Factor II deficiency

This factsheet is about a bleeding disorder related to problems with a blood-clotting factor called prothrombin or factor II (pronounced factor 2). 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 II deficiency?

Factor II is also called prothrombin, so you may hear this condition called prothrombin deficiency. It's a bleeding disorder that happens because either:

- your body produces less prothrom bin than it should, or
- the prothrombin your body makes does not work properly.

This causes problems because the clotting reaction (how the body usually reacts to the stop any bleeding) is blocked too early. So your body doesn't make the blood clots it needs to stop bleeding. Factor II deficiency is very rare. Doctors estimate that it only affects about one in every two million people.

What causes factor II deficiency?

Factor II deficiency 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 and can be found in all ethnic groups. It is frequently found in areas of the world where marriage between close relatives is common.

If you carry one copy of the gene fault for factor II 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 prothrombin levels, which can cause more minor symptoms.

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

Getting factor II deficiency later in life

It is also possible to develop a factor II deficiency later in life. This is called acquired factor II deficiency. It may be temporary and can be caused by:

- a lack of vitamin K
- some antibiotics or diseases that can affect how well things are absorbed in the gut,

or

- by liver disease

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Symptoms of factor II deficiency

The symptoms of factor II deficiency can vary a lot. They depend on how much prothrombin your body can make. The less prothrombin you have in your blood, the more often you are likely to have symptoms and the more severe they will be.

Common symptoms include:

- bleeding when the umbilical cord is at separated
- bleeding after circumcision
- bleeding during or after surgery
- bleeding in the mouth, particularly after dental surgery
- bleeding into joints
- bleeding into muscles
- easy bruising
- heavy periods or periods that last longer than normal (also known as menorrhagia)
- nosebleeds.

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

- Heavier than usual bleeding during or after injury or childbirth
- bleeding in the gut (also known as gastrointestinal haemorrhage)
- bleeding into the brain or spinal cord (the central nervous system).

These symptoms are rare:

- bleeding in the central nervous system (the brain and spinal cord)
- blood in urine (haematuria).

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 aren't 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 a low haemoglobin level because of the frequent loss of blood. Anaemia can make you feel tired and breathless. About half of all those with factor II deficiency also have anaemia.

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

Diagnosing factor II deficiency

Factor II deficiency is diagnosed with blood tests. If your baby had bleeding from the umbilical cord, or when they were circumcised, your doctor may suggest these blood tests at birth.

The blood tests are usually done at a haemophilia centre as the tests that are needed are very specialised. Once your blood has been taken it is sent off to the lab for testing straight away.

The lab will test your blood for lots of different things, including how quickly your blood clots and a specific test that measures the amount of prothrombin in your blood.

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Once they have your results, your haemophilia centre or your doctor will be in touch to talk you through what they found.

To find out if you or your children are carriers of the gene fault, you need to have DNA testing.

Treatment for factor II deficiency

How often you need to have treatment will depend on how severe your condition is.

You will probably only need treatment if you have a bleed. But very rarely, people with severe factor II deficiency may need regular treatment to prevent bleeds.

Your doctor may suggest that you take tranexamic acid tablets (Cyklokapron) for minor bleeding or before minor surgery. These work by stopping the breakdown of blood clots.

If you have very heavy periods, your doctor may suggest that you take the contraceptive pill to lighten them. Or you may take tranexamic acid tablets during your periods.

There are two treatments available for more serious bleeding in factor II deficiency:

- prothrombin complex concentrate
- (PCC) contains factor II along with two or three other clotting factors
- fresh frozen plasma (FFP).

Both are made from donated human blood from plasma, the straw-coloured fluid that the blood cells are carried in. You have these through a drip into a vein (intravenously).

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

You may also have treatment with PCC before major surgery or, if you are having a baby, when you go into labour and for a few days after your baby is born.

If you have very low prothrombin levels, you may need regular treatment with PCC to prevent bleeds. Your doctor may call this prophylaxis (pronounced proff-ill-ax-iss). It is usually possible to teach you to self-treat or treat your child at home.

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.

Caution is needed for injections as well – immunisations should be given subcutaneously (under the skin) rather than intramuscularly (into a muscle) to reduce the risk of a painful bruised swelling (haematoma) developing.

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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.

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 and waysto make bleeding less likely

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