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OUR EXPERIENCE WITH ORTHOPEDIC SURGERY IN HEMOPHILIACS

Dan V. POENARU[1], Margit ŞERBAN[2], Ioan L. BRANEA[1], Jenel M. PĂTRAŞCU[1]

[1]Orthopedics and Trauma Clinic II, "Victor Babeş" University of Medicine and Pharmacy, Timişoara, Romania;
[2]Pediatrics Clinic III, "Victor Babeş" University of Medicine and Pharmacy, Timişoara, Romania

ABSTRACT

Introduction Patients having severe hemophilia (levels of deficient factor below 1%) frequently suffer from disabling chronic arthropathy. An adequate substitution treatment using the coagulation factor VIII or IX concentrates renders an elective surgery feasible.

Objective The objective of the study was to check the results of different surgical procedures in the treatment of hemophilic arthropathies, and to propose the best protocol of their treatment.

Methods This is a retrospective study on 26 hemophilic patients operated in the Orthopedics and Trauma Clinic II, Timişoara, from 2002 to 2005. Elective surgical procedures were mainly performed in the knee (21 arthroscopic procedures, 1 open arthrodesis), elbow (2 open synovectomies, 2 radial head excisions), ankle (1 arthroscopic synovectomy and debridement) and thigh (1 giant pseudo tumor excision, other minor procedures). The results after operations on moderate and severe chronic knee, elbow and ankle arthropathy were evaluated, with approximately 24-month follow-up period.

Results Arthroscopic procedures (22) yielded good and satisfactory results with significant improvement according to the evaluation criteria recommended by the World Hemophilia Federation (Gilbert clinical score, Pettersson radiological score, Nuss MRI score).

Conclusion Mini-invasive elective surgery in moderate to severe chronic arthropathy produces good results when performed in a specialized centre and with multi-disciplinary approach.

Key words: hemophilia; knee; synovectomy; arthroscopy; chronic arthropathy

INTRODUCTION

The most common manifestation of severe hemophilia (factor VIII/IX concentration less or equal to 1% of normal levels) is spontaneous or provoked joint and muscle hemorrhage, representing 80% of all bleeding episodes in hemophiliacs [1]. The knee joint is most frequently affected, followed by the ankle and the elbow. The cycle of joint deterioration starts with recurrent hemarthrosis that leads to synovial hypertrophy and chronic synovitis and evolves towards chronic arthropathy with disabling joint deterioration.

Conservative treatment for hemophilic arthropathy always includes substitution treatment (prophylactic – weekly administration, or "if required" treatment) with factor VIII (in A type hemophilia) or IX (in B type) concentrates, physiotherapy, anti-inflammatory medication and pain management. Avoiding the first hemarthrosis is the cardinal measure [2]. Other conservative measures comprise non-surgical chemical or radioactive synovectomy (synoviorthesis), intraarticular corticotherapy and intraarticular hyaluronic acid injections [3].

Perpetual surgical objectives were life and limb salvage. Anemia, massive hematoma, infection and hemorrhagic choc were common and often fatal complications. The advent and development of VIII and IX factor concentrates makes elective surgery feasible. Before any operation on a hemophiliac is considered, it is essential to determine hemophilia type and severity, the presence of factor VIII or IX specific antibodies (circulating inhibitors – risk of fatal complications) and viral status. The availability of sufficient amounts of factor concentrate and blood products (including by-pass agents as VIIa coagulation factor) in the event of complications is crucial [4].

Principles of surgery are multiple procedures during the same surgery, careful hemostasis, minimal and meticulous surgical approach, use of tourniquet whenever possible, local adjuvant measures (tranexamic acid, fibrin film) and additional surgical team safety measures due to high rate of viral infection (HIV, HCV, HBV) in these patients [5].

In recent years, non-surgical synovectomy (synoviorthesis) has increasingly become the method of choice in treating the recurrent hemarthrosis and chronic hemophilic synovitis [6, 7], but advanced stages of hemophilic arthropathy are associated with joint lesions that frequently require surgical management (Table 1).

Close cooperation of the orthopaedic surgeon and hematologist is the key to success when treating the hemophilic skeletal complications.

The benefits of open and arthroscopic synovectomy in hemophilia were highlighted in different studies [8-15]. Minimally invasive debridement and synovectomy procedures can be applied with good results in moderate and severe chronic hemophilic knee arthropathy, potentially evading the complications and high costs related to major surgery. Thus, one could avoid or substantially defer some of more radical procedures [5, 14, 16].

OBJECTIVE

The objective of the study was to check the results of different surgical procedures in the treatment of hemophilic arthropathies, and to propose the best protocol of their treatment.

TABLE 1. Surgical procedures for hemophilic arthropathies.
ТАБЕЛА 1. Хируршки поступци код хемофиличних артропатија.

