Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Prednisolone

First-line treatment

Corticosteroids or Steroids are the standard first-line treatment approach for Immune Thrombocytopenia (ITP) and usually include Prednisone or Dexamethasone.

Corticosteroids are treatments based on a naturally occurring hormone produced by the adrenal glands involved in controlling inflammation, stress response, metabolism, behaviour, electrolyte balance and more.

Corticosteroids work by suppressing the immune system to raise the platelet count.

Corticosteroids are widely used as an initial therapy in ITP, and it is recommended, based on the Consensus Guidelines for the management of adult Immune Thrombocytopenia in Australia and New Zealand, that this treatment not be used for longer than six to eight weeks to minimise toxicity.

BRAND NAMES

Predsolone, Predsone / Panafcortelone, Solone

HOW DOES THE TREATMENT WORK?

Prednisolone reduces the destruction of antibody-coated platelets in the blood and the bone marrow, thereby increasing effective platelet production.

It may reduce ITP bleeding through a direct impact on the blood vessels.

HOW IS IT ADMINISTERED?

Prednisolone is taken orally and comes in both tablet and liquid form.

It is recommended to:

- take this medication after meals or with food or milk to decrease gastrointestinal upset.
- prescribe a Proton Pump Inhibitor (PPI) with this medication to reduce gastrointestinal acid-related side effects.

DOSAGE

A recommended starting dose of 1 mg/kg/day for the first two weeks (usually rounded to the nearest 25mg), followed by a tapering plan over 6 weeks.

It is recommended to cap the dose to 75 to 80 mg once daily, even for patients weighing more than 80 kg.

COMMON SIDE EFFECTS

The side effects vary with dose and duration of administration.

Common side effects can include mood swings, anger, anxiety, insomnia (difficulty sleeping), weight gain, cushingoid face (also known as moon face and appears as a puffy face and rounded facial features), stomach irritation, ulcers, high blood pressure, high blood sugar, and fluid retention.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Prednisolone

First-line treatment

RARE SIDE EFFECTS

With repetitive cycles, side effects can include osteoporosis (weakening of bones), skin changes including thinning and senile purpura, hair loss, avascular necrosis (death of bone) of joints, psychosis, cataracts, infections, and adrenal insufficiency.

A rapid decline in platelet count may occur between cycles.

TREATMENT RESPONSE

Initial response time is between 5 and 14 days, with a peak response of 7 to 21 days.

LIKELIHOOD OF AN INITIAL RESPONSE

Approximately 75%. The response rate can be increased when combined with other agents, such as IVIg.

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

Around a third of adult patients treated with steroids will have sustained normal platelet counts. The majority of patients will experience a fall in platelet count as the dose is reduced or the drug is discontinued. There is no evidence that long-term use leads to sustained response.

OTHER CONSIDERATIONS

Mood alterations have been widely reported.

Use with caution if an existing mental health condition is present. Prednisolone can be used in pregnancy.

Prednisolone is favoured in older patients who are less likely to tolerate the side effects of Dexamethasone.

Patients requiring longer-term steroid therapy (steroid dependent after more than 8-10 weeks) or repeated courses of steroid therapy should be referred to a Haematologist specialising in Immune Thrombocytopenia. Visit ITPAustralia.org.au for more details.

REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Dexamethasone

First-line treatment

Corticosteroids or steroids are the standard first-line treatment approach for Immune Thrombocytopenia (ITP) and usually include Prednisolone or Dexamethasone.

Corticosteroids are treatments based on a naturally occurring hormone produced by the adrenal glands involved in controlling inflammation, stress response, metabolism, behaviour, electrolyte balance and more.

Corticosteroids work by suppressing the immune system to raise the platelet count.

Corticosteroids are standard of care in the initial therapy of ITP, and it is recommended, based on the Consensus Guidelines for the management of adult Immune Thrombocytopenia in Australia and New Zealand, that Dexamethasone not be used for more than 6 pulses/cycles of treatment.

BRAND NAMES

Dexamethasone

HOW DOES THE TREATMENT WORK?

Dexamethasone reduces the destruction of antibody-coated platelets in the blood and the bone marrow, thereby increasing effective platelet production.

It may reduce ITP bleeding through a direct impact on the blood vessels.

HOW IS IT ADMINISTERED?

Dexamethasone can be administered by intravenous injection or oral tablet.

