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NOVEL CRITERIA FOR CLINICAL, LABORATORY, MOLECULAR AND PATHOLOGIC (2018 CLMP) DIAGNOSIS AND STAGING OF PREFIBROTIC JAK2, CALR AND MPL MUTATED MYELOPROLIFERATIVE NEOPLASMS

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The broad spectrum of JAK2 V617F mutated trilinear myeloproliferative neoplasms (MPN) include essential thrombocythemia (ET), prodromal polycythemia vera (PV), erythrocythemic PV, classical PV, masked PV and PV complicated by splenomegaly and myelofibrosis (MF). ET heterozygous for the JAK2V617F mutation is associated with low MPN disease burden and normal life expectancy. JAK2V617F mutation load increases from less than 50% in prodromal and early stage PV to above 50% up to 100% in combined heterozygous/homozygous or homozygous JAK2V617F mutated PV, advanced PV and progressive myelofibrosis (MF). Pretreatment bone marrow morphology and cellularity distinguish JAK2V617F mutated trilinear MPN from calreticulin (CALR) and MPL mutated MPN. The morphology of clustered large pleomorphic megakaryocytes with hyperlobulated nuclei are similar in JAK2V67F ET and PV patients. CALR mutated thrombocythemia shows bone marrow characteristics of normocellular megakaryocytic (M) proliferation and subsequent dual megakaryocytic granulocytic (MG) myeloproliferation, first described in the 1990s as primary megakaryocytic granulocytic myeloproliferation (PMGM) without features of PV in blood and bone marrow. MPL515 mutated thrombocythemia is featured by monolinear proliferation of large to giant megakaryocytes with hyperlobulated staghorn like nuclei. Natural history and life expectancy relate to the degree of splenomegaly, myelofibrosis, constitutional symptoms and increased allele burden in JAK2V617F trilinear MPN and MPL515 thrombocythemia but CALR thrombocythemia runs a more favourable course during life-long follow-up. The acquisition of epigenetic mutations at increasing age predicts unfavorable outcome in JAK2, CALR and MPL mutated MPN. Low dose aspirin in ET and phlebotomy on top of aspirin in PV is mandatory to prevent platelet-mediated microvascular circulation disturbances. Pegylated interferon is the first line myeloreductive treatment option in prodromal and early stage JAK2V617F mutated PV and in CALR and MPL mutated thrombocythemia to postpone or obviate the targeted use of hydroxyurea and ruxolitinib as long as possible.

Biography

The first author is a founder of the Goodheart Institute & Foundation in Nature Medicine & Health, Rotterdam, The Netherlands, Freedom of Science and Education, European Free University Network. JJ Michiels is founder of the European Working Group on Myeloproliferative Disorder / Myeloproliferative Neopasms 1998-2018, co-founder of the Central European Vascular Forum (CEVF) and serves as consultant professor in the Bloodcoagulation, Hemostasis Research Laboratory (co-founder VWF-VWD research program) at the department of Hematology University Hospital, Antwerp. JJ Michiels serves as consultant professor in Hematology and Bloodcoagulation, Comenius University, Bratislava, Slovakia; consultant to the Dutch Society of Internal Medicine and Ministery of Public Health; consultant of quality driven Industrial and Pharmaceutical Medicine; as an editor of 2 Medical Journals and as a guest editor on request and by self initiation. Writing assistance was utilised in the preparation of this lecture, it was carried out at the Goodheart Institute and Foundation in Nature Medicine and Health, Rotterdam, The Netherlands. The author has no other relevant affiliations or financial involvement with any organization or entity with a financial interest in or financial conflict with the subject matter or materials discussed in the manuscript apart from those disclosed.

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