Clinical Practice Guidelines
for Physiotherapists working with persons with
Bleeding Disorders

2021

Companion document for
CPHC Standards of Physiotherapy Care
for Persons with Bleeding Disorders
Canadian Physiotherapists in Hemophilia Care Clinical Practice Guidelines

Revised January 11, 2021

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Introduction:

CPHC Clinical Practice Guidelines were originally developed in 2018 as a companion document to the CPHC *Standards of Physiotherapy Care for Persons with Bleeding Disorders*.

**Revisions to this 2021 version include:**

- Updated references, and inclusion of Levels of Evidence for all references
- Revised section on Physiotherapy before and after musculoskeletal surgery
- Conversion of the guidelines into Practice Statements
- Inclusion of “Grade of recommendation” for each Practice Statement

Additional modifications were made to align these guidelines with the 2020 Canadian Hemophilia Society *Canadian Integrated and Comprehensive Care Standards for Inherited Bleeding Disorders*.

https://www.hemophilia.ca/integrated-and-comprehensive-care-standards/

Key changes to the integrated and comprehensive standards include:

- The standards apply to “the entire family of inherited bleeding disorders”, not only to hemophilia
- The standards apply to all Persons with a Bleeding Disorder (PWBD) regardless of gender at birth
- The term Hemophilia Treatment Center (HTC) has been replaced with Treatment Center (TC)

Finally, in light of the new products coming available we have changed “factor replacement/coverage” to *Hemostasis Management*, which could also include non-infused products like DDAVP, and non-factor products such as Emicizumab.

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Standard 1: Each Canadian integrated and comprehensive bleeding disorders care team includes a physiotherapist.

Practice Statement 1.1
The TC physiotherapist should meet the PWBD /family soon after diagnosis*, and will provide assessment, treatment and education throughout the person’s lifespan.
Grade of recommendation: Theoretical

*Bleeding disorders that are associated with **Musculoskeletal (MSK) bleeding** include:

<table>
<thead>
<tr>
<th>Frequent</th>
<th>Common</th>
<th>Less Common</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemophilia A (FVIII deficiency)</td>
<td>Prothrombin deficiency</td>
<td>Factor V deficiency</td>
</tr>
<tr>
<td>Hemophilia B (FIX deficiency)</td>
<td>Factor X deficiency</td>
<td>Factor VII deficiency</td>
</tr>
<tr>
<td>Type 3 von Willebrand disease</td>
<td></td>
<td>Factor XIII deficiency</td>
</tr>
</tbody>
</table>

See Appendix A: Incidence and Clinical symptoms of Rare Bleeding Disorders

For individuals diagnosed with these conditions:

- The TC physiotherapist performs baseline and longitudinal assessments, and participates in the assessment of suspected MSK bleeds.  [See also Assessment of acute bleeds (S-2), Annual Assessment (S-4), and Pre-and post-operative assessments (S-6)]
- The TC physiotherapist arranges rehabilitation after acute bleeds, and for the sequelae of bleeding.  [See Treatment after Acute Bleeds (S-3) and Treatment of MSK Complications (S-5)]
- The TC physiotherapist will educate PWBD and their families regarding prevention, identification and management of bleeds and the potential MSK complications of bleeding disorders.

Rationale:
The WHO Chronic Care Model states that patients and families need to be informed about their chronic condition, including the expected course, potential complications, and effective strategies to prevent complications and manage symptoms.
PWBD need to be prepared with sufficient knowledge and skills to manage their condition at home. In addition to education about preventing and managing MSK bleeds and their sequelae, the physiotherapist will promote a healthy active lifestyle.
References:
Level of evidence: V- expert opinion

Level of evidence: V- expert opinion

Level of evidence: V- expert opinion

Level of evidence: Review

Level of evidence: Review

Level of evidence: V- expert opinion

Level of evidence: V- expert opinion
Practice Statement 1.2

The physiotherapist should consider using telerehabilitation, including online video conferencing platforms, for assessment, treatment and/or education of PWBD.

Grade of recommendation: Theoretical

Rationale:
The use of communications technology, if available and permitted by the TC/institution, can enhance self-management and decrease patient burden, such as lost time from school/work.

References:
Level of evidence: V-expert opinion

Level of evidence: III - Case controlled study
Standard 2: The physiotherapist participates in the assessment of acute musculoskeletal bleeds.

Practice Statement 2.1
Physiotherapy assessment of acute bleeds should include a detailed history of the present condition.
Grade of recommendation: Theoretical

Rationale/Discussion:
The earliest symptom of a musculoskeletal bleed is pain, BUT not all pain is due to bleeding.
History of present condition includes:
• details of bleed onset and progression of symptoms
• mechanism of injury
• hemostasis management used to date and response to treatment
• other treatment (first aid) used and response to treatment
• pain qualities, location, intensity, aggravating/alleviating factors
• pain with movement
By taking a detailed history, the physiotherapist might be able to recognize mechanisms of injury that correspond with the symptoms and subsequent physical exam findings.

References:
Key point: “the cause of the injury must be known, and the potential impact on the involved structures as well as their role in overall function must be recognized.”
Level of evidence: V- expert opinion

Key point: reports the words PWBD use to describe their sensation of bleeding in joints and muscles
Level of evidence: Cross-sectional study/Qualitative

Sørensen B, Benson GM, Bladen M, et al. Management of muscle hematomas in

Key point: “Cause of bleed should be determined by explicit questioning, especially in children”  
Level of evidence: V- expert opinion

Key point: Focus groups generated a list of items that patients and professionals use to differentiate between a joint bleed and a flare-up of Hemophilic Arthropathy.  
Level of evidence: Other/Qualitative

Key point: describes the language used by patients to describe and differentiate acute and persistent pain  
Level of evidence: Other/Qualitative

**Practice Statement 2.2**  
Physiotherapy assessment of acute bleeds should include past medical history.  
Grade of recommendation: Theoretical

**Rationale:**  
Past medical history includes:  
  - recent joint or muscle bleeds  
  - reasons for those bleeds  
  - previous target joints: location and number  
  - usual activities  
  - usual hemostasis management regime  
    - (factor or non-factor products)  
  - usual response to treatment for bleeds  
Repeated episodes of bleeding at the same site may indicate incomplete recovery of previous episodes, or shed light on a particular recurrent activity that contributes to re-injury.
References:
Key point: changes in bleeding frequency or severity should be thoroughly investigated and treatment modified as necessary
Level of evidence: V- expert opinion

doi:10.2147/JBM.S50644
Key point: “subacute hemarthroses arise after repeated episodes...in the same joint. Joint becomes a ‘target joint’ due to incomplete recovery.”
Level of evidence: V- expert opinion

doi:10.1111/j.1365-2516.2011.02720.x
Key point: “good history taking ... will allow planning of subsequent treatment and rehabilitation and enable an accurate prognosis.”
Level of evidence: V- expert opinion

Practice Statement 2.3
Physiotherapy assessment of acute bleeds should include a physical examination.
Grade of recommendation: Theoretical

Rationale/Discussion:
Physical exam should include:
- Observation/inspection:
  - posture/resting position
  - bruising** (see Notes)
  - swelling
  - redness
- Active Movement:
  - compare to contralateral limb or, in the case of chronic arthropathy, baseline measurements on file
• pain with movement

• Palpation:
  o warmth
  o swelling
  o tenderness
  o spasm

• Testing:
  o Joint end feel (within limits of pain)
  o Muscle length, especially two-joint muscles
  o Muscle strength, noting pain and/or weakness with active contraction
  o Sensation, especially if nerve compression is suspected

Physiotherapists routinely perform a physical exam to determine a clinical diagnosis and develop a treatment plan.

