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