

Multidisciplinary Treatment of a Haemophilic Person with Total Hip Replacement Surgery- A Case Report

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Abstract

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Background: Treatment of hemophilic arthropathy may be conservative and operative. Conservative therapy includes medications and physiotherapy, and surgery includes synovectomy, tendon release, capsulotomy, osteotomy and total joint arthroplasty. During physiotherapy, surgery and postoperative rehabilitation haemophiliacs need to receive clotting factor.

Aim: We present our first experience in multidisciplinary treatment of a patient with severe haemophilia A during and after total hip replacement surgery, the patient's outcome after postoperative rehabilitation treatment and 6 months later.

Material and Methods: In a patient, age 28, with severe Haemophilia A, hip replacement surgery with total hip cementless prosthesis on his left hip was undertaken. Prophylaxis was done with concentrate of human coagulation factor VIII. Patient assessment was made with clinical findings, VAS for pain, Harris Hip Score for surgery outcome.

Results: The postoperative rehabilitation treatment program consisted of exercise therapy, occupational therapy and education. After the rehabilitation Harris Hip Score was 79 points, and 89 after six months follow-up.

Conclusion: The arthroplasty of the hip can be a valuable option in the management of severe hemophilic arthropathy. The successful outcome of replacement surgery in haemophilia depends upon a close collaboration between the orthopaedic surgeon, hematologist/specialist of transfusion medicine, physiatrist and other team members.

Introduction

People with haemophilia (PWH) suffer from repeated bleeding episodes into the "target" joints. The most frequently affected joints are knees, elbows, and ankles [1].

This bleeding is painful and leads to long-term inflammation and deterioration of the joints, resulting in synovial hypertrophy, cartilage damage, narrowing of the joint space, and changes in bone structure. [1]. It leads to permanent deformities, limited range of motion, muscle wasting, misalignment, loss of mobility, and extremities of unequal lengths [2].

This kind of impairments of the musculoskeletal system causes problems for PWH in their activities of daily living, work, education and hobbies [2].

People with haemophilia need multidisciplinary team care. This team is consisted of a specialist of transfusion medicine/ hematologist, pediatric hematologist, orthopedic surgeon, physiatrist, nurse, physiotherapist, psychologist, social worker, dietician, vocational counselor and etc [3].

Primary prophylaxis, aiming to avoid recurrent joint bleeding is the most appropriate way to prevent this threat [4]. It has provided considerable improvement in the prognosis of patients with severe haemophilia [5]. On-demand therapy (treatment given when bleeding occurs) is effective in controlling acute bleeding but cannot halt the ongoing joint destruction process [6].

Pain and inflammation in hemophilic arthropathy may be reduced with non-steroid anti-inflammatory drugs [7, 8], physical therapy [7], and

aquatic training [9]. The aims of the rehabilitation procedure are prevention and reduction of joint and muscle contractures, improvement in muscle strength and coordination, pain reduction, improvement in functional status, education and improvement of quality of life [10].

Surgical treatment is considered when there is at least one single remarkable sign of chronic arthropathy and depending on what stage of the joint destruction the patient asks for medical treatment. Radiosynovierthesis, arthroscopic and open synovectomy, reduction of the osteophytes and removing of loose bodies, arthroplasty and arthrodesis are the main surgical options for the chronic haemophilic arthropathy [11]. Total joint replacement is used more often on knee and hip joints that may improve the quality of life of these patients [11, 12]. The postoperative rehabilitation is crucial for better outcome of these patients [13].

The Center for Haemophilia is part of the Institute of Transfusion Medicine in Skopje, Macedonia. Haemophilia Care in Macedonia is based on the collaboration between the Institute of Transfusion Medicine, the Clinic of Hematology and the Department of Hematology in Children Hospital. The Center for Haemophilia is a Day Hospital for adults who need treatment for bleeding in joints and muscles, as well as for prophylaxis before tooth extraction. The Center is responsible for Home treatment. Other obligations of the Center for Haemophilia are: laboratory diagnosis of bleeding disorders, registration of people with haemophilia and other bleeding disorders in the National Registry, regular check up of people with haemophilia and other bleeding disorders, centralized factor concentrates supply for the whole country, and protocols for secondary prophylaxis due to physiotherapy or surgical interventions for adults. The Center for Haemophilia collaborates very closely with hematologic pediatricians, hematologists, orthopedic surgeons, physiatrists and dentists, on daily basis.