**NOTES:**

**Bruising:**
  • May be an indication of a superficial soft tissue bleed.
  • Bruising is NOT a feature of joint bleeds
• Bruising MAY be a late feature of a muscle bleed, but is not always present. **Accurate muscle testing will differentiate superficial soft tissue bleeds from muscles bleeds.**

**Presentation of an iliopsoas bleed** may mimic that of appendicitis, or be confused with a hip bleed. Signs of an iliopsoas bleed can include:

• Pain in groin, thigh, hip, OR lower back
• Hip flexed, sometimes externally rotated
• Abdominal tenderness
• Tingling or numbness in distribution of femoral nerve
• Weakness of quadriceps and/or decreased patellar reflex
• Unable to stand straight: hyperlordosis, knee flexed, and unable to place foot flat on floor
• Unable to lie flat

[Also note that psoas bleeds can be associated with sexual activity. Inquiry regarding recent sexual activity should be included in ‘history of present condition’.]

**References:**


*Key point:* “Thorough assessment must follow any acute injury to ensure that comprehensive and complete rehabilitation is being pursued”

Level of evidence: V- expert opinion


*Key Point:* International group of Hematologists developed definitions of Joint Bleed, Target Joint, Muscle Bleeds based on presenting symptoms, in order to facilitate uniform documentation of outcomes in clinical studies.

Level of evidence: V- expert opinion


*Key point:* reports the words PWBD use to describe their sensation of bleeding in joints and muscles

Level of evidence: Cross-sectional study /Qualitative
MASAC Recommendations Regarding Physical Therapy Guidelines in Patients with Bleeding Disorders: Iliopsoas.
Accessed January 10, 2021

**Key Point:** describes presenting signs and symptoms, and differential diagnosis
Level of evidence: V-expert opinion


**Key point:** describes common symptoms of muscle bleeds, including iliopsoas
Level of evidence: V-expert opinion


**Key points:** Physical examination for joint changes (e.g., in joint circumference, muscle strength, joint effusion, joint angle, pain according to a visual analogue scale) should be conducted at all routine follow-ups. (Chapter 10: Musculoskeletal Complications)
Level of evidence: V-expert opinion


**Key Points:** symptoms of hemarthrosis partly overlap with symptoms of flare-ups of Hemophilic Arthropathy. “Differentiating based on imaging techniques or biomarkers causes practical difficulties. Clinical assessment still seems the most convenient and practical solution.”
Level of evidence: V-expert opinion


**Key point:** Focus groups generated a list of items patients and professionals use to differentiate between a joint bleed and a flare-up of Hemophilic Arthropathy
Level of evidence: Other/Qualitative
Practice Statement 2.4
Physiotherapists who have received appropriate training may use point of care ultrasound (POCUS), if available, to confirm diagnosis and monitor resolution.

Grade of recommendation: Moderate

Rationale:
There is considerable overlap between signs and symptoms of acute bleeding and arthritis, which can make accurate diagnosis of joint bleeds challenging. Deep muscle bleeds may not be easy to palpate or measure. POCUS may be used as an adjunct to clinical assessment to confirm diagnosis, guide the treatment plan, and confirm whether hematoma/synovitis is resolved.

References:
Key points: MSK-US is a valuable point-of-care imaging tool to distinguish whether acute joint and/or musculoskeletal pain episodes in adult PWH are due to bleeding or not. Only approximately 1/3rd of the painful musculoskeletal episodes were judged correctly, either by the patient or physician, and arthritis was being incorrectly treated with factor.
Level of evidence: IV-Case series

Key point: Point-of-care ultrasound (POCUS) has potential value in acute hemarthrosis to objectively identify presence of blood in joints, measure size, pinpoint location, assess evolution and confirm complete disappearance.
Level of evidence: V- expert opinion

Key point: US imaging complements the physical examination when screening and monitoring joint health of people with haemophilia at the point of care; inter-rater reliability between physiotherapists was good.
Level of evidence: I
Practice Statement 2.5
The physiotherapist should communicate assessment findings and treatment plans to the relevant other team members.
Grade of recommendation: Theoretical

Rationale:
Communication with the other core team members is essential, to confirm clinical diagnosis and to ensure adequate follow-up until full resolution and return to full activity.
In some situations, hemostatic management may need to be adjusted to provide optimum hemostasis during rehabilitation and/or during return to full activity.

References:
Canadian Alliance of Physiotherapy Regulators Core Standards of Practice – Updated December 2019
Accessed January 22, 2021
Key Points: Shares information with clients, team members, and other stakeholders about the roles and responsibilities of physiotherapists in client-centred care.

Key point: need to ensure adequate hemostatic management especially for people with inhibitors.
Level of evidence: V- expert opinion
Standard 3: The physiotherapist will become involved as soon as possible after acute bleeds to assist the individual to regain pre-bleed status while preventing new injury or re-bleeding.

Practice Statement 3.1
Early management of acute MSK injuries may include:
- Monitoring and education
- Hemostatic management, as prescribed by the hematologist
- Protection of the injury with splints, slings
- Complete rest of the injured area, including:
  - no weight bearing for joint or muscle bleeds
  - no ambulation for psoas bleeds
- Ice for pain relief
- Compression
- Elevation

Grade of recommendation: Theoretical

Rationale/Discussion:
The primary goal of Physiotherapy treatment following any acute musculoskeletal injury is to regain pre-injury status as soon as possible. Physiotherapy management of acute MSK bleeding in PWBD has been informed largely by the literature pertaining to other populations, such as sports injuries. In PWBD, attention must also be paid to ensuring that bleeding has stopped, and to preventing new bleeding during the rehabilitation period. The physiotherapist must be familiar with contraindications and safety precautions applicable to treating PWBD.

References:

Key point: Review by a physiotherapist is recommended as soon as a patient presents with or reports a joint bleed, to advise and educate on the anticipated timeline for rehabilitation.

Level of evidence: V- expert opinion


Key point: “Rest, ice, compression and elevation are generally recommended as an adjunct to factor replacement therapy during acute hemarthrosis....... evidence supporting the use of ancillary therapies (in addition to factor replacement) is lacking.”

Level of evidence: V- expert opinion


Key point: Protection, rest, ice, compression and elevation (PRICE) should be implemented in the initial acute stage. An exception is young children, in whom compression is not advocated.

Level of evidence: V- expert opinion

Practice Statement 3.2
After bleeding has stopped, the physiotherapist should introduce exercise to restore:

- Joint motion
- Muscle length (may also require serial splinting)
- Muscle strength
- Proprioception (upper limb as well as lower limb)
- Balance
- Gait
- Function (including ADL and sports)

Grade of recommendation: Theoretical

Rationale/Discussion:
Prior to modern clotting factor products, treatment of acute bleeds involved lengthy periods of immobilization and/or inactivity which resulted in muscle weakness and loss of sensorimotor skills.
It is now recognized that an exercise program is essential for return to full function and to minimize the risk of recurrent bleeding. The most effective post-bleeding exercise parameters for PWBD are unknown: exercise prescription guidelines for the general population should be followed, however, to decrease the risk of injury, exercise should commence in limited range, at low velocity and submaximal load, and within patient tolerance, while closely monitoring for signs of new bleeding.

References:
Key points: Describes rationale for exercise for Flexibility, Strength, Proprioception, Balance and Function within the context of hemophilia.
Level of evidence: V- expert opinion

Key point: An appropriate balance should be established between rest, early mobilization, and weight-bearing so as to prevent unwanted complications associated with immobilization
Level of evidence: V- expert opinion

Key point: Includes “Dos and Don’ts” for therapists who have limited experience with PWBD.
Level of evidence: V- expert opinion

Key point: Current literature divides physiotherapy and rehabilitation for muscle bleeds in athletes into four phases: (i) control of haemorrhage, (ii) restoration of ROM and strength, (iii) functional rehabilitation, and (iv) and return to normal living.
Practice Statement 3.3
The physiotherapist may consider using electrophysical modalities to enhance hematoma resolution.
Grade of recommendation: Research

Rationale:
Some authors have reported using Pulsed Ultrasound and/or Pulsed Short-Wave Diathermy to assist tissue healing and/or hematoma resolution in PWBD. There are no known references that specify parameters for treating acute bleeds in PWBD.
It is the opinion of the working group that any heat-generating modalities should be applied with caution.
Deep heating is not advisable, and pulsed settings should always be used.

