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MATRIC NO: 17/MHS02/014
LEVEL: 300L
DEPARTMENT: NURSING SCIENCE

ASSIGNMENT TITLE: IMMUNODEFICIENCY DISODERS
COURSE TITLE: MEDICAL SURGICAL NURSING II
COURSE CODE: NSC 306

QUESTION

Immunodeficiency disorder is the absence or failure of normal function of one or more elements of the immune system. There are two major types of immunodeficiency disorders: PRIMARY AND SECONDARY.

1. Identify and briefly explain 5 primary immunodeficiency disorders

2. Identify and briefly explain 2 secondary immunodeficiency disorders

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IMMUNODEFICIENCY DISORDER

1. PRIMARY IMMUNODEFICIENCY DISORDER

These disorders are usually present at birth and are genetic disorders that are usually hereditary. They typically become evident during infancy or childhood. However, some primary immunodeficiency disorders (such as common variable immunodeficiency) are not recognized until adulthood.

5 PRIMARY IMMUNODEFICIENCY DISORDERS INCLUDE:

- Chronic granulomatous disease (CGD)
- Common variable immunodeficiency (CVID)
- Selective IgA deficiency
- Severe combined Immunodeficiency (SCID)
- X-linked agammaglobulinemia (XLA)

A. CHRONIC GRANULOMATOUS DISEASE (CGD)

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.

People with CGD are unable to fight off common germs and get very sick from infections that would be mild in healthy people. This is because the presence of CGD makes it difficult for cells called neutrophils to produce hydrogen peroxide. The immune system requires hydrogen peroxide to fight specific kinds of bacteria and fungi.

These severe infections can include skin or bone infections and abscesses in internal organs (such as the lungs, liver or brain).

Aside from the defective neutrophil function in CGD, the rest of the immune system is normal. People with CGD can be generally healthy until they become infected with one of these germs. The severity of this infection can lead to prolonged hospitalizations for treatment.

Children with CGD are often healthy at birth, but develop severe infections in infancy or early childhood. The most common form of CGD is genetically inherited in an X-linked manner, meaning it only affects boys. There are also autosomal recessive forms of CGD that affect both sexes.

Therapeutic options for CGD include prophylactic antibiotics and antifungal medications, interferon-gamma injections, and aggressive management of acute infections. Bone marrow transplantation can cure CGD, however this therapy is complex and transplant candidates and donors must be carefully selected, weighing the risks and benefits carefully.

B. COMMON VARIABLE IMMUNODEFICIENCY (CVID)

Common Variable Immunodeficiency (CVID) is an antibody deficiency that leaves the immune system unable to defend against bacteria and viruses, resulting in recurrent and often severe infections primarily affecting the ears, sinuses, and respiratory tract. (Sinopulmonary infections). Permanent damage to the respiratory tract (bronchiectasis) may occur due to severe and repeated infections.

Both males and females are affected. It is one of the most common forms of primary immunodeficiency disease (PIDD).

CVID can be associated with autoimmune disorders that affect other blood cells causing low numbers of white cells or platelets, anemia, arthritis and other conditions such as endocrine disorders. Gastrointestinal problems including chronic diarrhea, weight loss, nausea, vomiting and abdominal pain can also be present. In some forms of CVID, patients develop granulomas in the lungs, lymph nodes, liver, skin or other organs.

People with CVID are also at an increased risk for certain cancers (lymphoid and gastrointestinal cancers primarily).

Treatment for CVID involves immunoglobulin replacement, preventative antibiotics, and if indicated management of autoimmune and granulomatous disease.

C. SELECTIVE IGA DEFICIENCY

Selective IgA Deficiency is the most common primary immunodeficiency disease (PIDD). People with this disorder have absent levels of a blood protein called immunoglobulin A (IgA). IgA protects against infections of the mucous membranes lining the mouth, airways and digestive tract.