It is recommended to be administered after meals or with food or milk to decrease gastrointestinal upset.

DOSAGE

A recommended starting dose for Dexamethasone is 40 mg or 0.6 mg/kg orally once daily for four days, known as a Dexamethasone pulse/cycle.

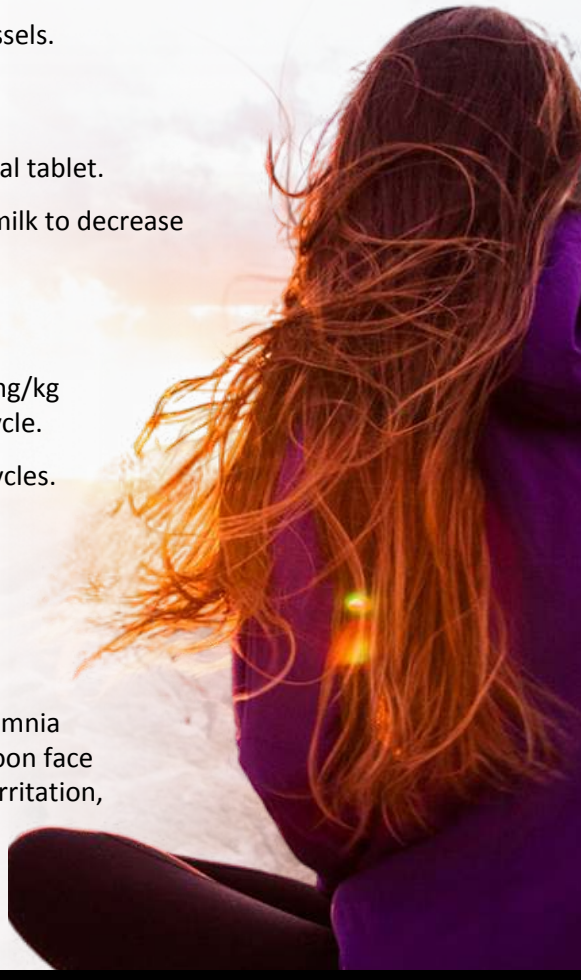
This treatment can be repeated every 14–28 days from one to six cycles.

Dexamethasone can be reduced to 20 mg for older adults.

COMMON SIDE EFFECTS

The side effects vary with dose and duration of administration.

Common side effects can include mood swings, anger, anxiety, insomnia (difficulty sleeping), weight gain, cushingoid face (also known as moon face and appears as a puffy face and rounded facial features), stomach irritation, ulcers, high blood pressure, high blood sugar, and fluid retention.



Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Dexamethasone

First-line treatment

RARE SIDE EFFECTS

With repetitive cycles, side effects can include osteoporosis (weakening of bones), skin changes including thinning and senile purpura, hair loss, avascular necrosis (death of bone) of joints, psychosis, cataracts, infections, and adrenal insufficiency.

A rapid decline in platelet count may occur between cycles.

TREATMENT RESPONSE

Initial response time is between 3 and 14 days, with a peak response of 4 to 28 days.

LIKELIHOOD OF AN INITIAL RESPONSE

Approximately 80%.

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

In general, initial treatment of ITP with steroid results in long term normalisation of platelet count in around a third of adult patients with ITP.

OTHER CONSIDERATIONS

Dexamethasone is favoured by clinicians and patients seeking a more rapid response with a shorter overall duration of steroid exposure.

