Selective IgA

Selective IgA deficiency (sIgAD)

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IMMUNO DEFICIENCY UK

Supporting families affected by primary and secondary immunodeficiency

About this booklet

This booklet provides information on selective IgA deficiency (sIgAD). It has been produced by the Immunodeficiency UK Medical Advisory Panel and Patient Representative Panel to help answer the questions patients and their families may have about this condition but should not replace advice from a clinical immunologist.

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Summary

Selective immunoglobulin A deficiency (sIgAD) is the most common of the primary antibody deficiencies, affecting about 1 in 600 people. It is not usually associated with bacterial infections but may be associated with autoimmune or allergic diseases. The genetic causes of sIgAD are unclear. People affected have very low or absent levels of one type of antibody, called immunoglobulin A (IgA), but produce protective antibodies of the other types of immunoglobulin (IgG and IgM). This could be why many people with sIgAD appear healthy.

IgA works to protect body surfaces, such as the respiratory tract and gut, that come into contact with outside organisms. So, if someone has infections, these are the most common sites of infection. Such individuals may have a partial IgG deficiency despite apparently normal IgG levels in the blood, and so should be investigated in an immunology clinic.

People with IgA deficiency may have allergies (e.g. allergic asthma) or an increased tendency to develop autoimmune conditions, where the immune system attacks itself, and this may result in conditions such as coeliac disease, which affects about 10 per cent of IgA-deficient patients. No specific treatment is required for sIgAD. Infections should be treated promptly with antibiotics.

How did I get sIgAD?

Clusters of cases can occur within families, so the doctor may want to ask you about your relatives' health. This is referred to as taking a family history. The genetic causes of sIgAD remain unknown. There may be a variety of causes and they may vary from individual to individual.

What are the symptoms of sIgAD?

In general, individuals who have sIgAD fall into one of four different types and can present with different symptoms:

- People who are asymptomatic. This covers the majority of people, in whom a lack of IgA in the blood would not have come to notice had they not had an immunoglobulin test for another reason. Such people are entirely healthy, probably throughout their lives, because compensatory mechanisms make up for the lack of IgA. They are referred to as being asymptomatic and do not require follow-up.
- People who have infections and IgA deficiency. These are individuals in whom sIgAD is a marker of other, more subtle, irregularities of their immune system, but having sIgAD is unlikely to be the cause. If associated with an IgG subclass or specific antibody deficiency, individuals may experience recurrent infections, often affecting the sinuses or ears.
- People affected by allergies.
- People who have health complications associated with autoimmunity because their immune system makes antibodies against their own tissues and organs. An example of autoimmunity is coeliac disease, a condition particularly associated with sIgAD.

Very few cases progress to common variable immune deficiency (CVID), and IgA deficiency may be present in those who have CVID in the family.

Most people with sIgAD do not have any symptoms. Others may have frequent infections of the ears, sinuses, lungs and gut, but IgA deficiency alone is not usually the cause.

These patients may have an additional partial IgG deficiency – known as an IgG subclass deficiency – resulting in failure to make some (but not all) protective antibodies; IgG subclasses and protective antibodies can be measured to assess this.

How is sIgAD diagnosed?

A clinical immunologist usually makes the diagnosis of sIgAD.

Making the diagnosis

To diagnose sIgAD the specialist immunologist may carry out blood tests to:

- Measure the levels of the different immunoglobulins: IgA, IgG and IgG subclasses, and IgM
- Test for the presence of antibodies to previous immunisations or infections. If antibodies are not present in the blood, you will be immunised and blood taken three to four weeks later to see if you have responded to the vaccines
- Count the numbers of T- and B-cells in the blood – these should be normal in sIgAD.

How will my sIgAD be treated?

Most people with sIgAD do not require treatment or regular immunology review.

Infections should be treated promptly with antibiotics, especially sinusitis, chest infections or diarrhoea. In rare cases, longer courses of antibiotics may be needed to prevent infections from coming back.

As in people who have healthy immune systems, recurrent sinusitis in sIgAD patients can be made worse by poor drainage of the sinuses. Obtaining the advice of an ear, nose and throat (ENT) specialist is often helpful to prevent further infections. Steroid nasal sprays may help to control ENT symptoms.

People with sIgAD who also do not respond to vaccination might benefit from immunoglobulin replacement therapy. This is done on a trial basis for one year in order to assess the clinical benefit in the patient as there is no published data available to advise on this treatment otherwise.

Iron deficiency anaemia is common if there have been gastrointestinal complications. In these cases iron tablets, or a gluten-free diet if coeliac disease is diagnosed, may be helpful.

Are there any associated health problems with sIgAD and how will my health be monitored?

Your clinical immunologist will be on the look out for the complications and will work with other clinical specialists to offer you the most appropriate advice and treatments.

