Definition:

Selective IgA Deficiency is the severe reduction or total absence of the IgA class of immunoglobulins in the blood. There are five types (classes) of immunoglobulins or antibodies in the blood: IgG, IgA, IgM, IgD, and IgE. The immunoglobulin class present in the largest amount in blood is IgG, followed by IgM and IgA. IgD and IgE are present in only very small amounts.

Of these immunoglobulin classes, it is primarily IgM and IgG that protect the bloodstream, body tissues, and internal organs from infection. It is also important that the body is protected at surfaces that come in close contact with the environment. These sites are the mucosa surfaces: the mouth and nose, the throat, the airways within the lung, the gastrointestinal tract, the eyes, and the genitalia. The IgA antibodies (which are transported in secretions to mucosal surfaces) play a major role in protecting mucosal surfaces from infection. IgG, IgM and IgE antibodies are also found in secretions at mucosal surfaces, but not in the same amount as the IgA antibody. This is why IgA is known as the secretory antibody. Individuals with Selective IgA Deficiency do not produce IgA but do, however, produce all the other immunoglobulin classes. In addition, the function of their T-lymphocytes, phagocytic cells and complement system are normal or near normal. Hence, this condition is known as "Selective" IgA Deficiency. The cause of Selective IgA Deficiency is unknown.

CLINICAL FEATURES:

Selective IgA Deficiency is the most common primary immunodeficiency disease. Studies have indicated that as many as one in every five hundred people may have Selective IgA Deficiency. The majority of people with Selective IgA Deficiency are relatively healthy and free of symptoms. However, there are also some people who suffer significant illnesses. Currently, it is not understood why some people have almost no illness while others are very sick.

The most common infections seen in patients with Selective IgA Deficiency are recurrent ear infections, sinusitis, bronchitis and pneumonia. This is because IgA normally protects these mucosal surfaces from infections. These infections may become chronic. Furthermore, the infection may not completely clear with treatment, and patients may have to remain on antibiotics for longer than usual.

A second major problem in IgA deficiency is the occurrence of autoimmune diseases. In autoimmune diseases an individual produces antibodies or T-lymphocytes which react with his/her own tissues with resulting damage to these tissues. Some of the more frequent autoimmune diseases associated with IgA deficiency are: Rheumatoid Arthritis, Systemic Lupus Erythematosus and Immune Thrombocytopenic Purpura (ITP). These autoimmune diseases may cause sore and swollen joints of the hands or knees, a rash on the face, anaemia (a low red blood cell count) or thrombocytopenia (a low platelet count). Other kinds of autoimmune disease may affect the endocrine system and/or the gastrointestinal system. Allergies may also be more common among individuals with Selective IgA Deficiency than among the general population. The types of allergies vary. Asthma is one of the common allergic diseases that occurs with Selective IgA Deficiency. It has been suggested that asthma may be more severe, and less responsive to therapy, in individuals with IgA deficiency than it is in normal individuals. Another type of allergy associated with IgA deficiency is food allergy, in which patients have reactions to certain foods. Symptoms associated with food allergies are diarrhoea or abdominal cramping. It is not certain whether there is an increased incidence of allergic rhinitis (hay fever) or eczema in Selective IgA Deficiency. Another unusual, but important form of allergy may also occur in IgA deficiency. In people whose blood contains no IgA, IgA from other individuals may be recognized by the immune system as a foreign protein. Because antibodies are normally made against foreign proteins, some people with Selective IgA Deficiency make an IgG or IgE antibody against IgA. In this situation, if an IgA deficient person who has antibodies against IgA receives a blood product that contains IgA, an allergic reaction may result. Although allergic reactions to IgA are very uncommon, it is important that every patient with Selective IgA Deficiency is aware of the potential risk of transfusion reactions if they receive blood or blood products.

DIAGNOSIS:

The diagnosis of Selective IgA Deficiency is usually suspected because of either chronic or recurrent infections, allergies, autoimmune diseases, or chronic diarrhoea. The diagnosis is established when tests of the patient's blood serum show a marked reduction or near absence of IgA with normal levels of the other major classes of immunoglobulins (IgG and IgM).

An occasional patient may also have IgG2 subclass deficiency and associated antibody deficiency. The numbers and functions of T-lymphocytes are normal.

TREATMENT:

It is not currently possible to replace IgA in IgA deficient patients. Therefore treatment is aimed at the problems associated with Selective IgA Deficiency. Patients with chronic or recurrent infections need appropriate antibiotics. It is not always possible to identify these organisms, however, and the use of broad-spectrum antibiotics may be necessary. Certain patients who have chronic sinusitis or chronic bronchitis may need to stay on long term antibiotic therapy. There are a variety of therapies for the treatment of autoimmune diseases. Anti-inflammatory drugs, such as aspirin or ibuprofen, and steroids may be helpful. Treatment of the allergies associated with IgA deficiency is similar to treatment of allergies in general.

As a matter of precaution, it may also be desirable to test for antibodies against IgA in case the patient may need a blood transfusion in the future. The most important aspect of therapy in IgA deficiency is open communication between the patient, family members and the physician so that problems can be recognized and treated as soon as they arise.

EXPECTATIONS:

Although Selective IgA Deficiency is one of the milder forms of immunodeficiency, it may result in severe disease in some people. Therefore, it is difficult to predict the long-term outcome in a given patient with Selective IgA Deficiency. In general, the prognosis in Selective IgA Deficiency depends on the severity of the associated diseases. It is important for physicians to continually assess and re-evaluate patients for the existence of associated diseases and the development of more extensive immunodeficiency.
For example, in some cases it has been seen that IgA deficiency can progress to become Common Variable Immunodeficiency with its deficiencies of IgG and IgM.

These booklets are designed to offer medical professionals, patients and their families' basic information about these rare disorders of the immune system.
For further information please contact your immunologist, paediatrician, physician or the National IDF Health Coordinator

Other booklets available:

Living with PID
What is IVIG Therapy
Recurrent infections
Common Variable Immune deficiency (CVID)
X-linked Agammaglobulinaemia (XLA)
Chronic Granulomatous disorder (CGD)
Genetic Testing & PID

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The Immune Deficiency Foundation Asia-Pacific Alliance, IDFAPA.

An alliance of not-for -profit PID Patient support groups across the Asia Pacific Region.

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Living with Primary Immune Deficiency Disorders

Selective IgA Deficiency

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