Common Variable Immunodeficiency (CVID)

Common variable immunodeficiency (CVID) is one of the most common primary immunodeficiency (PID) disorders, also known as inborn errors of immunity (IEI)**,** which affects both males and females.

Many people with CVID are not diagnosed until they are adults, however symptoms of CVID may appear in childhood.

CVID is a PID/IEI disorder

CVID and other PID/IEI disorders are caused by defects in cells of the immune system, and are usually inherited. PID/IEI disorders are different to AIDs (acquired immunodeficiency syndrome), that is due to human immunodeficiency virus (HIV).

In most cases the causes of CVID are not known. Studies have identified a small number of abnormal genes that are involved in immune cell development in around one in ten people with CVID.

How is the immune system different in people with CVID?

The main role of the immune system is to defend against infections and other invaders (such as cancer cells) whilst protecting the body's own cells.

Antibodies, also known as immunoglobulins, are proteins made by specialised white blood cells, called B cells (B lymphocytes). Antibodies recognise germs so they can be removed by the rest of the immune system.

For B cells to work effectively they usually need help from T cells (T lymphocytes), which are another type of specialised white blood cell in the immune system.

Most people with CVID have normal numbers of B cells. However, these B cells do not mature properly to produce effective antibodies, or they don't receive the help needed from T cells, to develop normal antibody responses.

People with CVID will vary in their ability to make effective antibody responses, due to decreased levels of:

- All three major types of immunoglobulins (IgG, IgA and IgM) or
- Immunoglobulins G and A (IgG, IgA) or
- Only Immunoglobulin G (IgG).

Diagnosis of CVID is usually confirmed by abnormal blood test results and medical history.

Reduced antibody responses in CVID lead to infections

Most people with CVID have frequent infections due to their reduced antibody responses. These infections usually occur in the ears, sinuses, nose and lungs. Other common infections in CVID include conjunctivitis, and persistent diarrhea. Unusual infections may also occur, including meningitis and blood stream infection.

Although people who don't have CVID can also have these infections, the difference in people with CVID is that the infections are unusually frequent, prolonged, severe or resistant to normal treatment.

Chronic infections can lead to organ damage

Infections that are not treated properly in people with CVID can result in damage to organs in the body, such as the sinuses, causing chronic sinusitis, or the airways of the lung (bronchi), causing bronchiectasis.

This organ damage can lead to tissue damage, causing ongoing mucus secretion, and the persistent need to clear phlegm (sputum) or thick white, yellow or green mucus from the nose. Once tissue damage is established, infection tends to become more persistent and difficult to clear.

Non-infectious complications of CVID

Autoimmune disease can affect some people with CVID. Autoimmunity occurs when the body doesn't recognise its own cells and attacks them. This can damage normal cells, including blood cells, skin, hair, bowel and hormone producing glands.

Granulomatous disease can cause organ damage that results from immune cells, which form small nodules in different tissues. These include the lungs, lymph nodes, liver and spleen.

Tumours of the immune system including lymphoma may occur in some people with CVID.

Treatment options for CVID

Treatment plans for CVID should consider several factors. These include antibody levels and responses, severity and range of infections and symptoms, and the ongoing need for treatment to prevent infections.

Treatment options include:

- **Immunoglobulin replacement therapy** (IRT), which is given as intravenous immunoglobulin (IVIg) or subcutaneous immunoglobulin (SCIg) infusions.
- **Antibiotics combined with IRT** can greatly improve the health and wellbeing of people with CVID, by reducing infections and preventing development of chronic lung disease.
- **Clearance of airway secretions.**
- **Corticosteroid therapy** for control of autoimmune disease.
- **Management of gastrointestinal inflammation.**

Support for people with CVID and their families

The following organisations provide support for people with CVID and their families:

- AusPIPS www.auspips.org.au
- Immune Deficiencies Foundation of Australia (IDFA) www.idfa.org.au
- Immune Deficiencies Foundation of New Zealand (IDFNZ) www.idfnz.org.nz