Results: A total of 224 patients, with median age at diagnosis 64.5 years (range 17-92) were included in the analysis. The initial diagnosis was CML (13%), ET (51%), PV (28%), PMF (5%) and myeloproliferative/myelodysplastic syndrom in 2%. Mean duration of follow-up was 8 years. In 20% of patients the presenting symptom was a thrombotic episode (arterial, venous or splenic infarct). During the follow-up, 83 events of interest were recorded. In the whole cohort, the incidence rate (in events per 100 patients per 10 years) of thrombosis was 13.42, hemorrhage 1.68, transformation to another hematologic malignancy 11.74, occurrence of solid tumor 5.59 and other events 13.98. Other events for which patients received medical attention included atypical chest pain, joint pain, arrythmias, pleural effusion and infections. Incidence of major events per diagnosis are presented in table 1. Cumulative incidence of thrombotic complications is illustrated in figure 1.

Table 1. Incidence rate (per 100 patient-years) and 95% confidence interval (CI) for the major events during follow up for different myeloproliferative neoplasms.

	CML	ET	PV	PMF
Thrombosis	2.39 (0.9-6.3)	1.27 (0.75-2.13)	1.29 (0.58-2.85)	0
Hemorrhage	0	0.27 (0.09-0.84)	0	0
Hematological malignancy	0.6 (0.08-4.2)	0.82 (0.43-1.56)	1.07 (0.45-2.57)	2.21 (0.32-15.37)
Solid tumor	0.6 (0.08-4.2)	0.36 (0.14-0.96)	1.07 (0.45-2.57)	0

All-cause mortality rates in deaths per 100 patient-years (95% CI) were for CML 1.8 (0.6-5.5), ET 0.9 (0.5-1.7), PV 1.9 (1-3.7), PMF 6.6 (2.2-19.8). Transformation to acute leukemia accounted for 46.7% of all deaths, solid tumors for 16.7% and other causes (cardiac death, sepsis) for 33.3% of deaths. All patients with atypical CML or other myeloproliferative/myelodysplastic syndrom died at a median of 16.4 months from diagnosis, while their disease had transformed to AML at a median of 14 months from initial diagnosis.

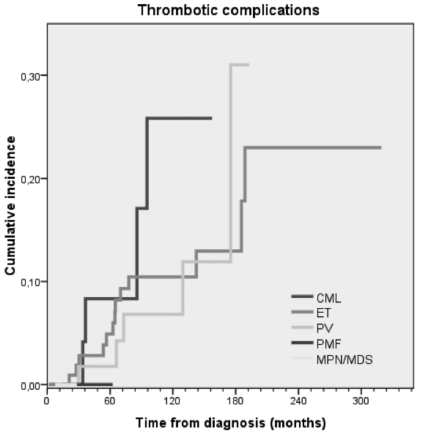

Figure 1.

Summary/Conclusion: Thrombotic complications account for the main burden of morbidity not only in patients with ET and PV, but also for CML patients on TKIs. Transformation to acute leukemia is the leading cause of death. It is crucial that future studies incorporate cardiovascular risk factors and estimate the effect of therapeutic interventions in both the thrombotic risk and the progression free survival.

