Pfizer Haemophilia

Let's talk about **physical activity in adults with Haemophilia**

Contents

Introduction

People with haemophilia are living longer lives thanks to the evolution of haemophilia care and the development of effective treatments. However, living longer does not necessarily mean living healthily and – just like the general population – people with haemophilia also suffer from age-related comorbidities such as diabetes, obesity and cardiovascular diseases. Leading a physically active and healthy lifestyle can help to prevent these comorbidities, but what does 'physical activity' really mean? For Sébastien it can be playing tennis with his friends; for Pål, it is skiing in the mountains; and for Thomas, simply mowing the lawn.

Selecting a physical activity that we are enthusiastic about is important in order to optimise ongoing participation. The choice of activity will be different for every person with haemophilia and is influenced by a number of factors: a person's preferences, where they live, their personal goals, the risks involved, their joint status, their treatment regimen, and a multitude of other individual characteristics. However, even after starting an activity, the fear of bleeding and the consequences of bleeding episodes can weaken the motivation to keep going. Strategies to minimise activity-related risks and keep people with haemophilia focused on their personal goals are key to achieving successful outcomes.

This booklet aims to provide the information to support conversations between healthcare professionals and adults with haemophilia to help encourage, maintain and inspire physical activity. It also highlights the considerations that should inform the design of a personalised physical activity / sports programme. The booklet will also be useful to physiotherapists, sports specialists, specialist nurses, sports instructors and others who want to know more about the benefits of physical activity for people with haemophilia and support a multidisciplinary team approach to the care of people with haemophilia.

This booklet is part of a series that started with Haemophilia and Sports – a booklet focused on the benefits and limitations of physical activity in children and adolescents with haemophilia.

Sébastien Lobet, PT, PhD
Haemostasis and Thrombosis Unit,
Cliniques Universitaires Saint-Luc, Brussels, Belgium

Pål Andrè Holme, MD, PhD
Department of Haematology and Institute of clinical medicine,
Oslo University Hospital, University of Oslo, Norway

Thomas Hilberg, MD, PhD
Department of Sports Medicine,
University Wuppertal, Wuppertal, Germany

Disclaimer: This booklet was produced by Dr S. Lobet (a Physical Therapist at the Saint-Luc University Clinics of Brussels), Professor P.A. Holme (Physician at the Department of Haematology, Oslo University Hospital and University of Oslo) and Professor T. Hilberg (Physician at the University Wuppertal, Wuppertal, Germany). This booklet was designed for use by healthcare professionals only, to support discussions with patients around physical activity. Neither the authors, the publishers, nor the sponsor are liable for the information given in this brochure, and they disclaim all liability in case of injury. Recommendations about physical activity should be assessed and tailored to each individual patient. Funding for the development of this booklet was provided by Pfizer. Pfizer did not influence the content development of this document. Editorial support to prepare this document was provided by SYNERGY (London, UK), a Medical Education provider.

Chapter 1
Living with haemophilia

As people with haemophilia enjoy longer lives, they are likely to experience similar age-related comorbidities as the general population, as well as facing some unique challenges due to the chronic nature of haemophilia. In this chapter, we review some of the age-related comorbidities that may not only impact a patient's ability to take part in physical activity but can also be improved or prevented through a more active lifestyle. Most comorbidities discussed in this chapter occur in older adults. However, as will be discussed further throughout this booklet, a sustainable physical activity programme can put people with haemophilia of all ages on the right track towards a healthier future.[1-4]

Haemophilia in adults

Increased life-expectancy in people with haemophilia

In the past, people with haemophilia had a shortened life expectancy when compared with the general population[5]; intracranial bleeds or other haemorrhages were the major cause of mortality.[6] Since the discovery of cryoprecipitate in the mid-1960s, several key developments – such as the introduction of plasma-derived clotting factor concentrates and, subsequently, recombinant products, as well as home treatment and primary and secondary prophylaxis – have meant that people with haemophilia are living longer than ever before.[7, 8] With continuing evolution in the management of haemophilia and the availability of new treatment options, people with haemophilia are expected to have a life-expectancy comparable to the general population.[9, 10]

However, an aging haemophilia population is likely to experience the same comorbidities as the general population, such as obesity[11] and cardiovascular disease,[11] as well as problems specific to haemophilia, such as haemophilic arthropathy[8] and the development of target joints.[12] Haemophilia may also put individuals at increased risk of certain comorbidities, including hypertension, osteoporosis and complications relating to viral infections (Figure 1.1).[8, 11]

Impact of haemophilia on quality of life

People with haemophilia report a lower quality of life when compared with the general population.[8, 13] The reasons for this appear to be multifactorial and vary with age, treatment regimen and the severity of comorbidities.[13, 14] In order to assess the full spectrum of consequences of the disease, use of the WHO International Classification of Functioning, Disability and Health (ICF) has been suggested for people with haemophilia.[15] The elements involved in this framework include joint function and structure, type of activities reported by the individual, participation level in society (e.g., days lost of school / work or paid employment), and economic burden (clotting factor consumption, haemophilia-related surgeries, hospital visits, days in hospital and utility assessment).[15]

Depression in the haemophilia population is not only associated with disease severity, presence of viral infections[16] – e.g. human immunodeficiency virus (HIV) / hepatitis B and C viruses (HBV, HCV) – age and/or functional impairment, but also a perceived lack of social support and unemployment. Data suggest that the promotion of social interaction, community involvement, education and self-reliance is therefore important for the well-being of people with haemophilia.[17] As we discuss later, one way to achieve this is by increasing physical activity through sports therapy and other sports activities.

Access to treatment appears to have a significantimpact on health-related quality of life, with improvements seen in both physical and mental assessments when patients switch from on-demand to prophylactic regimens.[14, 18, 19] These improvements in health-related quality of life have been attributed to reductions in pain, fewer restrictions on physical activities and a better perception of health.[19]

Despite the burden of living with a chronic disease, people with haemophilia are able to develop good coping strategies.[20] They can feel empowered by focusing on their strengths and by taking control of their condition. For these individuals, haemophilia is part of their identity, and they may regard their life and achievements in the context of the challenges that they have overcome.[18] With this in mind, involvement in physical activity could potentially give people with haemophilia feelings of empowerment and achievement, thus benefiting both their physical and mental health. The benefits of activity and steps to encourage and individualise physical activity are covered in Chapter 4 and Chapter 5, respectively.

Joint damage and haemophilic arthropathy

Arthropathy is a common challenge in adults with haemophilia.[11] It is particularly prevalent in those who have not benefited from routine prophylactic treatment from a young age.[11] Blood-induced joint damage is a multi-step process involving synovial inflammation, cartilage degeneration and bone alterations.[15] Repeated bleeding in the same joint can result in the development of target joint. Joint damage varies in terms of progression and symptoms,[21] and the relationship between the number of joint bleeds and joint status is not always clear. For example, in the Manco-Johnson study (2007), bone and cartilage damage detected on MRI was not correlated with the number of clinically evident haemarthroses.[22] Patients with joint damage will experience an impaired range of motion, joint instability, contractures, muscle atrophy, chronic synovitis and, ultimately, impaired mobility.[23] Once in a state of chronic synovitis, mechanical, chemical and enzymatic processes can result in irreversible damage, which may trigger osteoarthritic processes. At this stage, treatment of bleeding alone will not be able to prevent further progression.[24] It is important to acknowledge the impact of haemophilic arthropathy on patients' lives in order to help them manage their chronic pain and to reduce the risk of falls and fractures.[11] As discussed in Chapter 3, the strengthening of muscles and tendons around the joints that results from being physically active can help prevent haemophilic muscle atrophy, may reduce the number of bleeding episodes and minimise the risk of injury risk.[1, 3]

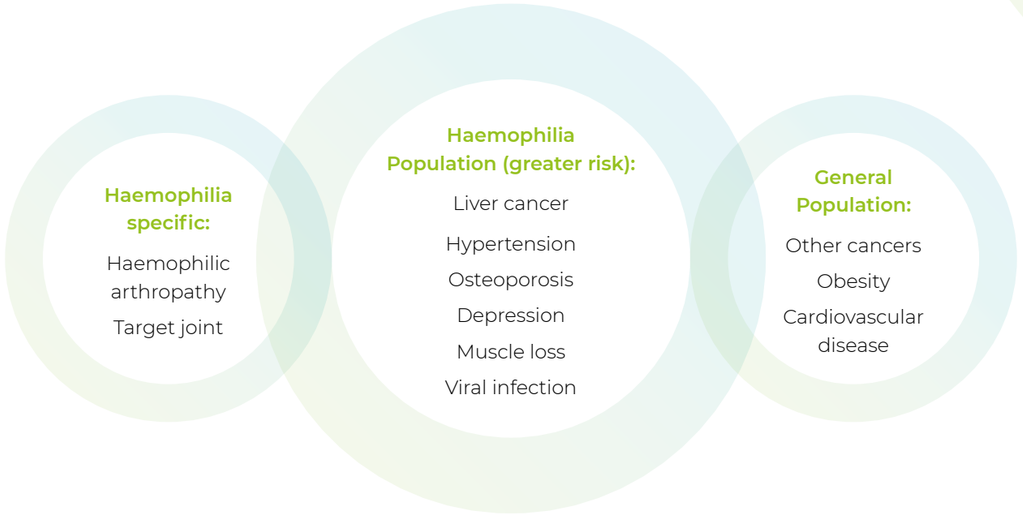


Figure 1.1
Age-related comorbidities seen in haemophilia patients and the general population.
People with haemophilia are likely to suffer from the same comorbidities as the general population (right circle), as well as from comorbidities directly related to their condition (left circle). This population is also at a greater risk of certain comorbidities (middle circle).[8, 11]

Age-related comorbidities in people with haemophilia

Overweight and obesity

Patterns of overweight and obesity vary across the globe but rates in people with haemophilia have been reported to be equal to or higher than those seen in the general population.[25, 26] Like the general population, people with haemophilia can be at risk of weight-related complications such as hypertension, diabetes, cancer and hyperlipidaemia.[11, 26] As discussed further in Chapter 3, these complications can be modified using exercise and physical activity to reach and maintain a healthy body weight.[2]

Obesity also adversely affects bone health through the alteration of bone-regulating hormones, increased oxidative stress and inflammation, and altered bone cell metabolism.[27] Obesity also affects bone quality, which may explain the higher than expected risk of fractures in obese individuals in the general population due to reduced bone mineral density (BMD; discussed later).[27] The relationship between adiposity and osteoporosis is complex. Further exploration to establish whether the risk of obesity-related complications is the same for people with haemophilia compared with the general population is required.

