

Medical surgical nursing

17/MHS02/023

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-Identity and briefly explain
5 primary immunodeficiency
disorders.

The types of primary
immunodeficiency disease
are far too numerous to list
all but few common ones:

- Wiscott-Aldrich
syndrome
- Severe combined
immunodeficiency disease
(SCID)
- Transient
hypogammaglobulinemia of
infancy
- Agammaglobulinemia
- Selective IgA
deficiency

1. **Wiscott-Aldrich**

syndrome: is an X-linked
recessive disease

characterized by eczema, thrombocytopenia (low platelet count), immune deficiency and bloody diarrhea (secondary to the thrombocytopenia). A.K.A the **eczema-thrombocytopenia-immunodeficiency syndrome** in keeping with Aldrich's original description in 1954.[2] The WAS-related disorders of XLT and XLN may present similar but less severe symptoms and are caused by mutations of the same gene.

2. **Severe combined immunodeficiency disease (SCID):** Infants with SCID appear healthy at birth but are highly susceptible to severe infections. The

condition is fatal, usually within the first year or two of life, unless infants receive immune-restoring treatments, such as transplants of blood-forming stem cells, gene therapy, or enzyme therapy. More than 80 percent of SCID infants do not have a family history of the condition. However, development of a newborn screening test has made it possible to detect SCID before symptoms appear, helping ensure that affected infants receive life-saving treatments.

3. Transient Hypogammaglobulinemia of Infancy:

Following birth, maternal immunoglobulin G (IgG) is catabolized, and IgG synthesized by the infant gradually accumulates. Serum levels typically reach their physiologic nadir in infants aged 3–6 months. THI is characterized by decreased serum IgG with or without decreased IgA and IgM levels less than 2 standard deviations from age-adjusted reference range levels in infants older than 6 months of age in the first years of life but with normal to near-normal antibody responses to protein immunizations.

These levels usually increase to the reference range by age 2–6 years in children with THI. Abnormalities of T-cell help have been identified as a cause of THI, but recent studies suggest that THI may be an intrinsic B-cell defect with abnormal antibody responses, especially to *Streptococcus pneumoniae*, respiratory viruses, and *Haemophilus influenzae* type B.

4. **Agammaglobulinemia:** Agammaglobulinemia is a group of inherited immune deficiencies characterized by a low concentration of antibodies in the blood

due to the lack of particular lymphocytes in the blood and lymph. Antibodies are proteins (immunoglobulins, (IgM), (IgG) etc) that are critical and key components of the immune system. The types of agammaglobulinemia are: XLA, the much rarer X-linked agammaglobulinemia with growth hormone deficiency and autosomal recessive agammaglobulinemia (ARAG).

5. **Selective IgA deficiency:** is an immune system condition in which one lacks or doesn't have

enough immunoglobulin A (IgA), a protein that fights infection (antibody). Most people with selective IgA deficiency don't have recurrent infections. However, some people who have IgA deficiency experience pneumonia, ear infections, sinus infections, allergies, asthma and diarrhea.

-Identify and briefly explain 2 secondary immunodeficiency disorders.

Secondary immunodeficiency disorders happen when an outside source like a toxic chemical or infection attacks your body and they include:

- cancers of the immune system, like LEUKEMIA

- MULTIPLE MYELOMA(cancer of the plasma cells, which produce antibodies)
1. LEUKEMIA: A cancer of blood-forming tissues, hindering the body's ability to fight infection. Leukemia is cancer of blood-forming tissues, including bone marrow. Many types exist such as acute lymphoblastic leukaemia, acute myeloid leukaemia and chronic lymphocytic leukemia. Requires a medical diagnosis. Lab tests or imaging always required. Many patients with slow-growing types of leukaemia don't have symptoms. Rapidly

growing types of leukaemia may cause symptoms that include fatigue, weight loss, frequent infections and easy bleeding or bruising. Treatment is highly variable. For slow-growing leukaemias, treatment may include monitoring. For aggressive leukaemias, treatment includes chemotherapy that's sometimes followed by radiation and stem-cell transplant.

2. **MULTIPLE MYELOMA:**
The plasma cells are a type of white blood cell in the bone marrow. With this condition, a group of plasma cells

becomes cancerous and multiplies. The disease can damage the bones, immune system, kidneys and red blood cell count. Treatment can help, but this condition can't be cured. Requires a medical diagnosis. Lab tests or imaging always required. Chronic: can last for years or be lifelong. Symptoms may not be present or may be non-specific, such as loss of appetite, bone pain and fever.

Treatments include medication, chemotherapy, corticosteroids, radiation or a stem-cell transplant.