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Department : nursing

**Medical and surgical nursing assignment**

1. Identify and explain 5 primary immune deficiency disorder.

Immunodeficiency disorders disrupt your body’s ability to defend itself against bacteria, viruses, and parasites.There are two types of immunodeficiency disorders: those you are born with (primary), and those that are acquired (secondary). Anything that weakens your immune system can lead to a secondary immunodeficiency disorder.

5 primary immune deficiency disorder includes;

1. X-linked agammaglobulinemia (XLA)
2. common variable immunodeficiency (CVID)
3. severe combined immunodeficiency (SCID), which is known as alymphocytosis or “boy in a bubble” disease.
4. Hyper immunoglobulin E syndromes
5. Hyper immunoglobulin M syndromes
6. **X-linked agammaglobulinemia (XLA)**

XLA is caused by an inability to produce B cells or immunoglobulins (antibodies), which are made by B cells. People with XLA develop frequent infections of the ears, throat, lungs, and sinuses.

**Causes**

The mutated gene responsible for XLA codes for the protein Bruton tyrosine kinase, or BTK, and is located on the X chromosome, and is an X-linked recessive disease. Because males only have one X chromosome, they are affected if they inherit an X chromosome containing a mutated BTK gene

**Treatment**

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

1. **Common variable immunodeficiency**

CVID is caused by a variety of different genetic abnormalities that result in a defect in the capability of immune cells to produce normal amounts of protective antibodies. People with CVID experience frequent bacterial and viral infections of the upper airway, sinuses, and lungs.

**Causes**

CVID is caused by a variety of different genetic abnormalities that result in a defect in the capability of immune cells to produce normal amounts of all types of antibodies. CVID is also linked to IgA deficiency, a related condition in which only the level of the antibody immunoglobulin A (IgA) is low, while levels of other antibody types are usually normal or near normal.

**Treatment**

CVID is treated with intravenous immunoglobulin infusions or subcutaneous (under the skin) immunoglobulin injection to partially restore immunoglobulin levels. The immunoglobulin given by either method provides antibodies from the blood of healthy donors.

1. **Severe combined immunodeficiency**

SCID is a group of rare, life-threatening disorders caused by mutations in different genes involved in the development and function of infection-fighting T and B cells. Infants with SCID appear healthy at birth but are highly susceptible to severe infections. Symptoms of SCID occur in infancy and include serious or life-threatening infections, especially viral infections, which may result in pneumonia and chronic diarrhea. Candida (yeast) infections of the mouth and diaper area and pneumonia caused by the fungus Pneumocystis also are common.

**Causes**

**Adenosine deaminase (ADA) deficiency,** in which infants lack the ADA enzyme necessary for T-cell survival. X-linked SCID, which is caused by mutations in a gene on the X chromosome, primarily affects male infants. Boys with this type of SCID have white blood cells that grow and develop abnormally. As a consequence, they have low numbers of T cells and natural killer cells, and their B cells do not function.

**Treatment**

**Hematopoietic (blood-forming) stem cell transplantation** is the standard treatment for infants with SCID. Ideally, infants with SCID receive stem cells from a sibling who is a close tissue match. Children who have SCID with ADA deficiency have been treated somewhat successfully with **enzyme replacement therapy called PEG-ADA.** **Gene therapy** can be an effective treatment for some types of SCID, including X-linked SCID.

1. **Hyper immunoglobulin E syndrome**

Many different syndromes are known to lead to high levels of an antibody called immunoglobulin E, or IgE. Many more such syndromes likely remain unknown. Collectively, these conditions are called hyper-IgE syndromes, or HIES. Symptoms may include; eczema,. Skin abscesses and infection, repeated sinus infections, repeated lung infections, etc.

Causes

This disease is often caused by a genetic change (mutation) that takes place in the STAT3gene on chromosome 17. How this gene abnormality causes the symptoms of the disease is not well understood.

Treatment

Antibiotics, usually trimethoprim/sulfamethoxazole, are given continuously to prevent staphylococcal infections. The rash is treated with moisturizing creams, antihistamines, and, if infection is likely, antibiotics. Respiratory infections are treated with antibiotics.

1. **Hyper immunoglobulin M syndrome**

Hyper-immunoglobulin M (IgM) syndromes are rare, inherited conditions in which the immune system fails to produce normal levels of the antibodies IgA, IgG and IgE but can produce normal or elevated levels of IgM. Various gene defects that impair communication between T cells and antibody-producing B cells can lead to hyper-IgM syndromes. Hyper-IgM syndromes can cause severe respiratory infections in infancy and a higher risk of rare infections throughout life.

Causes:

Hyper IgM syndromes are caused by very rare, one-in-a-million, and potentially life-threatening genetic mutations that severely compromise the immune system and resulting in the individual's inability to produce antibodies. Patients with hyper IgM are at significant risk for opportunistic and repeated infections.

Treatment

Treatment includes regular intravenous or subcutaneous antibody replacement therapy, anti-fungal prophylactics, and in some cases, bone marrow transplant from a healthy donor.

