Planning and undertaking elective surgery in people with haemophilia (PWH) is most effective with the involvement of a specialist and experienced multidisciplinary team (MDT) at a haemophilia treatment centre. However, despite extensive best practice guidelines for surgery in PWH, there may exist a gap between guidelines and practical application. For this consensus review, an expert multidisciplinary panel comprising surgeons, haematologists, nurses, physiotherapists and a dental expert was assembled to develop practical approaches to implement the principles of multidisciplinary management of elective surgery for PWH. Careful preoperative planning is paramount for successful elective surgery, including dental examinations, physical assessment and prehabilitation, laboratory testing and the development of haemostasis and pain management plans. A coordinator may be appointed from the MDT to ensure that critical tasks are performed and milestones met to enable surgery to proceed. At all stages, the patient and their parent/caregiver, where appropriate, should be consulted to ensure that their expectations and functional goals are realistic and can be achieved. The planning phase should ensure that surgery proceeds without incident, but the surgical team should be ready to handle unanticipated events. Similarly, the broader MDT must be made aware of events in surgery that may require postoperative plans to be changed. Postoperative rehabilitation should begin soon after surgery, with attention paid to management of haemostasis and pain. Surgery in patients with inhibitors requires even more careful preparation and should only be undertaken by an MDT experienced in this area, at a specialized haemophilia treatment centre with a comprehensive care model.

**KEYWORDS**
elective surgery, haemophilia, multidisciplinary, physical therapy, planning, recommendations
INTRODUCTION

Haemophilia is a chronic condition, requiring specialist treatment throughout the patient’s lifespan. It is therefore recommended that people with haemophilia (PWH) are medically managed in a comprehensive care setting, with access to an integrated multidisciplinary team (MDT) of appropriate specialists. Management of surgical interventions at, or in consultation with, a comprehensive haemophilia treatment centre (HTC) is established best practice. The HTC is an integrated expert MDT, comprising at its core a haematologist, nurse, physiotherapist, laboratory specialist, social worker and/or psychologist. The broader comprehensive care team includes specialists with experience in working with PWH, including surgeons (particularly orthopaedic, but other specialists/generalists may be involved), a physical medicine and rehabilitation (PMR) physician (where available), an anaesthesiologist, dentist and pharmacist. Planning for surgery is complex, and interactions between specialists are typically more extensive than those required for patients without underlying bleeding disorders. Close collaboration and communication among the MDT members and the patient/family is vital throughout. Surgery in inhibitor patients remains the most challenging area and should only be conducted at an HTC by an experienced, specialized MDT.

The importance and organization of the MDT has been discussed in depth elsewhere; however, a gap can exist between best practice perspectives and practical application. With this in mind, a multidisciplinary panel of specialists was convened to provide practical recommendations on the application of the established principles of multidisciplinary management of elective surgery and the comprehensive care model for PWH. Two hypothetical examples of best practice for surgical interventions in PWH are provided in Appendix S1.

MATERIALS AND METHODS

An expert panel comprising haematologists (MAE, AS, VJ-Y), orthopaedic surgeons (LPS, HC, ECR-M), specialist haemophilia nurses (LL, JOOO), specialist physiotherapists (AF, PM, SL) and a dental expert (AB) reviewed the topic of multidisciplinary surgical care in haemophilia. The outcomes of this meeting are reported here as a narrative summary with supporting references.

Principles of elective surgical planning for the MDT

Frequent, recurrent and uncontrolled joint bleeding resulting from chronic synovitis leads to persistent pain and joint deformity that may be addressed by radiosynovectomy and orthopaedic surgery, respectively. Delayed referral to an orthopaedic surgeon may result in chronic synovitis and joint damage, requiring more aggressive surgical intervention. Regular monitoring at the HTC of joint status of patients who experience frequent joint bleeds and/or pain can help to identify candidates for orthopaedic surgery and prevent delayed referral. Although not the focus of this article, fractures, septic joints and compartment syndromes must be treated as an emergency. With compressed preparation time these episodes are best referred to an HTC with an experienced MDT, where resources and specialist experience are available to respond to the urgency of the condition.

Ongoing communication among the MDT and early involvement of a physiotherapist and/or PMR physician are required for prompt referral to surgery. Before the patient provides consent it is important that they understand the planning process, surgical procedure and potential outcomes; discuss their personal goals; and have any concerns and fears allayed. In advance of surgery the haemophilia nurse, psychologist or social worker explores with the patient their mindset and assesses their social support structure. The physiotherapist may discuss realistic postoperative rehabilitation and disposition planning.

