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TRANSLATION OF PVSG/WHO INTO THE CLINICAL, LABORATORY, MOLECULAR AND PATHOLOGICAL (2018 CLMP) DEFINED MYELOPROLIFERATIVE NEOPLASMS CAUSED BY JAK2V617F JAK2EXON12, CALR, MPL AND TPO DRIVER MUTATIONS ARE DISTINCT BLOOD AND COAGULATION DISORDERS: PROGNOSTIC AND THERAPEUTIC IMPLICATIONS TOWARDS 2020 AND BEYOND

The JAK2V617F mutated tri-linear myeloproliferative neoplasms (MPN) include a broad spectrum of clinical laboratory and bone marrow features in essential thrombocythaemia (ET), prodromal polycythaemia vera (PV) and erythrocythemic PV, classical PV and advanced stages of masked PV and PV complicated by splenomegaly and secondary myelofibrosis (MF). Heterozygous JAK2V617F mutated ET is associated with low JAK2 allele and MPN disease burden and normal life expectance. In combined heterozygous and homozygous or homozygous JAK2V617F mutated tri-linear MPN, the JAK2 mutation load increases from less than 50% in prodromal and early stage PV to above 50% up to 100%in classical PV, advanced PV and PV with MF. Bone marrow histology features show various degrees of diagnostic erythrocytic, megakaryocytic and granulocytic (EMG) myeloproliferation in JAK2V617F mutated tri-linear MPN clearly differ from mono-linear megakaryocytic (M) in MPL or dual megakaryocytic granulocytic (MG) myeloproliferation in calreticulin (CALR) mutated thrombocythemia without features of PV. The morphology of clustered large pleomorphic megakaryocytes with hyper lobulated nuclei is similar in JAK2V67F thrombocythemia, prodromal PV and classical PV patients. Mono-linear megakaryocytic (M) myeloproliferation of large to giant megakaryocytes with hyper lobulated staghorn like nuclei is the hallmark of MPL515 mutated normocellular thrombocythaemia. CALR mutated thrombocythaemia usually presents with high platelet count around

1000x109/l and normocellular megakaryocytic (M) proliferation of immature megakaryocytes with cloud-like hyperchromatic nuclei or prefibrotic dual megakaryocytic granulocytic (MG) myeloproliferation followed by various degrees of bone marrow fibrosis. Natural history and life expectancy of MPN patients are related to the response to treatment and the degree of anaemia, splenomegaly, myelofibrosis and constitutional symptoms. The acquisition of epigenetic mutations at increasing age on top of MPN disease burden independently predicts unfavourable outcome in JAK2V617F, MPL515 and CALR mutated MPNs, which mutually exclude each other. Current treatment options in MPN include low dose aspirin in JAK2 and MPL mutated ET, phlebotomy on top of aspirin in PV, pegylated interferon in intermediate stages of PV and CALR and MPL mutated ET followed by hydroxyurea and or ruxolitinib in the hypercellular stages of PV and MF.