9 Advanced Hemophilic Arthropathy of the Ankle: Total Ankle Replacement or Ankle Fusion?

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9.1 Introduction

In the final stages of hemophilic arthropathy of the ankle, when the joint is severely destroyed and the intense joint pain and/or major functional disability do not respond to nonsurgical treatment (hematological prophylaxis, analgesics, anti-inflammatories, orthotics), we must consider the need to carry out surgical treatment to alleviate the patient's problems [1–4].

Among the surgical techniques for the treatment of ankle joint destruction in idiopathic degenerative disease, before reaching the elimination of the joint via arthrodesis or total ankle replacement (TAR), we can try to relieve the symptoms by using the following surgical techniques [5]: arthroscopic ankle debridement (Fig. 9.1), joint distraction (arthrodiastasis) using external fixation, or supramalleolar osteotomy for alignment (in cases with particularly bad alignment).

However, in some patients, none of these techniques sufficiently alleviates the problem, leading us to consider the elimination of the joint via arthrodesis (joint fusion) or TAR. Today there is much controversy, both in the general population and in hemophilia patients, about which technique is most advisable. The purpose of this article is to review the literature on arthrodesis and TAR in non-hemophilia population, as well as in hemophilia patients, to try to clarify the controversy over arthrodesis or TAR.

9.2 Search Strategy

In a literature search for articles published in English in PubMed (MEDLINE) that included from January 2000 to December 2013, we found 43 articles related to ankle arthrodesis and TAR in non-hemophilia patients and in hemophilia patients. Another three articles on survival of knee prosthesis (TKR) and hip replacements (THR) were included for comparison with the ankle prosthesis (TAR). The keywords used were hemophilia, ankle, arthrodesis, and TAR. In total 46 articles were analyzed.

9.3 Results of TAR and Arthrodesis

In 2005, Stengel et al. [6] published a systematic review and meta-analysis (included in the Cochrane Library) on the effectiveness of TAR on the general population. In 1,086 patients, 35.2

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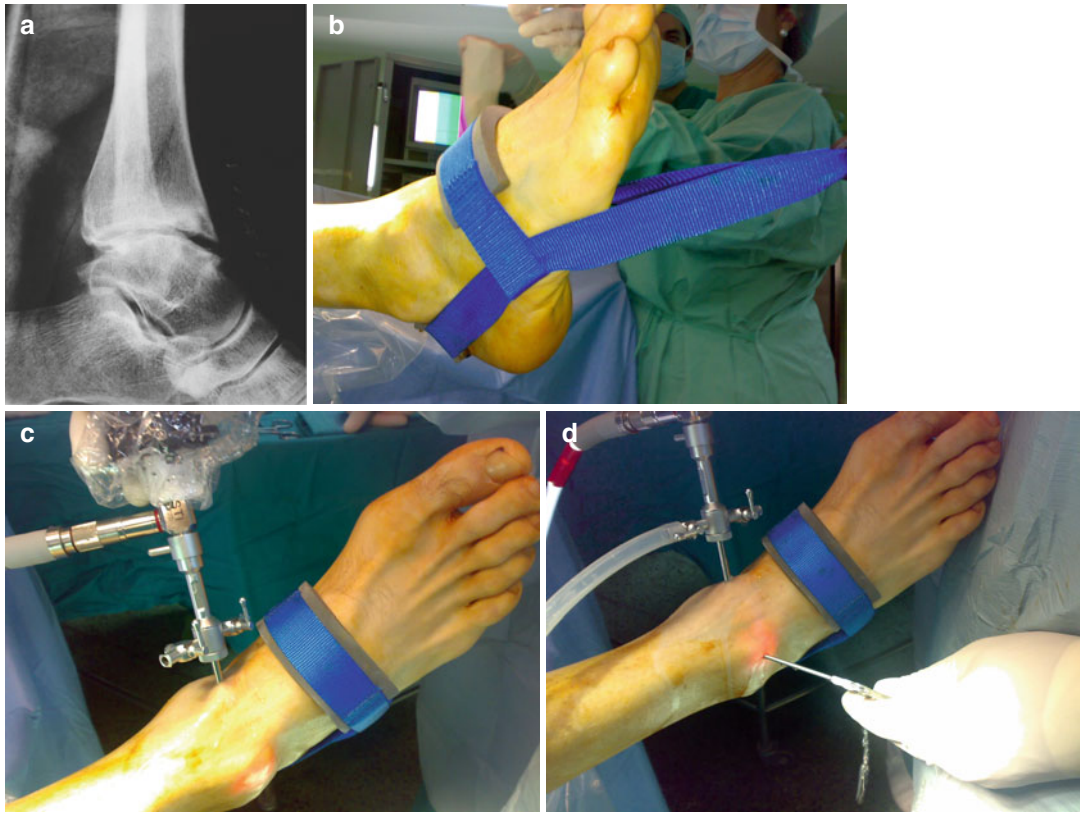

