

IMMUNOGLOBULIN REPLACEMENT THERAPY FOR PRIMARY IMMUNODEFICIENCIES

Primary immunodeficiencies (PIDs) are a group of rare, chronic disorders that weaken your immune system and increase your chances of serious infections and their complications.

This fact sheet explains how immunoglobulin (Ig) products can be used to treat PID, to help you make informed decisions about the care that is right for you.

What is immunoglobulin replacement therapy (IRT)?

Immunoglobulins, also known as antibodies, are proteins in the blood that help the immune system fight infections and diseases. The most common PIDs cause the body to produce lower than normal levels of Ig, leading to greater chances of infection.

Treatment with IRT is the most effective treatment for some people with PIDs. IRT uses Ig products that have been purified from blood plasma collected from healthy blood donors. The plasma is pooled together from thousands of donations and then manufactured into Ig products. These products are then carefully tested, screened, filtered, and treated to make sure they are safe for use.

Will immunoglobulin cure my condition?

Ig products will help treat the symptoms of PID, improving quality of life – but they won't 'cure' PID.

IRT replaces the Ig (antibodies) that your body should be making, but it does not correct the defect in antibody production or stimulate your immune system to make more antibodies.

IRT offers protection against many common and serious infections and can significantly reduce their frequency and severity. However, it will not prevent all infections, and you should still be careful, maintain good hygiene and carefully monitor any changes in your condition.

If you are receiving IRT and suspect an infection, contact your doctor. Antibiotics may be required to treat, and sometimes prevent, bacterial infections.

How is IRT given?

Ig products can be injected into a vein (intravenous, IVIg) or injected just below the skin (subcutaneous, SCIg).

- ▶ **IVIg** is usually given monthly (3–4 weekly) in a hospital day clinic
- ▶ **SCIg** requires frequent administration (1–3 times per week) by patients or their carers at home

Both methods are effective and safe. The best method for you will depend on factors such as your medical history and preferences. SCIg is accessed through hospitals approved to offer SCIg programs in the home.

Ig dosage is initially calculated according to weight, treatment method and treatment frequency. The dose may then be adjusted depending on your response to treatment, to ensure you receive the right level of Ig to protect against infection.

What are the side effects of Ig products?

Side effects differ according to the method used and your condition.

- ▶ Most people tolerate **IVIg** well but side effects from IVIg can include headache, fever, chills, nausea, fatigue, or flu-like illness ('systemic' effects) and are usually mild and short-lived. They often occur just after your infusion and can usually be reduced by a slower infusion rate or taking paracetamol. Serious adverse events are rare but include severe allergic reaction (anaphylaxis), aseptic meningitis, reduced kidney function, and blood clots.
- ▶ Systemic side effects from **SCIg** are much less common than from IVIg. Local injection site reactions (eg, redness, itching, and swelling) are common but improve with time.
- ▶ The risk of receiving a blood-borne infection from a plasma-derived blood product is close to zero due to various testing and screening stages and safety measures.

As with any treatment, you should discuss individual risks and benefits with your healthcare team.

How long will I need treatment for?

IRT for PID is usually lifelong. This is because IRT temporarily replaces the Ig that is missing but does not correct the underlying problem.

In addition, IRT only provides temporary protection. Most antibodies are used up or broken down by the body and need to be continually replaced. Repeat doses of IRT are required at regular intervals to maintain adequate levels of Ig.

The amount of Ig product you use may occasionally need to be changed, based on your individual need. This is particularly the case for growing children.

Monitoring and follow-up

As IRT is derived from blood (plasma), and reserved for people with confirmed abnormalities in antibody production, access to Ig products is highly regulated. Your doctor must register you and comply with specific criteria and policies to ensure that product is provided equitably and goes to those that need it the most. Regular contact with your healthcare team is a requirement and an important part of care for anyone receiving IRT. Regardless of your specific condition or the type of IRT you are receiving, your healthcare team will need to see you to monitor:

- ▶ your response to therapy
- ▶ side effects from treatment
- ▶ infections or other health concerns.

If you are self-administering SClg at home, your healthcare team will also want to assess your technique.

Depending on your response to IRT, your treatment team may also discuss adjusting dosage, treatment intervals, or trialling new formulations to ensure your treatment is tailored to you. Recording your symptoms and side effects in a treatment diary (such as the [immunoglobulin management and wellbeing plan](#)) can help you and your healthcare team monitor how you felt after each dose.

Important points to remember

- ▶ IRT is an essential, lifelong therapy for most PIDs that can replace antibody levels, so you are more able to fight infections.
- ▶ IVIg and SClg are two equally effective approaches to administering IRT. The best method is the one that works for your circumstances and preferences, and this may change over time.
- ▶ Ig is carefully tested and purified so the risk of getting an infection or virus from Ig is close to zero.
- ▶ Use a treatment diary. Keep a record of any symptoms and signs, good or bad, that you experience. Treatment diaries are useful to fill in treatment histories and help your specialist develop a more effective treatment plan.

Want to know more?

- ▶ The Criteria for the Clinical Use of Immunoglobulin in Australia outlines the conditions for access to government-funded immunoglobulin products. www.criteria.blood.gov.au/
- ▶ Find out more about Access to Subcutaneous Immunoglobulin (SClg) www.blood.gov.au/SClg
- ▶ Find out more about Access to Intravenous Immunoglobulins (IVIg). www.blood.gov.au/Intravenous-Ig
- ▶ The Immune Deficiencies Foundation Australia provides education, awareness, and advocacy on PIDs. www.idfa.org.au/
- ▶ AusPIPS provides advocacy and support for people living. www.auspips.org.au/
- ▶ ASCIA provides information and resources for consumers and health professionals about IRT and PID allergy.org.au/patients/immunodeficiencies/immunoglobulin-replacement-therapy
- ▶ Find out more about the Value in Prescribing [immunoglobulins program](#)

VALUE IN PRESCRIBING PROGRAM – IMMUNOGLOBULIN PRODUCTS

Increasing the awareness and understanding amongst health professionals of access to immunoglobulin products in Australia, and improving health outcomes for patients through access to better health information to manage their health conditions. Funded by the Australian Government Department of Health through the Value in Prescribing Program: Immunoglobulins Products Grant.

