

Christian Carulli¹
Silvia Linari²
Roberto Civinini¹
Massimo Innocenti¹

¹Orthopaedic Clinic,
University of Florence
(Italy)

²Center for Bleeding
Disorders, Careggi
University Hospital
Florence, Italy

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e-mail: ojhm@hemonline.it
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Case Report

Simultaneous bilateral total knee arthroplasty in a subject with severe haemophilia A

Abstract

Haemophilia is a rare blood disorder leading to haemorrhages and haemartrosis, and causing the so-called haemophilic arthropathy. Knees, ankles, elbows, and hips are the target joints. Knee arthropathy is specifically the most frequent and particularly debilitating for young haemophiliacs. Total Knee Arthroplasty is the gold standard of treatment in severe cases without relief after conservative treatments. A 51-year-old man affected by severe type-A Haemophilia was treated with a bilateral simultaneous Total Knee Replacement. Surgery was performed under prophylaxis with recombinant factor VIII, antibiotics, and general anaesthesia, after a careful multidisciplinary evaluation and full information to the patient. The outcomes were very good to the complete satisfaction of the patient at almost 10 years after surgery.

To date, it represents the first long-term report about a simultaneous bilateral Total Knee Arthroplasty in our country in a haemophilic patient and one of the very few successful and uncomplicated cases in literature.

Key-words

Total Knee Arthroplasty, Haemophilia, Haemophilic Arthropathy, bilateral joint replacement

Introduction

Haemophilia is an uncommon inherited disorder consisting of a lack of a specific coagulative factor, leading to haemorrhages and haemartrosis in male subjects since childhood [1]. Even though the modern haematological prophylaxis has significantly limited the haemorrhages, articular bleedings represent the most common complication, inducing severe degenerative changes in joints [1,2]. Knee arthropathy represents the most typical pattern: several non-surgical approaches have been reported during the last few years with high rates of success at early stages [3-5]. However, advanced stages may need a Total Knee Arthroplasty (TKA) to ensure relief from pain and functional recovery [6,7]. Haemophilic arthropathy is mostly bilateral but a simultaneous surgery is not generally feasible due to the higher rates of complications related to Haemophilia [8,9]. Thus, a one-stage bilateral TKA should be reserved to selected patients after a meticulous collegial assessment. This is the report of the first bilateral simultaneous TKA performed in our country in a Haemophilic patient with a long-term successful follow-up.

Case report

A 51-year-old man affected by severe A-type Haemophilia (FVIII:C <1%) and non-active C-type hepatitis was seen at our multidisciplinary office after years of treatment with analgesics, viscosupplementation, and a prophylaxis with recombinant FVIII concentrates. He referred a progressive functional impairment and continuous pain during daily life activities. When he was 21 years old, he underwent an open synovectomy on his left knee with acceptable results for several years. Both knees presented a severe flexion contracture,

a varus malalignment with coronal instability, and a symptomatic crepitus. The Range of Motion (ROM) of the right and left knees was 5°-75° and 10°-70° respectively. The Knee Society Score (KSS) at last pre-operative visit was 32 for the right knee, 27 for the left knee [10]. The Petterson score, a radiographic-based scoring system specific for haemophilia, was 11 for the right knee and 12 for the left knee [11]. After a careful assesment of the general health status and given an exhaustive informed consent to the strongly motivated patient, and respecting the principles of the Declaration of Helsinki, the multidisciplinary team decided to perform a one-stage bilateral TKA. The surgical procedure was performed by the Senior Surgeon (MI). The preoperative prophylaxis was conducted by a preoperative bolus of 5000 units of recombinant FVIII concentrate, 1000 mgs of tranexamic acid, and antibiotics (teicoplanin 400 mgs + amikacin 500 mgs). A general anaesthesia was induced; two separate operative draped fields and a tourniquet applied bilaterally, but singularly inflated. On the right knee an anterior longitudinal standard approach and a medial parapatellar capsulotomy were performed. The second surgical phase on the left knee was conducted using the previous scar of the open synovectomy during the wound closure of the other knee (Figure 1). The implant was the Genesis II system (Smith & Nephew, Memphis, TN), chosen for its peculiar features in haemophilic patients, as previously reported [6]. Posterior-stabilized cemented femoral components, fixed bearing tibial components with

