East Kent Hospitals University NHS Foundation Trust

Severe 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 faster, but may bleed for longer than expected.

Bleeding from scrapes and cuts do not usually cause too many problems for people with severe Haemophilia B. The main problems arise from bleeding that can happen internally, particularly in to your muscles and joints.

For further information, please see our information sheet **How to recognise a bleed**. Ask a member of staff for a copy or download it from www.ekhuft.nhs.uk/patient-information/haemophilia-treatment/

How does Haemophilia B affect blood clotting?

Factor IX is a protein made by your liver and it helps make your blood clot when there is blood vessel injury. If you have Haemophilia B you do not make enough of this protein for your blood to clot normally.

The normal level for Factor IX in the general population is 60 to 150 iu/dl or %. In severe Haemophilia B your level of Factor IX will be less than 1 iu/dl or %.

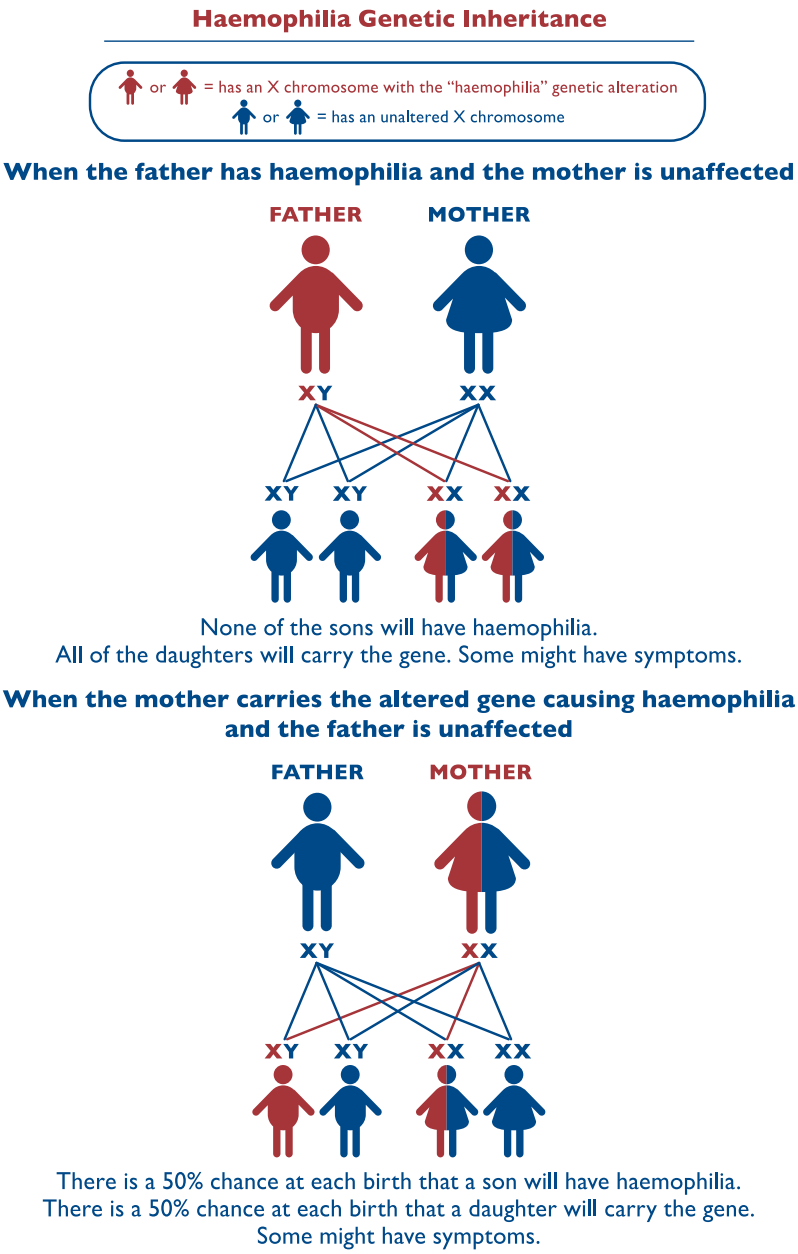
We care

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 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 B 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 out of 10, either the mother is unaware she is a carrier or the condition has happened spontaneously.



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

What are the symptoms of Haemophilia B?

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

How is Haemophilia B diagnosed?

- Haemophilia B is diagnosed through a blood test.

- If we were aware of your family history, it may be that we have tested your child's blood from the umbilical cord sample at the time of their birth.

- We always confirm a diagnosis with a second blood test and send blood to another specialist centre to find the genetic mutation causing your haemophilia. This information is useful to test other family members, particularly girls who may be carriers. Knowing the mutation can save a considerable amount of time for future searches.

How is Haemophilia B treated?

There is currently no cure for Haemophilia B but there are very good and effective treatments available to manage the condition. With the right treatment your child will be able to live a normal and active life. Today, a child born with severe haemophilia has a normal life expectancy.

Haemophilia B is treated by replacing the missing Factor IX. If there is a bleeding problem, this treatment is given straight into the vein in the short term; this is known as **on demand treatment**. Usually around the age of one year we will need to start regular replacement or "prophylaxis" and we would administer this more easily using a Portacath.

For more information see the information sheets **Implantable port (portacath or port)** and **Prophylaxis**. Ask a member of staff for a copy or go to the haemophilia web page www.ekhuft.nhs.uk/haemophilia-patient-leaflets/

- Replacement Factor IX
- Antifibrinolytic Agents (Tranexamic Acid)

- **Replacement Factor IX**
  Recombinant Factor IX replaces the missing Factor IX in your body. Recombinant means 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 directly into a vein in your arm) 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 uncommon. Some reported side effects include fever, headaches, or allergic reactions.

  There is an important risk you need to be aware of with replacement Factor IX, and that is the risk of inhibitors. 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.ekhuft.nhs.uk/patient-information/haemophilia-treatment/

- **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 clot stability. Tranexamic Acid stops the substances that destroy the fibrin within the clot.

  When your son is small we can get Tranexamic Acid syrup from the hospital pharmacy. It needs to be ordered specially and has a short expiry date.

  Once he starts eating solid food we would advise he takes the tablets. These come in 500mg tablets but are easy to break in half if necessary. They should not be chewed but can be crushed and then mixed with a small amount of soft food such as yogurt.

  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 colour vision. **If you do have any side effects, please contact the Haemophilia Centre for advice.**

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

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.ekhuft.nhs.uk/patientinformation

Information produced by the Haemophilia Centre
Date reviewed: October 2021 Next review date: February 2025 Web 418