Appropriate management of polycythaemia vera with cytoreductive drug therapy: European LeukemiaNet 2021 recommendations

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Summary

Polycythaemia vera is associated with a reduced quality of life, a high rate of vascular events, and an intrinsic risk of disease evolution. The results of several randomised trials for the treatment of this disorder are now available, and both a new ropegylated formulation of interferon alfa-2b (ropeginterferon alfa-2b; 2018) and ruxolitinib (2015) have been approved in Europe. European LeukemiaNet (ELN) investigators have therefore deemed it appropriate to provide recommendations for the use of these drugs in clinical practice. An expert panel of 14 senior haematologists from ELN centres that had actively participated in previous ELN projects or relevant randomised trials, chaired by a member of the ELN Steering Committee, developed a list of clinical questions, and a methodologist established three patient, intervention, comparator, outcome (PICO) questions and systematically reviewed the evidence. Recommendations were approved by six Delphi consensus rounds and two virtual meetings (on Jan 26, 2021, and June 24, 2021). The expert panel recommended that patients with polycythaemia vera who are younger than 60 years and have not had previous thrombotic events should start cytoreductive drug therapy if at least one of the following criteria are fulfilled: strictly defined intolerance to phlebotomy, symptomatic progressive splenomegaly, persistent leukocytosis (>15 × 10^9 white blood cells per L), progressive leukocytosis (at least 100% increase if baseline count is <10 × 10^9 cells per L or at least 50% increase if baseline count is >10 × 10^9 cells per L), extreme thrombocytosis (>1500 × 10^9 platelets per L), inadequate haematocrit control requiring phlebotomies, persistently high cardiovascular risk, and persistently high symptom burden. Recombinant interferon alfa, either in the form of ropeginterferon alfa-2b or pegylated interferon alfa-2a, is the recommended cytoreductive treatment for these patients. The expert panel suggested that either interferon alfa or ruxolitinib should be considered for patients who are being treated with hydroxyurea but require a therapy change.