cemented trays and pressfit keels, and cemented patellar components were bilaterally implanted. The surgical time was 60 minutes for the right knee (tourniquet time: 40 minutes; estimated blood loss: 250 cc), and 70 minutes for the left one (tourniquet: 50 minutes; estimated blood loss: 300 cc). All components were of equal size. At the end of both procedures, wound drains and compressive tapings were positioned (Figure 2). The postoperative management consisted in blood transfusions (two bags) and a tailored haematological prophylaxis. Antibiotics were maintained for three postoperative days and parenteral paracetamol and morphine were also administered. A daily evaluation of blood concentrations of the FVIII was tested. No intraoperative or postoperative complications were reported. Drains were removed on the first postoperative day (blood loss: 150 cc on right knee, 200 cc on the left knee) and active motion of the ankles and isometric quadriceps contractions were explained to the patient. Two days after surgery, a Continuous Passive Mobilization (CPM) protocol was introduced with progressive intensity. The patient reached a painless ROM of 0°-50° bilaterally on the fifth postoperative day, and was able to move independently in a wheelchair. After a week, the patient was discharged, and transferred to the institutional rehabilitation ward in the same facility. Three weeks after surgery, the patient was discharged with ROM 0°- 90° at right knee, 0°-100° at the contralateral and a good gait ability with a single crutch.



Figure 1: Preoperative clinical (a) and radiographic (b) aspect.



Figure 2: Intraoperative images before the osteotomies and after implant (a); end of surgery (b).

The first follow-up visit was performed one month after surgery: ROM 0°-95° at right knee, 0°-100° at the left one, no pain during daily activities, full recovery of daily activities, and return to work. The KSS reached a score of 82 for the right knee, 85 for the left knee. At the latest assessment (9.4 years after surgery), the patient is very happy, and able to move and work significantly better than before surgery (Figure 3).

Discussion:

Orthopaedic surgery in haemophiliacs is technically demanding. In case of a bilateral involvement, the patients' comorbidities or risks frequently limit the opportunity to perform a simultaneous procedure. TKA is one of the most successful procedures of modern Orthopaedics, with good long-term clinical results and survival in haemophiliacs [7,8]. Similar outcomes have been reported for simultaneous bilateral TKAs, with a substantial reduction of the risks and costs in patients affected by Osteoarthritis: very few publications deal with haemophilic arthropathy [12,13]. Reichel and colleagues reported about 6

cases treated with a one-stage bilateral TKA with variable functional outcomes, and significant rates of complications: they suggested strict indications due to high risk of complications [9]. Frauchiger and colleagues reported a case of a simultaneous bilateral TKA in a 40-year-old haemophiliac with inhibitors complicated by an aneurysm of the popliteal artery that requested further surgery [14]. Recently, Thes et al. reported a comparison between two groups of haemophiliacs undergoing simultaneous (5 patients) vs. staged (12 patients) TKA with favorable clinical result outcomes in terms of costs for the former group with respect to the latter [15].

In selected cases of motivated subjects with Haemophilia, balanced comorbidities, and with the absolute agreement of all specialists of the Haemophilia Treatment Center, a bilateral TKA procedure may be considered a valuable option. The advantages may consist of a single surgical procedure, faster recovery, reduction of costs for the haematological prophylaxis and hospitalization; the high risk of complications can be avoided only thanks to the close cooperation between Specialists



Figure 3: Last follow-up (more than 9 years after surgery): clinical (a) and radiographic (b) aspects

in dedicated Centers and full information given to patients, both keys to success in such challenging cases.

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