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## Arthroplasty in patients with rare conditions

# Staged Bilateral Total Knee Arthroplasty for Neglected Blount Disease Using a Gap Balancing Technique

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#### ABSTRACT

Blount disease is an acquired, asymmetrical disorder of proximal tibial growth that results in a complex three-dimensional proximal tibial deformity, with tibial varus being the dominating feature. Although the exact pathophysiology is unknown, Blount disease is separated into 2 clinical variants, infantile and adolescent, based on the onset of symptoms occurring before or after the age of 10 years. If recognized and treated early, affected patients generally have a favorable prognosis; however, if neglected, it can lead to progressive malalignment and premature osteoarthritis. We present a patient with bilateral neglected Blount disease who underwent successful bilateral total knee arthroplasty performed in a staged fashion using a gap balancing technique with constrained condylar knee implants.

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## Introduction

Blount disease is a rare, acquired disorder of the medial proximal tibial physis and epiphysis that results in a three-dimensional proximal tibial deformity [1]. Although the exact etiology is unknown, it is generally considered to be multifactorial, with an increased prevalence in obese patients, early walkers, and African American, Afro-Caribbeans, or Hispanic populations [1-3]. Through application of the Hueter-Volkmann principle, it is thought that mechanical overload of the posteromedial proximal tibial physis causes osteochondrosis and eventual physeal bar formation, which results in a proximal tibial varus, internal tibial torsion, procurvatum, and leg length discrepancy [2].

Two distinct clinical and radiographic forms of Blount disease have been recognized and separated based on age at onset, with 10 years used as the cutoff. The infantile form is generally seen in children under the age of 3 years, is more common in females, more likely to be bilateral, and often has higher degrees of proximal tibial varus and posterior epiphyseal slope [2]. Adolescent Blount disease is seen in children older than 10 years, is more common in males, and is more strongly correlated with obesity [1,3]. In the adolescent

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form, epiphysiodesis is rare, the deformity is typically less severe, and approximately one-third of the overall varus malalignment is attributed to distal femur varus [2,4].

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Blount disease generally has a favorable prognosis if recognized and treated early; however, if neglected, it can lead to progressive malalignment and premature osteoarthritis (OA). In adults with neglected or severe Blount disease, premature OA develops around 30 to 50 years of age [1,5]. Total knee arthroplasty (TKA) is typically reserved for adult patients with severe deformity and advanced degenerative changes. Given the three-dimensional proximal tibial malalignment and associated bone defects, which are often coupled with obesity-related comorbidities, TKA for Blount disease patients can be challenging.

To our knowledge, only one study has reported outcomes after TKA for patients with Blount disease: Natoli et al. reported successful outcomes for TKA in 5 patients with Blount disease or Blount-like deformity [6]. In order to highlight and expand on these findings, we report on a patient with neglected Blount disease who had bilateral TKAs performed in a staged fashion using a gap balancing technique with constrained condylar knee (CCK) implants. We believe our case to be novel in that regard and hope to provide more insight into technical considerations when addressing this type of cases.

The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

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## **Case history**

A 71-year-old otherwise healthy African American female from Nigeria presented to our clinic with a long history of bilateral knee pain, left worse than right, and severe progressive varus deformity. As a child, she endorses a long history of progressive knee deformity starting early in childhood. Owing to her limited access to health care in Nigeria, her condition was ultimately neglected and continued to progress early into her adult life. At the time of presentation to our clinic, she lived a semi-sedentary lifestyle and was limited in her activities of daily living secondary to pain and disability. She described her symptoms as severe bilateral knee pain, rated as 10/10, that was worse with activities. She was only able to ambulate indoors, with strict reliance on a front-wheeled walker. She had failed a series of conservative treatment measures including non-steroidal anti-inflammatory drugs, activity modification, medial unloading knee braces, corticosteroid injections, and physical therapy. She denied any history of childhood obesity, trauma, surgery, or infection to either knee.

