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. There are two major types of immunodeficiency disorders: PRIMARY AND SECONDARY

1. Identify and briefly explain 5 primary immunodeficiency disorders

1.Common Variable Immunodeficiency (CVID)

Common variable immunodeficiency (CVID) is a primary immune deficiency disease characterized by low levels of protective antibodies and an increased risk of infections. Although the disease usually is diagnosed in adults, it also can occur in children. CVID also is known as hypogammaglobulinemia, adult-onset agammaglobulinemia, late-onset hypogammaglobulinemia, and acquired agammaglobulinemia.

NIAID supports research to determine genetic causes of CVID that may lead to therapeutic approaches to address the disease. Researchers also are exploring how antibody-based drugs may lessen the severity of the condition.

2.Congenital Neutropenia Syndromes:Congenital neutropenia syndromes are a group of rare disorders present from birth that are characterized by low levels of neutrophils, a type of white blood cell necessary for fighting infections. NIAID supports basic scientific research on the nature and development of neutrophils, which may lead to insights for addressing congenital neutropenia syndromes.

Congenital neutropenia syndromes also may be referred to as congenital agranulocytosis, severe congenital neutropenia, severe infantile genetic neutropenia, infantile genetic agranulocytosis, or Kostmann disease.

Researchers have identified numerous genetic mutations that cause congenital neutropenia syndromes. Generally, mutations that result in congenital neutropenia affect the development, lifespan or function of neutrophils. Congenital neutropenia syndromes are inherited through autosomal recessive, autosomal dominant and X-linked inheritance patterns. The genes linked to these syndromes include the following:

ELANE

HAX1

G6PC3

GFI1

CSF3R

X-linked WAS

CXCR4

3.Glycosylation Disorders with Immunodeficiency

Glycosylation refers to the attachment of sugars to proteins, a normal process required for the healthy function of cells. Glycosylation can impact how cells communicate, respond to their environment, grow and function. Because glycosylation regulates a wide range of activities in cells throughout the body, defects in glycosylation can cause extensive and severe symptoms. In some cases, these impairments disrupt the immune system, resulting in immunodeficiency. In some cases, these impairments disrupt the immune system, resulting in immunodeficiency.

NIAID scientists research genetic causes for glycosylation disorders with immunodeficiency, a set of rare diseases, and investigate the role that glycosylation plays in bacterial and viral infections. This research benefits people affected by these disorders and increases scientific knowledge of pathogens.

Researchers have not yet identified the cause of many forms of glycosylation disorders with immunodeficiency. However, in 2014, NIH scientists discovered a disorder caused by mutations in the PGM3 gene, resulting in defective sugar metabolism that leads to problems with glycosylation, especially in immune cells.

People with PGM3 gene mutations may experience the following symptoms:

Susceptibility to bacterial and viral infections

Allergies, including food allergy

Asthma

Eczema

Autoimmunity

Developmental delays, including motor and cognitive impairments

4:Warts, Hypogammaglobulinemia, Infections, and Myelokathexis (WHIM) Syndrome

Warts, Hypogammaglobulinemia, Infections, and Myelokathexis (WHIM) syndrome is a rare genetic disease of the immune system. Its name is an acronym for its main clinical manifestations: warts, hypogammaglobulinemia, infections, and myelokathexis. Hypogammaglobulinemia is a deficiency in specific infection-fighting antibodies in the blood. Myelokathexis refers to the failure of neutrophils — infection-fighting white blood cells — to move from the bone marrow into the bloodstream where they can patrol the body. WHIM syndrome patients also have trouble distributing most other types of immune cells to the blood. Such defects in the immune system predispose WHIM syndrome patients to frequent bacterial and viral infections, persistent skin and genital warts, and an increased risk of developing cancer caused by human papillomavirus.

5.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.

2. Identify and briefly explain 2 secondary immunodeficiency disoders

1. HIV:HIV stands for human immunodeficiency virus. It is the virus that can lead to acquired immunodeficiency syndrome or AIDS if not treated. Unlike some other viruses, the human body can’t get rid of HIV completely, even with treatment. So once you get HIV, you have it for life.

HIV attacks the body’s immune system, specifically the CD4 cells (T cells), which help the immune system fight off infections. Untreated, HIV reduces the number of CD4 cells (T cells) in the body, making the person more likely to get other infections or infection-related cancers. Over time, HIV can destroy so many of these cells that the body can’t fight off infections and disease. These opportunistic infections or cancers take advantage of a very weak immune system and signal that the person has AIDS, the last stage of HIV infection.

No effective cure currently exists, but with proper medical care, HIV can be controlled. The medicine used to treat HIV is called antiretroviral therapy or ART. If people with HIV take ART as prescribed, their viral load (amount of HIV in their blood) can become undetectable. If it stays undetectable, they can live long, healthy lives and have effectively no risk of transmitting HIV to an HIV-negative partner through sex. Before the introduction of ART in the mid-1990s, people with HIV could progress to AIDS in just a few years. Today, someone diagnosed with HIV and treated before the disease is far advanced can live nearly as long as someone who does not have HIV.

2.Malnutrition – Protein-calorie malnutrition is the biggest global cause of SIDs which can affect up to 50% of the population in some communities in the developing world.vii T cell numbers and function decrease in proportion to levels of protein deficiency, which leaves the patient particularly susceptible to diarrhoea and respiratory tract infections. This form of immunodeficiency will usually resolve if the malnutrition is treated.

3.Drug regimens – There are several types of medication that can result in secondary immunodeficiencies, but these drugs also perform critical roles in certain areas of healthcare. Immunosuppression is a common side-effect of most chemotherapies used in cancer treatment. The immune system usually recovers once the chemotherapy treatment has finished. Another common use forimmunosuppressive drugs is the prevention of transplant rejection, where medication is required to suppress the transplant recipient’s immune system and prevent it from targeting the transplanted tissue. These drugs can have significant side-effects and often suppress more areas of the immune system than are required, leading to susceptibility to opportunistic infections. Use of a new generation of medicines called biologics are becoming more widespread in treating transplant rejection. These drugs are derived from biological sources like cells, rather than chemical structures. Monoclonal antibodies are one such class of biologics and these drugs are made by farming antibodies from B cells that will act against a specific part of the disease process. These agents are more specific in their action than traditional drugs and have fewer side effects on non-target immune cells.

4. cancers of the immune system, like 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

5. Multiple myeloma

Multiple myeloma is a type of cancer that affects plasma cells. Plasma cells are a type of white blood cell found in bone marrow, which is the soft tissue inside most of your bones that produces blood cells. In the bone marrow, plasma cells make antibodies. These are proteins that help your body fight off diseases and infections.

Multiple myeloma occurs when an abnormal plasma cell develops in the bone marrow and reproduces itself very quickly. The rapid reproduction of malignant, or cancerous, myeloma cells eventually outweighs the production of healthy cells in the bone marrow. As a result, the cancerous cells begin to accumulate in the bone marrow, crowding out the healthy white blood cells and red blood cells.