

Opis choroby *

Definicja

A rare, slowly progressive, chronic leukemia characterized by presence of abnormal B-lymphocytes (medium sized with abundant irregular pale cytoplasm, hair-like cytoplasmic projections/ruffled cytoplasmic border, a round or bean-shaped nucleus and absent nucleoli) in the blood or bone marrow, spleen and peripheral blood pancytopenia, notable monocytopenia, and marked susceptibility to infection. The characteristic immunophenotype is CD11c+, CD25+, CD103+ and CD123+ with a BRAF mutation in most cases.

Dane

Klasyfikacja

Choroba

Synonimy

HCL-C

HCL

Leukemic reticuloendotheliosis

Leukemic reticuloendotheliosis

Kod ORPHA

58017

Kod OMIM

-

Kod ICD10

C91.4

Kod ICD11

2A82.2

*Źródło

orphanet