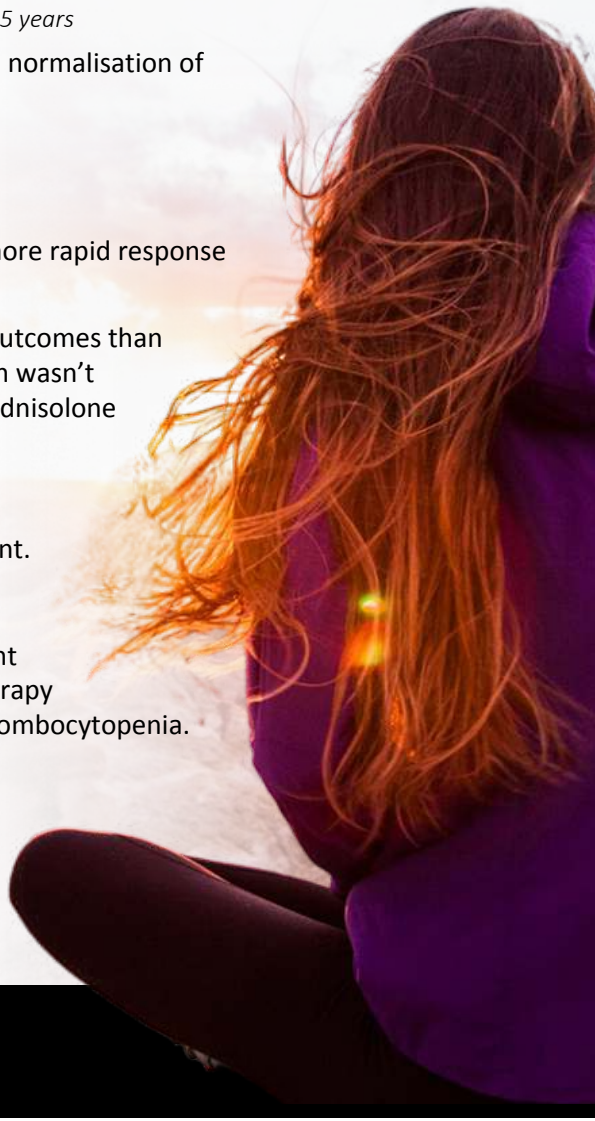
Some studies suggesting that dexamethasone resulted in better outcomes than prednisolone were published in the 2000 decade. This observation wasn't replicated with widespread use, and most haematologists use prednisolone in first-line treatment of ITP.

Severe mood alterations have been reported.

Use with caution if a pre-existing mental health condition is present.

Dexamethasone can be used in pregnancy.

Patients requiring longer-term steroid therapy (steroids dependent after more than 8 to 10 weeks) or repeated courses of steroid therapy should be referred to a Haematologist specialising in Immune Thrombocytopenia.



REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
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Information for the treatment of Immune Thrombocytopenia in Adults

Intravenous Immunoglobulin (IVIg)

First-line treatment

Intravenous Immunoglobulin (IVIg) is a solution comprising human plasma proteins, particularly IgG antibodies, with a broad spectrum of antibody activity. IVIg is prepared from large pools of human plasma collected from several thousand blood donors and is used for patients who need the replacement of antibodies and autoimmune disorders.

All IVIg products are tested and screened to protect patients from diseases and undergo at least two processes that destroy viruses. The risk of infection from this product is very low.

Intravenous Immunoglobulin is commonly called IVIg and can be used soon before, during or after medical procedures or on-demand as a first-line or rescue therapy. IVIg can be combined with other treatments, including corticosteroids.

IVIg is used to treat many immune deficiency disorders, and in ITP, it is used to elevate platelet counts temporarily. Although IVIg is not expected to result in a sustained, elevated platelet count, in rare cases, this does occur, and this therapy can be repeated for longer-lasting results.

BRAND NAMES

Privigen® AU (CSL Behring), Privigen® (CSL Behring) [Imported], Flebogamma® (Grifols), Gamunex® (Grifols), Octagam®10% (Octapharma), Kiovig® 10% (Takeda) Privigen® AU.

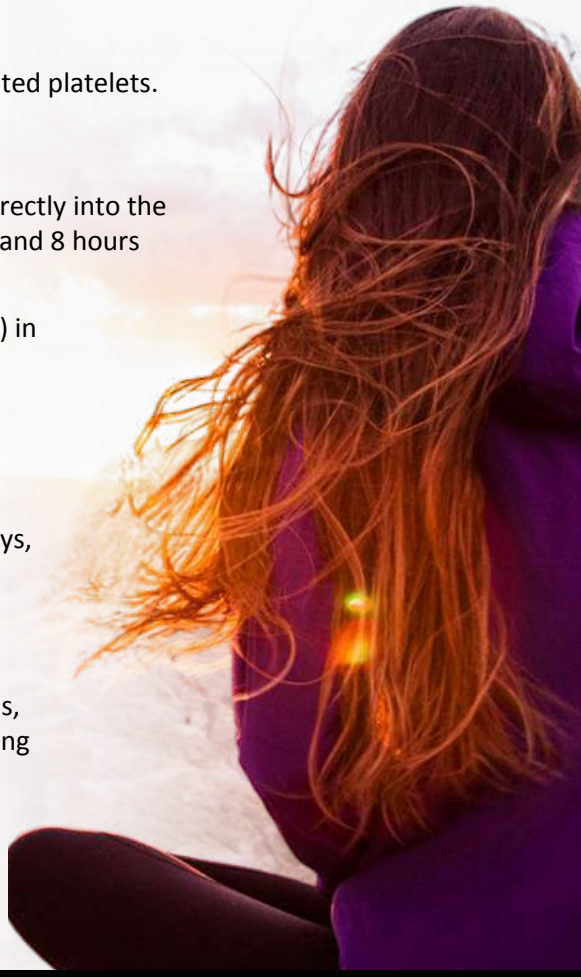
HOW DOES THE TREATMENT WORK?

