-
Stem (DePuy) was inserted with a ceramic head (Fig. 2). She was mobilised on the second post-operative day after the right hip replacement. A slide sheet and three people were needed to get her out of bed and she was mobilised with a pulpit frame. She had to sit out in a high chair with her legs supported because of the fixed deformities in the other joints. She progressed from a pulpit frame to two gutter crutches by the eighth post-operative day and she was walking independently with gutter crutches 11 days after operation. However, she still needed the assistance of two people for transfers. A similar protocol was followed after her left hip replacement.

At an interval of five months following bilateral THR, TKR was performed on the ankylosed left knee. A medial parapatellar approach was used. The fused patellofemoral joint was osteotomised from the medial side and a White-side tibial tuberosity osteotomy,\textsuperscript{23} hinged laterally, was performed to aid exposure. The tibiofemoral ankylosis was then divided with an osteotome. The bone was very soft, resulting in a fracture of the anteromedial tibial plateau extending into the osteotomy. The posterior cruciate and collateral ligaments were intact and the trochlea relatively normal. It was therefore decided to use a posterior cruciate ligament retaining implant. A stemmed tibial component was used because of the osteotomy of the tuberosity and the fracture of the plateau. Finally, the patella was resurfaced and a lateral retinacular release carried out to improve patellar tracking (Fig. 3). Post-operatively a continuous
passive motion machine was used with knee flexion between 0° and 20° for approximately two hours per day. She was mobilised on the third day, partially weight-bearing with a gutter frame. Active flexion was started on the fourth post-operative day and the range of flexion on continuous passive movement gradually increased. By 11 days after operation, she had achieved independent mobilisation on a gutter frame and then progressed to gutter crutches. Hydrotherapy was started on day 15 when the sutures had been removed. She was discharged home, partially weight-bearing, 23 days after operation when she was independent on gutter crutches with active knee flexion of 0° to 75° and active straight leg raising. She continued partial weight-bearing for six weeks and used the continuous passive movement machine at home.

Right TKR was performed three months after the left using the same surgical exposure, but the right side was more complex because of external rotation deformities of 30° in the distal femur and 20° in the tibia. The exposure was also more difficult and required an extensive release of the lateral collateral ligament, the iliotibial band and the posterolateral corner. The quadriceps was also mobilised from the anterior aspect of the femur proximally up to the mid-thigh. Great care was taken, when using the osteotome to separate the ankylosis, to avoid fracture of the tibial plateau as had occurred on the left side. A cemented femoral component and an offset tibial tray with a posterior stabilised insert were used (Fig. 4). Post-operatively, because of the extensive soft-tissue release, she was mobilised one week later using a canvas cricket pad splint with the knee in extension and a gutter frame. Continuous passive movement was used on both knees and active knee flexion was started on the sixth day. The range of movement on continu-
uous passive movement and active flexion was gradually increased to 0° to 80° and 0° to 70° respectively. She started to mobilise on gutter crutches on the tenth day, and was independent on them by discharge 21 days after operation. The padded cricket splint was discontinued by the second week.

At follow-up, 18 months after her second TKR, she was able to mobilise with one gutter crutch. The range of movement at the right knee was 10° to 100° with a 10° extensor lag, and 10° to 90° at the left. The right hip had flexion of 80°, abduction of 25°, and external and internal rotation of 10°. The left hip had 80° flexion, 35° abduction, and external and internal rotation of 15° and 10° respectively.

Discussion
Since the advent of joint replacement, the indications for primary fusion of the hip and knee joints have diminished considerably. Patients with a successful ankylosis in a good position usually maintain excellent function for a long period. Symptoms related to the adjacent joints become intrusive only after a lapse of 20 to 30 years. Conversely, with a malpositioned fusion, pain related to degenerative changes in the neighbouring joints develops more rapidly.4,25

There have been several reports of the conversion of ankylosed knees9,13,21-24 or hips1,13,5,8,10,11,13,14,25 to total joint replacement. In juvenile RA, THR or TKR are indicated for marked functional impairment and/or disabling pain.25 Our patient with juvenile RA and bilateral ankylosis of the hips and knees required total joint replacement of all four joints for marked functional impairment and pain.

The ankylosis of the hips was converted to THR for several reasons. Firstly, a TKR beneath a hip arthrodesis will be under abnormal stress.1,26,27 Secondly, tensioning of the muscles crossing both hip and knee joints is better addressed by treating the hip first.25 Thirdly, rehabilitation of the hip is possible with an ankylosed knee but it is very difficult to rehabilitate the knee with stiff hip,25 and lastly, recovery from THR is usually less painful and demanding than after TKR. This helps the patient to gain confidence for the second stage.25 Correction of the hip deformity re-aligns the knee and simplifies knee replacement, while treating both hip joints before the knees addresses the problems of abnormalities of pelvic obliquity and sitting. It had been intended to replace the right hip first, as it was the more severely deformed, and to proceed to the left hip a week later. However, the second hip was delayed for more than two weeks because the patient developed severe anaemia after the first operation. There was a delay of three months between the two knee joint replacements as the patient found it difficult to cope emotionally with repeated major surgery and long periods in hospital.

