let’s talk physiotherapy for haemophilia patients

An introductory guide for physiotherapists and haemophilia nurses

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Haemophilia B with inhibitors
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Haemophilia is a rare but serious inherited disorder which causes impaired blood clotting. Men and boys living with haemophilia are at risk of bleeds into their joints, especially knees, ankles and elbows. Joint bleeds are painful and reduce mobility and, over time, multiple episodes of bleeding can result in progressive and permanent joint damage. Physiotherapy has an important part to play in helping people with haemophilia recover from joint bleeds. A well-designed exercise programme can help patients maintain healthy joints, reduce pain and reduce the risks of future bleeding episodes. Ideally, a specialist physiotherapist should assess the patient’s current physical condition and tailor a programme to meet their needs.

This booklet provides a basic introduction to the current management of haemophilia and the roles that physiotherapy can play. It is aimed at healthcare professionals, including physiotherapists who manage people with haemophilia and haemophilia nurses who collaborate with physiotherapists to deliver musculoskeletal care. For a more in-depth consideration of the topics discussed here, please refer to the information sources listed in the ‘Helpful resources’ and ‘References’ sections.

Haemophilia is a hereditary disorder in which blood clotting is impaired. In normal blood clotting, thirteen different proteins – clotting factors – work together to form a blood clot. This series of events is known as the clotting cascade (Figure 1). People with haemophilia have a clotting factor that is missing or reduced, so blood clots fail to form properly. This makes bleeding last longer and be more difficult to control than normal.

Inheritance of haemophilia

Approximately 400,000 people are affected by haemophilia worldwide. Haemophilia A is the most common form, occurring in ~80% of all patients, and is caused by deficiency of Factor VIII (FVIII). Haemophilia B is caused by a lack of Factor IX (FIX) and occurs in ~20% of all people with haemophilia. The genes for Factor VIII and IX are located on the X chromosome, which is why the vast majority of people with haemophilia are male. A man with haemophilia can pass the affected gene to his unaffected daughter, who is known as a carrier, and through her to his grandson (Figure 2).

### Figure 2: Typical inheritance pattern of haemophilia showing how a haemophilia gene (x) is passed from grandfather to grandson through a carrier daughter.

![figure 2](image-url)
Symptoms of haemophilia
People with haemophilia do not bleed more than normal, but bleeding is prolonged and takes longer to stop. The severity of symptoms depends on the patient’s clotting factor levels: less than 1% of normal levels of FVIII or FIX is associated with severe disease, whereas higher levels are associated with moderate (1-5%) or mild (>5%) symptoms. People with severe haemophilia experience bruising, spontaneous bleeds and excessive bleeding following surgery or injury. People with haemophilia are prone to bleeds into their joints, with the majority of bleeds occurring in joints and known as haemarthrosis. Over time, cumulative bleeding into joints can cause long-term damage to the structure and function of the joint, known as haemophilic arthropathy. Medical treatment of haemophilia
Treatment options include replacement of the missing clotting factor, FVIII or FIX, which can be administered when a bleeding episode occurs (on-demand treatment) or at regular intervals to proactively prevent bleeding (prophylactic therapy). Prophylaxis is considered to be the ‘gold-standard’ treatment but is not always available or suitable for the individual patient. Replacement clotting factor can be recombinant (manufactured using genetic engineering) or derived from blood products (plasma-derived). Many patients with haemophilia are trained to administer clotting factor themselves, at home.

Inhibitors
Development of inhibitors (antibodies) to clotting factor therapy is an uncommon but serious complication that reduces the efficacy of haemophilia treatment. Risk factors for inhibitors include severity and type of haemophilia, family history, genetic and environmental factors as well as the clotting factor treatment exposure. Bleeding episodes are harder to manage in patients with inhibitors; management options include: higher doses of clotting Factors VIII or IX, immune tolerance therapy* and so-called bypassing agents.

