Opis choroby *

Definicja

A rare, primary combined T and B cell immunodeficiency characterized by early-onset of recurrent, invasive viral and bacterial infections associated with T and B cell lymphopenia, functional defects in T and B cells, poor antibody response and thrombocytopenia. Depending on the type of infectious agent, variable clinical manifestations commonly include recurrent pneumonia, bronchiolitis, otitis media, meningoencephalitis, colitis, and diarrhea, leading to fatal multiorgan failure in severe cases.

Dane

Klasyfikacja

Choroba

Kod ORPHA

447737

Kod OMIM 616433

Kod ICD10

D81.8

Kod ICD11 4A01.1Y

*Źródło

orphanet