THE HAEMOPHILIA SOCIETY

FACTSHEET

Factor VII deficiency

This factsheet is about a bleeding disorder that is related to problems with a blood clotting factor called factor VII (seven). 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 VII deficiency?

Factor VII deficiency 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 doesn't make the blood clots it needs to stop bleeding.

Factor VII deficiency is rare but is the commonest of the rare bleeding disorders.

Doctors estimate that it affects about one in 500,000 people.

The severity of symptoms ranges from mild to severe depending on the amount of factor VII present in the blood and its activity.

What causes factor VII deficiency?

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

If you carry one copy of the gene fault for factor VII 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. Carriers will not actually have the condition, though they may have lower than normal factor VII levels. About one in five carriers will have some symptoms.

It is also possible to develop a factor VII deficiency later in life. This is called acquired factor VII deficiency. This is rare and is most often caused by liver disease. Even more rarely, it can be caused by other conditions such as cancer, infection or after a bone marrow transplant.

For everyone affected by a genetic bleeding disorder

To find out more, visit haemophilia.org.uk or contact us on 020 7939 0780 or at info@haemophilia.org.uk

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Symptoms of factor VII deficiency

Symptoms of factor VII deficiency are very variable. Unlike other bleeding conditions, the severity of your symptoms does not seem to be entirely linked to the level of factor VII in your blood. Some people have no symptoms at all, others have minor symptoms, and some have very severe symptoms.

People with minor symptoms may have:

- bleeding after dental work
- bleeding at circumcision
- bleeding gums
- easy bruising
- heavy periods or periods that last longer than usual (menorrhagia)
- nosebleeds
- signs of blood in the urine.

People with severe symptoms may also have bleeding:

- from the umbilical cord stump at birth into joints
- into muscles
- into the gut (gastrointestinal system)
- into the brain or spinal cord (central nervous system)
- after childbirth (post-partum haemorrhage)
- bleeding in the head (newborns).

The age at which symptoms appear varies depending on the amount of Factor VII in the blood and how well it is working. Children with little or no factor VII may start to show symptoms soon after birth.

Some bleeds can cause long-term problems if they are not properly treated, such as bleeds into muscles or joints. Occasionally some can be life-threatening, such as a bleed into the brain or spinal cord.

A bleed into the brain may show as a headache that keeps getting worse, sickness, confusion and increasing drowsiness.

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

Diagnosing factor VII deficiency

Factor VII deficiency is diagnosed with blood tests. These include tests to measure how quickly your blood clots and tests to measure your factor VII levels. These are specialised tests, so you need to have them done at a haemophilia treatment centre.

People with factor VII deficiency that causes severe symptoms are generally diagnosed soon after birth, from cord bleeding or bleeding after circumcision. If you have the condition with milder symptoms, it may be diagnosed in young childhood. If you have the condition with no symptoms, it may only be picked up during a family study because someone else also has it, or during routine blood tests, for example before an operation.

Treatment for factor VII deficiency

The main treatment is factor VII concentrate. Generally, you only have this after a bleed. But some children and adults with severe symptoms have treatment three to four times a week to prevent serious bleeds. Your doctor may call this prophylaxis (pronounced proff-ill-ax-iss).

If you have mild symptoms, your doctor may suggest you take tablets before minor surgery or dental work. This is usually a drug called tranexamic acid (Cyklokapron), which helps to stop clots breaking down.

If you have a more serious bleed, you will need treatment with factor VII concentrate. This can now be made in the laboratory and is called recombinant factor replacement. As it doesn't have to be purified from donated human blood, it means there is no risk of infection. You have this treatment as an injection into a vein (intravenously). As an alternative to recombinant factor replacement, you might be given plasma-derived factor VII which stays in the blood for slightly longer than recombinant factor VII.

You may also have factor VII concentrate to prevent bleeding if you are going to have major surgery. You are most likely to have one dose before your operation and at least two afterwards.

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 will not 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:

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

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