Opis choroby *

Definicja

A rare myeloproliferative neoplasm characterized by sustained peripheral blood neutrophilia, bone marrow hypercellularity due to neutrophilic granulocyte proliferation, and hepatosplenomegaly. Other organs may be infiltrated in addition. Microscopically, the bone marrow shows an increase in proportion of myelocytes and mature neutrophils, but no significant dysplasia in any of the cell lineages. Peripheral blood neutrophils are mostly segmented, although band forms may also be substantially increased. Cytogenetic abnormalities are absent in most cases. The disease is slowly progressive with progredient neutrophilia followed by anemia and thrombocytopenia. Transformation to acute myeloid leukemia may occur.

Dane

Klasyfikacja
Choroba

Kod ORPHA
86829

Kod OMIM
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Kod ICD10
D47.1

Kod ICD11
2A20.1

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*Źródło

orphanet