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Information
FOR PATIENTS, CONSUMERS AND CARERS

Common variable immunodeficiency (CVID)

Common variable immunodeficiency (CVID) is one of the more common primary immunodeficiency diseases, and affects both males and females. Most people with CVID are not diagnosed until 30 to 40 years of age however around 1 in 5 people with CVID will have symptoms of CVID in childhood, from 2 to 16 years of age.

CVID is a primary immunodeficiency

Primary immunodeficiency diseases such as CVID are caused by defects in the development and maturing of cells in the immune system and are usually inherited. These diseases are called primary immunodeficiencies because no other causes for immunodeficiency such as infections, drugs or toxins can be identified.

The causes of CVID are not yet known. Studies have however identified a small number of abnormal genes that are involved in immune cell development in less than 1 in 10 people with CVID.

Reduced antibody responses in CVID lead to infections

Normal antibody responses are required to fight infections caused by germs such as bacteria, moulds, parasites and viruses. Most people with CVID have recurrent infections due to their reduced antibody responses. These infections usually occur in the ears, sinuses, nose and lungs, which may start in early childhood, adolescence or adult life. Other common infections in CVID include conjunctivitis and persistent diarrhoea. Unusual infections may also occur including meningitis and blood stream infection.

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

Chronic infections can lead to organ damage

When infections are unrecognised or not treated properly in people with CVID, they 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.

How is the immune system different in people with CVID?

The main role of the immune system is to fight infections from germs by producing immunoglobulins (antibodies) which respond to germs and either prevents the germs from causing illness or cause the illness to end.

B cells (B lymphocytes) are specialised white blood cells in the immune system that can produce immunoglobulins (antibodies) which recognise and inactivate germs such as bacteria and viruses, thereby preventing infection. For B cells to work effectively they usually need help from other immune cells such as the 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 normally to produce effective antibodies **or** they don't receive the help needed (e.g. from T cells) to develop normal antibody responses.

Different individuals with CVID will vary in their ability to make effective antibody responses. In some people with CVID there is a decrease in all three major types of immunoglobulins (IgG, IgA, IgM), whilst some people with CVID will have decreased levels of IgG and IgA and in others, only IgG is reduced.

How is CVID diagnosed?

Diagnosis of CVID is usually confirmed by abnormal blood test results. Blood tests include measurement of:

- Immunoglobulin levels including IgG, IgM and IgA
- Numbers of B and T cells
- B cell function by checking antibody responses to vaccines which may include Pneumococcus, Haemophilus influenza type B, diphtheria toxin and tetanus toxoid
- T cell function

Non-infectious complications of CVID

Autoimmune disease affects around 1 in 5 people with CVID. In these conditions, cells in the immune system can attack and damage normal cells, including blood cells, skin, hair, bowel and hormone producing glands.

Granulomatous disease can cause organ damage and results from collections of immune cells which form small nodules in different tissues, such as the lungs, lymph nodes, liver and spleen. The cause of granulomas is unknown.

Tumours of the immune system including lymphoma have been described, but only occur in a small minority of people with CVID.

Treatment options for CVID

Treatment plans for CVID require consideration of several factors including antibody levels, antibody responses, severity and range of infections, severity and range of symptoms and the ongoing need for treatment to prevent infections. Options include:

- **Immunoglobulin replacement therapy** - given as intravenous immunoglobulin (IVIg) infusions or subcutaneous (SCIg) infusions of immunoglobulin or gammaglobulin.
- **Antibiotics** - Immunoglobulin replacement therapy combined with antibiotic therapy has greatly improved the outlook for 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.**

Is there any support for people with CVID?

The following two foundations are part of an international alliance to provide support for people with primary immunodeficiency disease:

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

Further information

- Primary Immunodeficiency Resource Center (Jeffrey Modell Foundation) www.info4pi.org
- Primary Immunodeficiency Association UK www.pia.org.uk
- European Society for Immunodeficiencies www.esid.org

The Australasian Society of Clinical Immunology and Allergy (ASCIA) is the peak professional body of clinical immunology and allergy specialists in Australia and New Zealand.
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