Hip Arthroplasty in Patients with Osteogenesis Imperfecta

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Abstract

» Osteogenesis imperfecta (OI) is a rare congenital disorder that affects connective tissue.

» Modern medicine has mitigated the mortality that is associated with OI, allowing patients to live a near-normal life span.

» The degenerative process in OI is probably accelerated because of subclinical intra-articular fractures, joint laxity, and distorted femoral and acetabular anatomy.

» Total hip arthroplasty is seldom performed in patients with OI; it is technically difficult due to bone fragility, deformity, soft-tissue alteration, acetabular protrusion, the risk of intraoperative and postoperative fractures, and joint laxity.

» This review highlights that patients with OI need hip arthroplasty procedures at an early age and that early revision surgery can be expected. New-generation uncemented implants may improve implant survivorship.

Osteogenesis imperfecta (OI) is a rare congenital disorder that affects connective tissue. It is also known as "brittle bone disease."[1] It presents in 1 in 10,000 to 20,000 live births, with a genetic alteration that results in abnormal collagen structure, ossification, and mineralization. This results in various orthopaedic manifestations and inherent coagulation defects due to abnormal collagen within the endothelial capillary bed[2,3].

The described orthopaedic manifestations are growth retardation[4], reduced bone mass[1], acetabular protrusion (observed in 29% of patients), triradiate pelvis, joint laxity, osteoporosis, fragile bones, malunion, nonunion, and limb deformities such as a shepherd-crook's deformity[5], coxa vara, and limb-length discrepancies[4]. As a result of modern medical intervention, patients with OI have a near-normal life span, which has increased the incidence of osteoarthritis[6]. The joint laxity and subclinical insufficiency fractures with secondary joint degeneration lead to hip and knee osteoarthrosis that require joint arthroplasty[2].

Total hip arthroplasty (THA) in these patients is known to be technically demanding and challenging. Preoperative planning is mandatory for joint reconstruction to mitigate intraoperative complications. The technical difficulty is created by bone fragility, deformity, soft-tissue alteration, acetabular protrusion, the risk of intraoperative and postoperative fractures, and joint laxity. Primary procedures are usually performed in younger patients, and subsequent earlier revision surgery can be expected. Patients with OI have postoperative complications,

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Fig. 1
A preoperative pelvic anteroposterior radiograph demonstrating degenerative changes in the bilateral hip joints, with severe acetabular protrusion, a triradiate pelvis, nonunion of the right femoral neck, and hardware from a previous spinal fusion.

including hypertrophic callus and heterotopic ossification[3]. Prior to the intervention of hip arthroplasty, these patients tended to be severely disabled and have low Harris hip scores[6].

Case Report

A 29-year-old woman with OI sustained a fracture of the neck of the right femur 3 years ago. The patient was informed that information regarding her case would be submitted for publication, and she consented. At that time, she was advised that the fracture could not be treated operatively because of her abnormal anatomy, and she was treated nonoperatively. She presented to us with persistent pain and progressive difficulty in walking (Fig. 1).

We performed a THA to improve the pain and mobility. Technical problems included an acetabulum that was anteverted at 80° and a 140° backward bend of the femoral neck. A fully porous titanium acetabular cup with variable-angle locking screws was used to achieve the best osseous fixation. A polyethylene liner was cemented into the acetabular cup to obtain the best orientation. The acetabular cup inclination was approximately 80°, but this was improved to about 48° by cementing the polyethylene liner into the metal backing at the