Arnold-Hilgartner staging Класификација по Арнолд–Хилгартнеру	Interventions Хируршки поступци
I-II	Arthroscopic synovectomy (especially after failure of synoviorthesis) Артроскопска синовијектомија (нарочито после неуспеле синовиортезе)
III	Synovectomy (minimally invasive whenever possible) Синовијектомија (минимално инвазивна кад год је могуће)
	Debridement Дебридман
	Capsulectomies, ligamentoplasty Капсулектомије, лигаментопластика
IV-V	Arthroscopic (knee, ankle, shoulder) or open debridement (elbow) Артроскопски (колено, скочни зглоб, раме) или отворени дебридман (лакат)
	Osteotomy (particularly knee, hip) Остеотомија (нарочито колено, кук)
	Endoprosthetic arthroplasty (knee, hip, shoulder), arthrodesis (knee, ankle) Вештачки зглобови (колено, кук, раме), артродеза (колено, скочни зглоб)

METHOD

In the period January 1, 2002 to December 31, 2005, 32 surgical interventions were performed in 27 patients with severe and medium type A (23 patients) and severe B hemophilia (4 cases), at the II Clinic for Orthopaedics and Traumatology Timişoara. Mean age was 19 (9-34) years.

The following procedures were performed: 21 knee arthroscopies, 1 ankle arthroscopy, 2 open elbow synovectomies and 2 radial head excisions, 3 elbow bursectomies, 1 giant thigh pseudotumor ablation, 1 external fixation for distal femoral fracture and 1 biopsy of thigh osteosarcoma. Twenty-four patients suffered from moderate to severe chronic arthropathy at various levels. The majority (21) had several joints affected with various degrees of disability. Twenty-one patients were infected with HVC or/and HVB. None tested were positive to HIV. Low titers of circulating inhibitors (anti factor VIII antibodies) were found in five patients.

Recurrent hemarthrosis was present in 21 cases prior to surgery (average 2.5 episodes/month), and in 8 former surgical patients (appendicectomy, calcaneal tendinoplasty, operated psoas hematoma, femoral shaft fracture, knee arthrodesis, infected limb hematoma). In 3 youngest patients with knee arthropathy (aged 9, 10 and 12 years, respectively), the knee was involved unilaterally, probably due to a shorter evolution, while the majority patients had bilateral involvement. Functional disability varied from mild to severe, mainly associated with severe pain (9 cases) and significant limitation of joint movement. An average joint movement of operated knees was 67.5°, with an average flexion deformity of 11°.

Pronation-supination of the forearm on the side of the respective elbow was reduced to 45°. Flexion deformity of the operated elbows was also significant (50°-60°).

Preoperative deficient factor levels, viral status and inhibitor presence were determined. Radiographic evaluation included A-P and lateral views and 45° axial images of the knee joint. Magnetic resonance imaging of the affected joint and DXA bone densitometry of the lumbar spine and left hip were performed preoperatively in 22 and 23 patients, respectively.

The operated joints manifested moderate or severe stages of chronic hemophilic arthropathy, out of which there were 6 of stage VI according to Arnold and Hilgartner classification [1] (Table 2), 7 of stage V, 8 of IV and 2 of stage III. Radiographic Pettersson score [17] higher than 6 points out of maximum 13 was recorded in all operated joints.

Indication for elective surgery of hemophilic arthropathy was based on agreement between orthopedist and hematologist and consisted of abating the joint pain and marked reduction of joint movement, with/without the presence of recurrent hemarthrosis. The presence of circulating inhibitors in low concentration (less than 5 Bethesda units – B.U.) was not contraindication of the procedure. General contraindications were related to high concentrations of inhibitors and acute hepatitis.

The aim of the substitution protocol was to obtain 100% of deficient factor blood levels just prior to the operation using bolus administration of recombinant or monoclonal purified factor concentrates. This concentration was confirmed by preoperative factor dosage in patients with low titers of inhibitors (low responders). This blood level was maintained for the first 48 hours postoperatively using the continuous infusion (2-4 IU/kg/h) and then decreased gradually, with bolus administration bid. Substitution was continued throughout the rehabilitation period for 4-6 weeks, using 20-30 IU/kg twice a week in A hemophiliacs and three times a week in B type hemophiliacs. In 5 cases with low concentration factor VIII antibodies, the dosage of factor

TABLE 2. Classification of chronic hemophilic arthropathy after Arnold and Hilgartner.
ТАБЕЛА 2. Класификација хроничне хемофиличне артропатије по Арнолд–Хилгартнеру.

