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
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Kod ICD10
D47.5

Kod ICD11
2A20.3

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*Źródło

orphanet