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Commentary

Immunodeficiency disorders: An overview

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INTRODUCTION

Immunodeficiencies are inherited (primary) or acquired (secondary) disorders during which components of host immune defenses are either absent or functionally defective. In developed nations, most immunodeficiencies are inherited, which they're generally first seen within the clinic as recurrent or overwhelming infections in newborn children. However, on a worldwide scale, the foremost typical explanation for immunodeficiency is malnutrition which would be categorized as an acquired immunodeficiency. Acquired immunodeficiencies are more likely to develop later in life, and thus the pathogenic mechanisms of the numerous remain obscure.

PRIMARY IMMUNODEFICIENCY

Primary immunodeficiencies, which number quite 250, are caused by inherited imperfections of either nonspecific innate or specific adaptive immune defenses. In general, patients born with Primary Immunodeficiency (PI) commonly have an increased susceptible to infection. This susceptibility can become apparent shortly after birth or in infancy for kind of individuals, whereas other patients develop symptoms later in life. Some primary immunodeficiencies are caused due to a deformity of 1 cellular or humoral component of the immune system; others may result from defects of quite one component. Some of the common examples of primary immunodeficiencies include chronic granulomatous disease, X-linked agammaglobulinemia, selective IgA deficiency, and severe combined immunodeficiency disease.

Chronic granulomatous disease

The causes of Chronic Granulomatous Disease (CGD) are defects within the NADPH oxidase system of phagocytic cells, including neutrophils and macrophages that prevent the production of superoxide radicals in phagolysosomes. The inability to supply superoxide radicals impairs the antibacterial activity of phagocytes. As a result, infections in patients with Chronic Granulomatous Disease (CGD) persist longer, resulting in a chronic local inflammation called a granuloma. Microorganisms that are the foremost common causes of diseases in patients with CGD include *Aspergillus spp., Staphylococcus aureus, Chromobacterium violaceum, Serratia marcescens*, and *Salmonella typhimurium*.

Selective IgA deficiency

The most common inherited form of immunoglobulin inadequacy is selective IgA deficiency, affecting around one of every 800 individuals. People with particular IgA insufficiency produce typical degrees of IgG and IgM, but are not able to produce secretory IgA. IgA deficiency predisposes these people to lung and gastrointestinal infections for which secretory IgA is generally a crucial defense reaction. Infections within the lungs and digestive tract can involve a variety of microorganisms, including *H. influenzae, S. pneumoniae, Moraxella catarrhalis, S. aureus, Giardia lamblia*, or pathogenic strains of *Escherichia coli.*

SECONDARY IMMUNODEFICIENCY

A secondary immunodeficiency occurs as a result an acquired impairment of function of B cells, T cells, or both. Secondary immunodeficiencies can be caused by:

• Systemic problems such as diabetes mellitus, malnutrition, hepatitis, or HIV infection

• Immunosuppressive therapies like cytotoxic chemotherapy, bone marrow ablation before transplantation, or radiation therapy

• Prolonged critical illness because of infection, surgery, or trauma in the very young, elderly, or hospitalized patients.

Unlike primary immunodeficiencies, which have a genetic basis, secondary immunodeficiencies are frequently reversible

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if the underlying cause is resolved. Patients with secondary immunodeficiencies develop an increased susceptibility to an otherwise benign infection by opportunistic microorganisms like *Candida spp., P. jirovecii,* and *Cryptosporidium.*

HIV infection and other related Acquired Immuno Deficiency Syndrome (AIDS) are the well-known secondary immunodeficiencies. AIDS is characterized by profound CD4 T-cell lymphopenia (decrease in lymphocytes). The decrease in CD4 T cells is the results of various types of mechanisms, including HIV-induced pyroptosis (a kind of apoptosis that stimulates an inflammatory response), viral cytopathic effect, and cytotoxicity to HIV-infected cells.

The most well-known reason for secondary immunodeficiency worldwide is severe malnutrition, which affects both innate and adaptive immunity. More research and data are required for the more common causes of secondary immunodeficiency; however, the number of recent discoveries in AIDS research far exceeds that of the other single cause of secondary immunodeficiency. AIDS research has paid off extremely well in terms of discoveries and treatments; expanded research into the foremost common reason behind immunodeficiency, malnutrition, would likely be as beneficial.