Patient information

Thrombotic Thrombocytopenia Purpura (TTP)

Haematology Speciality

You have been diagnosed with Thrombotic Thrombocytopenia Purpura (TTP). This is a rare blood disorder and is regarded as a medical emergency.

Thrombotic: Small **clots** in the blood vessels.

Thrombocytopenia: Reduced number of **platelets** in the blood (platelets help the blood to clot).

Purpura: Bleeding in the skin causing purple spots or 'pinprick' rash.

Facts

- TTP is classified as acquired / idiopathic (no known cause) or congenital / familial (relating to families).
- TTP is a rare condition.
- TTP affects four to six people per million, per year.
- It affects women more than men.
- You cannot 'catch' TTP.
- TTP is one of the microangiopathic haemolytic anaemias (in which red cells are broken down as they pass through clots in small blood vessels).

What happens in the body?

People diagnosed with acquired / idiopathic TTP have a low level of the enzyme that breaks down a clotting factor called von Willebrand Factor, (vWF).

This enzyme is called ADAMTS 13. (ADintegrin- like And Metalloprotease with ThromboSpondin type 13). Often the ADAMTS 13 enzyme stops working because the body makes antibodies against it.

In a healthy person the von Willebrand Factor works with other clotting factors and platelets at the site of bleeding to help clotting. Large molecules of vWF are cut down into the correct size by the enzyme ADAMTS 13.

When a person has a low level of the ADAMTS 13 enzyme, the vWF remains large and causes lots of platelets to stick to it and clump together, this leads to extensive clots (thromboses) in small blood vessels throughout the body,(thrombotic microangiopathy).

As your red cells attempt to pass through the clots in your blood vessels, they may be damaged, this is known as fragmentation. This can lead to a low level of red cells (this is known as Anaemia) and can lead to extreme tiredness and fatigue.

Treatment for TTP

The best known treatment for TTP is plasma exchange **(PEX).**

Plasma is the fluid part of your blood. PEX involves removing the antibodies that destroy the ADAMTS 13 and replacing ADAMTS 13.

The procedure takes several hours and will be performed once or twice per day. Although plasma exchange must be started as soon as possible after diagnosis, immunosupression (steroids) are also usually required to produce a sustained remission.

An additional leaflet will be given to you explaining Plasma Exchange in greater detail before starting PEX.

Platelet transfusion should not be given to patients diagnosed with TTP (this can lead to an increase in further clots).

Medication used in TTP

- **Steroids**: Oral Prednisolone or Intravenous Methylprednisolone (These are given to suppress the antibodies).
- **Rituximab**: Given to target the specific cells which are involved in the making of antibodies in the body by the immune system.
- **Folic Acid**: To help you make red blood cells.
- **Aspirin** and **low molecular weight Heparin** when your platelet count is >50 (Greater than 50).
- **Omeprazole** / **Lansoprazole**: To protect the lining of your stomach whilst you are taking steroids.
- You will also have to wear Anti- embolic stockings, this is to help venous blood flow whilst you are unwell and in hospital.

Discharge home

Each patient's length of stay will vary according to the length of treatment and their response. When you no longer require Plasma Exchange and your platelet count is within 'normal levels' then discharge home is imminent.

(You will be given an additional leaflet PIF 1804 'Going Home Discharge Advice For Patients With Thrombotic Thrombocytopenic Purpura' when appropriate).

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

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