Stage Степен	Description Опис
0	Normal joint Нормалан зглоб
1	Soft tissue inflammation Упала меког ткива
2	Osteopenia and accelerated epiphysis growth probably as the result of hyperemia Остеопенија и убрзан раст епифизе због хиперемије
3	Bone contour modifications (widening of the intercondylar notch, flattening of the patellar and femoral condyle curves, the formation of bone cysts) Промене контуре кости (проширење интеркондиларног ноча, заравњење пателе и кондила фемура, костне цисте)
4	Narrowing of the joint space indicating cartilage erosion Сужење зглобног простора које указује на оштећење хрскавице
5	Substantial joint disorganization Дезорганизација зглоба

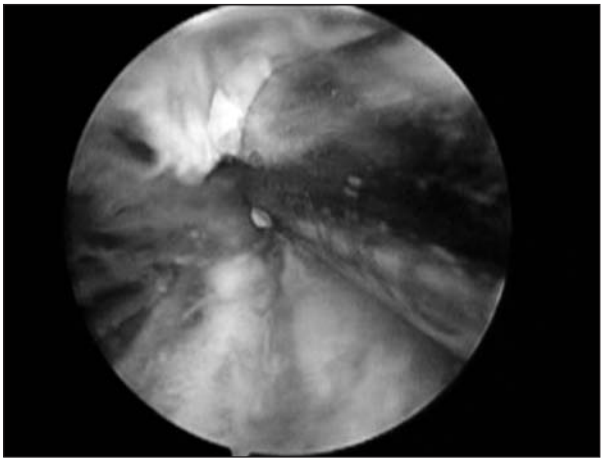

FIGURE 1. Arthroscopic knee synovectomy in hemophilia.
СЛИКА 1. Артроскопска синовијектомија колена код хемофилије.

concentrates was adjusted. The second generation antibiotic prophylaxis was systematic.

Arthroscopies were performed by the same surgeon. All procedures included synovectomy (Figure 1) and debridement, combined with the external and the internal patellar release in 4 cases and 1 case respectively, internal or external meniscectomy in 5 cases, osteophyte resection in 3 and loose body extraction in 5 cases. All arthroscopies were performed under general anesthesia and using the tourniquet. Anterolateral and anteromedial portals proved sufficient in all but one case of the knee arthroscopy. Synovectomy was carried out as thoroughly as possible with a shaver. Operated joints were immobilized for 24-72 hours postoperatively in a posterior splint or orthosis and full loading was allowed starting from day 2 or 3. Rehabilitation treatment was initiated on the second day and was continued in a specialized centre under supervision.

Joint alterations included chronic villous synovitis with hemosiderin impregnation and advanced chondropathy in all chronic arthropathy cases. The joint cartilage usually had severe macroscopic degenerative characteristics. Bone subchondral cysts were sometimes visible on arthroscopic examination.

Open synovectomy and radial head excision were performed in 2 elbows, with minimally 3cm-external incision. Synovectomy proved easier and more effective when arthroscopic shaver was used (Figure 2). The level of radial head resection was just below the ulnar radial facet preserving the annular ligament.

FIGURE 2. Open elbow synovectomy using the shaver in hemophilia. Postoperative X-ray of the same patient.
СЛИКА 2. Отворена синовијектомија лакта бријачем код хемофилије. Постоперациони радиограм код истог болесника.

One patient complained of severe pain, compressive vascular and functional impairment due to a giant (30/25/20 cm) pseudo-tumor in the posterior thigh muscular compartment. Surgery succeeded in complete tumor resection, in spite of difficult dissection. The tumor consisted of red-brownish liquid (unclotted and degraded blood) and the capsule was composed of compressed muscle tissue.

There were no infections or neurovascular complications. One patient had a wound dehiscence due to subcutaneous hematoma. Two patients complained of hemarthrosis during the rehab treatment at 2 and 3 weeks after surgery. There were 4 cases of joint stiffness that were treated conservatively with success. One patient who underwent the arthroscopic knee synovectomy, debridement, lateral release and the resection of large osteophyte needed repeated surgical drainage of an organized hematoma. One patient required revision arthroscopy due to degenerative internal meniscus tear.

A serious hemorrhagic complication developed in a 12-year old patient with very severe chronic knee arthropathy (13 points in the radiological Pettersson score), with a fixed 45° flexion deformity, patellar external and posterior tibial subluxation, in which case the knee arthrodesis was performed. The patient had low titers of factor VIII inhibitors (1.7 B.U.) preoperatively. A subcutaneous hematoma led to marginal necrosis and wound dehiscence. Four days after debridement and secondary suture, the intractable profuse bleeding which could result in hemorrhagic chock had to be managed. Repeated determination of inhibitors showed a ten-fold increase (16 B.U.). The bleeding was eventu- ally controlled through local hemostatic measures (elastic bandage, local tranexamic acid) and the administration of large amounts of VIIa factor concentrate (NovoSeven).

Average hospitalization period was 14.6 days (7-21), followed by rehabilitation in a specialized centre for 1-2 months. Substitution therapy averaged 11500 IU of factor VIII concentrate/arthroscopy and 14500 IU/case.

Follow-up visits at 1, 2, 6 months and then annual visits included clinical and radiographic examinations. An average follow-up period was 24 months (12-48). The retrospective evaluation was performed using the 1981 classification recommended by the Orthopaedic Advisory Committee of the World Federation of Hemophilia [17]. This included pain assessment (0-3 points, Table 3), bleeding (0-3, Table 4), clinical score (0-12 points, Table 5) and radiographic score (0-13, Table 6). In addition, the Nuss score [18] was used to assess joint damage viewed on the MRI (Table 7).