The purpose of this article is to present our first experience in multidisciplinary treatment of a patient with severe haemophilia during and after total hip replacement surgery, his outcome after postoperative rehabilitation treatment and 6 months later.

Case description

A patient K. Dz, 28 years old, body weight 60 kg, education elementary school, unemployed, who lives with his parents in a village, was admitted in the Institute of Physical Medicine and Rehabilitation for rehabilitation care after total hip replacement surgery on his left leg.

He has severe Haemophilia A with factor VIII basic level of 0.22% (normal values 50-150%). The diagnosis of haemophilia was confirmed in early

childhood, at three months of age, during surgical treatment of inguinal hernia. He also had spontaneous joints bleeding after different type of injuries. In his early age he was treated with cryoprecipitate, produced in the Institute of Transfusion Medicine, Skopje. Prophylaxis before any medical intervention and tooth extraction was performed with cryoprecipitate.

After the age of 10, he developed muscle weakness of lower extremities, limping and pain in his right thigh. Pain and limping were growing during the next years, but he never asked for medical care. In some occasions, he was treated with concentrate of clotting factor VIII in the General Hospital in Struga. He has never been on rehabilitation treatment.

In the last 4 years the difficulties increased, with extremely severe pain in both hips, and leg muscle weakness, so this patient with haemophilia was not able to walk and to transfer from a lying to standing position. He only was able to bend over and sit. He was spending his time only at home, he never left the premises hence, his quality of life was determined as poor.

He was examined in the Special Hospital for Orthopedics and Traumatology "St. Erasmo" in Ohrid. Following clinical and radiological examination, the diagnosis of hip osteoarthritis on both legs was established and a surgical treatment was recommended.



Figure 1: Osteoarthritis of the left hip.

Soon, the surgery intervention with total hip replacement on the left leg was performed in the Clinic for Orthopedic Surgery in Skopje. The assessment was made by taking a history, clinical findings, Visual Analog Scale (VAS) for left hip pain, Harris Hip Score [14] and standard radiography of the hips.

Before the surgery VAS for left hip pain was 10, and Harris Hip Score was 39 points. Preoperative radiologic findings of the left hip presented severe osteoarthritis (narrowing of the joint space and deformity of the femoral head) (Figure 1).

The standard lateral approach to the left hip was used. The implanted total hip endoprosthesis was cementless.



Figure 2: Total hip cementless arthroplasty of the left hip.

Prophylaxis was done with plasma derivate concentrate of human coagulation factor VIII, according to the protocol of the World Federation of Haemophilia. Forty-five minutes before the start of surgical intervention 3000 IU of factor VIII concentrate were administrated (50 IU/kg/BW). After the initial dose, he was treated with 1500 IU (25 IU/kg) every 12 hours until the fifth postoperative day. In spite of factor VIII level range of 73%-101%, he still had a huge

hematoma on the operative place. Because of that, the dose of 2000 IU was given three times a day until the ninth postoperative day. Factor VIII level was in range of 98-100%. From the tenth to the fourteenth postoperative day, the dose was decreased to 1000 IU, twice a day. Factor VIII level was in the range of 47-57% (sufficient level for second postoperative week).

Enoxaparin 20 mg s.c. as thromboprophylaxis was administrated during 12 postoperative days. Early postoperative rehabilitation program began on the first postoperative day. It included deep-breathing exercises, ankle pumping exercises, isometric exercises, and ambulatory training with crutches. There were no early postoperative complications.

