

Polycythemia Vera

PROGNOSTIC FACTORS AND MODELS IN POLYCYTHEMIA VERA

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Prognostic Models in Polycythemia Vera

Prognostic Score Definition	Prognostic variables	Risk Category
<i>Thrombotic Risk</i>	Conventional risk Stratification	<ul style="list-style-type: none">• age >60 years• thrombosis history <ul style="list-style-type: none">• low-risk (the absence of both risk factors): Thrombosis incidence 2.5x100 p/y• high-risk (at least 1 risk factor): Thrombosis incidence 5.0-10.9 x100 p/y
<i>Risk of Death</i>	MIPSS-PV	<ul style="list-style-type: none">• Mutation in SRSF2 (3 points)• Age >67 years (2 points)• Leukocytes count $\geq 15 \times 10^9 / L$ (1 point)• Thrombosis history (1 point) <ul style="list-style-type: none">• Low risk (0-1 point): median OS not reached• Intermediate risk (2-3 points): median OS 10.3 yrs (HR 4.6)• High risk (>3 points): median OS 4.6 yrs (HR 24.1)



Potential Novel Prognostic Factors in Polycythemia Vera

Thrombotic Risk

Prognostic variables	Risk Category	Use in clinical practice
<ul style="list-style-type: none"> JAK2 V617F VAF >50% Neutrophil-to-lymphocyte ratio (NLR) values ≥5 Absolute neutrophil count (ANC) ≥22x10⁹/L 	<ul style="list-style-type: none"> High Risk for Venous Thrombosis (HR 3.8) High Risk for Venous Thrombosis (HR 2.1) High Risk for Venous Thrombosis (HR 9.1) 	

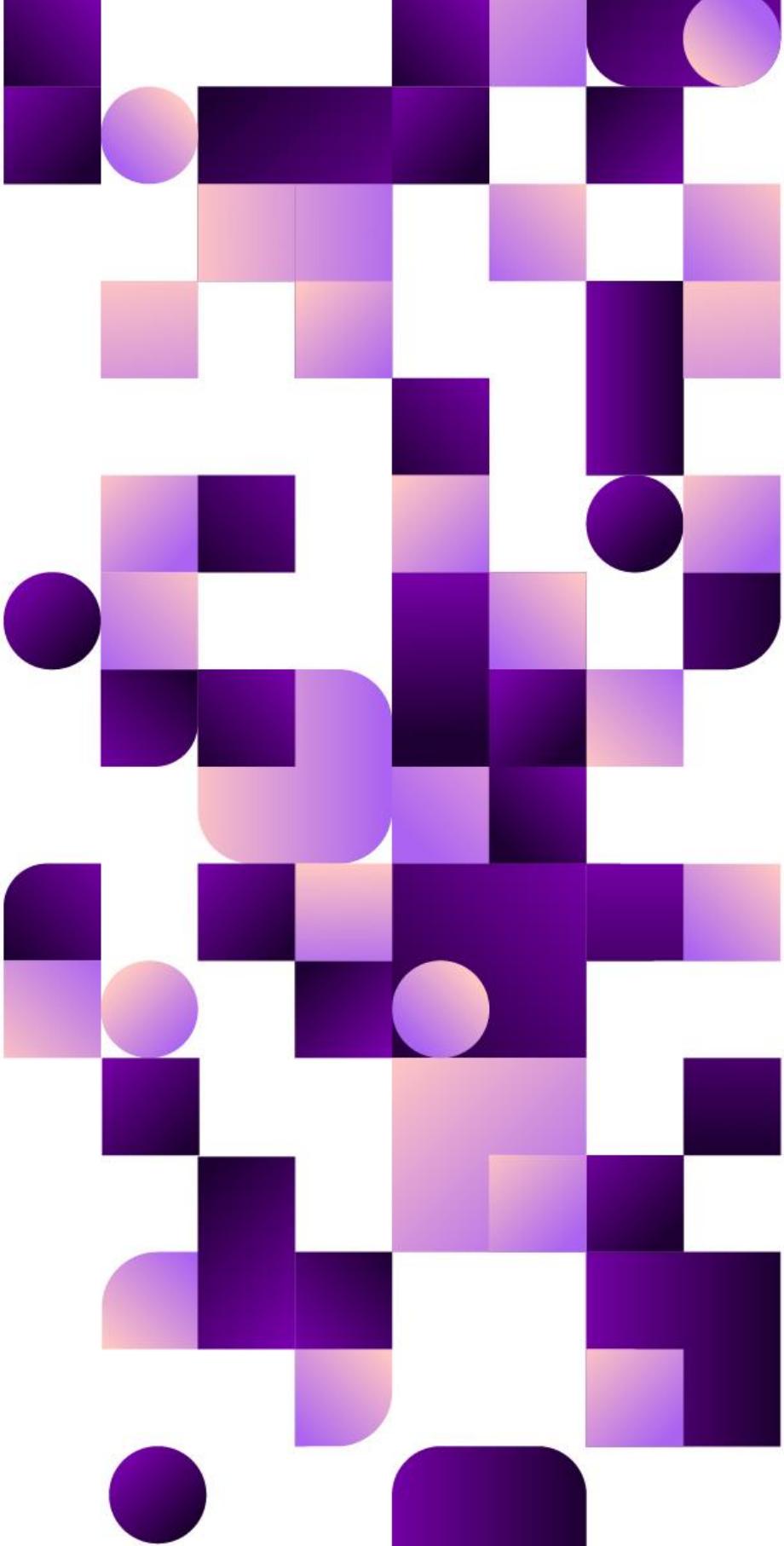
Bleeding Risk

<ul style="list-style-type: none"> Platelet count >1.000 x10⁹/L 	<ul style="list-style-type: none"> promotes the development of an acquired von Willebrand syndrome 	✓
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Disease Progression Risk

<ul style="list-style-type: none"> JAK2 V617F VAF >50% Mutations in ASXL1, SRSF2, IDH2 WBC ≥35x10⁹/L Treatment Exposure: Use of pipobroman or P32/chlorambucil 	<ul style="list-style-type: none"> High Risk for progression in PPV-MF (HR 1.04; a 10% difference in VAF between two samples corresponds to a 40% increase in risk of PPV-MF) High Risk for progression in PPV-MF or blast phase High Risk for Leukemia transformation (HR 24.2) High Risk for Leukemia transformation (HR 4.0) 	
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Il presente documento è il prodotto finale del progetto *Clinical Assessment of resistance and Intolerance to Hydroxyurea as Criteria for Second-line Treatment in patients with Polycythemia Vera*, condotto nel corso del 2023 e 2024 dal Working Party GIMEMA sulle Neoplasie Mieloproliferative Croniche.

EXPERT PANEL

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