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**DEPARTMENT: NURSING**

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**QUESTION**

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

ANSWERS

Immunodeficiency disorders are the absence or failure of normal function of one or more elements of immune system. it disrupt one’s body ability to defend itself against bacteria, viruses, and parasites.

There are two types of immunodeficiency disorders: those you are born with (primary), and those that are acquired (secondary).

Anything that weakens our immune system can lead to a secondary immunodeficiency disorder.

Immunodeficiency disorders prevent your body from fighting infections and diseases. This type of disorder makes it easier for someone to catch viruses and bacterial infections.

Immunodeficiency disorders are either congenital or acquired. A congenital, or primary, disorder is one someone was born with. Acquired, or secondary, disorders one get later in life. Acquired disorders are more common than congenital disorders.

**Examples of primary immunodeficiency disorders include:**

* **Common variable immunodeficiency (CVID)**
* **X-linked agammaglobulinemia(XLA)**
* **Severe combined immunodeficiency(SCID0**
* **Chronic Granulomatous Disease (CGD)**
* **Congenital Neutropenia Syndromes**
1. Common Variable Immunodeficiency (CVID)

CVID is caused by a variety of different genetic abnormalities that result in a defect in the capability of immune cells to produce normal amounts of protective antibodies. People with CVID experience frequent bacterial and viral infections of the upper airway, sinuses, and lungs.

1. X-linked agammaglobulinemia (XLA)

XLA is caused by an inability to produce B cells or immunoglobulins (antibodies), which are made by B cells. People with XLA develop frequent infections of the ears, throat, lungs, and sinuses.

1. Severe combined immunodeficiency(SCID)

SCID is a group of rare, life-threatening disorders caused by mutations in different genes involved in the development and function of infection-fighting T and B cells. Infants with SCID appear healthy at birth but are highly susceptible to severe infections.

1. Chronic Granulomatous Disease (CGD)

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. Mutations in one of five different genes can cause this disease.

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.

**Examples of secondary immunodeficiency disorders include:**

A secondary immunodeficiency occurs as a result an acquired impairment of function of B cells, T cells, or both. Secondary immunodeficiencies can be caused by:

Systemic disorders such as **diabetes mellitus, malnutrition, hepatitis, or HIV infection**

* HIV infection and the associated acquired immunodeficiency syndrome (AIDS) are the best-known secondary immunodeficiencies. AIDS is characterized by profound CD4 T-cell lymphopenia (decrease in lymphocytes). The decrease in CD4 T cells is the result of various mechanisms, including HIV-induced pyroptosis (a type of apoptosis that stimulates an inflammatory response), viral cytopathic effect, and cytotoxicity to HIV-infected cells.
* The most common cause of secondary immunodeficiency worldwide is severe malnutrition, which affects both innate and adaptive immunity .Severe protein malnutrition in newborns and infants is clearly associated with atrophy in the so called primary lymphoid organs, i.e., bone marrow and thymus. Consequences are devastating because these organs are generators of B and T cell repertoires. Furthermore, malnutrition clearly affects hematopoiesis, determining anemia, leucopenia and severe reduction in bone marrow. Production of IL-6 and TNF-α by bone marrow cells is also significantly lower in malnourished animals . The capacity of malnourished hematopoietic stroma to support the growth of hematopoietic stem cells (CD34+) in vitro is also decreased . This is a very relevant finding because CD34+ cells are able to generate multiple lymphohematopoietic lineages as myeloid, erythroid and lymphoid (B and T).

 Severe protein malnutrition, mainly in newborns and small children, also provokes thymus atrophy that, in turn, reduces thymus cell number and also severely affects the development of peripheral lymphoid organs . The immediate consequence of this atrophy is leucopenia, decreased CD4/CD8 ratio and increased number of immature T cells in the periphery.