Cardiovascular comorbidities

As individuals with haemophilia age, they are likely to be exposed to the same cardiovascular risk factors as the general population.[11] However, the prevalence of different cardiovascular comorbidities varies between the two populations.[11, 28] For example, multiple studies demonstrate a higher prevalence of hypertension in people with haemophilia,[28, 29] but there is conflicting evidence about whether the prevalence of other cardiovascular comorbidities / events – such as coronary artery / heart disease and stroke – is different in people with haemophilia when compared to the general population.[11, 28, 30] In one study, despite having similar cardiovascular risk factors, people with haemophilia had fewer cardiac events than the general population.[31] Some authors argue that bleeding tendency is associated with a lower risk of cardiovascular disease,[32] but whether and to what extent haemophilia exerts a protective effect on the incidence of thrombotic events is unclear.[11] People with haemophilia should be screened for cardiovascular risk factors[32] and can be managed with similar approaches as in the general population,[30] including participation in physical activity.[2]

Bone mineral density (BMD) and osteoporosis

People with haemophilia can experience a considerable reduction in BMD from childhood, with a significantly lower bone mass seen in the lumbar spine and hip.[8, 33] Several factors contribute to reduced BMD, including severity of haemophilia, type of treatment (on-demand, prophylaxis), severity of arthropathy, presence of HIV and / or HCV infection, level of activity (weight-bearing) and vitamin D deficiency.[8] The main cause is likely to be haemophilic arthropathy, which is associated with chronic pain and loss of joint function, leading to decreased weight-bearing physical inactivity.[8] While young patients treated on-demand tend not to reach the same peak bone mass as their healthy counterparts, prophylactic treatment can preserve BMD in people with severe haemophilia.[8] This protective effect appears to be connected to the ability to participate in physical activities.[8] Since older individuals with haemophilia are less likely to have received prophylaxis from a young age, they may have reduced BMD.[8] It is unsurprising therefore that while osteoporosis does occur in the general ageing population, it tends to occur at an earlier age in those with haemophilia.[8] Practising weight-bearing exercise from adolescence has been shown to improve BMD and may help to protect individuals with haemophilia from developing osteoporosis in later life.[1, 2] In the general population, optimisation of lifestyle factors is an important strategy to reduce the risk of osteoporosis or low bone mass later in life.[34]

Falls and fractures

Adults with haemophilia are at a higher risk of fractures and falls compared with the general population, and have poorer outcomes in balance and mobility tests, as well as demonstrating impaired leg strength and knee joint proprioception.[11, 35-37] Impaired proprioceptive function impacts patients' daily lives, particularly in situations where unimpaired proprioception is required, such as walking on a slippery surface.[37] The impaired physical and proprioceptive performance observed in some individuals with haemophilia may be due to joint bleeds or decreased usage of these joints, indicating the importance of specialised training to improve strength and proprioceptive performance.[37] The increased risk of fractures seen in people with haemophilia is believed to be correlated with disease severity and age.[11, 36]

Viral infections in people with haemophilia

The emergence and transmission of HIV, HBV and HCV through plasma-derived clotting factor concentrates resulted in high mortality rates in the haemophilia population in the 1980s and 1990s.[4] Thankfully, mortality and morbidity rates have now declined, but infection with HIV and HCV is still present in the older haemophilia population.[11]

Knowledge and expertise around the treatment of HIV-infected (HIV-positive) people with haemophilia is limited and treatment is mainly based on the guidelines developed for the non-haemophilia population.[4] Diagnosis, counselling, initiation of treatment and monitoring of HIV and treatment of HIV-associated complications in HIV-positive people with haemophilia is the same as for the HIV-positive non-haemophilia population.[4] Infection with HCV may affect the liver and increases the risk of chronic hepatitis, including end-stage liver disease and hepatocellular carcinoma, as well as affecting various non-hepatic tissues and organs.[5] The duration of HCV infection tends to be longer in people with haemophilia compared with other patient groups, since it was acquired at an earlier age as a result of receiving haemophilia treatment.[6] Thanks to recent treatment advances including direct acting antivirals, the cure rate for HCV has improved significantly.[38, 39] Where HCV eradication cannot be achieved, regular monitoring for end-stage liver complications is recommended.[4]

Viral infections and their associated comorbidities may impact a patient's ability to participate in physical activity and should be considered when planning a physical activity programme.†

† Author Recommendation

Transitioning from paediatric to adult care

As they approach adulthood, people with haemophilia undergo transition of care from paediatric to adult healthcare services. Ideally, transition should start as early as possible, such that by the time individuals reach the age of 18 years they understand all aspects of disease management and how to access the healthcare system.[40] The evolution of disease management over time should consider the natural medical history of the disorder, the cognitive and social development of the child and the psychosocial dynamics of the family setting.[40] As mentioned earlier, the International Classification of Functioning, Disability and Health (ICF) can be a useful tool to assess outcomes, particularly during the transition period.[15] The activity and participation dimensions of ICF are of particular interest for people with haemophilia since their activities may be restricted by their condition, which may in turn affect their ability to join in with social activities[15] (e.g. participating in a swimming club).

Often the biggest decision individuals are required to make during the transition of care is whether to continue with prophylactic therapy.[40] While some studies have demonstrated a benefit to continuing prophylaxis from the standpoint of preventing bleeding episodes, most adults discontinue prophylaxis for a variety of reasons[41] (e.g. infrequent bleeds, venous access, cost, etc.).[42]

Other factors that affect transition include: bleeding that results from typical activities; the timing of initiation of treatment (which is most effective when initiated early); practical challenges of administering therapy (e.g. intravenous infusion, or having treatment accessible); and the high costs of care.[40] A particularly important challenge is related to treatment adherence leading to a higher risk of recurrent joint bleeds, chronic pain and reduced quality of life.[43] With all of these challenges in mind, multidisciplinary comprehensive care is needed in order to ensure an easy transitional process for patients and their families.[40]

Summary: Impact of haemophilia in adults and considerations for sports/physical activity

Impact of haemophilia in adults	Considerations for sports/physical activity
Adults with haemophilia often experience feelings of depression and lower quality of life compared with the general population[8, 13, 16, 17]	Sports / physical activity may improve quality of life by promoting social interaction and self-reliance, as well as increasing feelings of empowerment and achievement[15, 44]
Joint damage is common in people with haemophilia[11]	Regular physical activity can help to prevent haemophilic muscle atrophy and may reduce the number of bleeding episodes, potentially protecting joints from damage[1, 3]
Similar rates of overweight and obesity are seen in the haemophilia and general populations, with the exception of older people with severe haemophilia who tend to have a lower body mass index (BMI)[25, 26] (likely to reflect reduced muscle mass)[45]	Exercise and physical activity can help people reach and maintain a healthy weight and benefit from the associated improvements in health[2]
Rates of hypertension are supposed to be higher than in the general population[11, 28-30]	Physical activities that improve cardiovascular fitness should be encouraged to help reduce cardiovascular risk factors and prevent cardiovascular disease[2, 46]
People with haemophilia experience a lower bone mass density (BMD) from an earlier age, putting them at increased risk of fractures and an earlier onset of osteoporosis[8, 11, 33]	Weight-bearing exercise can help build and maintain high BMD and may help prevent the development of osteoporosis in later life[1, 2]
In older people with haemophilia, and those with more severe disease, fracture rates are particularly elevated representing an important comorbidity to be considered during their care[11, 35-37]	Impaired physical and proprioceptive performance may be in part due to decreased usage of these faculties and may be improved through strength training[37]

Chapter 2

Psychosocial aspects of life with haemophilia

People affected by chronic health conditions, such as haemophilia, face challenges beyond purely medical issues.[47] Accordingly, haemophilia management guidelines and other sources recommend a comprehensive approach that addresses both clinical / physical and psychosocial dimensions.[4, 48] This approach includes, for example, an appropriate treatment regimen to control bleeding frequency and minimise joint damage.[4] In this chapter, we provide an overview of psychosocial challenges that people with haemophilia may face, together with a set of potential solutions to these issues, including physical activity. The psychosocial challenges faced by people with haemophilia may be divided into three broad, overlapping groups:

1. Impact on family and social interaction

2. Psychological aspects related to treatment and experiences with the healthcare system

3. Other psychological challenges

Impact on family and social interaction

Family interaction

Challenges for the family of an individual with haemophilia start with their reactions to diagnosis and their ability to learn to accept and adjust to life with the disease.[48] This may be particularly difficult in families with no history of familial haemophilia.[48] Initial reactions typically include disbelief, fear or guilt, combined with anger, denial, panic or confusion; these reactions usually decline over time.[48] All aspects of family life can be affected, such as relationships among family members, mobility, social activities and relationships, careers and finances.[48]

Adults with haemophilia may experience a sense of social stigma, leading to a reluctance to disclose their condition, fear of rejection in personal relationships and worries about starting their own family.[49] These concerns may be particularly difficult for those affected by HIV or HCV infection.[49] In addition to the psychosocial considerations, practical aspects of haemophilia can affect sexual relationships, including physical limitations and treatment-related infections.[49]

Some individuals with haemophilia may experience family overprotection during childhood, leading to problems with school attendance and social integration.[49] This may further result in poor academic achievement, low expectations and eventual difficulties at work.[49]

Transition from paediatric treatment to self-management

Adolescents with haemophilia can face psychological challenges in adapting to the constraints of their disease coupled with taking on the responsibility of managing their self-care.[40] The latter, in particular, is compounded by the transition from paediatric to adult haematology care services, when individuals with haemophilia need to build mature interpersonal relationships and establish the foundations of an independent career.[40, 43, 50] Parents of an adolescent / young adult with haemophilia also need to adapt to the change in situation and must manage their own psychological stress when the 'child' initiates self-care.[49] For more details on transition, see Chapter 1.

Social interaction

Adults with haemophilia need to decide when and how to reveal their diagnosis, as disclosure could result in negative social consequences, such as misunderstanding and misconceptions, social exclusion or stigma.[47] People with haemophilia may also perceive themselves as different due to physical limitations and the demands of treatment, both of which can impair the feeling of living a 'normal life'.[47]

Psychological aspects related to treatment and experiences with the healthcare system

Early prophylaxis and home treatment

Where appropriate and possible, people with haemophilia should be managed in a home therapy setting.[4] It allows immediate access to clotting factor and hence optimal early treatment, which in turn helps to control pain and minimise dysfunction and long-term disability.[4] Moreover, people with haemophilia can treat themselves at home in case of injuries sustained during physical activity. Psychosocial challenges related to maintaining prophylaxis include difficulties in engaging positively with the child, the time required to administer treatment and remaining motivated to treat even when haemorrhages are less frequent or absent.[48]

Continuing prophylaxis in adolescence and adulthood[40, 49]

Adherence issues in adolescents / young adults tend to relate to discordance with their lifestyle, teenage rebellion, lack of knowledge, and perceived benefits of treatment and difficulties. These may lead to demotivation and lack of attention to proper treatment. The routines required by regular prophylaxis – or on-demand treatment – affect the individual's sense of leading a 'normal life' and make him feel different. Fear of pain and frequent injections can also be a psychological barrier, even in adults who have experienced long-term treatment.[49]

Experiences with the healthcare system

An important aspect of managing haemophilia is the patients' experiences with the healthcare system. A lack of understanding from non-haemophilia-specialist clinicians may cause individual psychological distress and concerns about the appropriate management of their disease.[47, 49]

Other psychological challenges

Adults with haemophilia may be faced with uncertainty related to disease progression and emotional distress associated with symptoms, pain and other aspects of the disease.[47] People with haemophilia are at an increased risk of developing depression compared with the general population.[17] Depression can contribute to poor outcomes (e.g. functional impairment, decreased quality of life), decreased treatment adherence and an increase in risky behaviours (e.g. excessive use of alcohol and narcotics). An important risk factor for depression is perceived lack of social support and / or unemployment.[17]

Many people with severe haemophilia experience chronic and / or acute joint pain.[49, 51] Pain is a critical aspect of haemophilia, contributing to the psychosocial burden of the disease, and its effective management is a crucial element of haemophilia care.[52] For more details on pain, see Chapter 6.

