Total Knee Arthroplasty With Concurrent Femoral and Tibial Osteotomies in Osteogenesis Imperfecta

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Abstract

Three total knee arthroplasties (TKA) with concurrent femoral and/or tibial osteotomies in 2 patients with osteogenesis imperfecta were performed from 2004 to 2009. The 2 patients were followed for a mean of 6 years. One patient with concurrent TKA, and femoral and tibial osteotomies developed a nonunion of the tibial site that responded to open reduction and internal fixation with iliac crest bone graft. The second patient underwent right TKA with bi-level tibial osteotomies, which healed uneventfully, allowing pain free, unassisted ambulation. The same patient then elected to undergo left TKA with bi-level tibial osteotomies. Intraoperatively he sustained a minor tibial plateau fracture requiring the use of a stemmed component and postoperatively, he developed a nonunion at the proximal site and valgus malunion of the distal site. Revision of fixation was performed at both osteotomy sites, and both healed within 3 months. Both patients are now pain free and ambulate without assistance.

Osteogenesis imperfecta (OI) is a rare autosomal dominant disorder with variable expressivity occurring in approximately 1/20,000.[1] This disorder involves structural or quantitative defects in type I collagen resulting in fragile bones often accompanied by sensorineural hearing loss, abnormal dentin, and blue sclera.[1-3] Patients with OI represent a broad spectrum of severity, ranging from lethality in utero to milder forms where diagnosis may be equivocal in adults. Classic findings are characterized most notably by recurrent fractures, generalized osteopenia, limb deformities, and limb-length discrepancy.[1,2,4-7] Long bones characteristically demonstrate bowing due to malunions and nonunions, and narrowing of the intramedullary canal, presenting unique challenges. Adult OI patients commonly present with deformities of long bones, severe arthritis, scoliosis, and recurrent fractures.[5-8] Unfortunately, limited information exists regarding the treatment of adult patients with arthritis and significant deformity related to OI. We present 2 patients with OI who underwent bilateral total knee arthroplasties; concurrent osteotomies of the femur and/or tibia were performed in 3 extremities.

The patient provided written informed consent for print and electronic publication of the case report.

Case Report

Case 1

A 46-year-old male with a history of OI, and multiple level osteotomy of the right femur as a child, presented with mild right hip pain, bilateral knee pain, and leg length discrepancy. He was employed in a position that required standing, and required the use of a wheelchair on an increasingly frequent basis.

On physical examination, he had obvious lower extremity deformities and palpable osteophytes along the joint line with good range of motion of the knee, bilaterally. Radiographs revealed an intramedullary retrograde Rush rod and a malunion of the right distal femur with a 27° varus deformity and "bone-on-bone" medial compartment arthritis (**Figure 1**). Secondary to prior malunions, a long gentle "S" shaped deformity existed in the tibia with the distal aspect in approximately 20° of valgus and apex anterior bow. This deformity caused a limb-length discrepancy, for which the patient wore a shoelift (**Figure 2**). Radiographs of the left knee demonstrated bone-on-bone arthritis of the lateral compartment

Figure 1. AP radiograph of both knees.

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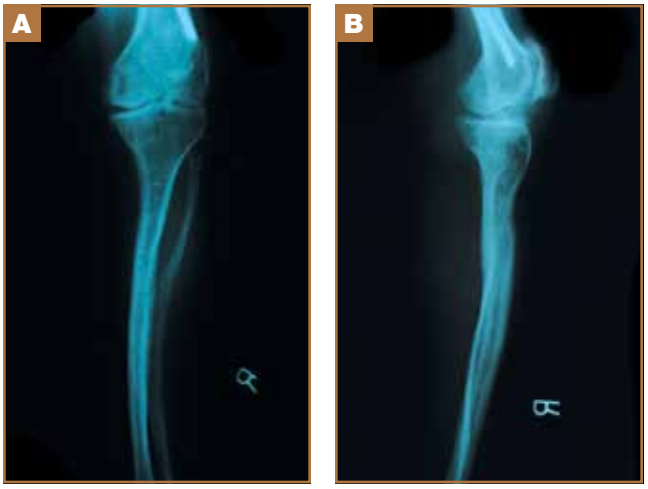

Figure 2. AP (A) and lateral (B) radiographs of right tibia.

