

Acquired Haemophilia

Advice for people who have been diagnosed
with acquired haemophilia

BAXTER & YOU

Shaping the future of haemophilia together



Introduction

This booklet is designed to give information and advice for people who have been diagnosed with acquired haemophilia and their families. If you, or someone you know is in this position, it is possible that you will not have heard of the condition or know much about it. You may be feeling apprehensive about the future and how the diagnosis could affect your life.

This information booklet will try to answer some of the most common questions and concerns. It is not intended to be a complete guide and individual questions should be directed to your own Haemophilia Centre.



What is acquired haemophilia?

Many people have heard of haemophilia but usually as an inherited blood disorder that affects boys and men from birth. This is sometimes called congenital haemophilia.

Acquired haemophilia shares some similarities with congenital haemophilia but is a distinctly different condition.

Normally, blood clots after injury or surgical procedures by a complicated process involving a number of different proteins. These are mostly made in the liver and then released into the blood.

In congenital haemophilia, boys are born with lower levels of one of these clotting factor proteins, most commonly one called Factor Eight (VIII). This is because they inherit a gene that results in lower levels of this clotting factor.

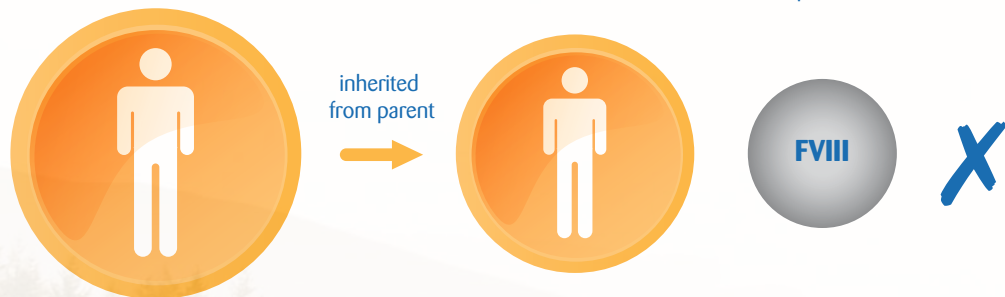
In acquired haemophilia, the gene for Factor VIII production is normal. Instead, the person's immune system suddenly starts to destroy their own Factor VIII as fast as it is made. Because Factor VIII is necessary for blood to clot normally, the person may bleed for no apparent reason. However unlike congenital haemophilia it is not passed on through families (because the gene is normal).

Acquired haemophilia is as likely to affect women as men and can theoretically occur at any age although in practice it seems to affect more elderly people.



Congenital Haemophilia

Factor VIII blood clotting protein not produced



Acquired Haemophilia

Factor VIII blood clotting protein produced



Note: Although most cases of acquired haemophilia are due to destruction of Factor VIII, more rarely other clotting factors, for example Factor IX, or von Willebrand factor are affected. The same general principles and advice apply to these conditions too.

What causes it?

In about half the cases of acquired haemophilia that are diagnosed, the cause is never established. Part of the immune system simply stops working properly.

The immune system (which consists of some of the blood cells, the lymph glands and the spleen) exists to protect the body from attack by bacteria, viruses and other proteins that it regards as foreign. It does this by making proteins known as antibodies to destroy the foreign “invaders”.

