Lancashire and South Cumbria Haematology CRG
GUIDELINES FOR THE INVESTIGATION AND MANAGEMENT OF ESSENTIAL THROMBOCYTHAEMIA

Proposed diagnostic criteria for essential thrombocythaemia. (BCSH)

Diagnosis requires A1–A3 or A1 + A3–A5

A1	Sustained platelet count >450 × 10^9/l
A2	Presence of an acquired pathogenetic mutation (e.g. in the *JAK2* , *CALR* or *MPL* genes)
A3	No other myeloid malignancy, especially PV*, PMF†, CML‡ or MDS§
A4	No reactive cause for thrombocytosis and normal iron stores
A5	Bone marrow aspirate and trephine biopsy showing increased megakaryocyte numbers displaying a spectrum of morphology with predominant large megakaryocytes with hyperlobated nuclei and abundant cytoplasm. Reticulin is generally not increased (grades 0–2/4 or grade 0/3)

1. * PV,polycythaemia vera ;excluded by a normal haematocrit in an iron-replete patient.
2. † PMF, primary myelofibrosis ; indicated by presence of significant marrow bone marrow fibrosis (greater or equal to 2/3 or 3/4 reticulin) AND palpable splenomegaly, blood film abnormalities (circulating progenitors and tear-drop cells) or unexplained anaemia.
3. ‡ CML, chronic myeloid leukaemia; excluded by absence of *BCR-ABL1* fusion from bone marrow or peripheral blood.
4. § MDS, myelodysplastic syndrome ; excluded by absence of dysplasia on examination of blood film and bone marrow aspirate.

* *BCR-abl* testing recommended if other molecular tests are negative and/or if positive and there are atypical features eg. basophilia

**Bone marrow examination is recommended by the World Health organisation classification to confirm the diagnosis but may not always be clinically indicated

*** The requirement for cytogenetic analysis on the bone marrow be guided by blood and bone marrow morphology.

Molecular testing should only be undertaken at HMDS Leeds

Treatment of Essential Thrombocythaemia (ET)

1. Risk Stratification :

High risk

A.Age > 60

B.Previous ET related haemorrhagic or thrombotic event

C. Platelet count > 1500 X 10^9/l

Intermediate risk

A. Age 40-60 with no high risk features

Low risk

A. Ag e<40 with no high risk features

Microvascular complications not responding to aspirin should be stratified as high risk.

2. Management:

- All patients should have cardiovascular risk factors reviewed and managed appropriately
- All patients should start on low dose Aspirin (75mg daily) unless contraindication (caution with platelet count above 1000 X 10^9/l due to the risk of bleeding)
- High risk patients should be started on cytoreductive treatment with a platelet target <400 X 10^9/L
- Low and intermediate risk patients should generally not be started on cytoreductive treatment outside the context of a clinical trial or if they are symptomatic: progressive splenomegaly; erythromelalgia not responsive to aspirin; uncontrolled bleeding with high platelet counts.

Cytoreductive treatment

- 1st line – Hydroxycarbamide
- 2nd line – Anagrelide
- Consider interferon alfa in young patients and in high risk pregnancy
- Consider radioactive phosphorus or busulfan in those with short life expectancies (risk of leukaemic transformation)

- **All patients starting on cytoreductive treatment or changing treatment must be discussed at an appropriate MDT.**

- **All molecular studies/ histology should be sent to HMDS Leeds for review.**

- **Previously diagnosed patients being represented to the MDT for change of treatment without results at HMDS Leeds, requires formal presentation of molecular studies for oversight by the MDT.**

- **Patients should have holistic support with clinical nurse specialist input and written information about their disease.**

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Diagnostic pathway for investigation of thrombocytosis