

Living life with
mild or moderate
Haemophilia



BAXTER & YOU

Shaping the future of haemophilia together

Is this book for me?

This book is intended for the parents of a child with mild or moderate haemophilia and for the children themselves when they are ready to learn more about their condition. It may also be helpful if you are a teenager or adult, especially if you have recently been diagnosed.

Often girls and women who are haemophilia carriers have lower than normal levels of the affected clotting factor. Although this does not mean they have mild or moderate haemophilia, it amounts to the same thing in terms of bleeding risk.

All the information in this book is for you too, and there is also a special section for you at the end.

There are many other books about haemophilia but they mainly concentrate on the severe form of the condition. Try not to let them alarm you. Certain information in these books can be very useful but you may find it helpful to read this book first.

It will help you to understand that much of the information available about treatment and complications of severe haemophilia does not apply to those with a milder form of the condition.

Remember, the treatment and complications associated with severe haemophilia do not all apply to mild and moderate haemophilia.

What is haemophilia?

When a person's blood cannot clot properly the condition is generally known as haemophilia. A person with haemophilia will not bleed any faster than anyone else following an injury but they can bleed for longer.

Worldwide more than 350,000 people have haemophilia. In 70% of all cases there is a family history of haemophilia. In as many as 30% of cases, either the mother may not be aware that she is a carrier, or the condition may have occurred spontaneously.

The two most common types of haemophilia are haemophilia A and haemophilia B. The blood of people with haemophilia A has reduced levels of a protein called factor eight (written as factor VIII). The people with haemophilia B have reduced levels of a protein called factor nine (factor IX).

Haemophilia is generally classified by how much of these proteins, known as clotting factors, there are in your blood. Mild haemophilia usually means a factor VIII or IX level of 6–40% of normal; moderate haemophilia usually means levels of 1–5% and severe haemophilia less than 1%. Once established, the severity of haemophilia does not usually change during a person's lifetime and people in the same family inherit the same level of severity of haemophilia.

Although haemophilia is a lifelong disorder, which will not go away, it can be managed as part of a normal active life.

Inheriting haemophilia – it's in our genes

Everyone's body is made up of tiny cells, each of which contains 46 chromosomes, arranged in 23 pairs. These chromosomes, and in particular the so-called 'genes' they contain, decide many of the things which make us into unique individuals, for example the colour of our hair and eyes.

We inherit one half of each pair of chromosomes from our mother, the other comes from our father. Consequently we are a mixture of our mother's and our father's genes. That is why, whilst we may have blue eyes like our mother, we may also have brown hair like our father.

One of the pairs of chromosomes decides what sex we become. The individual sex chromosomes are called X and Y. Women have two X chromosomes; men have an X and a Y chromosome.

Haemophilia is a 'sex linked' disorder. This means the gene for haemophilia is found on one of the sex chromosomes - the X chromosome.

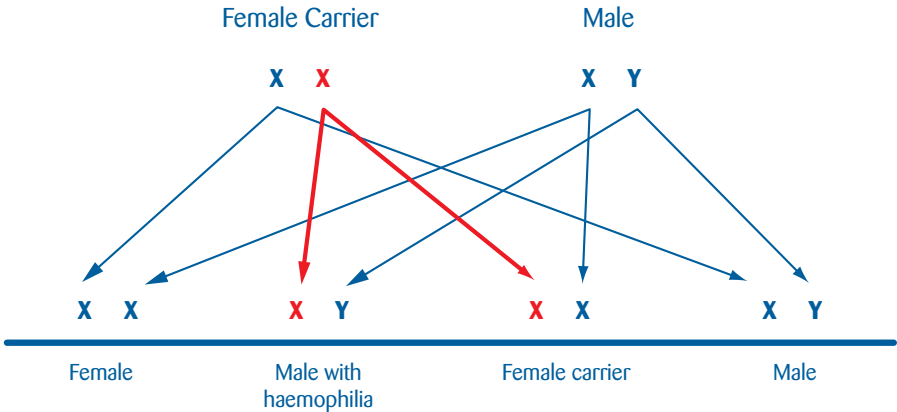
As men have only a single X chromosome, if it carries the haemophilia gene, he will have haemophilia.

