Opis choroby *

Definicja

A rare hemophagocytic syndrome characterized by excessive activation and proliferation of macrophages and T cells occurring in the context of a variety of diseases, including infections, neoplasms, rheumatic disorders, and leading to sudden onset of persistent fever, lymphadenopathy, and hepatosplenomegaly. Complications include profound depression of one or more blood cell lines with coagulopathy and pancytopenia, and impaired liver and renal function. Bone marrow examination reveals numerous well differentiated macrophages actively phagocytosing hematopoietic elements.

Dane

Klasyfikacja
Zespół kliniczny

Kod ORPHA
158061

Kod OMIM
-

Kod ICD10
D76.1

Kod ICD11
-

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*Źródło

orphanet