The patient's body mass index at initial presentation was 30.7 m/kg<sup>2</sup>. On examination, she had severe fixed varus deformities, with 10-85° of knee range of motion bilaterally. Her gait examination was significant for lateral thrust to her knees bilaterally. She exhibited 4/5 weakness to her quadriceps bilaterally and was otherwise neurovascularly intact distally. Her preoperative Knee Injury and Osteoarthritis Outcome Score for Joint Replacement score was 26 with an interval score of 15.9. Preoperative radiographs demonstrated severe tricompartmental knee arthritis bilaterally with approximately 35° varus malalignment, 18° of posterior tibial slope, and significant posteromedial proximal tibia bone loss, consistent with infantile Blount disease (Figs. 1 and 2). The patient's tibia metaphyseal-diaphyseal angle was measured to be 23° on the right and 18° on the left (Fig. 3). Also, the presence of several loose bodies, particularly on the left side, was noted (Fig. 1).

#### Preoperative considerations

The patient had a complete preoperative workup, as well as evaluations by her primary care physician and anesthesiologist to ensure that she was medically optimized before surgery. There was no evidence of vitamin D deficiency or other metabolic abnormalities on laboratory evaluation. We discussed continued nonoperative care and operative interventions including knee arthroplasty or fusion. Given the bilateral nature of her disease and severe limitations in activities of daily living, she elected for staged bilateral TKA starting with her left, more symptomatic, side.

The senior surgeon (R.P.R.) routinely uses a gap balancing technique for femoral and tibial components and resurfaces nearly all patellae with cemented patellar components. Given the patients' severe deformity, both primary and CCK implants (NexGen Legacy Constrained Condylar Knee [LCCK]; Zimmer-Biomet, Warsaw, IN) with stem extensions were made available on the day of surgery. As the patient was noted to have significant posteromedial tibial bone loss, a universal small fragment system (DePuy Synthes, Raynham, MA) and bone cement (Biomet Bone Cement R; Zimmer-Biomet, Warsaw, IN) were made available for possible use of a screw and cement rebar technique [7,8]. Given the unknown degree of bone loss and bone quality preoperatively, trabecular metal modular augments and cones were also made available.

## **Operative considerations**

The patient was positioned supine on a regular table, and a standard medial parapatellar approach was performed under hyperbaric Marcaine spinal anesthesia. Upon entering the knee, several loose bodies were removed from the suprapatellar pouch. She was noted to have a fixed deformity with severe wear on the medial tibia. The patella was provisionally cut then translated laterally, and the knee was flexed. As is routine for senior surgeon (R.P.R.), a gap balancing technique where the extension gap was balanced before the flexion gap was used. Resection of the distal femur was accomplished using an intramedullary cutting guide at 5 degrees of valgus. Given her wear on the femur, 2 mm of extra distal femur were cut to get down to a stable boney base medially. The tibia was translated anterior, and an extramedullary guide was used to resect the tibia at 3° of posterior slope and neutral alignment, with approximately 12 mm of bone taken off the lateral plateau. Tibial rotation was set based on the relationship of the medial third of the tibial tubercle to the center of the tibial tray. Resection depth was estimated based on preoperative anteroposterior radiographs using the proximal tip of the fibula as a guide for the level of maximum resection. This left a residual 3-mm uncontained defect in the posteromedial aspect of the tibial that was appropriate for screw and cement rebar technique.



Figure 1. Preoperative radiographs of the left knee with anteroposterior (a), lateral (b), and sunrise (c) views demonstrating severe varus malalignment, with excessive posterior tibial slope, and posteromedial tibial bone loss. Abundant loose bodies are also noted.



Figure 2. Preoperative radiographs of the right knee with anteroposterior (a), lateral (b), and sunrise (c) views demonstrating severe varus malalignment, with excessive posterior tibial slope, and posteromedial tibial bone loss.