References:
Key point: in the lab, pulsed Ultrasound seems more effective than LLLT.
Level of evidence: Experimental- Animal study

Key point: summarizes relevant studies on US and PSWD in athletes.
Level of evidence: V- expert opinion

Key point: a review of how modalities could be used in PWBD: hypothetically speaking
Level of evidence: V- expert opinion

Key Point: Expert review of theories and applications of electrotherapy intervention, with separate section on using modalities for PWBD.
Level of evidence: V-review, expert opinion
Standard 4: The physiotherapist will perform a complete musculoskeletal assessment annually on each patient*, regardless of severity of the bleeding disorder.

(*each PWBD with a potential for MSK bleeding- see intro)

World Federation of Hemophilia recommends a multidisciplinary check-up including hematologic, musculoskeletal, and quality-of-life assessments by the core comprehensive care team members at least yearly and every six months for children. (This applies to all bleeding disorders.) Individuals with no bleeds, no recent treatment changes and no musculoskeletal or psychosocial concerns may be reviewed in person less frequently as deemed to be appropriate by the comprehensive care team.


Practice Statement 4.1
The physiotherapist should review the individual’s bleeding history.
Grade of recommendation: Theoretical

Rationale:
Regular review of the history of bleeding episodes will identify sites of repeated bleeding, so-called ‘target joints’, and perhaps incomplete rehabilitation of some injuries. It may also help the individual correlate specific activities with their bleeds, and indicate if changes to hematologic management regime are needed.

Reference:
Key point: European consensus exercise regarding components and frequency of assessments.
Level of evidence: V expert opinion

doi:10.2147/JBM.S50644

Key point: “any patient with severe hemophilia should be followed-up by a hemophilia specialist, with one or two visits conducted per year. For patients with moderate or minor deficit, annual follow-ups are also essential. The evaluation of joint status, along with its evolution and the impact on QoL, represent an integral part of the hemophilia consultation”.

Level of evidence: V-expert opinion

Practice Statement 4.2
The physiotherapist should perform a detailed musculoskeletal examination.

Grade of recommendation: Theoretical

Rationale:
Assessment of Body Structures and Function should include, at a minimum:
- range of joint motion of all peripheral joints* (See note)
- joint swelling
- crepitus with movement
- muscle wasting
- functional muscle strength
- gait

Assessment of joint end feel, balance and proprioception is also recommended.

The Hemophilia Joint Health Score (HJHS) is a reliable, validated disease-specific assessment tool that was developed to assess early arthropathy in children. The use of HJHS for teens and adults, as well as for individuals with chronic arthropathy, is being investigated.

An instruction manual and video are available at https://www.ipsg.ca/portal/terms-and-conditions

*Note: the HJHS does NOT include hips or shoulders.

References:
Compendium of assessment tools. World Federation of Hemophilia.
Accessed December 14, 2020
Key point: Describes the psychometric properties of disease specific tools, including HJHS
Level of evidence: V expert opinion

Key point: Discusses use of ICF, including relevant “impairment measures”.
Level of evidence: V-expert opinion

Key point: Interobserver reliability of the HJHS1.0 in teenagers and young adults with limited joint damage is excellent. Preliminary data on validity were similar or better than those in children.
Level of evidence: I

Practice Statement 4.3
The physiotherapist should assess pain.
Grade of recommendation: Weak

Rationale/discussion:
Pain is a prominent feature in persons with bleeding disorders, and can be due to diverse factors including acute bleeding, venous access, and chronic joint disease. PWBD may experience pain that is acute, chronic or acute ON chronic. A large number of adults with bleeding disorders experience pain on a daily basis, which impacts their daily activities, quality of life and mental health.
Pain is a complex biopsychosocial phenomenon. A ‘static’ pain measurement such as the VAS, which was designed to measure intensity of acute pain, may not be adequate for assessing the complex pain experienced by PWBD.

Different tools will be necessary for children and adults.
Hemophilia-specific tools are available.

References:

Key points: Analysis of psychometric properties of 8 pediatric pain scales. Weak recommendations at best could be made across all measures for chronic pain. Measures were not recommended for children younger than 6 years, and sometimes up to 8 years old.

Level of evidence: I - Systematic review


Key points: Increased disability and pain were associated with increased age, lower employment, higher reported bleed frequency, and lower HRQoL.

Level of evidence: III- case controlled study


Key point: a patient-reported outcome tool, designed in Canada, to be used with adults attending outpatient bleeding disorder clinics. Objective: to facilitate discussions about pain management options as a means of promoting self-efficacy.

Level of evidence: III


Key point: how PWBD (hemophilia) describe their pain, as well as what they do (or don’t do) to manage it.

Level of evidence: Prospective survey (other)


doi:10.1111/hae.12349
Key point: Although the title says “management’, includes a good description of chronic pain in people with hemophilia.
Level of evidence: V- review

Practice Statement 4.4
The physiotherapist should will assess the individual’s activities and participation throughout the lifespan, including:

• gross motor development
• school/work
• physical activity/sports
• leisure/social activities
• self-care

Grade of Recommendation: Theoretical

Rationale:
The presence of a bleeding disorder will have different impacts on Activities and Participation at different life stages, and a combination of objective and self-reported instruments is necessary to give a complete picture of the impact of the condition on the individual.

Bleeds, or fear of bleeding, particularly in young children may result in some parents/caregivers restricting activities that promote normal development. Gross motor delays have been identified in children with hemophilia (Hernandez). Physiotherapists should monitor overall development, and ensure that children with BD are able to participate with their peers in school and leisure activities. During the lifespan, musculoskeletal bleeding may alter movement patterns and cause pain, which begin to limit activities and participation in normal social roles. As PWBD live longer, issues often associated with “normal aging” may appear - such as decreased activity levels, decline in muscle strength, issues related to balance. These may be amplified or accelerated by the bleeding disorder.

Therefore, regular review of activities and participation, using age-appropriate patient-reported measures, should be done to identify issues that are important to the individual, and assist with goal-setting and treatment planning.

References:

Key point: See Appendix 4: outcomes to be measured.
Level- V-expert working group


Key point: Emphasizes need to assess activities/participation as well as impairments.
Level of evidence: V- expert opinion


Key Point: recommends using Hemophilia Activities List once a year
Level of evidence: V- expert opinion


Key point: objective and self-reported instruments should be combined.
Level of evidence: V-expert opinion


Key point: more than 50% of the boys tested had gross motor delays. The percentage of boys showing deficits increased and persisted after age 7.
Level of evidence: III case-controlled study


Key points: Supports use of HAL, FISH, and PedHAL, with some caveats
Level of evidence: I - Systematic review
Practice Statement 4.5
Physiotherapists should use standardized and validated assessment tools, including patient-reported outcomes, to evaluate changes in condition, identify areas of concern, and assist with goal-setting.
Grade of Recommendation: Theoretical

Rationale:
Disease specific tools have been developed, including:
- Hemophilia Joint Health Score
- Hemophilia Activities List (HAL and PedHAL)
- Functional Independence Scale for Hemophilia (FISH)
- Pain Treatment Planning Questionnaire

Generic tools, or tools developed for other populations (e.g., osteoarthritis), may also be useful, and allow comparison between PWBD and other groups. Some examples of these are:
- Alberta Infant Motor Scale
- Peabody Developmental Gross Motor Scale
- SF-36
- Community Balance and Mobility Scale
- Timed Up and Go
- 6-minute Walk test

Work is being done to develop a Core Set of assessment tools specifically for Hemophilia, based on the ICF.
Different tools will be needed for pediatric and adult populations.
Selection of tools should be guided by reason for use (clinical vs research tools).

