Original Article

RESULTS IN OSTEOARTICULAR SURGERY OF THE HAEMOPHILIAC PATIENTS

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Abstract

Hemophilia is a hereditary disease due to a defect of chromosome X, which lead to impaired production of coagulation factor VIII in hemophilia A (85% of cases) and factor IX in hemophilia B.

The evolution of hemophilic arthropathy is almost always from haemarthrosis to chronic synovitis and extensive erosion of the articular surface and, ultimately, the final stage of joint destruction - chronic haemophilic arthropathy.

The present paper aims to analyse results obtained in treating 105 patients that received surgical care (a total of 107 surgical interventions) within the Department of osteoarticular surgery for haemophiliacs, of the Orthopaedics-Traumatology Clinic II Timisoara, between year 2001 and 2012.

The treatment protocol was performed in collaboration with the team of hematology specialists from the Clinic of Pediatric OncoHematology - Pediatrics Hospital"Louis Ţurcanu" from TimiŞoara.

With substitution treatment and correct surgical indication, osteoarticular surgery applied to hemophiliacs, in a specialized centre, by a multidisciplinary team, can lead to good results with acceptable risks.

Keywords: *haemophilia, chronic arthropathy, bleeding, rare diseases*

Rezumat

Hemofilia este o boala ereditara datorata unui defect al cromozomului X, care determina o productie deficitara a factorului de coagulare VIII in hemofilia A (85% din cazuri) si a factorului IX in hemofilia B.

Evoluţia artropatiei hemofilice este aproape întotdeauna de la hemartroză la sinovită cronică şi eroziuni extinse ale suprafeţei articulare şi, în cele din urmă, la stadiul final de distrugere articulară – artropatia cronică hemofilică.

Lucrarea de fata analizeaza rezultatele tratamentului aplicat pe un lot de 105 pacienți operați (în total 107 intervenții chirurgicale) în Compartiment de chirurgie osteo-articulară al bolnavilor hemofilici din Clinica II Ortopedie – Traumatologie în intervalul 2001- 2012.

Tratamentul chirurgical al hemofilicilor se realizează în colaborare în echipa cu Clinica de OncoHematologie Infantilă de la Spitalul de Pediatrie „Louis Țurcanu" din TimiŞoara.

Cu un tratament substitutiv Şi o indicație chirurgicală corecte, chirurgia osteoarticulară la hemofilici, efectuată într-un centru specializat, de către o echipă pluridisciplinară , poate da rezultate bune cu riscuri acceptabile.

Cuvinte cheie: *hemofilie, artropatie cronică, hemoragie, boli rare*

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INTRODUCTION

Hemophilia is a hereditary disease due to a defect of chromosome X, which lead to impaired production of coagulation factor VIII in hemophilia A (85% of cases) and factor IX in hemophilia B. There are described three types regarding the severity of the disease: mild, with a concentration of deficient factor greater than 5% of normal values, an average of a concentration between 1 and 5% and a severe, with more than half of cases of hemophilia, with a concentration of factor VIII or IX less than 1% of normal. Hemorrhages in muscles and joints occur spontaneously in severe form, running a minor injury in the moderate and only after a major injury or surgery in mild forms. (1)

The evolution of hemophilic arthropathy is almost always from haemarthrosis to chronic synovitis and extensive erosion of the articular surface and, ultimately, the final stage of joint destruction - chronic haemophilic arthropathy. End stage of arthropathy is complicated by severe limitation of joint range of motion secondary to the arthrofibrosis due to the replacement of the hypertrophic synovium with a dense fibrosis. Severe contractures, angular

deformation and loss of bone substance due to mechanical abrasion and bony cysts are common.(2)

Patients with severe hemophilia have haemarthrosis especially in knee, elbow and ankle with appearance of hemophilic arthropaty. Knee is most commonly affected (30% followed by elbow (25% 0, ankle (15%), hip (5%) and other joints (<2.5%). (1) Arthritis modifications are seen in approximately 90% of affected joints even in moderate forms of the disease. Evolution of hemophilic arthropaty depends on patient age with a maximum deterioration in the second decade.

