Opis choroby *

Definicja

A rare myelodysplastic syndrome characterized by macrocytic anemia (with or without other cytopenias and/or thrombocytosis), and with del(5q) occurring either in isolation, or with one other cytogenetic abnormality, other than monosomy 7 or del(7q). The bone marrow is typically hypercellular with erythroid hypoplasia and increased numbers of megakaryocytes, which show non-lobated and hypolobated nuclei. Myeloblasts constitute less than 5% of the nucleated bone marrow cells and less than 1% of the peripheral blood leukocytes. Auer rods are absent. Ring sideroblasts may be observed. Patients present with anemia and often thrombocytosis, while thrombocytopenia or pancytopenia are uncommon. Transformation to acute myeloid leukemia may occur in a small number of patients.

Dane

Klasyfikacja
Choroba

Synonimy
5q- syndrome
Zespół 5q

Kod ORPHA
86841

Kod OMIM
153550

Kod ICD10
D46.7

Kod ICD11
2A36

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*Źródło

orphanet