Information for the treatment of Immune Thrombocytopenia for Paediatrics



Splenectomy

Second Line Treatment

A splenectomy is a surgical treatment option for ITP, where, in most cases, patients undergo a laparoscopic (key hole) surgical procedure to remove the spleen.

This treatment has been widely used in Immune Thrombocytopenia (ITP) in the past, however, advancements in treatment options and their access, including Thrombopoietin Receptor Agonists (TPORAs), has meant that splenectomy is currently rarely undertaken to treat Immune Thrombocytopenia.

A discussion with patients and their families to identify patient Quality of Life (QoL) factors, including treatment preferences, age, lifestyle, comorbidities, and drug availability, should be considered before moving forward with this treatment.



WHERE IS THE SPLEEN?

The spleen is located on the left side of the abdomen. It is estimated that the spleen in children from toddlerhood to puberty can be calculated as follows: spleen length[cm] = 6 cm + 1/3 cm per year of age. In adults, the spleen weighs around 160g and is roughly the size of a clenched fist.



WHAT IS THE JOB OF THE SPLEEN?

The spleen helps the body fight infections by acting as a blood filter, removing bacteria from the bloodstream. In particular, the spleen helps to protect people from encapsulated bacterial infections.

As blood flows through the spleen, it filters any old or damaged blood cells (including platelets) that are then broken down and destroyed. Normally there is a store of red blood cells and platelets within the spleen. In the event of significant blood loss, the spleen releases this store into the bloodstream.



WHY REMOVE THE SPLEEN?

In people with ITP, the immune system can treat platelets as foreign and destroys them. The spleen is responsible for removing these damaged platelets; therefore, removing the spleen can help keep more platelets circulating in the body and improve the platelet count.



ABOUT THE SURGERY

A splenectomy is usually completed laparoscopically (keyhole) through four small incisions in the abdomen, which are between 5mm – 10mm in size.

Carbon dioxide gas is pumped into the abdominal cavity to provide a space to operate.

A fibre-optic telescope and a long instrument are inserted into the abdomen, and the spleen is separated from the stomach, kidney and colon. Once the blood supply has been cut from the spleen, the spleen is removed.

In some cases, a larger incision is required due to several factors, resulting in open surgery and a longer recovery period.



TYPICAL TIME TO RESPONSE

1-3 days.



LIKELIHOOD OF AN INITIAL RESPONSE

70-80% have an initial response, while 10-15% have no meaningful response.

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LIKELIHOOD OF A LONG-TERM RESPONSE? 3-5 years

Splenectomy is associated with the greatest probability of durable remission, with a long term response rate of 60–70%.



LONG-TERM COMPLICATIONS ASSOCIATED WITH SPLENECTOMY

- 1. The immune system will not work as well as it did before the splenectomy, and other organs, such as the liver, bone marrow and lymph nodes, will take over some of the functions of the spleen.
- 2. Approximately 55% of all splenectomised patients will develop a post-operative blood clot, most resolving naturally. However, some patients will require to be placed on blood thinning medications for a period of time. Other patients may develop other complications post-surgery, with long-term impacts.
- 3. Serious bacterial infections are increased in splenectomised patients, which should be considered when discussing this treatment.



OTHER CONSIDERATIONS

- Splenectomy should be delayed for at least 12 months after a new diagnosis of ITP if possible.
- If a splenectomy is the next course of the treatment plan, it is recommended that Spleen Australia be
 contacted to discuss pre-surgery and post-surgery requirements. Patients living in Queensland, Victoria
 and Tasmania are automatically registered with Spleen Australia; however, it is free
 for all Australia residents to be part of this register.
- Patients undergoing a splenectomy should:
 - Review the medical recommendations for people without a functioning spleen from Spleen Australia - https://spleen.org.au/vaccinations/
 - Ensure all vaccinations are updated prior to surgery. This includes extra vaccinations for Pneumococcal, Meningococcal, Haemophilus influenza type b and the annual influenza vaccine.
 - Be prescribed prophylactic antibiotics 20 mg/kg (up 250mg daily) for three years post-surgery.
 - Receive an emergency supply of antibiotics (22.5mg/kg/dose up to 875mg/dose) twice daily in case a medical review is not immediately available.
 - Follow the spleen being removed it is extremely important for patients to watch for signs of infection such as fever, vomiting and/or diarrhea, feeling cold, shivering, sweating, shaking and/or a severe headache or confusion. They must seek urgent medical attention if they notice any of these symptoms.
- There is no readily available testing to identify if a splenectomy will
 work on a specific patient, and this treatment may not eliminate ITP
 because the spleen is not the only organ that is responsible for platelet
 destruction in ITP.
- The possibility of an accessory spleen may be considered if a patient does not respond or if relapse occurs.

REFERENCES

https://itpaustralia.org.au/itp-in-children/

https://spleen.org.au/wp-content/uploads/2020/03/RECOMMENDATIONS Spleen_Registry_p.pdf http://itpaustralia.org.au/wp-content/uploads/2024/10/2023-ANZCHOG-Paediatric-ITP-Treatment-Guidelines.pdf

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