As women have two X chromosomes, if one carries the haemophilia gene, the other one will make up for some of the deficit. This is why a carrier may only have about half of the usual amount of clotting factor in their blood; however, this can vary from less than 10% to over 90% depending upon which of her two X chromosomes is most active.

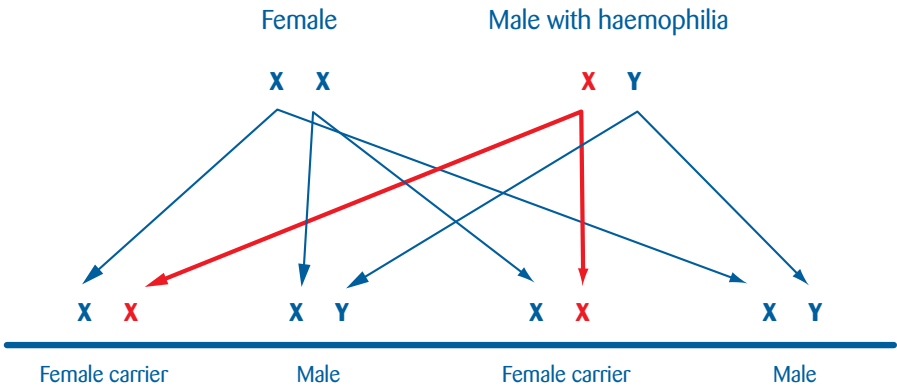
The diagrams on the right show what happens if a female carrier or a male with haemophilia has children. On both diagrams the X chromosome containing the haemophilia gene is shown in red.

A woman carrying the haemophilia gene has a 50:50 chance of her son having haemophilia or her daughter being a carrier.

This is what can happen if a female carrier has children:



Now look what can happen if a man with haemophilia has children:



All the daughters of a man with haemophilia will be carriers but none of the sons will have haemophilia.

If you are not aware of any previous history of haemophilia in your family your doctor may recommend that you and the rest of your family are tested.

This will not only help identify any carriers in the family but will also spot any other, as yet undiagnosed, haemophilia.

How does blood clot and what happens in haemophilia?

Typically when the body is injured, blood clots to form a scab. This stops the bleeding and provides protection while the body repairs itself. In mild and moderate haemophilia this also happens but sometimes the clot is weaker and breaks down before the repair process is finished which can mean that the bleeding restarts.

This clotting process is very complicated and is often described in books using relay races or toppling dominos to help explain it.

The different ways blood clots in severe, moderate and mild haemophilia

To illustrate the difference between severe, moderate and mild haemophilia it might help to think of the amount of clotting factor in a person's body like the amount of air in a football. Usually if the ball is fully inflated, it can be kicked long distances and it can be bounced really high. In mild and moderate haemophilia the ball is not fully inflated, in fact it's less than half full. The ball can still be kicked and bounced but not as far or as high.

It is not as easy to use as the fully inflated ball but it still works.

This is different to severe haemophilia where there is so little air in the ball that it is barely inflated at all. It cannot be used and it needs pumping up. This is why, in severe haemophilia, more treatment with factor is needed.

Understanding what treatment is suitable for you

Your doctor or nurse at the Haemophilia Centre will tell you what type of haemophilia you/your child has and what the clotting factor level is. They will use this information to work out what treatment may be needed. For example, people with mild or moderate haemophilia often have enough clotting factor to cope with minor injuries.

This means they may only need treatment if they have a serious accident or an operation. In fact some people only have their haemophilia diagnosed when these things happen because previous minor injuries have healed normally.

People with mild or moderate haemophilia often have enough clotting factor to cope with minor injuries and everyday life.



