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QUESTIONS

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 disorder: PRIMARY AND SECONDARY.

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

ANSWERS

1. Identify and briefly explain 5 primary immunodeficiency disorders.
* Immunoglobulin A deficiency
* Thymic hypoplasia
* Severe combined immunodeficiency disease
* Granulomatosis with polyangiitis
* Partial combined immunodeficiency disease

Immunoglobulin A deficiency: This is a genetic immunodeficiency, a type of hypogammaglobulienemia. People with this deficiency lack immunoglobulin A (IgA), a type of antibody that protects against infections of the mucous lining the mouth, airways , and digestive tract, It is define as an undetectable serum IgA level in the presence of normal serum levels of IgG and IgM, in persons older than 4 years. It is the most common of primary antibody deficiencies. Most such persons remain healthy throughout their lives and are diagnosed.

Thymic hypoplasia: Thymic refers to the thymus which is an immune organ that sits between the lungs, hypo- refers to under, and –plasia refers to development. So Thymic hypoplasia is a condition where the thymus is underdeveloped and has a reduced number of cells.

Severe combined immunodeficiency disease (SCID): is a group of rare disorders caused by mutations in different genes involved in the development and function of infection-fighting immune cells. Infants with SCID appear healthy at birth but are highly susceptible to severe infections. The condition is fatal, usually within the first year or two of life, unless infants receive immune-restoring treatments, such as transplants of blood-forming stem cells, genes therapy, or enzyme therapy.

Granulomatosis with polyangiitis: Previously known as Wegener’s granulomatosis (WG), is an extremely rare long-term systemic disorder that involves formation of granulomas and inflammation of blood vessels (vasculititis). It is a form of vasculitis that affects small- and medium- size vessels in many organs but most commonly affects the upper respiratory tract, lungs and kidneys.

Partial combined immunodeficiency disease: Thos can also be called Wiskott-Aldorish syndrome is an X-linked disorder characterized by an altered cell surface glycoprotein structure (CD43 or sialophorin) common to lymphocyte and platelets. Its clinical features include a microcytic thrombocytopenia that distinguishes this disorder from idiopathic thrombocytopenic purpura and other forms of normocytic thrombocytopenia. The immunologic findings are variable but usually include impaired humoral responses to polysaccharide antigens and elevated serum IgA and IgE levels.

1. Identify and briefly explain 2 secondary immunodeficiency disorders.
* Diabetes mellitus
* Burn

Diabetes mellitus: This is a metabolic disorder that affects the pancreas, it can be as a result of the pancreas not producing enough insulin, or the cells of the body not responding to the insulin produced. They are three types of diabetes mellitus they are:

* Type 1: This is known as insulin dependent diabetes mellitus (IDDM) with beta cell destruction or defect in function: immune mediated- presence of islet or insulin antibodies that identify the autoimmune process leading to beta cell destruction it is sometimes idiopathic.
* Type2: Non-insulindependentDM (NIDDM), insulin resistance with relative insulin deficiency
* Gestational(GDM): increase blood glucose levels during pregnancy

Burn: is a type of injury to skin, or other tissues, caused by heat, cold, electricity, chemicals, friction, or radiation. It can lead to infection they are four types of burn they are:

* Superficial burn or first-degree burns: they appear red without blisters and pain typically lasts around three days.
* Partial thickness or second-degree burns: This is when the injury extends into some of the underling skin layer. Blisters are often frequently present and they are often very painful. Healing can require up to eight weeks and scarring may occur.
* Full-thickness or third-degree burns: injury extends to all layer of the skin. Often they are no pain and burnt area is stiff. Healing typically does not occur on its own.
* Fourth-degree burns: this involves additionally injury to deeper tissues, such as muscle, tendons, or bone. The burn is often black and frequently leads to loss of the burned parts.