Psychosocial challenges in selected haemophilia populations

Patients with inhibitors generally report worse treatment-related outcomes, more comorbidities (such as joint disease) and may require high-intensity treatment and hospitalisation. These challenges can exacerbate the psychosocial burden already present in people with haemophilia who do not have inhibitors.[51, 53]

As they age, people with haemophilia face similar problems to the general ageing population, such as increased presence of comorbidities and psychological issues.[13] However, ageing with haemophilia comes with an additional set of psychosocial challenges. An individual's autonomy may be affected by the demands of treatment, uncertainty about disease evolution, emotional distress,[47] declining health, changes in support networks, loss of employment or early retirement.[49] Some previously established routines might need to be changed and the possibility of independent life might be at risk.[49]

Solutions to psychosocial challenges in people with haemophilia

Even though people with haemophilia are faced with numerous psychosocial challenges, they are able to develop excellent coping strategies that – together with tailored support, counselling and advances in haemophilia treatment – support their mental and emotional well-being.[49]

A supportive family plays a key role in the well-being and function of a person with haemophilia, promoting adaptation to disease, reducing its impact and encouraging treatment adherence.[49] Successful adherence to treatment requires a forward-looking and optimistic approach with the understanding that their future health is worth the present investment.[49, 53] Family members may also act as 'co-therapists', providing additional emotional as well as practical support.[49, 54] Social support through interpersonal relationships allows people with haemophilia to share their experiences and receive emotional support.[49] Employment helps adults with haemophilia to develop a strong self-image.[55] Promotion of social interaction, community participation, networking, education and independence have been shown to be crucial factors in maintaining an individual's well-being.[17]

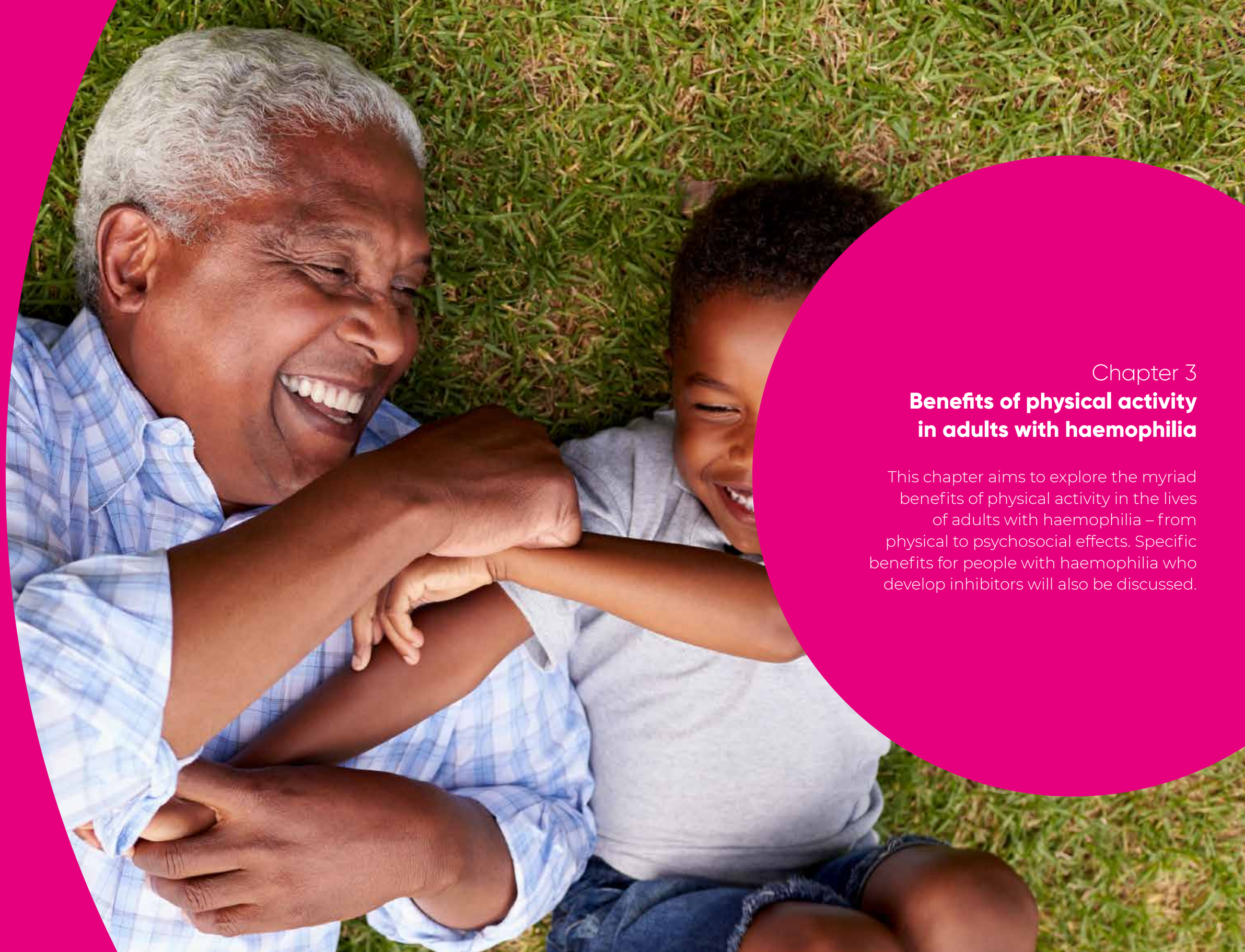
For people with haemophilia, a good therapeutic alliance with their multidisciplinary healthcare team is crucial. Higher patient satisfaction is likely to result in higher motivation and treatment adherence. Building trust in the physician–patient relationship may also help patients feel more comfortable communicating possible difficulties, barriers and emotional needs, which facilitates identification of solutions as quickly as possible.[49]

Psychosocial challenges and physical activity in people with haemophilia

Maintaining physical activity is crucial in people with haemophilia, as it exerts benefits beyond the physical domain, contributing to improved psychosocial outcomes (for more details, see Chapter 3). Physical activity may help control stress, improve self-esteem and body image, and potentially help to overcome any tendency to withdrawal and depression[55] (discussed in Chapter 4). Recommendations and practical advice should be tailored to a patient's individual needs (for more details, see Chapter 5).[56] It is therefore particularly important to involve people with haemophilia in decision-making and understand their perception of their health status.[19]

Summary: Key psychosocial challenges in people with haemophilia and potential solutions[13, 48, 49, 53]

Dimension	Challenges	Solutions
Family & social interaction		
Family interaction	• Initial reactions to diagnosis	• Tailored support, counselling • Promotion of social interaction, community participation, networking, education, independence • Seeking more information on the disease • Forward-looking and optimistic approach • Supportive family • Social support • Good therapeutic alliance with their multidisciplinary healthcare team • Maintaining physical activity
Social interaction	• Sense of social stigma and fear of rejection • Revealing the diagnosis • Family overprotection and / or absenteeism • Change in family dynamics	
Transition from paediatric treatment to self-management	• Taking responsibility for self-management • Establishing relationships with healthcare professionals	
Psychological aspects related to treatment & experiences with the healthcare system		
Early prophylaxis & home treatment	• Initiation of prophylactic treatment • Learning to administer intravenous treatment	
Continuing prophylaxis in adolescence & adulthood	• Adherence (demand of the treatment) • Fear of pain and injections	
Experiences with the healthcare system	• Lack of understanding from healthcare professionals	
Other psychological challenges		
	• Risk of depression • Chronic joint pain	



Chapter 3

Benefits of physical activity in adults with haemophilia

This chapter aims to explore the myriad benefits of physical activity in the lives of adults with haemophilia – from physical to psychosocial effects. Specific benefits for people with haemophilia who develop inhibitors will also be discussed.

Long-term benefits of physical activity in the general population

Long-term benefits of physical activity in the general population

The benefits of physical activity on long-term health are well recognised with the knowledge that "exercise is medicine"[57] expanding during the last decades.[58] High-level scientific evidence has shown that physical activity has the greatest impact on the reduction of cardiovascular disease risk by improving endothelial function, reducing high blood pressure, decreasing high cholesterol levels, and preventing metabolic syndrome and diabetes.[58-60] Physically active adults have lower rates of cancer, such as colon and breast cancer.[61, 62] In some musculoskeletal diseases, such as osteoarthritis and non-specific chronic low-back pain, physical activity may also be associated with an improvement in pain symptoms.[62, 63]
Physical activity also plays a key role in psychological well-being, reducing symptoms of anxiety and depression as well as conferring social benefits, ranging from building social skills in children to maintaining social interaction in the elderly.[58]

In addition to these health benefits, studies have consistently shown that exercise improves physical performance – including muscle strength and aerobic / functional capacity – in people with chronic health conditions, with no detrimental effect on disease progression.[62, 63] This is crucial in our ageing society where exercise therapy may result in reduced disability and an increased number of elderly people living independently.[62, 63]

Being physically active...

Figure 3.1
Health benefits of physical activity in the general population[58, 61, 73]
Physical activity plans should be tailored to each patient, be based on discussion between the patient and physician and, if possible, include the multidisciplinary team.

Benefits of physical activity in people with haemophilia throughout life

Although the benefits of physical activity are established in the general population (Figure 3.1), physical activity was not recommended for people with haemophilia until the 1970s.[1, 2] Thanks to the improved quality of life achieved by standard use of prophylactic therapy, the current standard of care is to recommend participation in certain low-risk physical activities, such as swimming, walking, cycling, rowing, table tennis, and weight training (for more details see Chapter 5).[1-4]

The rationale behind these recommendations is the observed benefits derived from leading an active lifestyle in people without haemophilia. Health advantages seen in the haemophilia population are similar to those described above for the general population. As mentioned earlier, physical activity helps to prevent chronic diseases such as heart disease and diabetes, and to promote normal neuromuscular development.[2, 4, 62] An extensive body of evidence in people with knee osteoarthritis has shown that exercise reduces pain, and improves physical function and QoL.[64]

Physical activity also has some specific positive effects on the challenges encountered most frequently by people with haemophilia:

- Physical activity improves strength, flexibility and endurance in people with haemophilia.[2] Through improving muscle strength around affected joints, being physically active can prevent muscle atrophy and may reduce the number of bleeding episodes, potentially protecting joints from haemophilic arthropathy.[1-3] Improving muscle strength, coordination and endurance is particularly important in people with haemophilia, as they experience reductions in these functions dependent on their joint status.[46, 65, 66] Greater muscle strength also helps to reduce joint pain, increase joint stability and reduce the risk of falls and injuries.[2, 3] The latter is likely to be increasingly important as the haemophilia population ages

- The need to rest a joint following a bleed and return to activities slowly can affect proprioception. As a result, along with muscle strength, proprioceptive function might be impaired in people with haemophilia[1, 37] To address this issue, sports therapy that is focused on proprioceptive performance might be used to improve muscle strength with minimal stress on the joints[67]