How do you feel about having haemophilia?

If you are the parent of a child with haemophilia:

If your child has haemophilia you may feel very sorry for them. You may even feel guilty if you find you were the carrier of the haemophilia gene. These are quite normal reactions but it is important that you learn to accept your child's condition. You may find it helps to talk to the nurse at your Haemophilia Centre or to other parents; your Centre can give you contact numbers for the Haemophilia Society and local support groups.

Children need to be children and, while you will inevitably need to be a little cautious, it is important to encourage your child to play and have fun. Remember if your child has a bruise or bleed it does not help if you become upset or angry. Instead, follow the guidance from your Haemophilia Centre on how to deal with bruises and bleeds.

It will also help if the rest of your family understands about haemophilia, especially any other children you may have.

In the early days they may feel that your child with haemophilia is getting special treatment.

On the other hand, they may feel guilty, particularly if you become cross when your child with haemophilia becomes bruised during play. Be aware that children with mild or moderate haemophilia will probably bruise more easily than their peers.

If you have haemophilia yourself:

If your haemophilia has recently been diagnosed it has probably been a shock. But although it may be difficult at first, remember there is much more to your life than haemophilia. Don't let your condition dominate everything you do. Instead learn as much as possible about haemophilia, so you know what to expect and what to do if problems arise.

It is important to learn as much as you can about haemophilia – your Haemophilia Centre and the Haemophilia Society will have plenty of information. The more you know, the easier it will be to make haemophilia part of your everyday life.

How will haemophilia affect me/my child?

People with mild or moderate haemophilia do not usually have bleeding problems in everyday life. They tend only to have problems after an injury, an operation or dental treatment.

Nevertheless be aware that bleeds could happen from less severe injuries and that nosebleeds may be more frequent and last for longer.

The term 'bleed' has a specific meaning in haemophilia which is slightly different from what most people think. A 'bleed' is often more likely to mean bleeding into a muscle as a deep bruise rather than open bleeding from a cut.

Make sure that you learn from the staff at your Haemophilia Centre what a bleed is, how to recognise the different types and what to do. The section '**Recognising and coping with bleeds**' later in the book may help you with this.

In contrast to those with mild or moderate haemophilia, people with severe haemophilia have spontaneous bleeds into joints and muscles even when they have not been injured. They need treatment to stop these bleeds or to prevent them.

Remember, if in doubt... contact your Haemophilia Centre.

How should I live my life day-to-day?

There are a few practical things you will need to consider to help you/your child in your everyday life. In some ways this is more important with mild/moderate haemophilia because you do not feel any different from anyone else and you can forget the simple things that can prevent a problem from happening:

Identify yourself

Your centre will supply you with a 'green card' which you should carry with you at all times, just in case you have an accident. There are also several companies who make necklaces or bracelets that you can wear to make sure, in an emergency, people know about your haemophilia. Details can be obtained from your Centre.

Care in the home

Try to provide a safe environment for your young child with haemophilia, just as you would for other children. If you can, limit the number of times you tell your child to be careful so he (or she) learns to approach life with confidence.

Like every parent, you will need to leave your child in the care of others from time to time. Make sure anyone who looks after your child knows when to be concerned and who to contact in case of emergency.

School and work

When your child starts school, make sure the staff are informed. If they do not understand about haemophilia they may place quite unnecessary restrictions on your child's day-to-day activities, but a child with haemophilia should not be treated any differently from anyone else at school.

The Haemophilia Nurse at your Centre will be able to provide you with information to give to the school and if necessary will talk to staff involved with your child on your behalf.

If you are an adult with mild or moderate haemophilia or the parent of such a child you might consider telling your employer in case you need time off work.

Sport and activities

Sport and activities are beneficial to everyone, including people with all types of haemophilia. Those with mild or moderate haemophilia can take part in most sporting activities but remember there is a risk of bleeding following injury so it is best to avoid contact sports such as rugby and boxing.

