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**17/MHS02/ 026**

**Nursing science**

**Medsurg Assignment**

Immunodeficiency disorder is the absence or failure of normal function of one or more elements of the immune system. There are two major types of immunodeficiency disorders: PRIMARY AND SECONDARY.

1. Identify and briefly explain 5 primary immunodeficiency disorders

2. Identify and briefly explain 2 secondary immunodeficiency disorders

Primary Immunodeficiency disorders refer to a heterogeneous group of disorders characterized by a poor or absent function in one or more components of the immune system. They include

1. **Wiscott-Aldrich syndrome:** This is a rare X-linked recessive disease characterized by eczema, thrombocytopenia, immune deficiency and bloody diarrhea

The estimated incidence is between 1-10 cases per million males worldwide and the condition is rarer in females.

**Signs and symptoms** includes frequent and easy bleeding that occurs from the nose, from the mouth and gums, in bowel movement, frequent and easy bruising, Petechiae, chronic infections, eczema.

**Treatment:** Antimicrobial therapy; splenectomy with continuous antibiotic prophylaxis; IV immunoglobulin and bone marrow transplantation

1. **Hyperimmunoglobulinemia E (HIE) syndrome:** also known as the Job syndrome. It is a rare, inherited disease.

**Major symptoms includes:** Bacterial, fungal and viral infections; deep-seated cold abscesses

**Causes:** recent research shows that the disease is often caused by the mutation in the stat3 gene on chromosome 17. How the gene abnormality causes symptoms is not well understood. However, people with the disease have a higher than normal level of antibody IgE.

**Treatment:** Antibiotic therapy and treatment for viral and fungal infections.

1. **Chronic mucocutaneous candidiasis:** it is an immune disorder of the T cells, it is characterized by chronic infections with candida that are limited to mucosal surfaces, skin and nails.

**Risk factors:** usually associated with certain endocrine conditions: hypoadrenalism, diabetes mellitus, hypothyroidism, hypoparathyrodism.

**Major symptoms:** Candida albicans infections of mucous membrane, skin, and nails; endocrine abnormalities (hypoparathyroidism, Addison’s disease)

**Treatment:** Antifungal agents: Topical: miconazole, Oral: clotrimazole, ketoconazole, IV: amphotericin B.

1. **Thymic hypoplasia (DiGeorge syndrome):** this is a condition where the thymus is underdeveloped or involuted. It is also known as deletion syndrome which is typically due to the deletion of 30-40 genes in the middle of chromosomes 22 at a location known as 22q11.2.

**Symptoms** usually varies widely even among members of the same family and affect many part of the body. These common symptoms include:

Recurrent infections, hypoparathyroidism, hypocalcemia, tetany, convulsions, congenital heart disease, possible renal abnormalities, abnormal faces, cleft palate, cyanosis, learning difficulties, hearing disorders, psychiatric disorders.

There is no cure for this syndrome but the symptoms are treated accordingly in order to manage it.

1. **Common variable immunodeficiency (CVID):** this is an antibody deficiency that leaves the immune system unable to defend against bacteria and viruses, resulting in recurrent and often severe infections primarily affecting the ears, sinuses, and respiratory tract.

Causes: the exact cause and genetic inheritance pattern of CVID is unknown but genetic mutations that leads to CVID have been identified.

Both male and females are affected and the severity of the symptoms from one person to another.

Symptoms: Hypogammaglobulinemia, recurrent infection, Pernicious anemia, chronic respiratory infections.

Treatment: IV immunoglobulin, Metronidazole (Flagyl), Quinacrine HCl (Atabrine), Vitamin B12, Antimicrobial therapy.

**Secondary immunodeficiency disorders (SID)**

 These are disorders that occur when the immune system is weakened by another treatment or illness. Common causes of secondary immunodeficiencies include malnutrition, chronic stress, burns, uremia, diabetes mellitus, certain autoimmune disorders, certain viruses, exposure to immunotoxic medications and chemicals, and self-administration of recreational drugs and alcohol.

1. **AIDS:** this results from infection with the human immunodeficiency virus 1. It is the advanced stage of HIV and untreated the life expectancy of AIDS is 3 years.

Common methods of transmitting the virus includes; unprotected sex with an infected partner, sharing needles with infected person, transmission from mother to child.

**Signs and symptoms:** weight loss, fever or night sweats, fatigue and recurrent infections, mouth ulcers, oral thrush, pneumonia, red bloches, swollen lymph nodes, tuberculosis, candidiasis, kidney disease.

**Treatments:** No cure exists for AIDS but antiretroviral regimens can dramatically slow disease progress as well as prevents secondary infections and complications. It also reduces the risk of infecting others.

1. **Multiple myeloma:** is a cancer of plasma cells, a type of white blood cells that normally produces antibodies.

**Symptoms:** bone pain, bleeding, frequent infections, anemia, weight loss, renal failure.

Causes are usually unknown

**Risk factors:** obesity, radiation exposure, family history, and certain chemicals, older age,

**Diagnostic method:** blood or urine test finding abnormal antibodies, bone marrow biopsy finding cancerous plasma cells and medical imaging finding bone lesions. Another common finding is high blood calcium level.

**Treatment:** steroids, chemotherapy, thalidomide, stem-cell transplant, bisphosphonates, radiation therapy.