IVIg is a blood product that reduces the destruction of antibody-coated platelets.

HOW IS IT ADMINISTERED?

The immunoglobulin is administered intravenously through an IV directly into the vein. Depending on the dosage, this treatment can take between 2 and 8 hours per infusion.

IVIg is administered by a health care professional (nurse or clinician) in hospital or day treatment centre.



DOSAGE

The Ig dose is calculated based on the patients weight.

Dosing options include 0.4g/kg daily for 3-5 days or 1g/kg for 1-2 days, with the latter having a faster response.

COMMON SIDE EFFECTS

The most common side effects of IVIg include headache, fever, chills, nausea or vomiting, muscle pain or chest pain. In many cases, slowing down the infusion rate can help reduce these side effects.

It is recommended that premedication with antihistamines or occasionally corticosteroids can also assist with reducing side effects. Some reactions may occur post-infusion, and these will generally present within 24 hours of the infusion.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Intravenous Immunoglobulin (IVIg)

First-line treatment

RARE SIDE EFFECTS

Aseptic meningitis, hypotension (low blood pressure), haemolysis (breakdown of red blood cells), kidney failure, thrombosis (blood clots), and anaphylaxis (severe allergy) have been reported.

TREATMENT RESPONSE

24 to 48 hours. The 5% and 10% formulations appear to have similar efficacy.

In some instances, the effectiveness of IVIg can reduce over time when used repeatedly.

LIKELIHOOD OF AN INITIAL RESPONSE

Approximately 75%.

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

Effects generally last several days to weeks. It has not been shown to induce a sustained response in adults.

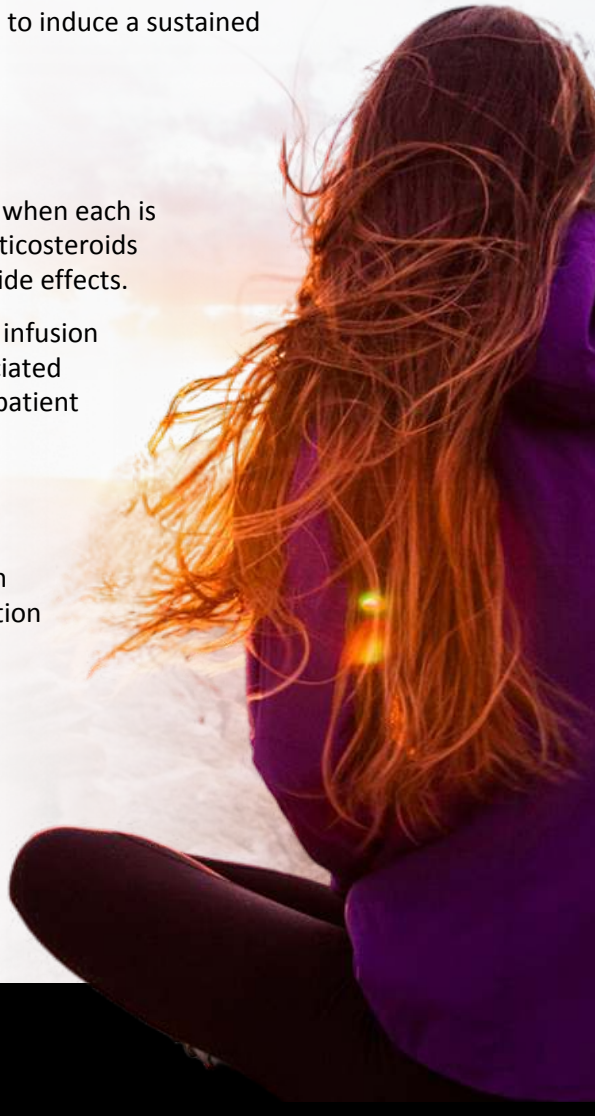
OTHER CONSIDERATIONS

IVIg may increase platelet count more rapidly than corticosteroids when each is used as a single agent. Pre-treatment and post-treatment with corticosteroids may be necessary to improve effectiveness and help to minimise side effects.