MATERIAL AND METHOD

The present paper aims to analyse results obtained in treating 104 patients that received surgical care within the Department of osteoarticular surgery for haemophiliacs, of the Orthopaedics-Traumatology Clinic II Timisoara, between year 2001 and 2012. This Compartment, the only one in our country, was founded in 2001, and was included in the Ministry of Health financing Programme for haemophilia.

In the mentioned time span, 158 patients from all over the country received a consult, of which 105 were further treated surgically resulting in a total of 107 surgical interventions.

Our patients were aged between 9 and 45 years old with a maximum between 11 and 20 years old. When discussing the number of joints that were affected by the disease: 43 patients had 3 or more, 36 patients had 2, and 26 patients had only one affected joint. Within our study group, 104 patients were suffering from hemophilia type A, 2 patients with hemophilia type B. We also included one patient with type III von Willebrand disease.

One of the particularities of hemophiliacs is the possibility of HVC/HVB infection or even HIV. In our study group 47 patients were found positive for HVC or HVB.

The investigations included X-ray , MRI and bone density tests for determining osteopenia or osteoporosis. We found that 53 patients were suffering from either ostopenia or osteoporosis.

We used Arnold and Hilgartner classification preoperatively, in order to stadialize the articular injury of our patients.

stage 0 : normal joint
stage I : no skeletal abnormalities, soft-tissue swelling is present
stage II : osteoporosis and overgrowth of the epiphysis, no cysts, no narrowing of the cartilage space
stage III : early subchondral bone cysts, squaring of the patella, widened notch of the distal femur or humerus, preservation of the cartilage space
stage IV : findings of stage III, but more advanced; narrowed cartilage space
stage V : fibrous joint contractures, loss of the joint cartilage space, extensive enlargement of the epiphyses with substantial disorganization of the joint

Table 1. Arnold and Hilgartner classification

We found 23% of the patients who underwent surgery in our Compartment to be in IV and V stages and 67 % of them in II and III stages.

We also used Petterson classification, which is presented in table 2.

Parameters	**Score**
Osteoporosis	0-1
Epiphiseal widening	0-1
Subchondral irregularity	0-2
Narrowed joint space	0-2
Subchondral cysts	0-2
Marginal erosion	0-1
Joint incongruence	0-2
Important dislocation, angulations	0-2

Table 2. Petterson classification

According to Petterson classification, all the patients had a score greater than 6.

RESULTS

Surgical interventions (107 interventions) performed in our Compartment are shown in table3.

Elbow sinovectomy	6
Radial head resection	4
Arthroscopic knee sinovectomy	55
Total knee arthroplasty (TKA)	10
Knee arthrodesis	6
Revision after total knee arthroplasty	1
Knee debridement (post infection)	1
Total hip arthroplasty	5
Arthroscopic ankle sinovectomy	3
Ankle arthrodesis	2
Neurolisys of cubital nerve	1
Resection of hemophilic pseudo tumors	2
Fracture stabilization	5
Supracondilar osteotomy	1
Other interventions –removal of implants	4
Von Willebrand disease- TKA	1

Table 3. Types of surgical interventions performed in our Compartment between 2001 and 2012

Removing the synovium from a joint in hemophilic patients is a major procedure of "unloading" the joint, because the hypertrophied synovium represent a source of hemorrhages due to its increased friability and decreased capacity of resorbtion.

