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Perspective

Management and Treatment of Polycythemia Vera

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ABOUT THE STUDY

The World Health Organization differentiates Polycythemia Vera (PV) from other Myeloproliferative Neoplasms (MPNs) mainly on the basis of erythrocytosis and activating mutations in Janus Kinase 2 (*JAK2*). Updated criteria now also include marrow trilineage myeloproliferation as determined by bone marrow biopsy and lower haemoglobin thresholds for erythrocytosis. Comparing patients with PV to others of same age and sex in the general population, they endure distressing symptoms and are at a higher risk of dying. There are an estimated 100,000 patients with PV in the US. The most frequent consequences and the main reasons for illness-related death are thromboembolic and hemorrhagic events, with disease transition into Myelofibrosis (MF) or Acute Myeloid Leukaemia (AML) being a less frequent cause of patient mortality.

Patients with PV frequently feel weariness, attention issues, itching, inactivity, as well as symptoms connected to splenomegaly (e.g., early satiety and abdominal discomfort or pain). *JAK2* activating mutations are present in almost all PV patients, most frequently *JAK2V617F* in exon 14 and less frequently *JAK2* exon 12 or other variants. The cause of the symptoms of PV disease, such as high hematocrit and blood counts, pruritus, splenomegaly, thrombotic risk, risk of fibrotic transformation, and systemic inflammation, is constitutively active *JAK2* signalling.

Patients with PV are frequently managed by community-based oncologists, and the best care involves current expertise in management techniques, treatment recommendations, and licenced medicines. Beyond the "textbook" PV patient, community-based healthcare providers must deal with difficult situations like patients with poor access to hematologic care, a lack of tools for evaluating PV-related symptoms, hydroxyurea resistance or intolerance, pregnancy, elective surgery, and concurrent immunosuppressant therapy. The usage of conventional treatments for PV patients, clinical trial data supporting the use of ruxolitinib in PV, disease management problems (particularly those specific to community practitioners), and potential therapeutic choices in the future are all outlined in this study.

PV management

The risk of thromboembolic events serves as the main guiding factor in the care of PV patients. Traditional risk factors for PV patients include age 60 years, thrombosis history, and hematocrit level 45%. White blood cell count $11*10^9$/L, female sex, molecular indicators such mutant JAK2 allele load, and standard cardiovascular risk factors are additional non-conventional risk factors that need further validation (i.e., diabetes mellitus, current smoking status, elevated cholesterol, and high systolic blood pressure). These several risk factors taken together imply that current risk classification approaches that only consider age and thrombosis history may not be able to identify all individuals at high risk. No matter their risk level, all patients get low-dose aspirin therapy and phlebotomy to maintain an iron shortage that keeps their haemoglobin levels below 45%. Additionally, some professionals support hematocrit levels in women of less than 42% because this would be more in line with the typical physiologic range. Additionally, cytoreductive therapy has been shown to provide additional clinical benefits for select patients, particularly those indicated by the aforementioned risk factors. According to European Leukemia Net (ELN) guidelines, hydroxyurea is the most often used cytoreductive medication for PV patients and is advised as first-line treatment. However, about 25% of patients will develop resistance or intolerance. Clinical benefits from traditional interferon (IFN-) therapy include decreased pruritus and normalised blood levels for certain individuals. However, IFN-associated toxicity, particularly at high doses, and the inconvenience of an injectable medication have a detrimental impact on some patients' long-term tolerability and adherence. Phase 3 clinical development of IFN-PEG variants with enhanced safety and tolerability profiles is in under research. After first-line therapy options have failed, busulfan or pipobroman may be considered, but because of their leukemogenic propensity, they are only appropriate for patients with a short life expectancy. The US Food and Drug Administration (FDA) has not yet authorized the use of hydroxyurea, IFN-, or busulfan in patients with PV.

It is still unknown whether these conventional therapies significantly enhance PV-related symptoms or Quality of Life

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(QoL). In a retrospective review of PV patients (n=538), hydroxyurea, IFN-, aspirin, and/or phlebotomy were not linked to a substantial reduction in the intensity of the symptoms as described by the patients. Additionally, in a prospective analysis, PV patients (n=1,334) who received phlebotomy and/or hydroxyurea continued to have more severe symptoms than individuals who did not get treatment. When taken as a whole, these data highlight the fact that some PV patients still have health issues which requires proper treatment.

CONCLUSION

Patients with PV endure a wide range of symptoms that affect their Quality of Life (QoL) and have a higher mortality risk than the general population. Phlebotomy, aspirin, and hydroxyurea cytoreduction all have positive clinical effects, but one in four patients develop hydroxyurea intolerance or resistance. The FDA has approved the *JAK1/JAK2* inhibitor ruxolitinib for the treatment of PV patients who did not respond well to hydroxyurea or who could not tolerate it. Ruxolitinib therapy lowers hematocrit, shrinks the spleen, normalises blood levels, and enhances symptoms and quality of life associated with PV. Many additional potential therapy alternatives are also being researched and developed.