NAME: BOBOLA PRAISE

MATRIC NO: 18/MHS02/200

 TYPES OF PRIMARY IMMUNODEFICIENCY DISORDERS

1. Wiscott-Adrich syndrome
2. Chronic granulomatous disease
3. Selective IgA deficiency
4. DiGeorge syndrome
5. Agammaglobulinemia
6. Wiscott-Aldrich syndrome: is a rare X-linked recessive disease characterized by eczema, thrombocytopenia (low platelet count), Immune deficiency, and bloody diarrhea (secondary to the thrombocytopenia). It is sometimes called the eczema-thrombocytopenia-immunodeficiency syndrome. The WAS-related disorders of X-linked thrombocytopenia and X-linked congenital neutropenia (XLN) may present similar but less severe symptoms and are caused by mutations of the same gene. Signs and Symptoms: petechae and bruising, resulting in low platelets counts. Spontaneous nose bleeds and bloody diarrhea are also common and eczema typically develops within the first month of life. Recurrent bacterial infections develops within three months
7. Chronic granulomatous disease: is an inherited primary immunodeficiency disease 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 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 needs 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 lung, liver or brain). 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. 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 affects both sexes.
8. Selective IgA deficiency: it is an immune system condition in which the bodies don’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.
9. DiGeorge syndrome: is a syndrome caused by the deletion of a small segment of chromosome 22. While the symptoms can vary, they often include congenital heart problems, specific facial features, frequent infections, development delay, learning problems and cleft palate. Associated conditions include kidney problems, hearing loss and autoimmune disorders such as rheumatoid arthritis or Graves’ disease. Diagnosis: diagnosis of DiGeorge syndrome can be difficult due to the number of potential symptoms and the variation in phenotypes between individuals. It is suspected in patients with one or more signs of the deletion. In these cases a diagnosis of 22q11.2DS is confirmed by observation of a deletion of part of the long arm of chromosome 22, region 1, band 1, and sub-band 2.
10. Agammaglobulinemia: is an inherited disorder in which a person has a very low level of protective immune system proteins called immunoglobulin. Immunoglobulin’s are a type of antibody. Low levels of these antibodies make you more likely to get infections. Causes: It is caused by a gene defect that blocks the growth of normal, mature immune cells called B lymphocytes. As a result the body makes very little (if any) immunoglobulin’s. Immunoglobulin’s play a major role in the immune response, which protects against illness and infection. People with the disorder develop infections again and again. Common infections include ones that are due to bacteria such as Haemophilus influenza, pneumococci (streptococcus pneumonia), and staphylococci. Common sites of infections include: Gastrointestinal tract, joints, lungs, skin, and upper respiratory tract.

TYPES OF SECONDARY IMMUNODEFICIENCY DISORDERS

1. Leukemia
2. Multiple myeloma
3. Leukemia: is a blood cancer caused by 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. Symptoms include: Weakness or fatigue, bruising or bleeding easily, fever or chills, infections that keep coming back, pain in your bones or joints, headaches, vomiting, seizures, weight loss, nights sweats, shortness of breath, swollen lymph nodes or organs like spleen. Risk factors: smoke, exposed to lots of radiations and chemicals, have a family history of leukemia, have a genetic disorder like Down syndrome.

TYPES OF LEUKEMIA

1. Acute lymphocytic leukemia: this is the most common form of childhood leukemia. It can spread to your lymph nodes and central nervous system.
2. Acute myelogenous leukemia: This is the second most common form of childhood leukemia and one of the most common forms for adults.
3. Chronic lymphocytic leukemia: This is the most common forms of leukemia. Some kinds of CLL will be stable for years and won’t need treatment. But with others, your body isn’t able to create normal blood cells, and you’ll need treatment.
4. Chronic myelogenous leukemia: with this form, you might not have noticeable symptoms. You might not be diagnosed with it until you have a routine blood test. People 65 and older have a higher risk of this type.
5. Multiple myeloma: this is a type of cancer that forms in the plasma, a type of white blood cell that normally produces antibodies. Often no symptoms are noticed initially. As it progresses, bone pain, bleeding, frequent infections, and anemia may occur. Complications may include amyloidosis; kidney problems bone fractures, overly thick blood. Risk factors: obesity, radiation exposure, family history, and certain chemicals. Multiple myeloma can develop from monoclonal gammopathy of undetermined significance that progress to smoldering myeloma. The abnormal plasma cells produce abnormal antibodies, which can cause kidney problems and overly thick blood. Diagnostic method: blood or urine test, bone marrow biopsy, medical imaging. Treatment: steroids, chemotherapy, thalidomide, stem cell transplant, bisphosphonate’s, and radiation therapy.