- People with haemophilia have a higher chance of lower bone mineral density (BMD) than the general population, and those with more severely affected joints and lower activity levels have lower BMD.[68] Undertaking appropriate weight-bearing exercises while young results in adequate BMD, which can protect from osteoporosis later in life.[1-3, 69] In the general population, improving peak bone mass and strength is an important strategy to reduce the risk of osteoporosis or low bone mass later in life. A recommended component of this strategy is optimisation of lifestyle choices, as they influence 20–40 % of adult peak bone mass[34]

- Another challenge in haemophilia care is the increasing rate of obesity and chronic health conditions among people with haemophilia, reflecting general worldwide trends. Being overweight can increase the risk of joint damage and regular exercise may reduce this risk by helping to achieve and maintain a healthy weight[1, 2]

- People with haemophilia frequently experience pain during their daily life,[66] which can potentially be reduced by physical exercise[70]

- Exercise exerts some beneficial effects not only through the physical aspects described above, but also on psychosocial aspects and quality of life. Sports activities enhance the feeling of social participation and inclusion, which is linked to improved self-esteem.[2, 71] Improvement of general quality of life was also observed, both in adults and children with haemophilia who participated in sports[2, 71]

- Studies have demonstrated that patients' self-assessment of their physical performance tends to be subjective. From a clinical practice perspective, physical performance in people with haemophilia should be examined with both objective (e.g. HEP-Test-Q) and subjective (self-assessment tools) measures[72]

Physical activity in people with haemophilia with inhibitors

Specific goals of rehabilitation and physical activity are similar across the entire haemophilia population; however, exercise therapy is particularly challenging in people with haemophilia with inhibitors as they may limit their physical activity due to the higher risk of recurring bleeding and arthropathy compared with people without inhibitors.[2, 74] Exercise programmes for people with haemophilia with inhibitors should be developed with specialist haematological and musculoskeletal input, carefully monitored once implemented, and have any adverse reactions addressed as soon as possible.[2, 74]

Contraindications to physical activity in people with haemophilia

Even though physical activity is recommended in haemophilia management, there are some important factors that should be considered when recommending specific activities. Some of the challenges for the haemophilia population include increased risk of injury, overloading of joints and potential bleeding.[2] With this in mind, children with haemophilia may restrict their physical activity due to their parents' concerns, and adults with haemophilia may also behave in a self-limiting manner.[2, 3] Other reasons for such behaviour may include musculoskeletal pain and deconditioning, particularly in older people with haemophilia who may have chronic arthropathy.[11] To address these challenges, a set of specific contraindications should be observed: high-risk sports including boxing, wrestling and hockey should not be recommended and any strenuous physical activities cannot be undertaken when not treated prophylactically.[4] Exercise or sports therapy should always be individualised (for more details see Chapter 5) and any sports or physical rehabilitation programme should be designed with the patient's haemophilia care team.

Did you know?

People with haemophilia should be advised to be physically active, as it offers a range of general-health and haemophilia-specific benefits.[1, 2, 71]

Summary: Physical activity targets in the general population and haemophilia outcomes improved by physical activity[1–3, 76]

Physical activity exerts multiple beneficial effects in the general population

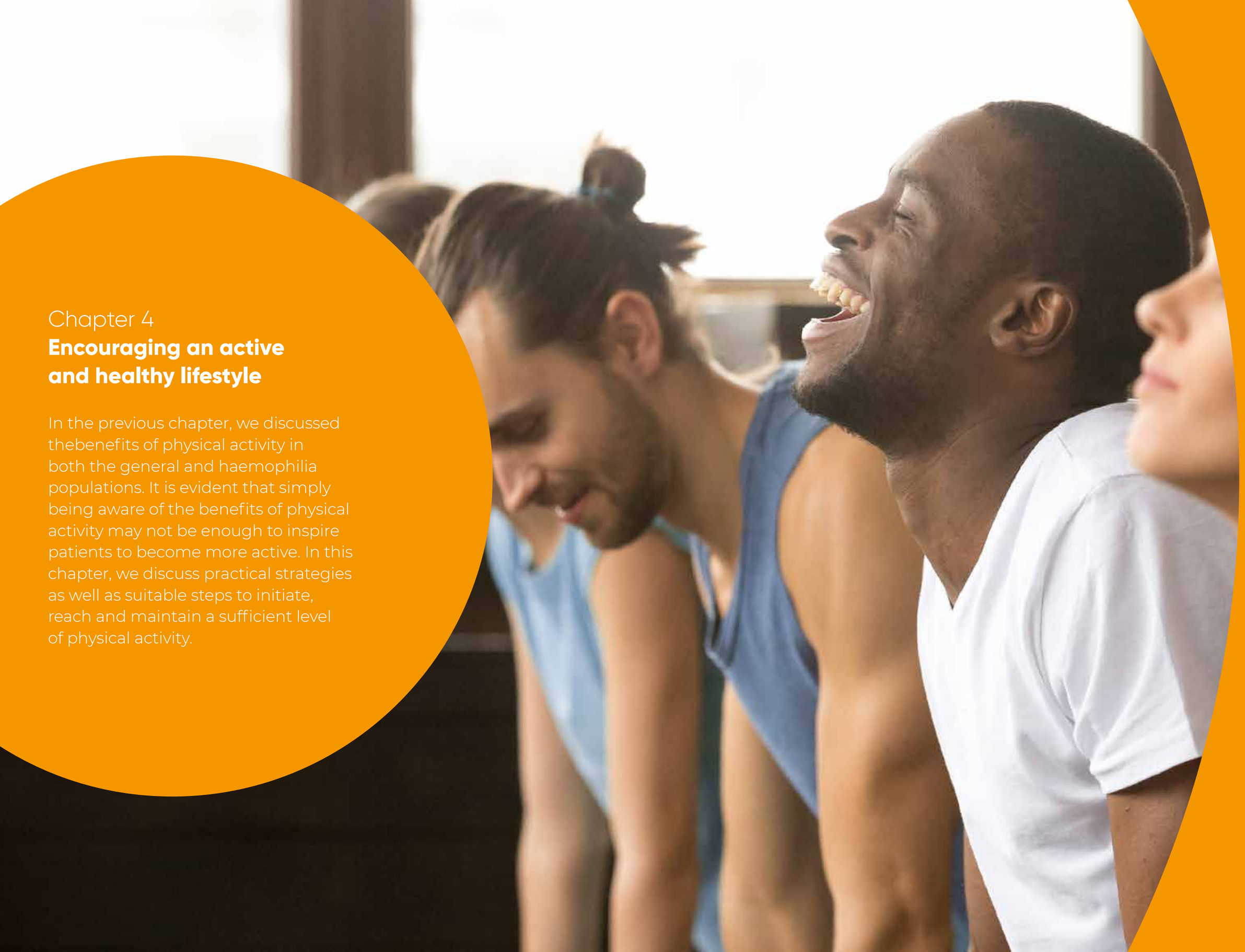
Physical targets in the general population

- ⊕ Muscle strength
- ⊕ Muscle tone
- ⊕ Proprioception
- ⊕ Range of movement of joints
- ⊕ Number of falls / injury risk
- ⊕ Lean body mass
- ⊕ Joint stability
- ⊕ Balance
- ⊕ Pain symptoms
- ⊕ Bone mineral density

People with haemophilia may also see improved outcomes for the specific challenges they encounter

People with haemophilia may also see improved outcomes

- ⊖ Haemarthrosis
- ⊖ Synovitis
- ⊖ Frequency of bleeding
- ⊖ Secondary osteoarthritis



Chapter 4

Encouraging an active and healthy lifestyle

In the previous chapter, we discussed thebenefits of physical activity in both the general and haemophilia populations. It is evident that simply being aware of the benefits of physical activity may not be enough to inspire patients to become more active. In this chapter, we discuss practical strategies as well as suitable steps to initiate, reach and maintain a sufficient level of physical activity.

How to address barriers to physical activity and get started?

Ideally, physical activity should be encouraged from childhood to help establish a positive attitude towards activity and the development of a lifelong habit of sport and / or participation in regular physical activity.[2, 77] When this is not possible, strategies to encourage the incorporation of exercise into the daily routine of people with haemophilia and promote adherence to an activity or exercise programme are recommended.[78] However, people with haemophilia, and those with severe haemophilia with inhibitors, may have adopted a more sedentary lifestyle to minimise bleeding risks, leading to a natural lack of motivation for exercise.[74, 79] Nevertheless, people with haemophilia can become more active and improve their physical performance when the benefits of sport and physical activity are successfully promoted.[2] Members of the multidisciplinary team may greatly help people with haemophilia to lead a healthier and active life by prescribing an appropriate duration, intensity and type of exercise.[4] But how can this be achieved, when there is so little time during consultations?[80]

Understanding the barriers to physical activity

The first step to motivate people with haemophilia to become more active is to fully understand the potential barriers to exercise.† For people with haemophilia these may include fear of bleeding and bleeding risk, concerns about management of bleeding, difficulty with exercise adherence and lack of motivation which may be influenced by the individual and their social environment.[2, 81] Physicians should be able to motivate individuals to open up about potential barriers and discuss how these can be addressed. Some individuals may also experience comorbidities (e.g. joint disease) that can limit their ability to engage in more intensive activities.† Although the level of physical activity may vary between adolescents and older adults, any activity counts: from a collective sport to mowing the lawn or gardening.[82] The following strategies to increase physical activity are a compilation of approaches applied to the general population and people with haemophilia, supported by clinical experience of haemophilia experts.

Measuring physical activity in routine consultations

Start asking your patients about the type(s) of physical activity they are involved in (e.g. running, gardening, swimming, etc.) and for how many minutes per week they participate.[81, 83] These questions let the individual know that exercise is an important part of health just like any other vital sign you may monitor, such as blood pressure or joint status.[81] Engaging in a physical activity can also make individuals feel more involved in their own treatment and encourage them to be more accountable for their own health.[84]

Promoting healthy behaviours with others

Behavioural interventions have been shown to be effective with diverse patient populations and in a variety of settings.[81, 85] Promoting healthy behaviours as targets (Figure 4.1) can be as simple as encouraging people to create triggers to become active during the day. For example, they may create a sign reminding them to use the stairs instead of the elevator or download a mobile application that sends push notifications reminding them to exercise at a convenient time. Suggest your patients encourage their partners, family members and / or friends to exercise together, e.g. a long walk every Sunday morning, attending a group class once a week, etc.† Setting up buddy systems between patients in your centre can also be an effective way to hold people accountable to each other to meet weekly activity goals together.[81]

Following a healthy diet can help people with haemophilia maintain a strong and well-nourished body.[86] Eating foods with saturated fat and / or added sugar will increase everyone's weight. Increased weight load in the joints, resulting from obesity and being overweight, can potentially increase the number of bleeds in people with haemophilia.[86, 87]

† Author Recommendation

Did you know?