The extension gap was then evaluated using spacer blocks to assess for mechanical alignment of the bone cuts and initial collateral tension and stability. Given her severe fixed varus deformity, a large release of the medial soft-tissue structures was performed to essentially skeletonize the medial tibia. This required release of osteophytes, posteromedial capsule, hamstrings, deep medial collateral ligament (MCL), and medial proximal tibial reduction osteotomy to help balance her knee in extension. After releasing all soft tissues medially except the superficial MCL, the extension gap was then checked using both spacer blocks and the



**Figure 3.** Anteroposterior radiograph of bilateral knees demonstrating a tibia metaphyseal-diaphyseal angle (TMDA) of 23° for the right knee and 18° for the left. The TMDA was measured as follows: (1) A reference line is drawn along the long axis of the tibia (dashed line); (2) a line is drawn perpendicular to this reference line that intersects the most distal aspect of the lateral beak of the proximal tibial metaphysis; (3) the angle between this perpendicular line and a line intersecting the most distal aspects of the medial and lateral beaks of the tibial metaphysis were measured.

kinematic balancer (FuZion; Zimmer-Biomet, Warsaw, IN) and found to still have varus instability in extension with the lateral hemi-gap looser than the medial hemi-gap. Medially the gap felt best with the 14-mm block but still had approximately 5 mm of opening laterally. Given that it was unlikely that further medial soft-tissue releases would be able to catch up to the loose lateral hemi-gap without total release of the superficial MCL, constrained nonhinged components were planned.

By holding to ankle and leg to remain in extension and neutral alignment, and not allowing it to fall into varus when tensioning the extension space, the gap opened to 14 mm at 30-40 lbs of torque; thus, 14 mm was the planned size of insert. The knee was flexed, and the kinematic balancer with the NexGen tower now attached was used to establish femoral component translation and rotation and drilled at a gap to match the 14-mm insert with 30-40 lbs of tension. By using the tensioner in flexion to match the forced neutral tension in extension, 3 of the 4 hemi-gaps with the final construct should be equal at a 14-mm insert; thus, the CCK constraint would only be "needed" to account for the lack of balance in the single lateral hemi-gap in extension.

After drilling the holes to match a 14-mm posterior referenced insert, the alignment and rotation was approximately parallel with the epicondylar axis and Whiteside's line. The anterior condylar cut was then made. The cutting block was removed, the 4-in-1 cutting block for the LCCK femur was placed, and the remaining condylar, chamfer, and box cuts were made as well as reaming performed for a 15  $\times$  30-mm femoral stem. Upon finishing the femur, the tibia was translated anterior, and after appropriate tibial sizing and rotation were established, trial implants were inserted, and a posterior-stabilized trial insert was inserted to assess the balance. As expected, 3 of the 4 hemigaps were balanced without constraint and left the lateral hemi-gap in extension loose; thus, CCK final components were inserted.

The wound was irrigated, and two 3.5-mm fully threaded cortical screws were placed for screw and cement rebar medially to her small residual posteromedial tibial defect. The tibia was then cemented, and the cement was allowed to fully harden before mixing a second batch of the cement for the femur and patella. The final femur and patellar components were then cemented, placed with a trial 14-mm

insert, and held with axial loading until final hardening of the second batch of cement was achieved. The tourniquet was released, hemostasis achieved, repeat irrigation was performed, and the final 14mm LCCK insert placed and screwed into the tibial component. The patella was noted to track well and did not require lateral release. Intraoperative range of motion demonstrated 0-135° of flexion with excellent stability in extension, mid-flexion, and at 90° of flexion. The patient recovered postoperatively without incident and was discharged home on postoperative day 1.

After discharge, the patient was followed up in our clinic postoperatively and progressed well with outpatient physical therapy. She was planned for staged right TKA 6 weeks later; however, her index left TKA was performed immediately before the COVID-19 pandemic, and owing to the resulting restrictions on elective procedures, her right TKA was not performed until approximately 3 months after her left side. During this time, it should be noted that she had approximately 20 mm of a leg length discrepancy (LLD) because of the correction of her malalignment. We were concerned she would develop a flexion contracture on the left side given her LLD; however, she was diligent with her home exercise program and achieved excellent range of motion. The patient recovered postoperatively without incident and was discharged home on postoperative day 1 (Fig. 4). Once COVID-19 restrictions were released, we proceeded with right TKA using an identical technique (Fig. 5). Owing to changes in hospital recovery protocols and expectations in response to the COVID-19 pandemic, her right TKA was able to be performed as an outpatient procedure.

