Secondary Immunodeficiency

A guide for patients, their families, friends and healthcare professionals

IDFA
Immune Deficiencies Foundation
www.idfa.org.au Australia

Raising awareness and supporting patients with immunodeficiencies in Australia.

Secondary Immunodeficiency

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IDFA is delighted to extend its support and advocacy activities to include patients with Secondary Immunodeficiency. This will include in the first instance, community members and other patients requiring immunoglobulin replacement therapy. As we continue to grow and with the help of our members and supporters, we are looking forward to being able to offer our services to a larger community.

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Immunodeficiency

Immunodeficiency is also commonly referred to as an immune deficiency, immune disease, immune disorder or immunodeficiency.

A common feature of many immunodeficiencies is a loss of **antibodies** (or immunoglobulins) or failure of antibody function. The decrease in antibodies causes the immune response to be deficient or absent, resulting in increased susceptibility to infection.

Immunodeficiency disorders are either **Primary** (of genetic origin) or **Secondary** (due to treatment, transplantation or disease).

Secondary Immunodeficiency (SI)

Secondary Immunodeficiency caused by a decrease in antibodies occurs most commonly as a consequence of:

- Chemotherapy
- Haematological malignancies
- Renal or gastrointestinal immunoglobulin loss
- Organ transplantation
- Infectious diseases
- Corticosteroid, anticonvulsant or immunosuppressive medications, e.g. Rituximab (RTX).

Secondary Immune Deficiency can be caused by:

- Chronic Lymphocytic Leukaemia
- Multiple Myeloma
- Non-Hodgkin Lymphoma

- Good's Syndrome
- Any lymphomas or cancers of the lymph nodes or immune system
- Protein Losing Enteropathy
- Lymphoreticular Malignancy
- Antibody Deficiency due to treatment for Autoimmune Disease using chemotherapy-like reagents

The Immune system

The immune system is a network of cells, tissues, and organs that work together to protect the body from infection. Our immune system is made up of B and T Cells, Phagocytes and Complement. They work together to provide the **immune response.**

B-Cells become plasma cells which make immunoglobulins (antibodies).

Immunoglobulins kill pathogens (virus, bacteria, fungi, parasites, protozoa and proteins) that attack our body. They also protect our blood, nose, lungs and intestines.

Immunoglobulin G molecule (IgG)

There are 3 main kinds of **T-Cells**:

1) Killer T-Cells kill pathogen infected and tumour cells

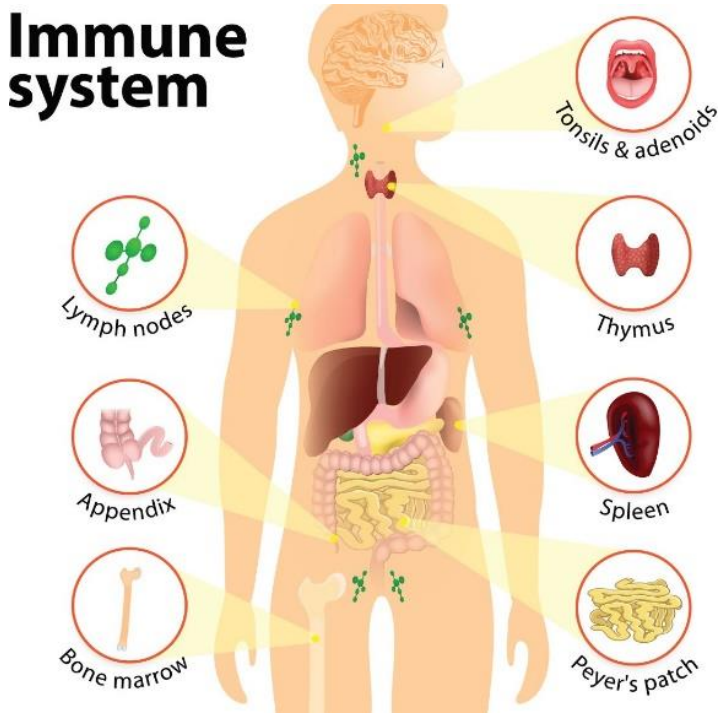
2) Helper T-Cells call in more Killer T-Cells to attack and also tell B-Cells when to make antibodies.

3) Regulatory T-Cells which communicate with the B-Cells and T-Cells to their activity when the body has recovered.

Phagocytes also kill pathogens by ingesting them.

Complement helps phagocytes clear, as well as directly destroying pathogens.

Because immunodeficiency is a disorder where the immune response (the body's defense system) is reduced or absent, **infections** are caused when these components are not present or do not function.



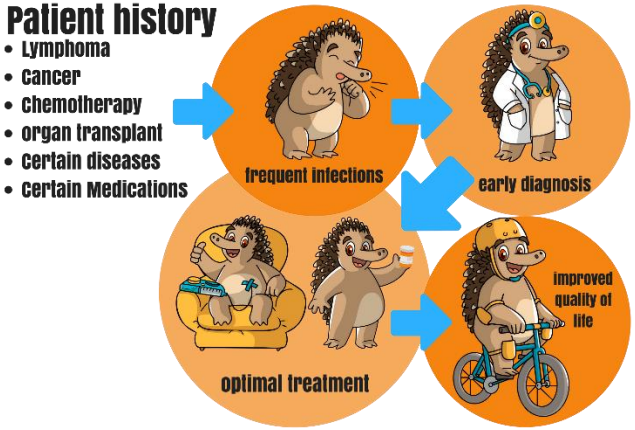
Organs of the Immune System

The organs affected by secondary immunodeficiency can include the Skin, Sinuses, Middle Ear, Lungs, Spleen, Bowel and Blood.

