Thrombosis UK

Awareness • Research • Care

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Clarity approved by Plain English Campaign

Thrombotic thrombocytopenic purpura

Thrombotic thrombocytopenic purpura (TTP) is a very rare disorder which affects around three people in one million. The name describes the effects caused in a person (see below). TTP is life-threatening without treatment but it can be controlled and, in the majority of cases, can be curable.

What is TTP

The name describes the effects.
Thrombotic - Small clumps (thrombi) of platelets that block or partially block the small blood vessels (capillaries).
Thrombocytopenic - The clinical term for a low platelet count of less than 100 billion cells per litre of blood.
A normal platelet count is between 150 and 400 billion cells per litre.
Purpura - Little pinprick spots due to bruising caused by a low platelet counts. artery, where it will block blood flow to the lungs.

What causes these effects?

The formation of platelet clumps is linked to a shortage of a protein called von Willebrand factor cleaving protease, also known as ADAMTS13.

Von Willebrand factor (vWF) is necessary for blood clotting and is made up of lots of small units that are linked together to form a long string. vWF cleaving protease breaks up this long string into smaller pieces. In someone with TTP, vWF cleaving protease is missing, and so the long strings of vWF are not broken up. These will bind to platelets and can cause platelet clumping which will partially or fully block blood flow. This affects the blood vessels of the brain, kidneys and heart in particular.

The blockage then causes two things to happen.

- First, the blockage prevents blood from getting to the affected areas - so blockages in the brain lead to symptoms such as headaches, blurred vision, confusion, weakness, seizures and, in extreme cases, can cause unconsciousness.
- Second, as the red blood cells try to pass the blockage some are broken up (known as haemolysis) and so the number of red cells falls and this leads to anaemia. Because red cells carry oxygen in your body, a lack of them leads to feeling tired, light-headed or short of breath.

What triggers the TTP process?

There are two main types of TTP - inherited and acquired.

In inherited TTP, people are born with a shortage of ADAMTS13 and tend to have TTP episodes from childhood. Episodes can be sparked off by infection, alcohol and, in women, pregnancy.

Acquired TTP is triggered by an unwanted antibody in the body. Your immune system normally protects your body from bacteria and viruses, which it identifies as harmful. Certain white blood cells (called lymphocytes) release proteins called antibodies, which attach to the bacteria and viruses and direct other cells to destroy them. In people with TTP, this process doesn't work properly and the antibodies target ADAMTS13. Some people describe having flu-like symptoms before the symptoms of TTP, suggesting that an infection may have triggered the episode of TTP.

Other causes:
Occasionally TTP is linked to a known condition such as cancer, or taking certain drugs, or a serious viral infection such as HIV.

How is TTP treated?

- First, further platelet clump formation must be prevented. To remove the antibody and replace the missing ADAMTS12 protein, plasma exchange is carried out by connecting the person to a machine called a blood-cell separator. This process constantly collects a small amount of blood from the body, removes the plasma (the clear fluid part of the blood) and then puts the cells back into the body. The plasma that has been removed is replaced with donated plasma. The donated plasma comes from the National Blood Service. Each unit has been tested to make sure it is not carrying HIV or hepatitis B and C and other diseases that can be passed on through transfusion.

- Second, in acquired TTP, the antibody formation may need to be suppressed. Various immunosuppressive drugs are used, such as prednisolone and ritoximab.

Each day, the clinical team looking after the person with TTP will assess a large number of blood results. These results, and the person's clinical condition, will indicate how well treatment is going. Each person will react to the treatment at a different rate and the blood results may be affected by other things.

The main blood results are monitored for:

- platelet count, to assess whether the platelets are still clumping;
- haemoglobin level, to see whether red blood cells are being broken up (however, plasma exchange will reduce haemoglobin levels in the short term);
- actate dehydrogenase (LDH), to measure damage to red blood cells; and
- reticulocytes (young red cells whose level indicates the state of blood-cell production in your bone marrow).

Can it happen over and over again?

In many cases TTP is a 'one off' event. However, people with inherited TTP and some people with acquired TTP may have several episodes of TTP and so it is important that they receive follow-up care. People who are at high risk of another episode may receive treatment to prevent this.

Striving To SPOT & STOP The Clots Through Education & Research

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For more information on how to help us, write to:

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