# Polycythemia Vera

# SWITCHING BETWEEN THERAPIES IN POLYCYTHEMIA VERA

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# Switching Between Therapies in Polycythemia Vera

## RECOMMENDED SHIFT

**Patients with polycythaemia vera who are receiving hydroxyurea are recommended to change to another cytoreductive drug if they meet at least one of the following criteria:**

### *Inefficacy*

- Development of vascular events: either clinically relevant bleeding, venous thrombosis, or arterial thrombosis (consensus: 80%; strength of the recommendation: weak)

### *Intolerance*

- Intolerance to hydroxyurea because of grade 3–4 or prolonged grade 2 non-haematological toxicity (eg, mucocutaneous manifestations, gastrointestinal symptoms, fever, or pneumonitis) at any dose (consensus: 100%; strength of the recommendation: strong. Note that the expert panel provided a strong recommendation if it was confident that the desirable effects of adherence to the recommendation outweigh the undesirable effects)
- Intolerance to hydroxyurea because of haematological toxicity (haemoglobin <100 g/L, platelet count $<100\times10^9$ cells per L, or neutrophil count $<1\times10^9$ cells per L) at the lowest dose of hydroxyurea to achieve a response[53] (consensus: 100%; strength of the recommendation: strong)
- Development of non-melanoma skin cancers (consensus: 80%; strength of the recommendation: weak)

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## SHIFT to be CONSIDERED

**Patients with polycythaemia who receive hydroxyurea should be considered to change to another cytoreductive drug if they show an insufficient clinical response to hydroxyurea (at ≥1·5 g per day for at least 4 months and without reporting intolerance), as defined by at least one of the following criteria:**

## *Inefficacy*

- Persistent disease-related symptoms: a total symptom score of at least 20 or an itching score of at least ten for at least 6 months (consensus: 92%; strength of the recommendation: strong)
- Persistent thrombocytosis: a platelet count >1000 × $10^9$ cells per L, microvascular symptoms, or both, persisting for more than 3 months (consensus: 92%; strength of the recommendation: weak)
- Symptomatic or progressive splenomegaly: increased in spleen size by more than 5 cm from the left costal margin in 1 year (consensus: 83%; strength of the recommendation: weak)
- Progressive (at least 100% increase if baseline count is <10 × $10^9$ cells per L or at least 50% increase if baseline count is >10 × $10^9$ cells per L) and persistent leukocytosis (leukocyte count >15 × $10^9$ cells per L confirmed at 3 months; consensus: 75%; strength of the recommendation: weak)
- Insufficient haematocrit control: need for six or more phlebotomies per year to keep haematocrit below 45% (consensus: 83%; strength of the recommendation: weak)

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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