CASE REPORT

Customised femoral stems in osteopetrosis and the development of a guiding system for the preparation of an intramedullary cavity

A REPORT OF TWO CASES

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Osteopetrosis is a rare disease caused by a major disturbance of bone metabolism. It is characterised by defective osteoclastic function, resulting in increased density of the bone and narrowing of the intramedullary canal. There are three types of this disease which can be broadly defined as infantile malignant autosomal recessive, which is fatal within the early years of life; intermediate autosomal recessive, which appears during the first decade of life but does not follow a malignant course, and the benign adult autosomal dominant type. Patients with the benign type may have a full life expectancy although they are more prone to fractures due to the increased brittleness of the bones. Also, the increased stiffness of the subchondral bone predisposes to osteoarthritis, particularly in the weight-bearing joints, and total hip replacement may be required. According to several reports, the obliteration of the femoral intramedullary canal and increased brittleness creates severe problems in the preparation of the proximal femur often leading to fracture. We have therefore developed a technique to secure gentle and accurate preparation of a cavity to accept a custom-made femoral component in a patient with benign autosomal dominant osteopetrosis with secondary osteoarthritis of the hip.

Case reports

Case 1. A 32-year-old woman presented with severe symptoms due to advanced osteoarthritis of her hips. Radiological investigation, including CT, revealed extreme density cortical and trabecular bone femurs with almost complete obliteration of the intramedullary cavities (Figs 1 and 2). A traditional free-hand technique for reaming would have carried a considerable risk of penetration and fracture of the femur, and the small external diameter of the proximal femur would make it difficult to use a standard femoral component. We therefore designed a CT-based, non-cemented customised component of suitable geometry and size which would allow the preservation of a relatively thick layer of dense bone around the periphery of the implant (Fig. 3). We also designed a broach for the final preparation of the cavity. A special computer-based guiding system for resection of the femoral

Case 1. Radiograph of the pelvis of a 32-year-old woman with benign autosomal dominant osteopetrosis, showing bilateral osteoarthritis and an almost complete obliteration of the intramedullary canal of the proximal femur in both hips.
Case 1. Scanogram a) of the left hip and CT scan b) through the femur at the level of the lesser trochanter. The intramedullary canal is almost completely obliterated with high density bone.

Figure 4a – Diagram showing the guiding system for resection of the femoral neck. The components of the device are produced with a CAD-CAM technique based on information from CT scans. The two clamps secure a correct rotational position, the interconnecting bar maintains a correct distance between the clamps and the rod at the top ensures a correct longitudinal position during mounting. Resection of the femoral neck is performed along the proximal clamp coloured violet. Figure 4b – Diagram showing a mounted guiding block for drilling of a cavity for the broach and the prosthesis after resection of the femoral neck. Drill-bits of specified length and thickness are used in the various holes to secure a suitable preparation of a cavity for the broach and the prosthesis. The cylinders, coloured violet, around the drill bits secure adequate depth of the drilling. The arrow indicates the hole for drilling the intramedullary canal.

Case 1. Post-operative radiograph after implantation of the customised prosthesis on the right hip, seven months after insertion of a similar component on the left.

neck (Fig. 4a) and preparation of the intramedullary cavity (Fig. 4b) was necessary to enable the use of the broach and insertion of the stem. This was developed using computer-aided design and computer-aided manufacturing (CAD-CAM) technique. The two hips were operated upon within a period of seven months, with the first operation on the left. A cemented Charnley acetabular component (DePuy, Warsaw, Indiana) was used on both sides.

There were no problems during the operations. The method enabled insertion of the femoral component with an accurate fit in both hips (Fig. 5). A good contact was achieved between the proximal hydroxyapatite-coated part of the stems and the surrounding bone. The lateral cortex close to the tip of the stem was thinner on the right. She had no pain and good function in both hips when reviewed three years after the first and 2.5 years after the second operation. Radiographs taken then showed, no signs of loosening.

However, at four years she sustained a fracture at the lower end of the right femoral component in a fall. At operation there were no signs of loosening of the prosthesis and the fracture healed after plating and allogeneic bone grafting. At the last follow-up, seven years after the arthroplasty on the left hip and six years and five months on the right, both hips functioned very well with no signs of loosening. The femoral fracture had healed well.

Case 2. The second patient was a 79-year-old woman. Earlier, she had been treated successfully by intramedullary
fixation for a fracture of the right femur. On the left side she had severe symptoms due to necrosis of the femoral head after internal fixation of a trochanteric fracture. The proximal intramedullary cavity was almost completely obliterated (Fig. 6a). A customised femoral component was inserted with a similar technique as used in case 1 (Fig. 6b). No technical problems were experienced during the operation and the hip was functioning adequately two months post-operatively.

Discussion
The technique uses a mechanical guiding system based on computerised information of the geometry and size of the proximal femur, along with an individually designed femoral broach and femoral component. It was successful in both cases and allowed the requisite accuracy to position the implant correctly.

Our system might be an alternative to a computer-controlled reaming system, such as that used by Egawa et al. The use of a surface replacement has also been reported but such a technique might increase the risk of early loosening and fracture of the femoral neck.

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References