

Moderate Haemophilia B

Information for patients, parents, and carers from the Haemophilia Centre

What is Haemophilia B?

Haemophilia B is a rare condition that affects blood clotting. It affects around 1 in 30,000 males in the UK.

People with Haemophilia B do not bleed any faster, but may bleed for longer than expected.

Having moderate Haemophilia B should not affect everyday life, but there may be more bleeding problems compared to someone with mild Haemophilia B. Most of the bleeding problems that happen tend to be after an injury, surgery, or dental treatment. Once it has been diagnosed we can treat before surgery or dentistry and prevent bleeding from happening.

How does Haemophilia B affect blood clotting?

Factor IX is a protein made by your liver and helps to make your blood clot when there is blood vessel injury. If you have Haemophilia B you do not have enough of this protein for your blood to clot normally, or what you do have of the protein may not work as well as it should.

The normal level of Factor IX in the general population should be 50 to 200 iu/dl or %. In moderate Haemophilia B your level of Factor IX could be anywhere between 1 and 5 iu/dl or %.



How do you get Haemophilia B?

Haemophilia B is an inherited condition, you are born with it, and the pattern of inheritance is X-linked. This means that usually men are affected by the condition and women are carriers. However, it is worth noting that some carriers can have low levels of Factor IX and may have some bleeding problems.

Women carry two X-chromosomes and men one X-chromosome and one Y-chromosome. The haemophilia gene is carried on the X-chromosome. Mutations can happen; the mutation means the haemophilia gene cannot make enough Factor IX.

In seven out of 10 cases of people born with Haemophilia B, there is a family history of the condition. In as many as three in 10 either the mother is unaware she is a carrier or the condition has happened spontaneously.

What are the symptoms?

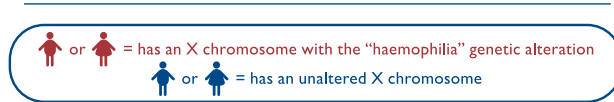
Common symptoms

- Nose bleeds
- Bleeding from gums
- Easy bruising
- Bleeding during and/or after surgery
- Bleeding after trauma/injury.

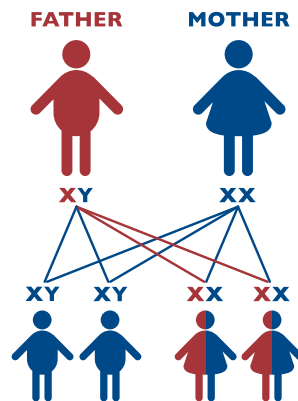
Rare symptoms

- Blood in your urine (haematuria)
- Bleeding in your stomach or intestines (blood in stools (poo) or black tarry stools)
- Bleeding in to your muscle
- Bleeding in to your joints (haemarthrosis)
- Bleeding in to your brain.

Haemophilia Genetic Inheritance

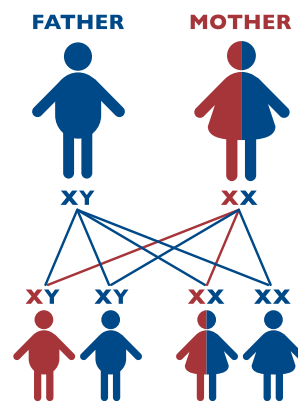


When the father has haemophilia and the mother is unaffected



None of the sons will have haemophilia.
All of the daughters will carry the gene. Some might have symptoms.

When the mother carries the altered gene causing haemophilia and the father is unaffected



There is a 50% chance at each birth that a son will have haemophilia.
There is a 50% chance at each birth that a daughter will carry the gene.
Some might have symptoms.

Source: Haemophilia Foundation
Australia, www.haemophilia.org.au (2013)

How is Haemophilia B treated?

Day to day treatment is not needed for Haemophilia B. Treatment may be necessary before and after surgery, dentistry, or trauma. The need for treatment will depend on:

- how severe your bleeding is
- the type of surgery or dental treatment you are having
- your previous bleeding history
- your family's history of bleeding; and
- your Factor IX level.

What are the treatments?

- Antifibrinolytic Agents (Tranexamic Acid)
- Replacement Factor IX

- **Antifibrinolytic Agents (Tranexamic Acid)**

Tranexamic Acid works by stopping the early breakdown of a clot that has been made after injury to your blood vessel. Fibrin gives the blood stability. Tranexamic Acid stops the substances that destroy the fibrin within the clot.

Often it is the only treatment needed for people with Haemophilia B. It is particularly useful for mouth bleeding and therefore very good preventative treatment when you are having a dental procedure.

The treatment is usually given as tablets. You may be asked to crush the tablets in a small amount of water and swish it around your mouth, before either swallowing it or spitting it out (you will be advised by the haemophilia team which is best for you).

Tranexamic Acid is not used when there is blood in the urine, as small clots can occur which can then block your urinary tract and cause you pain.

Are there any side effects?

Side effects are rare but include:

- nausea (feeling sick) and vomiting
- diarrhoea
- joint or muscle pain or cramps
- headache or migraine
- runny or stuffy nose
- stomach or abdominal pain.

Other side effects can include skin rash and changes to your colour vision.

- **Replacement Factor IX**

Factor IX products replace the missing Factor IX in your body. This is recombinant (meaning that it has been made in the laboratory) and does not come from blood donors.

It is given to you either through a small butterfly needle or a cannula (a small tube into a vein in your arm) directly into the vein over two to five minutes.

Are there any side effects or risks to this treatment?

Treatment is generally very well tolerated so side effects are not common. Some reported side effects include fever, headaches, or allergic reactions.

Replacement Factor IX does carry one important risk which you need to be aware of, and that is the risk of inhibitors, but with Haemophilia B the risk of developing an inhibitor is very small. Please see the **Inhibitors** patient information sheet for more information. Ask a member of staff for a copy or go to the haemophilia web page www.ekhufft.nhs.uk/patient-information/haemophilia-treatment/

Where can I find more information about Haemophilia B?

There are several sources of useful information about Haemophilia B, including the following.

- **NHS: Haemophilia**
Web: www.nhs.uk/conditions/haemophilia
- **The Haemophilia Society**
Web: www.haemophilia.org.uk
- **World Federation of Hemophilia**
Web: www.wfh.org

This leaflet has been produced with and for patients

If you would like this information in **another language, audio, Braille, Easy Read, or large print** please ask a member of staff. You can ask someone to contact us on your behalf.

Any complaints, comments, concerns, or compliments please speak to your doctor or nurse, or contact the Patient Advice and Liaison Service (PALS) on 01227 78 31 45, or email ekh-tr.pals@nhs.net

Patients should not bring in large sums of money or valuables into hospital. Please note that East Kent Hospitals accepts no responsibility for the loss or damage to personal property, unless the property had been handed in to Trust staff for safe-keeping.

Further patient leaflets are available via the East Kent Hospitals web site www.ekhufft.nhs.uk/patientinformation