THE HAEMOPHILIA SOCIETY

FACTSHEET

Factor XIII deficiency

This factsheet is about a bleeding disorder that is related to problems with a blood clotting factor called factor XIII (pronounced factor 13). 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 XIII deficiency?

Factor XIII deficiency 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 XIII deficiency is one of the rarest types of clotting disorder. Doctors estimate that it affects about one in every two million people. Factor XIII plays an important role in wound healing, pregnancy and formation of new blood vessels, though more research is needed to better understand this.

What causes factor XIII deficiency?

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

If you carry one copy of the gene fault for factor XIII 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. You will not have the condition yourself, but any children that inherit the gene fault from you will also be carriers of the condition.

It is also possible to develop factor XIII deficiency later in life. This is called acquired factor XIII deficiency. It can be caused by liver disease, some types of leukaemia, inflammatory bowel disease and an auto-immune disease called systemic lupus erythematosus.

Symptoms of factor XIII deficiency

Often, the first clinical sign of inherited factor XIII deficiency is a few days after birth or when the umbilical cord separates. Severity of symptoms ranges from mild to severe depending on the amount of factor XIII in the blood and its activity. Most people with factor XIII deficiency have some bleeding.

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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The commonest symptoms are:

- bleeding after surgery
- bleeding from the umbilical cord stump at birth
- bleeding into muscles or joints
- bleeding into the brain or spinal cord (central nervous system).
- easy bruising
- heavy periods that last longer than normal
- poor wound healing.
- Less common symptoms are:
- bleeding in the gut (gastrointestinal system).
- bleeding in the urinary system
- nosebleeds or bleeding into the mouth

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 XIII

Factor XIII deficiency is diagnosed with a specialised blood test to measure factor XIII activity. The usual clotting tests come out as normal with this particular disorder, so you do need to have the specialised test, which has to be done at a haemophilia centre.

Most people are diagnosed early in life, either at birth after bleeding from the umbilical cord stump or because of a bleed in childhood. Or your baby may be tested at birth because factor XIII deficiency runs in your family.

Treatment for factor XIII

Because bleeding can be common with factor XIII deficiency, many people have regular factor replacement to prevent bleeding from birth or as soon as they are diagnosed. Your doctor may call this prophylaxis (pronounced proff-ill-ax-is). You will need preventative treatment if:

- you have had bleeding episodes
- members of your family with the condition have a history or bleeds
- you have very low factor XIII activity.

This is given regularly as an injection, sometimes into a central venous access device such as an implantable port. Injections can be given at home which is less disruptive to family life.

Development of ‘inhibitors’ that mean the body fights off factor XIII injections can be a problem for people with factor XIII deficiency, though it’s extremely rare. This will be checked regularly at review appointments. If inhibitors develop, additional injections will be needed.

If you have mild bleeding, such as after dental surgery or a minor operation, your doctor

may ask you to take tranexamic acid tablets (Cyklokapron) as well as your monthly preventative treatment. If you have more serious bleeding or major surgery, you may need extra doses of factor XIII replacement.

It is very important to keep factor XIII levels up during pregnancy, so your doctor may suggest you increase your preventative treatment to fortnightly or three-weekly as soon as you know you are pregnant. You may also need extra doses when you go into labour.

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.

Any surgery (including dental) will need careful planning in advance, so it is important that all healthcare professionals involved are aware of the condition.

Factor XIII 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.

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.

Regular follow-up at a specialist centre is necessary to check that you or your child are responding to treatment and not experiencing any side effects. Regular check-ups with a dentist are also required.

How much your bleeding disorder affects your daily life will depend on how severe it is. Many people with factor XIII deficiency need regular treatment to prevent bleeding. You may need extra treatment for minor or more serious bleeds, or if you are going to have surgery.

Women with severe factor XIII deficiency should receive regular factor replacement throughout pregnancy to reduce the risk of early miscarriage.

Contact sports, which carry a high risk of head injury, need to be avoided but most day-to-day activities will cause few problems. Your haemophilia centre can help you choose activ-ities that are right for you or your child.

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.