

## CASE REPORT

# Rehabilitation of patients with haemophilia after orthopaedic surgery: a case study

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**Summary.** Preoperative and postoperative rehabilitation may be useful for improving the recovery of patients undergoing orthopaedic surgery, particularly in those with co-morbidity or special requirements. This case study, of a patient with haemophilia and inhibitors to factor VIII undergoing total knee replacement, demonstrates the benefits of 6 weeks' preoperative physiotherapy ('prehabilitation') combined with 6 weeks' postoperative rehabilitation.

The supervised physiotherapy regimen was individually tailored to specifically increase range of motion and muscle strength, enabling rapid mobilization and recovery of function, whilst minimizing the risk of bleeding.

**Keywords:** haemophilia, inhibitors, knee replacement, orthopaedic surgery, physiotherapy, rehabilitation

## Introduction

Recovery from orthopaedic surgery is a challenge for a fully healthy patient, but for those of advanced age or with concurrent disease, the problem is even greater. Patients with haemophilia comprise such a group, requiring orthopaedic surgery when conservative therapies have failed to relieve the symptoms and progression of disease-related haemarthrosis [1]. Bleeding into the haemophilic joint causes chronic arthropathy resulting in musculoskeletal deterioration, reduced range of motion, crepitus, pain and swelling. In addition, non-diseased joints may be affected by compensatory use. Combined with this preoperative morbidity, patients with haemophilia are necessarily confined to inactivity for extended periods postsurgery to minimize the risk of bleeding. This extended period of rest can further hamper recovery, as enforced immobility can lead to muscle atrophy, making attempts to walk on or move the joint difficult. Edgerton *et al.* [2] showed that muscle cross-sectional area decreased after just 5 days of restricted use, and Thompson [3] reported a 37% loss of muscle mass after just 4 days of inactivity.

Therefore, rehabilitation is essential in patients with haemophilia if they are to make a speedy and optimal recovery from orthopaedic surgery.

Rehabilitation of orthopaedic patients can incorporate both preoperative and postoperative physical therapy regimens:

- 1 Preoperative rehabilitation (also known as 'prehabilitation') aims to enhance and optimize functional capacity, making the patient as fit and strong as possible before they undergo surgery [4]. This should improve the ability to withstand the surgery itself, the associated immobilization and the recovery period, with minimal deterioration in function. Furthermore, this period can be used to teach the patient exercises to strengthen muscles and increase mobility after surgery, as well as how to use mobility aids such as crutches. It has been reported that prehabilitation does not improve postoperative outcome [5,6]. However, these studies focused on patients with monoarticular osteoarthritis; multiple joint involvement and high levels of disability, as seen in patients with haemophilia, were not differentiated. Prehabilitation may be more beneficial in these latter cases and, in particular, may help to reduce preoperative anxiety [6].
- 2 Postoperative rehabilitation may quicken healing and restore mobility following orthopaedic surgery in non-haemophilic patients [7–9], although some findings are contradictory [10]. Massage can be used to increase circulation and lymphatic

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drainage, while individually designed exercise regimens strengthen muscles and increase range of motion, which may be restricted by adhesion and contraction of soft tissues around the wound site and joint. This period of physiotherapy can improve the patient's proprioception, coordination and balance, and re-educate their gait, which may have become altered by compensation or surgery. In addition, daily activities may be modified to prolong joint function. The type and duration of postoperative rehabilitation depends on the surgical procedure and preferences of the orthopaedic surgeon. However, patient factors are also very important, and the patient's age and motivational status should be carefully considered when designing an exercise regimen. For example, a patient may be fearful of bleeding into the operated joint when they first take weight on it; in this case it may be helpful to use exercises that involve the patient pushing the leg into the bed or physiotherapist's hand to show them that they are able to take some weight through the joint.

