

Immunoglobulin Replacement Therapy in Primary Immune Deficiency (PID) Diseases

PATIENT INFORMATION

Immunoglobulin (antibody) replacement therapy is one of the most important and successful therapies for people with primary immune deficiency disease. Many of these people have insufficient antibodies to adequately fight infections, and this therapy can be life saving.



An information service of the Australasian Society of Clinical Immunology and Allergy (ASCIA)

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Why is immunoglobulin replacement therapy used?

In people with primary immune deficiency diseases, immunoglobulin replacement therapy can:

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

How is immunoglobulin replacement therapy 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.

How is immunoglobulin replacement therapy given?

There are two common ways that immunoglobulin replacement therapy can be given; intravenous

(into the vein) and subcutaneous (under the skin).

- **Intravenous immunoglobulin (IVIG)** - is currently the most common means of immunoglobulin replacement therapy in Australia and New Zealand. Infusions are delivered directly into the person's vein, usually in a hospital day clinic or sometimes in the person's home. The infusion takes approximately 2 to 4 hours. The dose and frequency vary, and depend on the person's weight and immunoglobulin levels. Most people receive doses once every month.
- **Subcutaneous immunoglobulin (SCIG)** - the last few years has seen a move toward subcutaneous immunoglobulin (SCIG) injections, especially in Europe. This involves slowly infusing the antibody preparation directly under the skin, which can be done at home using a special pump. This is usually done once or more each week as only 10-15mL can be infused into any one site. However, 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.

Are there any side effects of immunoglobulin replacement therapy?

Immunoglobulin replacement therapy is normally very well tolerated and serious side effects are very rare. However, there are some side effects that you need to be aware of:

- **Risk of blood-borne infections**

As immunoglobulin preparations are derived from blood donors, there will always be a theoretical risk of blood borne infections. In the past there have been cases of transmission of Hepatitis C and other viral illnesses, but there has never been a case of HIV transmitted by this means.

Transmission of Hepatitis C has not occurred in Australia or New Zealand. Current preventative measures have been greatly enhanced 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. Occasionally people experience hives or wheezing or

rarely severe headaches. Rare cases of severe allergic type reactions and abnormal kidney function have been described.

- **IgA reactions**

In rare cases, when a person lacks Immunoglobulin A (IgA) antibodies, reactions similar to severe allergy may occur when receiving blood products containing IgA. However, the great majority of IgA deficient people receive blood products without difficulty. These reactions are less likely with current IgA depleted Immunoglobulin products.

You should notify your doctor of any side effects you might experience.

Limitations of immunoglobulin replacement therapy

Immunoglobulin replacement therapy contains essentially only one of the important components of the immune system's response to infection. This therapy does not cure the antibody deficiency, and will seldom reverse long term organ injury from chronic infections. For this reason it is best to start before organ damage has occurred.

Availability of immunoglobulin replacement therapy

Changes in the way these products are supplied should reduce product shortages that have occurred in Australia in recent years. Such shortages have not been a problem to date in New Zealand. Regardless of product availability, immunoglobulin replacement therapy should be reserved for those people with confirmed abnormalities in antibody production, and who experience recurrent infections.

Immunoglobulin replacement therapy for other diseases

Immunoglobulin therapy 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.

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

Is there any support for people in Australia and New Zealand with primary immune deficiency diseases?

ASCIA has established a Register of people with primary immune deficiency diseases so these conditions can be better understood and managed.

The following two foundations are part of an international alliance to provide support for patients with primary immune deficiency disease:

- Immune Deficiencies Foundation of Australia (IDFA) www.idfaustralia.org
- Immune Deficiencies Foundation of New Zealand (IDFNZ) www.idfnz.org.nz

Further information

Common Variable Immune Deficiency

www.allergy.org.au/content/view/377/335/

Primary Immune Deficiency Diseases

www.allergy.org.au/content/view/157/270/

Severe Combined Immune Deficiency

www.allergy.org.au/content/view/150/317/

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