NHS

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Thrombotic Thrombocytopenic Purpura (TTP) – information for patients

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What is thrombotic thrombocytopenic purpura (TTP)?

TTP stands for thrombotic thrombocytopenic purpura, which is a rare disorder of the blood clotting system. It is considered to be a medical emergency requiring immediate attention in hospital.

TTP causes large strings of a blood protein (called von Willebrand factor) which are involved in clotting to be present in the plasma. These strings act as a type of “glue” to encourage blood clots to form in small blood vessels. In patients without TTP, these large strings would normally be broken down by an enzyme called ADAMTS 13, which works like a pair of scissors to split the large von Willebrand factor strings. In patients with TTP the ADAMTS 13 enzyme cannot work properly, putting the patient at high risk of blood clotting problems.

TTP only affects around one person per one million and is more common in women than men. It can affect people at any age.

What causes TTP?

TTP can be congenital (genetic or inherited) or acquired, whereby you develop the condition after birth (this usually due to a fault in the immune system).

It is not usually known what triggers acquired TTP but it can be triggered by pregnancy or a recent virus. In acquired TTP, the body suddenly starts to make antibodies (proteins) that block the activity of an important enzyme involved in clotting called ADAMTS 13.

Congenital (inherited) TTP (also called Upshaw-Shulman Syndrome) is very rare and means that the person was born with TTP but may not have been aware of it until they became unwell. In congenital TTP, the ADAMTS13 gene is faulty and does not prompt the body to make the normal ADAMTS13 enzyme which then causes clotting problems.

Congenital TTP is inherited which means that the condition is passed from parent to child through genes. A person who inherits TTP is born with two copies of the faulty gene—one from each parent. Usually the parents have one copy each of the faulty gene, but have no signs or symptoms of TTP.

If the doctors think you have TTP they will diagnose you by assessing your clinical condition for signs of blood clotting problems and by checking the levels of ADAMTS 13 in your blood using a specialist test.

How is TTP treated?

TTP is treated with a procedure called Plasma Exchange.

Plasma is the fluid part of the blood that holds all the blood cells, antibodies, nutrients, clotting factors and chemicals required for the body to function. In TTP it is the plasma which contains the large strings of protein which put the patient at high risk of clotting problems. Plasma exchange allows your own plasma to be replaced with donated plasma that contains the missing ADAMTS 13 enzyme. Plasma exchange is repeated every day until the risk of clotting problems has been reduced and your platelet count improves. A blood specialist doctor (haematologist) will decide when it is safe to stop the procedures. If you are diagnosed with TTP and are in an unstable condition you may be treated on a specialist ward or critical care unit so that you can be closely monitored and any other effects of TTP can be managed promptly.

What happens during plasma exchange?

A needle is placed into a large vein in your arm so that blood can be drawn out into a machine called a cell separator. In some cases, a tube (venous catheter) has to be inserted into a larger vein in the groin or neck to allow sufficient blood flow into the machine.

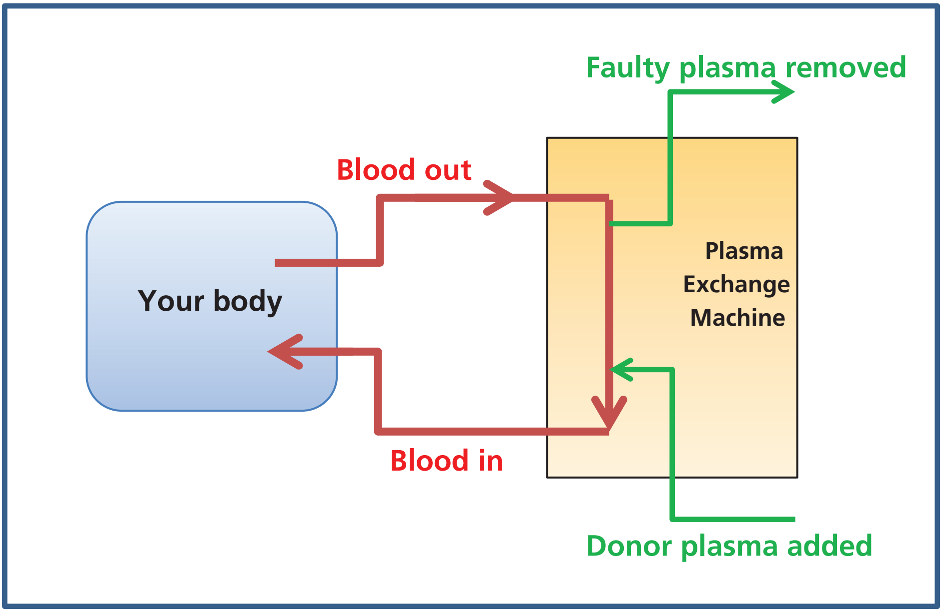
The blood is spun very quickly so that the different parts of the blood separate into layers. Your own faulty plasma is then removed and collected into a bag to be disposed of. The rest of the blood cells are returned to you along with donated plasma via a needle inserted into your other arm or via the venous catheter.