Patients with haemophilia undergo an annual joint assessment, and it is often at this time that a failure of conservative therapy (including activity modification, exercises, hydrotherapy, orthotic support) is identified. During this examination, the physiotherapist assesses a range of disease progression markers, including bleeding frequency, pain, swelling/synovitis, range of motion, crepitus and muscle power. In addition, functional elements such as gait and balance are assessed. By incorporating these markers into a functional outcome score and tracking changes over time, the clinician can detect deterioration in joint function and failure of conservative therapy. The Haemophilia Chartered Physiotherapist's Association (UK) have adopted a functional score combining the clinical joint score [11] and Colorado (PE 1) physical examination scoring instrument [12] where each criterion is scored from 0 (no impairment) to 3 (severe impairment; Fig. 1). The indications for orthopaedic surgery usually focus on chronic pain, increasing bleed frequency and deteriorating function in the problem joint. Deterioration of other joint function caused by altered function or movement patterns may also indicate a need for surgery. Objective tests are important during the evaluation because this patient group often under-reports the severity of symptoms, particularly those of pain, which often do not correlate with radiographic images [13]. Once the decision for orthopaedic surgery has been made, the patient with haemophilia undergoes approximately 6 weeks of rehabilitation prior to the operation. The role of



Fig. 1. Knee joint of a patient with haemophilia showing severe impairment (score of 3) according to the UK Haemophilia Chartered Physiotherapist's Association Joint Score.

prehabilitation cannot be overemphasized; this period is so important that, if the patient fails to comply with the preoperative rehabilitation regimen, surgery may be postponed. A detailed assessment of both the surgical and non-surgical joints is made at this time and includes range of motion, muscle strength, gait and the postoperative requirements for gait aids. In addition, the ease of performance of activities of daily living is evaluated. All of this information is vital in designing an appropriate preoperative and postoperative rehabilitation regimen.

The benefits of preoperative and early postoperative rehabilitation in patients with haemophilia, undergoing orthopaedic surgery are illustrated by the following case study.

### Case study

The patient, a 48-year-old Caucasian male (body-weight: 80 kg) with congenital haemophilia A (diagnosed at 18 months) and inhibitors to factor VIII ( $<1 \text{ IU dL}^{-1}$ ; inhibitor titre:  $>300 \text{ BU}$ ) was scheduled for total left knee replacement due to progressive deterioration of the joint following repeated haemarthrosis.

Although frequent bleeding had occurred into the left knee joint since infancy, there had been no

**Table 1.** Case study – functional status of the left knee in the 3 years prior to surgery.

	1999	2002
Mobility	>1 mile day <sup>-1</sup>	Wheelchair
Range of motion	10–90°	25–45°
Flexion contracture	10°	25°
Varus deformity	10°	15°

**Table 2.** Case study – preoperative joint scores.

	Right joint	Left joint
Ankle (24)	6	4
Knee (24)	0	21
Hip (24)	0	0
Elbow (21)	13	0
Shoulder (21)	0	0

A normal score in each case is 0, indicating that the joint is unaffected. Maximum scores (in brackets) in each case indicate severe impairment.

bleeding into the right knee joint. Table 1 shows the documented deterioration of the left knee over the 3 years prior to surgery. Mobility declined significantly: in 1999 the patient could walk the mile to and from work each day whilst in 2000 he could only manage this journey on certain days. By 2001, mobility had declined to journeys of 0.5 mile day<sup>-1</sup> and by 2002 he was only mobile indoors, using a wheelchair for outdoor activity. During this time, range of motion of the left knee decreased from 10–90°, 10° varus in 1999 to 25–45°, 15° varus in 2002; extension was decreased and contracture was increased, with varus deformity. In the year prior to surgery (2001–02), the joint score (Haemophilia Chartered Physiotherapist's Association Joint Score) for the affected knee deteriorated from 19 to 21. As the patient became less mobile, bleeding frequency into the left knee also decreased. Other preoperative joint scores are detailed in Table 2. The right elbow, left ankle and right ankle were affected by arthropathy, but other joints remained largely unaffected.

