CONDITION

Thrombotic thrombocytopenic purpura

A condition in which tiny blood clots lower platelets and restrict organ blood flow

WHAT IT IS
Rare blood-clotting condition affecting small blood vessels

BODY SYSTEM
Blood, brain, heart, kidneys, and circulation

MAIN SYMPTOMS
Bruising, extreme tiredness, headache, confusion

WHAT CAUSES IT
Immune or inherited severe ADAMTS13 deficiency

HOW IT IS DIAGNOSED
Severely low ADAMTS13 activity plus blood tests

MAIN TREATMENT
Plasma therapy and medicines that stop microclots

CAN IT BE CURED?
No - remission is possible, but relapse can occur

IS IT AN EMERGENCY?
Yes - any suspected new or relapsing episode

LONG-TERM OUTLOOK
Many recover, but relapses and lasting symptoms can occur

Overview

Thrombotic thrombocytopenic purpura, usually shortened to TTP, is a rare condition in which many tiny blood clots form in small blood vessels. These clots can reduce blood flow to the brain, heart, kidneys, and other organs.

The clots use up platelets, the blood cells that help stop bleeding. This causes a low platelet count, called thrombocytopenia. Red blood cells can also break apart as they squeeze past the clots. This is called microangiopathic haemolytic anaemia.

TTP usually happens because the body has too little activity from a blood enzyme called ADAMTS13. This enzyme normally cuts a large clotting protein into smaller pieces. Without enough ADAMTS13 activity, platelets can gather around the protein and form unwanted clots.

A new or relapsing episode of TTP is a medical emergency. Treatment often begins before all

test results are available because organ injury can develop quickly. With prompt specialist treatment, many people recover from an episode.

Symptoms

TTP does not always cause the same symptoms. They may appear suddenly or develop over several days.

Low platelets can cause:

The breakdown of red blood cells may cause severe tiredness, weakness, shortness of breath, pale skin, or yellowing of the eyes and skin. Urine may look dark.

Reduced blood flow to the brain can cause headache, confusion, trouble speaking, weakness, seizures, or loss of consciousness. Some people have symptoms that come and go.

Heart involvement may cause chest pain, breathlessness, a fast heartbeat, or fainting. Kidney injury can cause reduced urine, swelling, or changes found only on blood and urine tests.

Not everyone develops the traditional combination of low platelets, anaemia, fever, neurological symptoms, and kidney problems. Waiting for all five features can delay treatment.

- Unexplained bruises
- Tiny red or purple spots under the skin, called petechiae
- Bleeding from the nose or gums
- Heavier menstrual bleeding

Causes and risk factors

Most cases are immune TTP. The immune system mistakenly makes antibodies that block or remove ADAMTS13. This leaves too little active enzyme in the blood. Immune TTP is sometimes called acquired TTP because it is not present from birth.

An immune episode may occur without a clear trigger. In some people, it is associated with pregnancy, an infection, surgery, or another autoimmune condition. These associations do not mean that the person caused the episode.

Congenital TTP is caused by inherited changes in both copies of the ADAMTS13 gene. It may first appear in infancy, but some people do not develop clear symptoms until pregnancy, an infection, or another physical stress. Congenital TTP is lifelong and may affect other family members.

TTP is not contagious. It is not caused by eating particular foods, ordinary bruising, or everyday stress.

Other conditions and some medicines can cause similar damage in small blood vessels without severe ADAMTS13 deficiency. These are different types of thrombotic microangiopathy and may need different treatment.

Diagnosis

Doctors consider TTP when blood tests show a low platelet count and red blood-cell destruction without another clear explanation. Because treatment is urgent, initial decisions may be based on symptoms, examination, and routine blood tests while a specialised ADAMTS13 test is pending.

Tests commonly include:

Severely reduced ADAMTS13 activity, generally below 10% of normal, strongly supports TTP. Antibody testing helps distinguish immune TTP from congenital TTP. Genetic testing may confirm congenital TTP.

A scoring tool may estimate how likely severe ADAMTS13 deficiency is while the result is pending. It does not replace the ADAMTS13 test.

Doctors also check for other possible causes of low platelets and red-cell destruction. These include severe infection, pregnancy-related conditions, immune reactions, certain cancers, and other thrombotic microangiopathies.

- A complete blood count to measure platelets and red blood cells
- A blood film to look for fragmented red cells called schistocytes
- Tests for cell breakdown, including bilirubin, lactate dehydrogenase, and haptoglobin
- Kidney, liver, heart, and urine tests to check for organ injury
- An ADAMTS13 activity test, often with antibody or inhibitor testing

Treatment and management

TTP is treated in hospital by a team experienced in blood disorders. Treatment may begin as soon as TTP is strongly suspected rather than waiting for confirmation.

For immune TTP, plasma exchange is a central treatment. Blood is passed through a machine that removes the person's plasma and replaces it with donor plasma. This removes harmful

antibodies and supplies working ADAMTS13.

Corticosteroids like prednisone or hydrocortisone reduce the immune attack. Rituximab or another immune-targeting treatment may also be used to reduce antibody production and lower the chance of an early relapse.

Caplacizumab is a medicine that blocks platelets from attaching to the large clotting protein involved in TTP. It can reduce continued microclot formation while other treatments address the immune cause. Because it affects clotting, bleeding is an important treatment risk that the hospital team monitors.