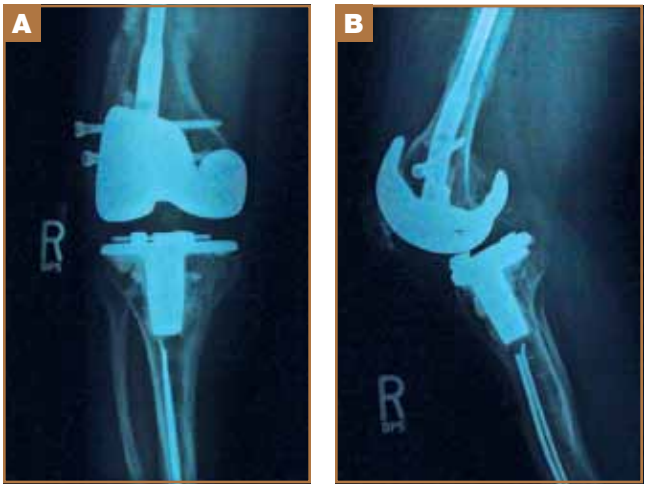

Figure 3. AP (A) and lateral (B) radiographs of right knee status post TKA and femoral and tibial osteotomies.

Figure 4. AP (A) and lateral (B) radiographs of right tibia status post tibial osteotomy and retrograde flexible nails with minimal callous formation.

with mild opening of the medial joint line. Over the next year, his pain progressed to the point where he could no longer ambulate and required a wheelchair for mobility. At this point, he requested surgical intervention to help with his pain.

In the operating room, tricompartmental changes with large osteophytes and significant erosion of the medial tibial plateau were noted. Using a radiolucent table and c-arm to confirm alignment, femoral and tibial cuts were made and a cemented posterior cruciate-retaining prosthesis was placed. The femoral rush rod was removed and a closing wedge osteotomy of the distal femur malunion site was performed through a separate lateral incision. An 8-mm diameter reamed intramedullary nail was introduced in a retrograde manner and locked, providing good stability of the femur.

The intramedullary canal of the tibia demonstrated an undulating course appearing to be approximately 5 mm at its thinnest diameter, with clinically visible valgus deformity occurring proximal to the ankle. A closing wedge distal tibial osteotomy through a small medial incision with osteoclasis of the fibula was performed. In order to span the whole tibia and avoid a stress riser, we placed 2 flexible titanium nails (3 mm on lateral side; 2.5 mm on medial side) in a retrograde manner through the medial malleolus and the anterolateral distal tibia. Clinically, the femur and tibia were well aligned and the leg length discrepancy corrected. While performing a trial range of motion, the lateral margin of the patella cracked, however, it remained in continuity within the quadriceps mechanism and the patellar component was stable on the medial portion of the patella (**Figure 3A, 3B**).

Postoperatively, he was placed in a fracture boot with toe-touch weight-bearing for 8 weeks and began physical therapy with a CPM machine. Secondary to concerns regarding the increased incidence of malunion and nonunion in OI patients, an ultrasound bone stimulator was ordered to facilitate healing. Three months postoperatively, he reported soreness at the tibial osteotomy site, but did not report knee pain or pain at the femoral osteotomy site, and range of motion was full extension to 120° of flexion. Radiographs showed bridging callous at the femoral osteotomy site and "blurring" of the tibial osteotomy site without much callus formation and maintenance of tibial alignment.

