

Background: There are controversies regarding on which cell parameter may be more conveniently used as a surrogate criteria of an increased red cell mass (RCM), that is, hemoglobin (HB) level and hematocrit (HCT) value. According to the 2007/2008 WHO classification, HB level should be >18.5 g/dl in men and >16.5 g/dl in women and/or increased HCT. It has been argued that application of these criteria may result in an underdiagnosis of PV by excluding patients with actual RCM mass that is 25% above mean predicted value, but whose HB and HCT levels are below the WHO guidelines. Therefore, recently the WHO revised the criteria and in 2015 made a new proposal lowering the HB/HCT threshold to 16.5 g/dl/49% in men and 16 g/dl/48% in women. **Aims:** We aimed to apply the proposed WHO 2015 criteria in order to determine whether this contribution in the identification of masked PV would compensate the work overload in our laboratory and clinical practise.

Methods: We selected samples of patients from routine analytical test that meet the new proposed WHO criteria from 22/12/2015 to 23/01/2016 and studied the presence of JAK2V617F mutation as mayor diagnosis criteria for PV. JAK2V617F mutation was determined qualitatively by amplification refractory mutation system polymerase chain reaction assay. We collected white blood count, erythrocytes and platelets levels as well. For statistical analysis has been used Shapiro-Wilk test to check the normality of the data of quantitative variables. The statistical program used was R Core Team (2014).

Results: In our study, there were 48 patients: 47 men and 1 woman. The median age of the patients was 48.5 years (range: 17-73). The median of HB was 17.7 g/dL (16.5-18.3) and the median HCT was 51.35% (48.1-55.4%). The number of positive JAK2V617F mutation was zero. The rest of analytical characters were: Erythrocytes (average $5.44 \times 10^6/\mu\text{L}$, 1.36-6.52), leukocytes (average $9.09 \times 10^3/\mu\text{L}$, 3.87-19.8) and Platelets (average $259.02 \times 10^3/\mu\text{L}$, 134-416).

Summary/Conclusions: Among the 48 patients, none of them was positive for JAK2V617F mutation. Admission of new criteria would mean a high increase in the number of patients to be evaluated, implying an enhance human and laboratory resource consumption, with a significant economic impact. Based on our results, we wonder whether these new proposed WHO criteria should be approved.

PB2030

PREVALENCE OF THE JAK2 V617F MUTATION IN SOME COHORTS OF THE CENTRAL SIBERIA (KRASNOYARSK REGION) POPULATION

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Background: Somatic mutation of the JAK2 V617F is associated with the pathogenesis of myeloproliferative neoplasms (MPNs) and it is an important diagnostic marker. However, V617F JAK2 was detected also at 0.2-1% of the adult population, without the MPN when using highly sensitive allelic load test (Xu X, et al., 2007; Nielsen C., et al., 2014). The V617F JAK2 mutation significantly increases the risk of both arterial and venous thrombosis, including cerebral vessels, visceral intestinal veins and especially Budd-Chiari syndrome (Smalberg JH, et al., 2012). These causes of hospitalization may be the first manifestations of MPN.

Aims: Evaluate the frequency of the JAK2 V617F mutation in different cohorts of hospital patients and blood donors.

Methods: Allele-specific RT-PCR was performed to detect of the JAK2 V617F allele load in whole blood samples among the following groups: healthy blood donors, patients who were included in the program of routine inspections, patients who were hospitalized in general hospitals, as well as those who were directed by hematologist with suspected to MPN.

Table 1. JAK2 V617F mutation frequency among cohorts.

Cohort	Total surveyed	JAK2-V617F positive patients, n (%)	JAK2-V617F allele load, (%) (Min-Max)	Age (Min-Max)
Blood donors	1148	0 (0.0)	0.47 (0.07-2.58)	30 (18-87)
In baseline medical examination	1815	17 (1.2)	0.38 (0.05-2.18)	35 (42-86)
From not hematological hospital departments	1280	34 (2.6)	0.34 (0.04-48.8)	57 (16-90)
Directed by hematologist	903	301 (33.3)	32.0 (2.06-97.0)	54.8 (18-100)

Results: The frequency of the JAK2 V617F mutation was maximal when the patients were directed by a hematologist with MPN suspected (Table 1). Minimum prevalence was observed in healthy blood donors. Among patients from non-hematological hospital departments 12% cases (3 of 24 patients) with JAK2-V617F had ischemic stroke. Participation in voluntarily medical examination reveals patients with mutation but who have no any hematological abnormalities in 95% cases were directed by hematologist with suspected to MPN.

Summary/Conclusions: High risk of thrombosis and of the MPN development, as well as the potential risk of transmission of the transformed cell clone to recipients of the bone marrow and blood, raises the issue of screening for JAK2 V617F among some cohorts of patients. An analysis of "benefit - harm", taking into account the effectiveness of preventive measures will be the subject of additional studies.

