

Immunodeficiency

What is immunodeficiency?

Immunodeficiency occurs as a result when part of the immune system is not working properly or is absent. Deficiencies which are hereditary and congenital are referred to as primary immunodeficiency disease or PIDD. Some immunodeficiency disorders can be acquired, such as secondary immune deficiency disease, where the immune system is compromised due to environmental factors, such as HIV or chemotherapy.

Your immune system is normally composed of many white blood cells, called B cells and T cells, which help to fight off foreign invaders such as germs, bacteria, and fungi. B cells are responsible for releasing antibodies, which are proteins produced to remove foreign invaders (antigens) from the body. T cells destroy foreign or abnormal cells. Complement proteins are also playing a protective role in the immune system. Immunodeficiency occurs when your body is unable to defend itself against antigens due to lack or dysfunction of disease fighting proteins. This results in a pattern of repeated infections, which can be severe and difficult to cure.

Who is at Risk?

Individuals with a family history of primary immunodeficiency have a higher risk for developing these disorders. Patients who have HIV or cirrhosis of the liver are also at a greater risk for developing immunodeficiency. Aging can weaken your immune system as organs which produce white blood cells can shrink and, therefore, produce fewer white blood cells. Finally, inadequate amounts of protein in your diet or an inadequate amount of sleep can weaken your immune system.

Symptoms/Diagnosis

Severe cases of PIDD can become apparent during infancy, such as T-cell or combined immunodeficiencies. Many cases of immunodeficiency are identified in adults and can be first suspected through a supporting clinical history of recurring infections. Important indications for immunodeficiency can include recurrent pneumonia, ear infections, or sinusitis, poor growth, weight loss, recurrent infections, multiple courses of antibiotics required to treat infections, family history, or autoimmune disease.

Your doctor will take a detailed history from you looking for signs of immunodeficiency. They will order blood work to evaluate your white blood cell count, T cell count, and immunoglobulin levels. Vaccines can also help by testing your immune system for antibody response. Your doctor will give you a vaccine then test the blood for its response. If your immune system is functioning normally, it will produce antibodies in response to the vaccine.

What are Treatment Options?

Treatment options for immunodeficiency have greatly helped to improve the quality of life for patients. These treatment options, such as Intravenous or Subcutaneous Immunoglobulin Therapy, provides your body with the antibodies which were deficient in order to help fight off infections. Other treatment options can include transplantation of bone marrow, stem cell, or thymus, preventative

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antibiotics, and strategies to manage autoimmune disease. Gene therapy is also on the horizon in treatment of specific types of PIDD.