CLINICAL GUIDELINES for Physiotherapists working with persons with Bleeding Disorders:

Appendix/companion document for
CPHC Standards of Physiotherapy Care for Persons with Bleeding Disorders 2018

These guidelines were developed by the Standards working group to augment the CPHC Standards and provide clinical guidance.

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Guidelines for Standard 1: The PT is a core team member

Each Canadian comprehensive bleeding disorders care team includes a physiotherapist with protected time dedicated specifically to the Bleeding Disorders program (1, 2).

- A physiotherapist is involved in the care of the PWBD from the time of diagnosis and will participate in the Assessment, Treatment, and Education throughout the person’s lifespan (1,2,3)

1.1 Assessment includes:
- Baseline and longitudinal assessments throughout the lifespan (1,2)
- Determining the site of an acute bleed and assisting with differential diagnosis (e.g. joint bleed vs muscle bleed; synovitis vs hemarthrosis; superficial soft tissue bleed vs muscle bleed) (2)
- Pre-operative and post-operative assessments (1,2,3)

1.2 Treatment includes management and rehabilitation of all musculoskeletal complications and sequelae of hemophilia (1, 2, 3) including
- Acute bleeds
- Acute and Chronic synovitis
- Chronic arthropathy
- Pain
- Limitations of Function and Participation
- Post-operative rehabilitation

Also see Guidelines for treatment for Standard 3 and Standard 5.

1.3 Education includes information provided to
- Patients and families regarding prevention, identification and management of bleeds and the musculoskeletal complications of bleeding disorders (1,2,3)
- Patients and families regarding healthy active lifestyle throughout the lifespan (1,2,3)
- School personnel with respect to the appropriate activities for the PWBD, and activity modifications that may be necessary to allow safe participation
- All people who are responsible for the patient’s safety, such as babysitters, teachers, coaches, etc. re: appropriate immediate treatment of a suspected bleed (1,2)
- Other core team members regarding identification and management of musculoskeletal complications of bleeding disorders (2)
- Other physiotherapists re: management of PWBD (2)
References:
Guidelines for Standard 2: PT assessment of acute injuries

The Physiotherapist participates in the assessment of acute musculoskeletal bleeds.

2.1 Musculoskeletal bleeding is a key characteristic of many bleeding disorders and can result in permanent damage to the joints and muscles (1,2). Accurate diagnosis is the key to ensuring correct treatment.

- The earliest symptom of a musculoskeletal bleed is pain BUT not all pain is due to bleeding (3,4).
- Symptoms of some musculoskeletal bleeds can mimic other conditions (e.g. psoas bleed presenting like appendicitis) (5)

2.2 Assessment of acute bleeds should include:

2.2.1 History of present condition
- details of bleed onset and progression of symptoms
- mechanism of injury
- factor replacement given 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

2.2.2 Past medical history
- recent joint or muscle bleeds
- previous target joints; location and number
- usual activities
- usual factor replacement regime and usual response to treatment

2.2.3 Specific objective criteria – least invasive first.
- Observation/inspection: posture, bruising** (see notes below), swelling, redness
- Active Movement:
  - compare to contralateral limb or baseline measurements on file
  - is there pain with movement? Location?
- Palpation: warmth, swelling, tenderness, spasm, paresthesia
- Testing:
  - Joints: end feel (within limits of pain)
  - Muscles:
    - muscle length especially two-joint muscles
    - pain and/or weakness with active contraction
  - Sensation especially if nerve compression is suspected
2.3 Clinical presentation of an acute bleed in a PWBD may include the following:

