Haemophilia

**Haemophilia is an inherited condition that affects the blood's ability to clot.**

Normally, when you cut yourself, substances in the blood known as clotting factors combine with blood cells called platelets to make the blood sticky. This makes the bleeding stop eventually.

However, in haemophilia, there aren't as many clotting factors as there should be in the blood. This means that someone with the condition bleeds for longer than usual.

The condition is passed to a child by one or both of their parents.

How it affects you

The symptoms of haemophilia can be mild to severe, depending on the level of clotting factors you have. Most cases are mild, but people with severe haemophilia experience symptoms, which require ongoing care.

People with severe haemophilia often experience internal bleeding. This usually occurs around the joints and muscles, causing pain and stiffness. It can also lead to joint damage over time.

Types of haemophilia

The two most common types of haemophilia are haemophilia A and haemophilia B, although haemophilia A accounts for the majority of cases.

Both types have the same symptoms, but they're caused by problems with different clotting factors and have slightly different treatments.

There's also a rarer form of haemophilia called acquired haemophilia. This isn't an inherited condition, but is caused by the immune system (the body's natural defence against infection and illness) attacking the clotting factors in the blood.

Who is affected?

There are about 6,000 people with haemophilia in the UK. Most of these are males because of the way the condition is inherited.

Worldwide, it's estimated that one boy in every 5,000 will be born with haemophilia A and one boy in every 30,000 will be born with haemophilia B.

However, some females who carry the haemophilia gene may also experience some bleeding problems, such as [heavy periods](http://www.nhs.uk/conditions/Periods-heavy/Pages/Introduction.aspx). There's also the chance that a girl could be born with haemophilia if both parents have the haemophilia gene.

If you have a family history of haemophilia, you can have tests before, during and after pregnancy to determine if your child has the condition. If there's no family history, haemophilia tends to be diagnosed when symptoms appear in childhood.

Treating haemophilia

Although there's no cure for haemophilia, treatment usually allows a person with the condition to enjoy a good quality of life.

In recent decades, genetically engineered clotting factor medications have been developed to prevent and treat prolonged bleeding.

These medications are given as an injection, the timing of which depends on how severe the condition is. Injections are usually only given in milder cases in response to prolonged bleeding, whereas more severe cases are treated with regular injections to prevent bleeding.

Symptoms of haemophilia

**The symptoms of haemophilia vary, depending on how severe the condition is, but the main sign is prolonged bleeding. The bleeding may occur spontaneously or after a medical procedure.**

The severity of the condition is determined by the level of clotting factors in the blood:

* **mild haemophilia** – where someone has between 5 and 50% of the normal amount of clotting factors
* **moderate haemophilia** – where someone has between 1 and 5% of the normal amount of clotting factors
* **severe haemophilia** – where someone has less than 1% of the normal amount of clotting factors

Most cases of haemophilia are severe.

Mild haemophilia

Children born with mild haemophilia may not have any symptoms for many years. The condition usually only becomes apparent after a significant wound, surgery, or a dental procedure such as having a tooth removed. These events could cause unusually prolonged bleeding.

Moderate haemophilia

As well as the above, children born with moderate haemophilia bruise easily. They may also have symptoms of internal bleeding around their joints, particularly if they have a knock or a fall that affects their joints. This is known as a joint bleed.

The symptoms usually begin with a tingling feeling of irritation and mild pain in the affected joint – most commonly the ankle joints, knee joints and elbow joints. Less commonly, the shoulder, wrist and hip joints can also be affected.

If a joint bleed isn't treated, it can lead to:

* more severe joint pain
* stiffness
* the site of the bleed becoming hot, swollen and tender

Severe haemophilia

The symptoms of severe haemophilia are similar to those of moderate haemophilia. However, joint bleeding is more frequent and severe.

Children with severe haemophilia have spontaneous bleeding. This means they start bleeding for no apparent reason. Spontaneous bleeding can take the form of nosebleeds, bleeding gums, joint bleeds and muscle bleeding.

Without treatment, people with severe haemophilia can develop:

* joint deformity – which may require replacement surgery
* soft tissue bleeding – which could lead to further complications
* serious internal bleeding

When to seek emergency medical help

There's a small risk of bleeding inside the skull, known as an intracranial haemorrhage. It's estimated that 3% of people with moderate or severe haemophilia will have an intracranial haemorrhage. However, spontaneous bleeding inside the skull is uncommon and is usually only caused by a [head injury](http://www.nhs.uk/conditions/head-injury-severe-/pages/introduction.aspx).

Bleeding in the skull should be treated as a medical emergency.

The symptoms of an intracranial haemorrhage include:

* severe headache
* stiff neck
* vomiting
* a change in mental state – such as confusion
* speaking difficulties – such as slurred speech
* changes in vision – such as [double vision](http://www.nhs.uk/conditions/double-vision/Pages/Introduction.aspx)
* loss of co-ordination and balance
* [paralysis](http://www.nhs.uk/conditions/paralysis/pages/causes.aspx) of some or all of the facial muscles

Call 999 for an ambulance if you suspect that someone is bleeding inside the skull.