Orthopedic approach of haemophiliacs

A single center experience in Romania

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Haemophilia, orthopaedic therapy, haemophilic arthropathy

Summary
Clinical expression of inadequately treated haemophilia is dominated by orthopaedic complications, requiring invasive or non-invasive interventions. Objective: In Romania, with under dosed and late introduced “on demand” substitution, we aimed at highlighting the experience of orthopedic treatment and its outcome. Patients, methods: Single center retrospective analysis regarding orthopedic interventions and their outcomes was conducted on 59 hemophilia A, B, and von Willebrand disease patients, between 2002 and 2007. Results: The majority of interventions, invasive (60.71%) and non-invasive (39.28%), were elective, only two being emergencies. Postoperative functional evolution after synovectomies was good in 68.28%, fair in 24.39%, satisfactory in 7.31%. Results of 33 non-invasive (extensive releases) procedures were very good in 27.27%, good in 63.63%, poor in 9.09%. Discussion, conclusions: The important number and complexity of orthopedic interventions are proving the precarious musculoskeletal state in persons with hemophilia, demonstrating the need of improving substitution, at least with discontinuous prophylaxis in patients with severe forms.

Schlüsselwörter
Hämophilie, orthopädische Behandlung, hämophilische Arthropathie

Zusammenfassung

Orthopädische Behandlung in Hämophilie Erfahrung eines rumänischen Hämophiliezentrums

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Clinical expression of inadequately treated haemophilia is dominated by orthopaedic complications, most of them requiring interventions for improvement of locomotor function that affects patients’ quality of life (1, 2).

In our country, poor substitution (~0.51 IU/capita/year), together with absence of prophylaxis and home treatment programs are responsible for frequent haemarthroses (18.5 ± 11.2/year in severe and 13.4 ± 8.1/year in moderate form) leading to chronic synovitis and severe haemophilic arthropathy (target joints: children: 44.11% and adults: 62.71%) (9).

Precarious musculoskeletal status, reflected in total orthopaedic joint score (in children: 22.96 ± 21.11, range: 0–154; in adults: 38.38 ± 20.79, range: 0–132), and disturbing complains, like frequent joint bleeds, pain and debilitating, imposed the orthopaedic interventions in our haemophilia patients. Because of the low availability of factor concentrates, elective orthopaedic approach is only a recent attempt to solve the major challenge of joint/skeletal complications in our country (6).

We aimed at providing an outline of our experience in orthopaedic treatment and at evaluating its outcome in these conditions, characterized by underdosed and late introduced on demand substitutive therapy.

Patients and methods
This is a single center, retrospective analysis, conducted on 59 patients from different regions of Romania, registered and treated in Haemophilia Center of Timisoara over a period of five years (2002–2007). We analyzed the indications and type of orthopaedic interventions and we performed the follow-up of outcomes from the perspectives of medical economy. Types of interventions were both
● invasive (synovectomy, which was undertaken mostly arthroscopic, arthrodesis, radial head resection, external fixation of distal femur fracture, pseudotumour surgery, drainage of a forearm haematoma) and
● non-invasive (knee and thigh extensive releases).

Indications for synovectomy were frequent recurrent haemarthroses (>3/month ± severe joint pain), which were present in 100% of patients (25.64% with severe pain, according to WFH pain score). Arthrodesis
was indicated in cases with intractable pain (100% severe pain estimated by WFH pain score) associated with severely damaged joint. We performed extensive release (ER) with gradual straightening in patients with flexion contracture. Substitution with FVIII/IX concentrates in surgical procedures was assured according to WFH indications (Tab. 1) (3–5), with daily monitoring of FVIII/IX concentration. In 68.5% the substitution was performed as continuous infusion. Substitution with FVIII concentrates in non-invasive interventions is represented in Table 2.

