Opis choroby *

Definicja

A rare myeloproliferative neoplasm characterized by a clonal proliferation of eosinophilic precursors with persistent increase of eosinophils in peripheral blood and bone marrow, accompanied by increased blasts (<20%) or clonal cytogenetic or molecular genetic abnormalities. Cases with BCR-ABL1, PCM1-JAK2, ETV6-JAK2, or BCR-JAK2 fusion, or rearrangement of PDGFRA, PDGFRB, or FGFR1, are not included in this entity. Infiltration of the liver and spleen, as well as a variety of other organs, is typical. Patients may present with constitutional symptoms and signs and symptoms of organ involvement, such as endomyocardial fibrosis, peripheral neuropathy, central nervous system manifestations, respiratory symptoms, or rheumatological findings. Acute transformation is common.

Dane

Klasyfikacja

Choroba

Kod ORPHA

168940

Kod OMIM

Kod ICD10

D47.5

Kod ICD11

2A20.3

*Źródło

orphanet