Opis choroby *

Definicja

A rare, genetic, lymphoproliferative syndrome characterized by early onset recurrent infections, lymphadenopathy with hepatosplenomegaly and variable autoimmune disorders, including hemolytic anemia, thrombocytopenia, neutropenia, enteropathy, type I diabetes, scleroderma, arthritis, atopic dermatitis, and inflammatory lung disease. Patients commonly have failure to thrive. Variable immunologic findings include decreased regulatory T-cells, hypogammaglobulinemia, and reduction in memory B cells.

Dane

Klasyfikacja
Choroba

Kod ORPHA
438159

Kod OMIM
615952

Kod ICD10
M35.8

Kod ICD11
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*Źródło

orphanet