VAS for the left hip pain after the total hip replacement was 0. The postoperative radiological findings of the left hip presented total hip cementless arthroplasty (Figure 2).

The patient was admitted in the Institute of Physical Medicine and Rehabilitation for postoperative rehabilitation treatment on the fourteenth day after surgery. He suffered from small pain in his right hip and left knee.

Patient's assessment was made with clinical findings, measurement of the length of the legs, measurement of the range of motion in the joints, measurement of the circumference of the legs, pain assessment with Visual Analog Scale (VAS), and Harris Hip Score. The assessment was made at admission, at discharge and at follow-up after 6 months.

At baseline the patient's clinical findings were as follows: his left leg was shorter 1 cm (umbilicus-malleol), with muscle weakness, with small restriction in range of motion in the left hip: flexion with extended knee was 60°, flexion with flexed knee was 55°, extension 0°, abduction 30°. The movements in the right hip were: flexion with extended knee 60°, flexion with flexed knee was 55°, extension 0°, abduction 30° (Table 1).

The knees were with slight enlargement on the left side, with no local pain on palpation or effusion, with normal range of motion on both sides and persistence of crepitus in the left knee. The movements in both ankles were: dorsal flexion 10°, plantar flexion 30°. His gait was with crutches with no weight-bearing on the left leg. VAS for right hip was 3. VAS for the left knee pain was 3. Harris Hip Score was 71 points.

Upper extremities were with no changes of the elbows on inspection, and no local pain on palpation. Muscles weakness was evident. The motions of both shoulders were in full range, both elbows flexion was in full range, but extension was limited to -10°, in wrists and joints of the both hands were in full range. He was able to perform the activities of daily living with both arms.

Table 1: Range of motion in the joints of lower extremities

| Left | | | | Right | | |
|------------------|------------------|------------------|----------------------------------|------------------|------------------|------------------|
| ROM*** | ROM** | ROM* | Movement | ROM* | ROM** | ROM*** |
| 75 ⁰ | 70 ⁰ | 60 ⁰ | Hip - flexion with extended knee | 70 ⁰ | 80 ⁰ | 80 ⁰ |
| 90 ⁰ | 90 ⁰ | 55 ⁰ | Hip - flexion with flexed knee | 70 ⁰ | 90 ⁰ | 90 ⁰ |
| 0 ⁰ | 0 ⁰ | 0 ⁰ | Hip - extension | 0 ⁰ | 0 ⁰ | 0 ⁰ |
| 35 ⁰ | 35 ⁰ | 30 ⁰ | Hip - abduction | 30 ⁰ | 35 ⁰ | 35 ⁰ |
| 130 ⁰ | 130 ⁰ | 130 ⁰ | Knee- flexion | 130 ⁰ | 130 ⁰ | 130 ⁰ |
| 0 ⁰ | 0 ⁰ | 0 ⁰ | Knee - extension | 0 ⁰ | 0 ⁰ | 0 ⁰ |
| 10 ⁰ | 10 ⁰ | 10 ⁰ | Ankle- dorsal flexion | 10 ⁰ | 10 ⁰ | 10 ⁰ |
| 30 ⁰ | 30 ⁰ | 30 ⁰ | Ankle- plantar flexion | 30 ⁰ | 30 ⁰ | 30 ⁰ |

*Range of motion (ROM) at admission in the Institute of PMR; ** Range of motion (ROM) at discharge from the Institute of PMR; *** Range of motion (ROM) at follow-up.

The rehabilitation treatment program was consisted of exercise therapy for both legs (isometric and dynamic exercise for hip flexors, extensors and abductors; dynamic exercises for knee extensors and flexors, and calf muscles), education about restricted movements of left hip (to avoid hip adduction and external rotation, flexion past 90 degrees), closed-chain exercises standing, ambulatory exercise with crutches on the stairs; occupational therapy. For analgesic effects he received dyadynamic currents for the right hip and ionophoresis with 2% Novocain on the left knee. In the last days he trained ambulatory exercises with crutches with partial weight-bearing. He also received a booklet with exercises for haemophiliacs, published by the World Federation for Haemophilia. Some exercises suitable for a home exercise program were chosen from the booklet.

