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Living with **Haemophilia B**

Date of Preparation: October 2021
NP-18906

What is Haemophilia B?

Haemophilia is a **rare bleeding disorder** that reduces the ability of the blood to clot and which is **hereditary** (passed on through genes).[1,2]

Haemophilia is caused by clotting factors in the blood being missing, inactive or present at low levels.[1,2]

	Haemophilia A[1,3]	Haemophilia B[1,3]
	The clotting factor missing or at low levels is **FVIII (Factor 8)**	The clotting factor missing or at low levels is **FIX (Factor 9)**
	More common: **1 in 5,000 males** affected	Less common: **1 in 30,000 males** affected
	Can require **more frequent** treatment	Can require **less frequent** treatment

The severity of haemophilia is determined by the amount of clotting factor that is present in the blood: lower levels of clotting factor equates to more severe haemophilia.[1-4]

Bleeding into muscles and joints is the **most common symptom** of both haemophilia A and haemophilia B.[4]

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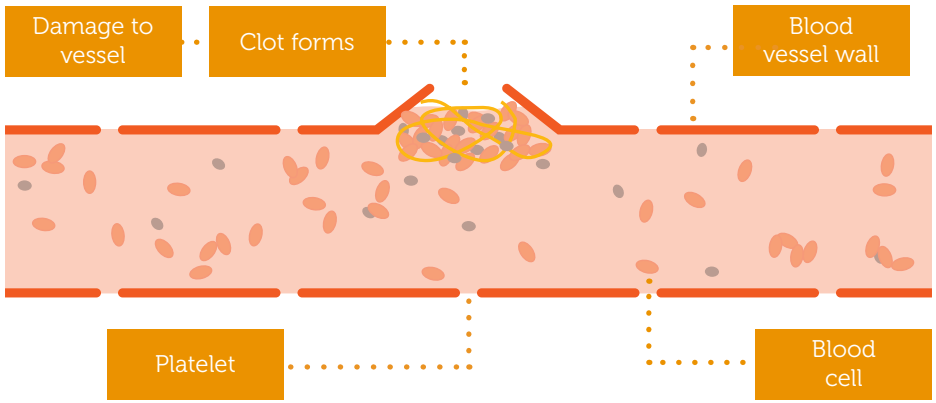
How does clotting work?[5]

Blood vessels carry blood to the muscles and tissues surrounding joints. Clotting factors, including FIX and FVIII, are present within the blood.

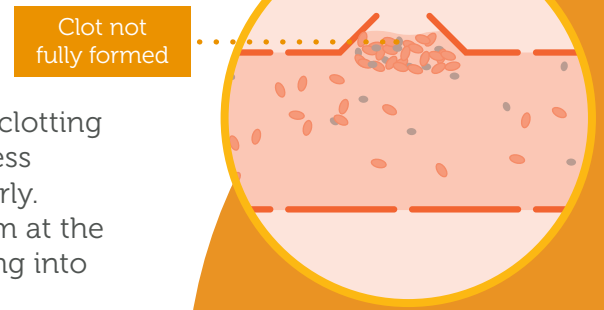
When a blood vessel is damaged, blood can escape – known as a bleed. This starts a process involving clotting factors and specialised blood cells called platelets. Platelets block the site of injury while the clotting process takes place.

Without haemophilia[5]



Once the clotting process has finished, a sticky mesh (a clot) covers the site of injury. This blocks the damaged vessel wall to stop further bleeding and allows the blood vessel to heal.

With haemophilia[5]

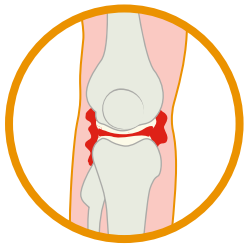


With a missing working clotting factor, the clotting process cannot take place properly. A stable clot doesn't form at the site of injury and bleeding into the tissues continues.

What is a joint bleed?

Bleeds are often not caused by an obvious injury – known as spontaneous bleeds.[4]

In people with haemophilia, bleeds can happen in the joints, commonly in the elbows, knees and ankles.[4]



During a joint bleed, blood enters the area between the bones of the joint which is already filled with fluid, putting extra pressure on the joint.[4]

Joint bleeds should be treated quickly to reduce the pain and to try to prevent further damage.

After several bleeds in the same joint:[6,7]

- **Important tissues** that protect the joint are damaged
- **Muscles** around the joint can become weakened
- **Bone** of the joint can start to break down
- This causes **permanent damage, loss of movement** and **severe, constant pain** in the joint
- To prevent further damage, **surgery** might be needed to fix the joint into place, or to replace the joint
- Joint damage can reduce the person with haemophilia's ability to take part in activities and can reduce their **quality of life**

How is Haemophilia B treated?

