www.mpnallianceaustralia.org.au

MYELOPROLIFERATIVE NEOPLASMS (MPNs): under diagnosed blood cancers

What are the 'classic' myeloproliferative neoplasms?

1. Essential Thrombocythaemia (ET) - high platelets
2. Polycythaemia Vera (PV) -high haematocrit and haemoglobin
3. Myelofibrosis (MF) – anaemia, variable platelet and white cell counts with immature cells on blood film, enlarged spleen

Could this be an MPN?

A patient with persistent abnormal blood counts - the single most important clue

Consider a myeloproliferative neoplasm in patients with:

- Blood count abnormalities
  - Persistent and unexplained elevation of haemoglobin, haematocrit
  - Persistent thrombocytosis
  - Leukoerythroblastic
- Thrombotic or vascular events
  - Macrovascular (e.g stroke,myocardial infarct)
  - Microvascular (e.g visual disturbances, fluctuating paraesthesia)
- MPN specific symptoms
  - Aquagenic pruritus
- Erythromelalgia (burning, red hands, feet or face)
- Splenomegaly, early satiety or constitutional symptoms
- Non-specific symptoms
  - Fatigue
  - Headaches, dizziness or brain fog
  - Unexplained weight loss

Symptoms that should raise suspicion

Many patients describe symptoms that are often attributed to ageing, stress, **menopause,** depression, or autoimmune disease.
Many patients experience symptoms for years before diagnosis.

Thrombosis – diagnostic clue & also the primary complication for MPN patients

An MPN should be considered in patients with vascular/thrombotic events and an abnormal FBC, especially if thrombotic events are otherwise unexplained.
Thrombotic events occur before or at the time of diagnosis in over 20% of patients.
All patients with a splanchnic vein thrombosis (e.g. hepatic, portal, splenic) should be screened for an MPN.

When should a GP investigate?

Persistent (>3 months) blood count abnormalities (e.g thrombocytosis, polycythaemia)
Consider reactive causes for a thrombocytosis (e.g infection, inflammation) or secondary causes for polycythaemia (e.g hypoxic lung disease, cigarette smoking, testosterone)

If abnormalities remain unexplained, referral to a haematologist is appropriate

Molecular testing for: JAK2 V617F mutation, CALR mutation and MPL mutation is often part of specialist assessment.

Article: MPN diagnostic information for General Practitioners was published in 2021

GP MANAGEMENT OF MPNS

Once diagnosed, under a haematologist's care and in partnership with their GP, most patients with an MPN can have a near-normal life expectancy. GPs should continue routine preventive care alongside MPN-specific management – see below.

Reduce thrombosis risk and cardiovascular risk

These are the major causes of morbidity in MPN patients.

Optimise:

- Weight management
- Smoking cessation
- Blood pressure control
- Lipid management
    - Increasing evidence of the benefit of statin therapy in MPN patients
- Diabetes management
- Physical activity
- Integrative oncology to improve symptoms and wellbeing – details in article

Screening for secondary cancers

All usual screening for secondary cancers is important as MPN patients have a higher rate of secondary cancers – in particular non-melanoma and melanoma skin cancers. **Regular and vigilant skin checks are especially important for MPN patients on hydroxyurea or ruxolitinib and possibly other JAK inhibitors.**

Situations of increased risk:

Obtain specialist advice in situations of increased risk such as surgery, pregnancy, oestrogen use, lower limb injury/immobilisation.

Pregnancy – many patients with an MPN can have a successful pregnancy but certain medications are not safe. Aspirin and interferon can be continued. There is also a need for closer monitoring in pregnancy by the haematologist and obstetrician.

Menopause and HRT

A review article recently published about management of menopause and MPNs.

In addition to above management issues, earlier review may be needed if:

Blood counts are changing significantly; symptoms are worsening; splenomegaly appears to be increasing; new thrombosis occurs; significant bleeding develops; treatment toxicity is suspected, constitutional symptoms emerge.

Red flags - do not delay if:

Stroke or TIA symptoms; chest pain; acute visual loss or major visual change; shortness of breath; suspected venous thromboembolism; severe bleeding; rapid clinical deterioration.

Preventative health matters: a coordinated care system improves outcomes

- Vaccinations
- Cancer screening
- Bone health assessment where appropriate
- Mental health support and management of comorbidities.