Behaviour change occurs when motivation aligns with ability and triggers.[81]

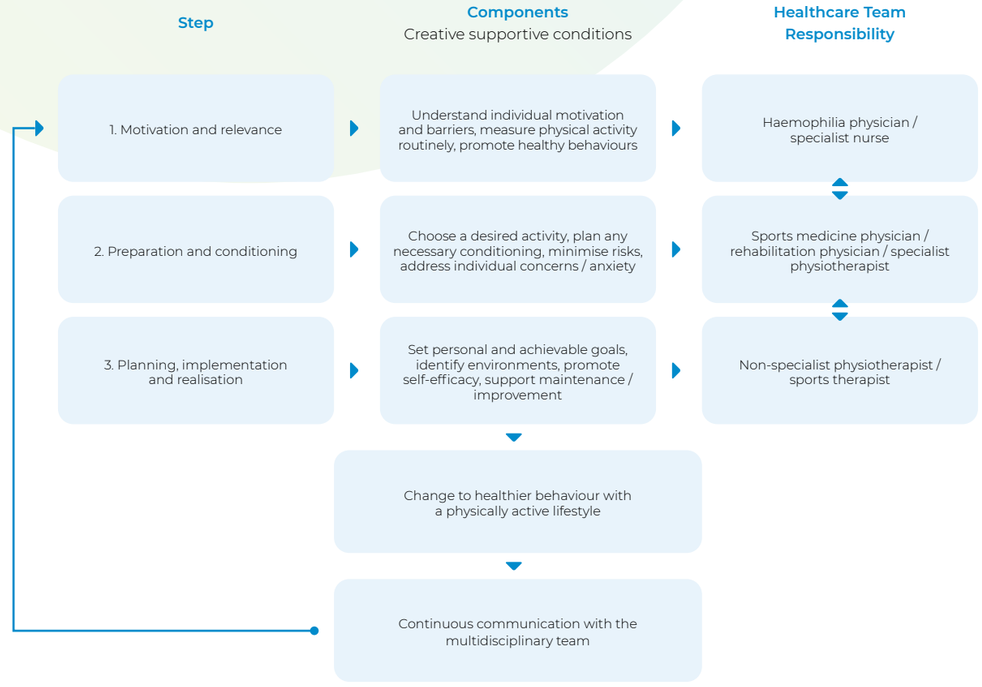


Figure 4.1
A step-wise model for encouraging a physically active lifestyle
The strategies to promote physical activity can be thought of as three main steps. These start with an understanding of the person with haemophilia's individual circumstances and motivation, and work through the realisation of a personalised treatment plan. Creating conditions that support physical activity – e.g. by sharing success stories and promoting multidisciplinary collaboration – will help to start the process. The endpoint should be the adoption by the person with haemophilia of a physically active lifestyle, with continuing reinforcement to ensure this is maintained. Different healthcare professionals have different, although overlapping, responsibilities at each step. Figure © 2019 T Hilberg

National recommendations on healthy nutrition can be an important tool to educate people with haemophilia and importantly, their families on how to live a healthier life.†

Choosing the activity and setting up realistic and meaningful goals

When selecting a suitable physical activity for people with haemophilia, consideration of the patient's preferences is essential.[2] Chapter 5 will be examining how to individualise an activity programme for people with haemophilia. Make sure to not propose a physical activity that the patient is reluctant to attempt. † If the patient is not immediately capable of starting their desired sport / activity – e.g. if they have mobility problems in a particular joint or a general lack of fitness – consider targeted physiotherapy / sports therapy to get them into a condition where the sport / activity is possible.†

The selection of activity is important but focusing on the result being achieved instead of the task / activity at hand can be as important.† Visualising the goal being reached can be a helpful way to enable its achievement. This can be easily implemented by creating a map and timeline of the goals to be achieved and make it visible on the daily basis (e.g. display the map on the fridge or any other surface / place the patient passes by every day) or by using a counting steps mobile application with daily goals. † The patient can also, with the support of the multidisciplinary team, put in writing the goals he wants to achieve as well as define key milestones.† Monitoring these can be helpful to identify if the patient is not reaching personal exercise goals (previously agreed with the treatment team) and support the multidisciplinary team to start a conversation about the importance of physical activity in the patient's overall health and adapt the exercise programme.[81]

Identifying suitable environments together

The multidisciplinary team and people with haemophilia should work together to identify environments where it is easier to become physically active.[81] This may include a family being active together or identifying a school-based or employer-based activity that fits the needs of each individual. Education on where to find suitable locations will help adults to be more physically active.[81] Sports therapy camps can be an ideal environment as they are supervised by sports professionals, camp counsellors and medical teams trained in haemophilia.[46] The camps can include theoretical as well as practical training such as endurance, strength and coordination as well as water-based activities, and others.[46] Practical aspects can include ways to discreetly modify / adapt activities to reduce risk.† Moreover, regular consultations with experienced specialist physiotherapists, sports therapists or rehabilitation physicians make it possible to supervise any changes to joint status, evaluate the risks and benefits associated with physical activities and (re)design / modify a physical fitness plan with the involvement of the patient.†

Promoting self-efficacy to build confidence in physical activity

People with haemophilia who understand that physical activity is important but do not have the confidence to perform it should be encouraged to enhance self-efficacy.[85] In this context, 'self-efficacy' refers to an individual's innate ability and belief to achieve goals. Self-efficacy can not only build the confidence of people with haemophilia but also encourage them to perform specific physical activities in particular environments.[85]

Possible approaches include:[85]

- Discuss activity-related challenges depending on the ability and successful experiences of the individual
- Share successful experiences with other people with haemophilia
- Praise the successful experience of the individual
- Raise awareness of improvement in physical function to the patient by comparing previous and present physical activity levels
- Provide a continuous cycle of feedback on physical activity levels†

† Author Recommendation

How to maintain / improve physical activity levels?

People with haemophilia may find it challenging to maintain participation in physical activities for several reasons, including consequences of bleeding episodes or the fear of bleeding.[2, 85, 88] Studies suggest that self-monitoring can be a valuable way to strengthen self-efficacy and keep people with haemophilia active.[79, 85] It can enable individuals to appreciate the benefits of physical activity by noticing the differences in their bodies, creating self-efficacy and improving exercise adherence.[85] Self-monitoring may involve tracking bleeding episodes, use of coagulation factor, physical activity and frequency of home exercise.[85] It may also include monitoring the goals and milestones previously agreed with the involvement of the patient and the multidisciplinary team.[85] These may be monitored digitally through mobile applications or in a more traditional way through a diary / notebook. Celebrating successes can be highly valuable to the patient and provide a reason to keep going.

If a patient is losing interest in physical activity, it is important to discuss with them the key factors influencing this. For some patients, starting with a coach / personal trainer who understands their lifestyle and has a close relationship with members of the multidisciplinary healthcare team can be helpful to support the individual's physical-activity journey.† Some people with haemophilia may prefer to start with a coach to build confidence before enrolling in a sport, club on gym membership, but the coach should be adequately briefed by the multidisciplinary team supporting that patient.†

Nevertheless, it is important to be realistic and assess the psychological and physical state of each individual following a bleeding episode before returning to daily activities, exercise and sports.†

How to minimise risks during physical activity

To minimise the risks associated with physical activity, people with haemophilia should consult with a musculoskeletal professional to discuss the appropriateness, protective gear, prophylaxis and physical activity skills required prior to beginning a certain activity.[4] This is particularly important for those with any problem / target joints that can be protected with braces or splints during the activity.[4] Where possible, discreet modifications / adaptations to reduce the risk of a particular activity are preferred.† Moreover, regular consultations with experienced specialist physiotherapists, sports therapists or rehabilitation physicians make it possible to supervise any changes to joint status, evaluate the risks and benefits associated with physical activities and (re)design / modify a physical fitness plan with the involvement of the patient.†

Identifying when patients usually engage in physical activity can be important from a treatment perspective. For example, a patient who goes swimming on two or three weekdays but is inactive during the weekend will require a different treatment regimen to a patient who is active only on the weekends. A tailored treatment regimen that considers the timing of physical activity (when peak clotting factor levels may be needed) may help minimise risks during physical activity.[89]

The next chapter will cover strategies to identify an adequate physical activity for people with haemophilia and how to individualise an activity programme.

† Author Recommendation

Summary: Encouraging an active and healthy lifestyle for people with haemophilia

Promote healthy lifestyle andbenefit of excercise[81]

Engage family support and 'body' networks[81]

Identify and overcome barriers to activity[2]

Set up realistic and meaningful goals[2]

Build confidence in physical activity[82]

Measure and monitor physical activity[61, 81, 84]

Give strategies to maintain physical activity[79,84]

Provide positive reinforcement

Identify suitable environments[81]

Chapter 5

Individualised activity programmes

While the benefits of physical activity in people with haemophilia have been discussed in Chapter 3, in this chapter we discuss which considerations are required to design an ideal individualised physical activity programme in this population.

Physical activity recommendations for the general population

Physical activity can be defined as any movement in the body produced by skeletal muscles that requires energy expenditure above resting level. Walking, cycling, sports and recreational activities (e.g. gardening, dancing, etc.) can be considered physical activity and accomplished at any level of skill.[58] The response to physical activity is influenced by the type of exercise and the "exercise dose", which encompasses three factors:[84]

- Frequency (how often the person exercises)
- Duration (how long is spent on the activity)
- Intensity (how hard / intense the session is)

The WHO recommends that adults (18–64 years) participate in at least 150 minutes (Figure 5.1) of moderate-intensity (e.g., brisk walking) or at least 75 minutes of vigorous-intensity (e.g., running) aerobic physical activity throughout the week (or an equivalent combination of the two), and perform muscle-strengthening activities on two or more days a week.[61]

Age Group	Recommended amount of physical activity
5–17	**Daily** 60 minutes of moderate-to-vigorous-intensity activity
18–64	**Weekly** 75 minutes of vigorous activity **OR** 150 minutes of moderately intense activity
>65	**Weekly** The same as above, but depending on what their abilities and conditions allow
More physical activity is likely to bring additional health benefits	

Figure 5.1
Global recommendations on physical activity for health[61]
Physical activity plans should be tailored to each patient, be based on discussion between the patient and physician and, if possible, include the multidisciplinary team.

† Author Recommendation

Did you know?

Activity intensity should be at least moderate which means that one is able to talk, but not to sing – heart rate and breathing will be increased.[84, 90]

What is an ideal physical activity programme for people with haemophilia?

When putting together an activity programme for a person with haemophilia, a comprehensive plan is needed, taking into account an individual's current physical ability, risk factors, joint status, aspirations and goals (Figure 5.2).[2] A physical activity plan is unlikely to succeed if the patient does not feel motivated or capable of achieving the targets set for them. With this in mind, a complete physical examination – with internal and orthopaedic / sports medicine aspects – should be carried out by appropriate medical staff.† If the patient is found to be unfit for a desired sport / activity, consider targeted physiotherapy / sports therapy to overcome physical barriers to participation in the sport / activity[2] (e.g. by addressing mobility problems in a particular joint). One practical approach to encourage participation is to design a structured programme of activities that is initiated with a high level of input from a sports therapist after which the patient is encouraged to manage their own training.[46]

Physical activity is increasingly being viewed as more than just conventional exercise, but instead includes everything from sport or planned exercise to recreational or leisure-time activities, walking or cycling as a means of transport, occupational activity (i.e., work), household chores, play and games.[58] This is of particular relevance to people with haemophilia, as their choice of physical activities can be limited by their condition.[2] Ideally the physical activity should be incorporated into a patient's lifestyle or already be part of the person's daily routine.[58] In general, adults are more likely to take up and continue with a physical activity if they are comfortable with it and find it enjoyable. Other motivating factors include a sense of achievement and skill development, health benefits, and the presence of support networks.[91] For more information on how to encourage people with haemophilia to lead a healthier and active life, see Chapter 4.