References:
Key point: There are presently no pain scales suitable for children under 6.
Level of evidence: I- systematic review
Compendium of assessment tools. World Federation of Hemophilia. 
Key point: Describes the psychometric properties of disease specific tools, including HJHS
Level of evidence: V –expert opinion

Key point: multiple domains must be assessed; tools are available
Level of evidence: V-expert opinion

Key point: highlights advances and limitations in current research, and calls for development of a core set of outcome measures that will enable future meta-analyses.
Level of evidence: V-expert opinion

Key point: A new disease-specific tool for adults.
Level of evidence: III
Standard 5: The TC physiotherapist will arrange for the provision of Physiotherapy treatment (in-house or elsewhere) to address the MSK complications associated with bleeding disorders.

Practice Statement 5.1
Physiotherapists should use therapeutic exercise to address hemophilic arthropathy.
Grade of recommendation: Strong

Rationale:
Features of hemophilic arthropathy include joint swelling, pain and stiffness, contracture and deformity, muscle imbalance and atrophy, as well as alterations in the individual’s ability to function and participate normally.

Exercises that have been studied in PWBD (particularly Hemophilia A and Hemophilia B) include:
- exercise in water (hydrotherapy)
- active, isometric, and resisted exercise
- aerobic exercise
- functional (closed chain) exercise
- treadmill walking
- supervised exercise for groups or individuals
- Tai Chi
- Nordic walking
- home exercise programs

In randomized controlled studies, exercise for PWBD has been shown to improve range of motion, strength, walking tolerance, gait, balance, and quality of life (Calatayud, Runkel, Strike), in addition to decreasing pain and anxiety (Firoozabadi). Exercise combined with education sessions also reduced illness behaviour (Cuesta-Barriuso 2017).

The findings are consistent with the many non-controlled reports in the literature, as well as the experience of the working group: exercise is safe and effective for people with bleeding disorders.

References:
Level of evidence: I- RCT

Level of evidence: I- RCT

Level of evidence: V- expert opinion

Level of evidence: II

Level of evidence: I – systematic review

Level of evidence: I-RCT

Level of evidence: I-RCT

Level of evidence: II - small study


Level of evidence: I - Systematic review

**Practice Statement 5.2**

**Physiotherapists may recommend orthotics for PWBD**

Grade of recommendation: Theoretical

**Rationale:**
Orthoses and footwear modifications can be used in PWBD to control or prevent joint movement, stabilize a specific joint or relieve joint stress or loading.

**References:**

**Key point:** a very good review article: Includes pertinent studies, discusses shoe modifications/inserts, and orthotics for ankles, knees, AND elbows

Level of evidence: V - review article


Level of evidence: V - expert opinion

**Practice Statement 5.3**

**Physiotherapists may use low load prolonged passive stretching and serial splinting to address flexion contractures.**

Grade of recommendation: Theoretical

**Rationale:**
Flexion contractures can develop in PWBD if prevention and management of bleeding are not optimal.
Repeated bleeding leads to progressive fibrosis of the synovium with development of fibrous intraarticular bands. Pain, spasm and shortening in the surrounding muscles, and eventually fibrosis of the joint capsule, will follow if not addressed.

In the early stages (contractures < 30 degrees), slow stretching and serial splinting can be used to improve extension. To maintain the improvement, this should always be accompanied by strengthening exercises for the extensor muscles.

References:
*Key point:* Describes uses of manual traction, joint mobilization techniques, muscle stretching.
Level of evidence: IV - case series

*Key point:* Quick review of conservative measures, and a precaution: Vigorous attempts to restore motion through manipulation or excessive force with serial casting should be avoided as it can result in avulsion of articular surfaces at sites of attachment of fibrous bands.
Level of evidence: V - review, expert opinion

*Key point:* “Stretching techniques in phase 2 of treatment should be progressive, of very light intensity and within the limits of pain”
Level of evidence: V-review, expert opinion

**Practice Statement 5.4**
**Physiotherapists may use manual techniques.**
Grade of recommendation: Strong
**Rationale:**
Manual techniques that have been studied include joint traction, joint gliding techniques, fascial therapy and manual stretching.
These techniques were found to be safe (no new bleeding) and effective in decreasing pain.
One group suggests that bleeding frequency may also be reduced (Donoso-Úbeda).
However, this was self-reported bleeding: as people with arthropathy sometimes have difficulty distinguishing bleed pain from arthritic pain, and vice versa, this finding should be interpreted with caution.

**References:**

*Manual therapy: joint traction, passive muscles stretching.*
Level of evidence: I RCT


Level of evidence: I RCT


Level of evidence: IV case series

**Practice Statement 5.5**
Physiotherapists may use electrotherapy, following the usual clinical indications and precautions.
Grade of recommendation: Theoretical
**Rationale:**
Applications of energy, including Short-Wave Diathermy, Ultrasound, LASER, magnetic therapy, TENS, IFC and NMES have been proposed for pain management, neuromuscular reeducation (and to promote tissue healing and hematoma resolution in acute phases).

It is the opinion of the authors of these guidelines that such modalities may be used for PWBD according to the usual clinical indications and precautions in the general population, but should be considered as ADJUNCTS to exercise, education and other self-management techniques.

**References:**

Key point: LASER with exercise and hydrotherapy/land-based exercise appears to have some positive effect on knee pain in PWBD (Hemophilia).

Level of evidence: I - Systematic review


Key points: a review of theory and possible applications; some clinical experience with EMG with PWBD

Level of evidence: V - Theoretical, review

**Practice Statement 5.6**
Physiotherapists who have the necessary training may use acupuncture.
Grade of recommendation: Theoretical

**Rationale:**
There is some evidence that acupuncture can be effective in decreasing chronic arthritic pain in PWBD. Some patients received factor prior to acupuncture, some did not, but no adverse effects or bleeding was reported in these studies.

Having a bleeding disorder may pose a small increased risk of bruising and mild bleeding with acupuncture treatment, but should not be a contraindication.
The working group suggests that acupuncture can be used with caution in PWBD; hematological management should be discussed with the team.

References:
Key point: Small study in 2 centers: Local and distal points were used; no bleeding and 50% of patients reported pain relief.
Level of evidence: IV case series

Key points: Small study: used a single skull point; decreased pain in lower extremities; no adverse effects.
Level of evidence: Level II- cohort study

Practice Statement 5.7
Physiotherapists should provide education to PWBD.
Grade of recommendation: Theoretical

Rationale:
WFH endorses ongoing education of PWBD and their families to enable self-management. Education from the physiotherapist should include prevention, identification and management of MSK bleeding and its sequelae. Education should be specific to the age and situation of the individual, although group education can also be useful (opinion of the working group).

There is some preliminary evidence that education can be effective in improving the health of PWBD, as well as decreasing parental stress. The education sessions in the research included anatomy, biomechanics, pathophysiology and treatment of bleeds, proprioception, and activity selection.

References:
Cuesta-Barriuso R, Torres-Ortuño A, Nieto-Munuera J, López-Pina JA. Effectiveness of an Educational Physiotherapy and Therapeutic Exercise Program in Adult Patients With
Practice Statement 5.8
The physiotherapist may recommend environmental adaptation and mobility aids.
Grade of recommendation: Theoretical

Rationale:
PWBD with advancing arthropathy and/or with inhibitors may need assistance such as mobility aids to enhance functional activities and participation. Modifications to the home and/or workplace may also be needed to ensure safety and optimal function. This may require consultation with a community-based physiotherapist and/or occupational therapist.

