

Selective IgA deficiency

This article has been translated from WikiSkripta; ready for the **editor's review**.

Selective IgA deficiency (OMIM: 137100 (<https://omim.org/entry/137100>)) is a genetic antibody immunodeficiency.

Etiology and pathogenesis

This is one of the **most common** pathological laboratory findings, which is classified as an immunodeficiency, but no disease may be present clinically. The cause of the IgA defect, as with conventional variable immunodeficiency (CVID), is not exactly known. As in the case of CVID, the genetic component is probably represented by a mutation in the section for III. HLA class (6p21.3). Rarely, mutations directly in the IgA heavy chain gene (IGHA1 , localization 14q32.33 or IGHA2, localization 14q32.33) may be the cause. The resulting defect already occurs at the level of stem cells. The disease may have an autosomal recessive inheritance type, but even here there may be a significant influence of external factors on the onset of the disease. Accurate resolution of CVID and selective IgA deficiency can sometimes be difficult.

Clinical picture

IgA defect may not be associated with **any clinical symptoms** and individuals with this laboratory finding are usually healthy. In the case of clinical symptomatology, the IgA defect is associated with a higher incidence of **autoimmune diseases, allergies** or a greater tendency to develop cancer.

Reduction of serum IgA below 0.5 g / l is often accompanied by the absence of secretory IgA. Manifestations then include various **recurrent infections of the upper and lower respiratory tract**. The most common incidence of respiratory infections is in childhood, followed by allergic manifestations. In adulthood, IgA deficiency is common in systemic autoimmunities. Secretory IgA in mucosal immunity can to some extent be replaced by secretory IgM rarely IgG, therefore the laboratory finding may not always be fully clinically manifested. Individuals in whom we detect a deficiency or a significant reduction in IgA in laboratory tests require increased health checks.

Links

Related Articles

- IgA
- Bruton's agammaglobulinemia
- Primary immunodeficiency

Source

- ws:Selektivní deficit IgA
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References

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