<table>
<thead>
<tr>
<th>Joint Bleeds (2,3,6,7)</th>
<th>Muscle Bleeds (8,9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Pain – often preceded by a ‘tingling’ sensation or ‘aura’</td>
<td>• Pain if muscle contracts or is stretched</td>
</tr>
<tr>
<td>• Warmth on palpation</td>
<td>• Muscle tightness and limited range of motion</td>
</tr>
<tr>
<td>• Rapid joint swelling</td>
<td>• Superficial Muscles:</td>
</tr>
<tr>
<td>• Joint held in a position of comfort (open pack position)</td>
<td>o Warmth</td>
</tr>
<tr>
<td>• Tender when touched or moved</td>
<td>o Palpable hematoma</td>
</tr>
<tr>
<td>• Reduced ROM</td>
<td>o Swelling</td>
</tr>
<tr>
<td>• Altered gait or Inability to weight-bear</td>
<td>• Deep muscles with potential for</td>
</tr>
<tr>
<td></td>
<td>o Compartment syndrome</td>
</tr>
<tr>
<td></td>
<td>o Paresthesia</td>
</tr>
<tr>
<td></td>
<td>o Distal pallor or pulselessness</td>
</tr>
<tr>
<td></td>
<td>e.g. forearm, calf, iliopsoas***</td>
</tr>
</tbody>
</table>

Notes:

*** Presentation of an iliopsoas bleed may include (5,8,9):
• 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 (lordosis), knee flexed, and unable to place foot flat on floor
• Unable to lie flat

**Bruising:
• May be an indication of a superficial soft tissue bleed.
• Bruising is NOT a feature of joint bleeds(10);
• 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. See table above.
2.4 Clinical assessment is usually sufficient to make the diagnosis. Ultrasound or other imaging techniques, if available, can be helpful to confirm the size and location of the hematoma and monitor resolution (4). There is increasing use of point of care ultrasound in Canadian HTCs to corroborate clinical assessment and monitor recovery (11).

2.5 Assessment findings and PT treatment plan should be communicated to the relevant other team members.

References:

Guidelines for Standard 3: Physiotherapy Treatment

3A: After Acute Musculoskeletal bleeds:

3.1 The primary goal of Physiotherapy treatment following a musculoskeletal bleed is to regain pre-bleed status as soon as possible while preventing new injury or re-bleeding (1,2,3).

This includes measures to:
- Reduce pain
- Reduce swelling
- Restore joint range of motion
- Restore muscle length
- Restore muscle strength
- Return to regular activity, including sports

3.2 Early management may include (3,4):
- Factor replacement, 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

3.3 Exercise must be progressed cautiously, within limits of pain, and with attention to signs or symptoms that would indicate new bleeding.

After the bleeding has stopped, exercise can be introduced (5-8) 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)

3.4 Manual techniques (9, 10), electrophysical modalities (4, 11) and acupuncture (12) can be used with caution once bleeding has stopped, and according to the usual clinical indications and contraindications.
References:
3B. Physiotherapy Treatment Before and After MSK Surgical procedures:

In many cases, it will be the HTC physiotherapist who suggests to the rest of the team that all conservative measures have been explored and that surgery should be considered to optimize Musculoskeletal function (1). Surgical options may range from simple (intraarticular steroid injection) to complex (osteotomy, debridement, arthrodesis, or arthroplasty). All members of the core HTC team need to participate in pre-operative planning (2).

The surgical team must realize that persons with hemophilia undergoing orthopedic surgery may be different than their usual population (3):
- They are often younger
- They may have widespread arthropathy and chronic pain
- They have been conditioned to interpret pain as a sign of bleeding
- Pain management may require a multimodal approach (MJP)
- Progression of rehabilitation is often slower due to fear of bleeding and the poor condition of the surrounding tissues.

3.5 Pre-op preparation (1)
The HTC physiotherapist should establish contact with the therapist from the surgical team to coordinate the pre-operative treatment and education which should include:
- Discussion with the patient and the surgeon regarding goals and expectations for the surgery.
- A program of physiotherapy to prepare the adjacent joints for the possible increased stress post-operatively
- A program of physiotherapy to prepare the operative joint, to optimize the recuperation and maximize the effects of the surgery- unless the joint is too painful.
- Determine need for ambulatory aids, adaptive equipment, environmental adaptations, and ensure these are in place.
- Education of the patient regarding anticipated length of hospital stay, post-operative therapy requirements, and time to resume physical activities such as driving or sports.
- Discuss the out-patient post-operative rehabilitation:
  - Location
    - ensure patient has transportation
  - Frequency
  - Duration
- With the HTC team, determine how factor coverage will be arranged.