Patients are recommended to remain well hydrated. Lowering the infusion rate can also assist with reducing common side effects. Risks associated with the use of a blood product should be discussed between the patient and the healthcare professional.

IVIg can be used in pregnancy.

NB: Allocation of domestic or imported immunoglobulin products, including Kiovig 10%, is being managed within BloodSTAR based on patients' specific conditions, according to a pre-determined allocation matrix that the NBA regularly adjusts.



REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new
https://itpaustralia.org.au/introduction-of-imported-immunoglobulin-product-kiovig/

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Rituximab

Second-line treatment

Rituximab is a manufactured antibody that targets B lymphocytes. It is not derived from blood donations. It is most commonly used if treatment beyond the first line is required.

Rituximab's other use is in the treatment of B cell lymphomas, usually in combination with chemotherapy. **Rituximab is not a chemotherapy drug** but is generally handled by cancer/oncology pharmacies because of its historical use in lymphoma. It is also increasingly used in a broad range of other autoimmune diseases.

Studies have reported around 25% long-term remission following rituximab treatment, with the initial response rates are about 60%.

It appears to be most likely a successful treatment in younger people, particularly females, with short duration of ITP and lower platelet counts. However, its use should not be limited to this group of ITP patients.

BRAND NAMES

Riximyo, Ruxience and Truxima.

Mabthera/Rituxan (Roche) is the original brand; however, over recent years, several biosimilar versions of rituximab have become available, resulting in a considerable drop in the cost of treatment.

HOW DOES THE TREATMENT WORK?

Rituximab targets CD20 on B lymphocytes, so depleting the cells that go on to produce the antiplatelet antibodies, which cause thrombocytopenia. Rituximab attaches itself to all the CD20 proteins it finds to mark them. This triggers the immune system cells to pick and destroy the marked cells.

Rituximab destroys both abnormal and normal B-cells. Once treatment is over, the body can replace the normal B-cells.

HOW IS IT ADMINISTERED?

Rituximab is administered as an intravenous infusion.

DOSAGE

100mg weekly for 4 weeks is the most commonly used dose.

The first infusion is given over 3-4 hours, but subsequent infusions can generally be given over 1 hour.

Rituximab is administered by a healthcare professional (nurse or clinician) in a hospital or day treatment centre.

Occasionally, the lymphoma dose of 375mg/m2 may be used, but there appears to be no advantage in using the higher dose in ITP.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Rituximab

Second-line treatment

COMMON SIDE EFFECTS

Rituximab is generally well tolerated. Common side effects are more likely with the first infusion at the standard dose or 1g rituximab infusions.

Common side effects include low-grade fevers, headaches or chills, nausea, stomach pain, diarrhea, heartburn, night sweats, muscle or joint pain and dizziness.

The most listed side effects of rituximab are those seen in the treatment of lymphoma and when combined with chemotherapy. Side effects are less frequently seen in ITP but may include a first-dose infusion reaction, diminished response to subsequent vaccinations, and a risk of worse outcomes with COVID-19 infection.

RARE SIDE EFFECTS

A rare yet serious complication of rituximab is progressive multifocal leukoencephalopathy (a rare viral disease characterised by progressive damage or inflammation of the brain's white matter at multiple locations).

Because B lymphocytes make our healthy immunoglobulins and the antibodies responsible for ITP, some patients develop hypogammaglobulinemia (stop making healthy immunoglobulins) after rituximab use and will require regular immunoglobulin (IVIg) infusions.

TREATMENT RESPONSE

The median time to response is 5.5 weeks.

LIKELIHOOD OF AN INITIAL RESPONSE

Approximately 60%.

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

Approximately 25%.

OTHER CONSIDERATIONS

Premedication of steroids and antihistamines is given before rituximab treatment to reduce the risk of infusion reactions.