Optia machine

Only a small amount (about 180mls) of blood will be outside of your body at any time, however the machine does require the blood to enter the machine quite quickly. The procedure does not normally make people feel unwell and you will be monitored closely throughout by a nurse trained in plasma exchange.

Some people need a tube called a VasCath to be inserted into the groin or neck to enable plasma exchange to take place. Experienced medical staff will insert the line in a procedure room or in theatre prior to plasma exchange taking place. The tube is inserted using local anaesthetic to numb the skin, and it will stay in place until the plasma exchange procedures are no longer required.

Plasma exchange takes between 3-5 hours each day and may continue for several weeks however this varies from patient to patient. The procedure may be done in your room on the ward, Intensive Care Unit or in the Apheresis Unit (an area where other similar blood procedures take place).



What are the side effects of plasma exchange?

Most people do not have any problems during the procedure and it is not usually painful but can be uncomfortable. It can be tedious as you are not able to move around whilst on the machine.

You may experience tingling in the lips, nose, feet or hands caused by a low calcium level in the blood. This is caused by a substance called sodium citrate that is mixed with your blood while it is in the machine to prevent it from clotting. If identified quickly, it is easily counteracted by giving you some calcium tablets. Occasionally, if the tingling does not improve following the tablets then calcium can be given as a drip directly into your vein. The tingling sensation will only happen during the procedure so should settle down once procedure has finished.

Occasionally, patients experience low blood pressure due to the small amount of blood that is outside the body whilst it is processed by the machine. The nurse conducting the procedure will monitor your blood pressure and can give additional fluid via a drip to bring the blood pressure back up if needed.

There is a small chance that people can react to the replacement plasma used in the procedure. Even though the fluid is carefully checked there is always the possibility of an allergic reaction. If this happens whilst you are on the machine, medication can be given to resolve this and the procedure can usually continue.

How will I feel after the plasma exchange?

Most people feel tired after the procedure. Even though you are reclined in a bed or chair during the procedure, your body has been through a lot and you will need to rest afterwards.

Depending on your clinical condition, you may have to stay in hospital to recover after the procedures have been completed. Each patient has a different response to treatment so length of stay can vary. You will usually be discharged home when your platelet count has increased to a safe level and plasma exchange is no longer required. TTP can relapse (return) so you still need close monitoring even when your condition has improved.

What other treatments are available to treat TTP?

It is common for doctors to give a few days of high dose steroids to patients at the time of TTP diagnosis. The number of days that steroids are required can vary from one patient to another. This can cause temporary problems with getting to sleep and mood changes. The medical team will keep an eye on your blood pressure and blood glucose at this time.

When you are recovering from TTP, you will also be given low dose aspirin and a blood thinning injection to prevent blood clots. A commonly used and very effective treatment for TTP (to speed recovery and help prevent relapse) is Rituximab. This drug is given as a drip at least once weekly, for up to 4 weeks. It is usually well tolerated but there is a small risk of allergy and delayed infection afterwards. If it is required your doctor will speak to you in more detail at the time and ask you to complete a consent form.

It is important to continue any prescribed medication for TTP unless you are told to stop and to inform your blood specialist doctor before starting any new medication. This includes the contraceptive pill and malaria tablets which may be given prior to travelling.

Returning home after being treated

It takes time for your body to recover after an episode of TTP, and you may need to rest at home for a while before going back to your normal activities. The medical team overseeing your TTP treatment can give you advice on when it is suitable to return to work and can provide letters/sick notes if needed.

Your GP will be informed of the diagnosis and the treatment given. Some people may feel overwhelmed when they return home from hospital and may feel tearful or upset. If you are concerned you can discuss this with your GP and if required get support from local counselling services.

Your blood specialist doctor should be informed if you wish to travel abroad so that they can ensure this is done safely and with correct insurance. As TTP can be triggered by pregnancy we strongly recommend you speak to your blood specialist doctor if you wish to start a family. Those who fall pregnant following previous episodes of TTP will require regular monitoring during their pregnancy.

You will need to attend clinic appointments with a blood specialist doctor (haematologist) and have regular blood tests so that you can be monitored closely, even when you are feeling well. The appointments will be sent out to you automatically but there will be a contact number on the letter if you need to change them for any reason.

If you have previously had TTP and experience any of the following symptoms you should **seek urgent medical advice:**

- Headaches not relieved by paracetamol
- Blurred vision, dizziness or confusion
- Easy bruising without obvious cause
- Dark urine
- Feeling unwell and concerned

If you are worried that your TTP is coming back, the on-call Haematology doctors can be contacted via Switchboard 24 hours a day on 0121 371 2000, or you should attend your nearest Accident and Emergency Department for assessment.

In most cases there is nothing to be worried about, however it is important get your blood count checked and confirm that your TTP has not recurred.

More information about TTP from patients and their families can be found at **www.ttpnetwork.org.uk**

The Apheresis Team (nursing team who perform plasma exchange procedures) can be contacted via switchboard on 0121 371 2000 between the hours of 08:00 - 18:00 Monday to Friday. If you need information outside of these hours or you are unwell please follow the instructions above.

With thanks to the TTP Network and TTP Team at University College London Hospitals NHS Foundation Trust.

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PI24/1834/03 Author: Annie Price
Date: November 2025 Review date: March 2027