3.6 Post-surgery (1,3,4)
After the surgery, the physiotherapist of the surgical team will become involved as soon as possible, as discussed with the medical team.
Prior to seeing the patient, the Physiotherapist must ascertain:
- the type of hemophilia and severity
- the factor replacement regimen
- the detailed procedure of the surgery
- the condition of the joint and surrounding muscles observed during the surgery
- the range of motion obtained during surgery (if appropriate to the procedure)

3.7 The post-surgical physiotherapist must be aware that:
- Rehabilitation times after surgery will vary by individual patient and extent of surgery.
- Longer than normal hospital admission and rehabilitation time should be expected. (1,3)
- PWBD are more likely to develop complications:
  - Pain: Pain management may be complicated: the patient may not be able to distinguish post-operative pain from pain due to new bleeding (3, 4)
  - Bleeding: The risk of bleeding may postpone the mobilizations and predispose to ankylosis
  - Delayed wound healing and infections (5)
  - Difficulty regaining pre-operative range of motion, especially after TKA
    - CPM may be useful after arthroplasty in PWH, even though the surgical team may not use it routinely (1).

- The physiotherapist should be extra cautious to prevent and watch for any signs of bleeding.
  - Spontaneous bleeds are common at the end of the first week post op.
  - If bleeding occurs, stop the rehabilitation program and rest the limb.
  - Watch for signs and symptoms of infection.
  - Understand that joint stiffness is more likely to occur due to slower progression and the state of the extra-articular structures (muscular tightness, joint damages, involvement of the others joints).

Recommended reading re TKA and Hemophilia:

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


References:


Guidelines for Standard 4: The Annual Assessment

A complete musculoskeletal assessment will be done annually on each patient regardless of the severity of the bleeding disorder (1, 2)

4.1 Review bleeding history:

Table 5. Information on bleeding episodes.

<table>
<thead>
<tr>
<th>Haemarthrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Number of episodes</td>
</tr>
<tr>
<td>- Time, location and reason for each haemarthrosis</td>
</tr>
<tr>
<td>- Type of treatment (including dose, number and interval)</td>
</tr>
<tr>
<td>- Response to treatment</td>
</tr>
<tr>
<td>Other major bleeds</td>
</tr>
<tr>
<td>- Number, type</td>
</tr>
<tr>
<td>- Time, location and reason for each bleeding</td>
</tr>
<tr>
<td>- Treatment and response to treatment</td>
</tr>
<tr>
<td>Minor bleeds</td>
</tr>
<tr>
<td>- Number, type</td>
</tr>
<tr>
<td>- Time, location and reason</td>
</tr>
<tr>
<td>- Treatment and response to treatment</td>
</tr>
</tbody>
</table>

Reference/source for Table 5:

4.2 Impairment:
The Hemophilia Joint Health Score (HJHS) includes impairments measures such as range of motion, swelling, strength, and crepitus and is sensitive to early changes in joint status (1,2,3). It was originally developed for use in children with minimal arthropathy, however “there is no theoretical or practical reason why it cannot be used in older patients too” (4) and validity studies are being done in adult populations (5).

However, the HJHS does NOT include hips or shoulders which should be assessed if joint damage is suspected. Published age-related reference values are available for range of motion (6).

References re Impairment:


4.3 Pain:
Pain is a prominent feature in persons with bleeding disorders, and can be due to diverse factors including acute bleeds, venous access, and chronic joint disease (1,2). Pain status should be monitored in PWBD so that appropriate management (non-pharmacological or pharmacological) can be instituted (3,4,5,6)

Examples of pain measurement scales commonly used in care of PWBD include:
- Visual Analog Scale/Numerical Pain Rating Scale (7,8)
- FACES (9,10)
- S-LANSS - Leeds Assessment of Neuropathic Symptoms and Signs (11)
- Brief Pain Inventory Scale (12,13)
- Functional Disability Inventory (14)
- McGill Pain Questionnaire (SF-MPQ) (15)

References re Pain:
4.4 Function and Participation:
Repeated musculoskeletal bleeding can cause altered movement patterns in the early years, and chronic hemophilic arthropathy with advancing age (1). Changes are often insidious and subtle. The Annual Physiotherapy assessment allows identification and early intervention as new problems arise (2,3).

