Appendix 1. Case histories of classical myeloproliferative neoplasms

Diagnosis	Case history	Assessment	Investigations	Management
PRV	Male regular blood donor, aged 40 years, with an incidental finding of a Hb 209 g/L when donating blood. WCC and platelet counts were normal.	• Assess for secondary causes • Assess for persistence and trend of blood counts • Look for thrombosis • Assess cardiovascular risk factors • Examine for organomegaly	• *JAK2* mutation positive • *BCR-ABL* mutation negative • LDH mildly elevated at 342 U/L	• Venesections with the aim to achieve a haematocrit of 0.45 • Aspirin 100 mg daily • Cardiovascular risk factor management
Essential thrombocytosis	Woman aged 64 years with headaches. Basic blood test showed a platelet count of 1000 × 10^9/L. Hb and WCC were normal.		• *JAK2* mutation negative • *CALR* mutation positive • *BCR-ABL* mutation negative • LDH mildly elevated at 308 U/L	• Hydroxyurea 1 g daily with the aim to achieve a normal platelet count • Aspirin 100 mg daily • Cardiovascular risk factor management
CML	Man aged 70 years presented for a routine check-up. FBE showed an elevated WCC of 34 × 10^9/L with a neutrophilia, basophilia, and left shift. Hb and platelet counts were normal.	• Assess for secondary causes • Assess for persistence and trend of blood counts • Examine for organomegaly	• *BCR-ABL* mutation positive • Bone marrow biopsy confirms CML in chronic phase.	• Commenced on dasatinib 100 mg daily • Ongoing monitoring with quantitative PCR *BCR-ABL*
PMF	Man aged 57 years presented with a 12-month history of drenching night sweats and worsening fatigue. Hb was 129 g/L, WCC 12 x 10^9/L, platelet count 189 x 10^9/L. Spleen was palpable at 10 cm below the costal margin.	• Assess blood film: a typical finding for myelofibrosis is a leucoerythroblastic blood film • Consider other causes of a similar presentation (eg other malignancies, autoimmune or infective causes)	• *JAK2* mutation positive • *BCR-ABL* mutation negative • Ultrasonography of the abdomen – 23 cm spleen • Bone marrow biopsy alongside positive *JAK2* mutation confirms myelofibrosis • LDH elevated at 620 U/L	• DIPSS-Plus score was 0 initially. Commenced on hydroxyurea 1 g daily. • Progressed two years later with worsening cytopenias. Commenced on ruxolitinib (*JAK1/2* inhibitor) and referred for an allogeneic transplant.

CML, chronic myeloid leukaemia; DIPSS-Plus, Dynamic International Prognostic Scoring System Plus; FBE, full blood examination; Hb, haemoglobin; LDH, lactate dehydrogenase; PCR, polymerase chain reaction; PMF, primary myelofibrosis; PRV, polycythaemia vera; WCC, white cell count