As for the general population, the main aims of a physical activity plan should be to improve cardiorespiratory and muscular fitness, improve bone / joint health, reduce the risk of diabetes and obesity, and improve quality of life (QoL).[46, 61] However, risk factors that specifically affect people with haemophilia also need to be considered.[1, 2, 46] The benefits of physical activity in people with haemophilia can be found in Chapter 3. These considerations will inform the types of activity that people with haemophilia will be able to perform – and which to avoid – while others may also contribute to the key goals of the programme.

Components of a physical activity programme for people with haemophilia

To optimise benefit, a physical activity programme aimed at people with haemophilia should combine body awareness, strength, balance / coordination, flexibility and endurance.[46] The intensity and duration will be determined by the patient's existing level of fitness and the presence of comorbidities, but should be similar to the recommendations for the general population.[2, 61, 92] It should be possible to perform all parts of the exercise programme without pain, and the patient should be reassessed regularly to allow necessary modifications to be made.[2]

Body awareness

Training of body awareness – and thus the optimisation of body perception and movement awareness – is often the most effective entry into physical activity for people with haemophilia. This training can facilitate the learning and practice of motor exercises.†

Muscle function and strength training

Both strength and endurance exercise can lead to significant health benefits.[61, 92] Conditioning to improve strength, flexibility and endurance is important in order to gain the appropriate level of fitness for a person with haemophilia to safely participate in a physical activity or sport.[77] Strength training has been recognised as a contributor to physical health in the general population,[61] and is important in people with haemophilia as it may prevent the progression of joint disease.[46] Increased strength improves work capacity and resistance of the muscles, bones, tendons and ligaments, reducing the risk of injury.[2]

"To safely build muscle strength, the resistance must be set appropriately: the higher the resistance, the bigger the increase in absolute muscle strength but the higher the risk of injury. Thus, to have a significant impact, the weight load should be sufficient but not so great that it puts the individual at risk of a bleed or unnecessarily burdens the joints."†

Strength training for the elderly in the general population can help maintain physical independence by mitigating or preventing functional limitations, as well as reducing the risk of falls and fractures.[93] For persons who have been physically inactive and / or bedridden due to Illness, and who have thereby lost both strength and aerobic fitness, it is especially important to train for strength to manage the strain they meet in their daily lives.[84] In people with haemophilia, the immobilisation and restrictions to activity required after a joint bleed can have significant consequences on muscle strength, particularly if the immobilisation is prolonged.[46]

Furthermore, improved joint and core muscle strength in people with haemophilia may help control exaggerated end-range of motion joint movements, thus potentially helping to prevent or decrease synovial impingement and associated haemarthroses or synovitis.[94]

When initiating a strength training programme, it is important to allow time for the adaptation of the musculoskeletal system and to practise good technique. The initial resistance should be set at a level that allows the participant to achieve the prescribed number of repetitions (10–15) without straining.[95] The training programme should involve the major muscle groups and alternate between upper- and lower-body work.[92, 95] In particular, people with haemophilia should focus on the abdominal and back muscles, and the muscles of the lower extremities.[2] As an individual improves, their training plan should reflect this to allow for continued improvements in muscular strength and endurance. An initial increase in the maximum number of repetitions is recommended before an increase in weight load. When the patient is able to achieve the upper limit of the repetition range, the load or the number of repetitions can be increased.[92, 94]

Progressive strength training often requires access to weights or various machines and apparatus that allow for adjustable resistance,[92] which is why training is best performed at a specially equipped gym with supervision by an experienced therapist.† Strength training can be conducted at home with tools such as small dumbbells or elastic exercise bands, or with the individual's own body load.† For elderly or disabled individuals, training can be carried out in the form of various functional elements, such as rising from a chair and climbing stairs.[84]

Coordination and flexibility

Balance and coordination / proprioception training are important to avoid sprains, reduce the risk of injury and improve physical performance when participating in physical activity as well as for activities of daily living. Coordination is important for everyone, but particularly for people with haemophilia who may have severe arthropathy in multiple joints, increased joint instability and / or balance dysfunction, all of which increase the risk of falls. Furthermore, falls are more likely to result in serious injury and fractures in people with haemophilia due to osteoporosis and osteopenia.[2] Proprioception exercises may be needed in preparation for a physical activity programme.†

Flexibility is defined as the joints' ability to maintain a range of motion, which is dependent on the function of the joint and the surrounding structures (muscles, tendons and ligaments).[84] To develop and maintain a good range of motion and prevent joint injury, it is important to include flexibility exercises in a physical activity programme. Flexibility exercises should be carried out two to three times a week and should stretch the major muscle groups.[96] Stretching as part of a warm-up and cool-down period is also advised for people with haemophilia undertaking a physical activity.[2, 77]

Endurance

From a physiological perspective, endurance is called either aerobic or anaerobic, depending on which form of metabolism is dominant. Endurance training improves cardiorespiratory fitness as well as metabolism. Depending on the intensity of training, muscle strength can also be influenced.[97, 98] Physical activity such as walking in hilly terrain often comprises both aerobic and anaerobic activities, again depending on the intensity.[84] If performed for a sufficient duration, endurance exercise stimulates the adaptation of the heart and the aerobic and anaerobic systems of the skeletal muscles.[84, 98] For people with haemophilia, endurance exercise should ideally be varied, attractive, progressive and individualised.[2] Moderate-intensity activities such as swimming, cycling and brisk walking, are recommended to improve endurance fitness and strengthen muscles without a high risk of joint damage.[77] Tools, such as the Borg scale,[99] can be used to assess the physical activity intensity level.[99]

"Fitness coaches should be advised of the individual's condition and limitations, and if possible, a member of the care team should be involved in such discussions."†

Role of sports therapy[46]

All the components discussed above (e.g. body awareness, strength, balance, etc.) should be considered when designing a physical activity programme aimed at people with haemophilia. These will inform decisions about the intensity and duration of this programme. Dependent on their individual limitations and health status, individuals should be encouraged to take part in organised sports therapy training supervised by a competent physiotherapist and / or sports therapist, perhaps as part of a sports therapy camp. Examples of this kind of camp are published in Hilberg 2018.[46]

† Author Recommendation

Important considerations when designing a physical activity programme for people with haemophilia

Considerations for an individual activity programme

- Age
- Aerobic fitness
- Bleeding risk
- Bone mineral density
- Cardiovascular complications
- Inhibitor status
- Joint damage
- Muscle strength
- Osteoarthritis
- Pain
- Proprioception
- Risk of falls
- Severity of disease
- Surgical rehabilitation
- Treatment regimen and adherence

Figure 5.2
Many aspects of the patient's physical condition should be taken into account when designing an individualised activity programme[1, 2, 46]

Treatment regimen

Thanks to advances in treatment, people with haemophilia receiving prophylactic therapy should be able to carry out tailored physical activity programmes. As prophylactic treatment is normally administered every few days, it is recommended to match the days of treatment with the days of activity.[2, 77] Although prevention of bleeding can be achieved by a prophylactic factor-replacement regimen,[4] it is also possible for patients receiving on-demand treatment to participate in physical activity. In this case, a factor infusion should ideally be given in the morning of the day that the physical activity is due to take place. Wearing appropriate footwear or orthotics can be a cost-effective approach to reducing the clinical manifestations of joint bleeds.[2] Although physical activity in patients with inhibitors remains a challenge, an exercise or sports therapy programme can be tailored to the individual patient and gradually extended in time and frequency.[2, 74] However, it is worth noting that compliance with treatment can decrease in some age groups (e.g. adolescents) and the need to adhere to treatment recommendations should be emphasised prior to the recommendation of a physical activity programme.[2]

Monitoring

Monitoring joint health is essential for identifying early signs of deterioration, allow adjustment of clotting factor replacement therapy, physical therapy, the current physical exercise plan, and the use of mobility aids to limit further decline.[100] The Haemophilia Joint Health Score (HJHS) and Haemophilia Early Arthropathy Detection with Ultrasound (HEAD-US) can be used as a tools for routine assessment of joint health and can be used by trained healthcare professionals.[4, 15, 100, 101] Monitoring progress and achievements allows for adaptation of physical activity programme and ensures goals are met.‡ People with haemophilia can monitor their physical activity progress using tools such as mobile apps or wearable fitness trackers.[102] Several validated questionnaires are available to monitor physical activity in people with haemophilia.[103, 104]

Risk–benefit profile of physical activity for individual patients

A report from the literature suggests that people with haemophilia, particularly those on long-term prophylaxis or presenting a mild / moderate bleeding phenotype, are as active as their healthy peers.[2] Although people with haemophilia experience the same benefits of exercise as the general population, the risks vary between patients therefore activities need to be chosen accordingly.[2] When selecting an activity for a person with haemophilia one needs to consider physical ability, aspirations and goals.[2] This will allow for a complete understanding of the risks involved and their management, translating into the greatest gain in QoL for each patient.[2] If a patient is interested in participating in an organised sport, the multidisciplinary team (particularly the sports therapists, when available) can be involved in the decision-making process to ensure safe selection of the most appropriate physical activity.†

Response to exercise training

Multiple factors influence how much a person improves when they increase their level of physical activity. For example, a person who is inactive or has a very low level of fitness will improve more in relative terms than an individual with a higher level of fitness. For someone who already has a good level of fitness, maintaining that level is also a successful outcome. Although some effects can be seen after just a few weeks, the effects of physical activity are considerably greater if training continues for several months to years.[84, 98] Therefore, it is important to include physical training in the daily routine of people with haemophilia throughout their lifetime, whatever their baseline level of fitness.‡

Home exercise planning

Some patients may prefer to exercise at home. Home exercise should be recommended by a physiotherapist or sports therapist and tailored to the physical condition of each patient.† If possible, the recommended exercises should be performed in the clinic and / or a sports therapy camp ahead of implementing a home exercise routine to ensure they are performed correctly.† For effective home exercise, behavioural changes and improvements in exercise adherence are required.[79] Self-monitoring techniques have proven efficacious in different patient populations, including in haemophilia.[79, 105]

Participation in competitive sports[77]

Competitive group activity can offer many positives, including learning the value of teamwork and getting recognition for individual accomplishments increasing one's self-esteem. However, in some cases there may be cumulative stress to joints and an increased risk of bleeding. Sports should be considered on an individual basis and take into account the desired effect, the way they are played, pace and duration, intensity, stresses on the joints, and above all, the condition of the patient's joints. Appropriate safety equipment should be used and coaching guidelines followed. Input from a specialist sports therapist may be helpful in informing both the choice of sport and any necessary preparatory exercises.

"Aim to follow-up with your patient's progress with the agreed activity programme at their usual consultation. Do not be afraid to create a new individualised plan if things are going better or worse than expected"†

† Author Recommendation; ‡ Authors' Clinical Expertise

"**Where appropriate, people with haemophilia should be encouraged to continue the activities that they enjoyed during childhood and adolescence.**"†

Physical activity after the transition to adult care

Monitoring of and education around physical activity is a key part of comprehensive care in children with haemophilia, and knowledge of a healthy lifestyle is considered a factor in a patient's readiness for transition.[40, 106] However, once into adulthood, the focus moves to management of joint disease, treatment decision making and challenges associated with treatment adherence and self-care.[40, 43] With so many important considerations at the time of transition, it is not surprising that discussions around the continued importance of activity throughout adulthood are not prioritised.‡ As many young adults discontinue prophylactic therapy after the transition,[107] it is important to tailor individual healthcare programmes around their lifestyles including planned physical activities and jobs. Therefore, it is important that the benefits of physical activity are communicated to young adults.†

Did you know?

