AFEBABALOLOA UNIVERSITY ADO-EKITICOLLEGE

COLLEGE

MEDICINE AND HEALTH SCIENECE

DEPARTMENT

NURSING

LEVEL

300

NAME

ENO SUSAN A.

MATRIC NO

17/MHS01/116

COURSE TTITLE

MEDICAL SURGICAL NURSING 11

COURSE CODE

NSC 306

ASSIGNMENT TITLE

IMMUNODEFICIENCY DISORDER

PRIMARY IMMUNODEFICIENCY

The primary immunodeficiency disorder are

- Wiskott-Aldrich syndrome (WAS)
- Serve combined immunodeficiency disease (SCID)
- X- Linked Agammaglobulinemia (XLA)
- Chronic Granulomatous Diseases (CGD)
- Leukocytes adhesion Deficiency (LAD)
- Job Syndrome
- Bruton's Disease
- Ataxia-Telangiectasia
- DiGeorge Syndrome
- Selective Deficiency of IgA

1. Wiskott-Aldrich syndrome (WAS)

It is a rare genetic disorder of the immune system that primary affect boys. WAS is an X-linked recessive disease caused by mutation in the WAS gene, which provide instruction for production of Wiskott-Aldrich syndrome protein. The disorder is cauterized by abnormal immune function and a reduced ability to from blood cloth. This can result in prolonged episode of bleeding, recurrent bacteria and fungi infection and increase risk of cancer and auto-immune disease.

It mostly occurs in males due to its X- linked recessive pattern of inheritance, affecting between 1 and 10 males per million. The first sign are usually petechiae and bruising, resulting from a low platelet count (i.e. thrombocytopenia).

2. Serve Combined Immunodeficiency Disease (SCID)

Serve combined immunodeficiency disease is a group rare disorder caused by mutation in different gene involved in development and function of infection –fighting immune cells. Infant in SCID appear healthy at birth but are highly susceptible to serve infection.

The condition is fatal, usually within the first year or two of life unless infant receive immune- restoring treatment, such as transplant of blood forming stem cells, gene therapy or enzyme therapy. More than 80% of SCID infant do not have a family history of the condition. However, development of a new born screening test as made it possible to detect SCID before symptoms appears, helping ensure that affect infant receive lifesaving treatment.

Symptom of SCID occurs in infancy and include serious or life treating infection, especially viral infection, which may result in pneumonia and chronic diarrhea. Candida yeast infection of the mouth and diaper area and pneumonia caused by the fungus pneumocystis jirovecii also are common.

3. X- Linked Agammaglobulinemia (XLA)

XLA is an inherited immune disorder caused by an inability to produce B cells or the immunoglobulins (antibodies) that the B cells make. XLA is also called Bruton type agammaglobulinemia, X-linked infantile agammaglobulinemia and congential agammaglobulinemia.

Infant with XLA develop infection of the ear, throat, lungs and sinuses. Serious infection can also develop in the blood stream. Centrall nervious system, skin, and internal organs. This children tends to cope well with most short term viral infection but are very susceptible to chronic viral infection such as haepittis , they usually lake or have very small tonsils.

People with XLA have extreme low number of braib cells, and blood celss will show all extremely low levels of all type of immunoglobulin (antibodies). People with XLA fail to develop antibodies to specific germs and will not produce protective antibodies against immunization treatment

People with XLA receive intravenous (through the vein) or subcutaneous (just under the skin) immunoglobulin regularly, as well as antibiotics to treat infection.

4. Chronic Granulomatous Diseases (CGD)

Chronic Granulomatous Diseases is a genetic disorder in which white blood cells phagocytes are unable to kill certain type of bacteria and fungi. People with CGD are highly susceptible to frequent and sometimes life treating bacteria and fungi infection.

CGD is caused by defect in an enzymes, NADPH oxidase that phagocytes need to kill curtain bacteria and fungi. Mutation in one of five different gene can cause this defect.

People in CGD are highly susceptible to infection with certain bacteria and fungi such as staphylococcus aureus, serratia-marcescens, burkholdderia capacia, norcardia species.

These people may develop abscesses (boil) in their lungs, liver, spleen, bones, bones, or skin; and masses of cells, called granulomas that can obstruct the bowel or urinary tract. In some people, granulomas can cause an inflammatory bowel disease similar to crohn's disease.in addition, heart or kidney problems, diabetes, and autoimmune disease may occur in people with CGD, but this varies depending on which gene is mutated.

People with CGD take lifelong regiment of antibiotics and antifungals to prevent infections. Infections with interferon's gamma, a protein that improves the activity of phagocytes, also may help reduce the number of severe infections

5. Leukocytes Adhesion Deficiency (LAD)

Leukocytes adhesion deficiency is a rare, inherited immune disorder in which immune cells called phagocytes are unable to move to the site of an infection to fight off invading pathogens. People with LAD experience sass, life-threatening infections and poor wound healing. LAD is caused by a mutation in the gene ITGB2, which provides instructions for the phagocytes surface, molecule CD18.treatmenets for LAFD include antibiotics to prevent and treat infections and, in some cases, bone marrow transplant from a healthy donor.

SECONDARY IMMUNODEFICIENCY

Secondary immunodeficiencies, also known as acquired immunodeficiencies, can result from various immunosuppressive agents, for example

- Malnutrition
- Metabolic disease: diabetes mellitus and uremia
- Cancer such as leukemia
- Absence of spleen

- Human Immune Virus

1. MALNUTRITION

This is the most common cause of immunodeficiency .malnutrition can result from limited access to food sources and chronic disease that induces cachexia, such as neoplastic diseases. Diarrhea caused by infections and respiratory tract infection are common.t-cells production and function decrease in proportion to the severity of hypoproteinnemia;however,specificantibody titers and immune response to vaccine can be detected in a malnourished subject for relatively prolonged period.

Eventually, these immune responses decrease if malnutrition persists. The deficiency of micronutrients (e.g., zinc and ascorbic acids) contribute to increased susceptibility to infections through the weakening of barrier mucosa, therefore facilitating a pathogens invasiveness.

Other essential molecules have been shown to have specific roles in the immune system; for example, vitamin D appears to be necessary in the macrophages activity against intracellular pathogens, remarkably mycobacterium tuberculosis.

2. METABOLIC DISEASE: DIABETES MELLITUS

Many disease processes originating from dysfunctional metabolic pathways significantly affect the cells involved in the immune response. Diabetes mellitus and uremia resulting from kidney or liver disease are 2 common metabolic disorders with known deleterious effects on immunity, optimal control of the metabolic abnormalities usually leads to improved immune function.