Figure 1. Complete destruction of the cartilage in tibial plateau - arthroscopic image

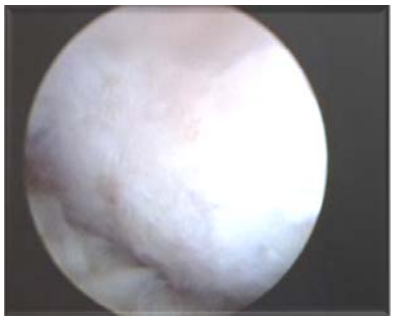


Figure 2. Chondropaty of the external femoral condile - arthroscopic image

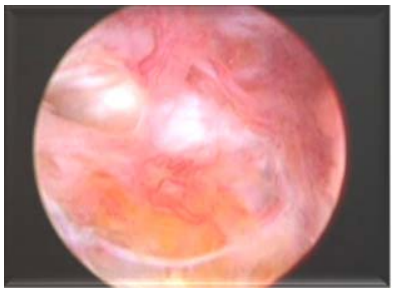
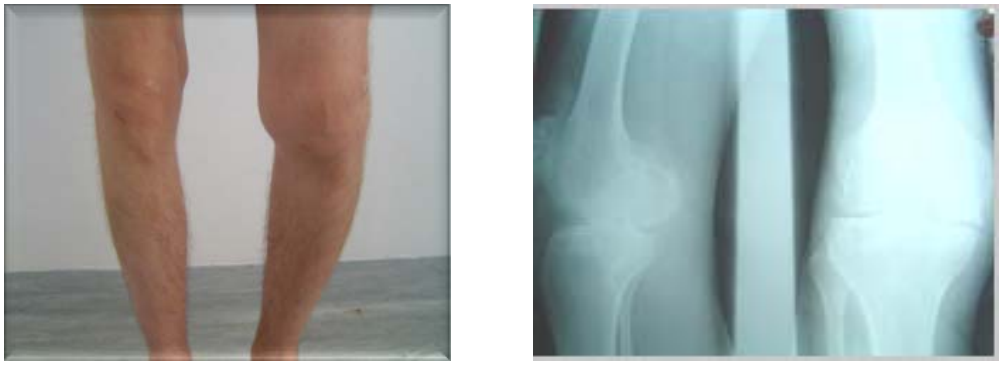


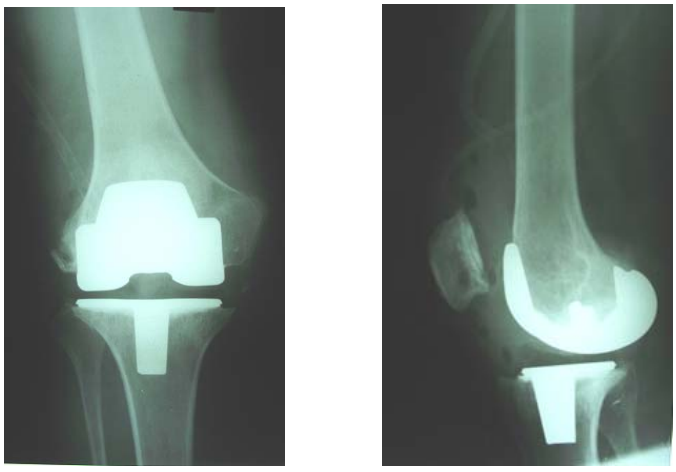
Figure 3. Hipertrofic synovitis - arthroscopic image

Total knee arthroplasty in hemophilic patients has indications which are severe pain, low range of motion, deformities, especially contracture in flexion and recurrent haemarthrosis.



(A) (B)

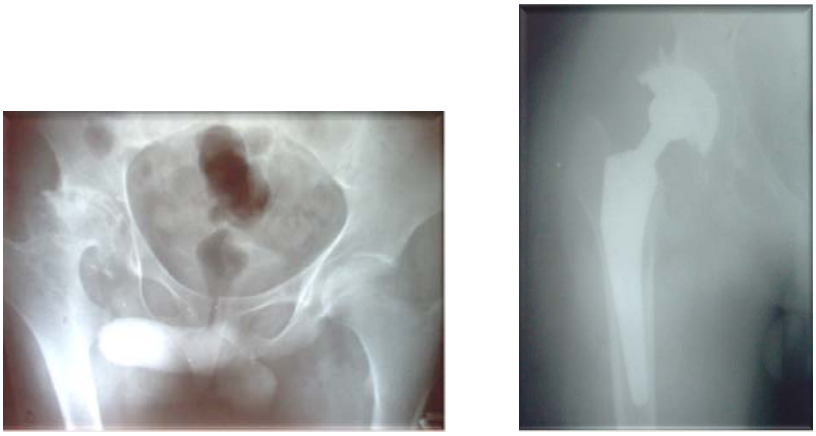
Figure 4. 42 years old patient with chronic hemophilic arthropaty
(A)Preoperative image (B)Preoperative X ray A-P and lateral



