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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

ANSWER

1. 5 PRIMARY IMMUNODEFICIENCY DISORDERS INCLUDE:
* Chronic granulomatous disease (CGD)
* Common variable immunodeficiency (CVID)
* Selective IgA deficiency
* Severe combined Immunodeficiency (SCID)
* X-linked agammaglobulinemia (XLA)

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. Chronic granulomatous disease (CGD) is an inherited disorder that occurs when a type of white blood cell (phagocyte) that usually helps your body fight infections doesn't work properly. As a result, the phagocytes can't protect your body from bacterial and fungal infections.

People with chronic granulomatous disease may develop infections in their lungs, skin, lymph nodes, liver, stomach and intestines, or other areas. They may also develop clusters of white blood cells in infected areas. Most people are diagnosed with CGD during childhood, but some people may not be diagnosed until adulthood.

The symptoms include: fever, Fever, Chest pain when inhaling or exhaling, Swollen and sore lymph glands, A persistent runny nose, Skin irritation that may include a rash, swelling or redness, Swelling and redness in your mouth, Gastrointestinal problems that may include vomiting, diarrhea, stomach pain, bloody stool or a painful pocket of pus near the anus.

Treatment

Antibiotics

Physicians often prescribe the antibiotic trimethoprim-sulfamethoxazole to prevent bacterial infections.[26] This drug also has the benefit of sparing the normal bacteria of the digestive tract. Fungal infection is commonly prevented with itraconazole, although a newer drug of the same type called voriconazole may be more effective. The use of this drug for this purpose is still under scientific investigation.

Immunomodulation

Interferon, in the form of interferon gamma-1b (Actimmune) is approved by the Food and Drug Administration for the prevention of infection in CGD. It has been shown to reduce infections in CGD patients by 70% and to decrease their severity. Although its exact mechanism is still not entirely understood, it has the ability to give CGD patients more immune function and therefore, greater ability to fight off infections. This therapy has been standard treatment for CGD for several years.

Hematopoietic stem cell transplantation (HSCT)Hematopoietic stem cell transplantation from a matched donor is curative although not without significant risk.

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.

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.

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.

X-Linked Agammagloblulinemia

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.

The most common treatment for XLA is an intravenous infusion of immunoglobulin every week, for life. IVIg is a human product extracted and pooled from thousands of blood donations. IVIg does not cure XLA but increases the patient's lifespan and quality of life, by generating passive immunity, and boosting the immune system. With treatment, the number and severity of infections is reduced. With IVIg, XLA patients may live a relatively healthy life.

1. Secondary immunodeficiency disorders include:

Multiple Myeloma:

Multiple myeloma is a cancer that forms in a type of white blood cell called a plasma cell. Plasma cells help you fight infections by making antibodies that recognize and attack germs.

Multiple myeloma causes cancer cells to accumulate in the bone marrow, where they crowd out healthy blood cells. Rather than produce helpful antibodies, the cancer cells produce abnormal proteins that can cause complications.

Some symptoms include; Bone pain, especially in your spine or chest, Nausea, Constipation, Loss of appetite, Mental fogginess or confusion, Fatigue, Frequent infections, Weight loss, Weakness or numbness in your legs, Excessive thirst etc.

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.

AIDS( Acquired Immunodeficiency Syndrome)

AIDS is disease in which there is a severe loss of the body's cellular immunity, greatly lowering the resistance to infection and malignancy. 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.

The symptoms include:

* Pain areas: in the abdomen
* Pain circumstances: can occur while swallowing
* Cough: can be dry
* Whole body: fatigue, fever, loss of appetite, malaise, night sweats, or sweating
* Gastrointestinal: nausea, persistent diarrhoea, vomiting, or watery diarrhoea
* Throat: difficulty swallowing or soreness
* Groin: sores or swelling
* Mouth: ulcers or white tongue
* Also common: opportunistic infection, headache, oral thrush, pneumonia, red blotches, skin rash, swollen lymph nodes, or wasting

AIDS has no cure but can be managed by adherence to Antiretroviral disease.