# IMMUNOGLOBULIN REPLACEMENT THERAPY FOR SECONDARY HYPOGAMMAGLOBULINAEMIA

Hypogammaglobulinaemias (HGGs) are a group of disorders that affect the immune system and prevent it from making enough antibodies. As antibodies – also known as immunoglobulins (Ig) – help protect the body from disease, this means that people with HGGs are more likely to get infections.

HGGs are categorised as primary HGGs or secondary HGGs. Primary HGGs have a genetic cause. Secondary HGGs can result from certain medical therapies, infections, protein loss or malnutrition.

This fact sheet explains how Ig products can be used to treat secondary HGGs. If you have been diagnosed with a secondary HGG, this information can help you make decisions about the care that is right for you.

#### What are the treatment options for my condition?

Secondary HGG can affect people in different ways. Some people will have no increase in infections compared to people who do not have HGG, while others will have more severe or recurrent infections that need treatment and may require hospitalisation.

For some, HGG may be successfully managed by addressing the underlying cause of their HGG. People with milder or no infections may respond well to regular oral antibiotics and recommended vaccinations to prevent infections, instead of receiving immunoglobulin replacement therapy (IRT). For people with very low levels of Ig, or for those who experience persistent serious infections requiring treatment, a trial of IRT may be recommended. IRT may be particularly beneficial for patients who developed secondary HGG from treatment associated with:

- ▶ blood cancers or haematopoietic stem cell transplantation
- ▶ B-cell depletion therapy, Good syndrome, or a solid organ transplant.

#### What is IRT?

Ig, also known as antibodies, are proteins produced by plasma cells in the blood that help the immune system fight infections and diseases. IRT provides additional antibodies to people who do not make enough on their own or produce lower than normal levels of Ig to maintain a healthy immune system. Ig products will help treat the symptoms of HGG, improving quality of life but they won't 'cure' HGG.

IRT can protect against many common and serious infections and reduce their frequency and severity. However, it may not prevent all infections and you should still be careful, maintain good hygiene and carefully monitor any changes in your condition. The lg used in treatment is made from the natural antibodies collected from the pooled blood plasma provided by thousands of healthy blood donors and are carefully tested, screened, filtered, and treated to make sure they are safe for use.

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

## How is IRT given?

Ig products can be infused into a vein (intravenous, IVIg) or injected just below the skin (subcutaneous infusion, SCIg). The best options for you will depend on factors such as your medical history and preference.

- ▶ IVIg is usually given monthly (every 3-4 weeks) while you are monitored in a hospital day clinic
- ▶ SCIg is usually required more often (weekly or multiple times a week) and is self-administered by patients or their carers at home. SCIg is accessed through hospitals approvided to offer SCIg programs in the home.

Both methods are effective and safe. The best method for you will depend on factors such as your comfort and ability to self-administer therapy. You should discuss your options with your doctor or hospital clinic.

Your doctor will work out the appropriate Ig dose for you according to your 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 and risks of Ig products?

Side effects differ according to how IRT is given and your condition.

- ▶ Most people tolerate IVIg well, but side effects can include headache, fever, chills, nausea, fatigue, or flu-like illness ('systemic' effects). These are usually mild and temporary.
- ▶ Side effects often happen during or just after your infusion. A slower infusion rate or taking paracetamol or antihistamines beforehand can reduce the chance of side effects. Serious adverse events are rare, but include severe allergic reactions (anaphylaxis), aseptic meningitis, reduced kidney function, and blood clots.
- ▶ Systemic side effects from SClg are much less common than from IVlg. Local injection-site reactions (eg, redness, itching and swelling) are common but improve with time.
- ▶ The risk of getting a blood-borne infection from a plasma-derived blood product is close to zero due to various testing, screening, and safety measures.

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

#### Monitoring and follow-up

As IRT is derived from the blood plasma of thousands of donors, its supply is limited, and products are expensive to manufacture. Its use is therefore carefully monitored, to ensure that there is equitable access, with priority given to those patients who will benefit from treatment the most, and where there are no or few alternative treatments. Your doctor will register you on an online system called BloodSTAR as part of the eligibility requirement.

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
- ▶ if the IRT is still providing benefit
- ▶ that your HGG is still present
- ▶ side effects from treatment
- ▶ infections or other health concerns.

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

#### Modifying or stopping treatment

Depending on your response to IRT, your healthcare team may discuss changing dosage, treatment intervals, or trialling new formulations to make sure your treatment is tailored to you.

Many people can safely reduce or stop their Ig therapy (under a doctor's guidance). Your doctor may suggest trying a short break from treatment if you have:

- ▶ a long period of time without infections
- ▶ a change in the underlying condition that caused your HGG
- ▶ an increase in antibody levels to an acceptable range, and you are well

Treatment breaks are often done in the warmer months when there is less chance of catching the common cold or flu.

It is recommended that you use a symptom diary (such as the <u>immunoglobulin management and wellbeing plan</u> to keep track of any signs or symptoms (good or bad) that you experience when changing any aspect of your health and wellbeing and treatment regime.

If frequent or severe infections return after stopping Ig products, your treatment may be restarted.

#### Important points to remember

- ▶ IRT is an essential therapy for some people with secondary HGG, helping them fight infections.
- ▶ Ig products have been carefully tested and purified so the risk of acquiring a blood-borne infection from Ig treatment is close to zero.
- ▶ IVIg and SCIg 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.
- ▶ Use a treatment management plan to keep a record of any symptoms and signs, good or bad, that you experience.

  Treatment plans are useful tools to complete and help you and your specialist develop a more effective treatment plan.





## Where to find support

▶ The Immune Deficiencies Foundation Australia provides education, awareness, and advocacy on immunodeficiencies.

#### Want to know more?

- ► For more information on Ig and information on access and consent visit the NPS MedicineWise webpage www.nps.org.au/immunoglobulins
- ▶ For more information about immunoglobulin products in Australia and how to access them see our factsheet
- ► The Criteria for the clinical use of immunoglobulin in Australia define eligibility for access to government-funded immunoglobulin products. <a href="https://www.criteria.blood.gov.au/">https://www.criteria.blood.gov.au/</a>
- ► Keep track of medicines and access important health information using the NPS MedicineWise app https://www.nps.org.au/medicinewiseapp
- ▶ Find out more about access to SCIg https://www.blood.gov.au/SCIg
- ▶ Find out more about access to IVIg https://www.blood.gov.au/Intravenous-Ig

#### **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.