Rituximab is favoured for patients without concomitant immunodeficiency and those at risk of blood clots. Rituximab should be considered in younger female patients with short disease duration (< 1–2 years).

Historically, most rituximab use was in treating lymphoma, so it is sometimes hard to separate the listed side effects of rituximab given in combination with chemotherapy for lymphoma from the generally lower dose given alone in ITP.

Lesser frequency and severity of side effects are likely seen in ITP.
As previously indicated, rituximab is not a chemotherapy/cytotoxic drug.

Rituximab use is avoided in pregnancy. It is generally advised that pregnancy is avoided for 12 months after rituximab treatment, although this is not based on any direct evidence.

REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Avatrombopag

Second-line treatment

Avatrombopag is a Thrombopoietin Receptor Agonist (TPORA) that increases platelet counts in most ITP patients.

Thrombopoietin (TPO) is a hormone produced by the liver that naturally controls the development of megakaryocytes, the large cell-producing platelets in the bone marrow.

TPO receptor agonists (TPO-RAs) bind to the same receptor and prompt megakaryocytes in the bone marrow to produce more platelets.

BRAND NAMES
Doptelet

HOW DOES THE TREATMENT WORK?
Avatrombopag stimulates the megakaryocytes in the bone marrow to produce more platelets.

HOW IS IT ADMINISTERED?
Oral Tablet

DOSAGE
The usual starting dose for adults is one 20 mg tablet with food at the same time each day.

The dose can be increased to a maximum of 40 mg daily.

With continued monitoring, some patients can switch to alternate daily dosing.

Note: there are no dietary restrictions with this medication.

TREATMENT RESPONSE
1 - 3 weeks

LIKELIHOOD OF AN INITIAL RESPONSE
Approximately 66% of patients have an initial response of a platelet count greater than 50,000 per microliter on day 8.

LIKELIHOOD OF A LONG-TERM RESPONSE? *3-5 years*
This is yet unknown.

COMMON SIDE EFFECTS
Fever, easy bruising or bleeding (nosebleeds, bleeding gums), purple or red spots on your skin, feeling tired, headache, joint pain, cold symptoms such as runny or stuffy nose, sneezing, sore throat, nausea, stomach pain, or swelling in your hands or feet.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Avatrombopag

Second-line treatment

RARE SIDE EFFECTS

Signs of a blood clot:

- chest pains, shortness of breath;
- fast heartbeats
- pain, swelling, and redness in one or both legs;
- stomach pain or tenderness;
- sudden fever or chills, yellowing of the skin or eyes, or
- bloody or tarry stools, coughing up blood or vomit that looks like coffee grounds.

Allergy or hypersensitivity:

- shortness of breath;
- wheezing or difficulty breathing;
- swelling of the face, lips, tongue or other areas of the body; or
- itching or hives

**These side effects were reported by patients in the initial trials. After you have received medical advice for any side effects you experience, you can report side effects to the Therapeutic Goods Administration online at www.tga.gov.au/reporting-problems. By reporting side effects, you can help provide more information on the safety of this medicine.*

OTHER CONSIDERATIONS

Treatment is required to be taken with food; however, there are no restrictions around the diet.

If the medication is not taken at the usual time, it is recommended to take the medication as soon as remembered. If it is almost time for the next dose, skip the missed dose. DO NOT take a double dose to make up for the missed dose.

Some patients with an extreme response to lactose may encounter a reaction to this treatment.

Not recommended for use during pregnancy.

If no response is seen in this treatment, switching to a different TPORA is recommended.

REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Eltrombopag

Second-line treatment

Eltrombopag is a Thrombopoietin Receptor Agonist (TPORA) that increases platelet counts in most ITP patients (ITP).

Thrombopoietin (TPO) is a hormone produced by the liver that naturally controls the development of megakaryocytes, the large cell-producing platelets in the bone marrow.

TPO receptor agonists (TPO-RAs) bind to the same receptor and prompt megakaryocytes in the bone marrow to produce more platelets.

BRAND NAMES

Revolade and Promacta (USA)

HOW DOES THE TREATMENT WORK?

Pathologically, ITP is classically characterised by increased platelet destruction and/or decreased platelet production due to inhibition of megakaryocyte function.

Revolade stimulates the bone marrow to encourage the production of more platelets. The molecule binds to the thrombopoietin receptor on megakaryocytes, which stimulates platelet production.