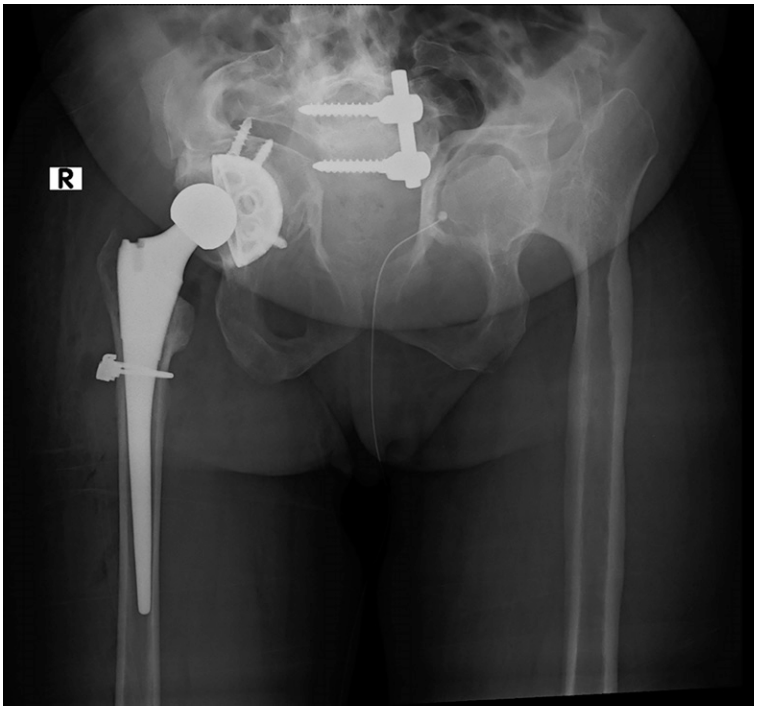

Fig. 2
A postoperative pelvic anteroposterior radiograph demonstrating the right total hip replacement. There is a prophylactic cable tie around the proximal aspect of the femur.

TABLE I Primary Hip Arthroplasty Performed in Patients with OI*

Article	Age *(yr)*	Indication	Acetabular Component	Femoral Component
Ramaswamy et al.[5]	54 (left) 41 (right)	Osteoarthritis	Left: impaction bone-grafting and uncemented cup Right: uncemented cup	Left: femoral allograft and cemented stem Right: uncemented stem
Sanz-Ruiz et al.[2]	63	Previous septic femur with open reduction internal fixation	Uncemented high-friction porous cup augmented with screws	Uncemented revision stem, structural allograft, and stabilization with a plate and trochanteric cerclage wires
Chaus and Heare[14]	16	Osteoarthritis	Uncemented cup	Uncemented stem
Liporace et al.[3]	47	Bicolumnar acetabular fracture	Impaction bone-grafting and uncemented cup augmented with screws	Cemented stem
Papagelopoulos and Morrey[6]	60	Osteoarthritis	Cemented cup	Cemented stem
	61	Osteoarthritis	Cemented cup	Cemented stem
	62	Fracture of the neck of the femur	Cemented cup	Cemented stem
	62	Osteoarthritis	Cemented cup	Cemented stem
Krishnan et al.[8]	NM	NM	Uncemented porous-coated cup augmented with screws	Uncemented stem with cabling
	NM	NM	Uncemented porous-coated cup augmented with screws and impaction bone-grafting	Uncemented stem with cabling
	NM	NM	Uncemented high-friction porous cup augmented with screws	Uncemented CAD-CAM stem with cabling
	NM	NM	Uncemented cup augmented with screws	Uncemented revision stem
Kermeci and Weinrauch[15]	46	Osteonecrosis	Cemented acetabulum	Cemented stem
Nishida et al.[10]	36 (right) 40 (Left)	Right: femoral neck fracture nonunion Left: osteoarthritis	Right: it was determined intraoperatively that the acetabulum was too small to accommodate an acetabular component, and a hemarthroplasty was performed Left: no intervention	Right: custom cemented stem Left: custom cemented stem
Ajlouni et al.[7]	14	Acetabular protrusion	Müller acetabulum reinforcement ring and cemented cup	Uncemented stem
Maeder et al.[13]	52	Femoral neck fracture	No intervention	Cemented stem
Carlson et al.[9]	60	Acetabular protrusion	No intervention	Cemented stem
	50	Osteoarthritis	Uncemented cup	Cemented stem
	61	Osteoarthritis	Cemented cup	Cemented stem
	58	Osteoarthritis	Cemented cup	Cemented stem
	63	Osteoarthritis	Cemented cup	Cemented stem
	66	Osteoarthritis	Cemented cup	Cemented stem
	57	Osteoarthritis	Uncemented cup	Uncemented stem
	38	Osteonecrosis	Uncemented cup	Cemented stem
	45	Osteoarthritis	Uncemented cup	Cemented stem
	64	Osteoarthritis	Uncemented cup	Cemented stem
	64	Osteoarthritis	Uncemented cup	Cemented stem
	26	Osteoarthritis	Uncemented cup	Cemented stem
	35	Left hip pain after hip arthrodesis	Uncemented cup	Uncemented stem

