



IMPLANTATION OF THE TOTAL KNEE ENDOPROSTHESIS IN PATIENT WITH HAEMOPHILLIA B – A CASE REPORT

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ABSTRACT

Inherited bleeding disorders constitute a group of rarely occurring coagulation disturbances caused by deficiency or improper function of blood clotting factors. The clinical course of inherited bleeding disorders is dominated by spontaneous bleeding episodes, leading to irreversible destruction of affected joints. Osteoarthritis is a multi-factorial disease that is the result of mechanical and biological changes in the cartilage and bones beneath the cartilage. Osteoarthritis of the knee is the leading cause of chronic disability in later life. Knee arthropathy is a frequent complication affecting hemophilic patients, which can cause severe pain and disability. When conservative measures fail, total knee arthroplasty may be performed. Total knee arthroplasty is the only way to eliminate the progressive pain and improve the quality of life in end-stage arthropathy of the knee joint. After appropriate medical optimization and with prompt rehabilitation, total knee arthroplasty can be performed in hemophilic patients with good clinical results and survivor rates comparable to nonhemophilic patients.

KEYWORDS: Hemophilia, knee osteoarthritis, inherited bleeding disorder, total knee arthroplasty.

INTRODUCTION

Inherited bleeding disorders (IBDs) constitute a group of rarely occurring coagulation disturbances caused by deficiency or improper function of blood clotting factors. The three most common inherited bleeding disorders are hemophilia A, hemophilia B (Christmas disease), and von Willebrand disease (VWD), which are triggered by the absence of coagulation factor VIII (FVIII), coagulation factor IX (FIX), and von Willebrand factor (VWF), respectively. The clinical course of IBD is dominated by spontaneous bleeding episodes, leading to irreversible destruction of affected joints. Severe arthropathy remains the major cause of morbidity for hemophilia, and the knee is the most commonly involved joint.^[1,7] Osteoarthritis is a multi-factorial disease that is the result of mechanical and biological changes in the cartilage and bones beneath the cartilage. The main symptoms of osteoarthritis are pain and loss of function of the affected region. Knee osteoarthritis is the leading cause of chronic disability in later life. If the cause is unknown then we are talking about the primary osteoarthritis, and sometimes that trigger is obvious, such as trauma, so we are talking about a secondary osteoarthritis.^[8] For more than 40 years, radiological

classification of osteoarthritis introduced by Kellgren and Lawrence was accepted as a "gold standard." It is based on a comparison of radiographic shots of the joint and joint regions. It is classified: First degree - narrowing of the joint space and possible creation of osteophytes. Second degree represents osteophyte occlusion and possible hinging of the joint. Third degree shows multiple medial osteophytes, hinging of the joint, with bone sclerosis and possible bone defects. Fourth degree shows large osteophytes with significant narrowing of the joint space and bone sclerosis.^[9] Knee arthropathy is a frequent complication affecting hemophilic patients, which can cause severe pain and disability. When conservative measures fail, total knee arthroplasty (TKA) may be performed. Total knee arthroplasty is the only way to eliminate the progressive pain and improve the quality of life in end-stage arthropathy of the knee joint. According to previously published reports, it can offer satisfactory results in majority of hemophilia patients, with 10-year prosthetic survival rate of 83–92%.^[10-15]

CASE REPORT

Patient M.S., born on 1991. with hemophilia B has been a patient of a Clinic for Orthopaedic and Traumatology

of the University Clinical Center Sarajevo for numerous years. During the conservative treatment period, clinical picture of the patient was worsening and X-ray picture resulted in stage IV of Kellgren classification (Fig 1.).

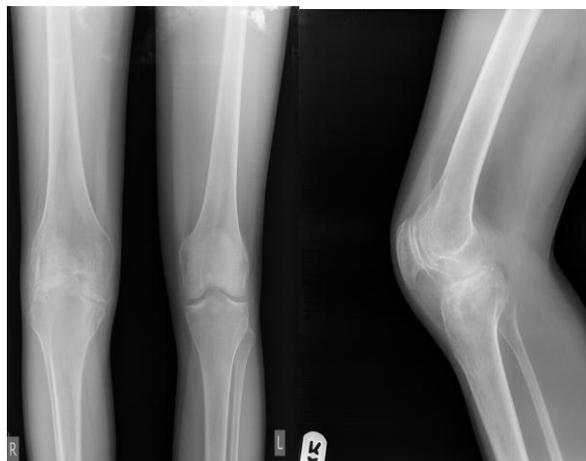


Figure 1. Preoperative X-ray of the patient's knee.