The patient was prescribed 6 weeks' preoperative rehabilitation. This involved two exercise sessions per week: a hydrotherapy session, during which reduced weight-bearing exercises were used to increase muscle strength and improve mobility; and a physiotherapy gym session. Prehabilitation focused on exercises specifically designed to increase the strength of the patient's quadriceps, hip extensors, hip abductors and trunk stabilizer muscles and incorporated exercises the patient would perform during the postoperative period. No prophylactic haemostatic cover was administered for the physiotherapy sessions during this prehabilitation period.

On the day of the operation, a peripherally inserted catheter (PIC) line was placed prior to surgery. After surgery, bleeding was controlled with recombinant activated factor VII (rFVIIa; Novo-Seven®, Novo Nordisk, Bagsvaerd, Denmark) 90 µg kg<sup>-1</sup> for 10 days using the following dosing intervals: 2-hourly for 2 days, 3-hourly for 2 days, 4-hourly for 2 days, 6-hourly for 3 days.

On return from surgery, the patient was nursed in the intensive care unit for 48 h. The operated leg was fully extended and wrapped in a compression bandage, and the foot of the bed was slightly raised to minimize swelling. Continuous passive motion was not used in this case. Postoperative rehabilitation began immediately. On day 0 (in the evening on the day of surgery), exercise of the left leg commenced and included static isometric exercises of the quadriceps and gluteal muscles, and foot and ankle movements. Movement of the right leg was also encouraged. On postoperative day 1, the static exercises of the left quadriceps and gluteus muscles, and foot and ankle movements were repeated as frequently as possible. In addition, five formal physiotherapy sessions were scheduled to coincide with the rFVIIa bolus delivery. Exercises included active/active-assisted knee flexion to 30°, inner range quadriceps strengthening, and hamstring and calf stretches. On postoperative day 2, the patient was transferred to the ward and surgical drains were removed. Five formal physiotherapy sessions, again timed to coincide with the rFVIIa bolus delivery, were scheduled. On day 2, the patient stood for the first time, placing weight on the left leg, and walking 10 m with a wheeled walker. Later that evening, the patient was encouraged to self-exercise when subsequent rFVIIa boluses were administered. On postoperative days 3–7, the number of formal physiotherapy sessions was gradually reduced (four physiotherapy sessions on days 3 and 4, three sessions on days 5 and 6 and two sessions on day 7) in parallel with the reduction in the number of rFVIIa boluses given. The duration of each physiotherapy session was, however, gradually increased. The types of strengthening and range of movement exercises were advanced as the patient's condition progressed. At all times, care was taken to minimize the risk of bleeding. By postoperative day 7 the patient had 10–70° of active knee flexion and twice daily walked 70 m on gutter crutches. Gutter crutches were necessary due to the poor condition of the patient's right elbow joint (Table 2). At this time, the continued presence of an active quadriceps extension lag during a straight leg raise focused physiotherapy on improving quadriceps strength. Recovery of both

extension and flexion of the knee joint is important; the former because full extension is necessary for walking (to reduce the stresses on the knee joint) and the latter because flexion of at least 60–70° is necessary for comfort during sitting and stair climbing. Although greater flexion can be achieved, this is often at the expense of extension. During postoperative days 8–10, the patient continued with two formal physiotherapy sessions per day. Mobilization was increased to 100 m, at least once daily, and left knee range of motion increased to 10–85°. Quadriceps strength improved, and by day 10 the previously observed quadriceps lag had been eliminated.

The patient was discharged home on postoperative day 10. He was treatment compliant and motivated, readily continuing the exercises in the home setting. During week 3 (postoperative days 15–21) he attended the Haemophilia Centre for a daily formal physiotherapy session. In week 4, these sessions were reduced to twice weekly, and in weeks 5 and 6 to once weekly. By week 6 (postoperative days 36–42) flexion of the left knee had increased to 7–90° and the patient was able to walk 400–500 m with a walking stick with good lower limb muscle strength. During week 6, recurrent bleeding into the gastrocnemius muscle occurred due to increased stretching of the calf muscle as knee extension in weight-bearing exercises increased. A temporary heel raise to reduce the knee extension was placed in the left shoe for 2 weeks and gradually removed over the next 2 weeks. At this point, hydrotherapy was initiated. Postoperative physiotherapy rehabilitation was also targeted at the deteriorating right elbow to increase the strength of biceps, triceps and wrist extensor muscles and to reduce the bleed frequency and improve function.