Elective surgery requires detailed, timely and coordinated preparation. The MDT members respond to changing situations that require plan amendments, even at short notice. Table 1 summarizes some of the key responsibilities of the MDT members surrounding elective orthopaedic surgery.

Throughout all phases of surgery, it is important that patient expectations of the outcome are communicated, recorded, understood and managed by the MDT, and aligned with the procedure.

The MDT coordinator

In addition to their specific responsibilities, a full-time permanent member of the MDT may take on a leadership role as a coordinator (summarized in Figure 1). The coordinator is responsible for ensuring that all steps associated with surgical preparation, procedure performance and required rehabilitation are executed efficiently and thoroughly. A checklist may be utilized (Table 2 and Table S1). The coordinator facilitates consistent communication among the MDT members and the patient/family, and confirms the availability of resources, services and facilities to support surgery and rehabilitation.

The role of coordinator may shift between MDT members, depending on the phase of planning. For example, the surgeon may be best suited to coordinate in the intraoperative period, and a physiotherapist suited for the outpatient period. In all cases, the MDT assures smooth handover between different phases, is aware of the patient’s requirements and expectations, and ensures continuity of care. The coordinator role may be held by a specialist local to the patient, with extensive communication back to the HTC.

Outpatient preoperative planning period

The preoperative planning period begins following an agreement to proceed to surgery. The duration of the planning phase varies case-by-case, according to urgency, pathology, needs and status of the patient, haemostatic complexity and resource availability. MDT members must agree on the clinical implementation of treatment protocols for surgery to assure team alignment translates into consistent communication with the patient/family, treatment plan development...
### TABLE 1 Roles and responsibilities of the multidisciplinary team specialists around elective surgery

| Specialist | Outpatient preoperative planning | Inpatient preoperative planning | Intraoperative | Postoperative recovery | Physical rehabilitation |
|------------|---------------------------------|--------------------------------|---------------|-----------------------|-------------------------|
| **Surgeon** |  |  | Full assessment of patient’s physical condition | Surgery Management of unexpected bleeding events | Follow-up with patient Discussing postoperative rehabilitation |
| Haematologist | Evaluation of patient’s suitability for surgery with patient/family on goals and realistic expectation of surgical outcome | Planning procedure Scheduling of surgery | Confirmation of haemostatic plan | Ensuring appropriate haemostatic cover during surgery Management of unexpected bleeding events | Ensuring appropriate haemostatic cover in postoperative period Assessment for risk of postoperative thrombotic events and inhibitor development Management of unexpected bleeding events Discussion of postoperative rehabilitation Ensuring appropriate haemostatic cover, tailored to patient’s needs, and communication of factor regimen to physiotherapist |
| **Haemophilia nurse** | Teaching ward/surgical nurses how to reconstitute and administer clotting factor concentrates | Understand patient concerns/willingness to undergo surgery Motivate patient, help to address questions and allay any fears Liaison with patient and anaesthetist to establish pain management plan | Evaluation of patient’s suitability for surgery Liaison with surgical team to ensure treatment plan is understood and carried out Briefing of ward staff to ensure patient needs (eg haemostasis and pain management) are understood | Ensuring appropriate haemostatic cover during surgery | Monitoring of patient and ensuring haemostatic cover is in place (regular and timely infusions) Regular meetings with patient/caregivers; motivate to maintain treatment regimen |
| **Surgical nurse** | Learning how to reconstitute and administer clotting factor concentrates | Preparation of patient for surgery |  |  |  |
| **Social worker** | Evaluation of patient’s suitability for surgery and home environment (family support, suitability of home for recovery) |  |  |  | Follow-up of suitability for surgery and home environment (family support, suitability of home for recovery) |
| **Physiotherapist and PMR physician** | Full assessment of patient’s physical condition Advise patient on prehabilitation exercises Preparation of rehabilitation plan (aligned with patient goals and expectations) involving family to explain long-term commitment to rehabilitation programme Assessment of aftercare needs, home situation and equipment needs | Full assessment of patient’s physical condition and communication to surgery team to better advise surgery team on positioning of patient on table | Observe surgery to prepare for future rehabilitation needs Implementation of rehabilitation plan Balancing range of motion and increasing functional mobility with the need to prevent bleeding. Possible use of splint/immobilization initially vs continuous passive motion | Implementation of rehabilitation plan Briefing of local physiotherapist to work with patient in rehabilitation Assessment of quality of life, pain, activities of daily living Observation of any bleeding and feedback of other needs for haemostatic cover to haematologist |

