

Opis choroby *

Definicja

A rare mature T-cell neoplasm characterized by proliferation of small to medium-sized prolymphocytes with a mature post-thymic T-cell phenotype, involving the peripheral blood, bone marrow, lymph nodes, liver, spleen, and sometimes the skin. T-cell receptor genes are clonally rearranged. Patients typically present with hepatosplenomegaly, generalized lymphadenopathy, high leukocyte count with normal serum immunoglobulins, anemia, and thrombocytopenia. HTLV-1 serology is negative. The disease course is aggressive with generally poor prognosis.

Dane

Klasyfikacja

Choroba

Synonimy

T-PLL

Przewlekła białaczka limfocytarna z komórek T

T-pIL

T-cell chronic lymphocytic leukemia

Kod ORPHA

86871

Kod OMIM

-

Kod ICD10

C91.6

Kod ICD11

2A90.0

*Źródło

orphanet