Haematology Department

Thrombotic Thrombocytopenic Purpura (also known as ‘TTP’)

Information for newly diagnosed patients

Introduction
This is an information leaflet for patients who have been diagnosed with Thrombotic Thrombocytopenic Purpura (often abbreviated to ‘TTP’). It explains the condition and its treatment.

What is TTP?
TTP is a rare blood disease. 6 -10 people per million may get this disease. It is a true medical emergency which has to be treated in hospital. It is caused by the formation of tiny blood clots in the blood vessels all over the body.

How does this happen?
Von Willebrand factor (VWF) is an important substance in the blood to help with normal blood clotting. Platelets are a type of cell found in the blood which help to form blood clots. After a cut or bruise, platelets and VWF work together to seal the injury forming a blood clot. Usually an enzyme called ADAMTS13 breaks down large VWF in the blood and regulates its levels. In TTP there is an excess of large VWF which should not be present. This large VWF causes platelets to get stuck in the small blood vessels throughout the body.

In the majority of patients with TTP the immune system is producing a rogue antibody that stops ADAMTS13 working properly. This is the cause of the accumulation of large VWF factor and this large VWF causes the formation of blood clots and leads to a depletion of platelets. The resulting low platelet count is one of the main features of TTP and is called thrombocytopenia. The low platelet count can also cause bruising, known as purpura. It is the blood clots, low platelet count and bruising that gives the condition the name Thrombotic Thrombocytopenic Purpura.

Sometimes we can find out why the immune system is producing a rogue antibody. This can result from various drugs, cancer, viral infections, thyroid problems or autoimmune problems (such as lupus). Most of the time we find no reason. We will do tests to try and find out about these conditions. If we find any of these diseases we will discuss this with you and make arrangements for any other treatments needed.
Occasionally we find that patients cannot make enough ADAMTS13 and this is called ‘congenital TTP’. This condition is extremely rare (less than 1 per million people have this). It is treated differently and we will explain this if you have this condition.

We can measure the ADAMTS13 level when you first come into hospital. We can then monitor it during treatment. This means that we can check up on your progress very closely.

**What are the symptoms of TTP?**

You may feel tired because TTP can make you anaemic (the number of red blood cells are low). Other symptoms of anaemia are looking pale and feeling breathless. TTP can cause bruising and bleeding. TTP can affect the heart (called cardiac involvement) and brain (neurological involvement). It can also cause a high temperature (fever) and affect the kidneys. We monitor for these extremely carefully so that they can be treated.

**How is TTP treated?**

Sometimes patients with TTP can be very unwell. This means that you may have to be looked after in intensive care department. Once you are more stable you can be looked after on the haematology wards (either D6 or C10).

There are different treatments for TTP and we will discuss them with you if they are needed. Nearly all patients receive the following treatments:

- **Folic acid.** This helps your blood make new red blood cells when you are anaemic. It is a vitamin tablet.
- **Aspirin.** This is an anti-coagulant that works on platelets and is given to protect you from the blood clots that can happen in TTP.
- **Low molecular weight heparin.** This is an anticoagulant that is given to protect you from the blood clots that can happen in TTP. It can be given in addition to aspirin.
- **Steroids.** These help stop the immune system from making antibody to ADAMTS13. This means that the ADAMTS13 level can return to normal. Steroids have side-effects that we will talk to you about.
- **Caplacizumab.** This is a once daily injection of an antibody fragment that helps prevent the formation of the clots that cause harm in patients with TTP. It has shown to help patients with newly diagnosed TTP in trials get better more quickly.
- **Plasma exchange.** A special machine (called an apheresis machine) can remove the plasma from your blood, which contains the rogue antibody, and replace it with plasma from blood donors. This usually means that you will have to have a central line put into the vein in either your neck or groin. This allows the treatment to be given. The procedure removes your plasma (with the antibody) and replaces it with normal plasma. Sometimes patients can feel a bit faint when having this treatment and have some tingling round their lips or fingers. There will always be a specialist apheresis nurse with you at all times when having this treatment. The treatment is usually given every day until the TTP gets better (sometimes this may just be a few days but can be longer). Each treatment lasts about two hours and can be given on a ward.
We monitor the platelet count every day. When you get better they return to normal. Usually they are more than 150 (x10^9/l) in the blood. Once they are back to normal (150) we give two more days of plasma exchange then can either stop or start to reduce how often it is given. This means you may be able to go home and have treatment as an outpatient. That exact timing of discharge depends on how well you are.

Sometimes we use a medication called rituximab to treat TTP. Usually this is if TTP affects the heart or the brain or the TTP returns after plasma exchange. This is an antibody that targets the cells in the immune system that make the antibodies. This means that immune system can no longer make the rogue antibody that causes TTP. It is often a very effective treatment for TTP.

**Benefits of treatment**
The treatment aims to get you better and back to normal.

**Risks of treatment**
The medications used to treat TTP have different side-effects. There is also a small risk of thrombosis or infection when you have a central line inserted for plasma exchange. We will discuss these with you.

**What happens after I go home?**
After you go home you may need to come into the apheresis unit for further plasma exchange until you are better. This may be several times a week for a period of time. This is not always necessary and depends on how you are. When the plasma exchange has finished then we will monitor you in clinic. This means we can see how you are but also arrange blood tests. We can monitor your ADAMTS13 level and platelet count. If there is any sign that the TTP is coming back (‘relapsing’) then we can arrange additional treatment.

**What is the long term prognosis?**
The majority of patients make a full recovery and the TTP does not come back. This is of course different between patients. We will tell you about the progress of your illness.

**Contacts/further information**

**TTP consultants:**
Dr Martin Besser (Secretary 01223 349 280)
Dr Will Thomas (01223 274 652)
Dr Emily Symington (01223 274 652)

Apheresis team nursing manager (Paul Boraks): 01223 256 272

24 hour help and specialist advice: 01223 245 151 (switchboard and ask for the ‘on-call haematology registrar’)

Haematology specialist nurse (Ruth Jolley) 01223 217717
Privacy and Dignity

Same sex bays and bathrooms are offered in all wards except critical care and theatre recovery areas where the use of high-tech equipment and/or specialist one to one care is required.

We are a smoke-free site: smoking will not be allowed anywhere on the hospital site. For advice and support in quitting, contact your GP or the free NHS stop smoking helpline on 0800 169 0 169.

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