(Continues)
### TABLE 1 (Continued)

| Specialist            | Outpatient preoperative planning                                      | Inpatient preoperative planning | Intraoperative | Postoperative recovery | Physical rehabilitation |
|-----------------------|-------------------------------------------------------------------------|---------------------------------|----------------|------------------------|-------------------------|
| Dentist/hygienist     | Dental checks for inflammation and tooth decay (risk of infection), including radiographic assessment of the dentition and mandible and maxilla |                                 |                |                        |                         |
| Psychologist          | Evaluation of suitability for surgery                                  |                                 |                |                        | Monitoring patient rehabilitation |
|                       | Understand patient concerns/willingness to undergo surgery             |                                 |                |                        |                         |
|                       | Motivate patient, help to address questions and allay any fears        |                                 |                |                        |                         |
| Special coagulation laboratory | Screening for inhibitor presence or inhibitor level                     | Prioritization of samples for measurement of baseline coagulation profile; screening of inhibitor presence or inhibitor level |                | Prioritization of samples to monitor plasma factor levels and screening for inhibitor presence or inhibitor level, including at nights and weekends if necessary |                          |
| Pharmacist/HTC        | Ensure adequate amount of factor/bypassing agents are available based on the haemostatic plan | Ensure concentrate is available as needed in the operating room |                | Coordinate with haemophilia nurse on daily dose of factor or bypassing agent (bolus or continuous infusion), factor levels and availability of concentrate |                          |

*Note: PMR, physical medicine and rehabilitation.*
and conduct. In some countries it is required to communicate with and obtain authorization from the healthcare insurer for haemostatic coverage and rehabilitation far in advance of surgery.

2.2.1 | Managing patient expectations

Meetings between the patient/family and the MDT are held early to ensure alignment between the patient’s and the MDT’s expectations of surgical outcomes, realistic functional goals and the patient’s role in their own recovery. Requirements differ between patients; the needs of a 20-year-old with a painful knee who desires to participate in activities with his friends will contrast with those of a 60-year-old with limited elbow motion. Motivations and expectations affect patient perception about surgery; reluctance, fears, individual capability for, and commitment to, rehabilitation is identified and discussed with the relevant MDT members. The haemophilia nurse is often best able to address these issues, but a social worker or psychologist may be involved to gain a deeper understanding of a patient’s concerns.

Discussion of “the road to surgery” between the MDT must include procedure description, associated risks, required commitment during the rehabilitation period, and expected outcome. Commitment during rehabilitation includes any practical difficulties the patient may face travelling to the HTC for rehabilitation appointments (eg limited mobility or access to transport after a knee replacement). Postoperative infusion requirements for patients with moderate haemophilia may include training on infusing clotting factor or insertion of a peripherally inserted central catheter for home treatment during the recovery period. With the patient, the physiotherapist and social worker evaluate the home recovery environment and physical aids required. The coordinator ensures that these meetings occur in a timely fashion and that outcomes are communicated between MDT members so that plans are established.

2.2.2 | Dental care

Adequate oral hygiene is important in PWH to prevent periodontal disease and other dental complications, which can be a source of postoperative infection, particularly in the case of total joint replacement. As resolution of oral infection may require several weeks, a dental surgeon should be involved early in the planning process to assess possible oral sources of infection. Prophylactic antibiotic treatment is typically not required. The MDT should keep apprised of issues identified by the dental surgeon and any need for invasive procedures, as these may create the need for an additional procedure and procedural planning.

2.2.3 | Pain management planning

The haematologist, anaesthetist and patient discuss the patient’s pain threshold and assess any increased tolerance to opioid medications; this information can influence plans for pain management, postoperative recovery and physical rehabilitation. Analgesics currently used should be identified and the need for increased doses in the perioperative period assessed. In patients with inhibitors, a nerve block would typically be avoided, owing to the risk of perforating a blood vessel and the consequences of bleeding into a confined space. A postoperative pain management plan is prepared, covering inpatient recovery and postdischarge. Preparations for postoperative pain management, both in the hospital and following discharge, are made. The anaesthetist liaises with the haemophilia nurse to confirm other medications the patient is taking. It is important for the haemophilia nurse to raise awareness that medication dosages for daily and acute pain may be greater than in patients without haemophilia. Guidelines from the World Federation of Hemophilia cover postoperative pain management.

2.2.4 | Physical assessment and prehabilitation

A comprehensive PMR baseline assessment is necessary to identify reduced muscle strength or damage to multiple other joints that may affect postoperative rehabilitation. This information may be used to predict postoperative function and ascertain rehabilitation limitations postprocedure.

In patients with advanced joint disease, the physiotherapist (or PMR physician if available at the site), haemophilia nurse, social worker and psychologist work together to ensure that the patient’s physical and mental condition prior to surgery is optimal. Assessments of multiple joint involvement and target areas can make the surgical team aware of special positioning or considerations for
the procedure. The physiotherapist, surgeon and haematologist work with the patient to determine the optimal approach to physical rehabilitation and assess progress.

