

Opis choroby *

Definicja

A rare autosomal recessive primary immunodeficiency characterized by susceptibility to Epstein-Barr virus (EBV)-related disorders (B-cell lymphoproliferative disorders including Hodgkin lymphoma) as well as dysgammaglobulinemia and recurrent infections. Patients can present with recurrent fever, lymphadenopathy, hepatosplenomegaly, Behçet-like stomatitis, pharyngitis, tonsillitis, adenitis, and viral encephalitis.

Dane

Klasyfikacja

Choroba

Synonimy

CID due to CD70 deficiency

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Kod ORPHA

538958

Kod OMIM

618261

Kod ICD10

D82.3

Kod ICD11

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*Źródło

orphanet