Six months postoperatively, the tibia had not healed and InFUSE recombinant bone morphogenic protein by Medtronic was placed in the operating room with continuation of the fracture boot and weight-bearing. At 11.5 months out from the original procedure the right tibial nonunion persisted (**Figure 4A, 4B**) and an open reduction internal fixation (ORIF) with iliac crest bone graft was performed. At the time of surgery, no obvious gap was present but minimal callous formation and visible rotational motion of the osteotomy were present. Three to 4 mLs of cancellous graft from the anterior iliac crest was obtained and packed around the osteotomy site; a 3.5 mm dynamic compression plate with 4 screws proximal and distal to the nonunion was placed. In order to avoid the titanium nails, 1 screw was placed

in compression and another in a non-locking manner; the remainder of the screws were locked.

Three weeks following ORIF with bone grafting, our patient finally reported no pain in the area. The patient continued to improve over the next few months and 15 months since the original total knee arthroplasty (TKA) and osteotomy, the tibial nonunion had healed (**Figure 5A, 5B**). He reported no pain in his tibia or right knee and was ambulating without assistance, and he was able to return to work. Five years later, his activity level was limited by pain in his left knee and we performed left TKA. On this leg, the extraarticular deformity was approximately 10° and did not require a concurrent osteotomy. He recovered without event, regained function, and is now 2 years after this second operation.

Figure 5. AP (A) and lateral (B) radiographs of right tibia after plating with healed osteotomy.

Case 2

A 37-year-old male with a history of OI and recurrent fractures presented with right knee pain and deformity of the right tibia. He was experiencing severe pain affecting his ambulation and ability to perform activities of daily living, and required increasing use of a wheelchair. Radiographs demonstrated significant anterolateral bowing of the tibia with varus alignment of the knee and anterior subluxation of the tibia (**Figure 6A-6D**). We discussed with him our limited prior experience, particularly complications, but felt surgery would offer significant chance for improvement in pain and function. Therefore, he agreed to undergo right TKA with bilevel tibial osteotomies.

In the operating room, tricompartmental arthritic changes with anterior subluxation of the tibia were noted. Routine preparation of the femur and tibia were performed and a trial reduction carried out. Prior to cementing the components, two 30° closing–wedge tibial osteotomies were performed at the location of maximal deformity through 2 medial incisions correcting the varus alignment and anterior bow. Again, the intramedullary canal of the tibia had an undulating course and very narrow diameter. Two flexible intramedullary nails (3 mm and 3.5 mm) were passed across the osteotomy site in an antegrade manner through the resected surface of the proximal tibia then cut off and driven below the level of the proximal tibial plateau. This did not provide adequate rotational stability as suspected, so a 3.5 mm dynamic compression plate with locking screws was placed on the medial aspect of the tibia, spanning both osteotomies. We supplemented the construct with InFUSE (rh-BMP-2) by Medtronic to stimulate healing of the osteotomy sites.

Postoperatively, the patient was placed in a hinged knee brace and was ambulating with toe touch weight-bearing. He was discharged on postoperative day 7 due to difficulties performing physical therapy. Three months after his procedure, he continued to ambulate with crutch assistance despite receiving physical therapy 3 times per week and reported only mild pain. On physical examination the right knee demonstrated full extension to 95° of flexion, which was relatively unchanged from his preoperative range of motion. Radiographs showed evidence of healing on the medial side, but limited callus formation on the lateral side of the osteotomies. Six months after surgery, he was pain free and ambulating without assistance. Radiographs demonstrated