Reference:
Level of evidence: V expert opinion

Practice Statement 5.9
The physiotherapist should arrange for referral of clients to other health professionals when indicated.
Grade of recommendation: Theoretical
Rationale:
Many PWBD have complex needs, and might require services of health professionals who are outside of the TC care team. These could include other Physiotherapists, Occupational Therapists, Pain Management Specialists, Orthotists, or other multidisciplinary programs (e.g., pain program)

[See also S 7: Consultation with other care providers]

References:
Key point: See Appendix 2: Core team and extended team members.
Level of Evidence: V- Expert opinion

Key point: Describes Core team and “others”
Level of evidence: V expert opinion
Standard 6: The TC physiotherapist must participate in discussions with the core team, the surgical team and the PWBD about optimal timing, pre-operative planning and post-operative rehabilitation for all MSK procedures.

This paper, by an international multidisciplinary panel of experts, outlines the roles and responsibilities of all team members, including the PWBD, during the pre-operative, intra-operative, and post-operative phases. It is relevant to all the practice statements related to Standard 6.

Level of evidence: V- expert opinion

A. Prior to MSK Surgery:

Practice Statement 6.1
The physiotherapist should ensure that the individual has had access to appropriate rehabilitation prior to recommending surgical intervention.
Grade of Recommendation: Theoretical

Rationale:
Surgical options may range from simple (intraarticular injection, radioactive synovectomy) to complex (arthroscopy, osteotomy, debridement, arthrodesis, or arthroplasty).

In some situations, it will be the TC physiotherapist who suggests to the team that it is time to consider surgery. In other situations, the request for surgical opinion might come from the PWBD or another team member.

In all cases, conservative therapy, including exercise, orthotic support, and environmental modification should be explored prior to considering surgery. (See Section 5: Physiotherapy Treatment of MSK Complications)

References: (in addition to Escobar)

**Key point:** International panel of surgical experts; summary of types of surgeries.

Level of evidence: V – expert opinion

**Practice Statement 6.2**

The bleeding disorders team, the surgical team and the PWBD should discuss goals and expectations for elective orthopedic surgery, including post-operative requirements.

Grade of Recommendation: Theoretical

**Rationale:**

The usual goals for surgical intervention are to **decrease bleeding, reduce pain and/or optimize function**.

Improved range of motion is **not always expected and can be difficult to achieve**, and the PWBD must understand this.

The PWBD must also understand that a lengthy period of out-patient rehabilitation may be required.

Pre-operative planning and coordination among all parties is imperative. The teams must discuss potential post-operative requirements such as:

- hemostatic management
- pain management
- anticipated length of hospital stay
- possible need for ICU (patients with inhibitors, complex surgeries with high risk of bleeding)
- anticipated time for restriction of usual activities, such as lifting, driving, work/school, or sports
- logistics regarding post-op rehab (e.g., location, frequency, transportation if the PWBD is unable to drive)

**References:**


Level of evidence: V-expert opinion
Practice Statement 6.3
After the decision is made to proceed with surgery, the physiotherapist should assess for indications for pre-operative physiotherapy.
Grade of Recommendation: Theoretical

Rationale:
PWBD often have multiple joint arthropathy which can limit their ability to maintain post-op weight-bearing restrictions. Consideration of upper extremity joint status should be made for weight-bearing status and use of mobility aids.

Depending on the procedure, pre-operative physiotherapy ("pre-hab") could include:

- preparation of other joints that may be subjected to increased stress post-operatively, by improving strength, ROM, or stability.
- preparing the operative joint, to optimize the recuperation and maximize the effects of the surgery
  - As long as the joint is not too painful, the physiotherapist should help the PWBD optimize range of motion and strength. For example, quads function may be poor due to prolonged pain, patellar adhesions and altered mechanics, making it difficult to achieve active knee extension.
  - It has been demonstrated that for Total Knee Arthroplasty, post-operative ROM correlates closely with pre-operative ROM (Moore 2016).
    - The working group feels that pre-operative preparation will optimize results of arthroplasty for other joints as well.
- ensuring that mobility aids, adaptive equipment, and environmental adaptations are in place to allow safe functioning after discharge

"Pre-hab" can also prepare the PWBD for the types of exercise he/she will need to do post-op.

References: (in addition to Escobar)

Key point: international panel of physiotherapy experts.
Level of evidence: V-expert opinion

Key point: Notwithstanding the title, they do address importance and suggest content of a “pre-hab” program.

Level of evidence: V expert opinion


Key point: Pre-op range and strength influences post-op results.

Level of evidence: II

Practice Statement 6.4
The physiotherapist from the TC should establish and maintain regular communication with other physiotherapists who may need to be involved post-operatively.

Grade of Recommendation: Theoretical

Rationale:
There will be situations where TC physiotherapist will not be supervising the post-op therapy: e.g.,
- the surgery and immediate post-op rehab may take place in a facility different from where the TC is located
- out-patient post-op rehab may take place a local clinic closer to the individual’s home.

The TC physiotherapist should be available to consult with the treating physiotherapist(s) who will plan and carry out appropriate therapy. Communication should emphasize the need to prevent new bleeding during mobilization and rehab.

Discussion could also include factors specific to the individual (e.g., status of other joints, environmental considerations for discharge, individual’s pain tolerance) that might impact post-op management.

All physiotherapists must be aware that:
• longer than normal hospital admission and rehabilitation time should be expected
• post-op bleeding may delay or interrupt mobilization, and make it difficult to optimize range of motion
• joint stiffness is more likely to occur in PWBD due to slower progression and the pre-existing state of the peri-articular structures
• PWBD are more likely to develop complications such as delayed wound healing and infection
• pain management may be complicated because the individual
  o may interpret post-operative pain as pain due to new bleeding
  o may already be using high levels of analgesics, including narcotics or cannabis, to manage chronic complex pain
  o may have developed other protective behaviours, such as avoiding activity, to manage persistent pain
  o may have strong and specific expectations about their complex pain and how it should be managed

(See also: Special Considerations for TKA)

**Reference:** (in addition to Escobar)

**Key point:** the TC physiotherapist ‘oversees’ the program.
Level of evidence: V


**Key point:** need to develop partnership with local physiotherapist who may be supervising the long-term rehabilitation.
Level of evidence: V-expert opinion

Key points: International surgeons and physiotherapists emphasize the need for long-term rehabilitation after several different procedures. Level of evidence: V-expert opinion

B. Following MSK surgery:

Practice Statement 6.5
Before beginning post-operative physiotherapy, the treating physiotherapist must be aware of:

- the type of bleeding disorder and its severity
- the hematological management plan
- the detailed procedure of the surgery including
  - the condition of the joint and surrounding muscles observed during the surgery
  - the range of motion obtained during surgery (if appropriate to the procedure)

Grade of Recommendation: Theoretical

Rationale:
All possible steps must be taken to prevent new bleeding while optimizing post-operative function. Therapy sessions should be coordinated with optimum hemostasis, analgesics, and other pain management strategies (e.g., relaxation).

The physiotherapist should discuss intra-operative findings directly with the surgeon. Sometimes surgery in PWBD may become more complex than was anticipated pre-operatively, e.g., need for wider field synovial resection, additional soft tissue or capsular releases, patellectomy.

