

This factsheet is about a bleeding disorder that is related to problems with a blood clotting factor called factor V (five). It is written to go with our **Rare bleeding disorders booklet**, where you will find much more information on living with one of these conditions.

What is factor V deficiency?

It is a bleeding disorder caused by the body producing less of a particular clotting factor than it should. This causes problems because the clotting reaction that would normally control any bleeding is blocked too early. So, your body does not make the blood clots it needs to stop bleeding.

Factor V deficiency is very rare. Doctors estimate that it affects about one in a million people worldwide.

What causes factor V deficiency?

This is an inherited genetic disorder. It is what is known as recessive, meaning you must inherit the gene defect from both parents to have the condition. It affects men and women equally.

If you carry one copy of the gene fault for factor V deficiency, you are known as a carrier. You can only pass the condition on to your children if your partner also carries the gene fault. Any children

that inherit the gene fault from you will also be carriers of the condition. Carriers will not actually have the condition but could have slightly lower than normal factor V levels, which can cause more minor symptoms.

As Factor V is autosomal recessive, if you inherit the faulty gene from both parents you will develop the severe form of the disease. Autosomal recessive disorders are more common in areas of the world where marriage between close relatives is common.

Our Rare bleeding disorders booklet has more information about how bleeding disorders are inherited.

It is also possible to develop a factor V deficiency later in life. This is called acquired factor V deficiency. This is rare but is most often caused by exposure to anti-clotting drugs during heart or brain surgery. It has also been found in a very small number of people who have had rheumatological conditions, cancer or treatment with some antibiotics.

Symptoms of factor V deficiency

The symptoms of factor V deficiency can vary in severity, even between members of the same family. This depends on how much factor V your body makes. The less factor V you have in

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your blood, the more often you are likely to have symptoms and the more severe they will be. Symptoms can be so mild that they cause no problems.

Common symptoms include:

- easy bruising
- nosebleeds
- bleeding gums
- heavy periods that last longer than normal (menorrhagia)
- bleeding after dental work, surgery or an accident.

You may also have these symptoms, but they are less likely:

- bleeding from the umbilical cord stump at birth
- bleeding into joints or muscles
- bleeding into the bladder or kidneys (genitourinary system)
- bleeding into the brain or spinal cord (central nervous system).

It is very important that you contact your doctor or haemophilia centre if you think you or your child are having a bleed.

Some bleeds can cause long-term problems if they are not properly treated, such as bleeds into muscles or joints. And some can be life-threatening, such as a bleed into the brain or spinal cord. If you have frequent bleeds, such as nosebleeds, you can become anaemic.

This means you have low haemoglobin levels because of the frequent loss of blood. Anaemia can make you feel tired and breathless.

There is more information in our **Rare bleeding disorders booklet** about how to spot the different types of bleeds.

If you are a carrier of the gene for factor V deficiency, you may have minor symptoms, such as bleeding for longer than normal after having a tooth or your tonsils out.

Diagnosing factor V

Factor V deficiency is diagnosed with blood tests. These include tests to measure how quickly your blood clots. They are specialised tests, so you need to have them done at a haemophilia centre. The level of factor V in the blood is checked, as well as the level of factor VIII, as these sometimes occur in combination, resulting in different bleeding. The gene mutation will also be identified as this can be helpful for planning pregnancies and for testing other family members.

Factor V deficiency can also be diagnosed before birth (prenatally) if there is a family history. There are several options for this including chorionic villus sampling (CVS) early in pregnancy or amniocentesis at around 15 to 20 weeks.

Factor V deficiency is usually diagnosed at a young age, because of a bleed, with many people being diagnosed in infancy or early childhood. If you have the condition mildly though, it may not show up until you have bleeding, for example after dental work or surgery.

Treatment for factor V

Generally, people with factor V deficiency only have treatment if they have a bleed or before planned surgery. In very rare cases, where you have a history of severe bleeding or very low factor V levels, your doctor may suggest regular treatment to prevent bleeds. Your doctor may call this prophylaxis (pronounced proff-ill-ax-iss).

For a nosebleed or other soft tissue bleed, you are most likely to have treatment with tranexamic acid (Cyklokapron) tablets. These work by stopping the breakdown of blood clots. You may also need to take these if you have heavy periods. Or your doctor may suggest that you take the contraceptive pill or have an intra-uterine device (IUD) inserted to make your periods lighter.

Replacement factor V concentrate is not yet available but is being developed. So treatment for a more serious bleed is usually with fresh frozen plasma (FFP), which contains all the clotting factors, including factor V. This is made from human blood – plasma is the straw-coloured fluid that the blood cells are carried in. You have this treatment through a drip into a vein (intravenously).

All blood products are now treated during manufacture to kill off any known viral infections such as hepatitis and HIV.

How often you need treatment will depend on how severe your condition is. But if you have a severe bleed, you may need daily treatment for a while, as factor V only lasts in the body for a few hours.

In the rare situations where people need treatment to prevent bleeding, your doctor is likely to suggest that you have FFP, which you have through a drip. You have this treatment regularly enough to keep your blood levels stable, which may be as often as every two days.

If you are having a baby, you may have FFP when you go into labour and for the next few days after your baby is delivered.

You should not use Non-Steroidal Anti-Inflammatory Drugs (NSAIDs such as ibuprofen) as this increases the risk of bleeding. Other methods of pain relief should be used instead. Speak to your doctor if you are unsure.

You should have immunisations or other injections subcutaneously (under the skin) rather than intramuscularly (into a muscle) to reduce the risk of a painful bruised swelling (haematoma) developing.

Coping with your condition

Finding out that you or your child has a bleeding disorder can be upsetting and bring on a range of different emotions. Of course, this will take time to accept. Finding out as much as you can about your condition can help you learn to cope with it.

How much your bleeding disorder affects your daily life will depend on how severe it is. For many people, it won't have much effect at all. It may only be an issue if you are having dental work, major surgery, are having a baby or have an accident.

Others may need treatment from time to time, to treat minor or more serious bleeding. A small number may need regular treatment to prevent bleeding.

Do find out as much as you can about how to prevent bleeding and when it is likely to cause a problem. Our **Rare bleeding disorders booklet** has a lot of information about what to look out for and precautions you can take to keep yourself healthy.

There is information on:

- ways to make bleeding less likely
- how to spot the early signs of a bleed
- dental care
- carrying medical information with you
- information for girls and women about problems with periods and pregnancy.

If you have factor V deficiency you may need to avoid some activities, such as contact sports that carry a high risk of head injury. Most day-to-day activities will cause few problems. Speak to your local haemophilia centre for advice.

If you have factor V deficiency you will need lifelong monitoring and treatment. As a child approaches their teenage years, they will start the transition process to move on to adult health services. This is a planned process so that they become more independent as they grow older and able to manage their own health.

A new diagnosis can feel scary or overwhelming but there's lots of great support available.