After her right TKA, the patient followed up at routine intervals of 2 weeks, 6 weeks, 3 months, and 12 months postoperatively without postoperative complications, readmissions, or emergency room visits. She was able to wean off her walker 8 weeks postoperatively and was ambulating without assist device by 3 months postoperatively. There was no evidence of component loosening or osteolysis noted on postoperative radiographs. The patient's final knee range of motion (ROM) was 0-120° bilaterally. She is extremely satisfied with the results of her surgeries, with a Knee Injury and Osteoarthritis Outcome Score for Joint Replacement score of 1 and an interval score of 91.9 at her most recent follow-up.



Figure 4. Postoperative radiographs of the left knee with anteroposterior (a), lateral (b), and sunrise (c) views demonstrating correction of alignment in the coronal, sagittal, and axial planes, respectively.



Figure 5. Postoperative radiographs of the right knee with anteroposterior (a), lateral (b), and sunrise (c) views demonstrating correction of alignment in the coronal, sagittal, and axial planes, respectively.

#### Discussion

Blount disease is associated with a complex, three-dimensional proximal tibial deformity that can result in early OA and severe malalignment if left neglected. To our knowledge, only one study has been carried out in recent literature that examined TKA in patients with Blount disease or Blount-like deformity [6]. Our case report of a patient with neglected Blount disease who had bilateral TKA performed in a staged fashion, using a gap balancing technique with CCK implants, highlights technical considerations to consider when approaching these cases.

The primary goals when approaching any TKA are implant stability, soft-tissue balance, and restoration of alignment [9]. To achieve a balanced knee, 2 distinct methods of knee prosthesis implantation have been described: measured resection and gap balancing. Several studies have shown comparable outcomes between these techniques for primary TKA [10-12]. One limitation of measured resection, however, is that it relies on bony landmarks followed by soft-tissue balancing to achieve rectangular flexion and extension gaps. In cases where bony landmarks are distorted, such as in neglected Blount disease, this may lead to inaccuracies in restoring normal balance [12]. With a gap balancing technique, ligament releases are performed early, without complete reliance on bony landmarks, which brings the limb into the correct approximate alignment before determination of femoral component rotation [11].

For both the affected knees in this report, there was severe varus malalignment with posteromedial bone loss, as is typical of neglected Blount disease [6]. After performing an extensive medial release, as well as proximal tibial reduction osteotomy, there was still significant varus instability in extension; therefore, the use of a constrained implant was planned. Although the need for a primary constrained implant over a posterior-stabilized or a cruciate-retaining implants is rare, increasing the amount constraint to achieve proper knee stability may be necessary in complex primary cases with severe deformity when persistent laxity occurs despite aggressive attempts at soft-tissue balancing. Historically, concerns for using constrained implants in TKA are related to the increased stress at the bone-cement-implant interface as well as the sacrifice of more bone, which have been associated with higher risk for aseptic

loosening and revision [13-15]. However, several studies have still demonstrated favorable outcomes for constrained TKA implants at the index surgery, if indicated [16-18]. Nonconstrained implants should be considered in Blount disease with less severe deformities; however, it should be noted that in the report by Natoli et al., 2 of the 6 knees in that series that underwent the surgery without constrained components were subsequently revised to hinge components [6]. Therefore, we argue that there should be a low threshold for considering primary CCK or hinge implants when approaching Blount disease cases, especially those with severe deformity.

The decision for using CCK implants over hinge components in this case was based on the integrity of the collateral ligaments and degree of bone loss. In situations, either primary or revision, where the collateral ligaments are incompetent, there is gross flexion-extension gap imbalance, or there is severe or relevant bone loss affecting the insertion site of the collateral ligaments, then a CCK would not provide adequate stability and a hinge component should be used instead [19,20]. In this case report, CCK components with fully cemented short stem extensions were used for both tibial and femoral components to increase the contact area of the bone-cement interface, which has been shown to reduce the rate of loosening and increase the longevity of the implant [21–23], although their necessity has been debated in literature [24].

For both the knees presented in this report, there were posteromedial small residual uncontained bone defects that were approximately 3 mm in depth. Given the size of defects and their contained nature, we elected to use a screw and cement rebar technique, which have been shown to have 30% less loosening of the prosthesis than cement alone in tibial defects [23,25]. The use of screws is intended to distribute the load away from the joint line and the cement-bone interface and has been proven to be a reliable, reproducible, inexpensive technique when addressing these defects [23,25]. In cases where there are large uncontained defects, however, modular metal augments, metaphyseal sleeves, or trabecular metal cones should be considered as an alternative [25].