RESULTS

The difference between the joint score prior to surgery and at the end of the follow-up was determined and summarized in table 8. Good result (according to the grading system proposed by Rodriguez-Merchan et al., 1994 [13]) was considered when the joint score was lessened by more than 5 points, satisfactory between 0 and 5 points and poor when the score was increased.

TABLE 3. Pain (total score on a scale 0 to 3).
ТАБЕЛА 3. Бол (градиран од 0 до 3).

Score Скор	Description Опис
0	No pain, no functional deficit Без отока и функционалног испада
1	Mild pain, does not interfere with normal limb use; occasional analgesia Благ бол који не угрожава функцију; понекад аналгезија
2	Moderate pain, some interference with normal use; occasional analgesia Осредњи бол, делимично угрожава функцију; понекад аналгезија
3	Severe pain, interference with normal limb use; frequent analgesia (narcotic) Јак бол, угрожава функцију; често аналгезија (наркотици)

TABLE 4. Bleeding – incidence in one year (total score on a scale 0 to 3). Minor bleeding: mild pain, minimal swelling, mild range of movement limitation, resolution in less than 24 hours; major bleeding: pain, swelling, movement limitation, lack of resolution after 24 hours of treatment.
ТАБЕЛА 4. Крвављење – годишња инциденција (на скали од 0 до 3). Мање крвављење: благ бол, минимални оток, благо ограничење покрета, опоравак за мање од 24 часа; веће крвављење: бол, оток, ограничење покрета, изостанак опоравка у року од 24 часа лечења.

Score Скор	Description Опис
0	No hemorrhage Без крвављења
1	No major hemorrhages, 1-3 minor hemorrhages Без већих крвављења, 1-3 мања крвављења
2	1-2 major hemorrhages, 4-6 minor hemorrhages Једно или два већа крвављења, 4-6 мањих крвављења
3	3 or more major hemorrhages, 7 or more minor hemorrhages Три већа крвављења или више мањих, седам мањих крвављења или више мањих

We obtained 15 good results (56.25%), 7 satisfactory (37.5%) and 2 poor (6.25%) results. Subjectively, 19 patients characterized the outcome as good, 4 as satisfactory and 1 as poor.

The best outcome was relative to hemarthrosis frequency which was consistently diminished to an average of 1 major episode/6 months. Joint mobility improvement was more modest, with an average increase of 12.6° in the knee arthroscopy patients. Among 4 patients with the chronic external patellar dislocation and lateral release, 3 had better joint movement. There was no increase of the radiological score at the end of the follow-up in any patient.

Both patients with radial head excision had significant pain reduction after surgery. Pronation-supination was improved by an average of 14° without marked advance for elbow flexion-extension. None manifested elbow instability.

Good statistical correlation of radiological Pettersson score and the MRI Nuss score ($p<0.01$) was found. There was no conclusive statistical correlation of the clinical and radiological score in both preoperative and follow-up period. The age at the time of surgery did not correlate with the improvement of the joint score, range of movement or bleeding frequency.

An interesting aspect was the lack of apparent correlation between the preoperative joint score and the outcome.

Lower bone marrow density (BMD) was omnipresent in these patients, while osteoporosis (T score lower than -2.5) was found in 15 patients. The incriminated mechanisms could be the lack of physical exercise and/or malabsorptive syndrome due to hepatic impairment secondary to HVC infection [19-21]. Statistical correlation of the incidence of C type hepatitis virus and osteoporosis could not be established as evidence supporting the latter hypothesis.

TABLE 5. Clinical evaluation score (0-12) of hemophilic arthropathy (excluding the joint pain) .
ТАБЕЛА 5. Процена хемофиличне артропатије (0-12) на основу клиничких знакова (зглобни бол искључен).

Clinical sign Клинички знак	Score Скор	Description Опис
Swelling Оток	0-2	0 – None / Без отока 2 – Present / Присутан (S) – If chronic synovitis is present / Уколико постоји синовитис
Muscle atrophy Атрофија мишића	0-1	0 – Absent / Изостаје 1 – Present / Присутна
Axial deformity Аксијални деформитет	0-2	0 – No deformity / Нема деформитета 1 – Less than 10 degrees / Мање од 10 степени 2 – More than 10 degrees / Више од 10 степени
Crepitus on motion Пуцкетање при покрету	0-1	0 – None / Без пуцкетања 1 – Present / Присутно
Flexion contracture Флексиона контрактура	0-2	0 – None / Изостаје 1 – Less than 15 degrees of fixed flexion contracture / Мање од 15 степени фиксиране флексионе контрактуре 2 – More than 15 degrees of fixed flexion contracture / Више од 15 степени фиксиране флексионе контрактуре
Range of motion Обим покрета	0-2	0 – Loss of up to 10% of full range of motion / Губитак покрета до 10% 1 – Loss of 10-33.33% of motion / Губитак покрета од 10 до 33.33% 2 – Loss of more than 33.33% of motion / Губитак покрета већи од 33.33%
Instability Нестабилност	0-2	0 – None / Изостаје 1 – Present but normal function possible / Присутна, али је нормална функција могућа 2 – Functional deficit, requires bracing / Са функционалним испадом, захтева ортотисање

TABLE 6. Radiologic evaluation score (Petersson score) of hemophilic arthropathy (0-13) [17].
ТАБЕЛА 6. Радиолошка процена хемофиличне артропатије по Петерсону (0-13) [17].