Because one of the most important functions of the normal immune system is to protect us against infection, patients with secondary immunodeficiency are more susceptible to infections in these organs and endure recurrent health problems.

Autoimmune conditions such as chronic sinusitis, bowel disease, thyroid disease, lupus and arthritis are also common.

Warning signs



People with secondary immunodeficiency can present in many ways, sometimes with **similar symptoms** to those in the **warning signs of primary immunodeficiency**. The 10 warning signs for primary immunodeficiency are:

1. Eight or more ***ear infections*** within one year
2. Two or more serious ***sinus infections*** within one year
3. Two or more ***months on antibiotics*** with little effect
4. Two or more ***pneumonias*** within one year
5. Failure of an infant to gain weight or grow normally
6. Recurrent deep skin or organ ***abscesses***
7. Persistent ***thrush*** in mouth or elsewhere on skin after age one
8. Need for ***intravenous antibiotics*** to clear infections
9. Two or more deep seated infections such as ***sepsis, meningitis or cellulitis***
10. ***Family history*** of primary immunodeficiency

Symptoms

Symptoms include:

- constant infections
  - upper respiratory tract
  - lungs
  - middle ear
  - sinus
  - skin
- bronchiectasis
- gastrointestinal symptoms
- immune dysregulation
- organ damage

Diagnosis

Tests potentially helpful for making a diagnosis include:

- History of frequency and type of infections
- Full blood count
- Immunoglobulin levels (+/- IgG subclasses)
- Specific antibody responses to pneumococcus, *Haemophilus Influenzae* and tetanus
- Chest and sinus X-Rays

In patients with low immunoglobulin levels, vaccination response to the pneumococcal polysaccharide vaccine should be checked to give an indication regarding immune function.

Diagnostic terms include Secondary Antibody Deficiency, Specific Antibody Deficiency, Hypogammaglobulinaemia or Common Variable Immune Deficiency.

Medical Management

Medical management includes:

- Monitoring of antibody levels
- Investigations for end organ damage (chest X-Ray, endoscopy, colonoscopy)
- Killed vaccines
- Microbiological screening for infections

It is likely patients may have several specialists in different fields.

IDFA recommends the following:

- Keep a file of your Specialist names and contact numbers, referrals, medical test results and specialist letters. Take this to appointments and Emergency.
- Keep a diary of your infections and treatments. Take this to appointments and Emergency.
- Keep your Medicare, Heath Fund, Concession and Medical Record Number (MRN) in your file and in your wallet.

Treatment

Where there is antibody deficiency, immunoglobulin replacement therapy has been shown to be effective in reducing the severity and frequency of infections.

Prophylactic antibiotics may also be used.

Immunoglobulin Replacement Therapy

Immunoglobulin replacement is the main treatment for the majority of immunodeficiencies.

Immunoglobulin products are made with plasma from blood donors. Plasma is sent to a specialised facility to produce immunoglobulin which contains antibodies. This is used for patients requiring immunoglobulin replacement therapy.

There are two ways immunoglobulin replacement therapy can be given; intravenously (into the vein) and subcutaneously (under the skin).

The dose and frequency vary dependent on the patient's weight and immunoglobulin levels.

Intravenous immunoglobulin (IVIg) infusions are delivered usually in hospital or sometimes in the person's home every 3-4 weeks.

Intravenous immunoglobulin (IVIg)

Subcutaneous immunoglobulin (SCIg) infusions are self-administered in the home using a syringe or special pump 1-2 times a week.

Subcutaneous immunoglobulin (SCIg) via pump

Subcutaneous immunoglobulin (SCIg) via push method

Some patients experience side effects such as low-grade fever, headache and leg or joint pains. These can be reduced by paracetamol or ibuprofen, hydrocortisone and a slower infusion rate.

Remember to hydrate well before and during infusions.

Lifestyle

A healthy lifestyle and protection as far as possible from infection is recommended. This includes having regular dental and physical check-ups.

Healthy eating, good hygiene, avoiding contact with people with infections and regular exercise contribute to both physical and mental well-being.

Eat healthy food

Maintain a healthy lifestyle

Fatigue

Fatigue is common, so ensure plenty of rest and hydration. The IDFA “Fatigue” Booklet may be helpful.

Isolation

Social isolation can be felt due to frequent infections, hospitalisation and treatment. This can lead to psychological issues. Explain your condition to family, friends and work colleagues. It is important to maintain friendships and seek professional support when needed.

IDFA

IDFA provides advocacy and educational, practical, social and emotional support through:

- Resources
- Teleconferences
- Conferences and patient meetings
- Social media including closed Facebook pages for adults and young adults
- Patient Support Officer

Patients, carers, family members and healthcare professionals can become members of IDFA.

Membership is free.

Join online www.idfa.org.au

Phone 1800 100 198 or email info@idfa.org.au

Glossary

Antibodies: or immunoglobulins, are large Y-shaped proteins which identify and help remove foreign antigens such as viruses and bacteria. They are produced by white blood cells known as B cells.

Bronchiectasis: Is usually a result of a chronic infection where damage to the airways result in some parts of the lung becoming scarred and dilated, which prevents the clearing of mucous.

Immune dysregulation: Uninhibited or unregulated immune response.

IVIg: Intravenous immunoglobulin (into the vein).

Killed Vaccines: Are made from microorganisms (viruses, bacteria, other) that have been killed through physical or chemical processes.

Primary immunodeficiency: Immunodeficiency disorder of genetic origin.

SCIg: Subcutaneous immunoglobulin (under the skin).

Secondary immunodeficiency: Immunodeficiency disorders due to treatment, transplantation or disease.

References

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