Comprehensive care in haemophilia
People with haemophilia should ideally be managed in specialist comprehensive care centres. These centres should be staffed by haematologists, specialist nurses and physiotherapists, and social workers. They have access to laboratory services and clotting factor treatment, and can draw on specialist expertise when needed (e.g. orthopaedic surgeons, dentists, occupational therapists and infectious disease specialists).

Serious bleeds should always be treated in hospital

*Immune tolerance induction therapy (ITI) is used to get rid of high-titre inhibitors to clotting factor. It involves giving the patient repeated doses of factor VIII or IX over a length of time (say, 18 months) to make the patient tolerant of the clotting factor. The process reduces the immune reaction to administered clotting factor and eliminates the inhibitor.
Bleeding into the joints (haemarthrosis) and subsequent joint damage (arthropathy) are commonly seen in haemophilia. By adulthood, approximately 23% of severe haemophilia A patients will have developed one or more ‘target joints’. A target joint is one which is affected by repeated bleeds, reduced function and increasing joint damage. It can arise when several bleeds occur within the same joint over a period of a few weeks and the joint is unable to return to its original function between bleeds. 'Arthropathy' is a general term for the degeneration or destruction of one or more joints in the body. Many factors and conditions can trigger arthropathy, including joint bleeds, and inflammation. The joints affected by arthropathy in haemophilia ('haemophilic arthropathy') are synovial or hinge joints, i.e. joints which enable movement at the point of contact of articulating bones. The knees, elbows, and ankles account for ~ 80% of joint bleeds (Figure 3). While the knee is generally the most affected joint, research in the UK found that bleeds in the ankle and elbow are becoming more common in some people with haemophilia. Recent observations suggest that people with haemophilia who have experienced no or just a few joint bleeds can develop arthropathy, suggesting that subclinical bleeding episodes can cause joint damage.

The development of haemophilic arthropathy appears to include both degeneration of cartilage and bone as well as inflammation of the synovium (joint lining) called synovitis. Without appropriate treatment, a vicious cycle of bleeding, inflammation, damage and more bleeding can set in. Proliferation of the synovium in response to blood within the joint creates an inflamed, vascular and fragile tissue. Further bleeds can destroy the cartilage. Destruction of cartilage leads to arthritic, fibrotic joints which are painful and have reduced range of motion. In the longer term, joint damage negatively affects the person’s mobility, functional activities and quality of life and may require elective orthopaedic surgery to correct or improve function. Despite advances in treatment and care, joint bleeding and arthropathy remain among the most common complications of haemophilia and are major concerns of both healthcare professionals and people with haemophilia.
Physiotherapists should be specially trained to manage patients with haemophilia. Special care must be taken in order to avoid causing a bleed or exacerbating symptoms during assessment or exercise. For example, the physiotherapist should handle the patient carefully and confirm factor replacement therapy has been administered before the session, if needed.7,14

Physiotherapy is also used to support recovery from surgical procedures, especially elective orthopaedic surgery (EOS).11 Pre- and post-operative physiotherapy are essential to optimise the outcomes of EOS.10,11

It is important to assess disease progression by evaluating markers including bleeding frequency, pain, degree of swelling/synovitis, range of motion, crepitus, muscle power, balance and gait.22 Indeed, the Hemophilia Joint Health Score (HJHS) – one of the main tools used to assess joint status in children – is designed to be used by specially-trained physiotherapists.22,23 The HJHS was recently shown to be 74% more efficient than the World Federation of Hemophilia (WFH) physical examination scale in differentiating boys receiving prophylactic treatment from those treated on-demand.23 Because it prevents bleeds, boys receiving prophylaxis generally have better joint status than boys who treat bleeds on-demand.1

Physiotherapy forms a vital component of the management of patients with haemophilia.5 Exercise programmes help to manage recovery from a muscle bleed or joint bleed and help prevent future bleeding episodes.3 The overall aim of physiotherapy in haemophilia is to restore normal motor function. Specific goals include:

- Maintaining healthy joints and avoiding deformity.6,14
- Reducing pain.14
- Increasing function by maintaining range of motion (ROM).6
- Improving balance and proprioception which help to avoid injury.4,6
- Helping the person remain fit/active and maintain a healthy weight.1,6

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Available functional and physical scoring systems that can be used to assess joint status in haemophilia are summarised in Table 1. These specialist scales should be used by physiotherapists and other healthcare professionals who regularly see haemophilia patients and who are experienced in the use of these assessments.

