FAST FACTS

Myeloproliferative Neoplasms

ESSENTIAL THROMBOCYTHEMIA[1]

Brief Overview

- Hyperproliferation of platelets in non-reactive bone marrow
- Can lead to thrombotic or hemorrhagic complications
- Most often presents with an incidental lab finding

Incidence and Mortality

- Clinicians diagnose approximately 6000 new cases/year
- Most often diagnosed in sixth decade of life; median survival 20 years (33 years for younger patients)
- 5% risk of leukemic transformation in 20 years

Signs and Symptoms

- Microcirculatory disturbances
- Migraines
- Thrombosis
- Bleeding

Diagnostic Criteria (World Health Organization [WHO] 2016)

All four major criteria **OR** first three major criteria and the minor criterion (improve standardization of bone marrow morphology and distinguishing between prefibrotic primary myelofibrosis [PMF] and masked polycythemia vera [PV]):

- **Major Criteria**
  - Platelets >450,000
  - Bone marrow with megakaryocytic proliferation with large and mature morphology. No significant left shift of neutrophil granulopoiesis or erythropoiesis and very rarely minor (grade 1) increase in reticulin fibers.
  - Not meeting WHO criteria for chronic myeloid leukemia, PV, PMF, myelodysplastic syndrome, or other myeloid neoplasm
  - *JAK2*, *CALR*, or *MPL* mutation
- **Minor Criterion:** Presence of a clonal marker (e.g., abnormal karyotype), or absence of evidence of reactive thrombosis

Treatment Goals

- Reduce risk of thrombotic or hemorrhagic complications
  - Control symptoms

1. Barbui T, et al. *Am J Hematol.* 2016;91:430-3; Barbui T, el al. *Blood Rev.* 2016;30:453-9.

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POLYCYTHEMIA VERA[2]

Brief Overview

- Hyperproliferation of all three cell lines
- Primary distinguishing feature: erythrocytosis

Incidence and Mortality

- Typically diagnosed in sixth decade, median survival 14 years (24 years in younger patients)
- Risk of transformation to myelofibrosis 7%–22% depending on presence of marrow fibrosis at diagnosis
- Leukemic transformation <10% at 20 years

Signs and Symptoms

- Impaired oxygen delivery
- Headaches
- Thrombosis
- Pruritus
- Splenomegaly

Diagnostic Criteria (World Health Organization 2016)

Requires three major **OR** the first two major criteria and one minor criterion:

- **Major Criteria**
  - Hemoglobin (Hgb) >16.5 or hematocrit (Hct) >49% (men) Hgb >16 or Hct >48% (women) or increased red cell mass
  - Bone marrow with age-adjusted hypercellularity and trilineage myeloproliferation with pleomorphic, mature megakaryocytes
  - Presence of *JAK2* mutation
- **Minor Criterion**
  - Subnormal erythropoietin level

Treatment Goals

Reduce incidence of thrombotic events by controlling:

- Cellular hyperproliferation
- Hct <45%
- Symptoms

2. Barbui T, et al. *Am J Hematol.* 2016;91:430-3; Barbui T, el al. Blood Rev. 2016;30:453-9; Bryan JC, et al. *Cancer Chemother Pharmacol.* 2016;77:1125-42.

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FAST FACTS

Myeloproliferative Neoplasms

PRIMARY MYELOFIBROSIS[3]

Brief Overview

Chronic myeloid neoplasm marked by:

- Progressive bone marrow fibrosis
- Ineffective erythropoiesis
- Excessive production of dysplastic megakaryocytes
- Extramedullary hematopoiesis
- Systemic inflammation with excess circulating levels of proinflammatory cytokines
- Shortened survival

Incidence and Mortality

- Typically diagnosed in sixth decade
- 1.5 cases per 100,000 patients in the United States annually
- Survival depends on disease characteristics (based on International Prognostic Scoring System):
  - High risk = approximately 2 years
  - Low risk = approximately 11 years

Signs and Symptoms

- Abdominal discomfort due to splenomegaly
- Cachexia
- Early satiety
- Bone pain
- Night sweats
- Pruritus
- Dyspnea
- Insomnia
- Fatigue

Diagnostic Criteria (World Health Organization [WHO] 2016)

Requires all three major criteria **AND** at least one minor criterion:

- **Major Criteria**
  - Megakaryocyte proliferation and atypia accompanied by either reticulin and/or collagen fibrosis (grade 2 or 3)
  - Not meeting WHO criteria for chronic myeloid leukemia, polycythemia vera, myelodysplastic syndrome, or other myeloid neoplasm
  - Presence of *JAK2*, *CALR*, or *MPL* mutation, or in the absence of another clonal marker or absence of evidence of reactive bone marrow fibrosis
- **Minor Criteria**
  - Anemia not attributed to comorbid condition
  - Palpable splenomegaly
  - Leukocytosis (≥11 x 10^9/L)
  - Elevated LDH
  - Leukoerythroblastosis

Treatment Goals

- Reduce symptoms and improve quality of life
- Ongoing research into reducing risk for leukemic transformation and improvement in overall survival

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3. Bryan JC, et al. *Cancer Chemother Pharmacol.* 2016;77:1125-42; MPN Research Foundation, Prevalence, http://www.mpnresearchfoundation.org/Prevalence.

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