After all the conservative treatment methods for gonarthrosis have been worn out, the operative treatment of the knee endoprosthesis implantation was the upcoming step. At the admittance at the Clinic for Orthopedics and Traumatology of the University Clinical Center Sarajevo, patient was evaluated by a multidisciplinary team constituted by a hematologist, an orthopedic surgeon, internal medicine specialist, anesthetist and a physical medicine and rehabilitation physician.

Immediately after the admittance, hematologist preparation of the patient began with everyday check up of the INR and A-PTT and administration of appropriate dose of factor IX therapy according to the values of INR and A-PTT. During the period of the patient optimization, the strict operative procedure was planned out as well as follow-up period. For the needs of operative and postoperative period in total of 100 000 I.U. of FIX have been prepared. After the optimization period, in the morning of the operation patient was administered with 6500 I.U. of FIX and with 500 I.U. for the every hour of the operative procedure. The procedure was carried out with standard surgical technique, through a medial parapatellar approach. The extensiveness of the treatment, including type of the prosthesis and degree of its constraint, depended on the level of intraoperatively assessed joint damage. In this particular case, Zimmer Nex-Gen endoprosthesis was implanted together with appropriate antibacterial cement and postoperative antibacterial treatment for prolonged period.^[16,17] A tourniquet was not used, and suction drainage was applied post-operatively for 48 hours. During the operation, samples for microbiological analysis were collected. Patient received peri-operative antibiotics in accordance with actual recommendations,^[18] and anti-thrombotic prophylaxis was not administered. In the postoperative period, patient was transferred at the

Intensive Care Unit for a supervision of the hematologist, orthopedic surgeon and physical therapy specialist. Twelve hours after the surgery, 5000 I.U. of FIX were administered with regular check up of the INR and A-PTT. For next two days patient received 4000 I.U. of FIX with constant monitoring of the hematologist. Wound was healed regularly with no signs of local infection. The physical treatment began immediately on the first postoperative day in terms of breathing exercises, passive and active exercises for the operated knee and verticalisation with full-weight bearing on the operated leg using crutches. The rehabilitation therapy was done twice daily in terms of increase of the range of motion in the knee. After the critical period of 72 hours postoperatively, patient was transferred at the department for the continuation of the physical therapy. This type of therapy must be carefully dosed, adjusted individually and supervised by experienced physical therapy specialist with constant check-ups,^[19,20] During the hospitalization, patient was monitored by the hematologist, orthopedic surgeon and physical therapy specialist. Sutures were removed 12th postoperative day with no local signs of infection (Fig.2).



Figure 2: Postoperative X-ray with total knee endoprosthesis.

DISCUSSION

Hemophilia patients are usually young at the time of operation and therefore more likely to need further reoperations, but they expect normal activity with pain-free joints, functional independence, and participation in social activities without constraints. As a result, although total knee arthroplasty remains the treatment of choice in patients with advanced, painful, and disabling knee joint, it carries a higher risk of complications in patients with co-occurrence of blood clotting disturbances, as confirmed by Cancienne et al.^[1] who found significantly higher rates of infections, venous thromboembolism, transfusion of blood products, medical complications, and revision TKAs in patients with IBD when compared with a cohort of patients without bleeding disorder. Given that IBDs are extremely rare and that joint replacement procedures in hemophilia patients are being conducted in only a few highly specialized orthopedics centers in selected countries, the worldwide experience and professional literature on this issue are very limited.

After appropriate medical optimization and with prompt rehabilitation, TKA can be performed in hemophilic patients with good clinical results and survivor rates comparable to nonhemophilic patients.

CONCLUSION

In light of the above results, avoidance of infection is paramount during the surgery, and the role of the surgeons performing the initial operation should be emphasized. By our opinion, all implantations of the total-knee endoprosthesis procedures in hemophilia patients should be performed only by highly skilled surgeons, in close cooperation with hematological specialists. Consistent with past studies, we also believe that all patients should be educated on the importance of meticulous antisepsis with self-infusions, regular medical check-ups, immediate reporting of any signs of infection, and the use of prophylactic antibiotics before procedures with potential for contamination, including dental work, to avoid late-onset infections.

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