Fig. 9.1 Arthroscopic debridement of the ankle in a patient with advanced hemophilic arthropathy: preoperative lateral radiograph (**a**). Traction device used for the procedure (**b**). Anteromedial and anterolateral portals used to perform debridement (**c**, **d**)

complications were found (1.6 % deep infections, 14.7 % impingement, 12.5 % secondary surgery, 6.3 % secondary arthrodesis). The prosthetic survival at 5 years was 90.6 %.

In 2011, in another systematic review, included in the Cochrane Library, Zhao et al. [7] analyzed 2,088 TARs with an average follow-up of 52 months. The failure rate was 11.2 %, whereby almost half of them occurred in the first year (5.2 % due to aseptic loosening, 1.7 % due to misalignments and 1 % of infections). The prosthetic survival at 5 years was 85.9 % and at 10 years was 71.1 %.

Also in 2011, Mann et al. [8] analyzed 84 TARs (in 80 patients), achieving a level of satisfaction with the result of 92 %. At an average of 9.1 years, 91 % of the TARs were still in place. They had a 25 % complication rate (including 14 secondary surgeries). The prosthetic survival at 5 years was 96 % and at 10 years was 90 %. Hendrickx et al. [9] analyzed 66 ankle arthrodesis (in 60 patients) with an average follow-up of 9 years, achieving a fusion rate of 91 %. There were seven (10 %) complications (six re-arthrodesis, one infection). 91 % of patients were satisfied with the result. Progressive osteoarthritis was detected in the adjacent joints, although its importance is still unknown.

In 2012, in another article, included in the Cochrane Library, Roukis et al. [10] analyzed 2,312 TARs with an average follow-up of 22.8 months. 9.7 % of them (224) had to be revised, leading to new prostheses in 182 cases (81.3 %), 34 in arthrodesis (15.2 %), and 8 in amputation below the knee (3.6 %).

In 2013, Noelle et al. [11] analyzed 100 TARs performed on 97 patients with an average

follow-up of 36 months. They had 27 complications and needed 21 revisions. Gordon et al. al [12] studied 82 ankles (73 patients) on which open ankle arthrodesis was performed using the anterior approach. 100 % of cases fused, with an average fusion time of 13.3 months. 80 % of patients were very satisfied or satisfied. The complication rate was 14.6 % (malalignment, healing problems, complex regional pain syndrome, delayed fusion).

A controversial topic regarding ankle arthrodesis is whether it should be tibiotalar (TT) or tibiotalocalcaneal (TTC). Ajis et al. [13] analyzed 100 TT arthrodesis, and they compared them with 173 TTC arthrodesis, with an average follow-up of 63 months (minimum 24 months). There were no differences in the results as regards three of the parameters studied: preoperative pain relief, return to previous work (74 %), and whether patients would be operated again (83 %), the results were similar. However, there was a difference in the desired level of activity, which was 58.5 % in TT arthrodesis and 66.5 % in TTC arthrodesis.

One of the most striking complications of TAR is periprosthetic fractures. In 2013, Manegold et al. [14] reported a rate of 4.2 % (2.2 % intraoperative, 2 % postoperative).