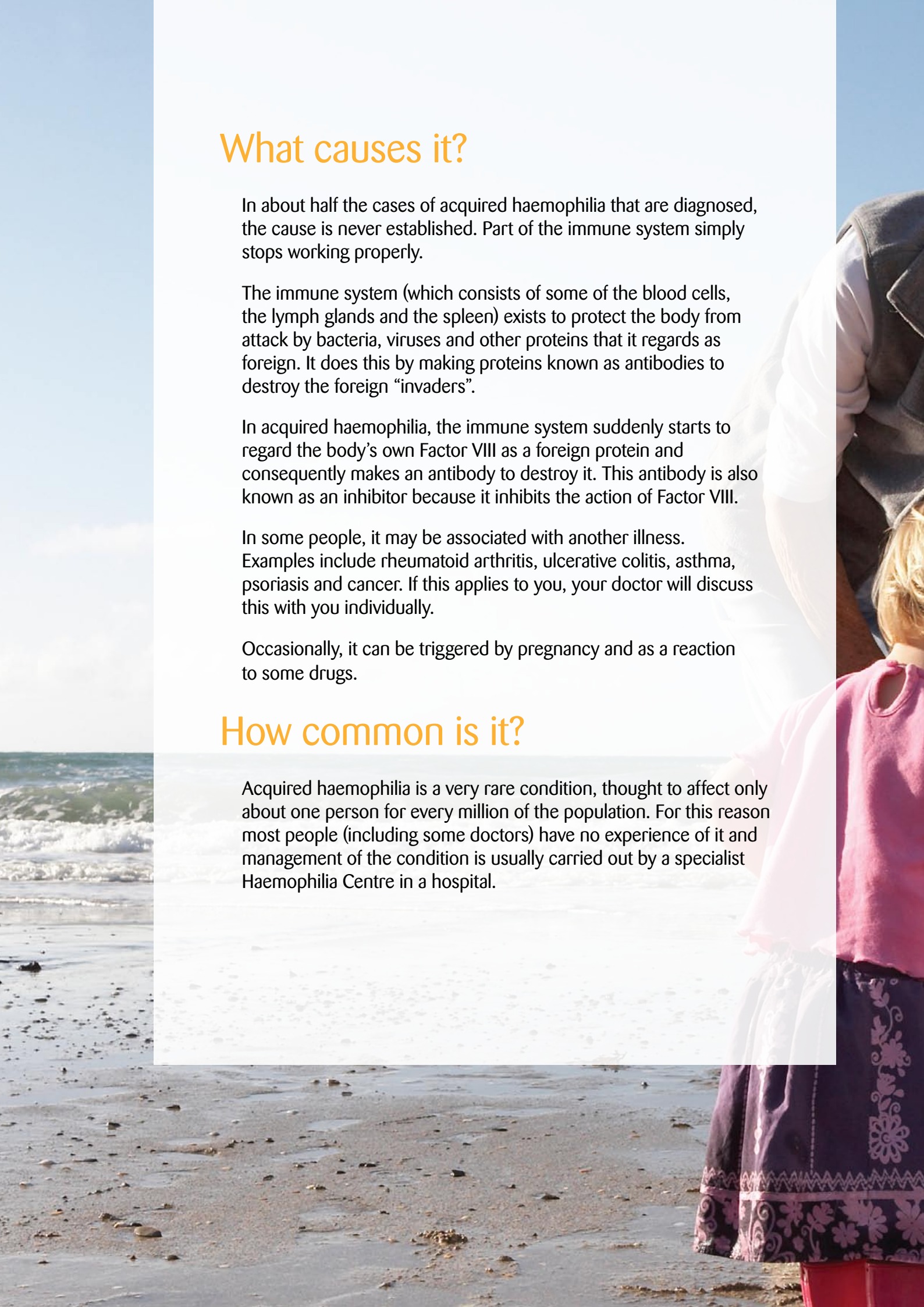
In acquired haemophilia, the immune system suddenly starts to regard the body’s own Factor VIII as a foreign protein and consequently makes an antibody to destroy it. This antibody is also known as an inhibitor because it inhibits the action of Factor VIII.

In some people, it may be associated with another illness. Examples include rheumatoid arthritis, ulcerative colitis, asthma, psoriasis and cancer. If this applies to you, your doctor will discuss this with you individually.

Occasionally, it can be triggered by pregnancy and as a reaction to some drugs.

How common is it?

Acquired haemophilia is a very rare condition, thought to affect only about one person for every million of the population. For this reason most people (including some doctors) have no experience of it and management of the condition is usually carried out by a specialist Haemophilia Centre in a hospital.





How is it diagnosed?

In most cases, the first sign is extensive bruising on the body, usually on the limbs or trunk, that is out of proportion to knocks or injuries received. The bruises may be very large, often extending the whole length of an arm or leg. Although they look alarming they fade with time.

Sometimes, bleeding can occur into muscles, which can be painful and cause temporary loss of movement of that part of the body.

Occasionally, acquired haemophilia may show as prolonged bleeding after a surgical procedure or as blood in the urine or from the bowels.

A simple blood test will show that the body's clotting system isn't working normally but the final diagnosis usually has to be made in a hospital laboratory attached to a Haemophilia Centre.

Is it serious?

Acquired haemophilia can be very serious, although once it is diagnosed a number of treatments are available which are often highly successful.

Occasionally, the antibody (inhibitor) against Factor VIII disappears without treatment but more often a period in hospital is necessary, followed by regular outpatient check-ups.