1. Identify and explain 2 secondary immune deficiency disorder.

Secondary immunodeficiency occurs when the function of the immune system is disrupted secondary to an underlying disease state, medications, surgery or other medical procedure.

Examples of secondary immunodeficiency disorders include:

1. AIDS.
2. cancers of the immune system, like leukemia.
3. immune-complex diseases, like viral hepatitis
4. multiple myeloma (cancer of the plasma cells, which produce antibodies)

**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 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 yearsTrusted Source. 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, cytomegalovirus, etc.

**Causes**

1. Majorly by unprotected sex (including anal and oral sex)
2. contaminated blood transfusions
3. hypodermic needles
4. mother to child during pregnancy, delivery, or breastfeeding.

**Symptoms**

Within a few weeks of HIV infection, flu-like symptoms such as fever, sore throat and fatigue can occur. Then the disease is usually asymptomatic until it progresses to AIDS. AIDS symptoms include weight loss, fever or night sweats, fatigue and recurrent infections.

**Prevention**

1. Safe sex( practice the use of condoms)
2. Male circumcising
3. Needles should be discarded immediately after use

**Treatment**

There is currently no specific cure for hiv/aids but it can be managed through the use of antiviral drugs: Antiviral drugs are a class of medication used for treating viral infections. Most antivirals target specific viruses, while a broad-spectrum antiviral is effective against a wide range of viruses.Unlike most antibiotics, antiviral drugs do not destroy their target pathogen; instead they inhibit their development. Most antivirals are considered relatively harmless to the host, and therefore can be used to treat infections.

**Diagnosis**

This is done trough blood test (antibody test, antibody and p24 antigen test and PCR)

**Pathophysiology**

Acquired immune deficiency syndrome (AIDS) is caused by the HIV or human immunodeficiency virus. The infection causes progressive destruction of the cell-mediated immune (CMI) system, primarily by eliminating CD4+ T-helper lymphocytes.Decreased immunity leads to opportunistic infections and certain cancers. Opportunistic infections are caused by organisms that do not cause infections in healthy individuals. HIV also directly damages certain organs like the brain.

**Cancer of the immune system (leukemia)**

Leukemia is a blood cancer caused by a rise in the number of white blood cells in your body.Those white blood cells crowd out the red blood cells and platelets that your body needs to be healthy. The extra white blood cells don’t work right.

Leukemia can be classified into two;

I**) Acute leukemia** happens when most of the abnormal blood cells don’t mature and can’t carry out normal functions. It can get bad very fast.

Ii) **Chronic leukemia** happens when there are some immature cells, but others are normal and can work the way they should. It gets bad more slowly than acute forms do.

**Causes**

There is no exact cause of leukemia but some factors may trigger it.Such risk factors includes;

1. Smoke
2. Consistent exposure to radiation or certain chemicals
3. radiation therapy or chemotherapy to treat cancer
4. Have a family history of leukemia
5. Have a genetic disorder like Down syndrome

**Prevention**

1. Be a non-smoker:Not smoking is the best way to lower your risk of leukemia.
2. Maintain a healthy body weight.
3. Avoid breathing in benzene and formaldehyde

**Symptoms**

1. Weakness or fatigue
2. Bruising or bleeding easily
3. Fever or chills
4. Infections that are severe or keep coming back
5. Pain in your bones or joints
6. Headaches
7. Vomiting
8. Seizures , etc

**Treatment:**

1. **Chemotherapy:** uses drugs to kill cancer cells in your blood and bone marrow. You can get the medicine through a shot into a vein, as a pill,Into the fluid around your spinal cord.
2. **Radiation:** uses high-energy X-rays to kill leukemia cells or keep them from growing.
3. **Biologic therapy:** helps your immune system find and attack cancer cells. Drugs like interleukins and interferon can help boost your body's natural defenses against leukemia.
4. **Targeted therapy:** uses drugs to block specific genes or proteins that cancer cells need to grow
5. **Stem cell transplant:** replaces the leukemia cells in your bone marrow with new ones that make blood.
6. **Surgery:** A splenectomy is surgery to remove the entire spleen, a delicate, fist-sized organ that sits under the left rib cage near the stomach.

**Diagnosis**

1. **Blood tests.** A complete blood count (CBC) looks at the number and maturity of different types of blood cells. A blood smear looks for unusual or immature cells.
2. **Bone marrow biopsy.** This test involves marrow taken from your pelvic bone with a long needle. It can tell your doctor what kind of leukemia you have and how severe it is.
3. **Spinal tap.** This involves fluid from your spinal cord. It can tell your doctor whether the leukemia has spread.
4. **Imaging tests.** Things like CT, MRI, and PET scans can spot signs of leukemia.

**Pathophysiology**

Sometimes an immature blast cell have two gene mutations which prevent it from maturing into a specialized blood cell and cause it to multiply out of control. These immature blast cells crowd the bone marrow and impair the ability of the bones to make healthy blood cells.