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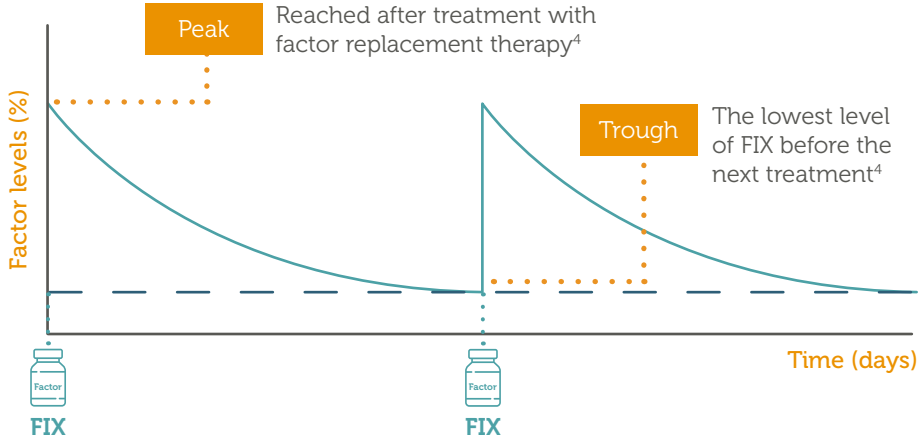
Treatment of haemophilia B aims to prevent or treat bleeds.[4]

The most commonly used treatments for haemophilia B are factor replacement therapies which replace the missing **FIX (Factor 9)** in the blood.[4]

Prophylaxis

Prophylaxis consists of **regular factor replacement injections to prevent bleeds** and is thought to be the most effective way to prevent bleeds. It should ideally **start at an early age** and **before the first bleed**.[4,8]

Measuring the amount of FIX in the blood can help to understand the protection level during prophylactic treatment, although other aspects should also be considered in order to personalise treatment.[4]



The curve on the factor level graph will vary depending on the patient, as well as the type of factor replacement therapy used for prophylaxis.[4]

Tailoring treatment[4]

Treatment is not always all about factor levels, and should be tailored for each person with haemophilia by their healthcare team.

Treatment should be based on a range of aspects in addition to the amount of FIX measured in the blood including: lifestyle, physical activity levels, age, number of bleeds and joint health.

On-demand treatment[4]

On-demand treatment consists of factor replacement injections *in response* to a bleed, rather than regular treatment to *prevent* bleeds.

Other treatment options

Research is ongoing into other treatment options, including gene therapy, which would aim to make an individual produce a functioning clotting factor.[9]

Other investigational treatment options aim to prevent bleeds without replacing the missing clotting factor (non-factor therapies).[10,11]

Everyone has their own lifestyle and personal goals, and haemophilia shouldn't hold anyone back. On the next few pages are some examples of patients who are learning to manage their haemophilia in order to lead full lives.

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Noah, 4

My son Noah has **severe haemophilia B**. As he has had prophylactic treatment from such a young age, we have a good weekly routine.

He loves playing with other children at the park. It is so important to me that he can play and make friends, but **I do worry** about him falling and hurting himself, which could lead to a bleed.

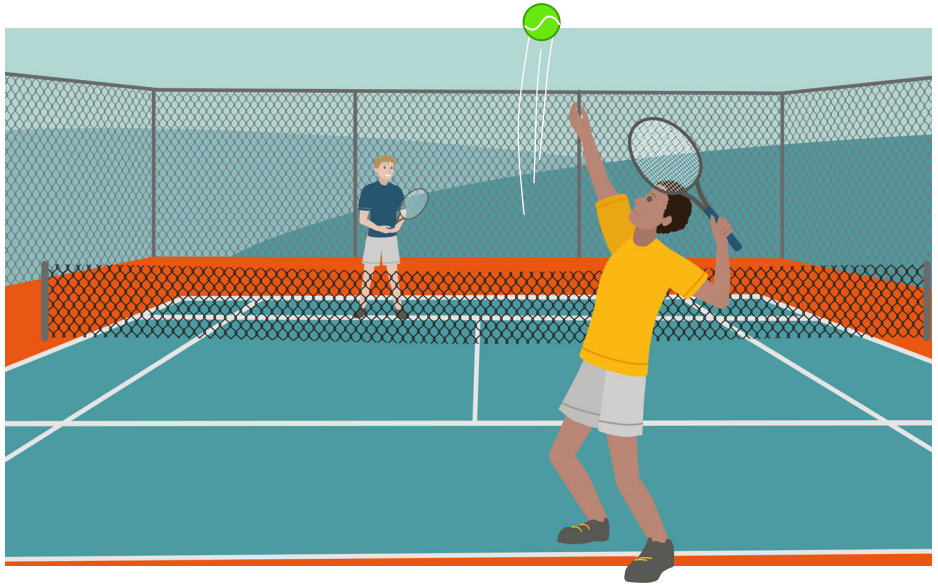
Kirsty, our haemophilia nurse, helped me understand how we can **ensure Noah is well-protected** from bleeds.

Kirsty is also helping us prepare for when Noah goes to school, so I can be sure his teachers understand his treatment and he doesn't have to miss out on any activities. It is all about working together to ensure that Noah is supported in **achieving his goals.**

Before engaging in physical activities or sports, consult with your doctor about your possibilities with haemophilia today.

James, 16

I really love **keeping fit**, especially playing tennis. I am grateful I can exercise to keep me active and strong.



There is a great community at my tennis club, so it's part of my social life too. It is important for me to **understand the risks** of playing tennis and how to **minimise them**.

