

Reference

Guideline for infection prevention in (functional) hyposplenism and asplenia. National Infectious Disease Control Coordination, Dutch public health authority (RIVM) - Infectious Disease Control Centre, November 2018.
(<https://lci.rivm.nl/richtlijnen/asplenie>)
(<https://www.itp-pv.nl/links>)

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.



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Measures after the surgery

A person can, in principle, live well without a spleen. This is partly because the liver can take over a lot of the spleen's functions. After a splenectomy, it is important to take measures against serious infections by taking antibiotics and getting revaccinated 3 to 5 years after primary vaccination.

Antibiotics

In adults and children over the age of 5, the advice is to take antibiotics as a precaution for a period of 2 years after spleen removal (antibiotic prophylaxis). For children under the age of 5, antibiotic prophylaxis is administered for at least two years up to age 5 and for up to 2 years after the splenectomy. Deviation from this rule is possible, such as in case of another disease that affects the patient's immune system.

Even when taking antibiotic prophylaxis, it is important to always see a doctor as soon as possible when a patient develops a fever or feels feverish. The doctor may then, if necessary, prescribe different antibiotics.

After two years, the antibiotic prophylaxis will be stopped. It is then important to stay alert to the following points:

- Always have a course of antibiotics available at home to be able to immediately go on antibiotics in case of an infection.
- Start on antibiotics immediately (<1 hour).
- Contact your GP after every acute fever (38.5°C or higher), especially when it also involves cold shivers. If you have to call another doctor because your own GP is unavailable, remember to tell them that you do not have a spleen.
- In case of dog and/or cat bites; clean the wound immediately and have a doctor look at it as soon as possible, and start on antibiotics.



Vaccinations

The Dutch public health authority (RIVM) has issued advice on vaccination ahead of a splenectomy (see Reference for the full list):

- If possible, complete the full recommended vaccination course at least 2 weeks before the surgery.
- The flu jab is given annually during the usual period.
- If vaccination before the surgery is not possible, do not start the full course of recommended vaccinations until at least 2 weeks have passed since the surgery.
- If the patient is on immunosuppressants or the patient's immune system is compromised, consult with a doctor about vaccination.



- People who had a splenectomy in the past and who have not been (fully) vaccinated yet are advised to get the recommended missing vaccinations after all.

Measures when travelling

- If you are planning to go abroad, you should get advice from a travel expert or specialist body in good time. The Dutch National Travel Advice Coordination Centre (Landelijk Coördinatiecentrum Reizigersadviesing) is one body you can turn to for more information.
- It is particularly important that you prevent mosquito bites in malaria-endemic areas. Careful use of malaria prophylaxis is important.
- Be alert to tick bites, especially when travelling in forested areas in the eastern part of the United States.

General information about spleen removal for ITP

(Immuun Trombocytopenie)



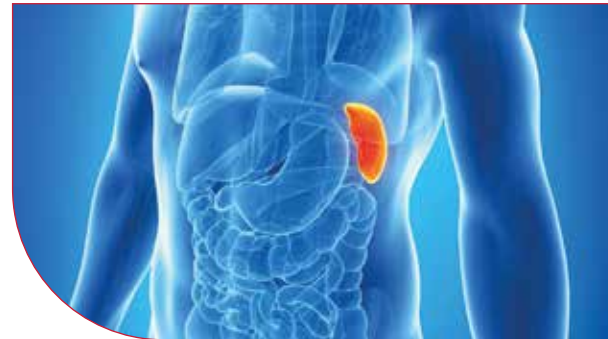
In this information brochure, we will go into the function of the spleen, the medical indications for spleen removal for ITP, vaccinations, and measures that have to be taken once the spleen has been removed.

This brochure is not intended to influence your decision on whether or not to have your spleen removed. This is a decision you should make in close consultation with your attending physician. To help you make that decision, you can use the online help tool that helps patients make medical decisions, the so-called 'Keuzehulp' at <https://www.keuzehulp.info/pp/itp>.

The function of the spleen

The spleen has several different functions, including:

- Defence against pathogens such as bacteria.
- Filtering out old or abnormal blood cells.