The THRs were undertaken via the posterior approach preferred by the senior author (MLP), with the patient in the lateral position, rather than the lateral or transtrochanteric approach described in a previous series of 54 patients with juvenile chronic arthritis.28 The anterior contractures were addressed by gradual release. For the knees a Whiteside tuberosity osteotomy was performed to improve surgical access and healed well.25,29,30 Although the peripheral pulses were present, there was evidence of a rheumatoid vasculitis which raised concerns regarding wound healing.

In view of the soft bone, cemented total joint replacements were used. The intra-operative fracture of the tibial plateau in the left knee was protected by a long stemmed implant and partial weight-bearing with crutches. The right knee was more complex, requiring extensive release of the lateral structures and the posterior cruciate ligament, which warranted the use of a cemented posterior cruciate substituting implant. A quadriceps release from the femur was also required which may have accounted for the extensor lag at the latest follow-up. In 1983, Sarokhan et al21 published their five-year results of 29 TKRs in 17 patients with juvenile RA. They used many types of prosthesis and had a survival rate of 97%, with improvement in pain, range of movement and function. Thomas et al32 described 17 cases of cemented TKR with no revision for aseptic loosening at a mean follow-up of six years. However, Rydholm, Boegård and Lidgren33 reported six revisions, four of which were required for loosening, in a series of only 11 patients. Cage, Granberry and Tullos34 reviewed 13 TKRs in seven patients with juvenile RA at 11 years and also found a high rate of revision (23%) secondary to peri-prosthetic fracture and loosening. One of the main concerns when performing TKR in these patients is their young age. In an effort to preserve bone stock and improve longevity, several authors have used uncemented press-fit components.35,36 In a series of 77 patients with juvenile RA, Lybäck et al35 found that cementless TKR afforded excellent pain relief and functional improvement, with a 99% survival at five years. Boublik et al36 also noted significant improvements in pain and function, with 100% survival at four years after cementless TKR. However, because of concern over poor bone quality and the need for tight interference fit, they cemented ten of their 22 tibial components.

Perhaps the most important question in this young population is the durability of the prosthesis. The largest study to date by Lehtimäki et al37 involved a registry of 186 hips followed for 15 years. They found 92% survival in patients who had a Charlnley type prosthesis. Chnell et al38 reported 67% survival of both components at a minimum of ten years. However, in a radiological review of cemented hips, Williams and McCullough39 noted impending failure because of radiological loosening in 43% of patients at five years, and Witt et al30 described aseptic loosening in 42% of their patients. Recently Wróblewski, Siney and Fleming40 reported that in 130 of 292 hips, who attended follow-up, where 33 hips could not be traced, and 61 patients (88 hips) had died, the survival was 74% at 25 years.
Colville and Raunio stated that the greatest risk of complication occurs during the operation. The femoral canals in these young patients are hypoplastic and osteopenic. There is a risk of fracture during exposure and femoral preparation. Severe proximal femoral torsion secondary to long-standing contractures and premature closure of the physes, can affect insertion of the implant and post-operative stability. Associated cortical thinning presents a characteristic metaphyseal and diaphyseal mismatch that should be considered when selecting a cementless implant. Access to a wide range of femoral components is crucial for successful surgical management. Haber and Goodman recommended the use of standard off-the-shelf cementless components. However, careful review of their patients suggests that this can lead to unpredictable fixation. In their study, 29 proximal femurs were prepared for cementless components but in only 20 was stable fixation achievable. The remaining nine had to be cemented, and only 17 of the 20 cementless components developed stable bony ingrowth. Scott reported the need to customise components intra-operatively to obtain initial fixation.

At our last follow-up, the ranges of movement achieved at the hip and knee were comparable to those reported in the literature. By 18 months after the last joint replacement, our patient’s mobility and quality of life have been improved dramatically. The functional outcome after THR for hip ankylosis has been reported to be encouraging. However, the post-operative range of flexion and hip stability were not as good as those following primary hip replacement in joints that had not been ankylosed. Mullenn and Cameron and Hu presented excellent results for TKR performed on stiff knee joints. However, Naranja et al had a complication rate of 57% after conversion of ankylosed knees to TKR with a satisfactory outcome in only 29% at a mean follow-up of seven and a half years. Clemens and Henkel also concluded that the procedure is technically demanding with a high complication rate.

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

