## Dutch ITP Patients' Association

The Dutch ITP Patients' Association (ITP Patiëntenvereniging Nederland) was founded in May 2003. Every year, a number of information sessions are organised to explain medical developments and give patients the opportunity to exchange experiences with each other. The association also publishes a newsletter 3 times a year. For more information and other brochures, please visit our website www.itp-pv.nl. Our website also provides details of how to support our association.

Patiëntenvereniging Nederland


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Precautionary treatment is virtually never started with those kinds of counts. If the platelet count is below 20, there is a much greater chance of bleeding. In that case, active treatment is often considered.

Between 20 and 50 platelets, a (temporary) treatment can be started as a precautionary measure. It depends on the patient and the circumstances (e.g. before surgery or childbirth). The attending physician will decide together with the patient what course of action to take. To be prepared for these kinds of decisions, you can use the online help feature that helps patients make these kinds of choices and then discuss the outcome with your attending physician (https://www.keuzehulp.info/pp, select 'ITP' under 'Haematology'). Other factors also come into it, such as age, other diseases, and any medication you are on. It may sometimes be that treatment of another disease or changing (or stopping) your medication could increase the platelet count.

## First-line treatment (in case of new ITP)

If treatment is necessary, the physician will start by prescribing corticosteroids (prednisone or dexamethasone) in high dosages. Prednisone is, in principle, taken in a high dosage for 3 and sometimes 6 weeks, and gradually reduced if successful.
Dexamethasone is taken in 4-day blocks at 40mg a day. In one of every three ITP patients, the disease will have gone away after that. In two of every three patients, the platelet count will drop again as soon as they stop taking prednisone or dexamethasone or the dosage lowered.

## Second-line treatment (in case of persistent and chronic ITP)

If prednisone is insufficiently efficacious or does not do anything (in the long term), there are various options for a follow-up treatment. The main options are:

- Thrombopoietin receptor agonists (TPO-RAs): this medicine stimulates platelet production in the bone marrow.
- Rituximab: this medicine inhibits certain white blood cells that produce antibodies that attack blood platelets.
- Spleen removal (splenectomy): platelets will then no longer be filtered out by the spleen. This option will only be on the table in case of chronic ITP and when other second-line treatments did not have an effect or did not have effect. Performing a spleen scan* can be considered prior to proceeding to a splenectomy.

*In some ITP patients, platelets are broken down not only in the spleen but also in the liver. A spleen scan is a test to check where in the body the most platelets are broken down. This scan can show, for example, that platelets are mainly broken down in the liver. If so, the chance of the platelet count going up significantly after a splenectomy is clearly smaller.

What to consider when choosing a certain treatment depends on various factors. These are partly medical factors such as physical fitness, underlying disease, and age. But the patient's own preference is also a factor to consider. Preference becomes a factor when there is not one option that is clearly the best, because two (or more) treatment options are medically equal. The ultimate decision will be made in consultation with the attending physician.

In acute situations (severe bleeding), treatment will always take place at the hospital and differ from the first-line and second-line treatments described above.



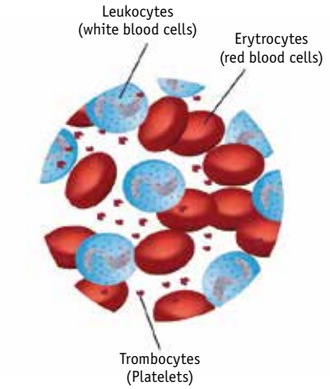


# General information brochure for adults with ITP

Patiëntenvereniging Nederland

## ITP

**Immune thrombocytopenia (ITP) is a disease that causes someone to bleed more easily and quickly. This is caused by a low platelet count in the blood. Blood platelets are cells that play a role in stopping bleeds. In ITP patients, the body produces antibodies that attack the body's own blood platelets. As a result, these platelets are broken down and the person suffers a shortage of platelets. This shortage is called thrombocytopenia (thrombocyte = blood platelet; penia = shortage).**




## Name

The disease used to have different names, such as Werlhof Disease (named after the man who discovered the disease in 1735) or Idiopathic Thrombocytopenic Purpura, which basically means bruising (purpura) caused by a low blood platelet count (thrombocytopenic) without a clear cause (idiopathic). We know now that it is an autoimmune disease. The abbreviation ITP has remained, but it now stands for something else, i.e. immune thrombocytopenia. Actually, AITP (autoimmune thrombocytopenia) would be a better name for this disorder, but ITP is used for convenience.

