Sickle Cell Trait

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Risk

- Incidence in USA: 3 million; 350 million in world
- Race with highest prevalence: African Americans

Perioperative Risks

- Increased risk of complications following CABG.
- Periop mortality rate in published cases of SA trait is 0.8%.
- Some increased risk of CVA and pulm infection but not well quantified.

Worry About

- Increased risk of vasoocclusive phenomenon with hypoxia and stress.
- Sudden death with stresses such as vigorous exercise make one worry because recovery from surgery may be considered a similar stress as vigorous exercise.

Overview

- Is not a disease
- Is not a cause of abnormalities in blood count
- Does not produce vaso-occlusive symptoms under physiologic conditions; painful crisis not a hallmark or concomitant of condition
- Does not adversely affect individual's life expectancy
- Dx established by Hgb electrophoresis
- See also Sickle Cell Disease

Etiology

- Heterozygous, in which individual has one beta S and beta A globin gene (SA disease); Sickle Thal is one beta S and one beta C (SC disease)

Usual Treatment

- None except iron supplementation (debated)

Assessment Points

System	Effect	Assessment by Hx	Test
RESP	Pulm embolism		
HEME	Hgb level usually 13–15 g/dL	Hx SOB: Poor exercise tolerance 10–40% of Hgb S; same cells as Hgb A	Hgb
GU	Painless hematuria and bacteriuria; pyelonephritis (especially with pregnancy) RO polycystic kidney disease		UA (culture if prosthesis planned)
CNS	Stroke	Migraine headache	

Key References: Djaiani GN, Cheng DC, Carroll JA, et al.: Fast track cardiac anesthesia in patients with sickle cell abnormalities, *Anesth Analg* 89(3):598–603, 1999; Tsaras G, Owusu-Ansah A, Boateng FO, et al.: Complications associated with sickle cell trait: a brief narrative review, *Am J Med* 122(6):507–512, 2009; Ingle SS, Ubale P: Anesthetic management of a patient with sickle cell disease for common bile duct exploration, *J Anaesthesiol Clin Pharmacol* 27(4):547–549, 2011.

Perioperative Implications

Preoperative Preparation

- Warm room.
- Consider prehydration.

Monitoring

- Temperature

Airway

- Occasionally distorted anatomy secondary to extramedullary erythropoiesis.
- Sinusitis possible.
- Prehydrate liberally if CV status will tolerate.

Induction

- Routine

Maintenance

- Keep warm.
- Keep vasodilated.
- Keep without stasis.
- High O_2 content.

Extubation

- Keep warm.

Adjuvants

- Vary if hepatic or renal insufficiency exists.

Postoperative Period

- Aggressively prevent pain, hypovolemia, and hypothermia.

Anticipated Problems/Concerns

- Stroke and/or pulm emboli or infection have been reported after CPB; 5 of 546 pts in literature of sickle trait disease died periop.

Silicosis

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Risk

- Silicosis is irreversible fibronodular lung disease caused by inhalation of dust containing crystalline silica (alpha-quartz or silicon dioxide) during occupational exposure.
- Currently, >1,000,000 workers are exposed, with 200–300 deaths/y; protection devices decrease incidence.
- Mostly >65 y of age
- Incidence higher in males than females.
- No racial predilection.

Perioperative Risks

- Hypoxemia, CO_2 retention with chronic respiratory acidosis, bronchospasm, pneumothorax, atelectasis, mycobacterium (30-fold increased risk for TB) and fungal infection, bacterial pneumonia, chronic bronchitis exacerbation
- Periop respiratory failure, especially following thoracic and upper-abdominal surgery
- Pulm Htn; cor pulmonale
- Renal insufficiency (tubular nephropathy)
- Steroid-induced diabetes (in cases of chronic steroid treatment)

Worry About

- In cases of associated scleroderma and/or rheumatoid arthritis, possible difficult intubation
- Bronchospasm and chronic bronchitis exacerbation
- Respiratory failure
- Cor pulmonale

Overview

- Silicosis-pulmonary fibrosis commonly occurs after 4–5 (acute, very rare), 5–10 (accelerated), or >10 y (chronic) of occupational exposure.
- In advanced stage, both obstructive (graduate loss of FEV_1, FVC and decrease of FEV_1/FVC ratio) and restrictive ventilatory defects, as well as decreases in diffusing capacity, are common; exertional dyspnea is the predominant symptom.
- CO_2 retention, pulm Htn, or cor pulmonale late in the course.
- Associated TB, lung cancer, connective tissue diseases (scleroderma, rheumatoid arthritis, Sjögren's syndrome), nephritic syndrome, and renal failure.

