End-stage Posttraumatic Osteoarthritis Treated With THA in Osteogenesis Imperfecta

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abstract

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Osteogenesis imperfecta is an incurable genetic disorder manifested with altered bone quality that predisposes patients to a multitude of fractures throughout their lives, including acetabular fractures. The management of acetabular fractures in patients with osteogenesis imperfecta remains a challenging clinical problem, with a paucity of literature supporting treatments and their outcomes. Limited reports in the literature validate the use of total hip arthroplasty (THA) in patients with osteogenesis imperfecta, and they describe the adult population only.

This article describes a case of delayed diagnosis of a transverse acetabular fracture and femoral head impaction fracture that led to posttraumatic end-stage hip osteoarthritis in a 16-year-old boy with osteogenesis imperfecta (Sillence Type I) that was sustained after minimal trauma. Clinical examination 3 months postinjury revealed a significant pelvic obliquity, severe pain with hip range of motion, and limited hip range of motion. Imaging studies revealed a complete loss of articular cartilage and significant joint effusion.

The patient underwent THA. No postoperative complications occurred. Two-year follow-up showed an excellent clinical result. The patient’s hip was pain free, and he was able to walk with no limp.

The authors are unaware of any reported cases of children with osteogenesis imperfecta undergoing THA. Based on the reported literature and the authors’ experience, THA can be a reliable surgical option for patients with osteogenesis imperfecta.

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In healthy adolescents or adults, acetabular fractures are often treated with open reduction and internal fixation. However, the current orthopedic literature lacks data on the treatment methods and results of these injuries in patients with osteogenesis imperfecta. Darmanis and Bircher reported 2 cases of acetabular fractures successfully treated acutely with open reduction and internal fixation in a 16- and a 25-year-old patient, both with osteogenesis imperfecta. No long-term results were presented in their report.

End-stage posttraumatic osteoarthritis after acetabular or femoral head fractures are often treated with total hip arthroplasty (THA). Good-quality data support the use of THA in a multitude of disease processes in children. However, data are limited on the use of THA in adult patients with osteogenesis imperfecta, and no data exist for its use in children with this pathology. Osteogenesis imperfecta poses additional challenges to the THA procedure, including the potential for fracture intra- and postoperatively, deformity of the skeleton, protrusio acetabuli or hypertrophic callus, osteopenia, bleeding diathesis, and potentially friable tissues. To the authors’ knowledge, few studies have been published concerning THA in patients with osteogenesis imperfecta; however, these studies were in adult patients. Outcomes and clinical experience of THA for pediatric patients with osteogenesis imperfecta would benefit the medical community and osteogenesis imperfecta patients due to the complex management and specificity of pediatric arthroplasty. The authors are unaware of cases of THA described in children.

This article describes a 16-year-old boy with osteogenesis imperfecta who sustained a left-side acetabular fracture and ipsilateral femoral head impaction fracture after minimal trauma. This patient rapidly progressed to end-stage posttraumatic osteoarthritis and underwent THA to address the pain and functional loss associated with degenerative arthritis. Total hip arthroplasty is a potential treatment option for children with osteogenesis imperfecta.

### CASE REPORT

A 16-year-old boy (weight, 41.8 kg; height, 156 cm) with osteogenesis imperfecta (Sillence Type I) had sustained multiple fractures over the course of his lifetime, predominantly in his upper extremity. Medical management of his genetic disease had been undertaken by a pediatric endocrinologist since birth. Treatment had included oral bisphosphonate therapy and, most recently, 1.5 years of intravenous pamidronate therapy. He had been fracture free for >1 year.

While on vacation, the patient sustained a fall from standing height that caused immediate severe left hip pain. He presented to the emergency department at another hospital. On admission, left hip plain radiographs and an ultrasound were reported as normal. These films and reports were not available for review by the current authors. The patient reported persistent left hip pain and walked with a limp. Two weeks later, the patient presented to an orthopedic surgeon at another hospital for a follow-up and repeat radiographs, which were reported as negative for fracture or acute injury (Figure 1). The patient was subsequently diagnosed with an adductor muscle avulsion and was allowed to bear weight as tolerated on the left lower extremity.

