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QUESTIONS: 1. Identify and briefly explain 5 primary immunodeficiency disorders

 2. Identify and briefly explain 2 secondary immunodeficiency disorders.

 **ANSWERS:**

 Immunodeficiency disorders involve malfunction of the immune system, resulting in infections that develop and recur more frequently, are more severe and last longer than usual. Immunodeficiency disorders usually result from use of a drug or from a long-lasting serious disorder (such as cancer) but occasionally are inherited.

There are two types of immunodeficiency disorders:

* **Primary immunodeficiency disorders:** These disorders are usually present at birth and are genetic disorders that are usually hereditary. They typically become evident during infancy or childhood. However, some primary immunodeficiency disorders (such as **common variable immunodeficiency)** are not recognized until adulthood.
* **Secondary immunodeficiency disorders:** These disorders generally develop later in life and often result from use of certain drugs or from another disorder, such as **diabetes or human immunodeficiency virus (HIV) infection.** They are more common than primary immunodeficiency disorders.
1. **PRIMARY IMMUNODEFICIENCY DISORDERS**
* Ataxia-telangiectasia (AT)
* Wiscott-Aldrich syndrome ( WAS)
* Agammaglobulinemia
* Chronic granulomatous disease
* Severe combined immunodeficiency disease ( SCID)
1. **ATAXIA-TELEGIECTASIA :**  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 in 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 eyes to look side-to-side (oculomotor apraxia). Small clusters of enlarged blood vessels called **telangiectases,** which occur in the eyes and on the surface of the skin, are also characteristic of this condition.

 Affected individuals tend to have high amounts of a protein called **alpha-fetoprotein (AFP)** in their blood. The level of this protein is normally increased in the bloodstream of pregnant women, but it is unknown why individuals with ataxia-telangiectasia have elevated AFP or what effects it has in these individuals. People with ataxia-telangiectasia often have a weakened immune system, and many develop chronic lung infections. They also have an increased risk of developing cancer, particularly cancer of blood-forming cells (leukemia) and cancer of immune system cells (lymphoma). Affected individuals are very sensitive to the effects of radiation exposure, including medical x-rays. The life expectancy of people with ataxia-telangiectasia varies greatly, but affected individuals typically live into early adulthood.

***CAUSES OF ATAXIA-TELANGIECTASIA:*** Mutation in the ATM gene cause ataxia-telangiectasia. The ATM gene provides instructions for making protein that helps control cell division and is involved in DNA repair. This protein plays an important role in the normal development and activity of several body systems, including the nervous system and immune system.

Mutations in the ATM gene reduce or eliminate the function of the ATM protein. Without this protein, cells become unstable and die. Cells in the part of the brain involved in coordinating movements (the cerebellum) are particularly affected by loss of the ATM protein. The loss of these brain cells causes some of the movement problems characteristic of ataxia-telangiectasia.

1. **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. Antibodies are proteins (immunoglobulins, (IgM), (IgG) etc) that are critical and key components of the immune system. It is caused by the gene defect that blocks the growth of normal, mature immune cells called B lymphocytes. As a result, the body makes very little immunoglobulins which play a major role in the immune response, which protects against illness and infection. There two types of agammaglobulinemia ,which includes; **X­-linked agammaglobulinemia (XLA), and autosomal recessive agammaglobulinemia (ARAG).**
2. **WISKOTT-ALDRICH SYNDROME (WAS):** Is a rare X-linked recessive disease characterized by eczema, thrombocytopenia, immune deficiency, and bloody diarrhea (secondary to the thrombocytopenia). It is also sometimes called the **eczema-thrombocytopenia-immunodeficiency syndrome.** Symptoms may include; Frequent and easy bleeding that can occur from the nose, from the mouth and gums, in bowel movements, eczema and chronic infections.
3. **CHRONIC GRANULOMATOUS DISEASE:** CGD is an inherited primary immunodeficiency disease which increases the body’s susceptibility to infections caused by certain bacteria and fungi. Granulomas are masses of immune cells that form at sites of infection or inflammation. It occurs when a type of white blood cell (phagocyte) that usually helps your body fight infections doesn’t work properly. As a result, the phagocytes can’t protect your body from bacterial infections. Symptoms include: **swollen lymph nodes, chronic runny nose, persistent diarrhea, abscesses that involve the (lungs, liver, spleen, bones or skin).**
4. **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, useless infants receive immune-restoring treatments such as transplants of blood-forming stem cells, gene therapy, or enzyme therapy. SCID is caused by genetic defect that affects the function of T cells. Depending in the type of SCID, B cells and NK cells can also be affected. These cells play important roles in helping the immune system battle bacteria, viruses, fungi that cause infections. Common signs and symptoms include an increased susceptibility to infections including ear infections, pneumonia or bronchitis, diarrhea, children with SCID do not grow and gain weight as expected (failure to thrive).
5. **SECONDARY (ACQUIRED) IMMUNODEFICIENCY DISORDERS:**
* HIV infection
* Malnutrition
1. **MALNUTRITION:** Malnutrition is a condition that results from nutrient deficiency or overconsumption. It includes undernutrition, overnutrition, both which can lead to health problems and nutrient deficiencies if not addressed.
* **UNDERNUTRITION:** This type of malnutrition results from not getting enough protein, calories or micronutrients. It leads to low weight-for-height (wasting), height-for-age (stunting) and weight-for-age (underweight).
* **OVERNUTRITION:** Overconsumption of certain nutrients, such as protein, calories or fat, can also lead to malnutrition. This usually results in overweight or obesity.

Common causes of malnutrition include:

* Excessive alcohol consumption
* Mental health disorders
* Digestive problems and issues with nutrient absorption
* Food insecurity or lack of access to sufficient and affordable food

Symptoms and signs of malnutrition depend on the type.

1. **HIV INFECTION:** 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 such that include: **blood, semen, vaginal and rectal fluids, breast milk.** The virus doesn’t spread in air or water, or through casual contact. 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.

 **Stages of HIV Infection**

1. **Stage 1: Infection** – HIV quickly replicates in the body after infection. Some people develop short lived flu-like symptoms for each headaches, fever, sore throat and rash within days to weeks after infection. During this time the immune system react to the virus by developing antibodies.
2. **Stage 2**: **Asymptomatic** – This stage of HIV infection does not cause outward signs or symptoms. A person may look and feel well but HIV is continuing to weaken their immune system. This stage may last several years (an average of 8 to 10 years) and without a HIV test many people do not know they are infected.
3. **Stage 3**: **Symptomatic** – Over time the immune system becomes damaged and weakened by HIV and symptoms develop. Initially they can be mild but they do worsen, symptoms include fatigue, weight loss, mouth ulcers, thrush and severe diarrhea. The symptoms are caused by the emergence of opportunistic infections because they take advantage of a person weakened immune system.
4. **Stage 4**: **Progression of HIV to AIDS –** There is no single test for AIDs; doctors will look at variety of symptoms including the CD4 count, the viral load and the presence of opportunistic infections in order to make an AIDS diagnosis.

**NOTE**: Most people get the virus by having unprotected sex with someone who has HIV. Another common way of getting it is by sharing drug needles with someone who is infected with HIV.