Thrombotic thrombocytopenic purpura (TTP)

This leaflet explains more about TTP, including information about signs and symptoms, as well as available treatments. If you have any further questions, please speak to a doctor or nurse caring for you.

What is TTP and why have I got it?
TTP is a rare and serious condition that occurs in about three to four out of every million people.

In TTP, an enzyme called ADAMTS13 does not work properly, which means that the levels of von Willebrand Factor, a blood-clotting protein regulated by ADAMTS13, increase. This causes platelets (the type of blood cells that help the blood clot) to stick to this protein, leading to:
- the formation of tiny blood clots in the small blood vessels throughout the body (called microvascular thrombosis). These blood clots make it difficult for the red blood cells to circulate around the body, breaking them up into small pieces. This can cause anaemia which can turn your skin yellow (jaundice), and lead to shortness of breath and fatigue (extreme tiredness)
- a reduced number of platelets in the blood (thrombocytopenia)
- small red dots on your skin (purpura).

In most cases, ADAMTS13 does not work properly because your body’s immune system makes antibodies to it, which means it attacks and destroys it. It is not known why this happens but sometimes it can be associated with other autoimmune conditions (where your immune system attacks healthy cells in your body by mistake), or it may be triggered by pregnancy.

Very rarely, people are born without the enzyme ADAMTS13 – this is called congenital TTP. In congenital TTP, the signs and symptoms of the condition usually present in childhood.

What are the signs and symptoms?
Some patients feel unwell for a little while before they are diagnosed with TTP. It can affect any organ in the body but most often it affects:
- the brain – leading to headaches, confusion, drowsiness, memory problems and sometimes strokes.
- the heart – leading to chest pain and damage to the heart.
- the kidneys – leading to abnormal function of the kidneys and low urine production. Your urine may also go dark.
- the bowel – leading to abdominal pain.
- the liver – leading to abnormal liver blood tests.

Other symptoms include:
- jaundice
- anaemia
- small red/purple dots on your skin (purpura).
Do I need any tests to confirm the diagnosis?
The diagnosis is usually made based on your symptoms and blood tests results, including a test to measure the levels of ADAMTS13 (which can take a few days to come back).

What treatments are available?
When you are unwell with TTP, you need to be admitted to hospital. The main treatment is plasma exchange therapy, where the levels of ADAMTS13 are replenished, and antibodies to ADAMTS13 removed. You will receive plasma exchange treatment every day and your doctor will discuss with you how long you may need it for. More information on plasma exchange therapy is available in our leaflet, Plasma exchange. Please ask your doctor or nurse for a copy, if you do not already have one.

In addition to plasma exchange, you may need steroid therapy and other medicines to reduce antibody production, such as rituximab. Your doctor will tell you if that is what you need.

If your red blood count is very low, you will require a blood transfusion.

You will also need to take the following medicines:
- aspirin to reduce the stickiness of your platelets
- heparin injections to reduce the risk of you having a large blood clot in your legs (deep vein thrombosis)
- folic acid to help you make more red blood cells and replace the ones that have been broken down.

What happens if I do not get treatment?
TTP is a very serious condition that needs treatment. Without treatment, patients usually do not survive.

How is treatment monitored?
During your treatment, your platelet count (test measuring the level of platelets) and your symptoms will be closely monitored. Once the platelet count is greater than 150 x 10⁹/L for more than two days, the plasma exchange will usually be stopped.

Other blood tests that tell us whether your TTP has settled down will also be monitored.

At the end of treatment, once plasma exchange has been stopped, we will also check your ADAMTS13 levels again.

Going home after TTP
When your platelet count and other blood test results are normal, and you are free from the more serious symptoms of TTP, it means that you are in remission and ready to go home.

Once you are at home, you may feel very tired as your body continues to recover. Even though your TTP is not active, it will take time for you to get back into your old routine. You will not be able to go back to work straight away and you should increase your activity levels gradually. Your doctor will be able to advise you on this.

Some people may feel quite upset following TTP. This is a normal reaction to having been unwell and we can arrange counselling for you if needed. Please ask your doctor for more information.
You will have regular long-term follow-up appointments in the haematology outpatient clinic to check your blood tests and assess your recovery.

Please tell your haematology doctor if you are planning to:
- start any new medicines, particularly the contraceptive pill, anti-malarial drugs or statins (medicines to lower your cholesterol levels)
- become pregnant
- travel abroad.

**Can TTP relapse?**
Most patients do not have relapses of TTP and make a full recovery once the treatment is finished.

However, a small number of people may have a relapse. This can happen at any time but might be triggered by pregnancy, infections or new medicines such as the contraceptive pill. If you do not feel well, it might not be TTP-related, but you will still need to get a blood test done to make sure your platelets are normal.

If you are worried about any symptoms, please contact us (details in the box).

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**Contact us**
If you have any questions or concerns, please contact:
- the **haematology day unit**, t: 020 7188 2727, Monday to Friday, 8.30am to 6pm, or
- the **thrombosis team**, t: 020 7188 2736, Monday to Friday, 9am to 5pm.

Out of hours, please call the hospital switchboard, t: 020 7188 7188 and ask to speak to the haematology specialist registrar on call.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.guysandstthomas.nhs.uk/leaflets

**Pharmacy Medicines Helpline**
If you have any questions or concerns about your medicines, please speak to the staff caring for you or call our helpline. t: 020 7188 8748, Monday to Friday, 9am to 5pm

**Your comments and concerns**
For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.
- t: 020 7188 8801 (PALS)  e: pals@gstt.nhs.uk
- t: 020 7188 3514 (complaints)  e: complaints2@gstt.nhs.uk

**Language and accessible support services**
If you need an interpreter or information about your care in a different language or format, please get in touch. t: 020 7188 8815  e: languagesupport@gstt.nhs.uk

**NHS 111**
Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day. t: 111