Approximately 3 months later, the patient presented to the authors’ institution. Examination revealed a significant pelvic obliquity, severe pain with left hip range of motion, and limited hip joint range of motion. With these new findings, magnetic resonance imaging, bone scan, and a workup for an infectious process were ordered (Figure 2). White blood cell count was 4.4×10³ µL (normal range, 4-10.5), erythrocyte sedimentation rate was 7 mm/hr (normal range, 0-15 mm/hr), and C-reactive protein was 0.2 mg/dL (normal range, 0-1 mg/dL), ruling out an infectious process. Bone scan revealed increased tracer uptake in the left acetabulum and femoral head (arrow) consistent with a healing fracture (A). Coronal T1 and coronal short T1 inversion recovery magnetic resonance image showing marked cartilage loss involving the left femoral head and acetabulum and a significant joint effusion (B).
On reexamination by the senior author (T.H.), the patient demonstrated significantly decreased left hip range of motion, pelvic obliquity on examination, and a new onset adduction contracture of his left hip. Repeat left hip radiographs revealed periosteal healing consistent with a previous acetabular fracture and a femoral head impaction fracture that had subsequently healed; however, the patient had progressed to rapid posttraumatic osteoarthritis (Figure 3). A retrospective examination of the 2-week postinjury images demonstrated callus formation medial to the acetabulum, indicating a healing fracture. Retrospectively, the initial injury was classified as a Salter-Harris Type 1 fracture vs a transverse acetabular fracture in favor of the latter, because it appeared that the triradiate cartilage had already closed on the contralateral side. Computed tomography scan of his hip joint evaluated the acetabular bone stock (Figure 4) and confirmed the healed transverse acetabular fracture, complete loss of cartilage in the hip joint, and abnormal contour of the femoral head. The patient was presented with 2 surgical options: hip arthrodesis and THA. After discussion of the risks, benefits, and alternatives of both procedures, the family elected to proceed with THA.

A Kocher-Langenbeck approach was used to access the hip joint, which demonstrated significant arthritic changes with a large area of cartilage loss consistent with the femoral head impaction fracture (Figure 5). A 48-mm ASR acetabular cup (DePuy, Warsaw, Indiana) was placed, followed by an S-ROM 14 B femoral component (DePuy), which was fit with a 30-mm standard neck with a +4 lateral offset and a 43-mm femoral head. The femoral and acetabular components were press fit successfully (Figure 6A). Total blood loss was 400 mL. The patient was allowed to bear weight as tolerated postoperatively, and the postoperative period was uneventful.

One month postoperatively, the patient had near complete pain relief, was ambulating with no gait aid, had a clinically equal leg length, and reported no surgical wound complications. Radiographs at 1-month follow-up showed well-seated components with no evidence of loosening (Figure 6B). At 2-year follow-up, the patient continues to show an excellent clinical result. The hip is pain free, and the patient walks with no limp. Radiographs show no evidence of prosthesis failure or loosening.

**DISCUSSION**

Acetabular fractures are uncommon in children. Watts\(^5\) reported that 10 pelvic fractures could be expected per year in a large children’s hospital, that 97%
would be stable, and that acetabular
fractures were rare. Acetabular fractures ac-
count for 0.8% to 15% of pediatric pelvic
fractures. Osteogenesis imperfecta is
an incurable genetic disorder manifested
with altered bone quality, predisposing
those affected to a multitude of fractures
throughout their lives, including acetabu-
lar fractures. The management of acetabu-
lar fractures in patients with osteogen-
esis imperfecta remains difficult, with a
paucity of literature supporting treatments and
their outcomes.

Osteogenesis imperfecta is a heritable
syndrome that results in ≥1 of the follow-
ing features: osteoporosis predisposing pa-
tients to fractures, defective dentition, blue
sclera, hearing loss, and hyperlaxity. Vari-
ability exists in the presence and severity of
the phenotypes of patients with osteogen-
esis imperfecta. Patients with this disease
often die during the newborn period. Pa-
pagelopoulous and Morrey described a
patient with osteogenesis imperfecta with
numerous deformities who underwent re-
section arthroplasty after significant intra-
pelvic protrusion by the component. Chi-
dren with a more severe phenotype pose a
more difficult treatment dilemma.

A thorough analysis of the available
literature concerning THA in patients with
osteogenesis imperfecta elicited 2 stud-
ies that discuss this treatment in the adult
population. Papagelopoulous and Morrey reported a series of 5 THAs and 3 total
knee replacements in 6 patients with os-
teogenesis imperfecta. All patients in this
study had reliable pain relief and were able
to ambulate with the use of gait aids. No
evidence existed of component loosening,
with the exception of 1 case of intrapelvic
protrusion of the acetabular component. Ramaswamy et al reported a case of a
54-year-old man with osteogenesis im-
perfecta who was successfully treated
with bilateral THA. Both of these studies
emphasized the increased complexity of
this procedure in this patient population
due to the anatomy of the pelvis and gross
distortion of the femur and acetabulum.

Patients with osteogenesis imperfecta have
also been reported as having protrusion ac-
etabuli or various deformities of the femur,
further complicating the surgical proce-
dure. Despite this array of complexities,
the 2 available studies have shown satis-
factory results after THA in patients with
osteogenesis imperfecta.

**CONCLUSION**

Total hip arthroplasty is a reliable option
for pain relief and improved function in pa-
tients with osteogenesis imperfecta that has
progressed to end-stage osteoarthritis. More
data are needed, specifically long-term fol-
low-up data in children with osteogenesis
imperfecta who have undergone THA.

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