APPENDIX 1. Erythrocytosis[1,2,20,21,53]

Hemoglobin: > 160 for females, > 165 for males
Hematocrit: > 0.48 in females > 0.49 in males

→ Urgent referral if:
Hemoglobin > 200
Recent thrombosis or symptoms of hyperviscosity

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Confirm result in 2–4 weeks
If patient is dehydrated, repeat when patient is euvolemic

→ Resolved → Relative erythrocytosis
No further workup or treatment needed

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If persistent erythrocytosis, conduct history and physical exam
Order serum erythropoietin (EPO)
Consider ordering JAK2 if EPO is low or normal

History	• Symptom review:
	  ○ Symptoms of hyperviscosity—**fatigue, headache, dizziness, transient blurry vision, paresthesias, burning sensation in hands/feet**
	  ○ **Pruritus after hot shower**
	  ○ Epistaxis, gastrointestinal bleeding, easy bruising
	  ○ **Non-specific abdominal pain (suggests peptic ulcer)**
	  ○ **Left hypochondrial pain and early satiety (suggests splenomegaly)**
	  ○ Constitutional symptoms (e.g., fever, night sweats, unintended weight loss)
	• Past medical history: **history of thrombosis**, congenital heart disease
	• Medication review: particularly testosterone or anabolic steroids
	• Family history: polycythemia
	• Social history: cigarette smoking, alcohol intake, extended stay at high altitude

Physical exam	• BP, BMI • Cardiac, respiratory, abdominal exams, including assessment for **hepatomegaly** and **splenomegaly**

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Primary erythrocytosis
Suspect polycythemia vera if:
- Low or normal EPO and JAK2 positive
- **Bolded** symptoms above (suggestive)

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Referral
or electronic/phone consult with hematology (if appropriate)

Secondary erythrocytosis
Suspect if:
High EPO
History and physical exam suggest underlying cause

Type	Causes
Hypoxia-driven	*Cigarette smoking, COPD, obstructive sleep apnea,* obesity, intracardiac shunting, high altitude
Local renal hypoxia	Renal artery stenosis, hydronephrosis, polycystic kidney disease, end-stage renal disease
Medication-related	Androgen therapy (testosterone, anabolic steroids), EPO
Pathologic erythropoietin production	Renal cell, hepatocellular, or parathyroid carcinoma Cerebellar hemangioblastoma, uterine leiomyomata, meningioma, pheochromocytoma
Others	Renal transplantation, idiopathic

Note: Causes in italics are the most common in primary care

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Further investigations and treatment guided by underlying cause
Refer as indicated

JAK2 = Janus Kinase 2
EPO = erythropoietin

Refer to the Reference List for cited sources

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