“Prehabilitation,” defined as a proactive approach to health and behaviour change to improve an identified outcome, should be initiated. Preoperative physiotherapy to prepare joints/muscles in the region of the procedure theoretically enhances a patient’s capacity to withstand surgery and preserve function in the postoperative period.15,23,24 Some patients may find exercise too painful to perform; prehabilitation then involves discussing what will happen before, during and after surgery, building familiarity with postoperative exercises and use of mobility aids, and further developing a trusting patient–MDT relationship. Proactive attempts to manage self-efficacy and patient expectations before surgery help to mitigate anxiety and decrease postoperative pain.25 Patient adherence, a key factor affecting outcomes, can also be assessed.23,26 Although the role of prehabilitation in the general population is controversial, there is some indication that the approach may have a small and short-lived effect on postoperative pain and function in joint replacement.27,28 Current data are lacking on the efficacy of preoperative physiotherapy in haemophilia patients with severe joint damage; however, prehabilitation retains value for the importance of rehabilitation education. Developments or decisions that affect the intraoperative or postoperative period are shared with the MDT.

### 2.2.5 Assessment of comorbidities

All patients should be assessed for comorbidities, including cardiopulmonary, renal or liver disease to ensure appropriate anaesthetic management.8,27 If the patient has hepatitis C, liver function should be assessed before surgery. If positive for HIV, CD4 levels are obtained and requirement for perioperative antibiotic prophylaxis assessed. The type of protease inhibitor the patient is receiving may require suspension of medications prior to anaesthesia to prevent possible interrelated drug toxicity. Special attention should be paid to older patients in whom the rate of viral disease or comorbidities (such as cardiovascular disease or cancer) may be greater than younger patients.30 Also, the risk of venous thromboembolism in older patients is elevated and the potential effects on thrombosis risk of excessive factor (F)VIII replacement during the peri-operative period are unclear.30

### 2.2.6 Haemostasis planning

After assessment of the patient’s underlying factor deficiency, inhibitor status and associated complications or comorbidities are aligned with the planned surgical procedure and the haematology team develops an individualized haemostatic management plan, including the preoperative, intraoperative and rehabilitation periods. Replacement therapy, doses and intervals are established, including method of administration (bolus/continuous infusion), as well as the need for concomitant antifibrinolytic therapy.1,31,32 A contingency plan should be in place (eg pharmacy and blood bank support) if unexpected bleeding occurs. The quantity of clotting factor product required for surgery is determined. Patients with mild haemophilia A undergoing surgery may not require clotting factor concentrate and may be treated with non-specific haemostatic agents (eg desmopressin acetate). Less-invasive procedures, such as radio- and chemical synovectomy, typically require smaller amounts of clotting factor concentrates. Prior to major elective procedures, an in vivo recovery and half-life study should be considered when the patient is not bleeding, using either a standard or a Bayesian approach to pharmacokinetic analysis.33,34

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**TABLE 2 Example multidisciplinary team coordinator checklist**

| Task                              | Completed (tick) or not applicable (N/A) |
|-----------------------------------|-----------------------------------------|
|                                  | C  | S  | SN | A  | H  | HN | Phy | D  | Pha | L  | Ps | SW | Notes |
| Outpatient planning               |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Indication for surgery            |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Inpatient preoperative planning   |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Patient assessment                |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Inpatient postoperative recovery  |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Postoperative assessment          |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Rehabilitation                    |    |    |    |    |    |    |     |    |     |    |    |    |        |
| Initiation of physical rehabilita- |    |    |    |    |    |    |     |    |     |    |    |    |        |
| tion plan                         |    |    |    |    |    |    |     |    |     |    |    |    |        |

Notes. See online Table S1 for a populated version of this example table. Fields in the table may be used by the coordinator when planning orthopaedic surgery in a person with haemophilia. The “task” column may be completed with the tasks relevant to the planning of surgery (see main text and Table 1). The columns in the “completed or not applicable” section contain tick boxes to indicate when individual tasks have been completed; if the task is not applicable to one or more of the listed specialists then “N/A” may be written by the coordinator. The “notes” column can be used to record comments for communication between team members on outcomes of discussions, points to followup and any concerns raised. The table should be customized according to the patient and procedure.