HOW IS IT ADMINISTERED?

Oral tablet.

DOSAGE

The usual starting dose for adults is one 50 mg tablet daily (or 25mg daily for people of Southeast Asian background).

The dose can be increased to a maximum of 75 mg daily.

With continued monitoring, some patients can switch to alternate daily dosing, or less.

COMMON SIDE EFFECTS

Nausea, diarrhea, upper respiratory tract infection (symptoms may include runny nose, stuffy nose, and sneezing), vomiting.

RARE SIDE EFFECTS

Higher risk for blood clots. New or worsened cataracts. Platelet count may drop suddenly if the drug is stopped abruptly. Increases the risk for serious liver problems.

Bone marrow reticulin (fibrous scarring) has been reported but is rare.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Eltrombopag

Second-Line Treatment

TREATMENT RESPONSE

1 to 3 weeks.

LIKELIHOOD OF AN INITIAL RESPONSE

Approximately 60-90% of patients have an initial response of a platelet count greater than 50,000 per microliter.

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

Approximately 80% of patients who respond initially maintain a platelet count of about 50,000 per microliter if treatment is continued.

The prevalence of sustained remission off treatment has yet to be established; however, it can be achieved.

OTHER CONSIDERATIONS

The dose should be taken **at least 2 hours before and 4 hours after** the following products:

- foods that contain calcium
- antacids used to treat stomach ulcers or heartburn,
- multivitamins, mineral supplements, or products that contain iron, calcium, aluminium, magnesium, selenium, and zinc.

Weekly blood tests are recommended until a regular dose is established to keep the platelet count levels stable, and then you can go to monthly blood tests or even less frequently.

Regular monthly monitoring of liver enzymes is recommended.

Regular monitoring of ferritin levels is recommended, with iron infusions recommended every 12-24 months as required.

Not recommended for use during pregnancy.

If no response is seen in this treatment, switching to a different TPORA is recommended.

It is extremely rare; however, a small percentage of patients may require a treatment holiday if this treatment stops being effective.

REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Romiplostim

Second-line treatment

Romiplostim is a Thrombopoietin Receptor Agonist (TPORA) That increases platelet counts in most patients with ITP.

Thrombopoietin (TPO) is a hormone produced by the liver that naturally controls the development of megakaryocytes, the large cell-producing platelets in the bone marrow.

TPO receptor agonists (TPO-RAs) bind to the same receptor and prompt megakaryocytes in the bone marrow to produce more platelets.

BRAND NAMES

NPlate

HOW DOES THE TREATMENT WORK?

Romiplostim stimulates the megakaryocytes in the bone marrow to produce more platelets.

HOW IS IT ADMINISTERED?

A weekly subcutaneous injection is initially administered by a healthcare professional.

In nearly all cases, with training, patients will be able to self-administer this treatment.

DOSAGE

Based on the patient's body weight, the Pharmaceutical Benefits Scheme (PBS) recommends that the initial dose for romiplostim is 1 µg/kg.

However, the average dose needed to maintain platelet counts in the initial clinical trials of romiplostim were around 3mcg/kg, so many ITP haematologists start at this dose.

Future dose adjustments are based on changes in platelet counts to achieve and maintain a platelet count in the range 50,000 - 200,000 per microliter.

The maximum allowed weekly dose from the PBS is 10 µg/kg.

COMMON SIDE EFFECTS

Headache, joint and muscle pain.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Romiplostim

Second-Line Treatment

RARE SIDE EFFECTS

The platelet count may drop if the drug is stopped abruptly.

Patients may have a higher risk of developing a blood clot when treated with a TPORA compared to some other ITP treatments.

People who take romiplostim may have an increased risk of developing new or worsening changes in the bone marrow called "increased reticulin."

TREATMENT RESPONSE

1 to 3 weeks.

LIKELIHOOD OF AN INITIAL RESPONSE

Between 60 to 90%.

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

Approximately 80% of patients who respond initially maintain a platelet count of about 50,000 per microliter if treatment is continued. The prevalence of sustained remission off treatment has yet to be established.

OTHER CONSIDERATIONS

Weekly blood tests until a dose of Romiplostim is established to keep the platelet count levels stable, and then go to monthly blood tests or even less frequently.

Some patients taking romiplostim will be able to cease treatment and maintain a safe platelet count on no treatment.

