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Matric No:17/MHS02/056

Course code: NSC306

Course title :Medical Surgical Nursing II

A .PRIMARY IMMUNODEFICIENCY DISORDERS;

They are also disorders in which part of the body’s immune system is missing or does not function normally. Most primary disorders are genetic disorders. They generally are considered uncommon. Primary immunodeficiencies include a variety of disorders that render patients more susceptible to infections. If left untreated, these infections may be fatal.

• Severe Combined Immunodeficiency (SCID); is a rare genetic disorder characterized by the disturbed development of functional T-cells and B-cells caused by numerous genetic mutations that result in differing clinical presentations. SCID are the most severe form of primary immunodeficiency. It is also known as Bubble Boy Disease and Bubble Baby Disease because its victims are extremely vulnerable to infectious diseases and patients are made to live in a sterile environment. SCID is as a result of highly compressed immune system to the point of complete absence. SCID has different types such as; X-linked severe combined immunodeficiency, adenosine deaminase deficiency, purine nucleoside phosphorylase deficiency etc. It is treated through bone marrow transplantation and prophylaxis against infection and Gene therapy.

• DiGeorge Syndrome; also known as Deletion Syndrome, is a syndrome caused by the deletion of a small segment of chromosome 22. While the symptoms can vary, they often include congenital heart problems, specific facial features, frequent infections, developmental delay, learning problems and cleft palate. Associated conditions include kidney problems, hearing loss and autoimmune disorders such as Rheumatoid arthritis or Graves’ disease. It is caused by genetic mutation. There is no cure for this disorder but treatments can improve symptoms which often includes Multidisciplinary approach with efforts to improve the function of the potentially many organ systems involved.

• Wiskott-Aldrich Syndrome (WAS): is a rare X-linked recessive disease characterized by eczema, thrombocytopenia (low platelet count), immune deficiency, and bloody diarrhea (secondary to thrombocytopenia). It is also called Eczema-Thrombocytopenia- Immunodeficiency. It is caused by; mutation of the same genes. WAS occurs most often in males due to its X-linked recessive pattern of inheritance. Signs and symptoms include; petechiae, bruising; resulting from low platelet count, spontaneous nose bleeds, bloody diarrhea and eczema. Recurrent bacterial infections develop by 3 months. Majority of children with WAS develop at least one immune disorders or cancer (mainly lymphoma and leukemia).

• Transient Hypogammaglobulinemia of Infancy; is a form of hypogammaglobulinemia appearing after birth, leading to a reduction in the level of IgG, and sometimes IgA and IgM. It can result in increased infections, but it can also present without symptoms. It has no treatment.

• Chronic Granulomatous Disease (CGD); it is also called Bridges-Good Syndrome, Chronic Granulomatous Disorder, and Quie Syndrome. Is a diverse group of hereditary diseases in which certain cells of the immune system have difficulty forming the reactive oxygen compounds (most importantly the Superoxide radical due to defective phagocyte NADPH oxidase) used to kill certain ingested pathogens. This leads to the formation of Granulomata in many organs. Symptoms of CGD include; Pneumonia, abscesses of the skin, tissues and organs, septic arthritis, osteomyelitis, bacteremia/fungemia, and superficial skin infections such as cellulitis or impetigo. Management of CGD revolves around 2 goals; (1). Diagnose the disease early and prescribe antibiotic prophylaxis, immunomodulation (Interferon, in form of interferon gamma-1b (acti-immune)), and Hematopoietic stem cell transplantation (HSCT). (2). Educate the patient about his or her condition so that prompt treatment can be given if an infection occurs.

B.

SECONDARY IMMUNODEFICINCY DISORDERS;

Secondary immunodeficiency occurs when the immune system is weakened by another treatment (the main medicine that causes problems are those that target the immune system including immunosuppressant drugs and chemotherapy), or illness. This disorder generally develops later in life. They are more common than Primary Immunodeficiency disorders.

• Leukemia (an example of cancer of the immune system); it is also called Leukaemia, is a group of blood cancers that usually begins in the bone marrow and results in high numbers of abnormal blood cells. These blood cells are not fully developed and they are called Blasts or Leukemia Cells. Symptoms may include; bleeding and bruising, fatigue, fever, in children; pale skin, an enlarged spleen or liver and an increased risk of infections (white blood cells, which are involved in fighting pathogens, may be suppressed or dysfunctional. This could cause the persons immune system to be unable to fight off simple infections or to start attacking other body cells). These symptoms occur due to the lack of normal blood cells; damage to the bone marrow, by way of displacing the normal bone marrow cells with higher numbers of immature white blood cells, results in a lack of blood platelets, which are important in the blood clotting process. Diagnosis; is made by Blood Tests or Bone Marrow Biopsy. The exact cause is unknown; leukemia like every other cancer, results from the mutations in the DNA. The risk factors include; smoking, ionizing radiation, some chemicals such as benzene, prior chemotherapy, down syndrome. Treatments may include; Bone marrow transplant, chemotherapy, radiation therapy, targeted therapy and supportive care.

• Human Immune Deficiency Virus ans Acquired Immune Deficiency Syndrome (HIV/AIDS); is a spectrum of conditions caused by infection with the human immunodeficiency virus. A person may not notice any symptoms, or may experience a brief period of Influenza-like illness. Typically, this is followed by a prolonged period with no symptoms. The progression of the infection interferes more with the immune system, increasing the risk of developing common infections such as tuberculosis, as well as other opportunistic infections and tumors. These late symptoms are referred to as Acquired Immunodeficiency Syndrome (AIDS). This stage is often associated with unintended weight loss. The risk factors include; exposure to infected blood, unprotected sex (anal and oral sex), sharp objects and breast milk, from mother to child during pregnancy. It could be prevented through; safe sex, screened blood, proper disposal of sharp objects etc. treatment used includes Antiretroviral Therapy.