Radiologic changes **Радиолошке промене**	**Score** **Скор**	**Description** **Опис**
Osteoporosis Остеопороза	0-1	0 – Absent / Изостаје 1 – Present / Присутна
Enlarged epiphysis Проширена епифиза	0-1	0 – Absent / Изостаје 1 – Present / Присутна
Irregular subchondral surface Неправилна субхондрална површина	0-2	0 – Absent / Изостаје 1 – Surface partially involved / Површина делимично захваћена 2 – Surface totally involved / Површина потпуно захваћена
Narrowing of the joint space Сужење зглобног простора	0-2	0 – Absent / Изостаје 1 – Less than 1 mm narrowing / Сужење мање од 1 *mm* 2 – More than 1 mm narrowing / Сужење веће од 1 *mm*
Subchondral cysts Субхондралне цисте	0-2	0 – Absent / Изостају 1 – One cyst / Једна циста 2 – More than one cyst / Више од једне цисте
Erosion of the joint margins Ерозија зглобних ивица	0-1	0 – Absent / Изостаје 1 – Present / Присутна
Gross incongruence of the articular bone ends Значајна неподударност зглобних површина	0-2	0 – Absent / Изостаје 1 – Slight / Блага 2 – Marked / Изажена
Joint deformity (angulation/ displacement) Деформитет зглоба (кривљење, ишчашење)	0-2	0 – Absent / Изостаје 1 – Slight / Благ 2 – Marked / Изражен

DISCUSSION

The advent of synoviorthesis pushed indication for arthroscopy to the second intention procedure, after failure of chemical or radioactive synovectomy in early stages of arthropathy. High percentage of good and satisfactory results (20 out of 21 patients) obtained after arthroscopic procedures in moderate and severe stages of the knee and ankle chronic hemophilic arthropathy indicates the potential value of arthroscopy in later stages of joint deterioration. The lack of correlation between the degree of improvement at the end of the follow up and the severity of the joint score suggests that debridement and other arthroscopic procedures can result in significant improvement regardless of the degree of joint impairment in hemophilia.

TABLE 7. MRI evaluation score (Nuss score) of hemophilic arthropathy (0-12) [16].
TABELA 7. Магнетнорезонантна процена хемофиличне артропатије према Нусовом скору (0-12) [16].

Disease **Обољење**	**Score and description** **Скор и опис**
Hemarthrosis Хемартроза	1 – Mild / Блага 2 – Moderate / Средње тешка 3 – Severe / Тешка
Hemosiderin Хемосидерин	1 – Present / Присутан
Synovium hypertrophy Хипертрофија синовије	1 – Discrete / Блага 2 – Moderate / Средње тешка 3 – Severe / Тешка
Subchondral cysts and erosions Субхондралне цисте и ерозије	1 – One cyst and partial joint surface erosions / Једна циста и парцијалне ерозије зглобне површине 2 – More than one cyst and partial joint surface erosions / Више од једне цисте и парцијалне ерозије зглобне површине 3 – More than one cyst and complete joint surface erosions / Више од једне цисте и ерозије целе зглобне површине
Joint cartilage defects Оштећења хрскавице зглоба	1 – Less than 50% / Мање од 50% 2 – 50% and more / 50% и више 3 – Complete / Потпуна

The presence of circulating inhibitors can contraindicate an elective open surgery [22, 23], and the treatment with factor VIII "by-passes", the agents such as an activated factor VII, is exorbitant [24] and insufficiently studied [21, 25]. Because of its low surgical trauma and factor replacement needs, arthroscopy can provide a solution in some patients with low concentration of factor inhibitors ("low responders" < 5 U.B).

Synovectomy coupled with radial head excision gave excellent results in 2 patients with severe chronic elbow arthropathy. Given the small incision, this procedure seems superior regarding the efficiency and surgical trauma to elbow arthroscopy in hemophilia.

The Nuss MRI score correlated well with the Pettersson radiological score, proving their relevance as tools of hemophilic knee image evaluation. However, in spite of its higher sensibility in detecting early modification of the joint, MRI seems redundant in moderate and severe (radiological) stages of hemophilic arthropathy. The incidence of lower bone density in these young patients (100%) is an aspect that needs further studies concerning the pathogenesis and treatment.

