Opis choroby *

Definicja

A rare and very aggressive neoplastic disease emerging after a primary acute or chronic active EBV infection. It presents with persisting fever and malaise, hepatosplenomegaly with or without lymphadenopathy, liver failure, severe pancytopenia and a rapid progression towards multi-organ failure and hemophagocytic syndrome with a fatal issue. It is characterized by clonal proliferation of EBV-infected T cells with an activated cytotoxic phenotype.

Dane

Klasyfikacja
Choroba

Synonimy
Systemic EBV+ T-cell LPD of childhood
Układowa choroba limfoproliferacyjna z komórek T EBV pozytywna wieku dziecięcego
Układowa LPD z komórek T EBV+ wieku dziecięcego
Systemic EBV-positive T-cell lymphoproliferative disease of childhood

Kod ORPHA
364033

Kod OMIM
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Kod ICD10
D47.9

Kod ICD11
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*Źródło

orphanet