

Primary immunodeficiencies and related diseases.

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Abstract

Primary Immune Deficiency Diseases (PIDDs) are a group of rare inherited diseases that affect the immune system. People with PIDDs may be susceptible to persistent, debilitating infections such as Epstein-Barr virus (EBV), which can raise the risk of cancer if the immune system is not functioning properly. Some PIDDs have the potential to be lethal. Depending on the severity, Primary immunodeficiencies can be diagnosed in infancy, youth, or adulthood.

Keywords: Primary immune deficiency diseases, Epstein-Barr virus.

Introduction

Primary immunodeficiency disorders, also known as primary immune disorders or primary immunodeficiency, weaken the immune system, making it more susceptible to infections and other health problems. Many people with primary immunodeficiency are born without parts of the immune system or with a faulty immune system, rendering them more vulnerable to infection-causing microorganisms.

Researchers have discovered around 300 different types of primary immunodeficiency diseases thus far. Some types are so little that they go undiscovered until they reach adulthood. Other types are so severe that they're found quickly after a baby is born with them [1-3].

A variety of primary immunodeficiency illnesses can be benefited from treatments that enhance the immune system. People with Alzheimer's disease are benefiting from continuous research that is leading to better therapies and a higher quality of life.

Ask your doctor to check for the likelihood of a primary immunodeficiency whenever you have an infection that is recurrent, persistent, unusual, severe, and/or shared by family members (PI).

Signs of Primary Immunodeficiency

Although being more susceptible to infection than the normal person is the most prevalent symptom of a PI, there are other signs that you may have one. Swollen spleen, liver, or lymph nodes, blood vessel inflammation, and autoimmune symptoms such as inflammatory bowel disease are all examples [1].

One of the most prominent signs of primary immunodeficiency is illnesses that are more common, last longer, or are more difficult to treat than infections in persons with a sound immune system. You could potentially catch infections that a person with a robust immune system would be immune to (opportunistic infections).

Primary immunodeficiency disorders have different signs and symptoms depending on the type, and they also differ from person to person. The following are some of the signs and symptoms of primary immunodeficiency:

- Pneumonia, nose & ear infections, meningitis, or skin infections that occur recurrently
- Internal organ inflammation and infection
- Low platelet count or anaemia are examples of blood diseases.
- Cramping, loss of appetite, nausea, and diarrhoea are all symptoms of digestive disorders.
- Growth and development are slowed.
- Autoimmune diseases, such as rheumatoid arthritis

Causes

Many primary immunodeficiency illnesses are hereditary, meaning one or both parents are affected. Many of these immune system flaws are caused by problems with the genetic code, which serves as a blueprint for manufacturing the body's cells (DNA) [4].

There are around 300 different forms of primary immunodeficiency illnesses, and researchers are continually discovering new ones. They can be divided into six categories according to whatever element of the immune system is affected:

- Deficiencies in B cells (antibodies)
- Deficiencies in T cells
- B and T cell deficits in combination
- Phagocytes with a defect
- Deficiencies in complement

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- Unidentified (idiopathic)
- Factors that are at risk

Having a family history of a primary immune deficiency disorder is the sole recognised risk factor, which enhances your chances of developing the disease.

If you have a major immune deficiency illness and want to start a family, you should obtain genetic counselling.

Complications

Complications are different based on the type of primary immunodeficiency condition you have [5]. They may include the following:

- Infections that recur
- Disorders of the immune system
- Heart, lungs, neurological system, or digestive tract damage
- Growth is slowing.
- Cancer risk is higher.
- Death due to a severe infection

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