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Case report

Custom hemiarthroplasties for retention of existing hardware associated with osteogenesis imperfecta

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ABSTRACT

Osteogenesis imperfecta is a rare genetic disorder that presents with heterogeneous phenotypes ranging from brittle bones to impaired hearing. Because of the decreased bone mineral density frequently observed in this patient population, many patients experience recurring and long-term fractures, which often require orthopaedic management. With the advancement of nonsurgical and surgical management and increased longevity of patients with osteogenesis imperfecta, the incidence of osteoarthritis has risen, presenting new orthopaedic challenges. However, compromised bone integrity and size combined with frequent existing hardware render traditional surgical therapies for osteoarthritis technically challenging in this patient population. In this report, we present a case in which we retained a portion of the patient's existing hardware, while performing staged bilateral custom hemiarthroplasties in a patient with osteogenesis imperfecta.

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Introduction

Osteogenesis imperfecta is a rare genetic disorder of type I collagen, which results in a wide range of clinical symptoms. This often manifests as decreased bone mineral density, short stature, skeletal deformity, and an increased susceptibility to mild trauma, which can lead to frequent and recurring fractures [1]. One orthopaedic treatment option for children with osteogenesis imperfecta is the insertion of telescoping rods. These rods expand as the bone grows, thus providing additional structural support and reducing the need for multiple orthopaedic surgeries [2-4]. In the past few decades, improvements in care such as these have led to increased life spans for this patient population [5]. However, it has also been correlated with an increased incidence of osteoarthritis, with most affected adults reporting either an established diagnosis of arthritis or arthritis-like symptoms [6-8]. Because of the nature of this

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patient population's bone fragility, size and frequent existing hardware, traditional surgical therapies for osteoarthritis are technically challenging. Patient-specific and custom implants provide a unique approach to specifically accommodate an individual's anatomy [9-12]. Here, we present a case in which we retained a portion of the patient's existing hardware, while performing staged bilateral hemiarthroplasties in a patient with osteogenesis imperfecta using custom-designed femoral components.

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Case history

A 36-year-old female with a history of osteogenesis imperfecta type II presented with a nonunion of a right femoral neck fracture (Fig. 1a), which was sustained during a motor vehicle accident 6 months prior. Her medical history reveals several fractures and a multitude of previous orthopaedic procedures including the placement of Bailey Telescoping Rods (Zimmer, Warsaw, IN) in both of her femurs. On physical examination, the patient experienced severe pain with any internal and/or external rotation, and forward flexion of the hip was limited to 80°. Radiographs showed significant degenerative changes, and she experienced progressive hip pain from the long-term fracture, eventually being unable to ambulate and having difficulty transferring independently.

The Bailey Telescoping Rod comprises of 3 components: an obturator pin fixed to either the proximal or distal region of the

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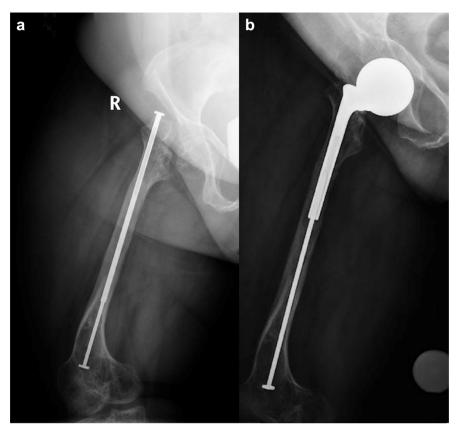


Figure 1. Anteroposterior views of the presurgery nonunion of the right femoral neck (a) and postsurgery custom fit right hemiarthroplasty, which cannulates the existing telescoping rod (b).

bone, a sleeve fit over the obturator pin, and a T-piece screwed on the sleeve to fit over the opposite end of the bone. Removing the Bailey Telescoping Rod posted 2 problems: (1) The rod was providing structural support to mitigate further risk of femoral fracture and (2) Removal of the rod would require an additional incision at the knee and potential damage to the joint. Because of these issues, we opted to retain the obturator pin (located distally on both femurs) and design custom cannulated femoral stems (Biomet, Warsaw, IN), which would be fit over the obturator pins. The stems and internal cannula were designed for cemented use because of the patient's underlying condition and to help with engaging the obturator pin and stem cannula (Fig. 2).

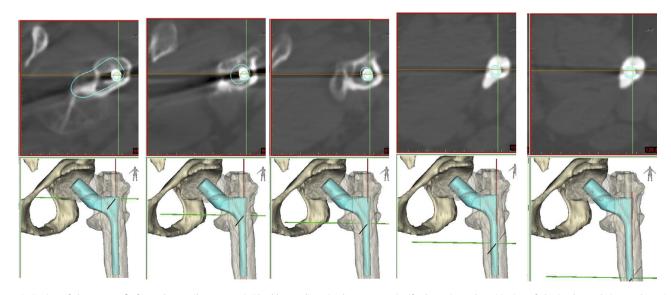


Figure 2. Design of the custom fit femoral stem (bottom row). The blue outlines in the top row signify the estimated positioning of the implant relative to the existing telescoping rod.

