Orthopaedics & Traumatology: Surgery & Research xxx (2017) xxx–xxx

Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com/en

Original article

Ankle fusion in hemophilic patients

D. Eichler[a], M. Ehlinger[a,*], A. D'Ambrosio[a], D. Desprez[b], G. Bierry[c], P. Adam[a], F. Bonnomet[a,b]

[a] *Service de chirurgie orthopédique et de traumatologie, hôpital de Hautepierre, hôpitaux universitaires de Strasbourg, 1, avenue Molière, 67098 Strasbourg cedex, France*
[b] *Service d'hématologie clinique, centre régional du traitement des hémophiles (CRTH), hôpital de Hautepierre, hôpitaux universitaires de Strasbourg, 1, avenue Molière, 67098 Strasbourg cedex, France*
[c] *Service de radiologie osseuse, hôpital de Hautepierre, hôpitaux universitaires de Strasbourg, 1, avenue Molière, 67098 Strasbourg cedex, France*

ARTICLE INFO

Article history:
Received 3 March 2017
Accepted 22 August 2017

Keywords:
Ankle
Fusion
Hemophilia
Clotting factors

ABSTRACT

Introduction: Hemophilic arthropathy is painful and disabling. We report a retrospective study of ankle fusion with intra- and peri-operative clotting factor perfusion. The objective was to assess the efficacy of maintaining perioperative clotting factor rates close to 100%, and report long-term results. The study hypothesis was that results would be good, without early hemorrhagic complications.
Material and method: Between 2000 and 2013, 12 ankle fusions were performed in 9 patients, with a mean age of 39 years (range, 19–58 years). Anti-hemophilic factor perfusion was controlled by the reference physician of the Regional Hemophilia Treatment Center. Clinical AOFAS and Olerud scores and the Pettersson radiologic score were used for assessment. Mean preoperative AOFAS score was 22 (range, 2–55) and mean Olerud score 7 (range, 5–12). Mean preoperative factor VIII concentration was < 1% (range, < 1–3%).
Results: Mean follow-up was 8 years (range, 2–16 years). Mean AOFAS score at follow-up was 69 (range, 35–92) and mean Olerud score 70 (range, 30–100). Improvement mainly concerned the Pain dimension. Statistical analysis found a significant difference between pre- and post-operative clinical scores (AOFAS, $P = 0.004$; Olerud, $P = 0.004$). Mean factor VIII concentration at surgery was 90% (range, 24–117%), and 109% (range, 75–152%) the day following surgery. There were no cases of hematoma or surgical site infection. Radiologic fusion was systematic at a mean 3.5 months (range, 3–4 months).
Conclusion: The study hypothesis was confirmed. Ankle fusion in advanced hemophilic arthropathy improved function and quality of life. Perioperative clotting factor perfusion contributed to these good results, providing supplementary prevention of hemorrhagic risk.
Level of evidence: IV, retrospective study.

1. Introduction

Hemophilic arthroplasty is painful, destructive and disabling. It predominantly affects the weight-bearing joints, especially knee and ankle, and less frequently the hip [1]. It is the main complication of severe hemophilia, and is secondary to iterative hemarthrosis, occurring spontaneously or due to minor trauma [2]. Recurrent hemarthrosis leads to chronic synovitis, with initially hypervascularized synovial membrane and reduced cell apoptosis, the membrane becoming progressively fibrous [3]. A vicious circle develops between hemarthrosis and synovitis [4]. When bleeding occurs 3 times in the same joint within 6 months, the joint is called "target" [5].

We report a retrospective series of 12 ankle fusions performed in 9 hemophilic patients by the same surgeon (FB). The principal study objective was to report long-term results; the secondary objective was to assess the benefit of maintaining factor VIII concentration close to 100% by controlling clotting factors. The study hypothesis was that long-term results would be good, without major and notably hemorrhagic complications.

* Corresponding author.
E-mail address: Matthieu.ehlinger@chru.strasbourg.fr (M. Ehlinger).

http://dx.doi.org/10.1016/j.otsr.2017.08.016
1877-0568/

2. Material and method

2.1. Type of study and inclusion criteria

All patients with advanced hemophilic ankle arthropathy managed by fusion were included in a retrospective study. The study period was 14 years (2000–2013), with minimum 2 years' follow-up.

2.2. Assessment

Clinical assessment was based on preoperative and last-follow-up AOFAS hindfoot [6] and Olerud [7] scores. Pettersson radiologic score, recommended by the Orthopedic Advisory Committee of the World Federation of Hemophilia [8], was assessed preoperatively. Preoperative and follow-up radiologic assessment comprised 3 weight-bearing views: AP, lateral and Méary cerclage. Analysis concerned fusion rate and time to fusion and realignment quality on AP Méary view. Fusion was defined as complete bone continuity between tibia and talus on plain X-ray; CT was not systematically used. Fusion positioning was considered good when the tibial axis projected to the junction between the mid-third and lateral two-thirds of the weight-bearing zone.