Simple checklist for assessing joint status

A checklist has been developed to allow non-specialist physiotherapists and other healthcare professionals to monitor the status of their patients’ joints. The checklist asks a series of questions about the joint, with a combination of scoring of the joint condition and simple assessment of change over time. The checklist is a subjective assessment that is intended to help quickly identify any changes in function based on professionals’ observations (Figure 5). **

The checklist is included in the TalkingJoints® educational toolkit, available from Novo Nordisk.

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Figure 5: Joint status checklist

<table>
<thead>
<tr>
<th>Question</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>How severe is the swelling?</td>
<td>0-2</td>
</tr>
<tr>
<td>If yes how long has it been swollen?</td>
<td></td>
</tr>
<tr>
<td>Does the joint move freely?</td>
<td>Yes/No</td>
</tr>
<tr>
<td>Can the joint be fully flexed?</td>
<td>Yes/No</td>
</tr>
<tr>
<td>Can the joint be fully extended?</td>
<td>Yes/No</td>
</tr>
<tr>
<td>Does the joint hurt at rest without painkillers?</td>
<td>Yes/No</td>
</tr>
<tr>
<td>If yes, how long have you been taking painkillers?</td>
<td></td>
</tr>
<tr>
<td>If yes, which painkillers?</td>
<td></td>
</tr>
<tr>
<td>How severe is the pain when weight bearing?</td>
<td>0-10</td>
</tr>
<tr>
<td>Do you need to take painkillers to manage the pain?</td>
<td>Yes/No</td>
</tr>
<tr>
<td>If yes, how long has it been painful?</td>
<td></td>
</tr>
<tr>
<td>When is the pain worst during the day?</td>
<td></td>
</tr>
<tr>
<td>How severe is the pain during the day?</td>
<td>0-10</td>
</tr>
<tr>
<td>Sketch on the diagram (opposite) how the pain varies during the day</td>
<td></td>
</tr>
</tbody>
</table>

Scoring: 0 = Improved or no change 1 = Worsened

Management of joint bleeds: P.R.I.C.E.

• PROTECTION – protect the injured joint from undue stress, perhaps by using a sling or crutches.
• REST – rest the joint for at least 24 hours or longer.
• ICE – applying ice can limit joint damage as it reduces the temperature of the tissue at the site of the injury and can help ease pain.1
• COMPRESSION – wrap the injured joint in a compression bandage or strapping for the first 24 hours. Check frequently to make sure the wrap is not too tight, and use with caution in small children. Compression helps to control the amount of swelling and may lead to a quicker recovery.1, 7
• ELEVATION – elevate the injured area to lower the pressure in the local blood vessels and help to limit the bleeding into the area. Elevating the area will also help to increase drainage of fluids from the injury, which limits swelling.11

Exercise programmes for people with haemophilia can also help prevent bleeding episodes from occurring.1, 5 Studies have shown that children with strong musculoskeletal systems have fewer spontaneous bleeding episodes.16 The best prevention is to restore full functional activity after bleeding episodes (Figure 6 – overleaf).
Physiotherapy management of haemophilia

Physiotherapy exercise programmes form a vital part of the management of people with haemophilia. Physical benefits of exercise include improved strength, proprioception, joint range of motion and lean body mass, as well as reduced frequency of bleeding. Exercise strengthens muscle, and strong muscle protects joints. Proprioceptive (balance) training may promote joint stability and function. Weight-bearing exercise can improve bone health in children with severe haemophilia who have reduced bone mineral density. Exercise programmes for people with haemophilia are designed and implemented for a number of reasons, including: Reactive management of recovery after a muscle bleed, a joint bleed (haemarthrosis) or surgery Proactive prevention of future bleeding episodes General health and psychosocial well-being.

