

## 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