References: (in addition to Escobar)

Key point: “Patients going to surgery with very limited flexion may require quadricepsplasty, which is often associated with an extensor lag for six months or longer”.
Level of evidence: V- expert opinion
C. Special Considerations regarding Total Knee Arthroplasty:

Practice Statement 6.6
The treating physiotherapist should be aware that PWBD may present challenges that are not seen in other populations undergoing TKA.
Grade of Recommendation: Theoretical

Rationale:
Since the knee joint is a common site for hemarthrosis in PWBD, many PWBD require TKA due to degenerative changes. There are several differences in the characteristics of PWBD, compared to other populations, that may require special consideration during the post-operative period, including (but not limited to):

- PWBD undergoing TKA are often younger
- PWBD may have significant arthropathy of several joints, and may undergo multiple procedures during one admission
- PWBD may be undergoing revision of a previous TKA
- PWBD have been conditioned to interpret pain as a sign of bleeding
- PWBD may already be using high levels of narcotic analgesics due to severe arthropathy
- Complication rates, including bleeding and infection, may be higher than in other populations
- PWBD may achieve good post-op range of motion initially, but have difficulty maintaining it
- PWBD may benefit from ‘pre-hab’, and extended post-operative rehabilitation including hydrotherapy even though these may not be used routinely with other populations undergoing TKA.

References: (in addition to Escobar)

Key Point: Description of components of In-patient and on-going out-patient rehabilitation programs.
Level of evidence: V-expert opinion
MASAC Recommendations Regarding Physical Therapy Guidelines in Patients with Bleeding Disorders: Total Knee Replacement 2015
Key points: From USA; describes Physiotherapy treatment, challenges and long-term expectations.
Level of evidence-V-expert opinion

Key point: “progressive postoperative loss of motion can occur in the most cooperative patients”.
Level of evidence: V- expert opinion

Practice Statement 6.7
The treating physiotherapist must actively and regularly watch for any signs of bleeding or infection.
Grade of Recommendation: Weak

Rationale:
Post-operative bleeding is not unusual, especially during the first week. Active bleeding will delay rehabilitation.
If bleeding occurs, the physiotherapist should stop the rehabilitation program, allow the limb to rest, and discuss with the hematologist and the surgeon.

References: (in addition to Escobar)
Key points: Fairly high rate of post-op bleeding and infections; difficulty achieving good knee extension
Level of evidence: IV
MASAC Recommendations Regarding Physical Therapy Guidelines in Patients with Bleeding Disorders: Total Knee Replacement 2015
Accessed January 10, 2021

Key points: From USA; describes signs of infection after TKA

Level of evidence: V - expert opinion


Key point: higher rate of bleeding and infection than in PWOB

Level of evidence: II

Practice Statement 6.8
The physiotherapist should consider use of hydrotherapy after hip or knee arthroplasty.
Grade of Recommendation: Weak

Rationale:
Several authors recommend the use of exercise in water during the post-operative period.

Advantages:
- Buoyancy can be used to assist or resist movement, and provide support and tactile stimulation.
- Warmth of the water assists with relaxation.
- Weight bearing can be progressed by varying depths of water as strength and confidence improve.

Prior to starting hydrotherapy, the wound must be completely healed, with no signs of infection.

References: (in addition to Escobar)
De Kleijn P, Fischer K, Vogely HC, Hendriks C and Lindeman E. In-hospital rehabilitation after multiple joint procedures of the lower extremities in haemophilia patients: clinical
doi:10.1111/j.1365-2516.2011.02527.x
Level of evidence: IV Case series and expert opinion

doi:10.1111/j.1365-2516.2008.01748.x
Level of evidence: V-expert opinion

doi.org/10.1111/hae.13748
Level of evidence: IV Case series

**Practice Statement 6.9**
The TC physiotherapist should ensure that on-going out-patient rehabilitation has been arranged.
Grade of recommendation: Theoretical

**Rationale:**
Although it may not be common practice in other populations, an extended program of supervised progressive rehabilitation may be useful for PWBD to help regain range of motion and strength, and re-educate posture and gait.

PWBD often have more difficulty regaining knee extension after TKA, which will affect their gait and overall function.

Referral for out-patient rehab should be organized by the in-patient physiotherapist OR the TC physiotherapist, depending on local practice. During the rehab program regular communication between the TC physiotherapist and the treating physiotherapist is recommended for optimal care.
(See also Practice Statements 6.4 and 7.1)

**References:**
Key point: all recommend extended course of post-op rehab
De Kleijn P, Fischer K, Vogely HCh, Hendriks C, Lindeman E. In-hospital rehabilitation
Level of evidence: IV Case series and expert opinion

Level of evidence: V- expert opinion

doi:10.2147/JBM.S50644
Level of evidence: V-expert opinion

MASAC Recommendations Regarding Physical Therapy Guidelines in Patients with Bleeding Disorders: Total Knee Replacement. 2015.

Level: V - expert opinion
Standard 7: The TC physiotherapist will be available for consultation with other care providers.

Practice Statement 7.1
The TC physiotherapist should be available for consultation with physiotherapists outside the TC who may be seeing PWBD.

Grade of recommendation: Theoretical

Rationale:
There are many circumstances where PWBD may receive physiotherapy assessment and treatment from physiotherapists who are not affiliated with a comprehensive treatment center (TC).

Reasons include, but are not limited to:

- The protected time allocation of the TC physiotherapist is not sufficient to meet all the needs of the PWBDs within the program
- The PWBD requires therapy outside the skill set of the TC physiotherapist
- The PWBD undergoes surgery in a facility that is different from the TC
- The PWBD lives far away from the TC, and is receiving treatment from a physiotherapist closer to home.

In these situations, the TC physiotherapist will ensure that the treating physiotherapist:

- is familiar with contraindications and precautions to be observed when treating ANY PWBD
- is aware of known chronic impairments and limitations specific to the PWBD being treated that may not be amenable to treatment
- is aware of the hemostasis management plan recommended by the TC team
- is aware of signs and symptoms of new bleeding
- is aware of whom to contact in the event of new bleeding

References:

Key point: “provide education and treatment recommendations to other community professionals who provide services to PWBD (e.g., surgery, dentistry).”

Level of evidence: V Expert working group
Key point: “The overall responsibility of physiotherapy management should remain with the specialist physiotherapist with effective communication between professionals”
Level of evidence: V -Expert working group

Canadian Alliance of Physiotherapy Regulators Core Standards of Practice – Updated December 2019
Accessed January 22, 2021
Key points: “Consults with/refers to the appropriate team member when aspects of clients’ goals are best addressed by another provider.”
“Takes appropriate actions (e.g., referral to another physiotherapist or health care provider, courses, mentorship) in situations where he/she does not have the required competence to deliver quality client-centred care.”

Practice Statement 7.2
The physiotherapist should be available for consultation with coaches, teachers.
Grade of recommendation: Theoretical

Rationale:
All people who are responsible for a child’s safety, such as babysitters, teachers, coaches etc. need to have sufficient information to minimize risk of bleeding and respond appropriately to suspected bleeds.
The physiotherapist can provide information about
- prevention of bleeding, including activity selection and modification (See S-8: Health promotion)
- identification of acute bleeding
- first aid management of bleeding
- return to activity following a bleed

There are several suitable educational resources at www.hemophilia.ca that can assist with this.
Practice Statement 7.3
The physiotherapist should be available to consult with other team members regarding identification and management of musculoskeletal complications of bleeding disorders
Grade of recommendation: Best Practice

Rationale:
Physiotherapists have specific training in identifying subtle changes in MSK health, and management of acute and chronic MSK conditions.

As the MSK expert on the team, the physiotherapist will assist other team members, other physiotherapists, and students to develop the skills they need to work with PWBD.

References: Opinion of the working group
Standard 8: Physiotherapists will encourage PWBD of all ages to lead active, healthy lifestyles.

Practice Statement 8.1
The Physiotherapist should encourage all PWBD to participate regularly in physical activities that are enjoyable to the individual, and discuss ways to minimize injury.
Grade of Recommendation: Theoretical

Rationale:
An active healthy lifestyle promotes cardiovascular fitness, healthy body weight, psychological well-being, and ability to maintain independence.