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CLINICAL FEATURES OF LATENT/MASKED POLYCYTHEMIA VERA (SINGLE CENTER EXPERIENCE)

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Background: Polycythemia vera is a heterogeneous group of diseases. In patients who don't meet the World Health Organization (WHO) criteria for overt polycythemia vera (PV), a diagnosis of latent/masked PV (mPV) can be determined. mPV is characterized by JAK2V617F positive, morphological features of bone marrow for PV according to WHO, but hemoglobin <18.5 g/dL in men and <16.5 g/dL in women.

Aims: The aim of this study was to identify clinical features of mPV as a separate group of PV.

Methods: The study included 81 patients observed in the outpatient department of National Research Center for Hematology from 2014 to 2015, 50 patients with PV and 31 patients with mPV.

Results: Distribution of patients by gender was statistically comparable. Patients with PV was older compared to mPV: median age was 56 and 44. Between the groups of patients with mPV and PV obvious difference in red blood cells ($5.37 \times 10^{12}/\text{L}$ (4.1-6.5 $\times 10^{12}/\text{L}$) vs $6.94 \times 10^{12}/\text{L}$ (5.4-8.8 $\times 10^{12}/\text{L}$)); hemoglobin (14.8 g/dL (10.0-16.7 g/dL) vs 17.8 g/dL (13.6-24.7 g/dL)); hematocrit (45% (30-52%) vs 53% (42-70%)). Median platelet counts higher in the group of patients with mPV compared with PV: the median was $644 \times 10^9/\text{L}$ (179-1978 $\times 10^9/\text{L}$) vs $636 \times 10^9/\text{L}$ (137-2437 $\times 10^9/\text{L}$). Differences of white blood cells was not revealed in the two groups. All the patients were V617F JAK2 positive. Determination of allele burden JAK2V617F performed 29 patients with mPV and 37 patients with PV. JAK2 allele burden was significantly higher in patients with PV compared to mPV: median 14% (3-57%) and 55.5% (24-86%) respectively. Thrombosis revealed in 38% (12 patients) with mPV and in 16% (10 cases) in the PV. It was mainly venous in the case of mPV with a high frequency of splanchnic vein thrombosis. Arterial thrombosis detected only in 4 cases.

Summary/Conclusions: Masked PV is a separate nosological variant of PV.

PB2032

CLINICAL FEATURES AND MOLECULAR MARKERS IN ESSENTIAL THROMBOCYTHEMIA

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Background: Particular molecular marker detection (JAK2 V617F, MPL, CALR) or its absence (triple-negative (TN)) in essential thrombocythemia (ET) can be served as a basis of different biological neoplasm behavior.

Aims: The aim of this study was to investigate interactions (differences) between the presence of each molecular marker, clinical features and course of ET.

Methods: One hundred and fifty ET patients, who had been diagnosed at our institution according to WHO 2008 criteria. The following parameters were assessed: age, gender, complete blood count, clinical symptoms (weakness, headache, dizziness, arthralgia, constitutional symptoms, erythromelalgia, splenomegaly), thrombotic complications and bleeding. Overall survival (OS) in ET patients with different molecular markers were analyzed by Kaplan-Meier method and compared between groups. Log-rank, ANOVA Kruskal-Wallis test and the Chi-square test with Yate's contingency correction were used for statistical analysis.

Results: Median age was 56 years (range 19-82), 110 patients (73%) was female. Median follow-up was 28 months (range 1-168). The following mutations were detected: JAK2V617F (JAK2+) n=115 (76.7%), MPL+ n=1 (0.7%), CALR+ n=14 (9.3%) and triple-negative (TN) molecular status was registered in n=20 (13.3%) patients. Complete blood count mean values (standard deviations) at initial ET diagnosis were: JAK2+: Hb 14.2 (17.6) g/dL, WBC 9.7 (3.7) $\times 10^9/\text{L}$, PLT 831 (273) $\times 10^9/\text{L}$, MPL+: Hb 12.3 g/dL, WBC 7.1 $\times 10^9/\text{L}$, PLT 2079 $\times 10^9/\text{L}$, CALR+: Hb 13.8 (16.7) g/dL, WBC 9.3 (3.9) $\times 10^9/\text{L}$, PLT 1086 (453) $\times 10^9/\text{L}$, TN: Hb 13.6 (15.6) g/dL, WBC 11.4 (4.7) $\times 10^9/\text{L}$, PLT 777 (230) $\times 10^9/\text{L}$. The presence of CALR mutation was associated with significant higher platelets level (p=0.03). The symptoms frequencies according molecular markers groups were as followed: JAK2+: weakness-34.8%, headache/dizziness-20.9%, arthralgia-19.1%, splenomegaly-16.5%, erythromelalgia-8.7%, pruritus-5.2%, constitutional symptoms-4.4%, MPL+: weakness only (100%), CALR+: weakness-21.4%, headache/dizziness-7.1%, splenomegaly-21.4%, erythromel-