(A) (B)

Figure 5. Postoperative X ray: A-P (A) and lateral (B)

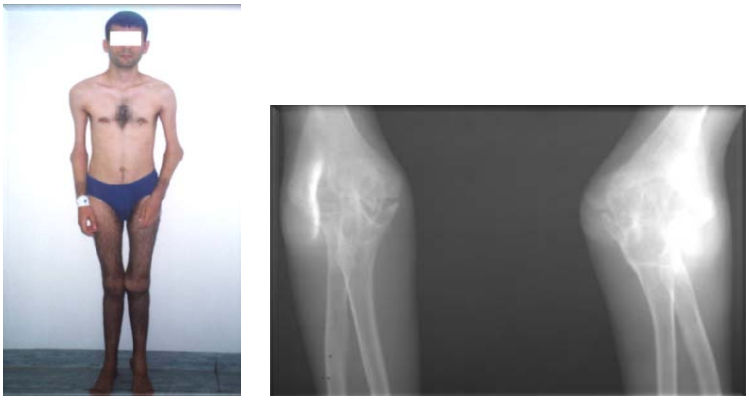
In patients with important lesions of the hip joint, total hip arthroplasty was extremely efficient.



(A) (B)

Figure 6. - Chronic hemophilic arthropaty of the hip,
(A) preoperative X-ray, (B) postoperative X-ray

31 years old patient- A type severe hemophilia, bilateral chronic hemophilic arthropaty shoulder -elbow-hip-knee-ankle.



(A) (B)

Figure 7. (A) Clinical aspect (B) preoperative X-ray, notice the widening and deformity of the epiphysis, disappearance of the normal joint space.

Open sinovectomy was performed using the shaver and radial head resection with ablation of the olecranon bursae. (figure 8)

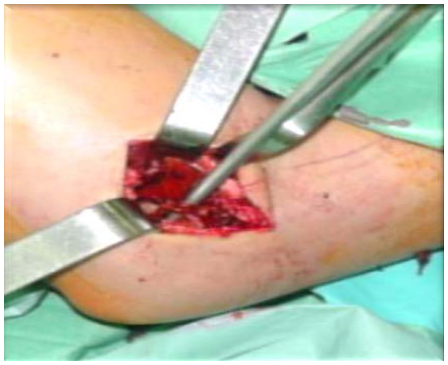
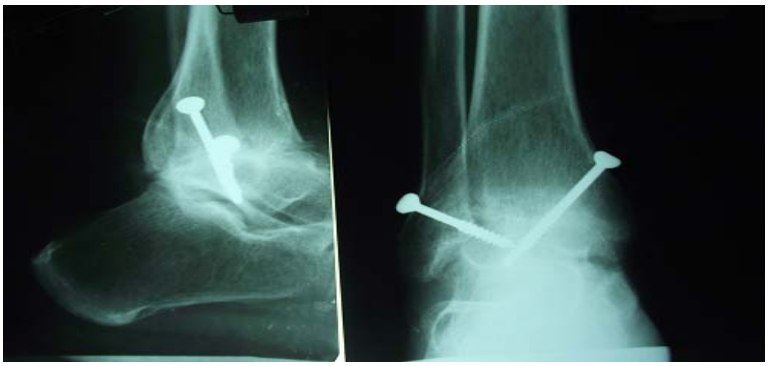


