■ CASE REPORT

Bilateral total hip replacement in osteogenesis imperfecta with hyperplastic callus

R. Ramaswamy,
Y. Kosashvili,
H. Cameron

*From Holland
Orthopaedic and
Arthritic Centre,
Toronto, Canada*

Osteogenesis imperfecta is a rare inherited disorder of connective tissue which may present with recurrent fractures which are prone to nonunion and malunion resulting in deformity. Some patients develop osteoarthritis of the hip. Formation of hyperplastic callus after recurrent fractures may deform the shape of the femur and preclude the use of standard implants at joint replacement. Replacement can thus be technically demanding.

We present a case of bilateral hip replacement in a patient with osteogenesis imperfecta and hyperplastic callus which was treated by the use of long femoral allografts and cemented femoral stems.

Osteogenesis imperfecta is a rare inherited disorder of connective tissue with varying degrees of severity. Orthopaedic complications include osteopenia, recurrent fractures which are prone to nonunion and malunion resulting in deformities, protrusio acetabuli and malalignment of the limbs. Ligamentous laxity is seen frequently. In the past few decades there has been an increase in the lifespan of patients with osteogenesis imperfecta.¹ This in turn has produced additional problems including osteoarthritis of the hips and knees. One complication of osteogenesis imperfecta is the formation of hyperplastic callus after recurrent fractures. Replacement and reconstruction of the hip in these patients can be challenging and technically demanding. We present a case of bilateral hip replacement in a patient with osteogenesis imperfecta and hyperplastic callus.

Case report

A 54-year-old man with osteogenesis imperfecta who had multiple fractures in childhood presented with pain in both hips. Radiographs showed severe osteoarthritis of the hips with exuberant formation of callus in both femora, suggesting previous fractures. Both femora were deformed and severely osteopenic (Fig. 1).

A primary uncemented right total hip replacement (THR) (S-ROM; Depuy, Warsaw, Indiana) had been performed approximately 13 years previously. This had involved an extended trochanteric osteotomy to facilitate surgical exposure. A histopathological examination of the specimen confirmed the

diagnosis of osteogenesis imperfecta with the formation of hyperplastic callus.

The patient underwent a left THR (Fig. 2). A single-stage procedure was performed using an extended femoral osteotomy. The acetabulum was reconstructed with large quantities of femoral-head allograft and autograft. An uncemented acetabular component was inserted with good primary stability. A femoral allograft was shaped and trimmed after appropriate measurements of the host femur had been made. The allograft was then reamed to accept a Richards femoral stem (Smith and Nephew, Memphis, Tennessee) which was cemented with a cement mantle of at least 2 mm. This composite

Fig. 1

Anteroposterior radiograph of the pelvis showing bilateral protrusio acetabuli and hyperplastic callus in both femora.

■ R. Ramaswamy, MSc,
FRCS(Trauma & Orth), Clinical
Fellow in Lower Extremity Joint
Reconstruction
■ Y. Kosashvili, MD, MHA,
Clinical Fellow in Lower
Extremity Joint Reconstruction
■ H. Cameron, MD, FRCSC,
Orthopaedic Surgeon
Holland Orthopaedic and
Arthritic Centre, 43 Wellesley
Street East, Toronto, Ontario,
Canada M4Y 1H1.

Correspondence should be sent
to Dr Y. Kosashvili;
e-mail: yonasofi@gmail.com

©2009 British Editorial Society
of Bone and Joint Surgery
doi:10.1302/0301-620X.91B6.
22061 \$2.00

J Bone Joint Surg [Br]
2009;91-B:812-14.

Received 14 November 2008;
Accepted after revision 13
February 2009

Fig. 2

Radiograph six months after left total hip replacement (THR) and 13 years after right THR. There is abundant callus in both femora and acetabuli.

Fig. 4

Anteroposterior radiograph of the right femur. The abundant callus allowed for fixation of the osteotomy by multiple screws.

Fig. 3

Radiograph showing loosening of the femoral stem. The white arrows demonstrate the radiolucent line around the stem.

Fig. 5

Radiograph at the latest follow-up 12 years after the left total hip replacement (THR) and two years after revision of the right THR.

was then wedged distally into the host bone. The trochanteric osteotomy was reattached by AO screws and washers. He initially mobilised partially weight-bearing for six weeks. There were signs of union as early as 12 weeks and complete

union was achieved by 20 weeks. At this point he was walking with two sticks.

Three years after this operation he complained of pain in the anterior right thigh and groin. Radiographs showed

loosening of the femoral component on the right side (Fig. 3). He underwent revision of both components. The femoral host bone had poor residual stock compromising reliable distal fixation. Therefore a femoral allograft into which an Exeter component (Howmedica, Rutherford, New Jersey) was cemented was used to reconstruct the proximal femur which was split in the coronal plane and the allograft-cemented stem composite press-fitted into the distal host femoral pedestal. The abundant hyperplastic callus allowed the use of screws to augment the allograft-host bone junction (Fig. 4). The host bone was placed around the composite and secured by large cancellous screws. Satisfactory union and incorporation of the allograft were seen by 24 weeks.

