

Polycythemia vera (PV) and essential thrombocythemia (ET) progress to myelofibrosis (MF) at a rate of 10% and less than 4% over 10 years, respectively.² Careful monitoring of these patients can facilitate early identification of disease progression of post-PV MF and post-ET MF.

Mark the patient's major and minor clinical presentation criteria, add the number of boxes checked in each column and compare them against the totals required to meet IWG-MRT guidelines for diagnosis of post-PV MF and post-ET MF.

			POST-PV MF		POST-ET MF	
Source		Criteria	Major	Minor	Major	Minor
Patient history		Prior diagnosis of WHO-defined PV				
		Prior diagnosis of WHO-defined ET				
Clinical exam		Increased splenomegaly [*]				
		Developing one or more of the following:				
		greater than 10% weight loss in six months,				
		night sweats, or unexplained fever				
Bone marrow biopsy		Bone marrow fibrosis ⁺				
Other Lab Results	Complete blood	Anemia [§]				
	count					
	Biochemistry	Increased serum LDH [#]				
	Blood film	Leukoerythroblastosis				
	(smear)					
TOTAL NUMBER OF CRITERIA						
To meet IWG-MRT						
diagnostic criteria for		Must have at least $ ightarrow$	2	2	2	2
this MPN $ ightarrow$						
ET=essential thrombocythemia: IWG-MRT=International Working Group for Myeloproliferative Neoplasms						ns

ET=essential thrombocythemia; IWG-MRT=International Working Group for Myeloproliferative Neoplasms Research and Treatment; LDH=lactate dehydrogenase; PV=polycythemia vera; MPN=myeloproliferative neoplasm; WHO World Health Organization

*An increase in palpable splenomegaly of \geq 5 cm (distance of the tip of the spleen from the left costal margin) or the appearance of a newly palpable splenomegaly.

[†]According to the European classification (grade 2/3): diffuse, coarse fiber network with no evidence of collagenization (negative trichrome stain) or diffuse, coarse fiber network with areas of collagenization (positive trichrome stain). According to the standard classification (grade 3/4): diffuse and dense increase in reticulin with extensive intersections, occasionally with focal bundles of collagen and/or focal osteosclerosis or diffuse and dense increase in reticulin with estensive intersections with coarse bundles of collagen, usually associated with significant osteosclerosis.

[§]Below reference range for appropriate age, sex, and altitude of residence. For Post–PV MF, sustained loss of requirement for phlebotomy in the absence of cytoreductive therapy is sufficient. For Post–ET MF, must be accompanied by ≥2 g/dL decrease from baseline hemoglobin level.

[#]Above reference level.



REFERENCES

- Barosi G, Mesa RA, Thiele J, et al. Proposed criteria for the diagnosis of post-polycythemia vera and post-essential thrombocythemia myelofibrosis: a consensus statement from the International Working Group for Myelofibrosis Research and Treatment. *Leukemia*. 2008;22:437-438. Epub 2007 Aug 30.
- 2. Tefferi A. Essential thrombocythemia, polycythemia vera, and myelofibrosis: current management and the prospect of targeted therapy. *Am J Hematol*. 2008:83(6):491-497.