During the inpatient rehabilitation treatment, the patient with haemophilia received plasma derivate concentrate of human coagulation factor VIII, 1000 IU, as intravenous injection, early in the morning, from Monday to Friday (days of sessions). He underwent a rehabilitation program with 15 days sessions, five sessions per week.

Results

Ten days after the physical therapy treatment, the patient with haemophilia had no pain in the right hip and left knee (VAS 0). At discharge, fifteen days after the rehabilitation treatment, he was with improved muscle strength on the left leg, increased range of motion in the left hip (flexion with extended knee 70⁰, flexion with flexed knee 90⁰, extension 0⁰, and abduction 35⁰), his gait was with partial weight-bearing with crutches, and he was trained to climb the stairs. Harris Hip Score was 79 points.

At follow-up assessment, six months later, he had no pain, he was with improved muscle strength, increased range of motions in both hips, and his gait was without crutches at distances as long as he liked. He performed exercises regularly and he felt very well. Harris hip score was 86 points.

Discussion

Despite the tremendous benefit offered by primary prophylaxis, recurrent joint bleeding with progression to chronic synovitis and hemophilic arthropathy is still a daily concern for the multidisciplinary health care teams managing people with severe haemophilia or haemophilia complicated by inhibitor development. Regular assessment of musculoskeletal status allows for early detection of symptoms and consecutive implementation of appropriate physiotherapy and medical training could prevent advanced stages of arthropathy [1].

Despite the fact that our patient has severe haemophilia, with affection of knees, hips and elbows, he has never received rehabilitation treatment for arthropathy. Hemophilic arthropathy infrequently affects the hip joints [15]. The hip is considered to be one of the main load bearing joints in the body. In the people with haemophilia, joint bleeds can be catastrophic, leading to long term degeneration and accompanying arthritis [16].

Physical therapy and rehabilitation play a major role in both prevention and treatment of haemophilic arthropathy and are integral parts of multidisciplinary care of people with severe haemophilia [17]. Strength, flexibility, balance and proprioception are important factors in reducing the occurrence of injuries, and thus bleeds [18]. In all cases, the guiding principle of physiotherapy is that it should not induce pain [1]. Also, it's important first to relieve pain in arthopathic joints, and next to decrease the functional limitations [19]. Vallejo et al. in clinical study with 13 persons with haemophilia reported that 27 sessions, three times per week of aquatic training, had a positive effect on their motor performance and considerably improved their aerobic and mechanical capacity without causing adverse effects [9].

In people with haemophilia synovectomy, tendon release, capsulotomy, osteotomy and total joint arthroplasty are commonly used [7]. It is well known that the indication for elective orthopaedic intervention with total joint replacement is severe degenerative change in a joint with increasing disability and incapacitating pain [7, 13, 20]. The general surgical principles are the same but tissue

handling must be careful with particular attention paid to securing haemostasis [13, 20]. The total joint replacement surgery is technically demanding, and complications are commonly described, particularly of the hip and knee [11]. There are many studies that demonstrated the outcome after total knee arthroplasty, because the knees are one of the targets joints in people with haemophilia [11].

The implantation of a prosthesis is frequently indicated in young persons (the mean age at the time of knee replacement is about 20 years less than that of patients with osteoarthritis or rheumatoid arthritis). Therefore, the risk of wear or loosening of the prosthesis is increased [1].

Rehabilitation of people with haemophilia after a single joint procedure with an appropriate supply of concentrate is a challenging proposition [21].

Our patient with severe Haemophilia A received rehabilitation treatment on an inpatient basis. It included exercise therapy, physical agents, occupational therapy and education.