Good results, low costs and risks using the arthroscopic or minimal open procedures in moderate and severe chronic hemophilic knee, elbow and ankle arthropathy led us to consider these interventions as precursor steps to potential major surgeries as osteotomy, arthrodesis or total arthroplasty.

Hemorrhagic complications can be avoided and controlled through an expert hematological surveillance and care. Preoperative level of deficient coagulation factor of 80%-100% must be achieved and maintained using the continuous infusion in the first 48 hours.

The severity of the manifestations depends on the rest activity of factor VIII or IX and on the quality of the substitution treatment. The management of hemo-

TABLE 8. Main data and results.
TABELA 8. Одлике болесника и резултати.

Case Болесник	Age (years) Старост (године)	Follow-up period (months) Период надгледања (месеци)	Procedure Поступак лечења	Pain score[1] Степен бола[1]	Bleeding score[2] Степен крварења[2]	Clinical score[3] Клиничко стање[3]	Radiological (Pettersson) score[4] Радиографски налаз (Петерсон)[4]	Joint score (1+2+3+4) Стање зглоба (1+2+3+4)	MRI Nuss score МР стање	T score (spine/hip) Т однос (кичма/кук)	Result (objective/subjective) Резултат (објективан/субјективан)
1	26	48	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/1	3/0	10/8	11/11	26/20	8	-0.5/-2.5	G/G
2	15	42	Knee synovectomy, debridement, internal meniscectomy, lateral patellar release Синовијектомија колена, дебридман, медијална менисцектомија, спољашње ослобађање чашице	2/0	3/1	6/3	7/7	18/11	7	-1.1/-2.5	G/G
3	16	42	Knee synovectomy, debridement, internal meniscectomy, loose body extraction, osteophyte resection Синовијектомија колена, дебридман, медијална менисцектомија, вађење слободног тела, ресекција остеофита	2/1	2/0	4/2	6/6	14/9	5	-2.3/-2.3	G/G
4	26	36	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/0	3/0	6/3	7/7	18/10	5	-2.3/-2.3	G/G
5	11	36	Knee synovectomy, debridement Синовијектомија колена, дебридман	1/1	2/1	7/6	9/9	19/17	7	-4.3/-2.1	S/S
6	10	36	Knee synovectomy, debridement, loose body extraction Синовијектомија колена, дебридман, вађење слободног тела	1/0	2/0	7/4	8/8	18/12	7	-2.7/-1.5	G/G
7	31	24	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/1	2/0	5/3	7/7	16/11	6	-4.2/-3.8	S/S
8	10	24	Knee synovectomy, debridement Синовијектомија колена, дебридман	1/1	2/0	5/5	6/6	14/12	6	-3.2/-3.0	S/G
9	25	18	Knee synovectomy, debridement, revision arthroscopy Синовијектомија колена, дебридман, ревизиона артроскопија	2/0	3/0	7/6	9/9	21/15	6	-2.6/-2.3	G/G
10	25	18	Knee synovectomy, debridement, internal meniscectomy, loose body extraction Синовијектомија колена, дебридман, унутрашња менисцектомија, вађење слободног тела	3/1	3/0	7/6	8/8	21/15	7	-2.8/-2.3	G/G
11	10	18	Knee synovectomy, debridement, lateral release Синовијектомија колена, дебридман, спољашње ослобађање	2/2	2/1	6/8	6/6	16/17	5	-4.3/-3.3	P/S
12	14	18	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/1	2/1	6/5	6/6	16/13	4	-4.2/-2.2	S/G
13	29	12	Knee synovectomy, debridement, external meniscectomy Синовијектомија колена, дебридман, спољашња менисцектомија	2/0	2/0	7/3	7/7	18/12	7	-0.7/-1.5	G/G
14	10	12	Knee synovectomy, debridement, loose body extraction Синовијектомија колена, дебридман, вађење слободног тела	3/1	2/0	8/6	9/9	22/16	6	-2.8/-2.7	G/G
15	12	12	Knee synovectomy, debridement, lateral release Синовијектомија колена, дебридман, спољашње ослобађање	3/1	1/0	8/9	10/10	22/20	9	-4.8/-3.8	S/S
16	12	12	Knee synovectomy, debridement, internal meniscectomy, loose body extraction, osteophyte resection, lateral release Синовијектомија колена, дебридман, медијална менисцектомија, вађење слободног тела, ресекција остеофита, спољашње ослобађање	2/0	1/0	9/7	11/11	22/18	9	-5.1/-3.8	S/G
17	32	12	Knee synovectomy, debridement Синовијектомија колена, дебридман	3/1	2/0	7/4	9/9	21/14	9	-4.1/-3.2	S/G
18	22	12	Knee synovectomy, debridement, lateral release Синовијектомија колена, дебридман, спољашње ослобађање	3/1	3/1	7/4	9/9	22/15	8	-4.2/-4.1	G/G