*OI = osteogenesis imperfecta, NM = not mentioned, and CAD-CAM = computer-aided design and computer-aided manufacturing.

optimal orientation. An uncemented stem was utilized with appropriate anteversion, ignoring the native anatomy of the proximal aspect of the femur. The impinging osseous anatomy (the extended greater trochanter) was trimmed down to ensure ongoing stability (Fig. 2). The reduction was stable intraoperatively. Postoperatively, the patient was able to mobilize comfortably. By 6 weeks, she was able to walk with only 1 crutch, and she walked unaided by 9 weeks. The patient reported no residual pain. Clinically, she had no fixed flexion deformity; she had flexion to 100°, internal rotation of 10°, external rotation of 20°, and abduction/adduction of 30°.

To our knowledge, this is the first report of a new-generation high-friction porous acetabular component with a polyethylene liner that was cemented into the acetabular shell in the patient population with OI. This allowed optimal acetabular orientation.

Material and Methods

A systematic review of the literature was performed to identify any studies that reported cases of hip arthroplasty in patients with OI. A search of the PubMed and Google Scholar databases was conducted with the following keywords: "osteogenesis imperfecta," "hip arthroplasty," and "hip replacement." We did not set any limitations regarding the year of publication. A total of 778 results were found. The title and the abstract of all of the articles were reviewed. The last search was performed in July 2020. Case report data were identified for 12 articles.

Discussion

A review of the English-language literature revealed only 31 previously reported cases in which patients with OI needed primary hip arthroplasty (Table I) and 19 cases in which they needed revision hip arthroplasty (Table II). Based on the literature search, the patient in our case report is the thirty-second patient with OI to undergo primary hip arthroplasty and the fifty-first patient with OI requiring any type of arthroplasty.

Primary Arthroplasties

There are 31 recorded cases of primary hip arthroplasty in patients with OI in the literature (Table I). In 4 of these cases, the age of the patient at the time of the procedure was not recorded. In the remaining 27 procedures, the mean age was 49.7 years. That mean age is much older than our 29-year-old patient; however, as shown in Table I the youngest patient to undergo hip arthroplasty was 14 years old[7]. The indications for arthroplasty were osteoarthritis in 17 patients (54.8%), not specified in 4 patients (12.9%), fracture-related in 4 patients (12.9%), and femoral septic pseudoarthrosis in 1 patient (3.2%), acetabular protrusion in 2 patients (6.5%), a hip arthrodesis conversion to a THA in 1 patient (3.2%), and osteonecrosis in 2 patients (6.5%). Osteoarthritis is the most common indication for arthroplasty. The degenerative process in OI is probably accelerated because of subclinical intra-articular fractures, joint laxity, and distorted femoral and acetabular anatomy[8]. The anatomical distortions at the joint are subchondral insufficiency and periarticular osseous dysplasia[4]. The indication for our patient was nonunion of a femoral neck fracture.

Revision Arthroplasties

We found 19 cases of revision hip arthroplasty in patients with OI that had been recorded in the literature (Table II). In the case series by Krishnan et al., THA patients with OI underwent revision at a median of 5.2 years (range, 2.8 to 11.0 years) after the primary procedure[8]. In the case series by Carlson et al., the mean time to revision was 6.6 years (range, 4.1 to 9.4 years)[9]. This survivorship is shorter in the population with OI than in the general population.

As shown in Table II, the indications for revision included aseptic loosening in 10 cases (52.6%), acetabular protrusion in 1 case (5.3%), pain with absence of infection and loosening in 3 cases (15.8%), septic loosening in 1 case (5.3%), pelvic discontinuity in 1 case (5.3%); the indication for the remaining 3 cases was not recorded. Aseptic loosening and osteopenia result in a further reduction of bone stock in these already compromised patients, making revision surgery even more challenging[8]. These 19 revisions were mostly not linked to the primary cases that are reported in Table I, indicating that there probably are primary hip arthroplasties and revision surgeries that have not been reported.

