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**17/MHS02/069**

**300LEVEL**

**NURSING DEPARTMENT**

**NSC 306 ASSIGNMENT**

PRIMARY IMMUNODEFICIENCY DISORDERS.

1. **Autoimmune Lymphoproliferative Syndrome (ALPS).**

Autoimmune lymphoproliferative syndrome (ALPS) is a rare immune disorder first described by NIH scientists in the mid-1990s that can cause numerous autoimmune problems, such as low levels of red blood cells, clot-forming platelets, and infection-fighting white blood cells. These problems can increase the risk of infection and hemorrhage. Most cases of ALPS are caused by mutations in the FAS gene. FAS produces a receptor that, when activated, leads to programmed cell death, or apoptosis. This programmed death is an important part of the normal cell lifecycle. When cells do not receive the message that it is time for them to die, an abnormal buildup of cells can result.

1. **Caspase eight deficiency state (CEDS).**

This is a very rare genetic disorder of the immune system caused by mutations in the CASP8 gene. CEDS is characterized by an enlarged spleen and lymph nodes, recurrent sinus and lung infections, recurrent viral infections, and a low level of infection-fighting antibodies. CEDS is caused by mutations in the CASP8 gene, which provides instructions for production of the protein caspase eight, which is also abbreviated as CASP8. The CASP8 protein is involved in programmed cell death, or apoptosis. The body must maintain a careful balance between proliferation of immune cells and apoptosis to defend against pathogens and avoid autoimmunity. The mutations that cause CEDS destabilize the CASP8 protein and block its function, leading to buildup of immune cells.

1. **Chronic Granulomatous Disease (CGD).**

Occurs when white blood cells called phagocytes are unable to kill certain bacteria and fungi, making people highly susceptible to some bacterial and fungal infections. CGD is caused by defects in an enzyme, NADPH oxidase, that phagocytes need to kill certain bacteria and fungi. Mutations in one of five different genes can cause these defects.

1. **Congenital Neutropenia Syndromes:**

Congenital neutropenia syndromes are a group of disorders present from birth that are characterized by low levels of neutrophils, a type of white blood cell necessary for fighting infections.

Congenital neutropenia syndromes also may be referred to as congenital agranulocytosis, severe congenital neutropenia, severe infantile genetic neutropenia, infantile genetic agranulocytosis, or Kostmann disease. People with congenital neutropenia experience bacterial infections early in life. These may cause inflammation of the umbilical cord stump, abscesses (or boils) on the skin, oral infections and pneumonia. Congenital neutropenia also increases one’s risk for developing myelodysplastic syndromes (MDS), blood disorders that are distinguished by low levels of various blood cells, this can lead to a type of blood cancer called acute myeloid leukemia.

1. **CTLA4 Deficiency.**

CTLA4 deficiency is a rare disorder that severely impairs the normal regulation of the immune system, resulting in conditions such as intestinal disease, respiratory infections, autoimmune problems, and enlarged lymph nodes, liver, and spleen. CTLA4 deficiency is caused by mutations in a gene called CTLA4, which gives cells instructions for making the CTLA4 protein. This protein functions as a brake to slow down and control the action of the immune system. People with CTLA4 deficiency often experience autoimmune problems that can affect various organs and tissues, including the blood, thyroid, skin, and joints.

SECONDARY IMMUNODEFICIENCY DISORDERS.

1. **AIDS(Acquired Immunodeficiency Syndrome):** This is the most advanced stage of 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 has no cure. However, with medical care, including treatment called antiretroviral therapy, it’s possible to manage HIV and live with the virus for many years. HIV is transmitted through bodily fluids that include: blood, semen, vaginal and rectal fluids, breast milk. If AIDS does develop, it means that the immune system is severely compromised. It’s weakened to the point where it can no longer fight off most diseases and infections. The life expectancy of someone with AIDS is 3 years.
2. **LEUKEMIA:** This is a cancer of the white blood cells(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 do not function like normal WBCs. They can also divide too quickly and eventually crowd out normal cells. The types of leukemia are; (I) The onset of leukemia can be acute (sudden onset) or chronic (slow onset). In acute leukemia, cancer cells multiply quickly. In chronic leukemia, the disease progresses slowly and early symptoms may be very mild.

(II) Leukemia is also classified according to the type of cell. Leukemia involving myeloid cells is called myelogenous leukemia. Leukemia involving lymphocytes is called lymphocytic leukemia. There are four main types of leukemia:

1. Acute myelogenous leukemia (AML).
2. Chronic myelogenous leukemia (CML).
3. Acute lymphocytic leukemia (ALL).
4. Chronic lymphocytic leukemia (CLL).

Symptoms of leukemia are;

1. Excessive sweating, especially at night.
2. Fatigue and weakness.
3. Weight loss.
4. Bone pain and tenderness.
5. painless, swollen lymph nodes (especially in the neck and armpits).
6. Enlargement of the liver or spleen.
7. Red spots on the skin, called petechiae.
8. Bleeding easily and bruising easily.
9. Fever and chills.
10. Frequent infections.