Whatever sport your child chooses the occasional injury will be impossible to avoid. Encourage your child to tell you when they get hurt and be honest about how it happened. This will help you and the doctor to decide what treatment they need.

Following a few important guidelines will help you to live life to the full.



What general health issues should I be aware of?

Although people with mild or moderate haemophilia don't generally have bleeds in everyday life, care is needed with dental problems, vaccinations and operations.

Vaccinations

You/your child should have all the usual routine vaccinations, plus any others that your centre may recommend. This also applies to travel vaccination at all ages.

You need to ensure that the person carrying out the vaccination knows that you/your child has haemophilia. They should be aware that they may need to administer the vaccine in a different way than usual and they should contact your Haemophilia Centre for advice.

Dental

It is important for everyone to have regular dental checkups so that potential problems are dealt with early. This is especially true for people with haemophilia. Before you have any dental treatment you should speak to the staff at your Haemophilia Centre, so they can advise you. They may even recommend a particular dentist to you. You should always make sure that your family dentist knows that you/your child has haemophilia, as there is a risk of swelling and prolonged bleeding following procedures.

When children lose their baby teeth there may be bleeding. If bleeding occurs you will need to check that the bleeding stops. If it continues for a long time or keeps stopping and starting, ask your Haemophilia Centre for advice.

Remember to notify the staff at your Haemophilia Centre if you/your child is having any kind of procedure. They can advise you if any treatment is required before or after the procedure.

Surgery

Make sure the team looking after you/your child knows about your haemophilia and that they must contact your Haemophilia Centre for advice well in advance of the operation.

Bumps, bruises and minor accidents

As soon as a child starts to crawl and climb bruises are unavoidable and, even in a child with haemophilia, are not usually painful. Bruises should therefore not alarm you but you should keep an eye on them. They should fade and disappear within two weeks. If a bruise grows larger over a few hours you should contact your Haemophilia Centre for advice.

The only exception is for bruises that appear on the head, neck, throat, joints or groin. If you see bruising in any of these areas you should contact your Haemophilia Centre straight away.

The Haemophilia Doctor should check all head injuries, whether there is any outward sign of damage or not.

IMPORTANT

A Haemophilia Doctor should check suspected head injuries. Follow the usual emergency procedure if the injury occurs out of hours.

How do I recognise and cope with bleeds?

If you/your child has moderate haemophilia it is possible that you may experience bleeds without an apparent cause (as people with severe haemophilia do) or as a result of a minor injury. People with mild haemophilia are unlikely to have bleeds as a result of everyday minor accidents.

Because bleeds are rare, it is important to learn how to recognise them if they happen.

Here are some tips to help you:

Firstly, if you are a parent of a small child with moderate haemophilia it is a good idea to get into the habit of observing them carefully.

At changing or bath time

- Look at your child's skin.

Are there any new bruises?

- Are the legs equal in size? And arms? Are they moving the same way?
- Is there any bruising or swelling in the nappy area?

In general

- Is your child miserable or crying for no apparent reason (not because they are hungry, thirsty, need a nappy change or want a cuddle)?
- Do they avoid reaching for things with the nearest hand?
- Are they avoiding particular actions?
- Are they favouring one leg?

If you have any problems that you think may be related to haemophilia, contact your Haemophilia Centre for advice. It is always better to seek advice than let a potential problem go untreated.

What more do I need to know about bleeds at any age?

Other signs of bleeding or issues that may be encountered in moderate haemophilia:

Bleeds in muscles and joints

- The area feels tight, warm or swollen
- The limb is painful, stiff or difficult to extend

Bumps to the head

- There may be obvious swelling, lumps or bruising
- There may be no mark at all - but always seek help from the Haemophilia Centre

Bleeds in the mouth, stomach and intestines or urinary tract

- Visible bleeding from mouth or tongue
- Red or brown urine
- Bloody or black tar-like motions

Nose Bleeds

- These can go on for longer than normal and can be more frequent

What to do?