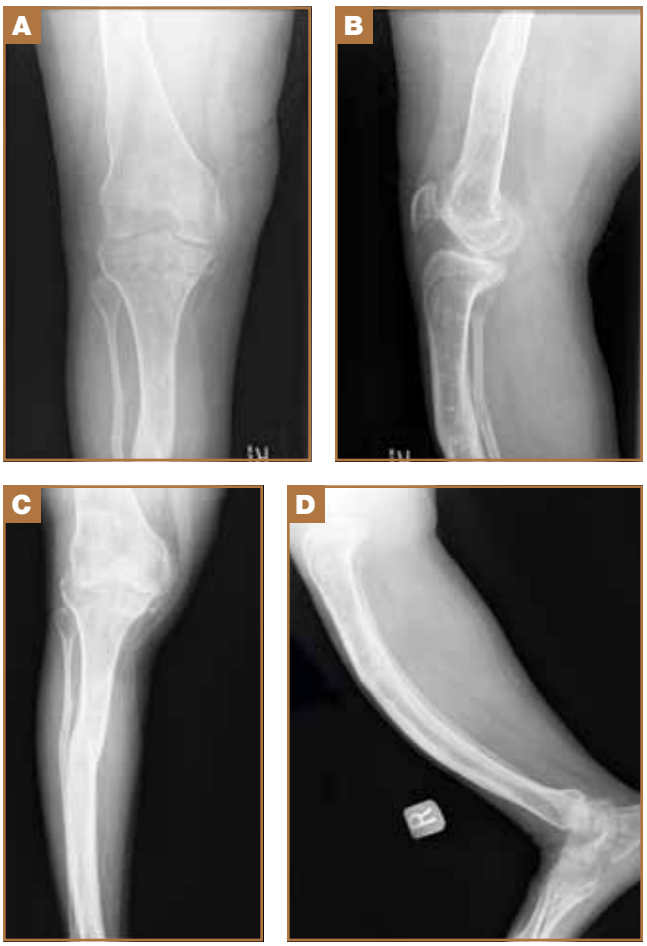

Figure 6. AP and lateral radiographs of the right knee (A, B) and right tibia (C, D).

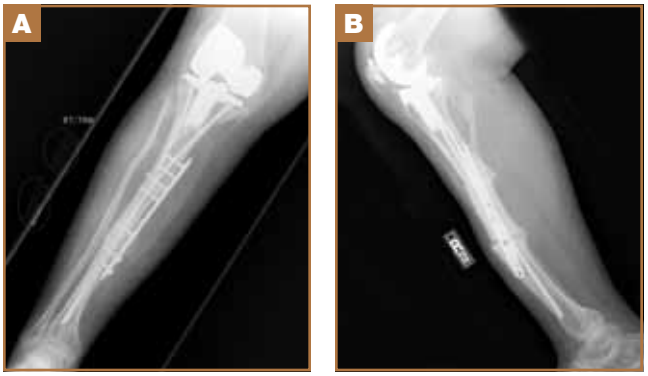

Figure 7. AP (A) and lateral (B) radiographs of the right knee and tibia after TKA and tibial osteotomies demonstrating healing of osteotomy sites.

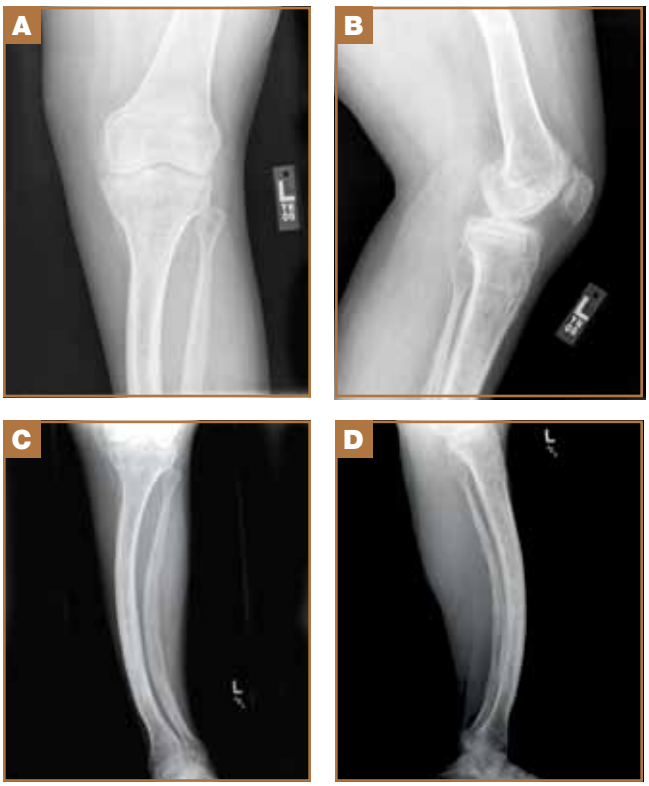

Figure 8. AP and lateral radiographs of the left knee (A, B) and tibia (C, D).

