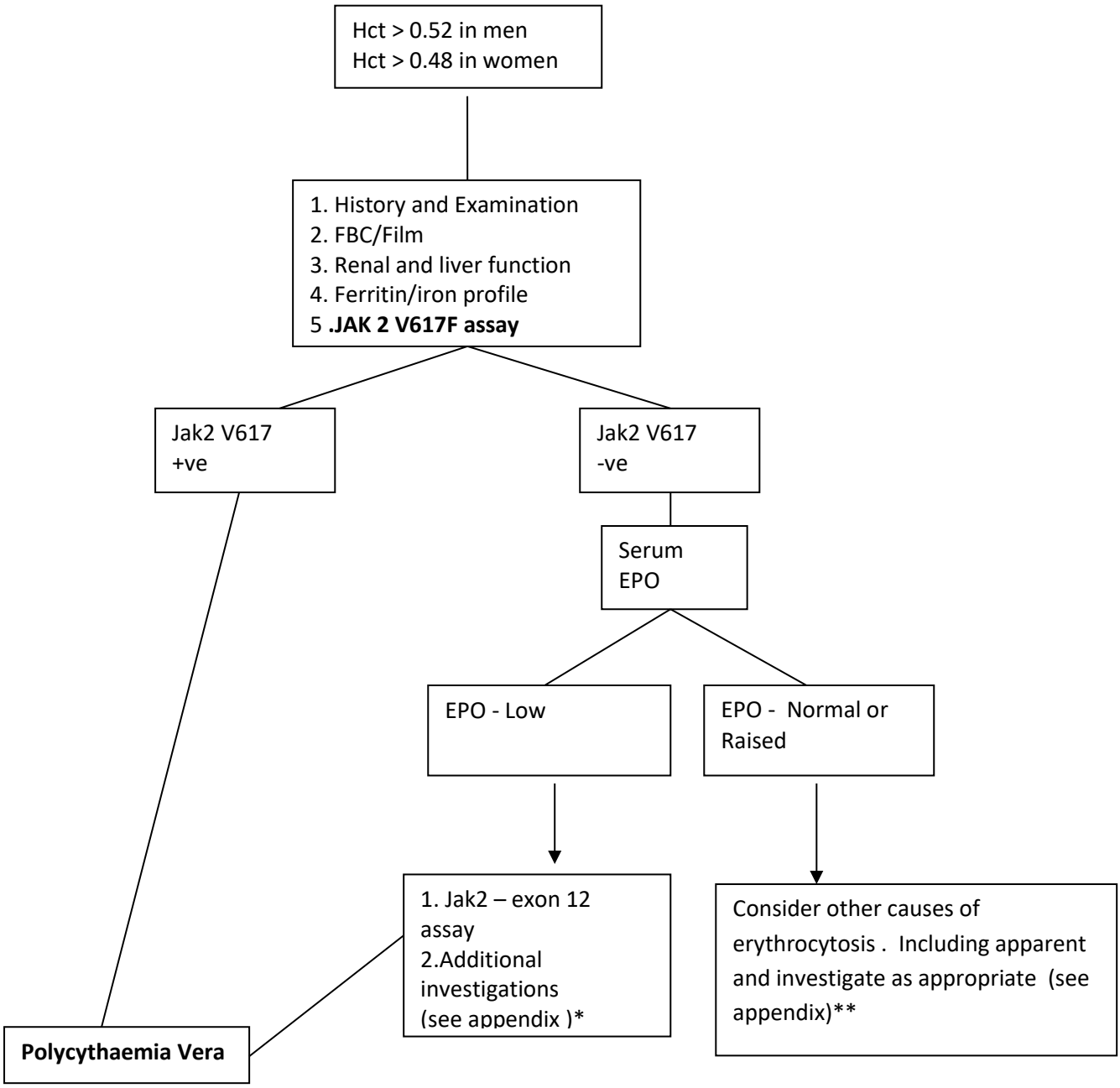
Lancashire and South Cumbria Haematology CRG
GUIDELINES FOR THE INVESTIGATION AND MANAGEMENT OF POLYCYTHAEMIA VERA

Investigation algorithm:



JAK 2 mutational assay should only be undertaken at HMDS LEEDS

Note: A Hct > 0.56 in women and > 0.60 in men is evidence of true erythrocytosis

Diagnosis

(Modified BCSH diagnostic criteria for polycythaemia vera)

JAK2-positive polycythaemia vera

A1 High haematocrit (>0.52 in men, >0.48 in women) OR raised red cell mass (>25% above predicted)

A2 Mutation in JAK2

Diagnosis requires both criteria to be present

JAK2-negative polycythaemia vera

A1 Raised red cell mass (>25% above predicted) OR haematocrit >0.60 in men, >0.56 in women.

A2 Absence of mutation in JAK2

A3 No cause of secondary erythrocytosis

A4 Palpable splenomegaly

A5 Presence of an acquired genetic abnormality (excluding BCR-ABL) in the haematopoietic cells

B1 Thrombocytosis (platelet count >450 X 10^9/l)

B2 Neutrophil leucocytosis (neutrophil count > 10 X 10^9/l in non-smokers; >12.5 X 10^9/l in smokers)

B3 Radiological evidence of splenomegaly

B4 Endogenous erythroid colonies or low serum erythropoietin

Diagnosis requires A1 + A2 + A3 + either another A or two B criteria

Treatment of Polycythaemia Vera

1. Risk Stratification :

High risk

A. Age > 65

B. Previous Thrombosis

Low risk

None of the above risk factor

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)
- Low risk patients – Venesect to a target HCT < 0.45 (consider cytoreductive treatment if poor tolerance of venesection, symptomatic or progressive splenomegaly, significant thrombocytosis, progressive leucocytosis)
- High risk patients – Cytoreductive treatment, +/- Venesection

Cytoreductive treatment

- Age < 40 - 1st line : Inteferon alpha ; 2nd line Hydroxycarbamide
- Age > 40 – 1st line : Hydroxycarbamide; 2nd line Inteferon alfa, anagralide
- Busulfan as second line treatment in those over 75 (increased risk of transformation to acute leukaemia – Radioactive Phosphorus is currently unavailable)

- **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.

Appendix 1

Secondary investigations of erythrocytosis:

- Red cell mass studies *
- Abdominal ultrasound*
- Bone marrow aspirate and trephine with cytogenetic analysis*
- Erythroid burst forming unit cultures*
- Arterial oxygen dissociation studies **
- High affinity Hb variant studies**
- Sleep studies**
- Lung function tests**
- EPOR, VHL, PHD2 mutational analysis**

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