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# Fibrinogendeficiency

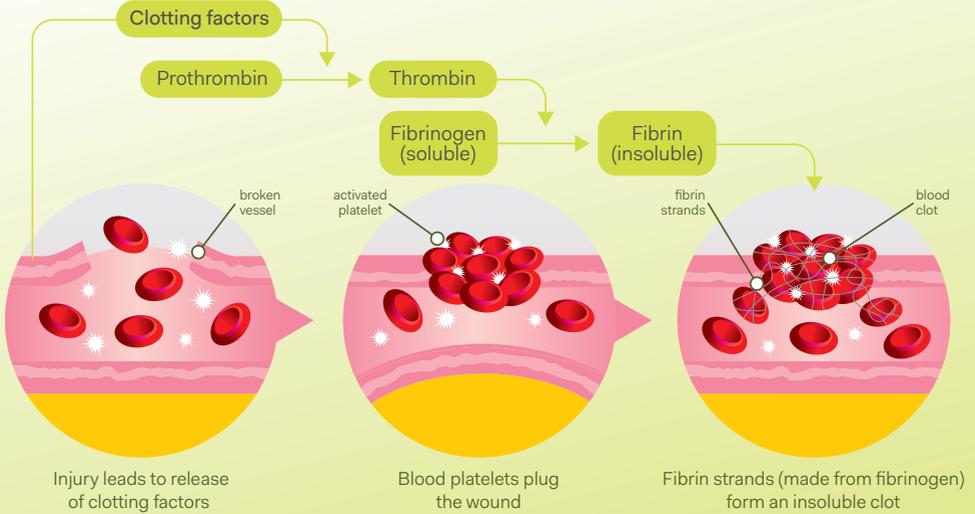
# The role of fibrinogen

Fibrinogen (or factor I) is one of the blood's clotting factors. Clotting factors help the body to control bleeding through a process called coagulation (clotting).

When the body is injured, a series of reactions occur and clotting factors work together to form a blood clot.

A particular clotting factor called fibrinogen plays an important role during the process. It is converted to a protein called fibrin, which forms a mesh trapping platelets to produce a clot.

# The normal process of clotting after an injury



## The problem of fibrinogen deficiency

When the body does not produce enough fibrinogen, or the fibrinogen that is produced does not work properly, this is known as fibrinogen deficiency.

Without fibrinogen, the clotting process is disrupted and clots cannot form properly. This can cause excessive bleeding, or blood clots inside blood vessels.

# Causes and characteristics of fibrinogen deficiency

Congenital fibrinogen deficiency is a very rare, inherited condition that affects about 1 in a million people. It affects men and women equally.

## Inherited deficiency

There are three types of fibrinogen deficiency:

1. Afibrinogenaemia – when the body does not produce any fibrinogen
  - A *recessive* condition, meaning both parents need to carry the specific faulty gene in order to pass it onto their child
2. Hypofibrinogenaemia – when the body has low amounts of fibrinogen, but not enough for normal clotting
  - A *dominant* condition, so only one parent needs to pass on the specific faulty gene to their child
3. Dysfibrinogenaemia – when the body produces enough fibrinogen, but it does not work properly
  - Can be inherited through either a dominant or recessive gene

Inherited fibrinogen deficiency results in a diverse range of bleeding issues in different patients.

## Acquired deficiency

More commonly, fibrinogen deficiency can be acquired rather than inherited. This could be the result of huge blood loss that reduces the amount of fibrinogen in the body – following significant physical injury, major surgery or an infection like sepsis. It could also be due to dilution of blood as a result of some medical treatments.

Fibrinogen deficiency can be inherited or acquired

# Recognising the signs of fibrinogen deficiency

The characteristics of fibrinogen deficiency depend on the form and the severity of the condition. As a general rule, the less fibrinogen that is in the blood, the more frequent and/or severe symptoms you will have.

The three different types of fibrinogen deficiency may be recognised at different stages of life.

# Afibrinogenaemia and hypofibrinogenaemia

These conditions are usually diagnosed at birth, following excessive bleeding from the umbilical cord or after circumcision. However, symptoms may appear later in life and can vary from mild to severe. Both afibrinogenaemia and hypofibrinogenaemia present with similar symptoms, although the severity can vary depending on fibrinogen levels. Patients may present with different symptoms, which include:

- Heavy bleeding during or after injury, surgery, or childbirth
- Bleeding of the mouth (particularly after dental surgery)
- Complications with pregnancy, including spontaneous miscarriages
- Long, heavy menstrual periods
- Joint and muscle bleeds
- Nosebleeds
- Abnormal thrombotic events (formation of blood clots)
- Abnormal bleeding after circumcision or cutting of the umbilical cord
- Easy bruising

## Dysfibrinogenaemia

Symptoms of dysfibrinogenaemia depend on how well the fibrinogen is functioning.

Some people have no symptoms at all, some will experience similar bleeding characteristics to afibrinogenaemia, and others will experience abnormal clotting in the blood vessels.

Some people may initially be asymptomatic, but they can develop symptoms in later years depending on their fibrinogen levels

# Diagnosing fibrinogen deficiency

A variety of blood tests can identify fibrinogen deficiency. These can include a specific test measuring the level of fibrinogen in the blood, and/or the time it takes for blood to clot.

If your doctor suspects that you have inherited fibrinogen deficiency, they may look at the genetic sequence that makes up fibrinogen to see why yours is different. If there is a chance that your children might also be fibrinogen deficient, this information can help their doctor treat them in the future.

Sometimes other diseases that also cause bleeding problems may need to be ruled out before a diagnosis of fibrinogen deficiency can be made.

## Treatments for fibrinogen deficiency

To control or prevent bleeding in people with fibrinogen deficiency, the missing fibrinogen in the blood may need to be replaced.

Fibrinogen concentrate is the first choice to replace fibrinogen, as it provides a consistent and defined concentration of fibrinogen. The amount needed will be determined by your doctor.

You may be treated with fibrinogen concentrate during major surgery or following severe bleeding, or you may be given weekly doses of fibrinogen as a preventative measure. If you are pregnant, your doctor may recommend that you take fibrinogen concentrate as a preventative measure throughout your pregnancy.

If fibrinogen concentrate is not available, less pure products such as cryoprecipitate or fresh frozen plasma may be used instead.

Your doctor will be able to give you more detailed information about any treatments you may be given

# Getting the right treatment for fibrinogen deficiency

- Fibrinogen deficiency is a rare bleeding disorder that means the blood is unable to clot properly, and may lead to uncontrolled bleeding
- There are three different inherited types, each of which can be confirmed with a blood test
- There are three possible types of treatment, but fibrinogen concentrate is the preferred treatment
- Acquired deficiency can also occur after severe blood loss

For further information about  
fibrinogen deficiency:

- Visit the World Federation of Hemophilia website: [www.wfh.org](http://www.wfh.org)
- Visit [www.haemophilia.org.uk](http://www.haemophilia.org.uk)
- Refer to any treatment booklets your doctor or nurse may have given you
- Talk to your doctor or nurse



Further information available at:

<http://www.haemophilia.org.uk>

<http://www.wfh.org>

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