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1. PRIMARY IMMUNODEFICIENCY DISORDERS

1.Severe Combined Immunodeficiency (SCID)

Severe combined immunodeficiency (SCID) is a group of rare disorders caused by mutations in different genes involved in the development and function of infection-fighting immune cells. Infants with SCID appear healthy at birth but are highly susceptible to severe infections. The condition is fatal, usually within the first year or two of life, unless infants receive immune-restoring treatments, such as transplants of blood-forming stem cells, gene therapy, or enzyme therapy. More than 80 percent of SCID infants do not have a family history of the condition. However, development of a newborn screening test has made it possible to detect SCID before symptoms appear, helping ensure that affected infants receive life-saving treatments.

More than a dozen genes have been implicated in SCID, but gene defects are unknown in approximately 15 percent of newborn-screened SCID infants, according to an NIH-funded study. Most often, SCID is inherited in an autosomal recessive pattern, in which both copies of a particular gene (inherited from the mother and one from the father) contain defects. The best-known form of autosomal recessive SCID is caused by adenosine deaminase (ADA) deficiency, in which infants lack the ADA enzyme necessary for T-cell survival. X-linked SCID, which is caused by mutations in a gene on the X chromosome, primarily affects male infants. Boys with this type of SCID have white blood cells that grow and develop abnormally. As a consequence, they have low numbers of T cells and natural killer cells, and their B cells do not function.

Symptoms of SCID occur in infancy and include serious or life-threatening

infections, especially viral infections, which may result in pneumonia and chronic diarrhea. *Candida* (yeast) infections of the mouth and diaper area and pneumonia caused by the fungus *Pneumocystis jirovecii* also are common.

2.Interferon Gamma, Interleukin 12 and Interleukin 23 Deficiencies

Interferon gamma, interleukin 12 and interleukin 23 deficiencies are rare, inherited immune disorders in which the body fails to produce one or more of these signaling molecules, which allow infection-fighting immune cells to communicate. Deficiencies in these molecules lead to increased susceptibility to bacterial and viral infections. Many people with these deficiencies develop granulomas, or inflammatory lesions that form in tissues and organs because of recurring infections. While many of these deficiencies begin to cause symptoms in infancy or childhood, some symptoms appear later in life. Treatment includes antibiotic therapy to prevent infections and, in some cases, bone marrow transplant from a healthy donor.

3.Leukocyte Adhesion Deficiency (LAD)

Leukocyte adhesion deficiency (LAD) is a rare, inherited immune disorder in which immune cells called phagocytes are unable to move to the site of an infection to fight off invading pathogens. People with LAD experience recurrent, life-threatening infections and poor wound healing. LAD is caused by a mutation in the gene ITGB2, which provides instructions for the phagocyte surface molecule CD18. Treatments for LAD include antibiotics to prevent and treat infection and, in some cases, bone marrow transplants from a healthy donor.

4.Hyper-Immunoglobulin M Syndromes

Hyper-immunoglobulin M (IgM) syndromes are rare, inherited conditions in which the immune system fails to produce normal levels of the antibodies IgA, IgG and IgE but can produce normal or elevated levels of IgM. Various gene defects that impair communication between T cells and antibody-producing B cells can lead to hyper-IgM syndromes. Hyper-IgM syndromes can cause severe respiratory infections in infancy and a higher risk of rare infections

throughout life. Treatment includes regular intravenous or subcutaneous antibody replacement therapy, anti-fungal prophylactics, and in some cases, bone marrow transplant from a healthy donor.

5.PI3 Kinase Disease

PI3 kinase (PI3K) disease is a rare disorder that severely impairs the immune system's ability to fight bacterial and viral infections. The disease also increases a person's risk of lymphoma, a type of immune cell cancer. PI3K disease is sometimes called PASLI disease (short for PI3K-p110δ activating mutation causing senescent T cells, lymphadenopathy, and immunodeficiency) or APDS (for activating PI3K delta syndrome).
NIAID scientists and their collaborators identified PI3K disease in 2013. NIAID supports research to learn more about the genetic cause of the disease and to investigate potential therapies for its symptoms and complications.
PI3K disease is caused by mutations in the genes *PIK3CD* or *PIK3R1*, which provide instructions for producing a protein called PI3K-p110δ. These mutations can affect the immune system by overactivating an important immune system signaling pathway. This overactivation launches a chain reaction leading to disruptions in the normal development of B and T cells, which play a key role in fighting pathogens, and to increased susceptibility to infection.

2.SECONDARY IMMUNODEFICIENCY DISORDERS

1.HIV(HUMAN IMMUNODEFICIENCY VIRUS)

HIV is a virus that damages the immune system. The immune system helps the body fight off infections. Untreated HIV infects and kills CD4 cells, which are a type of immune cell called T cells. Over time, as HIV kills more CD4 cells, the body is more likely to get various types of infections and cancers.

HIV is transmitted through bodily fluids that include:

- blood

- semen
- vaginal and rectal fluids
- breast milk

The virus doesn't spread in air or water, or through casual contact.HIV is a lifelong condition and currently there is no cure, although many scientists are working to find one. However, with medical care, including treatment called antiretroviral therapy, it's possible to manage HIV and live with the virus for many years.

Without treatment, a person with HIV is likely to develop a serious condition called AIDS. At that point, the immune system is too weak to fight off other diseases and infections. Untreated, life expectancy with AIDS is about three yearsTrusted Source. With antiretroviral therapy, HIV can be well-controlled and life expectancy can be nearly the same as someone who has not contracted HIV.

AIDS is a disease that can develop in people with HIV. It's the most advanced stage of HIV. But just because a person has HIV doesn't mean they'll develop AIDS.HIV kills CD4 cells. Healthy adults generally have a CD4 count of 500 to 1,500 per cubic millimeter. A person with HIV whose CD4 count falls below 200 per cubic millimeter will be diagnosed with AIDS.

2.LEUKEMIA

Leukemia is a cancer of the blood cells. There are several broad categories of blood cells, including red blood cells (RBCs), white blood cells (WBCs), and platelets. Generally, leukemia refers to cancers of the WBCs.

WBCs are a vital part of your immune system. They protect your body from invasion by bacteria, viruses, and fungi, as well as from abnormal cells and other foreign substances. In leukemia, the WBCs don't function like normal WBCs. They can also divide too quickly and eventually crowd out normal cells.

WBCs are mostly produced in the bone marrow, but certain types of WBCs are also made in the lymph nodes, spleen, and thymus gland. Once formed, WBCs circulate throughout your body in your blood and lymph (fluid that circulates through the lymphatic system), concentrating in the lymph nodes

and spleen.

Risk factors for leukemia

The causes of leukemia aren't known. However, several factors have been identified which may increase your risk. These include:

- a family history of leukemia
- smoking, which increases your risk of developing acute myeloid leukemia (AML)
- genetic disorders such as Down syndrome
- blood disorders, such as myelodysplastic syndrome, which is sometimes called "preleukemia"
- previous treatment for cancer with chemotherapy or radiation
- exposure to high levels of radiation
- exposure to chemicals such as benzene