A controversial issue is the revision of the TAR when it fails. Hinterman et al. [15] in 2013, presented 117 TAR revisions (in 116 patients), noting that the survival of the prosthetic revision was 83 % after 5 years, with similar results to those of primary TAR. There were 19 complications (16 %) highlighting 1 malleolar fracture, 1 dislocation of the prosthetic polyethylene, and 15 secondary revision surgeries.

It should be noted that although ankle arthrodesis is usually performed as an open procedure [1–5], it can also be carried out by arthroscopy. In fact, Lee et al. [16] in 2011 and Townshend et al. [17] in 2013 stated that arthroscopic ankle arthrodesis has better fusion rates, fewer complications, less postoperative pain, and a shorter hospital stay than open arthrodesis.

9.4 Comparative Studies: TAR vs Arthrodesis

In 2009, Saltzman et al. [18] noted that TAR provides better function than arthrodesis; however, from the pain perspective, the relief is comparable. Their study (included in the Cochrane Library) compared 158 TARs and 66 ankle arthrodesis, with an average follow-up of 24 months.

Noelle et al. [11] obtained a similar complication rate between TAR and arthrodesis. In an ongoing study, Flavin et al. [19] noted a clear improvement after either procedure (TAR or arthrodesis) with similar postoperative results.

In a comparative study published in 2012 by Schuh et al. [20], in which TARs (20 cases) and arthrodesis (21 cases) were compared with an average follow-up of 34.5 months, the authors found no differences in sports, recreational activities, and function.

According to Terrell et al. [21], the number of TARs increased by 57 % from 2004 to 2009, although the number of ankle arthrodesis did not change in that time period. However, as the previously studied literature highlights, prosthetic survival of the TAR (Table 9.1) is far from that of hip replacements (THR, total hip replacement) and knee (TKR, total knee replacement) [22–26]. TAR survival at 14 years is 62 %, while that of THR is 93 % and 88 % at 15 and 20 years, respectively [27]. TKR survival is somewhat less than that of THR, 84 % and 71 % at 10 and 20,

Table 9.1 Survival rates for total ankle replacement (TAR) in the general population according to literature (2005–2013)

Author	Survival at 5 years (%)	Survival at 10 years (%)	Survival at 14 years (%)
Stengel [6]	90.6	–	–
Zhao [7]	85.9	71.1	–
Mann [8]	96	90	–
Henricson [22]	81	69	–
Pinar [23]	86	–	–
Barg [24]	94	84	–
Brunner [25]	–	70.7	45.6
Angthong [26]	–	–	77

respectively [28, 29]. Obviously there are no TAR survival studies with over 14 years of follow-up. Our opinion coincides with that of Henricson et al. [22], who stated that TAR survival will not come close to that of THR and TKR in the near future in the population with advanced ankle arthrosis. We must not forget that TAR currently presents a high complication rate, failures, and revision (Table 9.2). In comparative studies, Saltzman et al. [18] indicate better function after TAR but equivalent pain relief. Schuh et al. [20] found no difference between TAR and arthrodesis as regards sports, recreational activities, and function. Flavin et al. [19] did not find any differences in postoperative progress either. Thus, even the literature with a high level of scientific evidence does not clarify the controversy over TAR or ankle arthrodesis, but it does state that TAR has not reached the levels of survival of THR and TKR.

Table 9.2 Rates of complications, failures, and revision for total ankle replacement (TAR) in the general population according to literature (2005–2013)

Author	Complications (%)	Failures (%)	Revision (%)
Stengel [6]	35.2	–	–
Mann [8]	25	–	–
Zhao [7]	–	11.2	–
Angthong [26]	–	4.9	–
Henricson [22]	–	–	22
Roukis [10]	–	–	9.7
Noelle [11]	27	–	21
Barg [24]	–	–	8.4
Brunner [25]	–	–	38

9.5 Total Ankle Replacement (TAR) or Arthrodesis of the Ankle in Hemophilia

According to Ling et al. [30], the incidence of arthropathy is very high in hemophilia patients, whereby 47 % of them were in pain and 52 % had positive radiological signs. In hemophilia, patients with ankle arthropathy present a modification in progress that improves recovery via the pendular mechanism, in order to save energy. This modification is proportionately greater in cases of greater arthropathy [31].