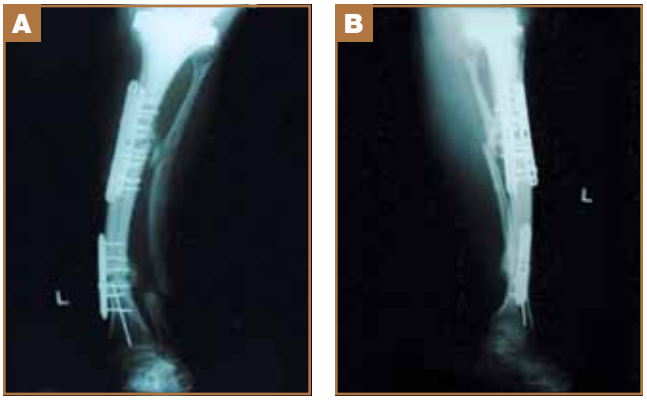

Figure 9. AP (A) and lateral (B) radiographs of left knee with tibia and osteotomy nonunion.

Figure 10. AP (A) and lateral (B) radiographs of left tibia after plating with healed osteotomies.

satisfactory healing of the osteotomy sites (**Figure 7A, 7B**).

He was extremely grateful for the improvement in daily function and requested surgery for his left knee and tibia (**Figure 8A-D**). Therefore, 8 months after the right side was performed, he was scheduled to undergo left TKA with bilevel tibial osteotomies.

During the procedure, bone-on-bone contact in the medial compartment was noted along with erosion of the patellofemoral joint. In the same manner as the right leg, standard preparation for a total knee and 2 closing wedge osteotomies were performed; 2 flexible titanium intramedullary nails (2.5 mm and 2.5 mm) were passed across the osteotomy sites through the resected edge of the proximal tibia prior to cementing. Two dynamic compression plates were placed medially at the osteotomy sites for torsional stability. Following cementing of the prosthesis, the knee was noted to hyperextend and appeared unstable. Evaluation revealed a fracture of the proximal tibia causing the tibial prosthesis to slope anteriorly. A portion of the posteromedial and pos-

terolateral tibial plateau also had a minor fracture, yet the soft tissues were in continuity providing collateral stability. Revision of the tibial component with an 80 mm cemented stem and 10 mm augments provided proper stability with good range of motion.

To place the stemmed tibial component, the intramedullary nails were shortened, and one of the nails broke distally during extraction, probably due to the screw placement and nail damage. The osteotomy sites appeared stable despite the broken nail and we felt that the benefits of replacing the broken nail did not outweigh the risks. Again, InFUSE bone morphogenic protein by Medtronic was placed at both the proximal and distal tibial osteotomy sites.

His left leg improved over the next 8 months. However, he continued to experience occasional mild discomfort at the osteotomy sites and was not placing full weight on the extremity. Radiographs 9 months postoperatively revealed a radiolucent gap at the proximal tibial osteotomy site, and the distal osteotomy site appeared to be healing with a valgus deformity (**Figure 9A, 9B**). After a lengthy discussion of the options, our patient decided to proceed with revision of the left tibial malunion and nonunion. One long 3.5 mm dynamic compression plate was placed and, 2 months following the osteotomy revision, our patient was finally pain free with full range-of-motion of the left knee and a small amount of callus formation. His ambulation continued to improve and osteotomies were healed 3 months after his revision surgery. Two years after his last operation, he continues to ambulate without pain and without ambulatory aids (**Figure 10A, 10B**).

Discussion

OI results in numerous fractures that frequently heal with some degree of malunion and would be expected to result in progressive arthritis. Very little information exists to guide orthopedic surgeons care for these patients.