The Physiotherapist will use standardized tools to assess participation in
- school/work
- physical activities and /or sports
- leisure activities
- social activities
- self-care

Hemophilia Specific tools include:
- Hemophilia Activities List and Pediatric Hemophilia Activities List (4,5,6)
- Functional Independence Scale for Hemophilia (4)

Other tools, that are not specific to hemophilia, may also be useful to assess overall physical functioning.
Some examples of these are:
Children:
- Alberta Infant Motor Scale (7)
- Movement Assessment of Infants (7)
- Peabody Developmental Gross Motor Scale (7)
Adults:
- Community Balance and Mobility Scale (8)
- Timed Up and Go (9)
- 6 minute Walk test (9)

References re Function and Participation:

4.5 Patient Reported Outcomes
These tools are useful because they allow individuals to identify issues that are important and meaningful to them.

Examples include:
- Patient Specific Functional Scale, https://www.physio-pedia.com/Patient_Specific_Functional_Scale
- Canadian Occupational Performance Measure: http://www.thecopm.ca/
- Goal Attainment Scale.
Guidelines for Standard 5: PT Treatment of Musculoskeletal Complications of Bleeding Disorders

5.1 Features of arthropathy include joint pain and stiffness, contracture and deformity, muscle imbalance and atrophy, and alterations in the individual’s ability to function and participate normally (1).

5.2 Most physiotherapy interventions can be applied according to the usual clinical indications and precautions and as long as there is no active bleeding, including:

- Exercise program to maintain or improve range of motion, strength, flexibility, balance or exercise tolerance (1,2,3,4,5)
- Joint protection techniques such as proper body mechanics, ergonomic adaptations, orthotics or bracing (1,6)
- Hydrotherapy (7)
- Prescription and instruction regarding mobility aids
- Consultation with Occupational Therapy regarding adaptations to the individual’s environment.
- Consultation with orthopedics and preparation for surgery (8,9,10)
- Use of splinting/low load prolonged passive stretching techniques/devices (serial splinting/casting) (11,12)
- Manual therapy (12,13)
- Electrophysical agents (14)
- Acupuncture (15)

References:


Guidelines for Standard 6: Consultation with other care providers

There are many circumstances where PWBD may receive Physiotherapy assessment and treatment from physiotherapists who are not affiliated with a comprehensive Hemophilia treatment center (HTC). These circumstances may include, but are not limited to:

- the PWBD requires therapy outside the skill set of the HTC therapist (e.g. pediatric therapist seeing adult patients and vice versa, orthopedic therapist seeing PWBD with neurological impairment)
- Surgery is done in a facility different from the HTC, and the therapists at the surgical center have specialized skills and knowledge re the post-op rehab requirements.
- The PWBD lives too far away from the HTC and will see a therapist in his/her home community
- The FTE allocation of the HTC therapist is not sufficient to meet all the needs of the PWBDs within the program: (e.g. time is allocated for annual review only so patients requiring ongoing treatment must access physiotherapy services elsewhere)

In these circumstances the HTC therapist will be available for consultation (1) to assist the treating therapist to:

- be familiar with contraindications and precautions to be observed when treating any PWBD
- be familiar with known chronic impairments and limitations specific to the PWBD being treated that may not be amenable to treatment
- verify the factor replacement regimen/schedule recommended by the HTC team for the patient
- recognize signs and symptoms of new bleeding and modify treatment accordingly
- facilitate communication between the treating therapist and the rest of the team including a plan for prophylactic factor replacement if needed, and a plan for treatment of new bleeding
- be aware of educational resources and other leaning opportunities pertinent to the management of bleeding disorders

Reference:
Guidelines for Standard 7: Education and health promotion

An active healthy lifestyle promotes cardiovascular fitness, healthy body weight, psychological well-being, and ability to maintain independence (7,8,9).

Physiotherapists provide education regarding selection of sports and activities that are enjoyable to the individual but minimize the risk of injury (1-5).