Although individuals with Selective IgA Deficiency do not produce IgA, they do produce all the other kinds of immunoglobulin. This is why many people with IgA deficiency appear healthy or only have mild illness such as gastrointestinal infections.

A common problem in IgA deficiency is susceptibility to infections. A second major problem in IgA deficiency is increased occurrence of autoimmune diseases. Also, many people with Selective IgA Deficiency also have allergies or asthma.

D. SEVERE COMBINED IMMUNODEFICIENCY (SCID)

Severe Combined Immunodeficiency (SCID) is an inherited primary immunodeficiency disease (PIDD) that typically presents in infancy results in profound immune deficiency condition resulting in a weak immune system that is unable to fight off even mild infections. It is considered to be the most serious PIDD.

SCID is caused by genetic defects that affects the function of T cells. Depending on the type of SCID, B cells and NK cells can also be affected.

These cells play important roles in helping the immune system battle bacteria, viruses and fungi that cause infections.

There are several forms of SCID. The most common type is linked to a problem in a gene on the X chromosome, affecting only males. Women may carry the condition, but they also inherit a normal X chromosome.
Other forms of SCID are caused by a deficiency of the enzyme adenosine deaminase (ADA) and a variety of other genetic defects.

E. X-LINKED AGAMMAGLOBULINEMIA (XLA)

X-Linked Agammaglobulinemia (XLA) is an inherited immunodeficiency in which the body is unable to produce the antibodies needed to defend against bacteria and viruses.

Frequently called Bruton's Agammaglobulinemia, XLA is caused by a genetic mistake in a gene called Bruton's Tyrosine Kinase (BTK), which prevents B cells from developing normally. B cells are responsible for producing the antibodies that the immune system relies on to fight off infection.

The most common bacteria causing infection in XLA are Streptococcus, Staphylococcus and Haemophilus.

2. SECONDARY IMMUNODEFICIENCY DISORDERS

These disorders generally develop later in life and often result from use of certain drugs or from another disorder, such as diabetes or human immunodeficiency virus (HIV) infection. They are more common than primary immunodeficiency disorders.

2 SECONDARY IMMUNODEFICIENCY DISORDERS INCLUDE:

- AIDS
- Multiple myeloma (cancer of the plasma cells, which produce antibodies)

A. AIDS

AIDS is a disease that can develop in people with HIV. It’s the most advanced stage of HIV.
HIV kills CD4 cells. Healthy adults generally have a CD4 count of 500 to 1,500 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
- Oral thrush, a fungal infection in the mouth or throat
- Cytomegalovirus (cmv), a type of herpes virus
- Cryptococcal meningitis, a fungal infection in the brain
- Toxoplasmosis, a brain infection caused by a parasite
- Cryptosporidiosis, an infection caused by an intestinal parasite
- Cancer, including kaposi's sarcoma (ks) and lymphoma

The shortened life expectancy linked with untreated AIDS isn't a direct result of the syndrome itself. Rather, it's a result of the diseases and complications that arise from having an immune system weakened by AIDS.

B. MULTIPLE MYELOMA

Multiple myeloma is a type of cancer that affects plasma cells. Plasma cells are a type of white blood cell found in bone marrow, which is the soft tissue inside most of the bones that produces blood cells. In the bone marrow, plasma cells make antibodies. These are proteins that help your body fight off diseases and infections.

Multiple myeloma occurs when an abnormal plasma cell develops in the bone marrow and reproduces itself very quickly. The rapid reproduction of malignant, or cancerous, myeloma cells eventually outweighs the production of healthy cells in the bone marrow. As a result, the cancerous cells begin to accumulate in the bone marrow, crowding out the healthy white blood cells and red blood cells.

Like healthy blood cells, cancerous cells try to make antibodies. However, they can only produce abnormal antibodies called monoclonal proteins, or M proteins. When these harmful antibodies collect in the body, they can cause kidney damage and other serious problems.