Transient hypogammaglobulinemia of infancy

GENERAL INFORMATION

Description:

Transient hypogammaglobulinemia of infancy (THI) results from a delay in the maturation of immunoglobulin production.This disorder, affecting both males and females, presents at 5-6 months of age and can last as long as 3 years. The infant does not synthesize IgG, although the levels of IgM (e.g. the natural isohemagglutinins) and IgA are normal.

Alternative names:

- THI
- Transient hypogammaglobulinemia of infancy (THI)

Classification:

- Deficiencies predominantly affecting antibody production
  - Other antibody deficiencies

Inheritance:

Unknown

OMIM:

- %240500 Common variable immunodeficiency

Cross references:

Phenotype related immunodeficiencies:

- IDR factfile for common variable immunodeficiency

Incidence:

It is not known.

CLINICAL INFORMATION

Description:

Disease has an early onset. Patients present bacterial infections occuring after 6 months of age. It may last up to 36 months. Disease is characterised by recurrent respiratory infections and otitis media, bronchitis and/or bronchial asthma and recurrent gastroenteritis. During the first years of lives there is a high incidence of recurrent upper respiratory infections but usually not pneumonias or life-threatening infections. The presence of normal B-cell numbers differentiates THI from XLA. IgM is frequently normal.

Diagnosis:

Diagnostic laboratories:

Clinical:

- Common Variable Immunodeficiency (CVID), eMedicine
- Common Variable Immunodeficiency (CVID), eMedicine

Therapeutic options:

- (Intravenous)immunoglobulins may be required and should always be used for a fixed period and be withdrawn to check for spontaneous recovery. All patients recover after treatment. If there is no recovery, than the patients has CVID.
- Transient hypogammaglobulinemia of infancy, eMedicine

Research programs, clinical trials:

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

GENE INFORMATION

Names:

HUGO name:

Localization:

Maps:

(Map View)

Other gene-based resources:

Ensembl: , GENATLAS: , GeneCard: , UniGene: , LocusLink: , euGenes: , GDB:

PROTEIN INFORMATION

Description:

Other features:

Expression pattern for human:

Tissue	Exp. (%)	Clones

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