University of Vermont Health

UVM Health HEMATOLOGY ERYTHROCYTOSIS Referrals/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:
    - **Increased hemoglobin:** >16.5 g/dL in men or >16.0 in women
    - **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

- *Myeloproliferative neoplasm (polycythemia vera etc) (JAK2 mutation)*
- *Inherited mutations (e.g., VHL gene, epo receptor) – extraordinarily rare.*
- *Populations from South America from high altitudes*

Secondary Erythrocytosis

- *Hypoxia*
- *Exogenous testosterone*
- *Sleep apnea*
- *Smoking*
- *Carbon monoxide poisoning*
- *Autonomous epo production (e.g., Hepatocellular carcinoma, renal cell, fibroids)*
- *Renal artery stenosis; post-kidney transplant*
- SGLT-2 inhibitors (canagliflozin, empagliflozin, dapagliflozin, ertugliflozin)

University of Vermont Health

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)