Opis choroby *

Definicja

Hemophagocytic syndrome (HPS) is a rare immune disease (see this term) and a potentially life-threatening disorder characterized by cytokine storm and overwhelming inflammation causing fever, hepatosplenomegaly, cytopenia, hypertriglyceridemia, hyperferritinemia, and hemophagocytosis in bone marrow, liver, spleen or lymph nodes. It can be either primary due to a genetic defect (primary hemophagocytic lymphohistiocytosis ; see this term), or secondary to malignancies, to infections, most commonly with viruses such as Epstein-Barr virus or cytomegalovirus, human immunodeficiency virus, or to autoimmune disorders such as systemic lupus erythematosus or adult-onset Still disease (secondary hemophagocytic lymphohistiocytosis) (see these termes).

Dane

Klasyfikacja
Kategoria

Synonimy
HLH
HLH
Limfohistiocytoza hemofagocytarna
Hemophagocytic lymphohistiocytosis

Kod ORPHA
158032

Kod OMIM
-

Kod ICD10
-

Kod ICD11
4A01.23

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*Źródło

orphanet