References:
Key point: Comprehensive review of relevant literature.
Level of evidence: II

Practice Statement 8.2
The physiotherapist should work with the individual and the TC team to develop a personalized activity plan which includes a plan for injury management.
Grade of recommendation: Theoretical

Rationale:
Personalized activity plans should take into account each person’s bleeding tendency, physical capabilities and interests. The prophylactic hemostasis management plan and a plan for management of injuries must be discussed with the whole team.

A tool has been developed by CPHC to assist this process: In the Driver’s Seat: A Workbook is designed to guide personalized decision-making about physical activities.
https://www.hemophilia.ca/files/In%20the%20Drivers%20Seat%20-%20EN_LR.pdf
accessed Jan 14, 2021

References:

Key point: “Studies have reported between 50% and 80% of PWH avoid exercise and activity due to pain and arthropathy, but there is reasonable evidence emerging that exercise via its effect on pain, joint ROM, strength and mobility can have a positive impact on maximizing mobility and function as well as quality of life in PWH”.

Level of evidence: V

Standard 9: Continuing Competence: The TC Physiotherapist will maintain clinical competence

Practice Statement 9.1
The TC Physiotherapist should participate in learning activities specific to bleeding disorders, and be familiar with current relevant literature.
Grade of recommendation: Best Practice

Rationale:
Bleeding disorders are rare, and recently there have been recent rapid changes in assessment and management options. The TC Physiotherapists should take an active role in maintaining their knowledge and competence to treat PWBD.

Examples of resources include:

Journals:
- The Journal of Haemophilia Practice

Websites:
- Canadian Hemophilia Society [www.hemophilia.ca](http://www.hemophilia.ca).
- National Hemophilia Foundation (USA) [https://www.hemophilia.org/Bleeding-Disorders](https://www.hemophilia.org/Bleeding-Disorders).

Congresses and Meetings:
- CHS New team member workshops: designed for Treatment Center team members with 3 years or less of experience
- CPHC education days and annual meetings
- World Federation of Hemophilia:
  - Congress
  - Musculoskeletal Congress
  - Global Forum
• Congress of the European Association for Haemophilia and Allied Disorders. http://eahad.org/
Appendix 1: Rare Bleeding Disorders: Incidence and clinical symptoms

Modified from:

- WFH elearning Introduction to Hemophilia [https://elearning.wfh.org/elearning-centres/introduction-to-hemophilia/#hemophilia_faq](https://elearning.wfh.org/elearning-centres/introduction-to-hemophilia/#hemophilia_faq)

<table>
<thead>
<tr>
<th>Deficiency</th>
<th>Incidence</th>
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<tbody>
<tr>
<td>Factor VIII (Hemophilia A)</td>
<td>1/10,000</td>
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<tr>
<td>Factor IX (Hemophilia B)</td>
<td>1/50,000</td>
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<tr>
<td>Von Willebrand Type 3</td>
<td>1 in 1 million</td>
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<tr>
<td>Fibrinogen</td>
<td>1 in 1 million</td>
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<tr>
<td>Prothrombin</td>
<td>1 in 2 million</td>
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<tr>
<td>Factor V</td>
<td>1 in 1 million</td>
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<tr>
<td>Combined FV and FVIII</td>
<td>1 in 1 million</td>
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<tr>
<td>Factor VII deficiency</td>
<td>1 in 500,000</td>
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<tr>
<td>Factor X deficiency</td>
<td>1 in 1 million</td>
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<td>Factor XI</td>
<td>1 in 1 million</td>
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<tr>
<td>Factor XIII</td>
<td>1 in 2 million</td>
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<tr>
<td>Vitamin K-dependent factors deficiency</td>
<td>&lt; 50 families</td>
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</table>
Table 2. Clinical symptoms in severe RBDs

<table>
<thead>
<tr>
<th>RBD</th>
<th>Affibrinogenemia, hypofibrinogenemia</th>
<th>Prothrombin deficiency</th>
<th>FV deficiency</th>
<th>Combined FV and FVIII deficiency</th>
<th>FVII deficiency</th>
<th>FX deficiency</th>
<th>FXI deficiency</th>
<th>FXIII deficiency</th>
<th>Vitamin K-dependent factors deficiency</th>
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<tbody>
<tr>
<td>Main bleeding symptoms for severe deficiencies</td>
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<td>Common:</td>
<td>Menorrhagia</td>
<td>Epistaxis</td>
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<td></td>
<td>Prolonged post-injury</td>
<td>Skin</td>
<td>Menorrhagia</td>
<td>Umbilical cord</td>
<td>Intracranial</td>
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<td></td>
<td>Mucosal tract</td>
<td>Mucosal tract</td>
<td>Gingival postoperative</td>
<td>Gum bleeding</td>
<td>Umbilical cord</td>
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<td></td>
<td>Hemorrhage</td>
<td>Postoperative</td>
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<td>Menorrhagia</td>
<td>Gum bleeding</td>
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<td>Skin</td>
<td>Common: Menorrhagia</td>
<td>Common:</td>
<td>Common: Gum bleeding</td>
<td>Common: Intracranial</td>
<td>Common: Intracranial</td>
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<td>GI</td>
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<td>Gastro-urinary tract</td>
<td>Less common:</td>
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<td>CNS</td>
<td>Common: Menorrhagia</td>
<td>Gum bleeding</td>
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<td>Umbilical cord</td>
<td>Menorrhagia</td>
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<td>CNS</td>
<td>Common: Menorrhagia</td>
<td>Gum bleeding</td>
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<td>Muscle/extent</td>
<td>Rare:</td>
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Risk of thrombosis

Affibrinogenemia: reported
Dy fibrinogenemia: reported
Inherited dysprothrombinemia due to G20210A mutation and linked to slightly increased levels of circulating prothrombin, there is a significantly higher risk to develop thrombocytopenia

Thrombotic episodes, particularly deep vein thrombosis post-treatment reported (3%-4% of patients)
Spontaneous thrombosis may occur

Cause of myocardial infarction and venous thrombosis reported (disseminated or after VKLA infusion)

Although protein S/C levels decreased, no reports of venous or arterial thrombosis

CNS = central nervous system; GI = gastrointestinal.
Appendix 2: Levels of Evidence

Levels of evidence were assigned using this system:

<table>
<thead>
<tr>
<th>Level</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Evidence obtained from high-quality diagnostic studies, prognostic or prospective studies, cohort studies or randomized controlled trials, meta-analyses, or systematic reviews (critical appraisal score &gt;50% of criteria)</td>
</tr>
<tr>
<td>II</td>
<td>Evidence obtained from lesser-quality diagnostic studies, prognostic or prospective studies, cohort studies or randomized controlled trials, meta-analyses, or systematic reviews (eg, weaker diagnostic criteria and reference standards, improper randomization, no blinding, &lt;80% follow-up) (critical appraisal score &lt;50% of criteria)</td>
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<tr>
<td>III</td>
<td>Case controlled studies or retrospective studies</td>
</tr>
<tr>
<td>IV</td>
<td>Case studies and case series</td>
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<tr>
<td>V</td>
<td>Expert opinion</td>
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</table>


For these guidelines, Qualitative studies and animal studies were classified as “Other”.