A, anaesthetist; C, coordinator; D, dentist; H, haematologist; HN, haematology nurse; L, laboratory; Pha, pharmacist; Phy, PMR (physical medicine and rehabilitation) physician or physiotherapist; Ps, psychologist; S, surgeon; SN, surgical nurse; SW, social worker.
Attention to the possibility of inhibitor development during and following surgery requiring replacement therapy is imperative. A factor assay monitoring schedule must be coordinated with the coagulation laboratory to ensure timely and accurate results to permit dosing adjustments. Expertise in inhibitor identification and treatment must be available. Some patients with non-severe haemophilia A may have mutations that increase their risk of inhibitor development after intensive exposure to FVIII. 35 The haemostatic plan must be approved by the haematologist, communicated to all teams involved in the surgery, and a copy filed in the medical records. Patients with existing inhibitors require additional attention, and the quantity and mode of administration of bypassing agents and need for adjunctive therapy (eg antifibrinolytics) agreed upon. 31

2.2.7 | Laboratory testing

Appropriate laboratory testing is needed to confirm patient status. An inhibitor titre measurement should be available within 2 months of scheduled surgery. 31 Assessment of other coagulation parameters (eg prothrombin time) is performed to assure an otherwise intact haemostatic system. Baseline laboratory evaluation including complete blood count and chemistries is obtained to determine adequacy of haemoglobin and platelet count, and overall liver/renal function. The MDT should be aware if a patient is immunocompromised, so that proper precautions are taken to prevent infection. The specialized laboratory team is informed by the haematologist in advance of anticipated factor assay samples and need for prioritized results, including capacity to analyse samples during nights and weekends, if required.

2.2.8 | Scheduling of surgery

Surgery should ideally occur early in the week and day to ensure the availability of all MDT members and access to required specialized laboratory testing. Availability of resources such as hospital beds and blood bank support is also considered. If the patient requires multiple surgical interventions it may be possible to schedule them for the same period, to optimize resources.

Once scheduled, the haematologist communicates with the relevant department (usually the pharmacy or blood bank) to ensure there is sufficient clotting factor concentrate/bypassing agent for the entire perioperative period. The haemophilia nurse communicates with the ward staff and pharmacist regarding the specific requirements of a postoperative haemophilia patient with particular attention to reconstitution, administration and timing of delivery of factor concentrate or bypassing agent.

2.3 | Inpatient period

2.3.1 | Perioperative planning

When the patient is admitted for surgery, perioperative planning and checks begin. The surgeon takes the lead as the individual with direct patient responsibility during surgery. The haemophilia nurse assures adequate haemostasis is achieved and maintained by confirming administration of the prescribed clotting factor concentrate immediately before surgery. Coagulation factor assay results are provided to the haematologist, who confirms that the haemostatic plan is appropriate.

2.3.2 | Intraoperative period

The intraoperative period begins when the patient is transferred to the operative room and ends with completion of the procedure and transfer to the recovery (postanaesthesia care) unit. The surgeon and the haemophilia coordinator (ie haematologist, nurse) remain involved to assure adequate haemostasis, required laboratory testing and optimal outcome. The surgical team should always remain prepared to manage and communicate unexpected bleeding, with the haematologist available to work with the surgical team as required.

Preoperative assessment can inform a checklist for the position of the patient on the operating table, and used to place gel pads necessary to avoid external compressions and prevent forced positions. Other MDT members may observe the procedure; for example, the treating physiotherapist may be able to prepare for future rehabilitation needs, particularly in case of unanticipated events.

After completion of the patient’s surgery and transferal to the recovery room, factor activity levels should be assayed if bypassing agents are not being used, and clotting factor administered as required. Other necessary routine checks are made, such as intravenous drips and appropriate function of monitors. The nurses in the recovery room should be made aware of any specific requirements previously established.

2.4 | Postoperative recovery period

The main goal of an inpatient stay is maintenance of adequate haemostasis and initiation of required rehabilitation to assure a safe discharge. For PWH without inhibitors, rehabilitation begins immediately; as early as the transfer to the postanaesthesia care unit. The role of the MDT coordinator may shift from the surgical service to the medical or haematology service, although the surgeon will remain involved to respond to surgical queries.

2.4.1 | Haemostasis and thrombosis prophylaxis

The haemophilia nurse ensures that the patient’s haemostatic plan is carried out and that haemostasis is adequately monitored, particularly for patients with severe haemophilia requiring frequent or continuous infusions of clotting factor concentrate. Patients with inhibitors require careful monitoring to ensure that they receive consistent, timely treatment with the appropriate bypassing agent. 6 The haematologist is available to determine changes to the haemostatic plan as required. The surgical and haematological teams monitor wound healing and drainage. The MDT should be fully aware of concomitant medications the patient receives, their potential impact
on coagulation, and need for continuity throughout the hospital stay. Patients receiving clotting factor concentrate by continuous infusion are monitored to ensure appropriate product delivery and adequate factor levels. Inhibitor and clotting factor monitoring should be performed more frequently in patients with mutations that increase their risk of inhibitor development after intensive exposure to FVIII.