#### Current controversies and future considerations

Other considerations for this case are the options for staged vs simultaneous bilateral TKAs and the possibility for outpatient TKA in severe Blount disease. Although some studies have shown higher risk of medical complications, thrombotic events, and need for blood transfusions with simultaneous bilateral TKA [26-28], other studies have shown that simultaneous bilateral TKA has an acceptable risk, especially in appropriately selected patients [29,30]. For this case, we performed staged TKAs: however, it should be noted that due to the severity of the patient's deformity. the patient had an approximately 20-mm LLD after the first TKA. which made rehabilitation and ambulation difficult; however, this may have possibly avoided the medical complications associated with simultaneous TKA. Ultimately, the final decision on staged vs simultaneous should be left to discretion of the patient and surgeon. In addition, given her success of the first TKA and necessity of outpatient surgery in the setting of COVID-19 restrictions, the second-staged TKA was performed as an outpatient procedure. These technically complex cases that are able to be discharged home the same day of surgery may further allude to the success of rapid recovery TKA protocols, even in complex deformity cases.

Future considerations and limitations in preoperative planning for these cases included the assessment of mechanical alignment and torsional deformity. As long-leg radiographs are not available at the authors' institution, the angle between the femoral and tibial anatomical axes on standard anteroposterior weight-bearing knee radiographs was used as a surrogate to predict the true mechanical axis. The tibia and femoral anatomic axes were determined by lines running from the midpoint of the tibial spines to a point midway between the medial and lateral cortices 10 cm above the tibial spines for the femur and 10 cm below the tibial spines for the tibia [31,32]. Although this is one alternative to the hip-knee-ankle angle in the assessment of mechanical alignment in the absence of longleg radiographs [31,32], it can be associated with potential error in estimation of the true mechanical alignment [33,34]. Furthermore, assessment of torsional deformity that may be present in Blount disease [1] may be further evaluated preoperatively by the use of computerized tomography scans for visualization of the anatomic relationship of the patellofemoral joint and measurement such as tibial tubercle to trochlear groove distance [35]. However, in this report, torsional deformity did not significantly impact the rotation of tibial component, which was ultimately set based on the relationship of the medial third of the tibial tubercle to the tibial tray.

## Summary

This report describes a 71-year-old female with neglected Blount disease who underwent successful bilateral, staged TKAs using cemented CCK components with a gap balancing technique. Although this report is limited to the findings of one patient with Blount disease, we show that TKA can be successfully performed in these patients through extensive preoperative planning and execution of the appropriate operative technique. When addressing these cases, surgeons should be prepared to manage posteromedial tibial bone loss and use constrained prothesis at the primary procedure. We advocate for the use of a gap balancing technique, especially when bony landmarks for a measured resection are deformed. Further studies are needed to show the long-term outcomes of primary TKA in this patient population.

#### **Conflicts of interest**

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this article.

### **KEY POINTS**

- Blount disease is an acquired disorder of the medial proximal tibial physis and epiphysis that results in a three-dimensional proximal tibial deformity [1]. There are 2 clinical variants, infantile and adolescent, based on the age of onset with 10 years being the cutoff.
- If recognized and treated early, Blount disease generally has a favorable prognosis; however, if neglected it can lead to progressive malalignment and premature osteoarthritis. TKA is typically reserved for adult patients with severe deformity and advanced degenerative changes.
- When performing TKA in these cases, surgeons should be prepared to manage posteromedial tibial bone loss and have a low threshold for use of a constrained prothesis at the primary procedure.
- A cement with rebar technique is appropriate for addressing small residual uncontained defects in these cases; however, in cases where there are large uncontained defects, surgeons should be prepared to use modular metal augments, metaphyseal sleeves, or trabecular metal cones.
- We advocate for the use of a gap balancing technique in these cases, especially when bony landmarks for a measured resection are deformed. Through this approach, ligament releases are performed first, without complete reliance on bony landmarks, which brings the limb into the correct approximate alignment before determination of femoral component rotation.

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