Basic first aid procedures apply in haemophilia as with everyone. Bleeding from cuts will stop more quickly if direct pressure is applied to the wound. Pain and swelling affecting a joint or tissue group can be reduced by wrapping a bag of frozen vegetables or some ice cubes in a towel and applying to the area.

If you think that an injury is complicated because of haemophilia then contact your Centre for specific advice.

Never use any medicine that contains aspirin (acetylsalicylic acid) or other Non Steroidal Anti-Inflammatory (NSAIDs) drugs such as Ibuprofen, unless advised to do so by your Haemophilia Centre. These medicines can interfere with blood clotting.



If I have a bleed what will the treatment be?

There are several alternative treatments for bleeding:

DDAVP

DDAVP (also known as desmopressin) may be used for people with mild or moderate haemophilia A. It makes the body release stored factor VIII so that it can help the blood to clot. DDAVP can be given as an infusion into a vein, as an injection under the skin of the tummy or as a nasal spray. As DDAVP releases stored factor VIII, it doesn't work for haemophilia B.

Tranexamic acid

Tranexamic acid works by stopping blood clots from breaking down after they've been formed. It is called an "anti-fibrinolytic" and can be taken as tablets, syrup, or injection. It is often used to help treat bleeding resulting from dental treatment, mouth and nosebleeds.

Factor concentrates

Factor concentrates work by replacing the missing clotting factor and need to be injected directly into a vein. They can be made using recombinant technology to 'grow' copies of human factor VIII or IX, or they can be harvested from blood donated from carefully screened donors.

Factor concentrates are sometimes needed to treat mild or moderate haemophilia, especially if you have a serious accident or if you need an operation. In some cases, people with moderate haemophilia have problems with bleeding into muscles and joints similar to someone with severe haemophilia and may need regular prophylactic injections with factor VIII concentrate to prevent painful and potentially damaging bleeds.

These treatment alternatives may or may not apply to you. Your Haemophilia Centre will advise you on what treatment is best for you.

Special advice for girls and women

Many girls and women who are carriers of haemophilia have low levels of a clotting factor, so they may have the same sort of problems with bleeding as those with mild or moderate haemophilia.

For women this may lead to extra problems with periods, which may be very heavy. The doctor at the Haemophilia Centre will be able to suggest treatment to control this heavy bleeding.

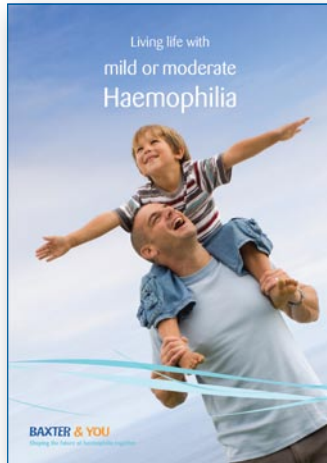
You will also need to take certain precautions during childbirth. Make sure that your Haemophilia Centre knows when you are pregnant so that they can arrange specialist care for you. Problems with bleeding are unusual during pregnancy because levels of clotting factors tend to be higher at that time. You may, however, need treatment to prevent or control bleeding when giving birth and extra care will be taken with the baby in case he/she also has haemophilia.

You are not alone

Finally, remember you are not alone. Your haemophilia, or your child's haemophilia, is now part of your life, but it does not need to dominate every day. Get to know the staff at your Haemophilia Centre. Although you should not need to contact them very often, it helps to have a familiar person to talk to when you are concerned. Remember they are always there to help you if you need them.

Although you may not need help from your Haemophilia Centre very often, keep in touch. They can provide you with a wide range of information, contacts and reassurance to help you live life to the full.





Thanks to the UK Haemophilia Nursing Community
for their help with this publication.

Haemophilia Society Helpline – 0800 018 6068

www.haemophiliacare.co.uk

BAXTER & YOU

Shaping the future of haemophilia together

ADV08/2264B

October 2008