Activities that may have been considered appropriate in the past may become more dangerous as your patients age.[77]

Type of activity (recommended vs contraindicated)

Although it is important to consider the patients' preferences for certain physical activities, discussions on safe exercise are essential for people with haemophilia. Suitable alternative activities and appropriate preventative measures can be recommended with the patients' interests in mind.[1] Table 5.1 outlines the pros and cons and haemophilia-specific considerations for a number of popular sports.

† Author Recommendation; ‡ Authors' Clinical Expertise

Summary: Tips and advice for safe exercising for people with haemophilia

These tips and advice should only be seen as recommendations that need to be individualised to each patient.

Key steps for safe exercise†

1. Ensure shared decision-making when selecting an activity and sport for your patient
   This should involve the risk-benefit balance, reason for choosing a sport (e.g. becoming active, increase social interaction, etc.), patient interests and goals, treatment regimen and joint health status
2. Encourage your patient to follow the programme agreed with the multidisciplinary team, including adherence to treatment
3. Decide whether your patient should slow down or stop exercising in case any problems are encountered (e.g. pain)
4. Educate your patient on how to identify and manage bleeds while being active
   Timing of infusion before activity and preventive measures in case of accident should be considered
5. Agree goals with the patient and make sure to monitor progress on a regular basis

Identifying and managing joint bleeds while being active

Despite the health benefits, being physically active will always carry a certain degree of risk for people with haemophilia. Although joint bleeds can be difficult to recognise, people with haemophilia usually recognise early symptoms even before the manifestation of the physical signs, which may include:[4, 108]

- Gradual joint swelling
- Tingling
- Joint heaviness
- Gradual discomfort
- Reduced range of motion
- Acute pain in the muscle or joint

Although pain can be considered a sign of a joint bleed, the type of pain[108] (types of pain are covered in more detail on Chapter 6) needs to be considered in order to differentiate between a joint bleed and an acute flare-up of arthropathic joint pain.[108] If the pain occurs after inactivity, or increases after prolonged activity, this may indicate a flare-up of haemophilia arthropathy.[108] However, if the patient is feeling local pain (vague or tingling sensation) and then this spreads as a pressing pain over the entire joint, there is a chance that this pain can be linked to a joint bleed. If it is a bleed, clotting factor replacement or other therapies as recommended by the patient's physician should be used† [108]

A combination of replacement therapy and protection, Rest, Ice, Compression and Elevation (R.I.C.E.) of the joint may also be helpful:

- **Protection** – protect the injured joint from undue stress, perhaps by using a splint or crutches[4]
- **Rest** – rest the joint[4] for at least 24 hours or longer.‡ – take advice from a healthcare professional
- **Ice** – applying ice (covered with a towel to prevent frostbite) can help ease pain[4, 23]
- **Compression** – wrap the injured joint in a bandage or compression stocking for the first 24 hours. Check every 2 hours to make sure the wrap is not too tight.‡ Compression helps to control the amount of swelling and may lead to a quicker recovery[4, 23]
- **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[4, 23]

After a bleed, physical therapy can help the recovery of joint function. It is essential to monitor the joint throughout the recovery process.[35] Once recovered, the patient may gradually start a sport or sports therapy that is appropriate for their condition, in order to build up long-term joint health.†

Table 5.1: Considerations for choosing appropriate sports

This table includes a selection of sports that can be considered for most adults with haemophilia provided that individual factors such as joint status and treatment regimen are addressed (see 'colour coding'). The list is not intended to be comprehensive, and deliberately excludes sports which are discouraged for people with haemophilia (these are reviewed extensively in other publications).[77, 109] The sports listed below, along with the pros and cons and 'considerations for people with haemophilia', have been selected based on published literature and the authors' experience. The risks of undertaking any sport depend not only on the sport itself but also on individual situations and experience.[2] They should be considered alongside programmed sports therapy or other interventions to address the individual's needs. Before undertaking any sport, the person with haemophilia should consult their healthcare team / musculoskeletal professional and always follow their directions, particularly as they relate to the timing of doses of clotting factor treatment and any necessary precautions.

Colour coding:

- Activities that generally have very low risks of bleeding
- Activities that generally have low risk of bleeding
- Activities that generally have a moderate to high risk of bleeding

Determination of 'risk' will depend in part on patient-specific characteristics including the treatment regimen and timing of treatment, severity of haemophilia and bleeding phenotype, and joint status.[2]

Sporting activity	Pros	Cons	Considerations for the person with haemophilia
Aquatic exercise, including swimming	• Provides aerobic exercise while reducing weight-bearing on weight-bearing joints for easier movement[77, 78] • May help people to get started with a physical therapy programme[78]	• Risk of repetitive strain / overexertion injuries[77]	• Proper technique is important for avoiding repetitive strain injury[77] • Devices can be used to increase resistance for a more challenging workout[77] or to improve buoyancy[78] • Diving should be avoided[109]
Dance-based exercise[110]	• Social aspects‡ • Provides an alternative option e.g. for people who are not interested in organised sports‡	• Risk of injury from falls or twisting, especially in those with existing joint damage‡	• To be done with supervision from an instructor (e.g. in the context of a dance class)[77] • Consideration of the style of dance is important: ballroom may be more suitable than street dance‡
Rowing	• Provides an excellent total-body, non-impact aerobic workout[77] • Rowing machines offer the benefit of an all over workout with little impact on the joints and can strengthen arm, back, shoulder and abdominal muscles[77] • Can row alone or in groups‡	• Risk of strain injury to the knees and lower back[77]	• Wearing a life jacket when rowing in open water‡ • Modification of movements to accommodate joints that lack full range of motion can be done‡ • Proper position, selection of an appropriate range of motion, and use of the back is important to prevent injury[77]

‡ Authors' Clinical Expertise

Sporting activity	Pros	Cons	Considerations for the person with haemophilia
Walking Hiking	• Can be performed in a variety of locations[78] • Can be done individually or in a group, and throughout the lifespan[77] • Aerobic effort, impact and muscular workout increase with more challenging terrain[77]	• May be challenging for those with joint damage‡ • Risk of falls for those with balance problems[78]	• Appropriate boots or shoes should be worn[77] • Walking indoor or on a treadmill reduces the likelihood of falls due to uneven terrain[78] • Braces, orthotics and assistive devices (e.g. walking sticks or poles) may be needed to provide support and stability[78] • In order to obtain sufficient energy expenditure, walking must be done at a good pace and for a sufficiently long time‡
Golf	• Low-impact, lifetime sport that provides a good workout for those who walk the course rather than use a cart[77] • Social aspects‡	• Expense and access to a course are limiting factors[77]	• Proper physical conditioning beforehand can minimise risks[114] • Joint status (especially the elbows) should always be evaluated when considering golf as an activity*
Road cycling	• Provides aerobic exercise with minimal joint impact[77] • Offers opportunities for participation (e.g. in cycling clubs)‡	• May be challenging for individuals with joint damage (especially knees or ankles). Elbows may experience increased pressure‡ [7] • Risk of falls and collisions with obstacles‡ [77]	• When outdoors, cycling on roads or cycle paths as well as wearing proper safety gear, including a helmet (this is mandatory)[78] • Proper adjustments to the bike and seat at height[109] • Individuals with limited knee range of motion may need to adjust seat height or use a recumbent bike to increase comfort[78] • Mountain biking especially downhill is not recommended as it can carry moderate to dangerous risk[77] • Cycling on a stationary bike at home or in a gym is an alternative to road cycling‡
Skiing (Cross-country) Skiing (Downhill)	• Cross country skiing is an excellent aerobic activity that can be performed by all ages[77]	• The difficulty of the course will directly affect the degree of stress on joints and overall risk such as risk of falling[109] • Risk of collision from other skiers cannot be controlled‡	• Wearing a helmet and suitable boots, as well as using proper length skis and poles[109] • Recommendations include to ski in relaxed and safe conditions‡ • Ski jumps and moguls are not recommended[77] • Downhill skiing is considered high-risk because the chance of head injury and traumatic bleed is high from falls and collisions[109]; it is not recommended for people with haemophilia
Yoga	• Can improve balance, posture and flexibility, and reduce stress[78, 113] • Can be done at home (using video or online tutorials) or in a group class[77]	• May be challenging for people with multiple joint involvement[78] • Weight-bearing on one leg may be difficult and painful[78]	• Proper technique is important to avoid injury; for example, special attention to alignment to prevent over-stretching is needed[77, 113] • Modifications to the exercise routine may be done to and / or gradually progressed over time to accommodate individual challenges e.g. with joint flexibility[78] • Ideally, yoga should be combined with strength training‡

* Recommendations by authors based on clinical experience; ‡ Authors' Clinical Expertise

Sporting activity	Pros	Cons	Considerations for the person with haemophilia
Indoor climbing	• Offers a full-body workout[112] • Strengthens the muscles of the lower and upper limbs in a secure way because muscle contractions are programmed and progressive‡ • As it is an individual sport, the person can immediately stop in case of pain or other problem‡	• Risk of falls / chronic repetitive injuries[77, 112] • Requires two to practise the activity because the other one must hold the harness / ropes[111]	• Attention to safety and climbing with appropriate equipment, including harness, ropes and climbing shoes[77, 112] • Proper supervision[77, 112] and recommendation to climb in relaxed and safe conditions‡ • Outdoor climbing is high-risk for people with haemophilia[77, 112] and is not recommended
Running	• Can be done alone or in groups‡ • Good cardiovascular exercise‡	• High impact to weight-bearing joints, may increase the number of bleeds and contribute to severity of joint disease[77] • Risk of falls / injuries depending on the running surface (e.g. concrete or uneven ground) and intensity of running (such as distance, speed, frequency)[109]	• Joint status (especially of the ankle) should always be evaluated by a specialist before considering running as an option, as existing damage may be exacerbated‡ • Wearing appropriate footwear (need good shock absorption, firm heel-counter, arch support)[109]
Weight training (in a fitness centre)	• Can improve strength, bone strength and even boost mood[111] • May result in weight loss,[111] improved body image and self-confidence[77] • Fitness centres are generally accessible, so that sessions can be scheduled‡ • Little risk of injury if exercises are performed properly and patients have good control of the range of motion‡	• Over exertion may cause joint or muscle bleeds‡ • Risk of injury from weights or other equipment if devices are not used properly or if the programme is not well adapted to the patient[111]	• Requires proper technique and regular training[109] • Collaboration with a fitness trainer who is aware of the limitations of haemophilia‡ • Power lifting (where the goal is to lift the heaviest weight possible) is not recommended for people with haemophilia[77]

