

## Opis choroby \*

### Definicja

A rare primary immunodeficiency characterized by infantile onset of generalized lymphadenopathy, splenomegaly, and lymphocytosis, with excessive polyclonal expansion of B-cells. Patients present recurrent infections and impaired T-cell and antibody responses, while overt autoimmune manifestations are usually absent. Occurrence of B-cell malignancy later in life has been reported.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

B-cell expansion with NF-kB and T-cell anergy disease

B-cell expansion with NF-kB and T-cell anergy disease

#### Kod ORPHA

464336

#### Kod OMIM

616452

#### Kod ICD10

D81.8

#### Kod ICD11

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#### \*Źródło

orphanet