Conservative management, such as serial casting, bracing and orthotics (e.g. shoe inserts, crutches) may be used to correct deformities and support painful and unstable joints. Patients may need clotting factor administration prior to undertaking physiotherapy. It is important that the physiotherapist discusses this with the Haemophilia Treatment Centre (HTC), not just with the patient. The factor replacement guidelines set by the HTC should be followed before undertaking any physiotherapy or exercise programme, with infusion ideally before activity. Regular replacement therapy may not completely prevent bleeds, so it is important to monitor for any bleeding or other injuries, and follow HTC guidelines to manage these. It is especially important to monitor for any bleeding or injury in patients with inhibitors, as they are more likely to have musculoskeletal problems than people without inhibitors.

Table 2 lists some ‘do’s’ and ‘don’ts’ that should be kept in mind by healthcare professionals and patients.

Table 2: ‘Dos’ and ‘dons’ for exercise programmes in haemophilia

<table>
<thead>
<tr>
<th>DO</th>
<th>NOT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treat any joint bleed immediately with clotting factor, ideally within 2 hours and until the bleed resolves.</td>
<td>Exercise a joint during an active bleed.</td>
</tr>
<tr>
<td>Allow an appropriate rest period to allow for bleed resolution and healing, and prevention of early re-bleeding.</td>
<td>Avoid physical activity, as it can help prevent joint bleeds.</td>
</tr>
<tr>
<td>Put an exercise programme in place to help patients get mobile again following a joint bleed and help prevent future bleeds.</td>
<td>Return to a sport/regular activity following a bleed without discussing with the HTC.</td>
</tr>
<tr>
<td>Incorporate a range of exercises into exercise programmes.</td>
<td>Do high-impact sports or power lifting that may precipitate a joint bleed.</td>
</tr>
<tr>
<td>Ensure appropriate clotting factor cover during exercise if recommended by the HTC.</td>
<td>Forget to exercise other body parts as able, while resting the joint that is healing.</td>
</tr>
<tr>
<td>Set functional goals that assess progress tailored to the patient.</td>
<td>Do not.</td>
</tr>
<tr>
<td>Follow HTC guidelines for the individualised management of any bleeding or injury that may occur.</td>
<td></td>
</tr>
</tbody>
</table>

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Elective Orthopaedic Surgery (pre- and post-op physiotherapy)

When carried out by an experienced and well-equipped multidisciplinary team, with appropriate physiotherapy support, elective orthopaedic surgery (EOS) can be safely performed in patients with haemophilia, even those with inhibitors.43 EOS procedures can offer a number of benefits to patients with haemophilia:14, 44 – 47

• Significantly improve joint function
• Reduce pain due to chronic arthropathy
• Improve mobility
• Reduce the frequency of bleeding episodes
• Improve quality of life.

There is a trend towards a more aggressive approach to surgery in younger patients based on the rationale that these patients can benefit from optimal physical health. In addition, the musculature of these younger patients is still in a good condition and thus can usually be more easily rehabilitated following orthopaedic surgery than that of older patients.48

Table 3 lists important considerations for developing an exercise programme that is tailored to the individual needs of the person with haemophilia.13 Specialist physiotherapy input should always be sought when establishing a new programme for a patient with haemophilia, especially if the patient has inhibitors.