‡ Authors' Clinical Expertise

Chapter 6

Pain and physical activity

In this chapter, we provide a brief introduction to pain in people with haemophilia and how physical activity can play a role in its management. Although guidance on assessing and treating pain is important and should be considered in the management of people with haemophilia, this is out of scope of the current booklet. For more information on pain-related treatment options please refer to the WFH Guidelines for the Management of Haemophilia.[4]

Understanding pain management and pain mechanisms

Pain can be defined as an unpleasant sensory and emotional experience associated with actual or potential tissue damage.[115, 116] People with haemophilia often experience pain and functional impairment resulting from acute and chronic manifestations of the disease (haemarthrosis / arthropathy).[20, 116]

Acute pain in haemophilia

In haemophilia, acute pain is often caused by bleeding episodes in the joints and muscles. However, pain is not always a reliable indicator of acute bleeding.[51, 116] People with haemophilia, as with the general population, may suffer injuries that can also cause acute pain. Moreover, healthcare professionals and patients with advanced arthropathy often find it difficult to distinguish between pain caused by an acute bleed, inflammation or joint damage, and chronic arthritic pain.[116, 117] If activity-related pain improves on resting, then we might be looking at arthritic pain and not bleed related.†

Chronic pain in haemophilia

In haemophilia, chronic pain is characterised as being long-lasting and recurrent, resulting from arthropathy and / or other long-term complications in haemophilia (i.e. synovitis and arthritis).[53, 117-119] A recent study in people with haemophilia suggested that structural changes such as synovial tissue alterations as well as the presence of osteophytes, may enhance pain sensitivity.[120] Unlike acute pain, chronic pain is associated with neurobiological, psychological and social changes that can maintain pain, making it more complicated than acute pain.[119] Assessing these different pain mechanisms is important to avoid insufficient pain treatment.[121]

Impact of pain in people with haemophilia

Studies have shown that pain related to haemophilia is associated with reduced health-related quality of life. Chronic pain is associated with an increased incidence of depression, anxiety, irritability, anger and frustration.[51] When left untreated, pain can lead to patients seeking pain relief in risky behaviours including excessive alcohol intake and narcotics.[53] Pain is an individual experience[51] that varies based on emotional and[115] social factors and physical sensitivity, as well as the strategies used to deal with pain.[51, 115]

When in pain, patients may modify their movements which can lead to further problems in the long term.*[120] As an example, if a patient is feeling pain in the ankle then he might start walking differently, perhaps taking the weight off walking on that particular joint. This change in movement could lead to further problems, including bleeds, due to the decreased shock absorption.[120, 124] With this in mind, it is essential to know which tissue is causing pain. Identifying these will enable physicians to tailor the type and intensity of physical activities for each patient.† [124]

*For further reading on the peripheral and central changes induced by pain please refer to Bank PJM, et al. Eur J Pain 2013;17:145–157.125

Did you know?

The term "flare-up" is commonly used and refers to pain symptoms related to haemophilic arthropathy. Acute pain does not always need to be bleed-related.[116]

† Author Recommendation

Pain management

It is vital to stress to patients and their families that pain can be managed and that patients should not suffer in silence.[51]

Although pain is briefly addressed as part of the Guidelines for the Management of Haemophilia (WFH),[4] there are no specific guidelines for pain management in haemophilia. Nevertheless, guidelines and precautions from the general pain literature are relevant and can be applied to the haemophilia population (Table 6.1).[126] A comprehensive approach to pain management should consider the involvement of all members of the multidisciplinary healthcare team,[127] including haematologists, orthopaedic surgeons, physical therapists, nurses, psychologists, counsellors, pain specialists and pharmacists.[52, 127] Selection of pain treatment should be tailored to the patient, considering his expectations, anxieties about treatment choice and social environment.† Optimising treatment outcomes relies on open communication between all members of the healthcare team, adequate training of the healthcare professionals and confident use of the available guidelines (e.g. WFH).[52]

General guidance[126]	Considerations for people with haemophilia ★
Pain diagnosis with appropriate differential	Identify the cause of pain: bleed or arthropathy, nociceptive-mediated inflammatory or / and neuropathic
Pain diagnos Psychological assessment, including risk of addictive disorders is with appropriate differential	Assessment of depression and identification of addictive disorders (including the reliance on narcotics and alcohol to manage pain at home)
Informed consent about treatment plan	
Treatment agreement	Chronic pain is usually associated with joint degeneration or other long-term complications of haemophilia, which may require orthopaedic surgery to alleviate the pain[51]
Pre-intervention and post-intervention assessment of pain level and function	
Appropriate trial of opioid therapy ± adjunctive medication	Ensure that pain management done at home by the patient is discussed to avoid addiction risk. Refer to haemophilia-specific guidelines
Reassessment of pain score and level of function	
Regularly assess the four 'A's of pain medicine: • Analgesia (pain relief) • Activities of daily living (psychosocial functioning) • Adverse effects • Aberrant treatment behaviour	
Periodic monitoring and review of pain diagnosis as well as comorbid conditions including addictive disorders	Periodically review cause of pain (e.g. target joint or ongoing arthropathy)
Documentation of consultation	

Table 6.1.
Universal precautions of pain management

Understanding pain management and pain mechanisms

Physical activity may influence the experience of pain in a number of different ways.[116] Evidence from the general population suggests that different forms of physical activity lead to pain relief, positively influence a patient's mood,[116, 128, 129] reduce social isolation, improve quality of sleep[130] and increase functional capacity.[84] Physical activity can also improve self-esteem and confidence by an improved perception of being healthier.[84] All these effects can increase the likelihood of an individual being able to cope with and handle pain.

Distinct effects of physical activity:[84]

1. "Direct" pain-relieving effects of physical activity
2. "Non-direct" effects on fitness and mood, reduced stress sensitivity and improved sleep, with potentially even greater effects on the pain situation of the patient
3. Positive effects of physical activity on lifestyle-related diseases in patients who tend to be inactive

"Whatever treatment option is chosen for your patients, it is important to monitor pain and adjust management if necessary. Ongoing collaboration with the multidisciplinary team, including pain specialists is crucial."†

The type of physical activity a person with haemophilia can benefit from will depend on his pain status, joint status and initial physical fitness.† Physical activity may be accompanied by the risk of traumatic bleeding, making the type of physical activity an important consideration.[105] Physical activity in patients with long-term pain is often very low and, consequently, the intensity of the chosen activity should ideally be gradually increased, starting at a low-intensity level.[84] Available studies from the general population and people with haemophilia suggest that it is possible to achieve pain relief through short sessions of physical activity.[131, 132] Additional strategies for coping with pain besides exercise can include massage, physiotherapy, relaxation techniques, pain medication and active coping strategies.† The benefits of activity and steps to encourage an individualised physical activity are covered in Chapters 3 and 4.

† Author Recommendation

Summary: Pain and physical activity

Understand pain management and pain mechanisms[104, 105]

Identify type of pain - acute vs chronic - and underlying factors associated with pain for an individualised multidisciplinary management[46, 48, 105]

Assess the impact of pain on the person's ability to undertake their chosen physical activity[76]

Evaluate and monitor impact of physical activity on pain and adjust management and / or activity as needed[114]

In conclusion...

Appropriate physical activity – which can be anything from mowing the lawn to swimming in a competition – has multiple health benefits for people with haemophilia. The choice of activity is important and will be different for every person, depending on their personal preferences and physical capabilities. The desired activity and any necessary preparations or precautions should be discussed with the physician and other members of the multidisciplinary team, including a musculoskeletal specialist if possible.

We hope that this booklet has provided insights, information and advice to help people with haemophilia realise the many benefits of a more physically active and healthy lifestyle.

Glossary

Aerobic: Any moderate exercise / activity that uses large muscle groups, can be maintained continuously and is rhythmic in nature (e.g. cycling, dancing, hiking, aerobic classes, swimming and walking)[133]

Anaerobic: Intense physical activity of very short duration, fuelled by the energy sources within the contracting muscles and independent of the use of inhaled oxygen as an energy source (e.g. sprinting, high-intensity interval training [HIIT], etc.)[133]

Atrophy: Decrease in muscle mass[134]

Balance: Even distribution of weight enabling someone or something to remain upright and steady[135]

Bone health: Includes bone quality that refers to the capacity of bones to withstand a wide range of loading without breaking. Bone health also includes bone mineral content, structure, geometry and strength[60]

Bleeding episode: Also known as a haemorrhage, refers to blood escaping from the circulatory system from damaged blood vessels.[136] See also 'haemarthrosis'

Bone mineral density (BMD): Measurement of the amount of calcium in bone. Methods for taking measurements are fast, non-invasive and painless, for example X-rays or CT scans[137]

Body mass index (BMI): An index that relates weight to height, calculated as a person's weight in kilograms divided by his or her height in meters squared[138]

Conditioning: A type of resistance training which activate cells that build up the muscle tissues to prepare muscles for similar exercise movements in the future[139]

Deconditioning: The multiple, potentially reversible changes in body systems brought about by physical inactivity and disuse[139]

Exercise dose: Duration and intensity of exercise sessions[140]

Haemophilic arthropathy: Permanent joint damage occurring in people with haemophilia as a long-term consequence of repeated joint bleeds. In later stages, it can lead to wearing away of the bone around the joint, causing pain and limiting mobility[141]

Haemophilia Joint Health Score (HJHS): Sensitive validated score for physical examination of joint health in haemophilia[142]

Haemarthrosis: Bleeding into a joint, most commonly into the knee, ankle or elbow joints)[143]

Haemostasis: Physiological process that stops bleeding at the site of an injury while maintaining normal blood flow elsewhere in the circulation[144]

Hyperlipidaemia: Hyperlipidaemia is the state of raised serum levels of either total cholesterol, low-density lipoprotein cholesterol, triglycerides or both total cholesterol and triglycerides. Hyperlipidaemia is associated with increased risk of cardiovascular disease[145]

Hypertension: Also known as high blood pressure, places strain on organs and can increase the risks of serious and life-threatening conditions. A blood pressure of 140/90 mmHg or greater is considered to be high[146]

Obesity: Abnormal or excessive fat accumulation that presents a risk to health. A person with a BMI of 30 or more is generally considered obese[147]

Osteoporosis: Condition that weakens bones, making them fragile and more likely to break[148]

Osteophytes: Bony lumps (bone spurs) that grow on the bones of the spine or around the joints. They often form next to joints affected by osteoarthritis, a condition that causes joints to become painful and stiff[149]

Overweight: Overweight is defined as abnormal or excessive fat accumulation that presents a risk to health. A person with a BMI equal to or more than 25 is considered overweight[147]

Physical performance: Ability to carry out activities[150]

Proprioception: Perception or awareness of the position and movement of the body[151]

Range of motion (ROM): The range through which a joint can be moved. It is influenced by several structures: configuration of bone surfaces within the joint, joint capsule, ligaments, tendons, and muscles acting on the joint[152]

Resistance: Training in which you're working against some type of force that "resists" your movement (e.g. in swimming, water is the force resisting the movement)[153]

Self-efficacy: A person's estimate or personal judgment of their own ability to succeed in reaching a specific goal[154]

Synovitis: Occurs when the synovial membrane thickens and grows more blood vessels, causing even more bleeding into the joint. Often results of repeated joint bleeds that are not treated early or correctly[155]

Target joint: Defined as a single joint experiencing ≥3 spontaneous bleeds within a consecutive 6-month period[117]

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