If no response is seen using romiplostim, switching to a different TPORA is recommended.

REFERENCES

https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Splenectomy

Second-line treatment

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

This treatment has been widely used in Immune Thrombocytopenia (ITP); however, changes were made in Australia in 2023 to bring treatment access in line with the Consensus Guidelines for the management of adult Immune Thrombocytopenia in Australia and New Zealand. This improved access to medical therapies, including Thrombopoietin Receptor Agonists (TPORAs), **PRIOR TO** this treatment, meaning that splenectomy should rarely be required to treat ITP.

A discussion with patients 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.

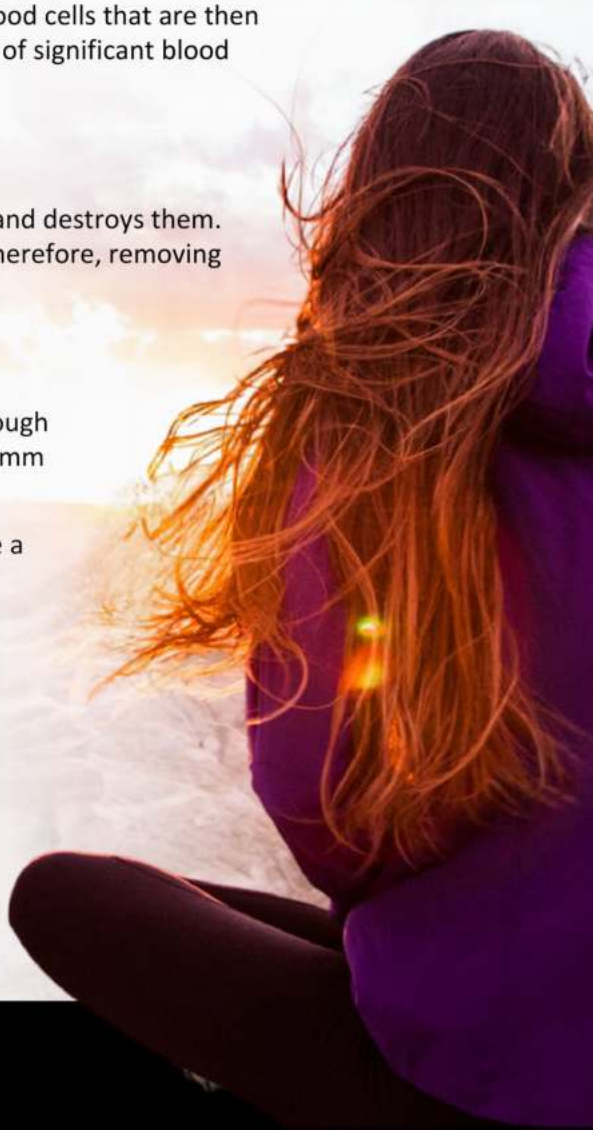
WHERE IS THE SPLEEN?

The spleen is located on the left side of the abdomen and weighs around 160g in the average healthy adult or 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 that are then broken down and stores red blood cells and platelets. 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 treats platelets as foreign and destroys them. The spleen is known to be the site where platelets are removed; therefore, removing the spleen can help keep more platelets circulating in the body.

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 day.

Information for the treatment of Immune Thrombocytopenia in Adults

ITP Australia New Zealand

Splenectomy

Second-line Treatment

LIKELIHOOD OF AN INITIAL RESPONSE

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

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

Splenectomy is associated with the high 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. 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.
4. Patients over 65 years more susceptible to many of these complications.

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*: **based on information from Spleen Australia*
  - Ensure all vaccinations are updated prior to surgery. This includes Pneumococcal, Meningococcal, Haemophilus influenzae type B and the annual influenza vaccine.
  - Be prescribed prophylactic antibiotics (250mg daily) for three years post-surgery.
  - Receive an emergency supply of antibiotics (3000mg) in case a medical review is not immediately available.
  - 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.
- 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 regulates platelet counts.
- The possibility of an accessory spleen may be considered if a patient does not respond or if a relapse occurs.

REFERENCES

- https://itpaustralia.org.au/thanz-aus-nz-itp-guidelines/
- https://www.mja.com.au/journal/2021/216/1/consensus-guidelines-management-adult-immune-thrombocytopenia-australia-and-new
- https://spleen.org.au/