



Immunoglobulin Replacement Therapy

Frequently Asked Questions

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Q 1: What is immunoglobulin (antibody) replacement therapy?

Immunoglobulin (antibody) replacement therapy (IRT) is one of the most effective and commonly used treatments for some inborn errors of immunity (IEI), including primary immunodeficiencies (PID).

Many people with PID/IEI disorders have insufficient antibodies to adequately fight infections, and this therapy can be lifesaving.

Q 2: Why is IRT used?

In people with PID/IEI disorders, IRT can:

- Treat existing infections.
- Prevent new infections from occurring.
- Prevent long term damage from chronic infections (such as bronchiectasis in the lung).

Q 3: How is IRT made?

Immunoglobulin products are purified from pooled plasma of many healthy blood donors. Plasma is the liquid part of blood that remains when all red blood cells have been removed.

When donors give blood, the red cells and plasma are separated.

The plasma is pooled together and processed in highly specialised and regulated facilities to produce immunoglobulin, which contains a wide variety of antibodies.

Q 4: How is IRT given?

There are two ways that IRT can be given:

- **Intravenous immunoglobulin (IVIg)** infusions are delivered directly into the person's vein, usually in a hospital day clinic. The infusion can take approximately two to four hours. The dose and frequency vary and depend on the person's weight and immunoglobulin levels. Most people receive IVIg doses once every month.
- **Subcutaneous immunoglobulin (SCIg) injections** involve slowly infusing the antibody preparation directly under the skin, which can be done at home, often by using a pump. This is usually done once each week, as only 10-15mL can be infused into any one site. A 10mL infusion can be delivered in half an hour. It may be necessary in some cases for infusions to be more frequent, particularly during introduction to therapy. When beginning SCIg therapy, red lumps may form under the skin. These usually disappear quite quickly, and after a few weeks of therapy usually stop appearing.

Q 5: Are there any side effects of IRT?

IRT is normally very well tolerated and serious side effects are rare. You should tell your doctor if you have any side effects.

Side effects of IRT may include:

- **Risk of blood-borne infections**

Current preventative measures have been greatly improved so that the risk of infection from antibody therapy is now close to zero. Nevertheless, you may wish to discuss this risk with your medical team.

- **Other side effects**

Some people get minor side effects such as low grade fevers or headaches, which can usually be reduced by a slower infusion rate, or treated with paracetamol. People may sometimes experience side effects such as hives, wheezing, or severe headaches. Severe allergic reactions (anaphylaxis) and abnormal kidney function due to IRT are rare.

- **IgA reactions**

Most IgA deficient people receive blood products without difficulty. In rare cases, when a person lacks Immunoglobulin A (IgA) antibodies, reactions similar to anaphylaxis may occur when receiving blood products containing IgA. These reactions are less likely with IgA depleted immunoglobulin products.

Q 6: What are the limitations of IRT?

IRT does not cure the antibody deficiency and does not usually reverse long term organ injury from chronic infections. IRT contains only one of the important components of the immune system's response to infection. For these reasons it is best to start IRT before organ damage has occurred.

Q 7: Who should receive IRT?

IRT is derived from blood (plasma), are in limited supply, and access is restricted. Doctors must follow specific guidelines to ensure that the product goes to people most in need.

IRT should only be used in these cases where scientific and clinical evidence supports its use, and where other therapies are considered less favourable.

IRT is reserved for those people with confirmed abnormalities in antibody production, and who experience recurrent infections.

IRT is also of great benefit for patients with certain autoimmune diseases (such as immune thrombocytopenia and Guillain-Barre syndrome), where it is used to alter the course of the disease (immunomodulation), rather than to replace antibodies that are deficient.

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