Case report

Total femur arthroplasty for revision hip failure in osteogenesis imperfecta: limits of biology

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A B S T R A C T

Osteogenesis imperfecta (OI) is a rare congenital disease characterized by alterations in bone quality, with susceptibility to fractures, instability, deformities, and osteoarthrosis. Prosthetic surgery in these patients is associated with an abnormally high rate of implant failures. On the other hand, abnormal bone fragility adds to the complexity of revision surgery in such individuals—thus representing a genuine challenge for the orthopaedic surgeon. We present a case of femoral reconstruction in a patient with OI and prosthetic loosening after reconstruction secondary to femoral septic pseudoarthrosis. Intramedullary total femoral reconstruction was carried out after exceeding the biological reconstruction limits. This is the first reported instance of the use of an intramedullary total femur arthroplasty as salvage technique in an OI patient. This technique should be considered when we have exceeded biological limits for femoral fixation.

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Introduction

Osteogenesis imperfecta (OI) is a rare congenital disease (1 case per 20,000 births) that affects the connective tissue and results in increased fragility of the bone. The disorder can manifest with different degrees of severity [1]. Nine clinical subtypes have been defined according to the underlying genetic alteration involved, with different impacts on collagen structure, mineralization, and ossification.

The orthopaedic manifestations include osteoporosis, fragile bone, recurrent fractures, misalignment of the limbs, and acetabular protrusion [2]. However, in recent decades, the medical advances have contributed to prolong the life expectancy of these patients. This fact, and the increased risk of joint degeneration secondary to joint fractures and joint laxity, have increased the incidence of hip and knee osteoarthrosis [2,3].

Primary arthroplasty in patients with OI is technically very demanding because of the extreme bone fragility, deformity, soft tissue alterations, and laxity of the ligaments. However, no information is available on alterations in biological implant integration capacity. Papagelopoulos et al. [4] performed 5 total hip arthroplasties and 2 total knee arthroplasties in 6 patients with OI (one patient subjected to total hip arthroplasty and contralateral total knee arthroplasty), and recorded a high acetabular protrusion rate. Krishnan et al. [5] described the usefulness of customized implants in treating the anatomical anomalies of patients of this kind.

We present a case of total femoral arthroplasty in a patient with OI, following loosening of a hip revision stem implanted after 2-step surgery for femoral septic pseudoarthrosis with associated coxarthrosis.

Case history

A 63-year-old male with OI and a history of multiple fractures in childhood reported with pain in the middle third of the thigh. The patient was unable to walk and also presented with a fistula on the
lateral surface of the thigh (Fig. 1a). He had undergone surgery 1 year prior for a femoral fracture. The radiographic study revealed atrophic septic pseudoarthrosis with disruption of the osteosynthesis material and severe coxarthrosis, as well as deformation of the femoral neck (Fig. 1b).

Two-step surgery was carried out. In the first step, we eliminated the fistula and removed the osteosynthesis material, the bone margins and all the poorly vascularized tissues, followed by implantation of an antibiotic-loaded cement spacer: 1 g of clindamycin and 1 g of gentamycin per package of 40 g (Copal; Heraeus Medical GmbH, Wehrheim, Germany), reinforced with a Rush pin (Fig. 2). The microbiological analysis of the samples obtained proved positive for methicillin-resistant *Staphylococcus aureus*. The patient was maintained without loading of the limb and intravenous antibiotic treatment was provided for 15 days, followed by oral levofloxacin and rifampicin for an additional 6 weeks. After confirming healing of the infection, second step surgery was performed, extracting the spacer, with tissue debridement and femoral reconstruction in the form of total right hip arthroplasty using a Continuum trabecular metal acetabular insert and Wagner SL stem (Zimmer, Warsaw, IN). A structural allograft was required to reinforce the bone defect in the femoral diaphysis, stabilizing it with a LISS plate (Synthes, Zuchwil, Switzerland) with trochanteric cerclage (Fig. 3).

Loading was avoided for 6 weeks, followed by partial loading until week 12, when allograft consolidation was noted, and then progression to normal loading with the help of 2 walking sticks was allowed. After initial stem collapse at loading of the limb, the patient received treatment with teriparatide (FORTEO, Eli Lilly, Indianapolis, IN) for 24 months. During this time, the patient was able to walk with the help of 2 crutches, without pain or radiographic changes.

