
Idiopathic thrombocytopenic purpura (ITP)

Summary

- Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder, in which a person's blood doesn't clot properly, because the immune system destroys the blood-clotting platelets.
 - The cause of ITP is not known, but it is thought that some kinds of viral infection may cause the immune system to malfunction and start producing antibodies that attack platelets.
 - Treatment options for ITP include ongoing monitoring of platelet levels, medications, and surgical removal of the spleen in severe cases.
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What is ITP?

Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder that causes you to have low platelet levels. Platelets are cell fragments that are found in the blood and normally help the blood to clot. In people with ITP, the body produces antibodies that attack and destroy the platelets. Antibodies are produced by cells of the immune system, and are normally part of our system for fighting infection.

The two categories of ITP are:

- acute ITP – this is mostly a disease of childhood, and accounts for nine out of 10 cases of ITP. It is far less common in adults, who are more likely to have chronic ITP. Sometimes a viral infection (such as a cold) seems to trigger the condition. The disease goes away by itself within two to six months
- chronic ITP – this ongoing form accounts for most ITP seen in adults and is far less common in children. Chronic ITP has similar symptoms to acute ITP, except that it lingers for longer than six months.

Malfunction of the immune system and ITP

The body's immune system is a specialised system of cells and chemicals that fight infections. Normally, the immune system recognises 'self' and doesn't attack tissues or organs of the body.

Doctors think that some types of viral infections trigger ITP. For reasons unknown, these infections prompt the person's lymph tissues and spleen (an organ that helps filter the blood) to make antibodies that attack the platelets in their blood. Antibodies that attack the body are called autoantibodies. In other cases, the cause is unknown (idiopathic).

Platelets are made in our bone marrow. Since ITP targets mature platelets as they circulate through the spleen, the 'newborn' platelets inside bone marrow are healthy and normal. In many cases of acute ITP, the platelet count will rise again within a few weeks and return to normal within a few months.

Who does ITP affect?

ITP is more common among children than adults, most often occurring around two to four years of age. Estimates suggest that ITP affects one in every 10,000 children. Among adults, young women are more likely to develop ITP than any other group, for reasons unknown.

What causes ITP?

The cause of ITP is not known. It is thought that viral infections might make the immune system become overactive, and start producing abnormal antibodies.

Symptoms of ITP

The normal level of platelets in the blood is between 150,000 and 400,000 per mL of blood. A person with ITP may have a platelet count of 20 000 or lower.

In most people with mild ITP, there are no symptoms and they will feel perfectly well. However, if the platelet count drops very low, they may experience an increase in bruising or bleeding. These symptoms may include:

- skin that bruises very easily
- a skin rash of small red dots (petechiae), which does not blanch (go pale) with pressure
- bleeding from any area of the body
- bleeding from the gums
- frequent nosebleeds that take a long time to stop
- internal bleeding
- long or heavy menstrual periods.

Diagnosis of ITP

Most of the time ITP has no symptoms and may be discovered during blood tests for an unrelated medical matter.

ITP is diagnosis of exclusion – once a low platelet count has been discovered, more tests are used to rule out any other causes. These tests may include:

- blood tests – such as a full blood examination (FBE) to check for platelet numbers, to see if there are abnormalities in other blood counts, or to see if the blood cells look normal under the microscope
- bone marrow biopsy – doctors remove a small sample of bone marrow through a needle and then check it in a laboratory. In a person with ITP, the platelets produced in the bone marrow should be normal. This test is rarely needed to diagnose ITP, and is used only when strictly necessary (such as when there is some uncertainty of the diagnosis)
- other tests – to rule out other conditions that may cause a low platelet count, such as acute leukaemia and aplastic anaemia.

In addition to these tests, doctors diagnose ITP by taking the person's medical history and doing a physical examination.

Treatment for ITP

Treatment for ITP includes time and close observation. In some cases, ITP goes away by itself. In cases where the platelet count is very low or there is evidence of bleeding, some form of treatment should be started.

First-line treatments (the preferred treatments, or the ones that are tried first) for ITP include:

- corticosteroids – these medications are used to reduce the activity of the immune system. They may be given as intravenous injections or tablets
- intravenous immunoglobulin (IVIG) – this is a blood product that consists of concentrated antibodies. It is thought that the autoantibodies might be 'swamped' by IVIG, reducing their ability to target platelets. IVIG has to be given by an intravenous infusion and may take a couple of hours.

Second-line treatments (those used if the first-line treatments do not work) include:

- splenectomy – surgical removal of the spleen. This operation cures ITP in about 70 per cent of chronic cases
- thrombopoietin analogues – treatment to increase production of new platelets in the bone marrow
- monoclonal antibodies to CD20 – an injection treatment targeting antibody-producing cells.

Occasionally, a person who appears to have been cured of ITP will experience a relapse, perhaps months or even years after the initial episode. If this happens the person will need repeat treatment. In most cases, the relapse seems to be triggered by a viral infection.

Where to get help

- Your **GP (doctor)**
- If you are bleeding, go to your nearest emergency department

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