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.
- We do not know the cause of ITP, but some kinds of viral infection may cause the immune system to malfunction and start producing antibodies that attack platelets.
- Treatment options include ongoing monitoring of platelet levels, medications, and surgical removal of the spleen in severe cases.

Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder. The person’s blood doesn’t clot properly because antibodies destroy the platelets that help blood to clot.

Blood is partly made up of platelets (thrombocytes) – small, cell-like bodies that are important in blood clotting. A healthy person has a count of between 150,000 and 450,000 platelets, but a person with ITP may have a count of 20,000 or lower.

We do not know the cause of ITP, but viral infections might make the immune system malfunction and start producing ‘rogue’ antibodies.

There are two broad categories of ITP: one that usually goes away by itself with time, and the other that lingers beyond six months. 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.

Symptoms of idiopathic thrombocytopenic purpura (ITP)

In general, the person appears and feels perfectly well. They may have no symptoms of ITP until their platelet count is extremely low. 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
- Abnormal menstruation.

Two categories of ITP

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.

**Diagnosis of ITP**

Very often, ITP has no symptoms and may be discovered during blood tests for an unrelated medical matter. ITP is a diagnosis of exclusion, where more tests are used to rule out any other causes of a low platelet count.

Doctors diagnose ITP by taking the person’s medical history and doing a physical examination, plus laboratory tests which can include:

- Blood tests – such as a full blood examination (FBE) to check for platelet numbers, to see if there are abnormalities in other blood-cell counts, or to see if the blood cells look normal under the microscope
- Other tests – to rule out other conditions that may cause a low platelet count, such as acute leukaemia and aplastic anaemia
- 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).

**Treatment for ITP**

Treatment includes time and close observation. In some cases, ITP goes away by itself. In severe cases, the symptoms of the disorder (such as the tendency to spontaneously bleed) must be managed.

First-line treatments include:

- Corticosteroids – medications to reduce the activity of the immune system. Corticosteroids may be given as intravenous injections or tablets
- Intravenous immunoglobulin (IVIG) – the antibodies are ‘caught’ by the IVIG and removed from the bloodstream. Autoantibodies might also be ‘swamped’ by IVIG, reducing their ability to target platelets.

Second-line treatments 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.
- Ongoing monitoring – doctors monitor the person’s platelet count regularly. They may re-start the treatment every time the platelet count drops below a critical figure (usually when it is less than 20,000) or before surgical procedures to reduce the risk of bleeding. The person may have relapses.

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

**Where to get help**
- Your doctor

**Things to remember**
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