Polycythemia vera (PV) is a rare progressive myeloproliferative cancer with significant symptom burden. Patients with PV often experience symptoms that adversely affect quality of life, work productivity, and functional status. Oncology nurses are well suited to assess for symptom burden and to provide educational interventions that support patients and their families.

AT A GLANCE

- PV is characterized by elevated blood counts and symptom burden, which often influences quality of life.
- Oncology nurses can assess patients with PV for symptom burden while monitoring for thrombotic and vascular events.
- Oncology nurses can support patients with PV and their families by providing ongoing education.

KEYWORDS
myeloproliferative neoplasms; thrombosis; symptom burden; polycythemia vera

DIGITAL OBJECT IDENTIFIER
10.1188/20.CJON.575-578

Polycythemia Vera

Symptom burden, oncology nurse considerations, and patient education

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Polycythemia vera (PV) is a rare hematologic cancer characterized by elevated blood counts. The driver mutation for most patients is the *Janus kinase 2* (*JAK2*) gene, which causes the bone marrow to overproduce blood products, such as red blood cells (Arber et al., 2016). Some patients may have elevated white blood cells and platelets. PV is one of three myeloproliferative neoplasms (MPNs), including essential thrombocythemia and myelofibrosis, which share a tendency for clotting and bleeding that increases patients' health risks (Arber et al., 2016). Patients with essential thrombocythemia have an overproduction of platelets, and patients with myelofibrosis suffer from anemia, bone marrow fibrosis, splenomegaly, and elevated white blood cells (Tefferi, 2018; Tefferi & Barbui, 2019; Tefferi et al., 2017). It is essential that oncology nurses have sufficient knowledge of MPNs to assess risk factors and disease progression and to provide appropriate educational support and interventions as they assist patients toward wellness.

Diagnosis and Prognosis

Hematologists diagnose PV using the World Health Organization (WHO) classification system. A patient must meet all three major criteria or the first two major criteria and the minor criterion (Arber et al., 2016). The first major criterion is (a) hemoglobin greater than 16.5 g/dl in men or greater than 16 g/dl in women, (b) hematocrit greater than 49% in men or greater than 48% in women, or (c) increased red blood cell mass. The second major criterion is having bone marrow results that include tri-lineage proliferation with pleomorphic mature megakaryocytes. The third major criterion is the presence of a *JAK2* mutation. The minor criterion is a subnormal serum erythropoietin level (Arber et al., 2016; Tefferi & Barbui, 2019).

It is not unusual for abnormal laboratory values, vascular complications, and thromboembolic events to occur before diagnosis of PV (Enblom et al., 2015). Because PV may progress slowly, nurses need to report abnormally elevated hemoglobin, hematocrit, and platelet levels, even if the patient appears asymptomatic, because complications may be avoided with early diagnosis and treatment.

Although PV is a progressive disease, it presents differently across populations. Most adults are diagnosed after age 60 years, so prognosis may be influenced by comorbidity and age-related health changes. Prognosis is also influenced by thrombotic and vascular events (Kamiunten et al., 2018). Survival estimates are 15 years, and less if the patient progresses to myelofibrosis (Szuber et al., 2019).

Treatment

Treatment priorities for PV are to prevent or reduce risks for thromboembolic events; therefore, nurses must prioritize the management of risk factors. The National Comprehensive Cancer Network's (NCCN's) clinical guidelines for patients with MPNs provide consistent and standardized care for patients with PV. All patients are monitored for