I have regular sessions with my physiotherapist, Julia, who is part of a multi-disciplinary team at the hospital. Julia knows my strength and fitness abilities, so can help me to **achieve my activity goals safely**.

If I do get injured, Julia can help me recover properly. Talking with Julia and other healthcare professionals helps me to **continue training whilst being protected** against bleeds.

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Charlie, 24

I got a tattoo a few years ago and now I really want to get a **body piercing**.

I would like to feel protected and safe if I do decide to get a piercing. I know that piercings **carry a larger risk of bleeds** than tattoos, so I went to see my doctor, Dr Roberts.

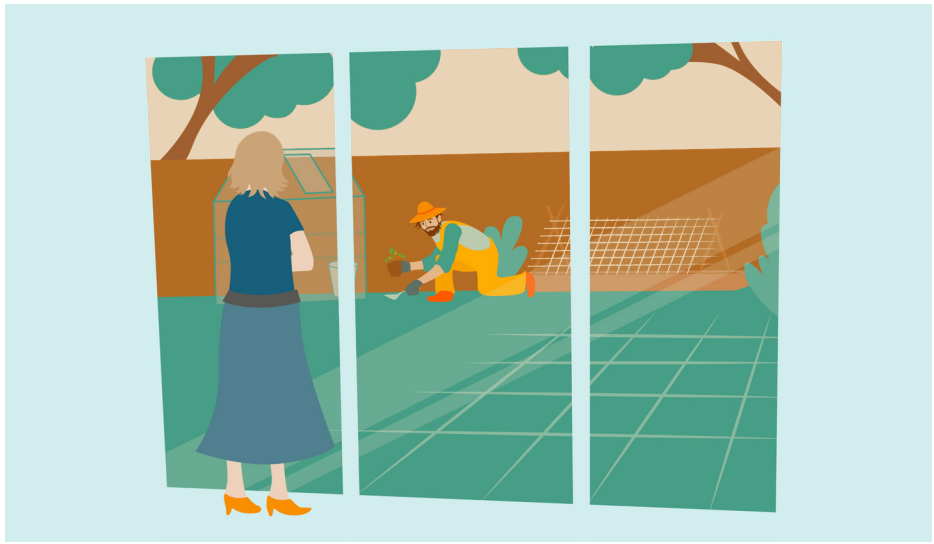
Dr Roberts helps me **consider the potential risks** against the benefits and advises me on how I can ensure my treatment is keeping me **well-protected** from bleeds.

Talking with Dr Roberts has made me aware of the considerations and possible risks associated with getting a piercing, enabling me to make **an informed decision**.

Tattoos or piercings can be risky. If you are considering getting a tattoo or piercing, consult with your haemophilia care team to discuss the possible risks.

Martin, 58

I have just retired and now that I have the time, I would really like to help my wife take care of our garden at home.



I know that working in the garden will mean I am **much more active** than I was when I was in the office. This means I need to **ensure I am managing my haemophilia well**, especially as my wife, Linda, is worried that I might injure myself.

My haemophilia team is very supportive. Armand, my physiotherapist, and Beverly, my doctor, have been working with me to make sure I am **prepared** for this new active lifestyle.

They encourage me to **reach my goals** and **help me to understand** how I can protect myself from bleeds, while giving Linda confidence that I can lead a more **active life safely**.

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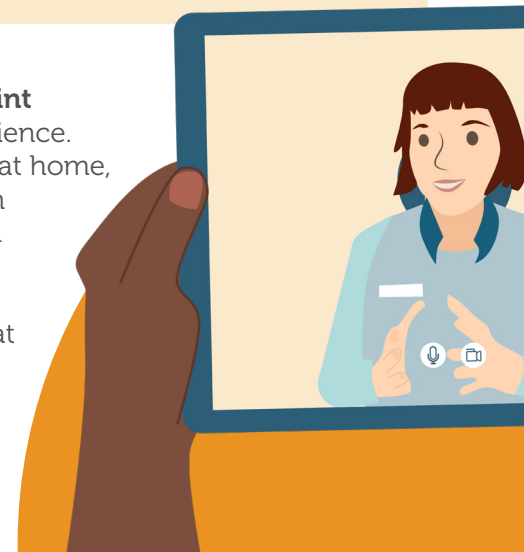
Arthur, 74

I am not as mobile as I used to be! My knee joints are often **stiff and painful** and this is making it difficult for me to play with my grandchildren.

I don't want my haemophilia symptoms to **affect my daily life** and happiness, so I am working with my physiotherapist, Sarah, via video call.

Sarah is helping me to **maintain my joint mobility** and **manage the pain** I experience. She recommended some **adaptations** at home, as well as some exercises I can do. I am working really hard on keeping up with the exercises, and I know that if I need support in other areas, I can ask to be referred to the right person to help at my **haemophilia treatment centre**.



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Patient stories were developed based on advice provided by Debra Pollard, Advanced Nurse Practitioner, Haemophilia & Inherited Bleeding Disorders at Katharine Dormandy Haemophilia & Thrombosis Centre, Royal Free London NHS Foundation Trust, London, UK.

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