Two months after ending treatment with Teriparatide (28 months after surgery), the patient developed progressive pain in the knee that gradually prevented him from walking over the subsequent months until walking became impossible 3 months after symptoms onset. The serial radiographic study showed loosening of the femoral component with progressive stem

![Figure 1. Sinus tract on the lateral surface of the thigh (a); radiograph views showing disruption and the presence of pseudoarthrosis (b and c).](image-url)
Figure 2. Views of first step surgery. Note the bone defect due to septic pseudoarthrosis (a and b), the segmental cement spacer (c) in the intraoperative views and X-ray control of the segmental spacer (d and e).

Figure 3. Intraoperative views of second step surgery. Observed in images a and b note the segmental bone defect after stem insertion. In images c and d note the reconstruction with cortical and morcellized allograft, in conjunction with a cable plate.
collapse—the tip of the latter protruding from the femoral trochlea (Fig. 4).

The acute phase reactant values were normal; reactivation of the infection was therefore excluded. Revision of the femoral component was decided. In view of the scant “native” distal femur available for correct anchoring of another diaphyseal stem, and the doubts raised about possible bone reconstruction (impacted graft technique) because of the limited biological potential of this patient with OI, we chose a more “mechanical” solution in the form of a Mega C intramedullary total femoral prosthesis (Waldemar LINK GmbH, Hamburg, Germany). During surgery, complete loosening of the femoral component was confirmed and 5 intraoperative cultures and histologic samples were obtained. We were able to remove it without the need for instruments and noted important bone sclerosis, with incorporation of the allograft. Using an independent double route (posterolateral in the hip and anterior in the knee), we prepared both joints—hip and knee—for placement of the megaprosthesis (Fig. 5a and b). Following extraction of the previous implant and cementing of the tibial component with PALACOS R® (Heraeus Medical GmbH, Wehrheim, Germany), we placed the new femoral component from distal to proximal, with prior cementing. After checking fixation of the implant in the diaphyseal and distal zone, we adjusted femoral neck length and version in the proximal segment of the femoral megaprosthesis, from the proximal approach. The postoperative course was uneventful, with negative cultures. Loading with crutches was allowed 48 hours after surgery.

The patient has undergone regular follow-up without complications. On the last visit, 2 years after the last operation, the patient was seen to be able to walk with the help of 2 crutches, without pain, and with a normal operated knee and hip joint range. The radiographic control showed good function of the right femoral reconstruction, without complications (Fig. 5c-e).

Discussion

The prolongation of life expectancy among patients with OI has resulted in an increased incidence of hip and knee osteoarthritis in these individuals.

The causes of osteoarthritis in these patients include intra-articular fractures (usually of a subclinical nature), ligament laxity, and deformities because of defective consolidation of the pathological fractures observed in these patients. In this regard, distortion of the pelvic, acetabular, and femoral anatomy is particularly important in such individuals [6]. Blackley et al. [7] reported a 29% acetabular protrusion rate in patients with OI, vs 66% according to Krishnan et al. [5]. The pelvis is often reduced and triradial in appearance, exhibiting a “shepherd crook” deformity [8]. The acetabular and femoral deformation, together with the bone weakness and high risk of intraoperative fractures constitute a genuine surgical challenge. This was evidenced with the first prosthesis, where after months of defunctionalization, associated to the background bone disease, the bone was seen to rupture throughout the proximal zone during reconstruction—hence the need to use the plate, graft, and proximal cerclage. The increase in primary arthroplasties in these patients, and the greater incidence of complications, lead to increasingly complex revision surgeries in young patients, because of the important bone loss, deformities, or sequelae of previous operations [8].
Papagelopoulos et al. [4] published the first series of patients with OI subjected to primary arthroplasty with a follow-up period of 7 years. In their series of 5 hips and 2 knees, all the patients experienced lessening of the pain, and only one reoperation proved necessary because of acetabular protrusion. Krishnan et al. [5], in a recent series, recorded a greater revision rate (5 of 6 patients),

Figure 5. Intraoperative and postoperative views after replacement of the femoral component. Note stem extraction without signs of integration (a); trochlear bone defect due to stem protrusion (b); full-length weightbearing x-ray (c) and lateral (d) view 2 years after surgery with no signs of fracture, subsidence or loosening; clinical image of patient (e). He can walk with the help of two crutches.
mainly as a consequence of acetabular protrusion with an important associated acetabular cavity defect. In our case, there were no complications at acetabular level. The absence of previous acetabular surgery/fracture, and the use of a trabecular metal implant, which has been shown to offer superior results in bone deficient cases (as in radionecrosis [9]), may explain the absence of complications.