He has been followed up regularly. At his recent visit, 12 years after the left THR and ten years after revision of the right hip, he was walking with two sticks with minimal discomfort. Radiographs showed that both hip reconstructions were working well with no adverse features (Fig. 5). He has recently started experiencing pain in his knees which are affected with osteoarthritis for which he is awaiting surgery.

Discussion

The formation of hyperplastic callus in osteogenesis imperfecta results in progressive enlargement of bone with symptomatic or asymptomatic fracture. It occurs most frequently in the femur but also occurs in the tibia and humerus.² The patients have usually had many previous fractures.^{3,4} The occurrence of hyperplastic callus may be an isolated event or there may be involvement of other bones at intervals of months or years.

The prolonged life expectancy of patients with osteogenesis imperfecta has increased the incidence of osteoarthritis of the hips and knees in these patients.¹⁻⁵ Sub-clinical fractures involving the joints and ligamentous laxity are two major causes of osteoarthritis in these patients. The anatomy of the pelvis, femur and acetabulum is grossly distorted. Protrusion of acetabuli has been observed in 29% of patients.⁶ The pelvis is triradiate and the femur develops the shepherd-crook's deformity.⁵ The presence of hyperplastic callus due to recurrent fractures or previous osteotomies further weakens the bony architecture and gives rise to progression deformity.

The deformity and the degree of osteopenia make replacement and reconstruction difficult. One study has described the outcome of replacement in five hips and three knees with a mean follow-up of seven and ten years, respectively.⁵ All patients had relief from pain and were able to walk with aids. No component had become loose. There was one instance of intrapelvic protrusion of the acetabular component.

In reconstructing large segmental defects the options are limited to substitution of the missing bone with metal or a bone allograft-prosthesis composite. There are concerns regarding this construct in terms of the development of

infection, fractures at the graft-host junction, nonunion at the graft-host junction and early resorption. Blackley et al⁷ in a series of 63 hips with large segmental defects used proximal cemented allograft-prosthesis composites carefully templated to avoid limb-length discrepancy, instability or gaps at the graft-host junction. A step-cut osteotomy in both the host bone and the allograft was performed to enhance the contact between the two before reinforcement of this construct using cortical struts and cerclage wires. They recorded success in 49 hips (78%) after a mean follow-up of nine years with success being defined as an increase in the Harris hip score after operation of more than 20 points, a stable implant and no need for additional surgery.

The structural allograft-prosthesis composite which was wedged into the host bone in our patient acted as a distal load-bearing device. This may have been a possible disadvantage since it resulted in stress shielding of the most proximal femur. However, the senior author (HC) chose this mode of treatment in order to maximise the area of contact between the host bone and the allograft and to minimise the chance of fracture at the host allograft-bone junction.

Successful union at the graft-host junction was achieved in both hips. This may have been due to the fact that the intact blood supply of the membrane bone along with the mechanical stability provided a suitable environment for the allograft to incorporate into the host bone. Distal fixation in this condition may not be reliable because of the distorted anatomy and lack of sufficient supportive diaphyseal bone. A total femoral replacement could have been considered, but the deformities at the knees precluded this as an option. The proximal femoral allograft gave enhanced junctional stability and a good blood supply.

Joint arthroplasty in osteogenesis imperfecta, especially in the presence of femoral and acetabular deformities, requires careful pre-operative planning and expertise in joint reconstruction.

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

1. Whyte MP. Heritable metabolic and dysplastic bone diseases. *Endocrinol Metab Clin North Am* 1990;19:133-73.
2. Roberts JB. Bilateral hyperplastic callus formation in osteogenesis imperfecta. *J Bone Joint Surg [Am]* 1976;58-A:1164-6.
3. Lamovec J, Mozina E, Baebler B. Hyperplastic callus formation in osteogenesis imperfecta. *Ann Diagn Pathol* 2003;7:231-5.
4. Burchardt AJ, Wagner AA, Basse P. Hyperplastic callus formation in osteogenesis imperfecta: a case report. *Acta Radiol* 1994;35:426-8.
5. Papagelopoulos PJ, Morrey BF. Hip and knee replacement in osteogenesis imperfecta. *J Bone Joint Surg [Am]* 1993;75-A:572-80.
6. King JD, Bohechko WP. Osteogenesis imperfecta: an orthopaedic description and surgical review. *J Bone Joint Surg [Br]* 1971;53-B:72-85.
7. Blackley HR, Davis AM, Hutchison CR, Gross AE. Proximal femoral allografts for reconstruction of bone stock in revision arthroplasty of the hip: a nine to fifteen year follow-up. *J Bone Joint Surg [Am]* 2001;83-A:346-54.