IgM hyperimmunoglobulinemia syndrome

IgM hyperimmunoglobulinemia syndrome is a heterogeneous group of several types of antibody primary immunodeficiencies , of which **type 1** is the best known and most common.

IgM hyperimmunoglobulinemia syndrome, type 1

This type (Hyper-IgM immunodeficiency X-linked, HIGM1, XHIM, OMIM: 308230) represents an X-linked immunodeficiency, caused by a mutation in the gene for the antigenic ligand **CD40** (CD40LG, localization Xq26). This ligand is crucial for the interaction of T and B-lymphocytes before the initiation of the isotype switch from IgM production to other classes of immunoglobulins . Studies have shown that T-lymphocytes are defective, as B-lymphocytes of carriers (heterozygous women) are properly stimulated even with a small amount of non-defective T-lymphocytes.

Clinincal picture

Manifestation includes **high levels of IgM** and the absence of other immunoglobulin classes (IgG, IgA). In addition to the antibody deficiency, there is **also a cellular deficiency** - due to defective T-lymphocytes and autoimmune-induced neutropenia (tendency to develop autoimmune diseases). Manifestations include susceptibility to various bacterial and viral infections, including opportunistic infections (for example, diarrhea caused by *Cryptosporidium parvum* is typical).

Other types

Other types of IgM hyperimmunoglobulinemia syndrome are mentioned on a separate page .

Links

Related Articles

- Antibodies
- Primary immunodeficiency
- Burton's agammaglobulinemia

Source

- ŠIPEK, Antonín. *Genetically determined disorders of the immune system* [online]. Last revision 9/6/2006, [cit. 5/12/2009]. < http://www.genetika-biologie.cz/primarni-immunodeficiency >.

References

- BARTŮŇKOVÁ, Jiřina. *Immunodeficiency.* 1st edition. Prague: Grada, 2002. 228 pp. ISBN 80-247-0244-4 .

Category: Immunology Category: Genetics Category: Pediatrics