Figure 8. Intraoperative image

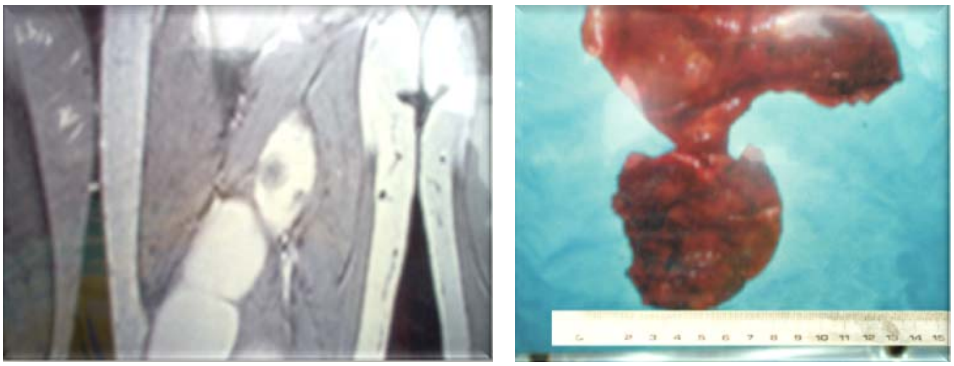
The resection of radial head was performed through a minimal anteroextern approach, preserving the annular ligament.

In cases of chronic arthropaty of the ankle, we performed sinovectomy and arthrodesis. (Figure 9)



(A) (B)

Figure 9. Postoperative X-ray showing ankle arthrodesis
(A) lateral image (B) A-P image



(A) (B)

Figure 10. Hemophilic pseudo tumor of tigh, resected
(A) MRI image, (B) intraoperative image of the resected specimen

In our study group, mean follow up period for patients who underwent knee arthroscopy was 18 month (6 to 36 month). Mean hospitalization period was 14, 6 days (7 to 21 days) and the recovery period was one month.

The treatment protocol was performed in collaboration with the team of hematology specialists from the Clinic of Pediatric OncoHematology - Pediatrics Hospital"Louis Țurcanu" from TimiŞoara.

COMPLICATIONS

Complications determined by anti-FVIII inhibitors made intra- and post-operatory administration of Novoseven necessary.

Septic complications: There was one septic complication in a patient with cronic hemophilic artropathy of the knee that underwent total knee artroplasty.

Treatment included debridement, antibiotherapy and revision artroplasty of the knee using revision prosthesis.

Mortality within our study group was zero.

DISCUSIONS

Osteoarticular surgery for hemofiliacs is highly pretentious, involving complex treatment applied in a specialised centre by a multi-disciplinary team that is well trained and has experience in this particular field.(14)

Sinovectomy as an isolated intervention was initially indicated for knee artropathy. Numerous studies were published comparing open and arthroscopic sinovectomy. Tryantafilou compared back in 1992, eight open sinovectomies with five done artroscopicaly and results clearly shows the benefits of arthroscopic techniques. (3) In 1996, Wiedel publishes a study with a follow-up of 10-15 years about the first arthroscopic vasectomies. Nine cases were monitored for that period and arthroscopy managed to maintain a low hemarthrosis rate.(4)

Complications involving arthroscopic surgery were described in 2 papers. In 1999 Heim reported 2 expansive hematomas at the arthroscopic portal site, and in 1992 he described an arteriovenous fistula after knee arthroscopy.(5)

Sinovectomy offers a lower hemarthrosis rate and pain relief with discrete or absent improvement of mobility. It is a low risk method with satisfying results. The only relative disadvantage is poor post-operatory bleeding control and the risk of additional hemarthrosis.

We found the shaver to be very efficient in removing the extremely brittle synovium, due to villous hypertrophy. We also successfully used it in open elbow sinovectomy with great results.