Autoimmunity

Some people affected by sIgAD may develop antibodies directed against their own body tissues. These are known as autoantibodies and they can cause symptoms. A typical disorder is coeliac disease, which affects the small intestine and is associated with sensitivity or intolerance to gluten found in wheat, barley and rye.

Allergy and anaphylactic shock

People with IgA deficiency may develop antibodies to IgA. This is only a problem if they are documented to have reacted to blood products containing IgA. No specific caution or testing is needed if a patient hasn't been transfused before: they should be treated as someone without IgA deficiency for a first transfusion or if they have been transfused without problem.

Immunisation

Most vaccines are safe to be administered to patients with sIgAD, provided that other tests of immune function are normal. The infant and childhood vaccination schedule can be followed as normal. Vaccinations required in adulthood can be discussed with your clinical immunology team.

Glossary of terms

allergy – an exaggerated sensitivity resulting from a heightened or altered reactivity of the immune system to an external substance.

anaphylactic shock – a widespread and life-threatening allergic reaction.

antibody – a type of protein (immunoglobulin) that is produced by certain types of white blood cells (plasma cells – a type of B-cell). The role of antibodies is to fight bacteria, viruses, toxins and other substances foreign to the body.

asymptomatic – disease is considered asymptomatic if a patient is a carrier for a disease or infection but experiences no symptoms.

autoimmune/autoimmunity – when an individual's immune system attacks the body's own tissues or vessels.

B-cell – a type of white blood cell (lymphocyte) that produces antibodies.

coeliac disease – a disease that is caused by the immune system reacting adversely to gluten, a product found in wheat, barley and rye. It causes inflammation of the bowel and leads to diarrhoea and/or malabsorption. The condition is reversible by avoiding gluten.

common variable immune deficiency (CVID) – a primary antibody deficiency. People with CVID have either no immunoglobulins in the blood or low levels and require immunoglobulin replacement therapy on a regular basis.

deficiency – a lack or shortage.

gastrointestinal – this refers to the lining of body parts that run from the mouth to the bottom. It can also be referred to as the gut.

genetic – something that relates to how your genes work and are inherited.

IgG subclass deficiency/immunoglobulin G subclass deficiency – a reduction in one or more of the IgG subclasses but with normal total circulating IgG with or without normal levels of IgA and IgM.

immune system – the structures and processes that protect the body against infection and disease.

immunoglobulin replacement therapy – a plasma-based treatment. The immunoglobulin contains antibodies that help fight infection. This treatment can be given through a vein or through the skin.

immunoglobulins – proteins (globulins) in the body that act as antibodies. They work to fight off infections. They are produced by specialist white blood cells (plasma cells/B-cells) and are present in blood serum and other body fluids. There are several different types (IgA, IgE, IgG and IgM), and these have different functions.

iron deficiency anaemia – a condition where a lack of iron in the body leads to anaemia.

lymph nodes – small bean-sized organs of the immune system distributed widely throughout the body. They are the home for the many types of cells that are important in fighting infections.

lymphocyte – a white blood cell that works to fight infection in the body. One type of lymphocyte is called a 'B-cell'. This type of lymphocyte makes antibodies.

plasma – the liquid component of blood without the cells (but with all the proteins).

plasma cell – a specific subtype of B-cell that is found within the bone marrow or lymph nodes. Plasma cells are responsible for the majority of high-quality antibody production.

primary antibody deficiency – covers a range of disorders resulting from the failure of the immune system to produce sufficient antibodies in the bloodstream to fight infections.

protein – one of the basic building blocks of life. Proteins make up the structure and determine the function of the cells that make up all the tissues of our bodies.

respiratory tract – the airway passage involved in breathing that leads from the mouth/nose to the lungs.

sinuses – air-filled space within the bones of the face and around the nose. Infection of the sinuses is called sinusitis.

steroid – sometimes known as a corticosteroid. A type of medicine used to reduce inflammation and which affects the way the immune system works.

T-cell – a type of white blood cell (lymphocyte) that helps the immune system work properly to fight infection.

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About Immunodeficiency UK

Immunodeficiency UK is a national organisation supporting individuals and families affected by primary and secondary immunodeficiency.

We are the UK national member of IPOPI, an association of national patient organisations dedicated to improving awareness, access to early diagnosis and optimal treatments for PID patients worldwide.

Our website has useful information on a range of conditions and topics, and explains the work we do to ensure the voice of patients with primary and secondary immunodeficiency is heard. If we can be of any help, please email us or call on the number above, where you can leave a message.

Support us by becoming a member of Immunodeficiency UK. It's free and easy to do via our website. Members get monthly bulletins.

Immunodeficiency UK is reliant on voluntary donations. To make a donation, please go to **www.immunodeficiencyuk.org/donate**

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