Factor XI deficiency

This factsheet is about a bleeding disorder that is related to problems with a blood clotting factor called factor XI (pronounced factor 11). 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 XI deficiency?

Factor XI deficiency was also sometimes called haemophilia C (pronounced heem-oh-fill-ee-ah) but has a very different pattern of bleeding and should not be confused with haemophilia A and B. It is a bleeding disorder caused by the body producing less of a 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 XI is important for producing thrombin protein that converts fibrinogen to fibrin during the clotting process.

Factor XI deficiency is rare. Doctors estimate that it affects about one in a million people. But it is much more common in some populations, including the Ashkenazi Jewish community where it affects up to one in 450 people.

What causes factor XI deficiency?

This is an inherited genetic disorder. It affects men and women equally.

It is generally what is known as recessive, meaning you must inherit the gene fault from both parents to have the disorder in a severe form.

If you carry one copy of the gene fault for factor XI deficiency, you do not have the disorder yourself but are a carrier. You can only pass on a severe form of the condition 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.

Although carriers will not have a severe form of the condition, they can have slightly lower than normal factor XI levels. This means they may be at risk of bleeding after surgery, following dental extractions or of having heavy periods.

It is also possible to develop a factor XI deficiency later in life. This is called acquired factor XI deficiency and is generally caused by liver disease.

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Symptoms of factor XI deficiency

Most people with factor XI deficiency don't have any symptoms. In those who do, the commonest symptoms are bleeding after surgery or an accident. Spontaneous bleeding is very uncommon, but some people have nosebleeds or bruise or more easily than normal.

A bleed is most likely after dental surgery or an operation on your mouth, throat, bladder, kidneys or prostate.

Carriers of factor XI deficiency are also at risk of bleeding after surgery. If you have factor XI deficiency or are a carrier, doctors recommend that you do not take non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen or aspirin as these could increase the risk.

Heavy periods are common in women with factor XI deficiency. They are also more at risk of bleeding after having a baby (a postpartum haemorrhage). 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 XI

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

Your doctor may suggest these blood tests at birth, because your baby had bleeding after a circumcision was done or because you have a family history of factor XI deficiency.

It is also quite common for factor XI deficiency to be diagnosed later in life, because of heavy periods or other bleeding symptoms – after dental work, surgery or an accident.

Treatment for factor XI

Most people with factor XI deficiency never need treatment. You are only likely to need it if you have an accident or to prevent bleeding before planned surgery or if you are having a baby.

For minor operations, you may have tranexamic acid tablets (Cyklokapron) to take beforehand and for a few days afterwards. These help to stop the breakdown of blood clots. If you have very heavy periods, you may have tranexamic acid tablets during your periods. Or your doctor may suggest that you take the contraceptive pill to make your periods lighter.

For more serious operations you may have treatment with fresh frozen plasma (FFP), which is often used to treat rare bleeding disorders. It contains all the clotting factors, including factor XI. This is made from human blood – plasma is

the straw-coloured fluid that the blood cells are carried in. You have it through a drip into a vein (intravenously).

Factor XI concentrate is available and is another possible treatment, but many doctors prefer to use FFP as factor XI concentrate can cause problems with blood clots.

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

If you are having a baby, whether you have treatment to prevent bleeding or not will depend on the usual factor XI level in your blood. Most women will not need any treatment.

If you have a low factor XI level, you may have tranexamic acid tablets to take when you go into labour. Or you may have factor XI concentrate.

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 most people with factor XI deficiency, 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.

Factor XI deficiency will require lifelong monitoring and treatment. As your child gets near to their teenage years, your haemophilia centre will start to talk to them about getting ready to move on to adult health services.

This is a slow process so that they become more independent as they grow older and more able to manage their own health.

It is possible to have genetic counselling before planning a family, both for affected individuals and unaffected carriers. You can discuss this with your haemophilia centre.

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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.