Incidence of symptomatic venous thromboembolism in PWH undergoing major orthopaedic surgery (some of whom received thromboprophylactict interventions) may be similar to that estimated in the general population not receiving thromboprophylaxis.36 Although there is no consensus in current haemophilia practice, a thrombosis risk assessment should be performed and postoperative prophylaxis considered. Minimally, the entire team should be aware of the potential for thrombosis and remain alert to clinical signs and symptoms.37

Mechanical prophylaxis with graduated compression stockings or impulse foot compression is commonly used. Chemical prophylaxis is uncommon.

2.4.2 Pain management

The pain management plan is implemented so that rehabilitation is not impeded. A patient may experience pain caused by both surgery and the posture required postoperatively. For instance, an immobilized patient with knees extended may experience pain due to stretching of posterior soft tissue elements, manifested as diffuse posterior knee and thigh pain/discomfort. Regular administration of pain medication contributes to a more consistent level of analgesia. The MDT, especially the haemophilia nurse, should ensure the ward staff are aware that PWH may require higher than expected doses of analgesics, which may include intravenous morphine or other narcotic analgesics, or an oral opioid such as tramadol, codeine or hydrocodone. Acetaminophen may be used when pain has decreased.1 If a patient is discharged on narcotics, the HTC staff should closely monitor use and possible side effects.

2.5 Physical medicine and rehabilitation

Physical medicine and rehabilitation are critical components of recovery following orthopaedic procedures and may be accomplished in the hospital or rehabilitation centre (as an inpatient), or the patient’s home (as an outpatient). When patients return to the community they often continue rehabilitation at the HTC or, if not possible, local providers can implement care plans communicated by the HTC physiotherapist. The importance of, and approaches to, postoperative physical rehabilitation have been discussed elsewhere.6,15,22,24,26,36–41 The patient should be aware that rehabilitation is likely to be lengthy and require significant time in the initial phases.

Physical rehabilitation after orthopaedic procedures may begin in the days after surgery; in patients with inhibitors, physical rehabilitation is often delayed by a few days to assure control of bleeding at the operative sites. The expected range of motion to be achieved should be communicated by the surgeon, or may be found in the medical chart/surgeon’s note. Postoperative postural pain may be addressed by physiotherapy and repositioning; measures taken should be communicated to the ward nursing staff for adequate ongoing care.

The physiotherapist plays a pivotal role in ensuring that the patient is trained and motivated to execute the established exercise plan. The physiotherapist and haemophilia nurse together confirm that haemostatic coverage is in place during physiotherapy, providing optimal balance between functional mobility recovery and minimization of bleeding risk. Reduction of postoperative infection risk is a high priority; prevention of bleeding by appropriate haemostatic cover and a surgical dressing to protect the incision site can contribute to this. All instances of bleeding should be recorded and reported promptly to the haematologist to assure required haemostatic rescue therapy.

Analgesia is made available to manage pain during rehabilitation. Cold compression, immobilization and splints may also be utilized in initial physiotherapy.2 Patients are encouraged to be as active as possible to positively influence pain responses, without being too aggressive and trigger a bleed.

People with inhibitors require extensive physiotherapy assessment to evaluate wound healing and assure adequate haemostasis before instituting therapy; these evaluations are communicated to the MDT so that treatment plans are adjusted as needed. Physiotherapy sessions are scheduled immediately following the administration of bypassing agents to reduce bleeding risk.

Patients may find the required exercises and pain experienced during the rehabilitation period challenging. The haemophilia nurse and physiotherapist offer support and motivation, emphasizing the benefits of adherence to the management plan. The physiotherapist intermittently assesses the patient for their current pain levels and progress in achieving identified functional activity goals and mobility. The social worker or occupational therapist may also be involved to ensure that the recovery environment is appropriate.

2.6 Relevance beyond the HTC

These recommendations are provided with the HTC in mind, where staff are experienced in surgery and rehabilitation of PWH, both with and without inhibitors, and there is access to resources and prompt on-site haemostatic monitoring.