Acetabular Components

In the primary cases, 17 acetabular components were uncemented and 10 were cemented. There were 4 hemiarthroplasties performed.

In the revision cases, there was a single cemented acetabular revision, 7 cases had no acetabular-sided intervention (it was not reported whether the liners were exchanged in the THA), a single cup removal was part of a Girdlestone procedure, there were 2 liner exchanges, and 8 cases had uncemented cups. This illustrates the use of modern components with either porous-coated or high-friction porous cups that were augmented with either impaction bone-grafting or screws for improved fixation. With acetabular revision for reconstructing large segmental defects, the options are limited to substitution of the missing bone with metal or a bone graft, but there are concerns regarding both nonunion and graft resorption[5]. To our knowledge, this is the first report of a new-generation high-friction porous acetabular component with a polyethylene liner that was cemented into the acetabular shell. This allowed optimum acetabular orientation. The ability to optimally align the component within the available bone stock should aid in optimizing outcomes.

The use of custom acetabular cups is applicable in the population with OI. Nishida et al. abandoned THA and converted to a hemiarthroplasty during surgery because the acetabular anatomy

TABLE II Revision Hip Arthroplasty Performed in Patients with OI*

Article	Age *(yr)*	Indication	Acetabular Component	Femoral Component	Time to Revision *(yr)*
Ramaswamy et al.[5]	NM	Aseptic loosening	Right: no intervention	Right: femoral allograft and cemented stem	16
Carlson et al.[9]	60	The acetabular component continued to migrate medially and resulted in pelvic discontinuity	Girdlestone and impaction bone-grafting	Removed	6.6
	66	Aseptic loosening	No intervention	Cemented stem revised to an uncemented stem	6.3
	26	Aseptic loosening	No intervention	Cemented stem revised to an uncemented stem	4.1
	45	Aseptic loosening	Polyethylene liner was revised to a lipped liner with retention of the acetabular component	Cemented stem revised to an uncemented stem	9.4
Sanz-Ruiz et al.[2]	65	Aseptic loosening with progressive femoral collapse with protrusion through the femoral trochlea	No intervention	Cemented intramedullary total femoral prosthesis	2.6
Schoof et al.[12]	62	Aseptic loosening	Polyethylene liner was revised to a lipped liner with retention of the acetabular component	Cemented stem revised to an uncemented long revision stem along with an osteotomy to correct femoral deformity	8
Papagelopoulos and Morrey[6]	66	Acetabular protrusion	No intervention	Removed	6
	NM		No intervention	Thompson hemiarthroplasty (cemented)	
			No intervention	Austin-Moore hemiarthroplasty (uncemented)	
			Cemented cup	Cemented stem	
Krishnan et al.[8]	NM	Septic loosening of the acetabulum and femur × 1	Uncemented porous-coated cup, impaction bone-grafting and augmented with screws	Cemented stem	
		Aseptic loosening of the acetabulum and femur × 1	Uncemented porous-coated cup augmented with screws	Uncemented CAD-CAM stem with cabling	
		Aseptic loosening of the acetabulum × 3	Uncemented porous-coated cup augmented with screws	Uncemented stem	
		Pain with no evidence of infection and loosening × 3	Uncemented porous-coated cup, impaction bone-grafting and augmented with screws	No intervention	
			Uncemented porous-coated cup, impaction bone-grafting and augmented with screws	Uncemented CAD-CAM stem	
			Uncemented high-friction porous cup augmented with screws	No intervention	
			Uncemented porous-coated cup augmented with screws	No intervention	
			Uncemented porous-coated cup augmented with screws	No intervention	

*OI = osteogenesis imperfecta, NM = not mentioned, and CAD-CAM = computer-aided design and computer-aided manufacturing.

was too small to seat a cup[10]. Custom-designed acetabular cups based on computed tomography have been shown to have superior accuracy in the setting of skeletal dysplasia, which presents with distorted acetabular anatomy[11].

