

IN BRIEF

Total Ankle Replacement in Hemophilia

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Abstract

The objective of this paper is to carry out a review of the literature on the contemporary situation of total ankle replacement (TAR) in people with hemophilia (PWH). A search was performed in PubMed on December 29, 2024, utilizing the keywords "hemophilia" and "total ankle replacement." To date solely 8 papers (136 TARs) in 103 PWH have been reported. All published series showed improvement in preoperative pain and function. However, the complication rate was 8.8% (intra-operative fractures of the medial malleolus, delayed wound healings, hematomas). Additional surgery is required in 5.1% of the TARs implanted (ankle arthrolysis due to painful arthrofibrosis, major surgeries, revision surgeries). The estimated 10-year survival free of any TAR removal or ankle fusion (AF) is 92.2%. Regarding the controversy AF or TAR in PWH, a recent systematic review and meta-analysis have found similar clinical results and rates of adverse events between TAR and AF in PWH.

Level of evidence: III**Keywords:** Ankle, Ankle fusion, Hemophilia, Hemophilic arthropathy, Total ankle replacement

Introduction

People with hemophilia (PWH) who are not appropriately managed from birth from the hematological viewpoint (proper replacement of the deficient clotting factor) will suffer recurrent hemarthroses in elbows, knees and ankles, and less commonly in shoulders and hips. As a result, between the 20 and 30 years of age they will develop severe degeneration of the involved joint(s) (hemophilic arthropathy).¹⁻⁴

When hemophilic arthropathy causes severe pain and functional disability of the ankle joint and they do not responded to conservative management [pain killers, cyclooxygenase-2 inhibitors (COX-2 inhibitors), intraarticular injections of hyaluronic acid and Physical and Rehabilitation Medicine procedures], the involved joint(s) will require surgical treatment [mainly ankle fusion (AF) or total ankle replacement (TAR)].⁵⁻¹⁶

Ankle problems in PWH have constantly been very uncommon, even when managed on demand from the hematologic viewpoint.⁵ The goal of this paper was to carry out a review of the literature on the outcomes and adverse

events of TAR in PWH.

Main body

The method of this review was the following: A search of the literature in PubMed on December 29, 2024, utilizing "hemophilia" and "total ankle replacement" as keywords was carried out. Of the 64 papers found, only 8 papers were focused on TAR in PWH. Therefore, this is a narrative review of the literature. Main information on TARs carried out in PWH is summarized in [Table 1].⁵⁻¹²

All published series showed improvement in preoperative pain and function. The mean age of the patients ranged from 43 years to 51.4 years. The mean follow-up was between 3 years and 9.6 years. Of the 136 TARs, 12 (8.8%) had complications: 2 intra-operative fractures of the medial malleolus, 2 delayed wound healings and 6 hematomas). Additional surgery was required in 7 (5.1%) of the 136 TARs implanted: 2 ankle arthrolysis due to painful arthrofibrosis, 2 major surgeries and 3 revision surgeries). The 10-year survival free of any TAR removal or AF was 92.2%.

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Table 1. Published series of total ankle replacement (TAR) in people with hemophilia (PWH)

Author, Year, Reference	Type of Bleeding Disorder	Number of Patients	Number of TARs	Mean Age of Patients and Range (Years)	Mean Follow-up and Range (Years)	Results	Adverse Events
Van der Heide et al, 2006 ⁵	Hemophilia	3	5	NA	4.3 (1-8.7)	At final follow-up all TARs were still in place and did not show any signs of loosening. Clinical scores showed a good to excellent result.	None
Barg et al, 2010 ⁶	Hemophilia	8	10	43.2 (26.7-57.5)	5.6 (2.7-7.6).	The AOFAS-hindfoot-score increased from 38 preoperatively to 81 postoperatively. All PWH were satisfied with the results. Four patients became pain free; in the whole patient cohort pain level decreased from 7.1 preoperatively to 0.8 postoperatively. All categories of SF-36 score showed substantial improvements in quality of life.	There were no intra- or peri-operative complications. In one patient, open ankle arthrolysis was carried out because of painful arthrofibrosis.
Strauss et al, 2014 ⁷	Hemophilia	10	11	49 (37-59)	3 (1.2-5.4)	In two patients deep prosthesis infection occurred leading to the removal of the prosthesis. In the remaining eight patients the mean AOFAS score improved substantially from 21.5 to 68 points, the VAS score decreased substantially from 7.6 to 1.9 points. ROM increased from 23.2° to 25°. At final follow-up all PWH without any complications were satisfied with the postoperative results.	None
Asencio et al, 2014 ⁸	Hemophilia	21	32	NA	4.4 (2.7-6.2)	The overall AOFAS score improved from 40.2 (pre-surgery) to 85.3 (post-surgery). The function score increased from 23.6 to 35.9 and dorsiflexion from 0.3° to 10.3°. Two patients experienced further ankle arthrodesis. On X-ray, both tibial and talar components were stable and correctly placed in all ankles. Alignment was good.	None
Barg et al, 2015 ⁹	Von Willebrand disease	18	18	47.3 (34.0-68.7)	7.5 (2.9-13.2)	Pain level (VAS) decreased from 8.2 preoperatively to 1.1 postoperatively. Substantial functional improvement including ROM was found. All categories of SF-36 score showed substantial improvement in quality of life.	One patient sustained an intraoperative medial malleolar fracture. In two patients delayed wound healing was found. Two secondary major surgeries were carried out.
Preis et al, 2015 ¹⁰	Hemophilia	14	14	51.4 (32.9-63.7)	5.8 (2-9.2)	VAS substantially decreased from 8.5 to 1.3. Significant functional improvement including ROM and AOFAS hindfoot score was found. The summarized components of the SF-36 physical and mental outcomes score substantially improved at the latest follow-up.	One patient sustained an intraoperative medial malleolar fracture. In two patients, delayed wound healing was observed. In one patient, open arthrolysis was performed due to painful arthrofibrosis.

