

# Bilateral Total Ankle Replacement as a Viable Treatment Option for Severe Ankle Arthritis Secondary to Hemophilia C

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### Statement of Purpose

- Hemophilia is a rare medical condition (1 in 100,000 live births). There are multiple types categorized by a deficiency in a specific bleeding factor. One rare sub-type is Hemophilia C (factor XI).
- A sequela of hemophilia is a condition called hemophilia arthropathy which often leads to severe deformation of joints, debilitating pain, and reduction of activity.
- The purpose of this presentation is to present a rare case of a patient with Hemophilia C that presented with bilateral debilitating ankle arthritis that was successfully treated with total ankle replacement (TAR) and proper perioperative planning.

### Literature Review

- Prior to development of TAR, the standard of care for patients with hemophilic ankle arthropathy was arthrodesis. Several studies evaluated these outcomes with good results, however not without negative outcomes such as difficulty walking and failure to return to high activity level. [4-5] With the emergence of TAR, options are now available for these patients.
- In the literature review there was no documented cases of Hemophilia C being treated with bilateral TAR. All studies reported on patients who had Hemophilia A or B. The results of these studies demonstrated significant functional improvements with longevity of the implant. [5-7]

### Case Study

- His bilateral radiographs and CT scan demonstrate extensive osteoarthritis with incongruent ankle deformity. There was a significant talar tilt, valgus deformity, and extensive degenerative joint disease of the ankle joint. (Fig 1 and 2)
- The patient selected to undergo bilateral TAR and gastrocnemius recession. The patient had right TAR preformed and five years later returned for the same procedure on the left.
- Preoperatively in both cases he received 4 units of fresh frozen plasma as recommended by his hematologist.
- A two component modular TAR with long tibial stem was implanted utilizing intramedullary alignment with success on both sides.
- Postoperatively patient had no issues with bleeding and did not require any more FFP.
- Radiographically, rectus ankle is appreciated on both sides. No complications occurred and an increase in 30 degrees ankle joint range of motion was noted bilaterally.
- Five years postoperatively on the right, patient was asymptomatic and pleased with his results. One year postoperatively on the left, patient has no discomfort and was back to high baseline activity.

### Analysis & Discussion

- We were able to successfully correct the patient's deformity on two occasions without complications and achieved great results. The patient is back to his baseline activity level, which includes 100+ mile bike rides per week with complete resolution of pain.

### Background Information

- The pathophysiology of hemophilia arthropathy stems from a minor trauma. This results in hemarthrosis which eventually leads to severe arthritis.
- Hemophilia C is unique from other types of hemophilia in such that the frequency of bleeding is not determined by the factor level. Furthermore, it is not typically associated with bleeding into joints and muscles.
- Due to the nature of condition, meticulous perioperative planning is mandatory. The surgeon should anticipate for intraoperative bleeding, replacement protocols and other such complications.

### Case Study

- A 70-year-old male with a past medical history of Factor XI deficiency, hypertension, hyperlipidemia, presented for bilateral severe ankle arthritis secondary to Hemophilia C. Patient was an avid bike rider (100 miles per week) and wished to return to that level of activity without pain. Both ankles were deformed, painful, and unresponsive to conservative treatment.
- On physical examination, he had pain and crepitus with manipulation, and no range of motion of his bilateral ankles.

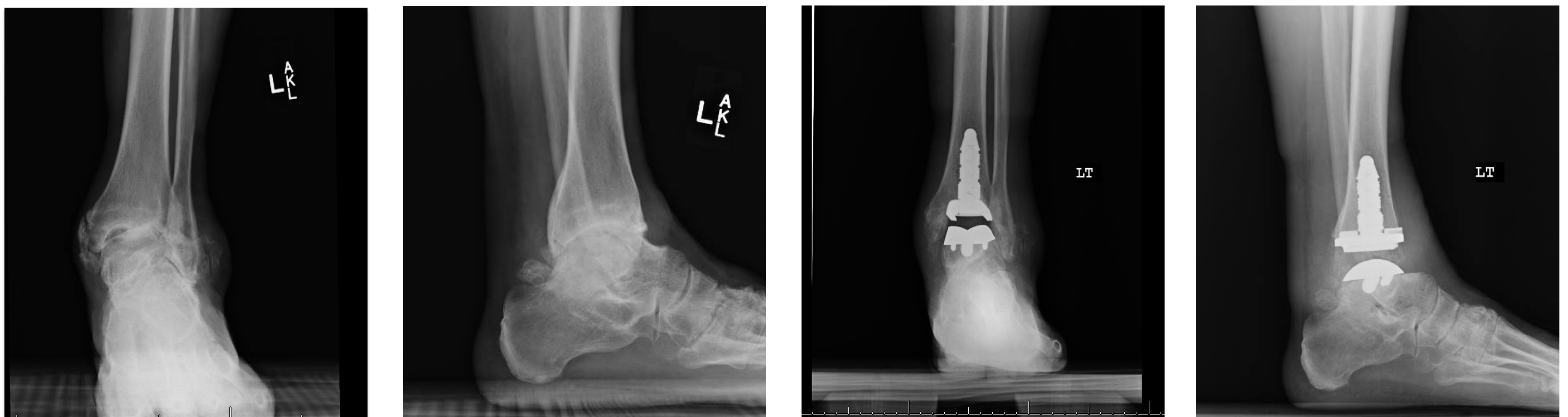
### Analysis & Discussion

- Hemophilia C is a rare condition with limited research that requires a multi-disciplinary approach. The condition results in severe deformity and arthritis of ankles.
- TAR is a viable treatment option for this severe deformity with emphasis on proper perioperative planning.
- Eckers et al. demonstrated ankle implant survivorship of 94% at 5, 85% at 10, and 70% at 15 years in 17 hemophilic patients.
- Barg et al. study showed significant functional improvement in range of motion and in level of pain in 14 patients with Hemophilia.



Fig. 2. Preoperative & Postoperative Radiographs of Right Ankle: AP & Lateral Views

Fig. 1. Preoperative & Postoperative Radiographs of Left Ankle: AP & Lateral Views



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