Consensus is lacking regarding the most appropriate type of implant at femoral level. Papagelopoulos et al. [4] obtained good results with cemented implants—only one requiring removal because of infection. Krishnan et al. [5] only used uncemented stems in primary surgery (2 customized implants) and recorded a 50% incidence of intraoperative fractures. However, these authors suggested the use of customized stems to reduce this problem and ensure better adaptation to the patient anatomy. These data illustrate the difficulty of achieving primary biological fixation in patients withOI. The problems in achieving adequate fixation of the femoral component are further increased in cases of femoral revision surgery, where bone deformity, sclerosis, and the absence of metaphyseal bone complicate adequate interdigitation of the cement layer. In most cases, biological anchoring stems are required, and uncertainty remains as to the outcomes and the possible biological limitations of these patients.

In the reconstruction of long segmental defects, the options are limited to replacing the lost bone with metal or graft-prosthesis compositions. There is concern about infections in the reconstruction of fractures around the graft-host interface, pseudoarthrosis, and early graft resorption. Blackley et al. [7] recorded success (increase Harris Hip score more than 20 points) in 78% after 9 years in 63 patients treated with graft-prosthesis compositions, performing stepped osteotomy and reinforcing the graft-host interface with allografts. None of their patients presented OI, however.

In the only case where this technique was performed in a patient with OI, Ramaswamy et al. [6] used a modification of the graft-prosthesis composition procedure, placing the latter bilaterally in a 53-year-old patient with OI and hyperplastic calluses in both femurs. This allowed insertion of the entire assembly within the canal, followed by fixation with multiple screws. Graft consolidation was observed after 20 months, with a follow-up period of 10 years. In our case, failure in achieving integration of the previous implant and the older age of the patient raised doubts about the advisability of this surgical technique. Furthermore, the presence of femoral trochanter damage secondary to stem collapse (Fig. 5b) made prosthetic knee arthroplasty necessary. The use of a graft-prosthesis technique might have caused a concentration of forces between the allograft and the implant in the distal femur, with a high risk of peri-implant fracture. On the other hand, the previous reconstruction with a diaphyseal fixation prosthesis plus allograft plus plate, which only functioned for the duration of supplementary treatment with Teriparatide, led us to consider this as representing the biological limit of the patient bone for repair purposes. We therefore chose a reconstruction strategy based mainly on mechanical principles [10]. The use of an intramedullary femoral component ensures adequate support. Incorporation of the trabecular metal acetabular insert had been achieved, and cementing of the tibial component in an intact bed ensured distal stability. Furthermore, maintenance of the bone structure makes it possible to use a conventional surgical approach to the knee and hip, avoiding an extensive transfemoral route, with the consequent decrease in morbidity-mortality. Likewise, postoperative function is improved, as the muscle insertions are respected; constrained implants at acetabular level are not required; and there is a lower risk of infection, as there is less effective joint space than in cases of total femoral arthroplasty [11].

Summary

Joint arthroplasty is a potential option for the management of osteoarthritis in patients with OI. However, the prosthetic revision rate is especially high in such individuals, particularly in the presence of acetabular protrusion. At femoral level, the presence of bone deformities, as well as femoral osteoporosis with a tendency to fracture, increase the probability that revision surgery will be needed. In such cases, it is particularly complicated to achieve primary stability of the femoral component, and there is concern about the viability of the graft-prosthesis technique, because of the biological limitations that characterize these patients. The use of an implant with femoral arthroplasty reduces the concerns about fixation, although it adds other potential problems such as infection, lesser function, and the possibility of tibial fractures. The utilization of an intramedullary component could lessen some of these concerns, such as infection and function, and does not affect the risk of failure at tibial level.

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