14. POLYCYTHAEMIA

14.1 SCOPE

An elevated haemoglobin / haematocrit has a wide differential diagnosis including:

- Primary proliferative polycythaemia, or polycythaemia vera (PV)
- Secondary causes (such as hypoxic lung disease, obesity/obstructive sleep apnoea, erythropoietin-secreting tumours, testosterone supplementation)
- Relative polycythaemia resulting from plasma depletion. (eg. dehydration, diuretics)

The JAK2 V617F mutation is detectable in over 95% of patients with PV. Where this mutation is absent, it is a reliable indicator that the patient does not have PV and that secondary causes are implicated.

14.2 INDICATIONS FOR TESTING

- Haematocrit* > 0.52 in males
- Haematocrit* > 0.48 in females

**Testing on at least two separate occasions > 3 months apart*

14.2.1 Blood tests to be sent in primary care

- Erythropoietin (clotted sample)
- JAK2 V617F mutation (EDTA sample)

14.3 REFERRAL CRITERIA

- All JAK2 V617F positive cases
- JAK2 V617F negative cases, and
    - Unprovoked thrombosis
    - High/low erythropoietin level AND no secondary cause**

14.4 CRITERIA FOR MANAGEMENT** IN PRIMARY CARE

- JAK2 V617F negative, and
- Normal erythropoietin level
- Secondary cause present irrespective of erythropoietin level

14.5 **MANANGEMENT OF SECONDARY POLYCYTHAEMIA IN PRIMARY CARE

Routine FBC monitoring is not indicated.

Aspirin is not indicated.

Venesection is rarely indicated in secondary polycythaemia, and management should be directed at the underlying cause. In select cases it can be considered if symptomatic; seek haematology advice if there is clinical concern.

(This Guidelines have come from the South West MPN Group with some slight amendments for the PCA.)