

## UVM Health HEMATOLOGY ERYTHROCYTOSIS REFERRAL/CONSULT

### Background:

- Erythrocytosis (also called polycythemia) refers to an increased hemoglobin concentration and/or hematocrit in peripheral blood. Red blood cells are produced by the bone marrow in response to erythropoietin (“epo”), produced by the kidney.
- Parameters for erythrocytosis:
  - o **Increased hemoglobin:** >16.5 g/dL in men or >16.0 g/dL in women
  - o **Increased hematocrit:** >49% in men or >48% in women
- Erythrocytosis can be (1) relative (e.g. dehydration); (2) secondary- appropriate response to erythropoietin signaling to produce more red blood cells, or (3) primary – production of red blood cells due to acquired genetic mutations (Polycythemia Vera/ Myeloproliferative Neoplasm) or inherited mutations.
- Treatment of secondary erythrocytosis is directed at the underlying driver. Treatment of Polycythemia Vera requires cytoreduction to mitigate risk of thrombosis.

Primary erythrocytosis	Secondary erythrocytosis
<ul style="list-style-type: none"><li>• <i>Myeloproliferative neoplasm (polycythemia vera etc) (JAK2 mutation)</i></li><li>• <i>Inherited mutations (e.g., VHL gene, epo receptor) – extraordinarily rare.</i></li><li>• <i>Populations from South America from high altitudes</i></li></ul>	<ul style="list-style-type: none"><li>• <i>Hypoxia</i></li><li>• <i>Exogenous testosterone</i></li><li>• <i>Sleep apnea</i></li><li>• <i>Smoking</i></li><li>• <i>Carbon monoxide poisoning</i></li><li>• <i>Autonomous epo production (e.g., Hepatocellular carcinoma, renal cell, fibroids)</i></li><li>• <i>Renal artery stenosis; post-kidney transplant</i></li><li>• <i>SGLT-2 inhibitors (canagliflozin, empagliflozin, dapagliflozin, ertugliflozin)</i></li></ul>

### Evaluation (within the prior 3 months):

- Complete Blood Count with differential x 2 (to show persistence over at least 1 month), Comprehensive metabolic profile, ferritin, erythropoietin level, urinalysis
- Pulse oximetry
- If epo is low --> send to heme and send JAK2 V617F mutation (requires prior authorization)
- If epo is normal to elevated --> do not send JAK2 mutation; review list of secondary causes

**Call Hematology consult attending through PAS (802 847-2700) to expedite an evaluation/referral if these exam reveals any of the following:**

- Signs of hyperviscosity (i.e., stroke-like symptoms and/or visual disturbances)
- Acute or recent cardiovascular disease or venous thrombosis (6 months)

- Other abnormalities on the CBC (thrombocytosis, blasts, metamyelocytes, myelocytes)