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QUESTION:

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

ANSWER:

1. Primary immunodeficiencies are disorders which part of the body’s immune system is missing or does not function properly. They are mostly genetic disorders. They include: -Agammaglobulinemia

-Wiscott-Aldrich syndrome

-Digeorge syndrome

-Ataxia- telengectasia

-Chronic granulomatous disease

-Complement deficiencies

* Agammaglobulinemia: is a group of inherited immune deficiencies characterized by a low concentration of antibodies in the blood due to the lack of particular lymphocytes in the blood and lymph. It affects 50% of male if mothers are carriers of the gene. Children are generally asymptomatic until 6-9 months of age when maternal IGb decreases, others include recurrent infections with streptococcus pneumonia, haemophilus influenze, mycoplasma pneumonia, hepatitis virus and enterovirus CNS infection.
* Wiscott-Aldrich syndrome: this is a rare X-linked recessive disease characterized by eczema, thrombocytopenia, immune deficiency and blood diarrhea.
* Chronic granulomatus disease: is a diverse group of hereditary disease in which certain cells of the immune system have difficulty forming the reactive oxygen compound used to kill certain ingested pathogens. This leads to the formation of granulomata in many organs. It is often characterized by pneumonia, abscesses, septic arthritis, oestomyelitis and other infections.
* Ataxia-telangiecta: this is a rare neurodegenerative, autosomal recessive disease causing severe disability. Ataxia refers to poor coordination and telangiectasia refres to small dilated blood vessels, both of which are hallmarks of the disease. It affects many parts of the body:
* It impairs certain part of the brain including the cerebellum, causing difficulty with movement and coordination.
* It weakens the immune system, causing a predisposition to infection.
* It prevents repair of broken DNA, increasing the risk of cancer.
* Digeorge syndrome: this is a chromosomal disorder caused by deletion of a small segment of chromosome 22. It results in poor development of severe bodily systems.

1. Secondary immunodeficiency disease: this are disease that occurs when the immune system is compromised due to environmental factor. These diseases include: -HIV/AIDS

-Cancers of the immune system (leukemia)

-Immune complex disease like viral hepatitis

-Multiple myeloma

* HIV/AIDS: HIV is a virus that damages the immune system. The immune system helps the body fight off infections. Untreated HIV infects and kills more CD4 cells, which are a type of immune cells called T cells. Over time the body is likely to get various types of infections and cancers. It is transmitted through bodily fluids ( blood, semen, vaginal and rectal fluids, breast milk). HIV is a lifelong condition and currently there is no cure, 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.
* Cancer of the immune system (leukemia): this is a group of blood cancers that usually begin in the bone marrow and result in high numbers of abnormal blood cells. These blood cells are not fully developed and are called blast or leukemia cells. Symptoms include bleeding and bruising, feeling tired, fever, and an increased risk of infections. They occur due to lack of normal blood cells.