

Selective IgA deficiency (sIgAD)

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PID UK

Supporting families affected
by primary immunodeficiencies

About this booklet

This booklet provides information on selective IgA deficiency (sIgAD). It has been produced by the PID 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 that come into contact with outside organisms, such as the respiratory tract and gut, 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 people in whom 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 as a result of the deficiency. These are individuals in whom sIgAD is a marker of other, more subtle, irregularities of their immune system. If associated with an IgG subclass 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.

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.

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.

About Primary Immunodeficiency UK

**Primary Immunodeficiency UK
(PID UK) is a national organisation
supporting individuals and families affected
by primary immunodeficiencies (PIDs).**

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

Our website at www.piduk.org provides useful information on a range of conditions and topics, and explains the work we do to ensure the voice of PID patients is heard.

If we can be of any help, please contact us at hello@piduk.org or on **0800 987 8986** where you can leave a message.

Support us by becoming a member of PID UK. It's free and easy to do via our website at www.piduk.org/register or just get in touch with us. Members get monthly bulletins and newsletters twice a year.

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