

Thinking about...

Haemophilia B



Thanks to the UK Haemophilia
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Haemophilia Society Helpline - 0800 018 6068
www.haemophilicare.co.uk



Thinking about haemophilia B

When you have haemophilia B in your family there is a lot to think about and a lot you need, and want, to know. This book is designed for parents of a child with haemophilia B, and for children with haemophilia B when you want to know a little bit more about your condition.

There are several booklets available about haemophilia but most concentrate on the more common form, haemophilia A. Nevertheless much of the information they contain applies equally to haemophilia B. In the later pages of this book you will find information on the similarities, and the differences, between haemophilia A and haemophilia B to help you make the most of all this information.



Parent of a newly diagnosed child?

Your doctor will have told you that your child has haemophilia B (sometimes called Factor IX deficiency or Christmas Disease) and you are probably rather concerned. So first a few words of reassurance.

Nowadays children with haemophilia can grow up fit and strong. You will have to learn about the special treatment your child needs, but when you understand how to manage his haemophilia you can help him enjoy a full and happy childhood.

You will need some help, particularly at first. As well as the staff at your Haemophilia Centre, there is information available from The Haemophilia Society. Also ask at your clinic about Parent Support Groups. These groups can put you in touch with other parents of children with haemophilia.

As he grows your child will begin to take care of himself and eventually there is nothing to prevent him enjoying life as a happy and healthy adult.

Haemophilia today

You have probably already heard of haemophilia. Often - but not always - there will be other men in your family who have the same problem. No matter what you have heard, or what you think you know, check with your clinic.

The treatment for haemophilia has progressed in recent years and children can live their lives without many of the restrictions imposed even a decade ago. For example, today children with haemophilia have fun with their friends and play lots of different sports.

How blood clots

When you cut yourself your blood clots to form a scab. This stops the bleeding and protects you while your body repairs itself.

Blood clotting is really quite complicated. Imagine a relay race. When you cut yourself the first runner starts the race and passes the message that your blood needs to clot to the next runner. The message is passed from runner to runner until, at the end of the race, the last runner tells your blood to clot.

But with haemophilia one runner - called a clotting 'factor' - is missing. So the instructions never reach the end of the race and no one tells the blood to clot.

Blood clotting is a bit like a relay race. When you have haemophilia one runner is missing.



Haemophilia A or haemophilia B?

The two most common forms of haemophilia are haemophilia A and haemophilia B. Different 'runners' or 'factors' are missing in each type of haemophilia:

In haemophilia A factor eight is missing - written as Factor VIII.

In haemophilia B factor nine is missing - written as Factor IX.

Haemophilia B is quite rare - only about 1,100 people in the UK have it.



How many people have haemophilia B?

In the UK about 6,500 people have haemophilia. Many of them have haemophilia A (about 80%). Of the rest - about 20% - most have haemophilia B. (A very, very small number are missing other factors such as factor VII and factor XIII.) This means that bleeding disorders like haemophilia are rare, so don't be surprised if family doctors, health visitors and teachers have not come across someone with haemophilia before.

Treating haemophilia B

For both haemophilia A and haemophilia B the treatment is replacement of the missing clotting factor, by injection into a vein. For haemophilia B it is clotting factor IX which is injected.

There are two ways of giving factor IX:

On demand treatment - Factor IX is given as soon as the person with haemophilia recognises that they have a bleed. This does not necessarily mean going to the hospital. Many families learn to give the injection at home and children with haemophilia can often learn to give the injections themselves.

Prophylaxis - Injections of Factor IX are given twice a week to prevent bleeding occurring.

A little bit of history

Doctors knew haemophilia existed for centuries but haemophilia B was only recognised for the first time in 1952.

Some people call haemophilia B 'Christmas Disease' but it has nothing to do with December 25th. It was christened Christmas Disease because when doctors recognised this new type of haemophilia (caused by missing factor IX) their patient's surname was Christmas.

Haemophilia A versus haemophilia B - the similarities and the differences

There are a lot of similarities between haemophilia A and B, and a few important differences.

First the similarities:

Inheritance

Both types of haemophilia are inherited in the same way. This means that, except in very rare cases, only boys are born with haemophilia B. Women can be 'carriers' but do not have haemophilia themselves. So, for example, a woman who is a carrier of haemophilia B has a 50:50 chance of having a son with haemophilia B or a daughter who is a carrier. Although women do not have haemophilia B themselves they may sometimes have lower than normal levels of Factor IX. This is sufficient for everyday life, but if you are a woman with haemophilia B in your family it is best to have your factor IX levels tested before an operation or a tooth extraction.

Treatment

Both types of haemophilia are treated by injection of factor, either on demand or prophylactically.

Life-style

Nowadays children and adults with haemophilia can lead an almost normal lifestyle. This is the same whether you have haemophilia A or B.

And now the differences:

Frequency

Approximately 1 person in every 10,000 will have haemophilia A. Haemophilia B is five times rarer so will affect only one person in every 50,000.

Factors

Because people with haemophilia B lack Factor IX, this is the factor used for their treatment. Until recently only Factor IX extracted from blood was available but now artificial factor IX produced by recombinant technology can be prescribed.

Prophylaxis

If you have haemophilia A, factor VIII needs to be injected three times a week. But for haemophilia B, injections of Factor IX are only needed twice a week. This is because Factor IX lasts longer in your blood stream.

Inhibitors

Finally, some people find that after a while their factor does not work so well. This is because they develop antibodies called 'Inhibitors' which destroy the factor in their blood when it is injected. This problem is most common for people with haemophilia A and fortunately developing inhibitors when you have haemophilia B is quite rare.

Having read this book, you hopefully know a little bit more about haemophilia B and in particular how it differs from haemophilia A. If you want to know more, the nurse at the haemophilia clinic will be happy to help.

If you need to know more about haemophilia B, ask at your clinic.