## Causes of ITP

ITP is caused by certain antibodies. Antibodies are proteins in the blood that play a big role in warding off bacteria and viruses. The antibodies adhere to these foreign intruders and break them down. With ITP, the defence system makes a mistake and produces antibodies that attack the body's own platelets. This is why ITP is an autoimmune disease, as 'auto' means 'self', i.e. targeted on itself. In roughly 80% of patients with chronic ITP, antibodies can be detected in the blood.

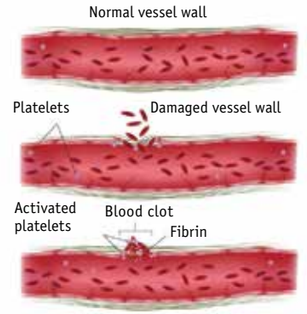
## Rare disease

ITP is a rare disease. The exact number of people who suffer from it is not known. The number of new ITP patients per year is estimated at 2 per 100,000 persons. In the Netherlands, this means around 400 new patients a year. This can be primary ITP or ITP in conjunction with another disease, i.e. secondary ITP.



Primary and secondary ITP

The term 'primary ITP' is used to denote that there is no other disease or cause that explains the thrombocytopenia. This is contrary to what is called 'secondary ITP,' where the thrombocytopenia is the result of another disease (such as systemic lupus erythematosus (SLE)) or a form of leukaemia/ lymphoma). In the vast majority of patients, a blood test shows that there is no other disease at play. Sometimes, however, the attending physician will have to perform a bone marrow puncture, especially to rule out a bone marrow disease.




Blood platelet shortage (thrombocytopenia)

Normally, people have between 150 and 350 platelets per millilitre of blood. After production in the bone marrow, these platelets circulate in the blood for between 7 and 10 days, following which they are detected as old and broken down. This mainly occurs in the spleen and liver.

There are various causes of thrombocytopenia:

- Increased platelet usage, due to bleeding
- Impaired platelet production, possibly due to a bone marrow disease or chemotherapy
- Increased platelet destruction, such as by antibodies in case of ITP or due to an enlarged spleen.

Thrombocytopenia symptoms

What you may experience due to a shortage of platelets:

- Small bleeds under the skin (petechiae)
- Nose bleeds and bleeding gums
- Heavy menstrual bleeding
- Large bleeds under the skin (haematomas)



In case of severe platelet shortage, bleeding may also occur in the gastrointestinal tract or urinary tract, primarily as a result of trauma or existing abnormalities. An ITP-related brain haemorrhage is very rare.

## Acute or chronic ITP

In young children, ITP (often associated with fever-causing infections) is mostly temporary, while in adults it is generally chronic. The literature often makes a distinction between patients with new/persistent ITP (the ITP is active for under a year) and patients with chronic ITP (active for over a year). Nowadays, this distinction is less relevant because 2nd-line treatment can often be started as early as after 3 months (see Treatment).

## The seriousness of ITP

The seriousness of ITP is determined by the blood platelet count and the presence of any other diseases (see secondary ITP). For the patient, however, it will generally be the degree of bleeding (more or less related to the platelet count) and fatigue (which may not be related to the platelet count at all) that determine how serious their ITP is for them. All these factors are different for different patients and need to be addressed during the patient's appointments with the attending specialist.

## Treatment

In principle, the treatment of ITP is based on suppressing and preventing bleeding and increasing the platelet count. Although active bleeding is always a reason to start a treatment, bleeding with platelet counts of over 50 is basically not more likely to occur in people with ITP than it is in people without ITP. Even surgery can be performed safely with a platelet count of over 50.