

# Recurrent AOM associated with selective IgA deficiency

## Abstract

Selective IgA deficiency is the most common primary immunodeficiency. A small percentage presents pathology, but at older age can associate deficiency of some subclass of IgG and greater susceptibility to infections, allergic diseases, autoimmune diseases and neoplasms.

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## Dr. Silvia Muriño

ENT specialist, Residence in Hospital Italiano of CABA, Argentina

**Correspondence:** Dr. Silvia Muriño, ENT specialist, Residence in Hospital Italiano of CABA, Certified Allergologist received at AAIBA, specialist title by U.B.A. Head of the ENT Service of H. P. Piñero, Argentina, Email [silviemurino@gmail.com](mailto:silviemurino@gmail.com)

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## Goals

To describe the association of DS IgA with middle ear infections in a pediatric population.

## Introduction

The deficit of IgA in the first years of life may be transient, and it is not definitively diagnosed until 4 years of age. Selective IgA deficiency is described for the first time in 1961 in patients with ataxia-telangiectasia. According to the consensus of the Pan-American Immunodeficiency Group (PAGID) and the European Society of Immunodeficiencies (ESID), selective IgA deficiency is defined as a situation in which blood IgA levels are lower than 7mg/dl (0.07g/L) with normal levels of IgG and IgM and an intact function of T cells in a patient older than 4 years. It was estimated until recently that up to 90% of selective IgA deficits remain asymptomatic; however, a long-term follow-up in blood donors with IgA deficiency showed that 80% develop symptoms throughout their lives.

## Anamnesis

**Age:** the older infant and the preschool can have a large number of annual infections, mainly upper respiratory tract, being usual 7 or 8, and decreasing from 5-6 years.

**Environmental factors:** The concurrence to daycare or having older siblings is associated with recurrent exposure to germs (especially viruses) will cause the child to present infections more frequently. Passive smoking increases respiratory morbidity, breastfeeding would protect above all from gastrointestinal infections.

Family history (recurrent or atypical infections, early childhood deaths, auto immunity or cancer) may give diagnostic clues at times.

## Physical exam

Upper respiratory tract infections and acute otitis media are common in healthy children. If the AOM exceed 6 to 8 a year or persist, so also maxillary sinus infections; look for local causes or rule out other processes (cystic fibrosis, immunodeficiencies of antibodies, ciliary dyskinesia) or local malformations. Persistent/recurrent airway processes should attract attention. Also take into account: intractable diarrhea, poor absorption, G. Lamblia, Cryptosporidium, which make us think of the presence of an ID. Any skin lesion can be impetiginized

by scratching. This is common in atopic dermatitis, with over-infection being a frequent cause of disease malaise; In turn, severe, atypical and/or over infected dermatitis can occur in ID syndromes.

## Physical exploration

Adequate nutritional status largely rules out relevant pathology, the absence of lymphoid tissue suggests immunodeficiencies, certain features point to specific diagnoses. After the initial evaluation with anamnesis and exploration, the child will be described as: 1 - certain infections (upper tract, otitis media, recurrent tonsillitis, acute gastroenteritis, nonspecific chronic diarrhea, "fast" intestinal rhythm associated with breastfeeding, eczema and impetigo of normal course, mild asthma/wheezing), common germs. Patients maintain a good general state between episodes and somatic development is globally adequate. Healthy child attending a normal event of his age.

## 2-Potentially pathological

Discard underlying respiratory pathology in the presence of persistent rhinorrhea/sinusitis, pneumonias (2 in a year or more than 2, without limit), persistent infiltrates, severe asthma or easily over infected. They also force to deepen the presence of recurrent or chronic diarrhea with repercussion pondoestatural, the presence of serious infections (especially if they recur in the same or different location) or by opportunists, suspicious family history. In addition, any combination of the above, associated phenotypic findings, as well as the absence of lymphoid tissue are suspect data.

## What are primary immunodeficiencies?

They are congenital diseases in which the immune effector mechanisms are diminished, so that the elimination capacity of microorganisms is reduced and the susceptibility to infections is increased. These effector mechanisms are also anomalous, which is why in these patients there is also increased susceptibility to allergic, autoimmune and oncological processes.

## When to suspect primary immunodeficiencies?

In the presence of recurrent infections, such as OMAR and pneumonia, by non-habitual or opportunistic germs,

Poor therapeutic response, Pondoestatural delay/prolonged diarrhea, Chronic diarrhea/poor absorption, Abscesses (recurrent

or internal skin), Recurrent fever, Late fall of cord, Adenopathies/absence of lymphoid tissue, Peculiar phenotypic traits. In our country, 200,000 cases of OMAR are calculated per year in children under 7 years of age.

## Discussion

Between 5 and 10% of all children suffer 4 or more episodes of otitis media per year. Before age 6, 80% of children have had at least one AOM and 50% have had 3 before 3 years. Usually the problem is temporary and is chronic only in a minority of cases in which complications such as mastoiditis can appear. Of all the primary immunodeficiency, those that affect the function of B cells are the most frequent. The selective deficiency of serum and secretory IgA is the most frequent defect; the incidents described range from 1: 223 to 1: 1000 cases in the general population. It may be partial or total. In general, the other immunoglobulins are normal but in some cases they can be associated with deficiency of IgG and its subclasses. It can be asymptomatic. When it is symptomatic, patients present recurrent infections and a poor evolution of the respiratory and gastrointestinal tracts, the severity of which depends on serum IgA levels. It is associated with a higher frequency of autoimmune diseases, malignant and allergic diseases and recurrent parotitis symptoms have also been described. In the pathogenesis of immunodeficiencies, genetic and environmental factors are associated. The laboratory is pathognomonic in revealing low or absent levels of IgA with normality of the other immunoglobulins. Secretory IgA may also be absent or traces may be detected with saliva being the most easily accessible source. It is also important to remember that the serum level does not always match the antigenic response capacity. The respiratory pathogens that prevail in these immunodeficiencies are: St. Pneumoniae, H. Influenzae and Mycoplasma. The polysaccharide portion of St. Pneumoniae was found in 55% of the material extracted from the OMAR of children and it is believed that this capsular presence predisposes to chronic otorrhea. A history of AOM associated with chronic otorrhea is frequently observed in this type of immunodeficiency.<sup>1-12</sup>

Gastrointestinal infections can be caused by whole virus and G. Lamblia. The treatment will depend on the clinical pictures that affect the patient since for now there is no specific therapy. The use of gamma globulin is not suggested for several reasons: the amount of IgA that it contains is very small and it does not serve as a substitute, anaphylactic reactions can be generated and finally, as IgG could provide anti-IgA antibodies, this would accentuate immunodeficiency. One of the recently established therapies is the administration of oral

immunostimulants, of the type of bacterial lysates, which in many cases manage to raise the level of IgA.

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## Conflicts of interest

The authors declare no conflicts of interest.

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