Before considering surgery, in severe hemophilic arthropathy of the ankle nonsurgical treatment including hematological prophylaxis, analgesics, anti-inflammatory, rehabilitation, and functional orthoses should be attempted [1–5, 32–34]. A surgical alternative used on only three hemophilia patients is arthrodiastasis (joint distraction) via circular external fixator by Ilizarov [34]. An alternative is supramalleolar osteotomy for realignment mentioned by Pearce et al. [35] who performed seven such interventions on six hemophilia patients.

As regards the controversy over TAR or ankle arthrodesis in hemophilia (Figs. 9.2, 9.3, and 9.4), back in 1976, Zimbler [36] mentioned the

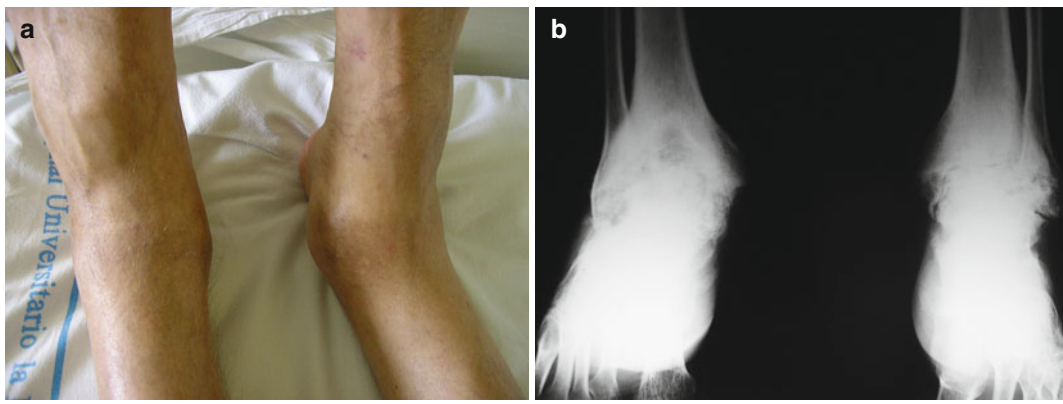

Fig. 9.2 Right ankle arthrodesis with a retrograde locking nail in a patient with severe hemophilic arthropathy of the tibiotalar and subtalar joints: preoperative view of the ankles (**a**). Anteroposterior radiograph of the ankles before surgery (**b**)