Additional tools, such as AGREE and AMSTAR 2 were also used when necessary to further refine the classification.
Appendix 3: Grading the Action Statements

<table>
<thead>
<tr>
<th>Grade</th>
<th>Recommendation</th>
<th>Quality of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Strong</td>
<td>A preponderance of level I studies, but at least one level I study directly on the topic support the recommendation.</td>
</tr>
<tr>
<td>B</td>
<td>Moderate</td>
<td>A preponderance of level II studies, but at least one level II study directly on topic support the recommendation.</td>
</tr>
<tr>
<td>C</td>
<td>Weak</td>
<td>A single level II study at &lt;25% critical appraisal score or a preponderance of level III and IV studies, including consensus statements by content experts support the recommendation.</td>
</tr>
<tr>
<td>D</td>
<td>Theoretical/foundational</td>
<td>A preponderance of evidence from animal or cadaver studies, from conceptual/theoretical models/principles, from basic science/bench research, or from published expert opinion in peer-reviewed journals supports the recommendation.</td>
</tr>
<tr>
<td>P</td>
<td>Best practice</td>
<td>Recommended practice based on current clinical practice norms, exceptional situations where validating studies have not or cannot be performed, and there is a clear benefit, harm or cost, and/or the clinical experience of the guideline development group.</td>
</tr>
<tr>
<td>R</td>
<td>Research</td>
<td>There is an absence of research on the topic, or higher-quality studies conducted on the topic disagree with respect to their conclusions. The recommendation is based on these conflicting or absent studies.</td>
</tr>
</tbody>
</table>


Appendix 4: Summary of Practice Statements

<table>
<thead>
<tr>
<th>Practice Statement</th>
<th>Grade of Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Comprehensive Interdisciplinary Care</strong></td>
<td></td>
</tr>
<tr>
<td>1.1 The Treatment Center (TC) physiotherapist should meet the PWBD /family soon after diagnosis, and will provide assessment, treatment and education throughout the person’s lifespan.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 1.2 The physiotherapist should consider using telerehabilitation, including online video conferencing platforms, for assessment, treatment and/or education of PWBD.</td>
<td>Theoretical</td>
</tr>
<tr>
<td><strong>Assessment of acute musculoskeletal bleeds</strong></td>
<td></td>
</tr>
<tr>
<td>PS 2.1 Physiotherapy assessment of acute bleeds should include a detailed history of the present condition.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 2.2 Physiotherapy assessment of acute bleeds should include past medical history.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 2.3 Physiotherapy assessment of acute bleeds should include a physical examination.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 2.4 Physiotherapists who have received appropriate training, may use point of care ultrasound (POCUS), if available, to confirm diagnosis and monitor resolution.</td>
<td>Moderate</td>
</tr>
<tr>
<td>PS 2.5 The physiotherapist should communicate assessment findings and treatment plans to the relevant other team members.</td>
<td>Theoretical</td>
</tr>
<tr>
<td><strong>Management of acute bleeds</strong></td>
<td></td>
</tr>
<tr>
<td>PS 3.1 Early management of acute musculoskeletal injuries may include monitoring and education, hemostatic management as prescribed by the hematologist, protection of the injury with splints or slings, complete rest of the injured area, ice for pain relief, compression, elevation</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 3.2 After bleeding has stopped, the physiotherapist should introduce exercise to restore joint motion, muscle length, muscle strength, proprioception, balance, gait and function (including activities of daily living and sports).</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 3.3 The physiotherapist may consider using electrophysical modalities to enhance hematoma resolution.</td>
<td>Research</td>
</tr>
<tr>
<td><strong>The annual assessment</strong></td>
<td></td>
</tr>
<tr>
<td>PS 4.1 The physiotherapist should review the individual’s bleeding history.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 4.2 The physiotherapist should perform a detailed musculoskeletal examination.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 4.3 The physiotherapist should assess pain.</td>
<td>Weak</td>
</tr>
<tr>
<td>PS 4.4 The physiotherapist should assess the individual’s activities and participation, throughout the lifespan.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 4.5 Physiotherapists should use standardized and validated assessment tools, including patient-reported outcomes.</td>
<td>Theoretical</td>
</tr>
<tr>
<td><strong>Physiotherapy treatment for musculoskeletal complications</strong></td>
<td></td>
</tr>
<tr>
<td>PS 5.1 Physiotherapists should use therapeutic exercise to address hemophilia arthropathy.</td>
<td>Strong</td>
</tr>
<tr>
<td>PS 5.2 Physiotherapists may recommend orthotics for PWBD</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.3 Physiotherapists may use low load prolonged passive stretching, and serial splinting to address flexion contractures.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.4 Physiotherapists may use manual techniques, including traction, joint gliding, fascial therapy and manual stretching.</td>
<td>Strong</td>
</tr>
<tr>
<td>PS 5.5 Physiotherapists may use electrotherapy.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.6 Physiotherapists who have the necessary training may use acupuncture.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.7 Physiotherapists should provide education to PWBD</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.8 The physiotherapist may recommend environmental adaptation and mobility aids.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 5.9 The physiotherapist should arrange for referral of clients to other health professionals when indicated.</td>
<td>Theoretical</td>
</tr>
</tbody>
</table>

**Physiotherapy management before and after musculoskeletal surgery**

| PS 6.1 The physiotherapist should ensure that the individual has had access to appropriate rehabilitation prior to recommending surgical intervention. | Theoretical |
| PS 6.2 The bleeding disorders team, the surgical team and the PWBD should discuss goals and expectations for elective orthopedic surgery, including post-operative requirements. | Theoretical |
| PS 6.3 After the decision is made to proceed with surgery, the physiotherapist should assess for indications for pre-operative physiotherapy. | Theoretical |
| PS 6.4 The Physiotherapist from the TC should establish and maintain regular communication with other physiotherapists who may need to be involved post-operatively. | Theoretical |
| PS 6.5 Before beginning post-operative physiotherapy, the treating physiotherapist must be aware of the type and severity of the bleeding disorder, the hematological management plan, the detailed procedure of the surgery including the condition of the tissues observed during the surgery and the range of motion obtained during surgery (if appropriate to the procedure). | Theoretical |
| PS 6.6 The treating physiotherapist should be aware that PWBD may present challenges that are not seen in other populations undergoing similar procedures. For example, PWBD may be younger, may have arthropathy at multiple joints, have been conditioned to interpret pain as a sign of bleeding, may be accustomed to using high levels of narcotic analgesia, and may have higher post-op complication rates. | Theoretical |
| PS 6.7 The treating therapist must actively and regularly watch for any signs of bleeding or infection | Weak |
| PS 6.8 The physiotherapist should consider use of hydrotherapy after hip or knee arthroplasty. | Weak |
| PS 6.9 The TC physiotherapist should ensure that on-going out-patient rehabilitation has been arranged. | Theoretical |

**Consultation with other care providers**

<p>| PS 7.1 The TC physiotherapist should be available for consultation with physiotherapists outside the TC who may be seeing PWBD. | Theoretical |
| PS 7.2 The physiotherapist should be available for consultation with coaches, school teachers (etc.) regarding activity modification, bleed prevention and injury management. | Theoretical |</p>
<table>
<thead>
<tr>
<th>PS 7.3 The physiotherapist should be available to consult with other team members regarding identification and management of musculoskeletal complications of bleeding disorders.</th>
<th>Best Practice</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Encouraging a healthy lifestyle</strong></td>
<td></td>
</tr>
<tr>
<td>PS 8.1 The physiotherapist should encourage all PWBD to participate regularly in physical activities that are enjoyable to the individual, and discuss ways to minimize injury.</td>
<td>Theoretical</td>
</tr>
<tr>
<td>PS 8.2 The physiotherapist should work with the individual and the TC team to develop a personalized activity plan which includes a plan for injury management.</td>
<td>Theoretical</td>
</tr>
<tr>
<td><strong>Continuing Competence</strong></td>
<td></td>
</tr>
<tr>
<td>PS 9.1 The TC physiotherapist should participate in learning activities specific to bleeding disorders, and be familiar with current relevant literature.</td>
<td>Best Practice</td>
</tr>
</tbody>
</table>