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Translation of WHO into improved clinical, laboratory, molecular and pathological (CLMP) criteria for the diagnoses of JAK2 trilinear myeloproliferative neoplasm, MPL thrombocythemia and CALR thrombocythemia and myelofibrosis: From Dameshek to Vainchenker Green, Kralovics and Skoda

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A new set of International Clinical, Laboratory, Molecular and Pathological (CLMP) criteria for myeloproliferative neoplasms (MPN) define the JAK2 V617F trilinear MPNs as a broad continuum of essential thrombocythemia (ET), polycythemia vera (PV), masked PV and post-ET or post-PV myelofibrosis (MF). Normal vs increased erythrocyte counts (5.8x1012/L) on top of bone marrow histology separate JAK2 V617F ET and prodromal PV from early and classical PV are diagnostic for JAK2 trilinear MPN obviating the need of red cell mass measurement. The JAK2 V617F trilinear MPNs, JAK2 exon 12 PV, MPL515 thrombocythemia and calreticulin (CALR) thrombocythemia and MF mutually exclude each other. JAK2 exon 12 PV is a rare benign distinct MPN entity with low risk of MPN disease. MPL515 mutated thrombocythemia is a monolinear megakaryocytic proliferation, whereas CALR mutated thrombocythemia and myelofibrosis result from dual megakaryocytic granulocytic proliferation without PV features. JAK2, MPL and CALR MPN disease burden is best reflected by the degree of anaemia, splenomegaly, mutation allele burden, bone marrow cellularity and myelofibrosis. Bone marrow histology of the JAK2 V617F trilinear MPNs show variable degrees of erythrocytic (E), megakaryocytic (M), and granulocytic (G) myeloproliferation, peripheral cytoses and splenomegaly related to low intermediate and high JAK2 allele burden. MPL515 thrombocythemia displayed predominantly normocellular megakaryocytic (M) proliferation. CALR thrombocythemia intially presents with megakaryocytic (M) followed by dual granulocytic and megakaryocytic (GM) myeloproliferation without features of PV. The morphology of clustered medium to large pleomorphic megakaryocytes are similar in JAK2 V617F normocellular ET, and prodromal PV, classical PV and masked PV. MPL515 thrombocythemia show increased clustered large to giant mature megakaryocytes with hyperlobulated stag-horn-like nuclei. CALR thrombocythemia is featured by dense clustered large immature dysmorphic megakaryocytes with bulky (bulbous) hyperchromatic nuclei, which are never seen in JAK2 V617F JAK2 exon 12 MPN and MPL515 thrombocythemia.

Biography

Jan Jacques Michiels is is the Professor of Nature Medicine and Health, Clinical and Molecular Genetics, Blood and Coagulation Research at the University Hospitals Antwerp, Brussels. He is the Editor of *Journal of Hematology & Thromboembolic Diseases, World Journal of Hematology and Editor in Chief of World Journal of Clinical Cases.*

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