

Patient information

Inherited Platelet Function Disorders

Haematology Liverpool

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.

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.

How do you get a platelet function disorder?

Platelet Function Disorders are a group of inherited conditions. This means that 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.

Please see the Platelet Function Testing Patient Information Leaflet for more information.

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.

Patients should avoid any drugs which may interfere with clot formation, such as aspirin, ibuprofen and naproxen.

How are platelet function disorders treated?

Day-to-day treatment is not needed for Platelet Function Disorders. However, treatment may 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?

Tranexamic acid, Desmopressin (DDAVP) and Platelet transfusions can all be used to prevent and stop bleeding in patients with Inherited Platelet Function Disorders.

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 inherited platelet function disorders for some procedures. It is particularly useful for mouth bleeding and therefore a very good preventative treatment when you are having a dental procedure.

This treatment is usually given as tablets. It can also be given by an intravenous drip if you are in hospital and can be made into a mouthwash.

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.

Common side effects are:

- 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 may include skin rash and changes to your colour vision.

Desmopressin (DDAVP)

Desmopressin (DDAVP) is a man-made substance that acts in a similar way to anti-diuretic hormone, which is produced naturally by the body.

It is given as a small injection under the skin and helps your blood to clot by releasing your own stores of various clotting factors and activating platelets. Please see the Desmopressin patient information sheet for more information.

Due to the way desmopressin works, the most common side effects include facial flushing and palpitations. It can also cause fluid retention, headaches, abdominal pain, and nausea.

Low blood pressure and increase in the heart rate have also been reported, which is why it is necessary to monitor these during the infusion.

Other side effects that have been reported in isolated cases of allergic skin reactions, and more seriously patients have suffered from seizures due to low sodium levels.

Please inform medical/nursing staff if you suffer from hypertension (high blood pressure), heart disease, renal disease, take anti-depressants or diuretics or have any of the above side effects.

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.

Feedback

Your feedback is important to us and helps us influence care in the future.

Following your discharge from hospital or attendance at your outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point. Your co-operation is greatly appreciated.

Further information

If you have any further questions, please ask a member of our medical / nursing team. Please contact the Haemophilia Team

Monday - Friday

9am - 5pm

Tel: 0151 706 3397

Useful websites

The Haemophilia Society:

<https://haemophilia.org.uk/>

World Federation of Haemophilia:

www.wfh.org

All Trust approved information is available on request in alternative formats, including other languages, easy read, large print, audio, Braille, moon and electronically.

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