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QUESTIONS

1. Identify and briefly explain 5 primary immunodeficiency disorders.
2. Identify and briefly explain 2 secondary immunodeficiency disorders.

ANSWER

Primary immunodeficiency disorder

- SCID
- Omenn syndrome
- Ataxia telangiectasia
- Di George syndrome
- Wiskott Aldrich syndrome
- Agamnaglobuninemia
- CVID
- Selective IgA deficiency
- Hyper IgM syndrome
- Transient hypergammaglobuinemia of infancy
- Chediak syndrome
- Familia Hemophagocytic
- Lymophoproliferative syndrome
- Fas deficiency

SCID

SCID is a syndrome that is caused by mutations in any of several Gene's whose product are crucial for development and function of both T and B lymphocytic cells and also affect the natural killer cells. The molecular defect happens only T cell deficiency, and B cells are intrinsically normal.

SCID is a primary immunodeficiency disorder which weaken the immune system, however, it leads the body's less active or inability to fight minor disease.

It affect babies of age 3 month, due to genetic hereditary from parent. It is caused as a result of the X chromosome, females are natural carries of these mutations as as result of 2 X - chromosome. It can be treated as a result of stem cell from bone marrow and it is given through IV. It affect sex and ethnicity groups.

2. OMENN SYNDROME

Omenn syndrome is an autosomal recessive form of severe combined immunodeficiency (SCID) characterized by erythroderma (skin redness), desquamation (peeling skin), alopecia (hair loss), chronic diarrhea, failure to thrive, lymphadenopathy (enlarged lymph nodes), eosinophilia, hepatosplenomegaly, and elevated serum IgE levels. People are highly susceptible to infection and develop fungal, bacterial, and viral infections typical of SCID. In this

syndrome, the SCID is associated with low IgG, IgA, and IgM and the virtual absence of B cells. There is an elevated number of T cells, but their function is impaired. It has been found to be caused by mutations in the RAG1 or RAG2 genes. It is treated with the stem cells transplant and bone marrow.

3. ATAXIA TELANGIECTASIA

Ataxia-telangiectasia is a rare inherited disorder that affects the nervous system, immune system, and other body systems. This disorder is characterized by progressive difficulty with coordinating movements (ataxia) beginning in early childhood, usually before age 5. Affected children typically develop difficulty walking, problems with balance and hand coordination, involuntary jerking movements (chorea), muscle twitches (myoclonus), and disturbances in nerve function (neuropathy). The movement problems typically cause people to require wheelchair assistance by adolescence. People with this disorder also have slurred speech and trouble moving their eyes to look side-to-side (oculomotor apraxia). Small clusters of enlarged blood vessels called telangiectases, which occur in th eyes and on the surface of the skin, are also characteristic of this condition.

4. CVID

Common variable immunodeficiency (CVID) is a group of disorders characterized by low levels of a type of protein known as immunoglobulins (Ig). Because of low level of Ig, the immune system cannot make antibodies that fight bacteria, viruses or other toxins in the body. leads to frequent infections, particularly in the sinuses, lungs, and digestive tract. Symptoms most commonly begin in early adulthood but can occur at any age. While in most cases the cause of CVID is unknown, a genetic change has been found in about one-third of cases. condition is diagnosed based on the symptoms, specific laboratory testings, and exclusion of other disorders. Treatment for CVID includes Ig replacement therapy, which stops the cycle of recurrent infections. The long term outlook for people with CVID varies depending on the severity of the symptoms and any underlying conditions.

5. Chediak syndrome

Chediak-Higashi syndrome is a condition that affects many parts of the body, particularly the immune system. This disease damages immune system cells, leaving them less able to fight off invaders such as viruses and bacteria. As a result, most people with Chediak-Higashi syndrome have repeated and persistent infections starting in infancy or early childhood. These infections tend to be very serious or life-threatening.

Chediak-Higashi syndrome is also characterized by a condition called oculocutaneous albinism, which causes abnormally light coloring (pigmentation) of the skin, hair, and eyes. Affected individuals typically have fair skin and light-colored hair, often with a metallic sheen. Oculocutaneous albinism also causes vision problems such as reduced sharpness; rapid, involuntary eye movements (nystagmus); and increased sensitivity to light (photophobia).

Many people with Chediak-Higashi syndrome have problems with blood clotting (coagulation) that lead to easy bruising and abnormal bleeding. In adulthood, Chediak-Higashi syndrome can also affect the nervous system, causing weakness, clumsiness, difficulty with walking, and seizures.

2. **Secondary immunodeficiency disorder**

- HIV and AIDS

- Severe burns
- Immuno complex disease(viral hepatitis)

VIRAL HEPATITIS

Hepatitis refers to inflammation of the liver. Inflammation is a tissue's reaction to irritation or injury which generally results in swelling and can cause pain.

There are many causes of hepatitis. Viral hepatitis is caused by a virus and can either be acute (lasting less than six months) or chronic (lasting more than six months). Viral hepatitis can be spread from person to person. Some types of viral hepatitis can be spread through sexual contact.

There are five known hepatitis viruses which are categorized by the letters A through E.

LEUKEMIA

Leukemia are blood cancer cause mainly by white blood cells. White blood helps in that fight against infection in the body.

Leukemia occur as a result of rise in white blood cells over the thr red blood and the platelets.