People with haemophilia represent a high-risk population for surgery, and operative planning is optimally performed in conjunction with an HTC. Decisions to undertake elective surgery in PWH at non-HTC hospitals should be made on a case-by-case basis, accounting for patient status, local experience/expertise, availability of resources, and impact on potential outcomes. When planning a procedure, it is advisable to request guidance from an HTC on the feasibility of a procedure, or resources. Referral to an HTC is recommended if there is any question of achieving a positive outcome. It is strongly advised that any invasive procedure in PWH with inhibitors be referred only to an HTC with an experienced MDT.
3 | CONCLUSIONS

The skill and experience in a specialist MDT are critical for safe, effective elective surgery in PWH. This is complemented by clear communication between all MDT members, which may be facilitated by an MDT coordinator. Application of an agreed checklist can ensure that all steps in planning are taken in a timely fashion and the outcomes communicated within the MDT. Patient-directed communications should be equally clear and benefit from the involvement of the coordinator. Institutions without required resources to support the comprehensive MDT should seek practical advice on how to maximize their available facilities and appropriate decision-making in PWH. Knowledge of when a patient should be treated at an HTC is critical to assuring optimal outcomes.

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REFERENCES

1. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. Guidelines for the management of hemophilia. Haemophilia. 2013;19:e1-e47.
2. Soucie JM, Nuss R, Evatt B, et al. Mortality among males with hemophilia: relations with source of medical care. Blood. 2000;96:437-442.
3. Rodriguez-Merchan EC. Musculo-skeletal manifestations of haemophilia. Blood Rev. 2016;30:401-409.
4. WFH. Factsheet: structure and functions of comprehensive hemophilia treatment centres. 2011. http://elearning.wfh.org/resource/structure-and-functions-of-comprehensive-hemophilia-treatment-centres/. Accessed August 9, 2017.
5. Caviglia H, Narayan F, Forsyth A, et al. Musculoskeletal problems in persons with inhibitors: how do we treat? Haemophilia. 2012;18(Suppl 4):S4-60.
6. Escobar M, Maahs J, Hellman E, et al. Multidisciplinary management of patients with haemophilia with inhibitors undergoing surgery in the United States: perspectives and best practices derived from experienced treatment centres. Haemophilia. 2012;18:971-981.
7. Giangrande PL, Wilde JT, Madan B, et al. Consensus protocol for the use of recombinant activated factor VII [epoetin alfa (activated); NovoSeven] in elective orthopaedic surgery in haemophilic patients with inhibitors. Haemophilia. 2009;15:501-508.
8. Kulkarni R. Comprehensive care of the patient with haemophilia and inhibitors undergoing surgery: practical aspects. Haemophilia. 2013;19:2-10.
9. Mingot-Castellano ME, Alvarez-Roman MT, Lopez-Fernandez MF, et al. Spanish consensus guidelines on prophylaxis with bypassing agents for surgery in patients with haemophilia and inhibitors. Eur J Haematol. 2016;96:461-474.
10. Rangarajan S, Austin S, Goddard NJ, et al. Consensus recommendations for the use of FEIBA® in haemophilia A patients with inhibitors undergoing elective orthopaedic and non-orthopaedic surgery. Haemophilia. 2013;19:294-303.
11. Rodriguez-Merchan EC. Orthopedic surgery is possible in hemophilic patients with inhibitors. Am J Orthop (Belle Mead NJ). 2012;41:570-574.
12. Rodriguez-Merchan EC, Jimenez-Yuste V, Gomez-Cardero P, Alvarez-Roman M, Martin-Salces M, Rodriguez de la Rua A. Surgery in haemophilia patients with inhibitors, with special emphasis on orthopaedics: Madrid experience. Haemophilia. 2010;16:84-88.
13. Shapiro A, Cooper DL. U.S. survey of surgical capabilities and experience with surgical procedures in patients with congenital haemophilia with inhibitors. Haemophilia. 2012;18:400-405.
14. Solimeno LP, Perfetto OS, Pasta G, Santagostino E. Total joint replacement in patients with inhibitors. *Haemophilia*. 2006;12(Suppl 3):113-116.
15. Teitel JM, Carcao M, Lillicrap D, et al. Orthopaedic surgery in haemophilia patients with inhibitors: a practical guide to haemostatic, surgical and rehabilitation care. *Haemophilia*. 2009;15:227-239.
16. Carcao M, Hilliard P, Escobar MA, Solimeno L, Mahlangu J, Santagostino E. Optimising musculoskeletal care for patients with haemophilia. *Eur J Haematol*. 2015;95(Suppl 81):11-21.
17. duTreil S. Physical and psychosocial challenges in adult hemophilia patients with inhibitors. *J Blood Med*. 2014;5:115-122.
