Allegato 4 : Terapia PV

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Obiettivi della terapia: prevenire trombosi ed emorragie, minimizzare l'evoluzione in MF o LA; controllare i sintomi della malattia.
I pazienti con PV vengono classificati in due categorie di rischio: alto rischio e basso rischio per eventi trombotici. (Barbui T et al. *Blood*. 2014; 124:3021-3023).
Per tutte le categorie di rischio si consiglia di monitorare e correggere i fattori di rischio cardiovascolare (fumo, diabete, dislipidemia, ipertensione, peso-BMI), monitorare la comparsa di trombosi/emorragie.

Non è raccomandata la supplementazione marziale neppure in carenza documentata TRANNE che in gravidanza; da valutare in terapia con ruxolitinib.

CRITERI DI RISPOSTA AL TRATTAMENTO

Figura 1. Criteri ELN di risposta alla terapia della PV (Barosi et al, *Blood* (2013) 121 (23): 4778–4781)

Response Grade	Response in PV
Complete response	1. Durable** resolution of disease-related signs* including palpable hepatosplenomegaly, large symptoms improvement **and** 2. Durable** peripheral blood count remission, defined as Hct lower than 45% without phlebotomies; Platelet count ≤ 400 < 10^9/L; WBC < 10x 10^9/L **and** 3. Without progressive disease, and absence of any hemorrhagic or thrombotic event **and** 4. Bone marrow histological remission defined as the presence of age-adjusted normocellularity and disappearance of trilinear hyperplasia, **and** absence of grade 1 reticulin fibrosis
Partial response	1. Durable* resolution of disease-related signs including palpable hepatosplenomegaly, largemsymptoms improvement **and** 2. Durable* peripheral blood count remission, defined as Ht lower than 45% without phlebotomies; platelet count <400 x10^9/L, WBC count < 10x10^9/L, **and** 3. Without progressive disease, and absence of any hemorrhagic or thrombotic event, **and** 4. Without bone marrow histological remission defined as persistence of trilinear hyperplasia
No response	Any response that does not satisfy partial response
Progressive disease	Transformation into post-PV myelofibrosis, myelodysplastic syndrome or acute leukemia

*"Disease-related signs" includono disturbi del microcircolo, prurito e cefalea
** Lasting at least 12 weeks

Figura 2. Criteri ELN di **resistenza/intolleranza** alla terapia con HU nella PV (Barosi G et al. *Br J Haematol*. 2010; 148:961-963)

Resistance	Need for phlebotomy (Hct < 45%) Platelets >400 x 10^9/L and WBC >10 x 10^9/L No reduction of spleen by 50% No reduction of spleen symptoms	⇨	After >3 months at MTD or 2 g/day
Intolerance	Cytopenias (any) - ANC < 1.0 x 10^9/L - Hb < 10 g/DL - Plt < 100 x 10^9/L Leg ulcers GI toxicity Fevers Mucocutaneous toxicity Skin cancers	⇨	At lowest dose to achieve either a PR or CR by ELN criteria