Congenital TTP is treated by replacing missing ADAMTS13. This may involve donor plasma or recombinant ADAMTS13, which is manufactured rather than taken from donated plasma. Treatment may be given regularly or around higher-risk situations, according to specialist assessment.

Supportive care may include treatment for seizures, heart problems, kidney injury, infection, or other complications. Red blood-cell transfusions may be used for significant anaemia. Platelet transfusions are generally reserved for particular situations, such as serious bleeding, because TTP already involves platelet-rich clots.

After recovery, follow-up blood tests track the platelet count and ADAMTS13 activity. A substantial fall in ADAMTS13 during remission can warn of increased relapse risk. Specialists may sometimes offer treatment before symptoms develop.

Complications

Tiny clots can interrupt blood flow and injure organs. Possible complications include stroke, seizures, heart injury, abnormal heart rhythms, kidney injury, and damage to other organs. Serious bleeding is less common than bruising but can occur because platelet levels are low or as a treatment side effect.

Some people continue to experience tiredness, headaches, reduced concentration, memory problems, low mood, or anxiety after blood counts return to normal. These effects can interfere with work, study, and daily activities.

Immune TTP can relapse months or years after an earlier episode. A relapse is not caused by something the person did. Regular monitoring helps identify falling ADAMTS13 activity, but a relapse can still develop unexpectedly.

Plasma exchange requires access to a large vein and can occasionally cause infection, bleeding, blood clots, allergic reactions, or changes in blood pressure. Immune treatments can increase susceptibility to some infections. The care team monitors for these risks.

Prevention

There is no proven way to prevent a first episode of immune TTP. It is not preventable through diet, exercise, or avoiding ordinary daily activities.

Congenital TTP is inherited and cannot be prevented by lifestyle changes. Genetic counselling can help affected people and their relatives understand inheritance and testing options.

For someone who has had immune TTP, planned blood tests can identify reduced ADAMTS13 activity during remission. Specialist treatment may sometimes lower the chance of relapse when the activity falls significantly.

Pregnancy can increase the risk of an episode in both immune and congenital TTP. Planning a pregnancy with haematology and maternity specialists allows closer monitoring and a treatment plan. This does not mean pregnancy is always unsafe.

Living with it and what to expect

An acute TTP episode usually requires a hospital stay and repeated blood tests. Treatment continues until platelet levels and signs of red-cell destruction improve. The time needed is different for each person.

Blood counts may return to normal before energy, concentration, or confidence fully recover. Fatigue and cognitive symptoms can continue for months. A gradual return to work, exercise, driving, or study may be more manageable than trying to resume everything at once.

People recovering from immune TTP need long-term, and often lifelong, follow-up. Appointments may include blood counts, ADAMTS13 measurements, and checks for treatment effects. The timing becomes less frequent during stable remission but should not stop without specialist advice.

Some people have one episode and remain well. Others experience one or more relapses. Congenital TTP usually needs continuing specialist management because the underlying ADAMTS13 deficiency remains present.

Many people return to usual family, work, and social lives. It can still be difficult to live with uncertainty about relapse. Keeping a written relapse plan and knowing where to seek urgent assessment may help.

When to get medical help

A suspected first episode or relapse of TTP needs emergency assessment. Call emergency services for:

If you have previously had TTP, follow your specialist relapse plan. Seek urgent same-day assessment for new unexplained bruising, petechiae, dark urine, marked tiredness, yellow skin or eyes, or a persistent headache. Do not wait for a routine appointment if these symptoms could represent a relapse.

Routine follow-up is for monitoring while you feel well, discussing ongoing fatigue or concentration problems, reviewing medicines, and planning pregnancy, travel, surgery, or dental procedures. Contact the treatment team sooner if symptoms change between appointments.

- New confusion, trouble speaking, facial drooping, or weakness
- A seizure, collapse, or loss of consciousness
- Chest pain, severe breathlessness, or fainting
- Sudden severe headache, especially with neurological symptoms
- Heavy or uncontrolled bleeding

Common questions

Can TTP go away completely?

An acute episode can resolve with treatment, and immune TTP may remain in remission for years. However, relapse remains possible. Congenital TTP is lifelong because the inherited ADAMTS13 deficiency does not go away.

Is TTP a type of cancer?

No. TTP is a blood-clotting condition, not a cancer. It can cause abnormal blood counts, but it does not involve uncontrolled growth of cancer cells.

Can TTP come back after treatment?

Yes. Immune TTP can relapse even after a long remission. Regular blood counts and ADAMTS13 testing may help identify increased risk. New symptoms still require urgent assessment, even if recent tests were normal.

Is TTP inherited?

Congenital TTP is inherited. A person usually receives one altered ADAMTS13 gene copy from each parent. Immune TTP is not usually inherited, although a tendency toward autoimmune conditions may run in some families.

How long does recovery take?

Platelets and red-cell breakdown may improve during hospital treatment, but complete recovery can take longer. Tiredness, reduced stamina, headaches, or concentration problems may continue for weeks or months. Recovery time cannot be predicted from the platelet count alone.

Can I live a normal life after TTP?

Many people return to their usual activities after treatment. Long-term monitoring remains important, and some people need support for fatigue, emotional distress, or cognitive symptoms. Congenital TTP may require regular preventive treatment.

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