Table 1. Continued

Eckers et al, 2018 ¹¹	Hemophilia	14	17	43 (27.4-57.6)	9.6 (3.3-17.8)	Estimated implant survival was 94% at 5, 85% at 10 and 70% at 15 years, respectively. At follow-up, 9.6 year postoperatively, the level of satisfaction was 76% and of pain 2/10 on the VAS. ROM had increased substantially. The SF-36 summary scores were comparable to those of a matched standard population. The AOFAS hindfoot score averaged 81 points.	Three cases required revision surgery.
Favelle et al, 2024 ¹²	Hemophilia	25	29	44.7 (26-65)	NA	The 10-year survival free of any prosthesis removal/arthrodesis was estimated to be 92.2% .	Six patients had hematoma.

NA = Not available; AOFAS = American Orthopedic Foot and Ankle Society; SF-36 = Short Form (36) Health Survey; ROM = range of motion; VAS = visual analog scale

Comparative studies: TAR versus ankle fusion (AF)

An important issue at present is whether it is better to perform AF or TAR in PWH. There are some studies that have

attempted to answer this dilemma, the main data and results of which are summarized in [Table 2].¹³⁻¹⁶

Table 2. Comparative studies: total ankle replacement (TAR) versus ankle fusion (AF) in people with hemophilia (PWH).

Author, Year, Reference	Type of Bleeding Disorder	Number of Patients	Number of TARs / AFs	Mean Age of Patients and Range (Years)	Mean Follow-up and Range (Years)	Results	Adverse Events
Dauty et al, 2015 ¹³	Hemophilia	2	1 / 1	NA	NA	TAR was recommended for PWH who present with a preserved ankle ROM.	NA
Ahn et al, 2020 ¹⁴	Hemophilia	29	16 / 13	44.1 ± 9.9	6.8 ± 3	Both TAR and AF in ESHAA exhibited significant improvement in pain based on VAS and FFI scales. Compared to AF, TAR resulted in superior outcomes in FFI disability and activity subscales, suggesting that TAR may be considered as a surgical option alongside AF for ESHAA.	Three cases of osteolysis and 1 case of heterotopic ossification were noted in the TAR group. No cases of nonunion were noted in the AF group. Progressive osteoarthritis of adjacent joints after AF was observed in 1 case.
Mussawy et al, 2021 ¹⁵	Hemophilia	19	11 (in 10 patients) 11 (in 9 patients)	49.4 (TAR group), 37.5 (AF group)	NA	Of the 11 ankles that underwent AF, 10 showed bone healing not later than 12 weeks after surgery, whereas one still showed nonunion after 6 months. VAS pain scores decreased significantly in both groups. Mean AOFAS scores also improved significantly, from 28.1 before to 80.3 after AF and from 21.5 before to 68 after TAR. No perioperative complications were observed in either group.	Late deep infection was observed in two patients that underwent TAR, which required removal of the implant.

Table 2. Continued

Arthur et al, 2023 ¹⁶	Hemophilia	SYSTEMATIC REVIEW	NA	NA	NA	Studies showed that the degree of improvement in AOFAS hindfoot-ankle score, VAS pain scores, and the mental and physical component summary scores of the 36-Item Short Form Health Survey were similar for both surgeries.	Complication rates were also similar between the 2 surgeries. Additionally, studies showed a significant improvement in ROM after TAR.
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NA = not available; ROM = range of motion; ESHAA = end-stage hemophilic ankle arthropathy; VAS = visual analog scale; FFI = Foot Function Index; AOFAS = American Orthopedic Foot & Ankle Society.

Conclusion

Preoperative pain and function improve after TAR. However, the complication rate of TAR is 8.8%. Besides, additional surgery is required in 5.1% of the TARs implanted. The 10-year survival free of any TAR removal or AF is 92.2%.

This is important to emphasize that TAR does not ensure the same long-lasting symptoms alleviation and range of motion increase as hip and knee replacements, as the related published articles show: this is the principal motive to pay attention for this indication in all PWH suffering from ankle arthropathy. Specifically as dedicated orthopedic surgeons know, for young individuals, it is essential to consider also the other joints (subtalar and mid-tarsal) and for adult individuals, to choose AF.

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