It is important that any exercise programme for a person with haemophilia is designed with their functional goals in mind.49 Functional goals can include returning to work or school, participating fully in social activities or taking up a new sporting activity. Specific exercises and physical activities should be recommended to help patients reach their goals, with progress assessed by how much these goals have been achieved.50

Control of arthropathic pain: analgesics

People with haemophilia need to be able to distinguish between pain from arthropathy and pain that could indicate an active bleed19 (see callout box). It is important that physiotherapists listen to the individual when he reports pain during exercise and do not encourage patients to over-exert themselves.13 The patient knows their condition best. For the control of arthropathic pain, analgesics are commonly used.

Non-steroidal anti-inflammatory drugs should be used with caution.1 A combination of analgesics (with their different mechanisms of action) may be the most effective way of relieving pain.51 As discussed above, protection, rest, ice and compression can also help reduce the pain of a bleeding episode.

Box 2: Characteristics of pain associated with a bleed

• Does not reduce when the joint is rested
• Is accompanied by one or more of:
  – Palpable swelling of the joint
  – Warmth of the skin over the joint
  – Sensation of tingling (‘aura’)
  – Progressive loss of range of motion
• Reduces with appropriate clotting factor treatment.

Characteristics of pain associated with a bleed52

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Compared with other patients undergoing EOS, patients with haemophilia tend to be younger, have greater and more widespread joint and soft tissue damage, require more intensive post-operative rehabilitation and be affected by pain from joint bleeds and from joint damage.\(^\text{19}\)

The physiotherapist plays a number of important roles in preparation for surgery (‘pre-habilitation’) and in post-operative recovery (rehabilitation), summarised in Table 4.

The goals of post-operative physiotherapy include recovery of strength and range of motion, re-education of gait and proprioception, and maintenance of unaffected joints and muscles.\(^\text{13,19}\) Individual motivation\(^ 1\) and inhibitor status\(^ 4\) should be considered when deciding timing of mobilisation. Early mobilisation delivers a better range of motion post-surgery but might trigger a bleed, especially in patients with inhibitors.\(^\text{21,48}\) To minimise the risk of bleeding during rehabilitation, physiotherapy should be timed to coincide with administration of factor replacement\(^\text{19}\) or covered by factor administration prior to a training session.\(^\text{17,18}\) Some experts recommend the use of continuous passive motion (isometric movement) to facilitate early mobilisation while reducing the bleeding risk.\(^\text{45}\)

### Table 4: Physiotherapist roles related to EOS in haemophilia

<table>
<thead>
<tr>
<th>Pre-operative roles: ‘pre-habilitation’</th>
<th>Post-operative roles: ‘rehabilitation’</th>
</tr>
</thead>
<tbody>
<tr>
<td>Evaluate the person’s adherence to prescribed treatments after surgery.(^\text{21})</td>
<td>Support the person in carrying out an individualised exercise regimen to strengthen muscles and increase range of motion.(^\text{20})</td>
</tr>
<tr>
<td>Determine a pre-operative rehabilitation programme in preparation for surgery.(^\text{19})</td>
<td>Tailored to the person’s needs and capability(^ 1)</td>
</tr>
<tr>
<td>• Pre-operative exercises to strengthen muscles and increase mobility after surgery.(^\text{13})</td>
<td>With appropriate haemostatic coverage and pain management.(^\text{19})</td>
</tr>
<tr>
<td>• Use of mobility aids such as crutches.(^\text{20})</td>
<td>Commence post-operative rehabilitation in a timely manner after surgery.(^\text{20})</td>
</tr>
<tr>
<td>Predict post-operative function.(^\text{19})</td>
<td>Determine plan for post-operative rehabilitation.(^\text{19})</td>
</tr>
<tr>
<td>Determine plan for post-operative rehabilitation.(^\text{45})</td>
<td>Secure the person’s commitment to the post-operative rehabilitation exercise programme.(^\text{20,48})</td>
</tr>
</tbody>
</table>

\(^1\) Individual motivation, \(^2\) Inhibitor status, \(^3\) Haemostatic coverage and pain management, \(^4\) Commence post-operative rehabilitation in a timely manner after surgery.
Exercise can improve not only physical well-being, but also the emotional and social well-being of people with haemophilia. Physical activity helps children develop socially and emotionally. In competitive sports, they learn teamwork. When people choose to participate in a sport they enjoy, it offers a number of benefits including better physical and mental health and improved bone and joint health and function.

