

## Idiopathic thrombocytopenic purpura

Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder, in which the blood doesn't clot properly because the platelets that help blood to clot are destroyed by antibodies.

Blood is partly made up of platelets (thrombocytes), which are small cell-like bodies that are important in blood clotting. In a normal body, platelets number between 150,000 and 450,000, but a person with ITP may have a platelet count of 20,000 or lower.

The cause of ITP is unknown, but it is thought that viral infections cause the immune system to malfunction and start producing 'rogue' antibodies.

There are two broad categories of ITP – one that usually resolves 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 one in every 10,000 children is affected. Among adults, young women are more likely to develop ITP than any other group, for reasons unknown.

### Symptoms of idiopathic thrombocytopenic purpura

In general, the person appears and feels perfectly well. There may be no symptoms of ITP until the platelet count is extremely low. Symptoms may include:

- The skin bruises very easily
- A skin rash of small red dots (petechiae), which does not blanch with pressure
- Bleeding from any area of the body, including:
  - 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 include:

- **Acute** – this is primarily a disease of childhood, which 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, the condition seems to be triggered by a viral infection (such as a cold). The disease resolves by itself within two to six months.
- **Chronic** – 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

The 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. It is thought that ITP is triggered by some types of viral infections. For reasons unknown, these infections prompt the lymph tissues and spleen to make antibodies that attack the platelets in the blood. Antibodies that attack the body are called autoantibodies. In other cases, the cause is unknown (idiopathic).

Platelets are made in the 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 further tests are required to exclude other causes of a low platelet count.

ITP is diagnosed by taking a medical history and physical examination, coupled with some laboratory investigations including:

- **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 exclude other conditions that may cause a low platelet count, such as acute leukaemia and aplastic anaemia.
- **Bone marrow biopsy** – a small sample of bone marrow is taken through a needle and then checked in a laboratory. In ITP, the platelets produced in the bone marrow should be normal. This test is rarely needed to diagnose ITP, and only used 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 resolves 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** – drugs to reduce the activity of the immune system. Corticosteroids may be administered as intravenous injections or tablets.
- **Intravenous immunoglobulin (IVG)** – the antibodies are 'caught' by the IVG and removed from the bloodstream.

Second-line treatments include:

- **Splenectomy** – is surgical removal of the spleen. This procedure cures ITP in about 70 per cent of chronic cases.
- **Thrombopoietin analogues** – is treatment to increase the production of new platelets in the bone marrow.
- **Monoclonal antibodies to CD20** – is an injection treatment targeting antibody producing cells.
- **Ongoing monitoring** – the platelet count is regularly monitored. Treatment is often recommenced every time the platelet count drops below a critical figure (usually when it is less than 20,000) or prior to surgical procedures to reduce the bleeding risk. Relapses may occur. Occasionally, the person who appears to have been cured of ITP will experience a relapse, perhaps months or even years after the initial episode. Repeat treatment is needed. In most cases, the relapse seems to be triggered by a viral infection.

## Where to get help

- Your doctor

## Things to remember

- Idiopathic thrombocytopenic purpura (ITP) is a rare autoimmune disorder, in which the blood doesn't clot properly because the blood clotting platelets are destroyed by the immune system.

- The cause of ITP is unknown, but it is thought that some kinds of viral infection 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.

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Australian Centre for Blood Diseases

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