

## Revised WHO criteria for primary myelofibrosis

### Major criteria

1. Presence of megakaryocyte proliferation and atypia,\* usually accompanied by either reticulin and/or collagen fibrosis, or, in the absence of significant reticulin fibrosis, the megakaryocyte changes must be accompanied by an increased bone marrow cellularity characterized by granulocytic proliferation and often decreased erythropoiesis (ie, prefibrotic cellular-phase disease)
2. Not meeting WHO criteria for PV, CML, MDS, or other myeloid neoplasm\*\*
3. Demonstration of JAK2617V>F or other clonal marker (eg, MPL515W>L/K), or in the absence of a clonal marker, no evidence of bone marrow fibrosis due to underlying inflammatory or other neoplastic diseases\*\*\*

### Minor criteria

1. Leukoerythroblastosis
2. Increase in serum lactate dehydrogenase level\*\*\*\*
3. Anemia\*\*\*\*|
4. Palpable splenomegaly\*\*\*\*

**Diagnosis requires meeting all 3 major criteria and 2 minor criteria.**

\* Small to large megakaryocytes with an aberrant nuclear/cytoplasmic ratio and hyperchromatic, bulbous, or irregularly folded nuclei and dense clustering.

\*\*Requires the failure of iron replacement therapy to increase hemoglobin level to the polycythemia vera range in the presence of decreased serum ferritin. Exclusion of polycythemia vera is based on hemoglobin and hematocrit levels. Red cell mass measurement is not required. Requires the absence of *BCR-ABL*. Requires the absence of dyserythropoiesis and dysgranulopoiesis.

\*\*\*Secondary to infection, autoimmune disorder or other chronic inflammatory condition, hairy cell leukemia or other lymphoid neoplasm, metastatic malignancy, or toxic (chronic) myelopathies. It should be noted that patients with conditions associated with reactive myelofibrosis are not immune to primary myelofibrosis and the diagnosis should be considered in such cases if other criteria are met.

\*\*\*\*Degree of abnormality could be borderline or marked.