

Jerry L. Spivak, MD, MACP

Professor Emeritus-Medicine
Johns Hopkins University School of Medicine
Director
Center for the Chronic Myeloproliferative Disorders
Baltimore, Maryland

How is risk assessed in PV and how does this risk inform treatment decisions ?

Current recommendations for polycythemia vera (PV) advise that risk stratification should be based on age and thrombosis history.¹ Patients under age 60 years are recommended to receive phlebotomy and aspirin, while those age 60 or older should receive hydroxyurea for cytoreduction and prevention of thrombosis. However, the evidence supporting these recommendations is weak, and fails to acknowledge 1) the impact of age on survival, which is independent of disease, 2) the very high risk of thrombosis in older adults, and 3) important differences in hematocrit phlebotomy targets for men versus women.² Also, most such recommendations are based on retrospective data or data obtained from a mixture of treated and treatment-naïve patients.

More recently, a genomic-based myeloproliferative neoplasm (MPN) prognostic calculator was developed.³ This calculator defines eight genomic subgroups derived from 63 clinical and genomic variables; each of these subgroups is associated with a unique distinct clinical phenotype, including blood counts, risk of leukemic transformation, and event-free survival. This calculator was able to accurately predict patient outcomes and may support the provision of personalized treatment of patients with an MPN, but this has not been validated clinically and may not reflect the behavior of indolent PV or essential thrombocythemia (ET).

References

1. Tefferi A, Vannucchi AM, Barbui T. Polycythemia vera treatment algorithm 2018. *Blood Cancer J.* 2018;8(1):3.
2. Spivak JL. How I treat polycythemia vera. *Blood.* 2019;134(4):341-352.
3. Grinfeld J, Nangalia J, Baxter EJ, et al. Classification and personalized prognosis in myeloproliferative neoplasms. *N Engl J Med.* 2018;379(15):1416-1430.

For more information on risk-adapted therapy in PV, please view the full newsletter by clicking [here](#).

This activity is supported by educational grants from Bristol-Myers Squibb and Incyte Corporation.