Polycythemia Vera

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Risk

- Prevalence: 22:100,000; twice as high among men as women.
- Higher prevalence in Jewish people of European origin.
- Prevalence increases with advancing age (rare in those <30 y).

Perioperative Risks

- Risk of deep venous thrombosis, pulmonary emboli
- Risk of coronary, cerebral thrombosis/ischemia
- Increased histamine release and prostaglandin production

Worry About

- Hyperviscosity from increased Hct
- Preop treatment should include phlebotomy to Hct of ≤45%
- Increased plt count and plt aggregation
- Thrombotic complications: MI, stroke, deep venous thrombosis, Budd-Chiari syndrome
- Bleeding diathesis

Overview

- A chronic myeloproliferative disease characterized by increased red blood cell mass.
- Often WBC and plt counts are increased.
- The resulting hyperviscosity of blood predisposes to thrombosis.
- Symptoms: Headaches, erythromelalgia (pain in hands/feet), pruritus.
- Predisposed to gouty arthritis, peptic ulcer disease.
- Accompanied by palpable splenomegaly.

Treatment

- Although incurable, treatment increases life expectancy from 1–2 y to 20 y
- Phlebotomy is first-line therapy
- Low-dose aspirin (81 mg/d) is often given to reduce thrombotic risk.
- Hydroxyurea (Hydrea): The most commonly used myelosuppressive agent for PV. Helps reduce both Hct concentration and plt count.
- Ruxolitinib (Jakafi): A Janus-associated kinase inhibitor, approved by the US FDA for treatment of pts with PV who have had an inadequate response to hydroxyurea or are intolerant of it.

Etiology

- Unclear, but a mutation in the *JAK2* (Janus kinase 2 gene) increases response to erythropoietin.
- Normal oxygen saturation (if low, may be secondary polycythemia).
- Low erythropoietin levels (if high, may be secondary polycythemia).

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT		Headaches, tinnitus, blurred vision or blind spots, dizziness or vertigo		
CV	Increased intravascular volume	Angina, coronary artery microthrombus	Reddened, purplish skin	Increased Hgb, Hct, red cell count Hct: >46% in women >52% in men May have increased WBC and plt count
RESP/HEME	Absence of hypoxemia	Poor exercise tolerance Smoking, high altitude not a cause Often asymptomatic	Signs/symptoms of pulm emboli	Normal arterial oxygen saturation *JAK2* mutation in blood cells
GI	Splenomegaly Budd-Chiari syndrome Peptic ulcer disease	Symptoms of liver disease secondary to Budd-Chiari syndrome	Splenomegaly	Liver enzymes
RENAL	Potential renal thrombosis			
ENDO	Low EPO levels			Bone marrow: Hypercellular, low iron stores
CNS		Headaches		
PNS	Erythromelalgia (pain in hands/feet)			
MS	Itching, gouty arthritis	Fatigue	Extremity edema, signs of DVT Bleeding, bruising in 25%	DVT assessment by US or venogram

Key References: Leukemia and Lymphoma Society: Polycythemia vera facts. FS13:1-7. <https://www.lls.org/sites/default/files/file_assets/FS13_PolycythemiaVera_FactSheet.pdf>, 2015 (Accessed 01.06.16); Finazzi G, Barbui T: How I treat patients with polycythemia vera, *Blood* 109(12):5104–5111, 2007.

Perioperative Implications

Preoperative Preparation

- Preop phlebotomy
- Hydration for hemodilution
- May benefit from periop hematology/oncology consult
- For low-risk blood loss cases, may continue aspirin, weighing the risk of thrombosis versus bleeding

Monitoring

- ST-segment changes for myocardial ischemia
- Large-bore access to facilitate additional phlebotomy

Induction/Maintenance

- Both RA and GA are options
- Caution with neuraxial anesthetics if there is a bleeding diathesis

Intraoperative Management

- Intraop hemodilution to reduce thrombotic risk
- Some pts at risk for increased bleeding and transfusion

Postoperative Period

- Increased risk for thrombosis and bleeding diathesis.
- Aggressive DVT prophylaxis is important.

Anticipated Problems/Concerns

- Vigilance for prevention, diagnosis, and treatment of thrombotic events
- Also must be prepared to treat bleeding diathesis
- Treat symptoms: Itching, fatigue, angina, heart failure, gout
- PV pts at greater risk for thrombosis than for secondary polycythemia

Polymyositis

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Risk

- Annual incidence in USA: 5.5:1,000,000, most prevalent among black women
- Annual incidence around the world: 1.9-7.7:1,000,000, although comprehensive epidemiologic data are lacking

Perioperative Risks

- Delayed recovery from muscle relaxation
- Aspiration pneumonitis
- Cardiac arrhythmias
- CHF

Worry About

- Increased risk of aspiration
- Respiratory muscle and/or diaphragmatic weakness
- Hyperkalemia following succinylcholine use; sensitivity to NMB
- Interstitial lung disease, progressive fibrosis, and/or difficulty with ventilation/oxygenation
- Cardiomyopathy with heart failure
- Chronic use of therapeutic steroids and anti-immunologic medications

Overview

- Rare form of an acquired inflammatory myopathy affecting adults and rarely children.
- Can often mimic many other myopathies and is a diagnosis of exclusion.
- Pts present with progressive and symmetric proximal muscle weakness.

