World Health Organization (WHO) Diagnostic Criteria
Primary Myelofibrosis (PMF)/Polycythemia Vera (PV)/Essential Thrombocythemia (ET)

Check off the major and minor criteria of the patient's clinical presentation and compare the results in each column to the WHO guidelines for diagnosing each myeloproliferative neoplasm (MPN)

***JAK2* mutation is common to PMF, PV, and ET**

Primary Myelofibrosis (PMF) Criteria	Major	Minor
Proliferation and atypia of megakaryocytes accompanied by reticulin and/or collagen fibrosis grades 2 or 3 (on a 0 to 3 scale)		
Not meeting BCR-ABL1+ CML, ET, MDS, PV, or other myeloid neoplasm WHO criteria		
Presence of CALR, JAK2, or MPL mutation (or presence of another clonal marker* in the absence of these mutations) or absence of reactive myelofibrosis†		
Anemia not attributed to a comorbid condition		
Leukocytosis ≥11 x 10^9/L		
Palpable splenomegaly		
LDH increased to above upper normal limit of institutional reference range		
Leucoerythroblastic		
TOTAL		
Diagnosis requires meeting all three major criteria and at least one minor criterion confirmed in two consecutive determinations		

*If any of the three major clonal mutations are absent, searching for the most frequent accompanying mutations (eg, ASXL1, EZH2, TET2, IDH1/IDH2, SRSF2, SF3B1) is helpful in determining the clonal nature of the disease.
† Bone marrow fibrosis secondary to infection, autoimmune disorders (or other chronic inflammatory conditions), hairy cell leukemia (or other lymphoid neoplasm), metastatic malignancy, or chronic toxic myelopathies.

Polycythemia Vera (PV) Criteria	Major	Minor
Hemoglobin >16.5 g/dL in men, >16.0 g/dL in women, *or* Hematocrit >49% in men, >48% in women, *or* Increased red cell mass >25% above mean normal predicted value		
Bone marrow (BM) biopsy showing hypercellularity for age with trilineage growth (panmyelosis), including prominent erythroid, granulocytic, and megakaryocytic proliferation with pleomorphic, mature megakaryocytes (differences in size)		
Presence of JAK2V617F or JAK2 exon 12 mutation		
Subnormal serum erythropoietin level		
TOTAL		
Diagnosis requires meeting either all three major criteria or the first two major criteria and the minor criterion*		

*BM biopsy may not be required in cases with sustained absolute erythrocytosis: Hb levels >18.5 g/dL in men (Hct 55.5 %) or >16.5 g/dL in women (Hct 49.5%) if mutation criterion 3 and the minor criterion are present. However, initial MF (present in up to 20% of patients) can only be detected by performing a BM biopsy; hypercellularity may predict a more rapid progression to overt MF (post-PV MF).

World Health Organization (WHO) Diagnostic Criteria

Primary Myelofibrosis (PMF)/Polycythemia Vera (PV)/Essential Thrombocythemia (ET)

Essential Thrombocythemia (ET) Criteria	Major	Minor
Platelet count ≥450 x 10^9/L		
BM biopsy showing proliferation mainly of the megakaryocyte lineage w/increased numbers of enlarged, mature megakaryocytes with hyperlobulated nuclei; no significant increase or left-shift in neutrophil granulopoiesis or erythropoiesis and very rarely minor (grade 1) increase in reticulin fibers		
Not meeting WHO criteria for BCR-ABL1+ CML, PV, PMF, MDS, or other myeloid neoplasms		
Presence of JAK2, CALR, or MPL mutation		
Presence of a clonal marker or absence of evidence for reactive thrombocytosis		
TOTAL		
Diagnosis requires meeting all four major criteria or the first three major criteria and the minor criterion		

CALR=calreticulin; CML=chronic myelogenous leukemia; JAK=Janus-associated kinase; LDH=lactate dehydrogenase; MPL=myeloproliferative leukemia virus oncogene; BM=bone marrow; Hb=hemoglobin; Hct=hematocrit; JAK=Janus-associated kinase

REFERENCE:

Arber DA, Orazi A, Hasserjian R, et al. The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia. *Blood*. 2016;127(20):2391-2405