The patient continued to benefit considerably from surgery until his death in 2004, which was caused by a hepatoma and unrelated to orthopaedic surgery. Despite his untimely death, this case report demonstrates the importance of successful orthopaedic surgery to the mobility and function of patients with haemophilia-related arthropathy.

## Conclusion

Successful outcomes following orthopaedic surgery depend on all members of the multidisciplinary team working closely together. This case study describes the principles of a successful programme of physiotherapy 'prehabilitation' and rehabilitation which can be utilized for patients with haemophilia undergoing orthopaedic surgery. Due to the risk of

postoperative bleeding and the unique multiple joint arthropathy found in patients with haemophilia, any physiotherapy rehabilitation programme needs to be tailored to the specific needs and function of the individual patient.

## References

- 1 Rodriguez-Merchan EC. Orthopaedic surgery in persons with haemophilia. *Thromb Haemost* 2003; **89**: 34–42.
- 2 Edgerton VR, Zhou MY, Ohira Y, *et al.* Human fibre size and enzymatic properties after 5 and 11 days of spaceflight. *J Appl Physiol* 1995; **78**: 1733–9.
- 3 Thompson V. Skeletal muscle adaptations with age, inactivity and therapeutic exercise. *J Orthop Sports Phys Ther* 2002; **32**: 44–57.
- 4 Ditmyer MM, Topp R, Pifer M. Prehabilitation in preparation for orthopaedic surgery. *Orthop Nurs* 2002; **21**: 43–51.
- 5 Ackerman IN, Bennell KL. Does pre-operative physiotherapy improve outcomes from lower limb joint replacement surgery? A systemic review. *Aust J Physiother* 2004; **50**: 25–30.
- 6 McDonald S, Hetrick S, Green S. Pre-operative education for hip or knee replacement. *Cochrane Database Syst Rev* 2004; **1**: CD003526.
- 7 Vervest AM, Maurer CA, Schambergen TG, de Bie RA, Bulstra SK. Effectiveness of physiotherapy after meniscectomy. *Knee Surg Sports Traumatol Arthrosc* 1999; **7**: 360–4.
- 8 Munin MC, Rudy TE, Glynn NW, Crossett LS, Rubash HE. Early inpatient rehabilitation after elective hip and knee arthroplasty. *J Am Med Assoc* 1998; **279**: 847–52.
- 9 Moffet H, Richards CL, Malouin F, Bravo G, Paradis G. Early and intensive physiotherapy accelerates recovery postarthroscopic meniscectomy: results of a randomized controlled study. *Arch Phys Med Rehabil* 1994; **75**: 415–26.
- 10 Rajan RA, Pack Y, Jackson H, Gillies C, Asirvatham R. No need for outpatient physiotherapy following total knee arthroplasty: a randomized trial of 120 patients. *Acta Orthop Scand* 2004; **75**: 71–3.
- 11 Hill FGH, Ljung R. Third and fourth workshops of the European Paediatric Network for Haemophilia Management. *Haemophilia* 2003; **9**: 223–8.
- 12 Manco-Johnson M, Nuss R, Funk S, Murphy J. Joint evaluation instruments for children and adults with haemophilia. *Haemophilia* 2000; **6**: 649–57.
- 13 Wallny T, Lahaye L, Brackmann H, Heb L, Sueser A, Kraft C. Clinical and radiological scores in haemophilic arthropathies: how well do these correlate to subjective pain status and daily activities. *Haemophilia* 2002; **8**: 802–8.