- Diagnosis confirmed by analysis of serum muscle enzymes, EMG findings, and muscle biopsy (most definitive test).
- A careful family history, medication list review, physical exam, blood test, and muscle biopsy are all crucial because they may help to exclude an alternative diagnosis, such as an inherited muscle disease or toxic myopathy.

Etiology

- An autoimmune etiology is suspected and hypothetically supported by an association with other autoimmune or connective tissue diseases and a response to immunotherapy.
- Drugs—especially D-penicillamine, statins, or zidovudine—may also trigger an inflammatory myopathy.
- Several viruses—including coxsackie, influenza, mumps, CMV, and Epstein-Barr virus—may also have an association.

Usual Treatment

- Treatment focuses on controlling inflammatory response through immunosuppression.
- Steroids, with prednisone as first-line agent.
- Immunosuppressive drugs, which include azathioprine, methotrexate, mycophenolate mofetil, rituximab, cyclosporine, tacrolimus, cyclophosphamide.
- IVIG.
- Physical therapy.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Neck muscles weakness	Headache Head drop	Neck ROM Head lift	EMG
RESP	Inspiratory muscles weakness Interstitial lung disease Aspiration pneumonitis	Dyspnea Chronic cough Limited exercise tolerance	Dyspnea/tachypnea Wheezing Hypoxia	PFTs CXR, CT scan ABG Bronchoscopy
CV	Conduction abnormalities CHF	Chest pain Dyspnea Palpitations	Arrhythmia Edema Inspiratory crackles	ECG TTE Stress test
GI	Pharyngeal muscle weakness	Dysphagia	Regurgitation Aspiration	Endoscopy CXR
CNS	Systemic manifestations	Malaise Fever	Hyperthermia	
HEME	Raynaud phenomenon	Cold digits	Digit vasospasm	
DERM	Only seen with concomitant disease, dermatomyositis	Rash	Erythematous and raised papules on extensor surfaces Heliotrope rash	Muscle/skin biopsy
MS/RHEUM	Proximal muscle weakness Arthralgias or arthritis Calcinosis of subcutaneous tissue Coexisting rheumatologic disorder (scleroderma, SLE)	Myalgia Muscle tenderness Skin ulceration Joint swelling	Muscle weakness, atrophy Delayed reflexes	CPK, CK, ALT, AST, LDH Autoantibodies EMG Joint x-ray Muscle biopsy

Key References: Strauss KW, Gonzalez-Buritica H, Khamashta MA, et al.: Polymyositis and dermatomyositis: a clinical review, *Postgrad Med J* 65(765):437–443, 1989; Gunusen I, Karaman S, Nemli S, et al.: Anesthesia management for cesarean delivery in a pregnant woman with polymyositis: a case report and review of literature, *Cases J* 2:9107, 2009.

Perioperative Implications

Preoperative Preparation

- Assess cardiovascular and pulm status.
- Consider use of RA in order to limit GA and use of NMB; there are some case reports of successful and safe neuraxial techniques and limited reports on peripheral nerve blockade.
- Concomitant steroid therapy and necessity of stress doses should be considered.

Monitoring

- Arterial line if indicated (either owing to CHF or frequent blood draws for ABG)
- TOF peripheral nerve stimulation with NMB use (consider baseline stimulation before NMB given)
- Foley catheter for urine output assessment if pt has cardiac disease

Airway

- Consider rapid sequence intubation if pt has dysphagia.

Induction

- Avoid use of succinylcholine (may cause hyperkalemia).
- Volatile agents and succinylcholine may serve as a trigger malignant hyperthermia and should be avoided in pts with baseline elevated CPK levels.
- If not necessary, avoid nondepolarizing NMB due to increased sensitivity (vecuronium and pancuronium associated with prolonged neuromuscular paralysis).
- Consider use of remifentanil to aid with intubation/for hypokinesis.

Maintenance

- Volatile anesthetics may potentiate the effects of muscle relaxation.
- Consider total IV anesthetic technique.
- Antagonism to NMB may cause additional muscle weakness and/or cardiac dysrhythmias.
- Consider stress-dose steroids.
- Avoid overuse of narcotics.
- Keep pt euvolemic to avoid heart failure.

Extubation

- If NMB is used, confirm complete reversal using twitch monitor and confirm full strength of pt with head lift (if able to do so preop).
- Confirm that pt is completely awake and able to breathe independently of ventilator prior to extubation.
- Consider NIF test to assess adequacy of strength of ventilation.

Postoperative Period

- If possible, keep head of bed elevated to assist with pulm function and to avoid an aspiration event.
- Increased susceptibility to infection if on immunosuppression.

Anticipated Problems/Concerns

- May need ICU stay postop to wean off ventilator.
- Pain control management; avoid overuse of narcotics, which may lead to oversedation and/or apnea.
- May need continued dose of stress dose steroids through periop period.
- Volume shifts may complicate cardiac status.
- Consider swallow study before oral intake to avoid unanticipated dysphagia and an aspiration event.

Pompe Disease

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Risk

- Combined incidence (infantile vs. late-onset): 1:40,000.
- Infantile form has higher incidence in African-American and Chinese populations.
- Late-onset disease has a higher incidence in the Netherlands.

Perioperative Risks

- Respiratory insufficiency
- Aspiration pneumonia
- Pulm edema
- Myocardial ischemia

Worry About

- Respiratory insufficiency, which may require prolonged mechanical ventilation
- Myocardial ischemia
- Arrhythmias, sudden death