The spleen is located in the top-left area of the abdomen. It is a vulnerable organ the size of a fist that is protected by the rib cage. It is an important part of the immune system. When it comes to defending the body against infections, the white blood cells in the spleen play a key role. They do that in 2 ways. Firstly, these white blood cells produce antibodies, which are proteins that can neutralise bacteria. In addition, certain white blood cells can also directly filter bacteria out of the blood.

Spleen removal (splenectomy)

The most common reason for spleen removal is to treat a ruptured spleen. Aside from that, it can also be a treatment option for certain blood diseases, such as the autoimmune disease ITP. With ITP, antibodies attack the body's own blood platelets. Blood platelets to which these antibodies have adhered are primarily broken down by the spleen.

Removing the spleen will then prevent platelets being broken down unnecessarily. While this does not eliminate the cause of ITP, it will improve ITP patients' platelet count by 60%-70% for the long term.

Sometimes a splenectomy has insufficient effect because accessory spleens may still remain or grow. Roughly 10%-20% of people have one or several accessory spleens. These are small (between 3mm and 20mm) and may be overlooked

during laparoscopic surgery or taken for lymph glands. This could reduce the positive effect of removing the spleen. Blood platelets may also be broken down mainly in the liver, which would mean that removing the spleen would not have the desired effect (see Spleen scan).

Indications for spleen removal for ITP

In the past, splenectomies were performed much more often than nowadays. This is related to the development of new medicines. Another reason that makes patients wary of a splenectomy is the heightened risk of infections and the 30%-40% chance that removing the spleen will not improve the situation in the long term.

A splenectomy can be considered when medication has proven to be insufficiently effective or when the disease has returned after previous successful treatment with medicines. A splenectomy can also be considered when a patient is particularly affected by side effects of medication or does not want to be on medication for the long term. Removing the spleen within 12 months of the diagnosis of ITP is not recommended, because the disease course can be very variable in the beginning and ITP can even resolve spontaneously.

In case of children with ITP, spleen removal is put off as long as possible because the ITP often has a more favourable course in children. Postponing spleen removal, preferably until after the 5th or 6th year of its life, will protect the young child against serious infections until the defence system has developed fully.

It is important that the patient/parents carefully weigh the pros and cons of a possible treatment with the attending physician.

The spleen scan

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. The result may predict whether a splenectomy might be useful. For example, if platelets are mainly broken down in the liver the chance of the platelet count going up significantly after a splenectomy is clearly smaller.

At present, spleen scans can only be performed at the Haga hospital in The Hague. There are plans, however, to also offer spleen scans at other hospitals in the Netherlands.

Living without a spleen

Persons who do not have a spleen are exposed to a heightened risk of serious infections that may even be life-threatening. These infections are caused mainly by:

- Bacteria; especially pneumococcal bacteria (streptococcus pneumoniae), haemophilus influenzae type B, and possible meningococcal bacteria (neisseria meningitidis).
- Infections after dog and cat bites.
- Parasitic infections such as malaria or babesiosis.

The infection generally begins with flu-like symptoms and fever. These symptoms can intensify rapidly in no time, causing the patient to feel increasingly sick. This can cause symptoms such as low blood pressure, cold shivers, fever, shortness of breath, and heart palpitations.

Most infections occur during the first two years after the splenectomy, but the heightened risk will remain for life. While vaccinations may reduce the risk, they cannot prevent all infections. As a result, someone who has had their spleen removed is strongly advised to adhere to a number of measures and advice (see Measures after the surgery).

Measures before the surgery

Due to the heightened risk of life-threatening infections, the patient is advised to complete the full recommended vaccination schedule at least 2 weeks before the splenectomy (see Vaccinations). Vaccinations offer protection against serious infection caused by pneumococcal bacteria, haemophilus influenzae type B, and meningococcal bacteria.

For people with coagulation disorders, vaccinations can be injected under the skin (subcutaneously) instead of in muscle (intramuscularly).

The surgery

Nowadays, splenectomies are performed laparoscopically, i.e. without cutting. In some cases, a splenectomy is still done laparotomically, which does involve an incision in the abdominal skin. These options will be discussed with you beforehand. After a laparoscopic splenectomy, you will often be discharged within a few days.