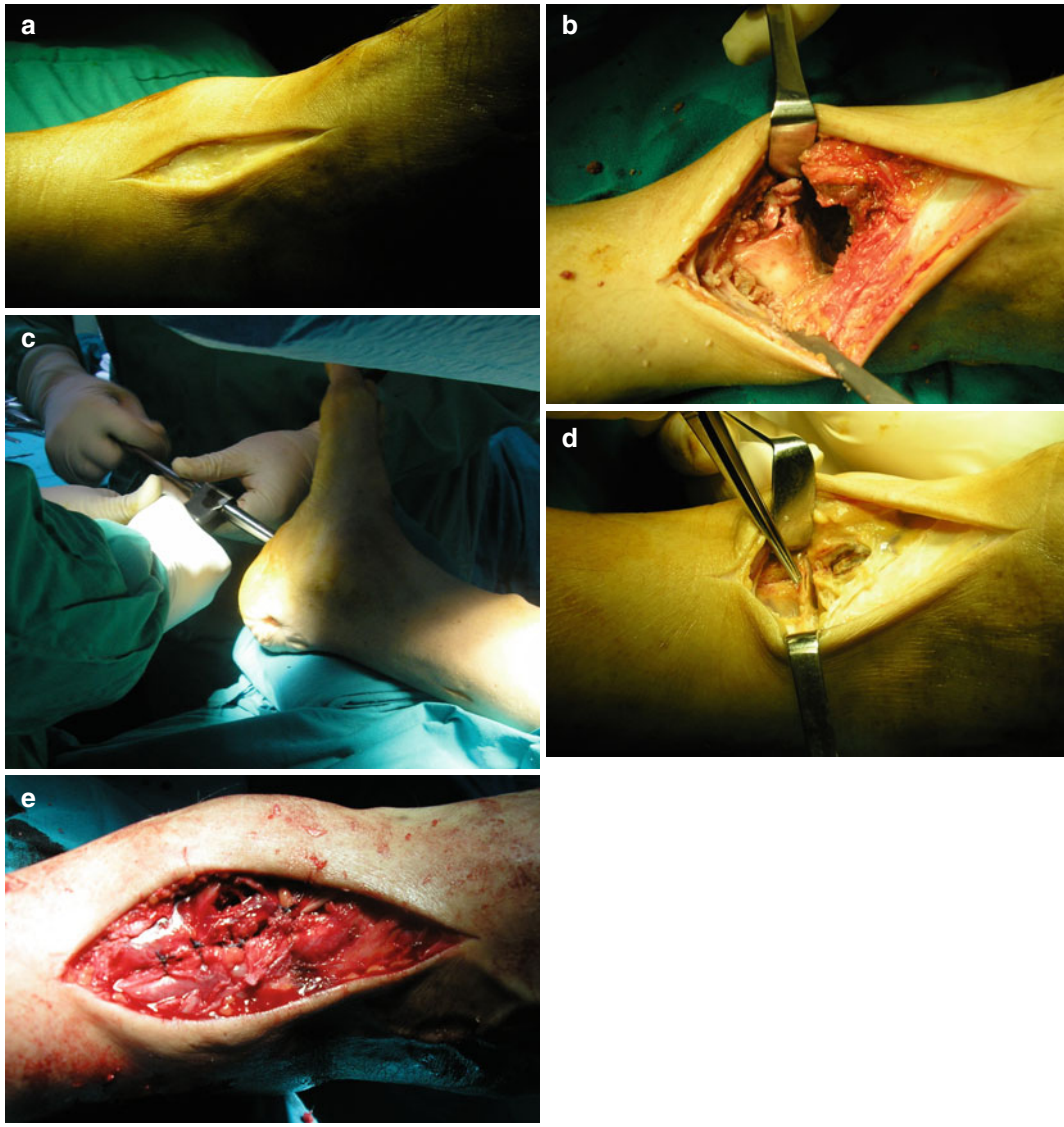

Fig. 9.3 Right ankle arthrodesis with a retrograde locking nail in a patient with severe hemophilic arthropathy of the tibiotalar and subtalar joints: intraoperative images of the procedure (**a–e**)

possibility of using TAR in these patients. In 1978 Houghton et al. [37] published seven ankle arthrodeses in hemophilia with satisfactory results. In 1991 Gambler et al. [38] noted that in elderly hemophilia patients, arthrodesis eliminates pain and bleeding, improving the deformity.

As regards ankle arthrodesis, in 2010, Tsailas and Wiedel [39] published 20 arthrodeses (in 13 patients with an average age of 38.7 years), 11 of them ankle (tibiotalar), 1 subtalar, and 8 combined. Average follow-up was 9.4 years. In 2011, Tsukamoto et al. [40] presented three arthroscopic ankle arthrodeses in two patients with hemophilia. In 2013, Bluth et al. [41] presented 54 ankle arthrodeses with an average of 6.6 years (in 45 patients). There was tibiotalar nonunion in 10.4 % of patients and subtalar nonunion in 8.3 % of them, but no further surgery was required. Their conclusion is that arthrodesis is a suitable surgical treatment. The ankle arthrodesis technique described in 2009 by Mann et al. [42] is of interest.