Papagelopoulos and Morrey,[5] reported 2 total hips and 3 total knees performed in patients with OI at the Mayo Clinic. They noted good pain relief and improvement of function. The patients in that series did not have extra-articular deformity to the degree that would require an osteotomy. We know of no other studies regarding knee replacement in this population.

Patients with extra-articular deformity at the time of TKA may be managed with either adjusted intra-articular resection or with extra-articular osteotomy, either as a staged procedure or concurrent with the knee replacement. Correction through intra-articular resection has several advantages, including the avoidance of nonunion and infection at the osteotomy site and an additional incision. The amount of coronal plane deformity that can be corrected with intra-articular resection has been reported in the range of 10° to 30°.[9,10,11] Wang and Wang[12] reported correction by intra-articular resection in patients with up to 20° of deformity in the femur and 30° in the tibia, if the line perpendicular to the mechanical axis of the femur at the femoral condyle did not pass through the insertions of the collateral ligaments, and if the line drawn from the medullary canal of the tibia distal to the angular deformity passed within the tibial condyle. Lonner and colleagues[11] reported simultaneous femoral osteotomy and TKA in 11 patients using either a blade plate, retrograde femoral nail, or a long-stemmed press-fit femoral component for osteotomy fixation, with one nonunion in the patient with the long-stemmed femoral component. As was seen in the 2 patients reported here, patients with OI frequently have deformity to the degree that it is not correctable with intra-articular resection.

Patients with OI present several challenges that are illustrated by the patients in this report. Despite attempting to handle the knee gently, both patients sustained a minor fracture during routine performance of the operation: a longitudinal nondisplaced fracture of the patella in one patient and a compression fracture of the proximal tibia in the other patient. Neither fracture negatively affected the outcome, but the proximal tibial compression fracture required a stemmed tibial component with augments.

The intramedullary canal of the tibia may be very narrow and preclude the use of a standard intramedullary nail. Intramedullary fixation with a Steinman pin or Rush rod as described in the Sofield osteotomy for children with OI[4,2,13] is usually supported with a cast, but it is ideal to avoid a long leg cast after a TKA to avoid stiffness. In the first patient reported, we used the largest flexible titanium nails that would fit in the canal to provide transaxial fixation for transverse bilevel osteotomies and to span the length of the bone, and a fracture boot for additional support. We later found small locking plates necessary to provide rotational stability and heal the osteotomy. Plates alone may be adequate to obtain stability and union but might lead to a future fracture at the end of or between the plates, especially in OI patients.

In the second patient, the flexible titanium nails were introduced through the resected surface of the proximal tibia rather than in a retrograde manner through the distal tibia. Although this method avoided the separate incisions and possible complications of distal insertion, it makes removal of the nails very difficult. When one of the nails was noted to be broken intraoperatively as described above, extraction would have been difficult. Consequently, retrograde placement may be preferable.

Another option to stabilize the osteotomy is external fixation. Ring and colleagues[5] reported deformity correction in 6 adult patients with OI using an Ilizarov fixator. Although all patients were satisfied with the result, there were 18 complications reported, including knee stiffness in 2 patients, peroneal palsy in 2, knee instability, knee infection, and deep pin tract infection.

Patients with OI have an increased incidence of delayed union and nonunion;[1,14] therefore, ultrasound (first patient) and bone morphogenic protein (second patient) were used at the time of the initial tibial osteotomies. These adjuncts may not have been necessary, were costly, and nonunion resulted nevertheless. In the first patient, the anterior iliac crest bone graft harvested at the time of nonunion surgery provided a

disappointing amount of cancellous bone.

Both of these patients were active and productive despite significant deformity, as was reported in adult OI patients by Moorefield and Miller,[8] but had become wheelchair dependent before their operations. Although both patients had complications, they both were able to regain their mobility and were happy with their pain relief and function, at least at short-term followup. Therefore, we believe knee replacement, with concurrent osteotomy as needed for extraarticular deformity, should be considered.

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