What is the treatment?

There are several types of treatment available but broadly these fall into two main areas:

- **Stopping immediate bleeding (usually short-term)**
- **Reducing or getting rid of the antibody (inhibitor) that is causing the problem (usually longer-term)**

Your doctor will decide what is the best combination of treatments for you depending on your individual circumstances.

Stopping the bleeding

This is the first priority. There are a number of products available that may be useful. Generally these involve injections of clotting factors directly into the veins, usually through a cannula (or drip). These may have to be given several times a day by doctors or nurses.

There are several types of clotting factors that may be used. Some are prepared from blood plasma donations whilst others are made in the laboratory using a method similar to that used for making many other medicines; these are called “recombinant” products.

Sometimes a drug called desmopressin (DDAVP) may be given if the bleeding is not too severe. This works by causing release of Factor VIII stores from the person’s body. It is given as an injection or in a drip.



A photograph of an older man with grey hair, wearing a grey sweater and light blue trousers, walking on a pebbly beach. He is pointing his right index finger towards the right. The background is a clear blue sky and the ocean.

Getting rid of the antibody

Again, several different treatments are used. Most involve taking tablets, sometimes for many weeks. This regime is usually started in hospital but may be continued after the person goes home.

Sometimes, a course of immunoglobulins (IVIg) is given first through a drip over 2–5 days. If this is successful, drug therapy may be unnecessary.

Occasionally, a technique called plasmapheresis is used which involves passing the patient's blood through a machine to try to filter out the antibody.

Examples of drugs used to eradicate the inhibitor are:

- **Steroids** – usually prednisolone
- **Cytotoxic therapy** – for example cyclophosphamide. Many people have heard of these drugs as treatment for cancer. It is important to stress that acquired haemophilia is not a form of cancer although occasionally people can develop it when they already have cancer.
- **Immunomodulatory therapy** – where drugs are used that prevent the body producing antibodies to clotting factor

Your healthcare team will give you more specific information on the particular treatment that you have been prescribed.

How long might I be in hospital?

This varies depending on how severe the bleeding episode is and the treatment chosen. It is often as much as a few weeks.

How will I know if I am bleeding?

The signs and symptoms you will get will depend on where the bleeding is coming from but will probably include one or more of the following:

- Pain
- Swelling
- Loss of movement in limb
- Obvious bruising
- Blood in the urine
- Blood from the back passage (Red or Black)
- Feeling faint or very tired
- Severe headache

If you experience any of these or any unusual symptoms, contact your Haemophilia centre at once.

Going home

After leaving hospital you will be monitored very carefully in the outpatient department.

The visits will usually include an examination by a doctor and blood tests.

What are the blood tests for?

- To check the inhibitor has not come back if it has been successfully cleared
- To check for any side effects of drug therapy if this is ongoing

You will be provided with the contact numbers for your nearest Haemophilia Centre so you can get advice and treatment at any time.



Is there anything I **should** do?

Contact your Haemophilia Centre immediately if any signs of bleeding or bruising develop or if you get any unusual symptoms. Also, remember that if you are worried about anything, the Centre staff will be able to advise you.

If the inhibitor has not been eliminated, try to protect yourself against knocks, falls and other injuries. Avoid activities which may put you at risk of injury and eat a normal healthy diet.

If you need dental work, contact your Haemophilia Centre for advice in advance as you may need some treatment beforehand.

Is there anything I **shouldn't** do?

Don't take aspirin or non-steroidal anti-inflammatory drugs eg ibuprofen, without specific medical approval.

Don't take other drugs not given to you by your Haemophilia Centre without checking with them first.

Don't have injections into your muscles if the inhibitor has not been eradicated.

And finally:

In at least half of the people affected by acquired haemophilia, treatment is successful, the inhibitor is eliminated and they can go back to leading a normal life. For the remainder, treatment will need to be ongoing. Once the inhibitor is gone, there is very little chance it will come back. The condition cannot be passed on to other members of your family.

Medical knowledge about acquired haemophilia is increasing all the time and new treatments may become available in the future. It is important to be positive.

Remember, if in doubt, ask!

Haemophilia Society contacts

Monday to Friday 9am to 5pm

Tel: 020 7831 1020

Fax: 020 7405 4824

Freephone Helpline: 0800 018 6068

info@haemophilia.org.uk

www.haemophilia.org.uk

Acknowledgements

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