Despite the surgical technique, rehabilitation is the cornerstone of treatment for people with haemophilia to achieve favorable orthopedic results through the maintenance of muscular function by limiting the period of immobility [13]. The postoperative rehabilitation includes physiotherapy with exercise [22, 23], prescription of orthoses, walking aids and education [21]. The physiatrist takes all aspects of impairment, disability, functioning, environmental factors, personal factors into account when making the rehabilitation, diagnosis and treatment plan. A multidisciplinary team is indispensable in this process [2]. There is difficulty in providing adequate rehabilitation for people with haemophilia on an outpatient basis [21].

De Kleijn P. et al. emphasized the role of physiotherapy before and after elective orthopedic procedures to achieve optimal functional outcome and therefore optimal quality of life for people with haemophilia [24].

During the operative period and postoperative rehabilitation, our patient with Haemophilia received plasma derivative concentrate of human coagulation factor VIII. All rehabilitation and physiotherapy period has to be covered with factor concentrate [13].

Because of the pain, dyadynamic currents were applied on the right hip and iontophoresis with Novocain on the left knee for 10 days. After 10 days of applications, the pain in these joints disappeared. Bossard et al. emphasized that the electrotherapy was not sufficiently evaluated, and that in practice, only two physical agents can be used: analgesic electrical currents (cutaneous stimulation below the pain threshold), effective for chronic pain, and excitatory motor currents, mostly useful immediately after surgery [1].

Immediately after surgery, the reported person with Haemophilia experienced relief of pain in his left hip. Postoperative outcome after total hip arthroplasty was good, and Harris Hip Score was increased from 39 to 79 points after rehabilitation. Six months later, patient's outcome was good, assessed with 89 points of Harris Hip Score. Marchetti P. et al. have made a range of outcome after hip replacement surgery according to Harris Hip Score [25]. Total joint arthroplasties of knees and hips can improve quality of life by relieving pain and improving mobility [7].

A clinical study evaluated the medium-term follow-up results, effectiveness, and suitability of arthroplasty for hemophilic arthropathy in group of patients with 26 total knee and 9 total hip arthroplasty operations on hemophilic patients. After their operations, patients experienced relief from pain and intra-articular bleeding in affected joints but only marginal improvement in the range of motion. The authors believed that total knee and hip arthroplasty was a good solution for haemophilic arthropathy before severe deformity occurred [26].

Other clinical study reported the long term outcome (mean 132 months) after total hip replacement of 15 hips in 13 patients. The main bleeding disorders were Haemophilia A in ten patients and severe von Willebrand disease in three patients. They demonstrated good long-term results, with only one aseptic loosening after 14 years and one septic loosening after 14 months in a HIV-positive patient. The Harris Hip Score increased from 48 points (32-66) preoperatively to 89 (76-100) postoperatively [27].

The multicenter retrospective study analyzed the results of hip arthroplasty in 27 male patients and thirty-four arthroplasties. The mean age of the patients at the time of operation was thirty-eight years. The mean duration of follow-up was eight years, with a minimum of two years. There were twenty-six total hip arthroplasties performed with cement, six total hip arthroplasties performed without cement, one was hybrid arthroplasty, and one bipolar arthroplasty was performed with cement. There were no infections after these thirty-four primary arthroplasties. There were three late infections around prostheses inserted with cement, and led all to a resection arthroplasty. Six (21%) of twenty-eight cemented femoral components and six (23%) of twenty-six cemented acetabular components were revised because of aseptic loosening [15].

The most important is that a patient should be evaluated fully and surgical treatment will only be indicated if pain does not respond to conservative management or if deformity does not allow activities of daily living or generates pain in another anatomical region [13].

A multidisciplinary approach is required to decipher the complexities that led to the deterioration of the musculoskeletal function in order to produce a

functional individual, with the ultimate goal of achieving social integration [21].

The arthroplasty of the hip can be a valuable option in the management of severe haemophilic arthropathy. The successful outcome of replacement surgery in haemophilia depends upon a close collaboration between the orthopaedic surgeon, haematologist/specialist of transfusion medicine, physiatrist and other team members.

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