

## MED SURG ASSIGNMENT

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MATRIC NO: 18/MHS02/199

DEPT: NURSING

LEVEL: 300LEVEL

### ASSIGNMENT

1. Identify and explain 5 primary immunodeficiency disorders
2. Identify and briefly explain 2 secondary immunodeficiency disorders

### PRIMARY IMMUNODEFICIENCY DISORDERS

1. AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME
2. APS-1 (APECED)
3. BENTA DISEASE
4. CASPASE EIGHT DEFICIENCY STATE
5. LRBA DEFICIENCY

#### 1. AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME

Autoimmune lymphoproliferative syndrome (ALPS) is a rare immune disorder first described by NIH scientists in the mid-1990s that can cause numerous autoimmune problems, such as low levels of red blood cells, clot-forming platelets, and infection-fighting white blood cells. These problems can increase the risk of infection and hemorrhage.

#### 2. APS-1 (APECED)

Autoimmune polyglandular syndrome type 1 (APS-1), also called autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED), causes a diverse range of symptoms, including autoimmunity against different types of organs and increased susceptibility to candidiasis, a fungal infection caused by Candida yeast.

#### 3. BENTA DISEASE

BENTA disease is a rare genetic disorder of the immune system caused by mutations in the gene CARD11. The disease is characterized by high levels of certain immune cells starting in infancy, an enlarged spleen, enlarged lymph nodes, immunodeficiency, and an elevated risk of lymphoma, a type of cancer

#### 4. CASPASE EIGHT DEFICIENCY STATE (CEDS)

Caspase eight deficiency state, or CEDS, is a very rare genetic disorder of the immune system caused by mutations in the CASP8 gene. CEDS is characterized by an enlarged spleen and lymph nodes, recurrent sinus and lung infections, recurrent viral infections, and a low level of

infection-fighting antibodies. NIH researchers first described this condition in two siblings in 2002.

## 5.LRBA DEFICIENCY

LRBA deficiency is a rare genetic disorder of the immune system caused by mutations in the LRBA gene. This disease impairs normal immune system function and results in autoimmunity, recurrent infections and an increased risk of lymphoma, a type of cancer. People with LRBA deficiency have excessive numbers of immune cells called lymphocytes, which sometimes enter and accumulate in organs where lymphocytes typically are not present in large numbers such as the gut, lungs and brain. This can cause a variety of symptoms

## SECONDARY IMMUNODEFICIENCY DISORDERS

1. HIV & AIDS
2. LEUKEMIA

### HIV & AIDS

AIDS is a disease that can develop in people with HIV. It's the most advanced stage of HIV. But just because a person has HIV doesn't mean they'll develop AIDS.

HIV kills CD4 cells. Healthy adults generally have a CD4 count of 500 to 1500 per cubic millimeter. A person with HIV whose CD4 count falls below 200 per cubic millimeter will be diagnosed with AIDS.

A person can also be diagnosed with AIDS if they have HIV and develop an opportunistic infection or cancer that's rare in people who don't have HIV. An opportunistic infection, such as pneumonia, is one that takes advantage of a unique situation, such as HIV.

Untreated, HIV can progress to AIDS within a decade. There's no cure for AIDS, and without treatment, life expectancy after diagnosis is about three years. This may be shorter if the person develops a severe opportunistic illness. However, treatment with antiretroviral drugs can prevent AIDS from developing.

If AIDS does develop, it means that the immune system is severely compromised. It's weakened to the point where it can no longer fight off most diseases and infections. That makes the person vulnerable to a wide range of illnesses, including:

- Pneumonia
- Tuberculosis

### LEUKEMIA

Leukemia is cancer of the body's blood-forming tissues, including the bone marrow and the lymphatic system.

Many types of leukemia exist. Some forms of leukemia are more common in children. Other forms of leukemia occur mostly in adults.

Leukemia usually involves the white blood cells. Your white blood cells are potent infection fighters — they normally grow and divide in an orderly way, as your body needs them. But in

people with leukemia, the bone marrow produces abnormal white blood cells, which don't function properly.