Case Болесник	Age (years) Старост (године)	Follow-up period (months) Период надгледања (месеци)	Procedure Поступак лечења	Pain score[1] Степен бола[1]	Bleeding score[2] Степен крварења[2]	Clinical score[3] Клинички статус[3]	Radiological (Pettersson) score[4] Радиографски налаз (Петерсон)[4]	Joint score (1+2+3+4) Стање зглоба (1+2+3+4)	MRI Nuss score MP стање	T score (spine/hip) T однос (кичма/кук)	Result (objective/ subjective) Резултат (објективан/ субјективан)
19	17	12	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/0	2/0	6/4	8/8	18/12	7	-3.2/-2.9	G/G
20	16	12	Knee synovectomy, debridement Синовијектомија колена, дебридман	2/0	2/1	7/4	8/8	19/13	6	-2.8/-2.8	G/G
21	34	48	Elbow synovectomy, radial head excision, debridement, bursectomy Синовијектомија лакта, вађење главе радијуса, дебридман, одстрањивање бурзе	2/0	1/0	4/1	8/8	15/9	-	-1.5/-1.2	G/G
22	31	48	Elbow synovectomy, radial head excision, debridement, bursectomy Синовијектомија лакта, вађење главе радијуса, дебридман, одстрањивање бурзе	3/1	3/0	10/7	13/13	29/21	-	-	G/G
23	12	18	Knee arthrodesis Артродеза колена	3/0	2/0	12/3	13	N/A	12	-4.7/-5.4	P/P
24	21	12	Ankle synovectomy, debridement Синовијектомија скочног зглоба, дебридман	3/0	3/0	3/3	7/7	16/10	8	-3.2/-3.4	G/G

/ – preoperational and end of follow-up value; N/A – not available; G – good; S – satisfactory; P – poor*
/ – вредност пре операције и на крају лечења; N/A – недоступан податак; G – добар; S – задовољавајући; P – лош*

philia needs multidisciplinary approach in a specialized centre and it is extremely expensive. However, with correct substitution treatment, orthopaedic surgery can be performed with acceptable risks and satisfying results.

CONCLUSION

Arthroscopic or minimal open procedures represent the best choice for the treatment of hemophilic arthropathies. More aggressive procedures (osteotomies, arthrodesis or total arthroplasty) have to be reserved for the most severe cases.

Hemorrhagic complications can be avoided and controlled through an expert hematological surveillance and care. Preoperative level of deficient coagulation factor of 80%-100% must be achieved and maintained using the continuous infusion in the first 48 hours.

REFERENCES

1. Arnold WD, Hilgartner MW. Hemophilic arthropathy. Current concepts of pathogenesis and management. J Bone Joint Surg Am 1977; 59:287-305.
2. Barnes C, Wong P, Egan B, Speller T, Cameron F, Monagle P. Reduced bone density among children with severe hemophilia. Pediatrics 2004; 114(2):177-81.
3. Luck JV, Silva M, Rodriguez-Merchan C, Galambor D, Zahiri C, Finn R. Hemophilic arthropathy. JAAOS 2004; 12(7-8):234-45.
4. Hedner U, Feldstedt M, Glazer S. Recombinant F VIIa in hemophilia treatment. In: Lusher JM, Kessler CM, editors. Hemophilia and von Willebrand's Disease in the 1990s. New York: Elsevier Science; 1991. p.283-93.
5. Poenaru D, Şerban M, Pătraşcu J, Branea I, Bîrsăşteanu F. Chirurgia electivă în artropatiile cronice hemofilice. Revista Societăţii Române de Ortopedie şi Traumatologie (SOROT) 2004; 24(4):3-9.
6. Heim M. The treatment of intra-articular synovitis by the use of chemical and radioactive substances. Hemophilia 2000; 8:369-71.
7. Eickhoff HH, Raderschadt G, Koch W, Brackmann HH. Control of the synovium in hemophilia. Hemophilia 1998; 4:511-3.
8. Storti E, Traldi A, Tosatti E, Davoli PG, Synovectomy. A new approach to hemophilic arthropathy. Acta Hemat 1969; 41:193-7.
9. Mannucci PM, de Franchis R, Torri G, Pietrogrande V. The role of synovectomy in hemophilic arthropathy. Israel J Med Sci 1977; 13:983-6.
10. Klein K, Aland C, Kim H, Giselle J, Saidi P. Long term follow up of arthroscopic synovectomy for chronic hemophilic synovitis. Arthroscopy 1987; 3:231-6.
11. Luck JV, Kasper CK. Surgical management of advanced hemophilic arthropathy. An overview of 20 years experience. Clin Orthop 1989; 242:60-82.
12. Triantafyllou SJ, Hanks GA, Handal JA, Greer RB. Open and arthroscopic synovectomy in hemophilic arthropathy of the knee. Clin Orthop 1992; 283:196-204.
13. Rodriguez-Merchan EC, Galindo E, Ladredat JMM, Pardo JA. Surgical synovectomy in hemophilic arthropathy of the knee. International Orthopaedics (SICOT) 1994; 18:38-41.
14. Wiedel JD. Arthroscopic synovectomy of the knee in hemophilia. Clin Orthop 1996; 328:46-53.
15. Eickhoff HH, Koch W, Raderschadt G, Brackmann HH. Arthroscopy for chronic hemophilic synovitis of the knee. Clin Orthop Relat Res 1997; 343:58-62.
16. Branea I, Poenaru D, Serban M, Birsasteanu F, Patrascu J. The management of osteoarticular complications in hemophilia. Timişoara Medical Journal 2005; 55(1):64-73.
17. Pettersson H, Gilbert MS. Classification of the hemophilic arthropathy. In: Pettersson H, Gilbert MS, editors. Diagnostic imaging in hemophilia. Springer: New York; 1985. p.56-68.
18. Nuss R, Kilcoyne RF, Geraghty S, Wiedel J, Manco Johnson M. Utility of magnetic resonance imaging for management of hemo-

