IgG subclass deficiency with or without IgA deficiency

GENERAL INFORMATION

Description:

Patients who suffer recurrent infections because they lack, or have very low levels of, one or two IgG subclasses (IgG1, IgG2, IgG3 and IgG4), but whose other immunoglobulin levels are normal, are said to have a "selective IgG subclass deficiency".

Alternative names:

- IgG subclass deficiency

Classification:

- Deficiencies predominantly affecting antibody production
  - Selective deficiency of IgG subclass, IgE and/or IgA class or subclass

Inheritance:

OMIM:

Cross references:

Phenotype related immunodeficiencies:

- IDR factfile for γ1 isotype deficiency
- IDR factfile for α1 isotype deficiency

Incidence:

Incidence is not known.

CLINICAL INFORMATION

Description:

Most patients with IgA and/or IgG subclass deficiency are asymptomatic but some may suffer from frequent mainly respiratory infections. Recurrent ear infections, sinusitis, bronchitis and pneumonia are the most frequently observed illnesses in patients with IgG subclass deficiencies. Both males and females may be affected. Some patients will show an increased frequency of infection beginning in their second year of life; in other patients the onset of infections may occur later. In general, the infections suffered by patients with selective IgG subclass deficiencies are not as severe as those suffered by patients who have marked deficiencies of IgG, IgA and IgM(for example X-linked agammaglobulinemia and common variable immunodeficiency). Occasionally, subclass deficient patients have suffered recurrent episodes of meningitis or bacterial infections of the bloodstream (e.g. sepsis).

Diagnosis:

Diagnostic laboratories:

Clinical:

- Hypogammaglobulinemia, eMedicine

Therapeutic options:

- Immunoglobulin G deficiency, eMedicine
- Hypogammaglobulinemia, eMedicine
- IgA and IgG subclass deficiencies, eMedicine

Research programs, clinical trials:

- Improved Healthcare for Patients with Primary Antibody Deficiencies through new Strategies Elucidating their Pathophysiology (IMPAD), IMPAD
- European Initiative for Primary Immunodeficiencies
- Immune Regulation in Patients with Common Variable Immunodeficiency and Related Syndromes, ClinicalTrials.gov

GENE INFORMATION

Names:

HUGO name:

Localization:

Chromosomal Location:

Other gene-based resources:

PROTEIN INFORMATION

Description:

Other features:

Expression pattern for human:

OTHER RESOURCES

Societies:

General:

- International Patient Organization for Primary Immunodeficiencies
- Immune Deficiency Foundation
- March of Dimes Birth Defects Foundation
- NIH/National Institute of Allergy and Infectious Diseases
- European Society for Immunodeficiencies