The elbow is a frequent location (second after the knee) for repeated bleeding followed by the widening of the radial head and arthrosis of the radio-capitellar and ulno-trochlear joints. The most disabling loss of movement amplitude with these patients is loss of supination. Possible supination lower than 45 degrees interferes wih the patient's abilities to feed, keep personal hygiene or other daily activities. In patients with post-operative limited prono-supination, removal of the radial head resulted in a 25 degree improvement (Poenaru et al) that brought significant improvement in their ability to deal with daily activities.(1)

Sinovectomy has proved to be useful in the ankle as well. Greene described results of ankle sinovectomy after failure of conservative treatment in 5 patients suffering from hemophilia that continued to present with hemarthrosis and palpable synovium hypertrophy. Post-operative follow-up has proven significantly lower hemarthrosis rate and also an improved range of motion in all patients.(6)

Indication for total knee arthroplasty in hemophiliacs is determined by pain and disabling loss of mobility that doesn't respond to conservative treatment. We

must keep in mind that end-stage arthropathy can be reached even during childhood. Severe pain and impairment are frequently present in the third or fourth decade of life.

Results include pain relief, discrete improvement of joint mobility, and deformity correction. Total knee arthroplasty in patients with hemophilia, although involving very high financial costs, allows improving quality of life in young disabled patients.

Severe hip arthropathy is less frequent than knee, elbow or ankle arthropathy. Radiologic aspect can take the form of juvenile arthritis or, more rarely, hip arthrosis. Coxa valga is the usual aspect; high intra-articular pressure secondary to hemarthrosis can lead to aseptic necrosis of the femoral head. Although these patients are more often too young for total hip arthroplasty , arthrodesis is not a viable indication due to associated knee pathology.

As opposed to the knee, cemented prosthesis does not show results as satisfying as with other types of arthropaties. Luck and colab. observed and described a unique aspect with hemophiliacs: due to ankle and knee arthrofibrosis these patients present with a stiff lower limb resulting in a distinct limp. This leads to higher forces being applied to the hip joint, together with function loss and low shock absorption provided by the impaired knee and ankle joints. The additional stress brought by this type of limp might be one of the causes for loosening of cemented prosthesis in hemophiliacs. (7) Another cause worth taking into consideration is the possibility of bleeding within the membrane formed where bone comes into contact with cement.

Fractures that appear in hemophiliacs raise discussions; some authors suggest that the main cause for the high rate of fractures in these patients is osteoporosis as well as low joint mobility.

Physical rehabilitation treatment was conducted in the Psychical rehabilitation Department from our Clinic and in the”Cristian Şerban” Centre for Hemophiliacs form Buziaş.

The rehabilitation protocols had two goals:

- First, tertiary prevention of disabilities by decreasing the deficiency caused by bleeding in joint or/and muscles

- Second, avoiding the handicap through actions which will have effects over the ostheoarticular disabling deficiencies

- rehabilitation procedures will address to muscular compartments which are affected by repeated bleeding and by atrophy due to decreased mobilization, as well as to the ostheoarticular compartment affected by spontaneous or provoked hemorrhages that occur in joint and will lead to various impairment forms of the joint, from chronic synovitis to chronic arthropaty.

Objectives of locomotory rehabilitation therapy are:

- To decrease the pain
- To increase the range of motion (ROM)
- Increase the muscle tonus and regain the joint stability
- To minimize the joint modifications such as ankilosis, fibrosis, ligaments and tendons shortening.
- Reintegration of the patients to normal life conditions

CONCLUSIONS

Hemoragic complications can be avoided with expert haematology monitoring.

Arthroscopic sinovectomy can be an indication in moderate stages of haemophiliac knee and ankle arthropathy. Arthroscopic procedures can be safely applied even in patients with a low concentration of inhibitors.

Knee arthrodesis remains an option in hemophiliacs with severe joint deformities.

Arthroplasty can be an indication even in young patients.

With substitution treatment and correct surgical indication, osteoarticular surgery applied to hemophiliacs, in a specialized centre, by a multidisciplinary team, can lead to good results with acceptable risks.

Pain, deformity, reduced excursion joint movements, muscular imbalance and consequent impairment of joint function are characteristic for more than 80% of patients with severe hemophilia.

Following the complex rehabilitation treatment we noticed overall reduction in the number of affected joints.

Recovery took place in several cycles and requires mandatory home rehabilitation program.

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