PB2281

HETEROGENEITY AMONG DIFFERENT MYELOPROLIFERATIVE NEOPLASMS ASSOCIATED WITH SPLANCHNIC VEIN THROMBOSIS

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Background: In *BCR-ABL1*-negative myeloproliferative neoplasms (MPNs) incidence of major thrombotic events ranges from 1.75 to 5.5% events patient-years according to the specific MPN subtype. Venous thrombosis account for about 30-40% of these complications and can occur also at unusual sites, including the splanchnic circulation (SVT) with a prevalence ranging between 1 and 23%.
Aims: To evaluate differences in MPNs associated with SVT.
Methods: We reported a consecutive monocentric series of 38 patients with a diagnosis of *BCR-ABL1*-negative MPN, who developed a SVT at diagnosis or during the follow-up between 1979 and 2016.
Results: We identified 18.4% of PV, 26.3% of ET, 34.2% of MF and 21.1% of MPN-U. The latter were all diagnosed at the time of SVT onset, and characterized most frequently by a PV- or PMF-like bone marrow morphology but lacked the clinical phenotype required for a complete diagnosis. The majority of the cases (81.5%) bear *JAK2*V617F mutation; however, five patients were characterized by other molecular markers, *i.e. MPL* mutations in three patients, and *CALR* type 1 mutation in the remaining two cases. Among patients with a previous diagnosis of MPN who developed SVT during the follow-up, a cytoreductive treatment was already on-going in 53.8% of the cases, whereas it was then started in all but four of the remaining cases, due to young age and a blood cell count in the normal range or even below. After thrombotic index event, anticoagulants were started in 29 patients (76.3%), including six cases (15.8%) with direct oral anticoagulants (DOACs). At a median follow-up from MPN diagnosis of 12.3 years, six deaths were recorded: it was due to leukemic transformation in four patients, intracranial bleeding and infectious complication in one patient each. According to the literature, 44.7% of the patients in our series suffered from recurrent vascular events, either involving the arterial (21.1%), or the venous district (23.7%): in particular, five patients experienced a recurrent SVT.
Summary/Conclusion: In this report patients with a diagnosis of MPN-U seem to represent a distinct clinical entity when compared to the other MPN subtypes. In particular, in all MPN-U cases, SVT was the initial manifestation which led to the diagnosis of the underlying MPN. They were all characterized by the presence of *JAK2*V617F mutation, except one case which bear an *MPL*W515L mutation, and showed a normal karyotype. In addition, any of these patients neither developed clinical features which could enable physicians to re-classify them among one of the so-called classical MPN subtype even according to the WHO 2017 classification, nor experienced a leukemic evolution. Being aware of the limits of the present study, we can speculate that SVT associated with MPN-U represents a disease with a more indolent course as compared with other cases associated with full-diagnosed MPNs which more frequently developed during the follow-up. Notably, all the cases of leukemic transformation were reported among patients with a previous MF diagnosis after a median follow-up of 17.9 years. Furthermore, about half of our patients developed recurrent vascular events, confirming the limited efficacy of conventional therapeutic approach in these particular patients. However, it is interesting to underline that none of the six patients treated with DOACs developed another thrombotic complication, so probably representing a more valid strategy in this setting.

PB2282

THE EPIDEMIOLOGY AND PRESENTING CLINICAL CHARACTERISTICS OF MYELOPROLIFERATIVE NEOPLASMS IN MALAYSIA

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Background: The evolution of molecular studies in myeloproliferative neoplasms (MPN) has enlightened us the understanding of this complex disease consisting of polycythaemia vera (PV), essential thrombocythemia (ET) and primary myelofibrosis (PMF). The epidemiology is well described in the western world but not in Asian countries like Malaysia.
Aims: To research the epidemiology of MPN in Malaysia in correlation with the clinical parameters and molecular studies.
Methods: This national registry of MPN was conducted from year 2009 to 2015 in Malaysia with description of clinical demographic in correlation to JAK2 V617F mutation, thrombosis, haemorrhagic complications and blood counts.
Results: A total of 1010 patients were registered over a period of 5 years. The mean age was 54 years with male predominance. The ethnic distribution revealed that Chinese had a relatively high weighted incidence proportion (43.2%), followed by Indian (23.8%), Malay (15.8%) and other ethnic groups (17.2%). The types of MPN reported were 40.4% of ET (n=408), 38.1% of PV (n=385), 9.2% of PMF (n=93), 3.1% of hypereosinophilic syndrome (HES) (n=31) and 7.9% of unclassifiable MPN (MPN-U) (n=80). Splenomegaly was only palpable clinically in 32.2% of patients. The positive JAK2 V617F mutation was present in 644 patients with 46.6% in PV, 36.0% in ET, 9.0% in PMF, and 7.4% in MPN-U, and had significantly