philic arthropathy in children. J Pediatr 1993; 123:388-92.
19. Monagle P, Barnes C, Wong P, et al. Bone density in children with severe hemophilia. Blood 2003; 102:795-8.
20. Poenaru D, Bîrsăşteanu F, Pătraşcu J, et al. Confrontation of clinical and imaging assessments in chronic hemophilic arthropathy. Abstract of the XXVI International Congress of The World Federation of Hemophilia. Hemophilia 2004; 10(3):103.
21. Poenaru D, Şerban M, Branea I. Artropatiile hemofilice. Bucureşti: Editura Academiei Române; 2001.
22. Luck JV, Lin JC, Kasper CK, Logan LJ. Orthopaedic management of hemophilic arthropathy. In: Chapman MW, editor. Chapman Orthopaedic Surgery. 3rd ed. Philadelphia: Lippincott William and Wilkins; 2001.
23. Rudowski WJ, Scharf R, Ziemski JM. Is major surgery in hemophiliac patients safe? World J Surg 1987; 11:378-80.
24. Schramm W, Royal S, Kroner B, et al. for the European hemophilia economic Study Group. Clinical outcomes and resource utilization associated with hemophilia care in Europe. Hemophilia 2002; 8:33-43.
25. Şerban M, Petrescu C, Ţepeneu P, et al. Evaluarea eficienţei clinice a rFVIIa în pediatrie. Documenta Hematologica 2000; 2:59-62.

НАШЕ ИСКУСТВО У ОРТОПЕДСКОХИРУРШКОМ ЛЕЧЕЊУ БОЛЕСНИКА ОБОЛЕЛИХ ОД ХЕМОФИЛИЈЕ

Dan V. POENARU[1], Margit ŞERBAN[2], Ioan L. BRANEA[1], Jenel M. PĂTRAŞCU[1]

[1]Друга ортопедско-трауматолошка клиника, Биомедицински универзитет „Виктор Бабеш", Темишвар, Румунија;
[2]Трећа педијатријска клиника, Биомедицински универзитет „Виктор Бабеш", Темишвар, Румунија

КРАТАК САДРЖАЈ

Увод Болесници оболели од тешке хемофилије (ниво недостајућег фактора нижи од 1%) често пате од хроничних артропатија. Коректна надокнада концентрованих фактора *VIII* или *IX* чине елективну хирургију могућом.

Циљ рада Циљ истраживања је био да се оцене резултати различитих хируршких поступака у лечењу болесника оболелих од хемофиличних артропатија, те да се предложи најбољи протокол за њихово лечење.

Метод рада Спроведено је ретроспективно истраживање које је обухватило 26 испитаника оболелих од хемофилије који су хируршки лечени на Другој ортопедско-трауматолошкој клиници у Темишвару од 2002. до 2005. године. Елективни хируршки поступци су углавном рађени на колену (21 артроскопски поступак, једна отворена артродеза), лакту (две отворене синовијектомије, две ресекције главе радијуса), скочном зглобу (једна артроскопска синовијектомија и дебридман) и бутини (одстрањење великог псеудотумора, други мањи хируршки поступци). Оцена резултата је рађена после наведених операција, а болесници су надгледани 24 месеца.

Резултати Артроскопски поступци (22) су дали добре и задовољавајуће резултате са значајним побољшањем према евалуационим критеријумима које је предложило Светско удружење хемофиличара – Гилбертова (*Gilbert*) скала за клиничку процену, Петерсонова (*Pettersson*) скала за радиолошку процену, Нусова (*Nuss*) скала за магнетнорезонантну процену.

Закључак Миниинвазивно елективно хируршко лечење средње тешких и тешких хроничних артропатија даје добре резултате уколико се примењује у специјализованим центрима и уз мултидисциплински приступ.

Кључне речи: хемофилија; колено; синовијектомија; артроскопија; хронична артропатија

Dan V. POENARU
10 Giurgiu Str., Timişoara
Romania
Tel.: +40 722 392 609
Fax: +40 256 497 824

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