Selective IgA Deficiency

Selective IgA deficiency is a congenital B-cell immunodeficiency, which is often asymptomatic, but can result in recurrent minor infections, diarrhea, and anaphylaxis in the most severe of cases. It results from an inability to class switch to IgA, thus limiting the amount of specific IgA that can be produced. Because IgA plays an important role in protecting mucosal surfaces, patients especially experience symptoms in the respiratory and GI tract.

PLAY PICMONIC

Pathophysiology

Defect in Class Switching

Broken Switch

In this disorder, there is defective class switching. The immune system is unable to class switch from non-specific IgM to specific IgA immunoglobulins.

Decreased IgA

Down-arrow Apple Globulin-Goblin

In selective IgA deficiency there is a lack of IgA, due to the inability to class-switch from non-specific IgM. These patients have normal levels of IgM and IgG, however.

Signs and Symptoms

Often Asymptomatic

Thumbs-up

Patients with selective IgA deficiency are often asymptomatic. Most commonly, selective IgA deficiency is not associated with clinically overt signs and symptoms, but rather it is discovered incidentally in patients with chronic lung disease, and in blood donors being screened.

Increase in Atopic & Autoimmune Diseases

Up-arrow A-tophat with Auto-in-moon

Patients with selective IgA deficiency are prone to allergic symptoms, including airway hyperreactivity and atopic dermatitis of the skin due to increased IgE levels.

Sinopulmonary Infections

Sinner-with-lungs and bacteria

IgA is essential for protection of mucosal surfaces. Without sufficient IgA protection, the mucosal surfaces of the respiratory tract are prone to infection. Thus, patients with this disorder develop recurrent sinopulmonary infections.

Increased Risk Giardia Lamblia Infections

Up-arrow Jar Lamb with bacteria

One of the more severe infections that IgA-deficient patients are prone to is giardiasis. Humoral immunity appears to play a significant role in the defense against infections by Giardia lamblia, and more specifically, secretory IgA immunity against these trophozoites when they are located in the intestinal lumen.

Inflammatory Bowel Disease

Bowel-bowl in-flames

While there is not a clear pathophysiologic relationship between selective IgA deficiency and inflammatory bowel disease (IBD), healthcare providers should be aware that there is an increased incidence of IBD in patients with this immunoglobulin deficiency.

Celiac Disease

Silly-yak

There is evidence that patients with selective IgA deficiency are at increased risk for developing celiac disease. Studies show there is approximately an 8% prevalence in selective IgA patients, which is a substantial increase over the 1% prevalence in the general population.

Considerations

Anaphylaxis From IgA Containing Blood Products

Anvil-ax and Blood-bag

Some IgA deficient patients develop anti-IgA antibodies, usually in the form of IgE or IgG. As a result, any blood product transfusion containing IgA, including whole blood, PRBCs, platelets, FFP, cryoprecipitate, granulocytes, and IVIG, can all lead to an immediate anaphylactic reaction. Extreme caution should be exercised when considering a transfusion in these patients, which commonly requires washing blood products prior to administration.