

Inherited platelet function disorders

Information for patients and carers from the Haemophilia Centre

What are platelets and what do they do?

Platelets are small cell fragments that circulate in your blood. When a blood vessel is damaged and blood leaks out, platelets rush to the site of the bleeding and form a temporary plug.

Platelets work by:

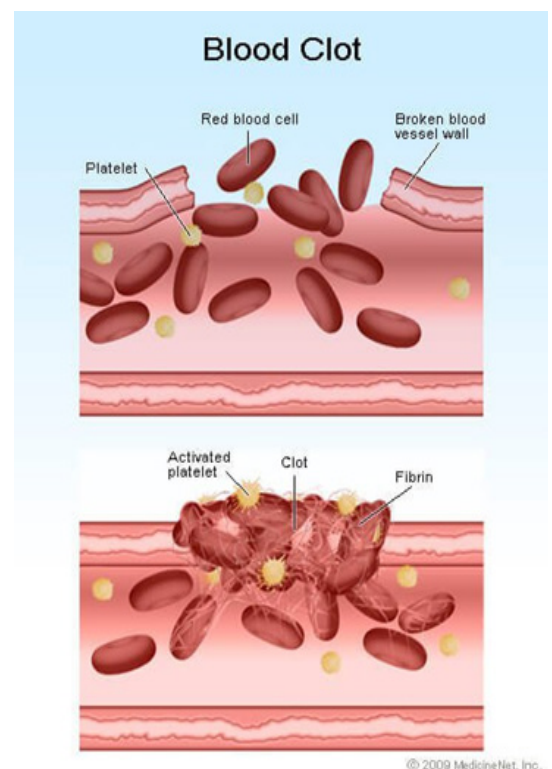
- sticking to the injured blood vessel wall to block the leak
- releasing chemicals that make more platelets gather at the site of injury; and then
- sticking together so they can form a plug.

The diagram shows how the plug is formed to stop the bleeding and the fibrin is made to make the plug that has made a more stable blood clot.

What are platelet function disorders?

Platelet function disorders are conditions where your platelets do not work the way they should, which means there is an increased risk of bleeding or bruising. Since the platelet plug does not form properly, bleeding can continue for longer. You should not bleed any faster, however you may bleed for longer than expected.

Having a platelet function disorder should not affect your everyday life. Most of the bleeding problems tend to happen after an injury, surgery, or dental treatment.



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How do you get a platelet function disorder?

Platelet function disorders are a group of inherited conditions, you are born with them. Both men and women can inherit them from their parents.

One set of our genes is inherited from our mother and the other set from our father. Both parents must carry the defective gene and pass it on for their child to have a more severe platelet function disorder.

How are platelet function disorders diagnosed?

Platelet function disorders are sometimes difficult to diagnose. This is done by a blood test, which will usually need to be repeated to confirm an abnormal result. The blood test involves separating out the platelets from the blood sample taken, adding substances to stimulate the platelets to stick together, and then recording the effects. How the platelets respond to a particular substance can give an indication of the type of platelet disorder a person has.

What are the symptoms of a platelet function disorder?

Most people with a platelet function disorder will have mild symptoms, which include the following.

- For women and girls, heavy and prolonged bleeding during menstruation (known as menorrhagia)
- Nose bleeds
- Bleeding from gums
- Easy bruising
- Bleeding after surgery
- Bleeding after trauma or injury
- Bleeding in the mother at childbirth.

How are platelet function disorders treated?

Day-to-day treatment is not needed for platelet function disorders. It may however be necessary before and after surgery, dentistry, or trauma.

The need for treatment will depend on:

- how severe your bleeding is
- the type of surgery or dental treatment you are having
- your previous history of bleeding; and
- your family's history of bleeding.

What are the treatments for platelet function disorders?

- Antifibrinolytic Agents (Tranexamic Acid)
- DDAVP (Octim)
- Platelet transfusion

- **Antifibrinolytic Agents (Tranexamic Acid)**

Tranexamic Acid works by stopping the early breakdown of a clot that has been made after injury to your blood vessel. Fibrin is a protein that gives the blood clot stability. Tranexamic Acid stops the substances that destroy the fibrin within the clot.

Often it is the only treatment needed for some people with platelet disorders and for some procedures. It is particularly useful for mouth bleeding and therefore very good preventative treatment when you are having a dental procedure.

This treatment is usually given as tablets. You may be asked to crush the tablets in a small amount of water and swish it around your mouth, before either swallowing it or spitting it out (you will be advised by the Haemophilia Centre which is best for you).

Tranexamic Acid is not used when there is blood in your urine, as small clots can occur which can then block your urinary tract and cause you pain.

Are there any side effects?

Side effects are rare but include:

- nausea (feeling sick) and vomiting
- diarrhoea
- joint or muscle pain
- muscle cramps
- headache or migraine
- runny or stuffy nose
- stomach or abdominal pain.

Other side effects include skin rash and changes to colour vision.

- **DDAVP (Octim)**

DDAVP (Octim also called Desmopressin) is a synthetic (artificial) medicine given as a small injection under the skin. This helps your blood to clot by making your platelets stickier. Please see separate information sheet **DDAVP** for more information.

Are there any side effects?

- Red or flushed face (this is only temporary)
- Headaches
- Dizziness
- Mild stomach pain
- Nausea (feeling sick)
- Allergic reactions (rare)
- Fits (very rare).

- **Platelet transfusion**

Platelets come from people who donate their blood. All blood donors are carefully screened to make sure they do not carry any viruses (such as Hepatitis and HIV) and tested for these before use, so that risks of you getting a virus from a transfusion are minimised.

The platelet transfusion is given to you as a drip through a cannula (a small tube into a vein in your arm). This usually takes 15 to 30 minutes and can be done in an outpatient unit or on a ward. If a platelet transfusion is necessary it would usually be given just before your procedure or if there were any bleeding problems. Once transfused, the new platelets have an effect straight away.

How will I feel after my platelet transfusion?

Most people having a platelet transfusion do not feel anything unusual. You will be monitored either in the outpatient clinic or ward before, during, and just for a short time after your platelet transfusion.

If you feel unwell at any time you should tell your healthcare professional straight away. Some people may develop a temperature, chills, a rash, or breathing difficulties. These reactions are usually mild and can be easily treated with paracetamol, an antihistamine, or simply slowing down the transfusion.

Severe reactions are extremely rare, but if they do happen staff are trained to recognise and treat them.

Where can I find more information on platelet function disorders?

The following websites provide useful information on platelet function disorders.

- **The Haemophilia Society**

Web: www.haemophilia.org.uk/

- **World Federation of Hemophilia**

Web: www.wfh.org

If you would like this information in **another language, audio, Braille, Easy Read, or large print** please ask a member of staff. You can ask someone to contact us on your behalf.

Any complaints, comments, concerns, or compliments please speak to your doctor or nurse, or contact the Patient Advice and Liaison Service (PALS) on 01227 78 31 45, or email ekh-tr.pals@nhs.net

Patients should not bring in large sums of money or valuables into hospital. Please note that East Kent Hospitals accepts no responsibility for the loss or damage to personal property, unless the property had been handed in to Trust staff for safe-keeping.

Further patient leaflets are available via the East Kent Hospitals web site www.ekhuft.nhs.uk/patientinformation