At this point, informed consent was obtained from the patient for the partial removal of hardware, followed by implantation of a custom fit total hip arthroplasty. The T-piece and sleeve assembly was removed without difficulty following a posterolateral approach. Trephines were used to make room for the distal stem, followed by a custom broach to prepare the canal for each implant. Fluoroscopy was used to confirm the engagement between the femoral stem cannula and the obturator pin. Simplex cement loaded with tobramycin was vacuum mixed and injected via a syringe into the femoral canal to hold the components in place. After the femoral component was completed, we determined that the acetabulum was too small to accommodate an acetabular component, and thus we continued with a hemiarthroplasty instead. We used a 32 mm-3 mm cobalt chrome head as the proximal femoral component, which provided adequate stability and tissue tension. As the original plan was to perform a total hip arthroplasty with a custom 22 mm stainless steel head, we did not have a 32 mm stainless steel femoral head and out of necessity used a cobalt head instead. As early as the 1-month follow-up postsurgery and now 5 years postsurgery, the patient remains ambulatory without any major issues. Thus, the procedure effectively resulted in a relief of her pain, and radiographs confirmed the successful positioning of the implant (Fig. 1b).

Four years after the initial custom fit right hemiarthroplasty, the patient presented with significant pain and degenerative changes in the contralateral hip (Fig. 3a). The previous custom fit right hemiarthroplasty was tolerated extremely well and remained stable. Thus, we performed a similar procedure on the left hip. We used the same design for the custom implant (Biomet), which again had a hollow femoral stem and allowed for the cannulation of the existing distal rod. After removing a proximal portion of the telescoping rod, we used custom broach-bodies to prepare the femoral canal and were able to cannulate the distal rod with the custom femoral stem without difficulty (Fig. 3b). Fluoroscopic imaging was used to confirm the reduction and cannulation. Using our experience with the first hemiarthroplasty, we prepared a custom 32 mm stainless steel head for the second side.

The patient has been followed up regularly since both surgeries. She uses a walker at home and is able to transfer on her own without issues. Five years after the initial custom fit right hemiarthroplasty and 1 year after the contralateral custom fit left hemiarthroplasty, the patient maintains functional range of motion in both hips. In addition, she has experienced a significant relief in pain and her radiographs continue to show good positioning of the bilateral hemiarthroplasties.

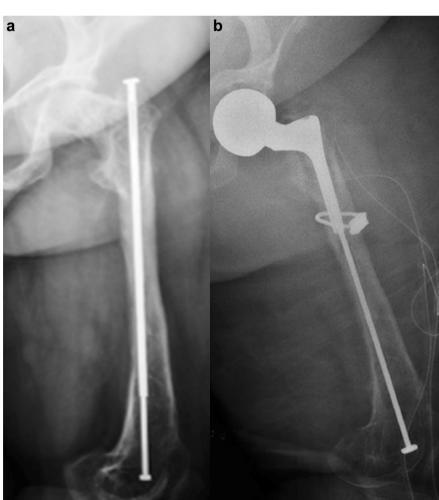
Because of many recent advances in the care of patients with

osteogenesis imperfecta, life expectancy and the incidence of

Discussion

b a

Figure 3. Anteroposterior views of the presurgery degeneration of the left hip joint (a) and postsurgery custom fit left hemiarthroplasty, which cannulates the existing telescoping rod, with a prophylactic cerclage cable (b).



osteoarthritis have increased among affected individuals. Standard surgical techniques in the treatment of osteoarthritis can be technically challenging in this patient population because of the nature of their bone integrity and fragility. Extra care and preoperative planning has been effective when using custom components [10] and standard components [7,13,14] in the treatment of end-stage osteoarthritis in patients with osteogenesis imperfecta. However, no case has been described having been performed in the setting of existing hardware.

The existence of hardware from previous orthopaedic procedures poses an additional technical challenge in this specific patient population. Traditionally, existing hardware is removed before performing a total hip arthroplasty. However, the removal of hardware poses its own risks including pain, creation of stress risers, and refracture [15,16]. Thus, several methods have been proposed to allow for the retention of hardware in cases of joint arthroplasty. Mont et al. [15] described the successful use of a short, neck-preserving femoral stem in a total hip arthroplasty to retain a previously implanted retrograde femoral nail. Goosen and Van Hellemondt [17] described the successful removal of a proximal segment and retention of the distal aspect of an existing intramedullary femoral nail in a primary total hip arthroplasty. However, neither case was performed in a patient with osteogenesis imperfecta or compromised bone.

Our use of custom fit implants in the current case proved effective for the retention of existing hardware in a particularly atrisk patient population. Designed with a hollow center stem, our implant allows for the cannulation of the obturator pin, preventing the necessity of its complete removal, which would have been technically difficult and left the patient susceptible to refracture. This unique approach is advantageous because it offers the benefit of a traditional arthroplasty, while simultaneously maintaining the stability previously established by the distal growing rod. In addition, a significant volume of host bone can be retained rather than removed, as is the case in the removal of the telescoping rod from the femoral canal. Considering the compromised bone integrity and fragility of this specific patient population, we believe it is beneficial to preserve as much existing bone as possible. An additional consideration was that by not removing the distal hardware, the integrity of the knees could be preserved.

The regulatory pathway for these implants falls under the Custom Device Exemption, last amended on July 2012 FDASIA Section 520(b). This exemption allows a device without 510k or premarket approval to be used to treat a sufficiently rare condition such that conducting clinical investigations on such a device would be impractical. The exemption is limited to no more than 5 units per year of a particular device type for each manufacturer, although bilateral cases such as this case report would fall under 1 unit [18].

Summary

To our knowledge, we present the first case illustrating that joint arthroplasty can be performed with the retention of existing hardware in a patient with osteogenesis imperfecta. The custom fit design we presented provides therapeutic relief in the setting of existing telescoping rods, and thus offers a unique solution to the complex treatment of this patient population.

Acknowledgments

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