After synovectomies, an aspirative drain was used in every case and maintained for 24–48 h postoperatively. In these cases, average hospitalization days were 14.6 (7–21) days. In case of surgical procedures, rehabilitation (mobilization and supervised physiotherapy) was started the second day after surgery and continued up to 90–180 days. After extensive releases immobilization was maintained up to three weeks, followed by wearing an orthosis and rehabilitation.

We monitored the following parameters: bleedings, pain, range of motion (ROM), and extension capacity for knee at one year post-intervention. We used an evaluation scale for haemarthroses. Results:

- very good (<3 haemarthrosis/year),
- good (3 haemarthrosis/year),
- fair (4–5 haemarthrosis/year) and
- poor (≥6 haemarthrosis/year).

Evaluation scale for pain considered the following results: very good (no pain); good (slight pain-no consequences on work or everyday activities, occasional use of non-narcotic analgesics); fair (moderate pain-occasional consequences on work/everyday activities; intermittent use of non-narcotic analgesics/ intermittent use of narcotics); poor (severe pain-consequences on work or everyday activities; frequent use of non-narcotic and narcotic analgesics) (7).


Results

A total of 86 procedures were performed in 59 patients (Fig. 1), including

- invasive: 53 (61.62%),
- elective: 51,
- emergencies: 2 (distal femur fractures),
- non-invasive: 33 (38.37%).

**Invasive procedures**

We performed 41 synovectomies: 3 surgical (for elbow), and 38 arthroscopic (35 for the knee and 3 for the ankle).

In four patients multiple simultaneous interventions have been performed: one with arthroscopic ankle synovectomy and ankle arthrodesis, one with open synovectomy of the elbow, arthroscopic knee synovectomy and knee arthrodesis and two patients with bilateral knee synovectomies.

After invasive procedures, we noticed complications in six cases: haemarthroses (4 cases), residual synovitis, requiring secondary removal and inhibitors development. In all cases, complications didn’t affect the late outcome. No perioperative vasculo-nervous or infectious complications were noticed. One year-follow-up after synovectomy showed for haemarthrosis

- very good results in 77.77% of patients,
- good results in 22.22%.

The impact on pain was similar:

- very good in 81.48% of patients and
- good in 18.5%.

One year-follow-up after synovectomy is represented in Table 3. ROM, flexion and extension loss showed very good results in 7.317% of cases, good results in 60.97%, fair results in 24.39% and poor results in 7.317% of cases.

**Non-invasive procedures**

A number of 33 extensive releases have been performed. They were done under gen-
eral anaesthesia, using substitution with FVIII concentrates (in bolus-in 9 patients and in endovenous perfusion-in 2 patients). This procedure was applied in 11 patients (10 in knee, 1 bilateral hip). Seven of them needed 3–4 serial procedures. In five patients, a single joint was involved, the rest of them having bilateral joint involvement.

In one patient we performed one unilateral extensive release, in three patients two unilateral extensive releases were performed respectively. In five patients, preoperative flexion contracture was between 45–90°, while six patients had flexion contracture less than 45°.

One year-follow-up after extensive release is represented in Table 3. One year follow-up after extensive releases showed poor results in 9.09% of cases, good results in 63.63% and very good in 27.27%.

After extensive releases, one patient developed inhibitors against FVIII (after 3 exposure days), but without bleeding events. Haemostatic control was achieved in all cases, treatment regimens were well tolerated, and there were no infectious or vascular-nervous complications.

### Discussion, conclusion

It is evident the precarious musculo-skeletal state in persons with haemophilia in our country. Despite the important shortage in substitutive treatment it was justified a significant number (17.2/year) of orthopaedic interventions. These have been beneficial in almost all cases from the point of view of pain, occurrence of joint bleeds and function in case of invasive, and in 90.9% of non-invasive procedures from the point of view of symptoms and function.

Disadvantages of late interventions (inability to stop the progression of arthropathy) and high costs recommend improvement of substitution, earlier orthopaedic interventions and, perhaps, a less expensive method (chemical synovectomy?).

### Conflict of interest

All authors declare, that there is no conflict of interest.

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