There are a number of resources that have been developed as guidelines for selection of physical activities (see list below). Many of these attempt to classify sports and activities according to risk and the recommendations made may not be suitable in all instances. “In the Driver’s Seat” is designed “to guide adults with bleeding disorders through an organized and thoughtful approach to decision-making around physical activities.”

It is the opinion of CPHC that personal activity plans should be INDIVIDUALIZED and should take into account each individual’s bleeding tendency, physical capabilities and interests. This requires discussion between the individual/family and the team.

Resources: Hemophilia Organizations:


- London Ontario treatment center http://www.lhsc.on.ca/Patients_Families_Visitors/Bleeding_Disorders/Physiotherapy/ActivityChoices.htm (London Ontario)
Resources: Pharmaceutical Companies:
- Bayer Pharmaceuticals; http://www.livingwithhemophilia.ca/managing/fitness-exercise.php

References:
Guidelines for Standard 8: Continuing Competence

There are many Hemophilia Specific learning opportunities available to physiotherapists.

Journal:  
Can be searched online via the WFH website:  
Some articles can be downloaded at no cost.  
CPHC members have full access to the journal: contact the Secretary for password information.

Educational Materials:  
- World Federation of Hemophilia: [www.wfh.org](http://www.wfh.org). Numerous resources are available for download, including:  
  - Guidelines for the management of Hemophilia  
  - Compendium of Assessment tools  
  - Treatment of Hemophilia monograph series  
  - Web-casts of congress sessions  
- Canadian Hemophilia Society [www.hemophilia.ca](http://www.hemophilia.ca). Resources include:  
  - All About Hemophilia  
  - Comprehensive Care Standards (2007)  
  - Contact information for all HTCs across Canada  
- National Hemophilia Foundation (USA) [https://www.hemophilia.org/Bleeding-Disorders](https://www.hemophilia.org/Bleeding-Disorders) Resources include:  
  - Medical and Scientific Advisory Council recommendations  
- Hemophilia Foundation of Australia  

Hemophilia-specific Conferences:  
In Canada:  
- CHS New team member workshops: designed for Hemophilia Treatment Center team members with 3 years or less of experience.  
- CHS Rendez-vous: held in even-numbered years. Medical symposium and combined meetings of all core team member groups:
Canada:

- Association of Hemophilia Clinic Directors of Canada (AHCDC)
- Canadian Association of Nurses in Hemophilia Care (CANHC)
- Canadian Physiotherapists in Hemophilia Care (CPHC)
- Canadian Social Workers in Hemophilia Care (CSWHC)

- Annual meetings of core team member groups: held in odd-numbered years.

Outside Canada:

- World Federation of Hemophilia (WFH) World Congress
  - Held in even numbered years.
  - Attended by care team members, researchers and people with bleeding disorders
  - Program has Medical, Multi-disciplinary, and Musculoskeletal streams.

- WFH Musculoskeletal Congress
  - Held in odd-numbered years.
  - Attended mainly by team members and researchers
  - Excellent opportunity for interaction between orthopedic surgeons and physiotherapists

- WFH Global Forum on Research and Treatment Products for Bleeding Disorders – held in odd numbered years in Montreal.
  - Topics vary. Check website for program details.

- Congress of the European Association for Haemophilia and Allied Disorders.
  - Annual meeting; recent formation of a physiotherapy committee
  - http://eahad.org/

- National Hemophilia Foundation (USA)
  - https://www.hemophilia.org/Events-Educational-Programs
  - Annual meeting and conference
  - Attended by people with bleeding disorders, team members and researchers

Other Hematology Conferences:

- International Society of Thrombosis and Hemostasis (ISTH)
  - https://www.isth.org/
  - “At the ISTH Congress, thousands of the world’s leading experts on thrombosis, haemostasis and vascular biology come together to present the most recent advances, exchange the latest science and
discuss the newest clinical applications designed to improve patient care.”
  o Congresses held in odd-numbered years

- American Society of Hematology (ASH) [http://www.hematology.org/](http://www.hematology.org/)
  o “The Society's mission is to further the understanding, diagnosis, treatment, and prevention of disorders affecting the blood, bone marrow, and the immunologic, hemostatic and vascular systems, by promoting research, clinical care, education, training, and advocacy in hematology.”