Greater Manchester Cancer Network Guidelines: Myeloproliferative Neoplasms (MPNs)

Title of paper	Greater Manchester Cancer Network Guidelines: Myeloproliferative Neoplasms (MPNs)
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Purpose of the paper	In this updated Greater Manchester Cancer guideline, the authors have not sought to replicate in long form existing national and international guidelines, but rather to point to well-regarded sources which the authors themselves appreciate, to guide patient therapy within region. This document will therefore give up-to-date links to the most important of these guidelines, and will be updated from time to time when new guidelines are published.
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Polycythaemia Vera (PV), including diagnosis of non-JAK2 mutated erythrocytosis

British Society of Haematology Guidelines

Diagnosis and management of polycythaemia vera (2018)

https://b-s-h.org.uk/guidelines/guidelines/diagnosis-and-management-of-polycythaemia-vera

- National guideline covering the diagnosis and management of PV
- Includes diagnostic algorithm, risk categorisation and management options.

Management of specific situations in polycythaemia vera and secondary erythrocytosis (2024)

https://b-s-h.org.uk/guidelines/guidelines/management-of-specific-situations-in-polycythaemia-vera-and-secondary-erythrocytosis

- National (UK) guideline
- Includes management of thrombosis, haemorrhage, pregnancy, pruritis.
- Includes management of secondary erythrocytosis

International Guidelines

Polycythemia Vera: 2024 update on diagnosis, risk stratification, and management

https://onlinelibrary.wiley.com/doi/full/10.1002/ajh.27002

- International (USA) guidance by Tefferi and Barbui (American Journal of Hematology)
- More recent guidance than BSH guidance but key differences include:
    - Slight differences in diagnostic classification (Hct > 49% in men/ 48% women)
    - Risk classification (>60 (c.f. 65 in UK guidance) as high risk)
    - Treatment: consider use of BD aspirin in patients with arterial thrombosis

JAK2 unmutated erythrocytosis: 2023 Update on diagnosis and management

https://onlinelibrary.wiley.com/doi/10.1002/ajh.26920

- Helpful guideline for investigation and management of secondary erythrocytosis

Essential Thrombocythaemia (ET)

British Society of Haematology Guidelines

Investigation and management of adults and children presenting with thrombocytosis (2010)

https://b-s-h.org.uk/guidelines/guidelines/investigation-and-management-of-adults-and-children-presenting-with-thrombocytosis

- National (UK) Guidance but from >10 years ago. Diagnostic criteria were updated in 2014 (https://b-s-h.org.uk/guidelines/guidelines/modification-of-diagnostic-criteria-for-essential-thrombocythaemia) but remain ~10 years old.
- Includes: diagnostic criteria, risk stratification and treatment.
- Includes: management of thrombocytosis of other causes e.g. reactive/ secondary to MDS/ MPN conditions
- Guideline does not recognise the differential thrombotic risk of diagnostic mutations (JAK2 vs CALR)

International Guidelines

Essential thrombocythemia: 2024 update on diagnosis, risk stratification, and management

https://onlinelibrary.wiley.com/doi/full/10.1002/ajh.27216

- International (USA) guidelines from Teferri, Vannucchi and Barbui
- Includes updated risk categorisation, taking into account mutation status (JAK2 versus CALR mutated).
- Includes a useful management flow diagram in Figure 5 including possible use of OD/BD aspirin

Myelofibrosis

British Society of Haematology Guidelines

Diagnosis and evaluation of prognosis of myelofibrosis (2023)

- Up to date national (UK guidance) for diagnosis and evaluation of myelofibrosis, including links to validated prognostic scoring systems and causes of secondary marrow fibrosis.

The management of myelofibrosis: A British Society of Haematology Guideline (2024)

- Up to date national (UK guidance) for management of myelofibrosis, including risk classification, indications for allogenic stem cell transplant, new agents and special situations

Summary of Treatment and Prescribing

All patients should be referred to the Haemato-Oncology MDT and formally discussed if active treatment is required. Please consider all appropriate patients for clinical trial options. Treatment should be in keeping with the above guidelines and the following NICE technical appraisals.

NICE Appraisals

Polycythaemia vera:

NICE: Ruxolitinib for treating polycythaemia vera. TA921. Published 18/10/2023

https://www.nice.org.uk/guidance/ta921

Myelofibrosis:

NICE: Momelotinib for treating myelofibrosis-related splenomegaly or symptoms. TA957. Published 20/03/2024

https://www.nice.org.uk/guidance/ta957/evidence/committee-papers-pdf-13363268509

NICE: Ruxolitinib for treating disease-related splenomegaly or symptoms in adults with myelofibrosis. TA386. Published 23/03/2016

https://www.nice.org.uk/guidance/ta386

NICE: Fedratinib for treating disease-related splenomegaly or symptoms in myelofibrosis. TA756. Published 16/12/2021

https://www.nice.org.uk/guidance/ta756/resources/fedratinib-for-treating-diseaserelated-splenomegaly-or-symptoms-in-myelofibrosis-pdf-82611378796741