Definition:

Common Variable Immunodeficiency (CVID) is a disorder characterized by low levels of serum immunoglobulins (antibodies) and an increased susceptibility to infections. The exact cause of the low levels of serum immunoglobulins is usually not known but is probably due to a genetic abnormality of some kind.

It is, as the name suggests, a relatively common form of immunodeficiency, with a prevalence of approximately 1 case per 50,000 in the general population.
The degree and type of deficiency, and also the symptoms, varies from patient to patient, hence, the word "variable." In some patients there is a decrease in IgG and IgA; in others, all three major types (IgG, IgA and IgM) of immunoglobulins may be decreased. The clinical signs and symptoms also vary from mild to severe. Frequent and unusual infections may occur for the first time during early childhood, adolescence or adult life. In the majority of patients, however, the diagnosis is not made until the 3rd or 4th decade of life.

SIGNS AND SYMPTOMS:

The presenting features of most patients with CVID are recurrent infections involving the ears, sinuses, nose, or lungs. When the lung infections are severe and occur repeatedly, permanent damage to the bronchial tree may occur and a chronic condition of the bronchi (breathing tubes) develops causing widening and scarring of these structures. This condition is known as bronchiectasis.
The organisms commonly found in these infections are bacteria that are widespread in the population and often cause pneumonia (Haemophilus influenzae, pneumococci, and staphylococci).

Patients with CVID may also develop enlarged lymph nodes in the neck, the chest or abdomen. The specific cause is unknown, but enlarged lymph nodes may be due to infection, immune dysregulation, or both. Similarly, enlargement of the spleen is relatively common as is enlargement of collections of lymphocytes in the walls of the intestine called Peyer's patches.
Some patients with CVID who may not be receiving optimal gamma globulin therapy may also develop a painful inflammation of one or more joints. This condition is called polyarthritis. In the majority of cases the joint fluid does not contain bacteria. The typical arthritis associated with CVID may involve the larger joints such as knees, ankles, elbows and wrists. The smaller joints (i.e. the finger joints) are rarely affected. Symptoms of joint inflammation usually disappear with adequate gamma globulin therapy and appropriate antibiotics.
Some patients with CVID report gastrointestinal problems such as abdominal pain, bloating, nausea, vomiting, diarrhoea and weight loss. Testing of the digestive tract may reveal malabsorption of fat and certain sugars or in some patients with digestive problems, a small parasite called Giardia may be identified in the stool samples. These parasites can be eliminated by medication. Involvement of the gastrointestinal tract may in some instances, interfere with normal growth in children or lead to weight loss in adults. Both males and females may have CVID.

DIAGNOSIS:

CVID is suspected in children and adults who have a history of recurrent infections involving ears, sinuses, bronchi, and lungs. The diagnosis is confirmed by finding a low level of serum immunoglobulins. IgG will be either low or absent, while IgA and IgM may be completely absent in some patients or normal in others.
Patients who have received complete immunizations against polio, measles, diphtheria and tetanus will usually have very low or absent antibody levels to one or more of these vaccines.
As more is learned about the genetic abnormalities that cause of CVID, there will eventually be available genetic testing to identify the specific gene defect for each patient. This will help to identify any other family members who may also have the defect but as yet have not had any symptoms.

TREATMENT:

For patients with CVID, gamma globulin treatment almost always brings improvement of symptoms. Gamma globulin is extracted from a large pool of human plasma consisting mostly of IgG and containing all the important antibodies present in the normal population. It is given either intravenously (IV) or subcutaneously (SUBCUT.).
Patients with chronic sinusitis or chronic lung disease may also require long term treatment with broad-spectrum antibiotics.

If bronchiectasis has developed, physical therapy and daily postural drainage are needed to remove the secretions from the lungs and bronchi. Patients with gastrointestinal symptoms and malabsorption are evaluated for the presence of Giardia, rotavirus and a variety of other gastrointestinal infections. Most patients with immunodeficiency and arthritis respond well to treatment with gamma globulin.

LONG TERM:

Gamma globulin replacement combined with antibiotic therapy has greatly improved the outlook of patients with Common Variable Immunodeficiency. The aim of the treatment is to keep the patient free of infections and to prevent the development of chronic lung disease. The long term picture depends on how much damage to the lungs or other organs occurred before diagnosis and treatment was started, and how successfully infections can be prevented in the future by using gamma globulin and antibiotic therapy

These booklets are designed to offer medical professionals, patients and their families' basic information about these rare disorders of the immune system.
For further information please contact your immunologist, paediatrician, physician or the National IDF Health Coordinator

Other booklets available:

Living with PID
What is IVIG Therapy
Recurrent infections
Genetic Testing & PID
X-linked Agammaglobulinaemia (XLA)
Chronic Granulomatous disorder (CGD)
Selective IgA Deficiency

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Immune Deficiencies Foundation
Asia Pacific Alliance

The Immune Deficiency Foundation Asia-Pacific Alliance, IDFAPA.

An alliance of not-for -profit PID Patient support groups across the Asia Pacific Region.

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Living with Primary Immune Deficiency Disorders

Common Variable Immune Deficiency

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