18. Morfini M, Benson G, Jimenez-Yuste V, et al. Tailoring care to haemophilia patients’ needs: which specialty and when? *Blood Transfus*. 2015;13:644-650.
19. Anderson JA, Brewer A, Creagh D, et al. Guidance on the dental management of patients with haemophilia and congenital bleeding disorders. *Br Dent J*. 2013;215:497-504.
20. Freedman M, Dougall A, White B. An audit of a protocol for the management of patients with hereditary bleeding disorders undergoing dental treatment. *J Disabil Oral Health*. 2009;10:151-155.
21. Sollecito TP, Abt E, Lockhart PB, et al. The use of prophylactic antibiotics prior to dental procedures in patients with prosthetic joints: evidence-based clinical practice guideline for dental practitioners – a report of the American Dental Association Council on Scientific Affairs. *J Am Dent Assoc*. 2015;146:11-16.e8.
22. De Kleijn P, Blamey G, Zourikian N, Dalzell R, Lobet S. Physiotherapy following elective orthopaedic procedures. *Haemophilia*. 2006;12(Suppl 3):108-112.
23. Forsyth A, Zourikian N. How we treat: considerations for physiotherapy in the patient with haemophilia and inhibitors undergoing elective orthopaedic surgery. *Haemophilia*. 2012;18:550-553.
24. Jimenez-Yuste V, Rodriguez-Merchan EC, Alvarez MT, Quintana M, Fernandez I, Hernandez-Navarro F. Controversies and challenges in elective orthopedic surgery in patients with hemophilia and inhibitors. *Semin Hematol*. 2008;45:S64-S67.
25. Auerswald G, Dolan G, Duffy A, et al. Pain and pain management in haemophilia. *Blood Coagul Fibrinolysis*. 2016;27:845-854.
26. Stephensen D. Rehabilitation of patients with haemophilia after orthopaedic surgery: a case study. *Haemophilia*. 2005;11(Suppl 1):26-29.
27. Kwok IH, Paton B, Haddad FS. Does pre-operative physiotherapy improve outcomes in primary total knee arthroplasty? – a systematic review. *J Arthroplast*. 2015;30:1657-1663.
28. Wang L, Lee M, Zhang Z, Moodie J, Cheng D, Martin J. Does pre-operative rehabilitation for patients planning to undergo joint replacement surgery improve outcomes? A systematic review and meta-analysis of randomised controlled trials. *BMJ Open*. 2016;6:e009857.
29. Shah UJ, Narayanan M, Graham Smith J. Anaesthetic considerations in patients with inherited disorders of coagulation. *BJA Educ*. 2015;15:26-31.
30. Philipp C. The aging patient with hemophilia: complications, comorbidities, and management issues. *Hematology Am Soc Hematol Educ Program*. 2010;2010:191-196.
31. Ingerslev J, Hvid I. Surgery in hemophilia. The general view: patient selection, timing, and preoperative assessment. *Semin Hematol*. 2006;43:S23-S26.
32. Negrier C, Shapiro A, Berntorp E, et al. Surgical evaluation of a recombinant factor VIII prepared using a plasma/albumin-free method: efficacy and safety of Advate in previously treated patients. *Thromb Haemost*. 2008;100:217-223.
33. Escobar MA. Products used to treat haemophilia: dosing. In: Lee CA, Berntorp E, Hoots WK, eds. *Textbook of Hemophilia*, 3rd edn. Chichester, UK: Wiley-Blackwell; 2014:180-184.
34. Morfini M. The history of clotting factor concentrates pharmacokinetics. *J Clin Med*. 2017;6:35.
35. Eckhardt CL, van Velzen AS, Peters M, et al. Factor VIII gene (F8) mutation and risk of inhibitor development in nonsevere hemophilia A. *Blood*. 2013;122:1954-1962.
36. Buckner TW, Leavitt AD, Ragni M, et al. Prospective, multicenter study of postoperative deep-vein thrombosis in patients with haemophilia undergoing major orthopaedic surgery. *Thromb Haemost*. 2016;116:42-49.
37. Ozelo MC. Surgery in patients with hemophilia: is thromboprophylaxis mandatory? *Thromb Res*. 2012;130(Suppl 1):S23-S26.
38. Blaney G, Forsyth A, Zourikian N, et al. Comprehensive elements of a physiotherapy exercise programme in haemophilia—a global perspective. *Haemophilia*. 2010;16(Suppl 5):136-145.
39. De la Corte-Rodriguez H, Rodriguez-Merchan EC. The role of physical medicine and rehabilitation in haemophilic patients. *Blood Coagul Fibrinolysis*. 2013;24:1-9.
40. Lobet S, Pendeville E, Dalzell R, et al. The role of physiotherapy after total knee arthroplasty in patients with haemophilia. *Haemophilia*. 2008;14:989-998.
41. Negrier C, Seuser A, Forsyth A, et al. The benefits of exercise for patients with haemophilia and recommendations for safe and effective physical activity. *Haemophilia*. 2013;19:487-498.

**SUPPORTING INFORMATION**

Additional supporting information may be found online in the Supporting Information section at the end of the article.

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