2.3. Patients

The series comprised 9 patients, a mean age of 39 years (range, 19–58 years). Twelve ankle fusions (7 right, 5 left) were performed by the same surgeon (FB). Eight patients had severe hemophilia A and 1 minor hemophilia A. Patients had been symptomatic for a mean 14 years (range, 4–40 years) before surgery. Mean preoperative AOFAS score was 22/100 (range, 2–55) and Olerud score 37/100 (range, 20–85). Mean Pettersson score was 7 (range, 5–12). One patient (n° 1, ankles n^{os} 1 and 2) had preoperative bilateral but completely asymptomatic radiologic subtalar joint involvement. Adjacent joints were likewise asymptomatic in all cases. Ten total knee replacements were performed in 6 patients; in the other 3, knee radiographs were normal but meniscectomy was performed. One total hip replacement was associated, and there were also 2 asymptomatic radiologic hip lesions (Table 1).

2.4. Coagulation control

A pharmacokinetic test was systematically performed during the weeks before surgery, with injection of 50 IU/kg factor VIII to determine the recovery rate to be used to adapt the initial rate of continuous perfusion. The aim was to achieve the factor-VIII levels recommended by the *Coordination médicale pour l'étude et le traitement des maladies hémorragiques constitutionnelles* (COMETH [Medical Coordination for the Study and Treatment of Constitutional Hemorrhagic Diseases]) for orthopedic surgery: i.e., 80–100% during the first postoperative week [1]. Daily clinical and biological follow-up was provided by the hemophilia reference physician of the Regional Hemophilia Treatment Center (*Centre régional de traitement de l'hémophilie* [CRTH]). The following parameters were analyzed: blood count, prothrombin and fibrinogen levels, presence of a factor-VIII inhibitor, and factor-VIII level. Two administration protocols were implemented: continuous perfusion, and bolus injections; the choice between the two was left up to the CRTH physician after consultation with the patient and analysis of biology data.

Continuous clotting factor perfusion was used in 8 fusions for a mean 7 days (range, 5–9 days).

Bolus injection was used in 4 fusions:

- 1 patient (n° 3) had an injection every 8 hours on the day of surgery and the following day, then every 12 hours the next days (and had had continuous perfusion for fusion of the other ankle);
- 1 patient (n° 9) with minor hemophilia had injection every 12 hours;
- 1 patient (n° 8, bilateral fusion) with a factor-VIII inhibitor (37 Bethesda units/mL) had factor-VII injection.

Preoperatively, the severe hemophilia A patients had a mean factor-VIII level less than 1% (range, < 1–3%), whereas the normal level would be 50–150%. At the moment of surgery, factor-VIII level was boosted to 90% (range, 24–117%), reaching 97% (range, 63–195%) at end of surgery and 109% (range, 75–152%) the following day; levels were 99% (range, 52–134%) on postoperative day 2, 102% (range, 42–131%) on day 3 and 105% (range, 87–130%) on day 4 (Fig. 1).

2.5. Surgical technique

Two open ankle fusion techniques were used, with a pneumatic tourniquet on the thigh: Méary technique (9 ankles) and Crawford-Adams technique (3 ankles) [9,10]. The latter was chosen in case of bone contact between the lateral malleolus and calcaneus. Tibiotalar fixation used a 6.5 mm cannulated screw and fibular fixation used 3.5 mm screws. No grafts were performed. Percutaneous lengthening of the calcaneal tendon was necessary in 1 case (patient n° 9) to achieve 90° ankle positioning for surgery. 6 weeks' ankle-cast immobilization without weight-bearing was prescribed, followed by 6 weeks' cast immobilization with weight-bearing.

2.6. Statistical analysis

Statistical analysis was basically descriptive, but quantitative data (clinical scores) were compared on Wilcoxon test, with a significance threshold of $P < 0.05$ (SAS software).

3. Results

Eleven ankles (91.7%), in 8 patients, were followed up for a mean 8 years (range, 2–16 years). One patient died after 18 months' follow-up.

3.1. Complications

There were no intraoperative complications.

There were no postoperative hemorrhagic or infectious complications. No difference in hemorrhage risk could be shown between protocols. One patient (ankle n° 7) developed symptomatic secondary joint involvement (Fig. 2), but no revision procedure was undertaken due to the patient's obesity (BMI = 33 kg/m^2) and dietary non-compliance. In 1 patient (n° 1, ankles n^{os} 1 and 2) with bilateral infraclinical preoperative subtalar joint involvement, complementary fusion was performed 2 years after ankle fusion, with good clinical outcome at last follow-up.

3.2. Clinical assessment

Mean AOFAS score was 69 (range, 35–92) and mean Olerud score 70 (range, 30–100).

Seven of the 8 patients (87.5%) reported resolution of pain.

Seven of the 11 ankles (64%) (6 patients) showed no limp. Hindfoot motion was normal in 3 of the 11 ankles (24%) (3 patients), limited in 4 (36%) (3 patients) and severely limited in 4 (36%) (4 patients).

Please cite this article in press as: Eichler D, et al. Ankle fusion in hemophilic patients. Orthop Traumatol Surg Res (2017), http://dx.doi.org/10.1016/j.otsr.2017.08.016