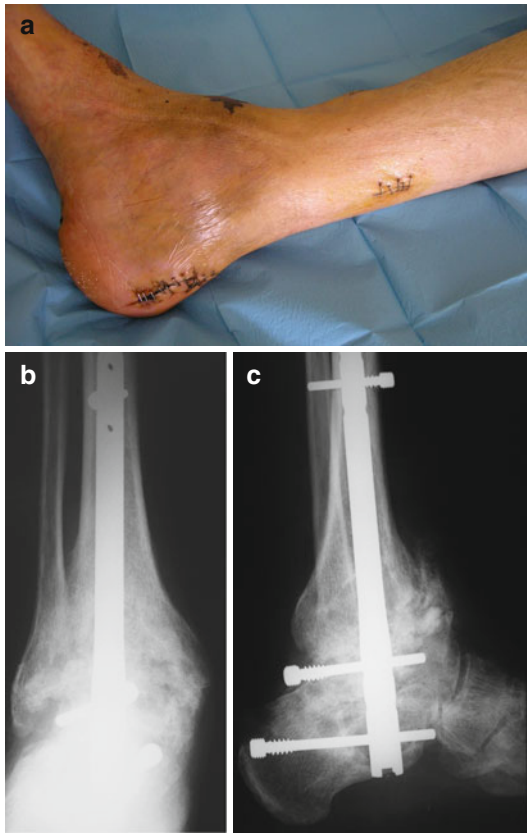

Fig. 9.4 Right ankle arthrodesis with a retrograde locking nail in a patient with severe hemophilic arthropathy of the tibiotalar and subtalar joints: lateral postoperative view of the ankle (**a**). Anteroposterior radiograph after arthrodesis (**b**). Lateral view after ankle fusion (**c**)

Bai et al. reported a series of ten patients (ten ankle joints) who underwent arthroscopically assisted ankle arthrodesis for the treatment of end-stage hemophilic arthropathy [43]. The fusion rate was 100 %. The average time to fusion was 10.5 weeks. Superficial wound infection occurred in one patient. There were eight good to excellent results and two fair results. All patients were satisfied with the outcome of the operation. Arthroscopic ankle arthrodesis was an effective alternative to open technique with established advantages in hemophilic arthropathy [43].

As regards TAR in hemophilia, in 2006, van der Heide et al. [44] published five cases (in three patients) with an average of 4.3 years. In 2010, Barg et al. [45] published ten TARs in eight patients, with an average age of 43.2 years with an average follow-up of 5.6 years (minimum 2.7 years). They had only one complication (painful fibrosis that required open arthrolysis).

The literature on TAR versus ankle arthrodesis in hemophilia is very limited and has a limited degree of evidence. Therefore, the literature does not tell us which of the two techniques is most suitable in advanced hemophilic ankle arthropathy. Before TAR or arthrodesis, we should think about arthroscopic debridement [1–4], arthrodiastasis [46], or supramalleolar osteotomy for realignment [35].

9.6 Author's Experience

In a period of 40 years, the authors performed 454 orthopedic surgical procedures on 398 hemophilia patients. Of these, only six were ankle procedures: four arthroscopic debridement and two ankle arthrodesis (one tibiotalar, one tibiotalocalcaneal). To date, we have not performed any TARs given its short survival in the long term and high rate of complications. In our experience, ankle arthropathy can be sufficiently relieved in a large percentage of cases with nonsurgical treatment, and the need for surgical intervention on this joint is very rare. For us, removing the ankle joint (via arthrodesis or TAR) is always the last option: we always try arthroscopic debridement before considering arthrodesis or TAR. On the other hand, given the controversy in the literature on ankle arthrodesis or TAR and the lack of literature on the subject in hemophilia, we currently tend to prefer arthrodesis to TAR in people with hemophilia. If the subtalar joint is affected, we prefer open tibiotalocalcaneal arthrodesis with locking nail. If not, we would opt for open tibiotalar arthrodesis with crossed cannulated screws or staples.

Conclusions

The current literature concerning the controversy on ankle arthrodesis or total ankle replacement (TAR) in non-hemophilia patients

is not definitive (it does not clarify the controversy). As regards hemophilia patients, the uncertainty is even greater, as there is very little literature available. Based on all of this and on our 40 years of experience treating people with hemophilia, our advice is to exhaust all types of nonsurgical treatment. When surgical treatment is considered absolutely necessary, my recommendation is to conserve the joint at all costs using arthroscopic debridement, arthrodiastasis, or supramalleolar osteotomy for realignment, according to each surgeon's preferences. If these techniques fail, when in doubt on whether to perform arthrodesis or TAR, in hemophilia, we would always opt for arthrodesis, as the current results for TAR quite frankly have much room for improvement. It is likely that in the mid-long term, new TAR designs will allow these results to improve.

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