Several components make up a well-rounded, complete fitness programme, including endurance or cardiovascular activities, muscle strengthening and flexibility exercises, and balance and stability exercises. Starting exercise at an early age can build a strong routine that will be beneficial throughout life.

When selecting sports activities for people with haemophilia, safety is an important consideration: activities must be age-appropriate and properly supervised. Ideally activity should take place soon after clotting factor treatment, although exercise can be safe even in the absence of replacement factor. Other safety considerations include appropriate warm-up and cool-down exercises, the using of protective gear, and choosing organised, supervised sports programmes rather than unstructured activities.

The National Hemophilia Foundation book ‘Playing it Safe’ lists a number of sports activities graded by whether they can or cannot be recommended for people with haemophilia when carried out at an appropriate intensity for the individual’s level of fitness and experience. Less risky sports include archery, stationary bike, fishing, golf, hiking, tai chi and swimming. Recent evidence shows the benefits for people with haemophilia of a range of activities, including sports, aquatic exercise and resistance training.

There are risks and benefits to all activities, and advice should be sought from the Haemophilia Treatment Centre before recommending a new or changed activity.

Specialist input should be sought whenever there is doubt about the patient’s treatment or prognosis. Specific circumstances include:

- Initiating a physiotherapy regime
- Assessing the patient’s baseline function
- Poor recovery in response to a physiotherapy programme
- Inadequate pain management
- When starting or choosing a sport or exercise programme

TalkingJoints
feelsomething, do something
**Glossary of terms**

**Arthropathy** Degeneration of one or more joints.

**Bypassing agents** Agents or products designed to bypass deficient clotting factor.

**Clotting factor** One of thirteen proteins involved in the blood clotting cascade. The factors missing or deficient in haemophilia are Factor VIII or IX.

**Crepitus** A grinding sound or sensation produced by friction between bone and cartilage.

**Elective orthopaedic surgery (EOS)** A term which covers a range of surgical procedures including synoviectomy and joint replacement.

**Factor replacement therapy** In haemophilia, the injection of clotting factors to substitute for the body's missing clotting factors. Alternatively, if the patient is ITI or ITT refractory, transfusion of replacement therapy in haemophilia B. Factor B is used.

**Haemarthrosis** Bleeding into a joint, most commonly a hinge joint (knee, ankle or elbow).

**Haemophiliac arthropathy** Joint damage associated with repeated bleeding into the joint in people with haemophilia. In later stages, it can lead to wearing away of the bone around the joint, causing pain and crippling mobility.

**HIV** Human Immunodeficiency Virus, a validated joint assessment tool for specialist physiotherapists.

**Immune tolerance** The most common way to get rid of inhibitors to clotting factor. It involves giving the patient repeated doses of Factor VIII or IX over a length of time (a few months to two years).

**Inhibitor** One of thirteen proteins involved in the blood clotting cascade. The factors missing or deficient in haemophilia are Factor VIII or IX.

**Joint Health Score** a global perspective. Haemophilia, 2010; 16 Suppl 5: 136–45.


**National Hemophilia Foundation. Playing It Safe: Bleeding Disorders, Sports and Exercise 2005, New York:**

**Reference list**


“let’s talk physiotherapy” is part of the TalkingJoints® programme to encourage better understanding of how physiotherapy can help look after the body and joints of patients with haemophilia.

TalkingJoints® is a programme of information, education and support that talks to healthcare professionals, patients and carers about haemophilia and its impact on joint function. TalkingJoints® aims to help individuals detect changes early (feel something) and act accordingly (do something). We hope that by helping people with their joint function we can help improve the way they live with haemophilia, for the better.