A transverse fracture of the acetabulum has been reported on cup impaction and managed with a multi-hole revision cup and screws[8]. Acetabular protrusion is common and results in superomedial migration of the hip center; the aim of surgery should be to lateralize the hip center[4]. There are high revision rates in cases where there has been preoperative acetabular protrusion; the population with OI has higher complication rates compared with the general population[8]. Reconstruction of the acetabulum can be achieved with femoral-head allograft and autograft[5,6]. The underlying collagen disorder, even after operative intervention, predisposes the patient to recurrence of the acetabular protrusion due to the poor quality of the bone[6]. Papagelopoulos and Morrey presented a case of acetabular protrusion after a hemiarthroplasty that was managed with resection arthroplasty[6]. In the same series, a patient had hemiarthroplasty revision twice before undergoing conversion to a cemented THA, highlighting the limited role of hemiarthroplasty in patients with OI.

Gross medial acetabular defects, especially in the revision setting, require reconstruction with a cup-cage construct or a large new-generation high-friction porous cup that is augmented with screws and bone graft[8].

Femoral Components

There is no consistency regarding femoral component selection[2]. In the reported primary cases, 21 femoral components were cemented and 10 were uncemented. Four femoral prostheses were cemented in the revision cases, and there was a single cemented intramedullary total femoral prosthesis. In 2 of the revision cases, the femoral component was removed, and there was no femoral intervention in 4 cases. There were a total of 8 uncemented femoral components.

With revision surgery due to sclerosis, deformity and metaphyseal bone loss prevent adequate interdigitation of the cement mantle, suggesting a role for uncemented revision components[2]. However, the use of cemented femoral stems in patients with poor bone stock has been suggested[5]. Papagelopoulos and Morrey reported good results with cemented components, with only a single case of infection[6]. In their case series, Carlson et al. described 3 cases of cemented stems that were successfully revised to uncemented stems[9].

Mandatory preoperative planning and templating is required as patients can have abnormally small and narrow femoral canals and acetabular bone deficiencies[6]. Computer-aided design and computer-aided manufacturing (CAD-CAM) components assist with the abnormal and distorted anatomy of patients with OI because standard components can be inadequate for some patients, especially in the setting of narrow canals and thin cortices[8]. Nishida et al. demonstrated the use of custom implants in the setting of existing hardware[10]. The use of intraoperative fluoroscopy is an adjunct to aid the surgeon in the setting of a pseudocanal[12]. Corrective valgus and shortening osteotomies have been reported to correct the distorted femoral anatomy in order to allow for femoral component implantation[12,13].

The underlying poor quality of the bone in the postoperative setting may lead to femoral stem subsidence[6]. To mitigate against subsidence, the use of cemented femoral stems has been advocated[4]. Iatrogenic femoral fractures are common and sometimes require the use of cerclage cabling[8]. The intraoperative fracture rate with primary hip arthroplasty in patients with OI is as high as 50%[8].

In the revision setting, poor residual bone stock can be addressed with a femoral allograft[5]. However, stress-shielding in the proximal aspect of the femur may result from use of this allograft-prosthesis structural graft[5]. Nonunion remains a risk with the use of grafts; however, favorable union results have been reported[5]. In a patient with distorted anatomy with no diaphyseal support or in the setting of comorbid knee pathology, a total femoral replacement remains a viable option[5].

Outcomes

Despite the complexities of surgery, the literature shows favorable outcomes following hip arthroplasty. Krishnan et al. reported that the median Oxford Hip Score increased from 15 (range, 14 to 24) preoperatively to 41 (range, 37 to 46) at the time of the final follow-up[8]. In their case series, Papagelopoulos and Morrey reported good outcomes in 5 patients who underwent THA (independent ambulation, although some needed gait aids) and an improvement in the Harris hip score from a mean of 32 to a mean of 78 on follow-up[6]. Additional long-term follow-up data are required to adequately assess the outcomes of hip arthroplasty in patients with OI.

Overview

Few cases of hip arthroplasty in patients with OI have been reported in the literature, and most have only short-term follow-up. The improved medical management of patients with OI has led to an increased life span; therefore, we can expect an increase in the number of patients who will require hip arthroplasty. This review article highlights that these patients need hip arthroplasty procedures at an earlier age, and early revision is common. New-generation uncemented implants may improve implant survivorship. Our technique of achieving the best osseous fixation with a high-friction cup that is complemented by a cemented polyethylene liner to achieve the best orientation may offer an acetabular solution for these complex patients.

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