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**MATRIC NO:** 17/MHS01/136

**DEPARTMENT:** NURSING

**LEVEL:** 300LEVEL

**MEDICAL SURGICAL ASSIGNMENT**

1. Identify and briefly explain 5 primary immunodeficiency disorders
2. Identify and briefly explain 2 secondary immunodeficiency disoders
3. Primary immunodeficiency disorders include :
* Wiscott-Aldrich syndrome
* Severe combined immunodeficiency disease (SCID)
* DiGeorge syndrome
* [Ataxia-telangectasia](https://www.medicinenet.com/ataxia/article.htm)
* Chronic granulomatous disease
1. **Wiscott-aldrich syndrome:** 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 the thrombocytopenia). It is also sometimes called the eczema-thrombocytopenia-immunodeficiency syndrome in keeping with Aldrich's original description in 1954. The WAS-related disorders of X-linked thrombocytopenia (XLT) and X-linked congenital neutropenia (XLN) may present similar but less severe symptoms and are caused by mutations of the same gene.
2. **Severe combined immunodeficiency disease:** 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 involves defective antibody response due to either direct involvement with B lymphocytes or through improper B lymphocyte activation due to non-functional T-helper cells. Consequently, both "arms" (B cells and T cells) of the adaptive immune system are impaired due to a defect in one of several possible genes. SCID is the most severe form of primary immunodeficiencies, and there are now at least nine different known genes in which mutations lead to a form of SCID. It is also known as the bubble boy disease and bubble baby disease because its victims are extremely vulnerable to infectious diseases and some of them.
3. **Digeorge syndrome**: DiGeorge syndrome, also known as 22q11.2 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.

DiGeorge syndrome is typically due to the deletion of 30 to 40 genes in the middle of chromosome 22 at a location known as 22q11.2. About 90% of cases occur due to a new mutation during early development, while 10% are inherited from a person's parents. It is autosomal dominant, meaning that only one affected chromosome is needed for the condition to occur. Diagnosis is suspected based on the symptoms and confirmed by genetic testing.

1. **Ataxia telangiectasia**: 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
2. **Chronic granulomatous disease:** **Chronic Granulomatous Disease** (CGD) is an inherited primary immunodeficiency **disease** (PIDD) 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.
3. Secondary immunodeficiency disorders include:
* AIDS
* Leukemia
	1. **AIDS:** This is a spectrum of conditions caused by infection with the human immunodeficiency virus (HIV). Following initial infection 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. If the infection progresses, it interferes more with the immune system, increasing the risk of developing common infections such as tuberculosis, as well as other opportunistic infections, and tumors which are otherwise rare in people who have normal immune function. These late symptoms of infection are referred to as acquired immunodeficiency syndrome (AIDS).This stage is often also associated with unintended weight loss.
	2. **Leukemia:** Leukemia is a cancer of the blood or bone marrow. Bone marrow produces blood cells. Leukemia can develop due to a problem with blood cell production. It usually affects the leukocytes, or white blood cells.