Etiology

- Prolonged occupational exposure may occur from mining, stone cutting, sandblasting, abrasive industries, granite quarrying, packing silica flour; this causes dose- and time-related development of pulmonary fibrosis.
- Alveolar macrophages engulf inhaled free silica particles and enter lymphatics and interstitial tissue. The macrophages cause release of cytokines (tumor necrosis factor-α, IL-1), tumor growth factor-β, and oxidants, stimulating parenchymal inflammation, collagen synthesis, and ultimately fibrosis.
- Initial lesions are silicotic nodule mostly located near the respiratory bronchiole. The nodule is composed of refractile particles of silica surrounded by whorled collagen in concentric layers, with macrophages, lymphocytes, and fibroblasts in the periphery. Emphysematous blebs surround the silicotic nodule, especially in the subpleural area. Bleb and bulla formation, and airways and vascular bed distortion by these nodules complicate advanced disease.

Usual Treatment

- Discontinue occupational exposure.
- In cases of silicoproteinosis, the whole lung lavage, with double-lumen tube intubation, may be indicated; rarely, lung transplantation.
- In some cases, oral corticosteroids.
- Empiric use of bronchodilators and inhaled corticosteroids for obstruction.
- If symptomatic/hemodynamically significant, pulm Htn treatment may include sildenafil; endothelin receptor antagonists (bosentan); high-dose calcium channel blockers, such as diltiazem, amlodipine, and in selected cases, verapamil. Epoprostenol infusions are rarely indicated.
- Prophylaxis for complicating infections (pneumococcal and influenza vaccines, TB treatment).
- Smoking discontinuation.
- No cure so far.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Pulm Htn Cor pulmonale	Dyspnea Exercise tolerance Leg swelling	S_3 Peripheral edema Distended neck veins	ECG CXR TTE
RESP	Pulm fibrosis Bulla/bleb formation	Cough Sputum production Dyspnea Exercise tolerance	Rales, rhonchi, Wheezing Cyanosis Use of accessory respiratory muscles RR	CXR ABG PFTs Inspiratory force Diffusing capacity Lung biopsy/lavage fluid microscopy
GI/ENDO	Weight loss Hyperglycemia (in chronic steroid treatment)			Body weight monitoring Blood glucose
MS	Generalized weakness			
RENAL	Renal insufficiency		Hypertension Peripheral edema Oliguria	Serum creatinine, BUN, potassium Creatinine clearance
IMMUNE	Hilar adenopathy (eggshell calcification) Increased susceptibility to infection, especially pulm	Cough Fever Sputum production		CXR Sputum culture and sensitivity

Key References: Rose C: Silicosis. In King TE Jr, Hollingsworth H, editors: *UpToDate*, Waltham, MA. www.uptodate.com/contents/silicosis. (Accessed 09.06.16); Stafford M, Cappa A, Weyant M, et al.: Treatment of acute silicoproteinosis by whole-lung lavage, *Semin Cardiothorac Vasc Anesth* 17(2):152–159, 2013.

Perioperative Implications

Preoperative Preparation

- Lung condition optimization: Treat bronchospasm (if present), bronchitis, and other pulmonary infections; possible lung lavage.
- Consider steroids (short course).
- Stop smoking at least 24 h before surgery.

Monitoring

- Preop and postop: Consider repetitive ABGs, lung mechanics (RR, TV, MV, FVC, NIF, etc.).
- Intraop: Arterial line; CVP is controversial. Consider PA catheter if pulm Htn is present and/or significant fluid shifts are expected.

Pre-induction/Induction

- Caution with IV agents that depress ventilation and regional techniques that affect accessory muscles of respiration (e.g., high epidural and interscalene blocks).
- Maintain adequate preload, and optimize cardiac output. Avoid hypoxemia, hypercapnia, and acidosis (both respiratory and metabolic), as these may increase PA pressures and worsen cor pulmonale.

Airway

- In case of difficult airways, consider techniques with spontaneous respiration preservation (e.g., awake FOI).

Maintenance

- Consider pressure-controlled mode of ventilation, for poor lung compliance may require increased airway pressures to reach the adequate TV. Observe for spontaneous pneumothorax, especially in severe disease.
- Optimize volume status, while avoiding crystalloids overload; rather, use colloids. If possible, minimize blood products use to avoid lung injury.
- Avoid hypotension. Treatment may include low doses of vasopressin, which decreases PA pressures while maintain systemic BP, rather than norepinephrine, which increases PAP and promotes acidosis; phenylephrine, while safe at low rates (0.2–0.6 mcg/kg/min), may exacerbate pulm Htn in higher rates.
- For severe metabolic acidosis treatment, consider THAM solution. Bicarbonate should be avoided because of excessive CO_2 production and hypernatremia.
- Consider use of remifentanil. Caution with long-acting opioids.
- For muscle relaxation, short-acting agents titrated to effect may be preferred.
- Any of inhalational agents are adequate options.

Extubation

- Consider temporary postop mechanical ventilation, especially for upper abdominal and thoracic surgery, until stringent criteria are met.

Postoperative Period

- Pain management is critical for adequate respiration and to avoid worsening pulm Htn.

Adjuvants

- Bronchodilators, supplemental O_2, incentive spirometry may improve ability to wean.

Anticipated Problems/Concerns

- Increased risk of respiratory failure and complications, especially after upper abdominal and thoracic surgery.
- Pts with pulm Htn with or without cor pulmonale are at increased risk of cardiac complications.

Single (Including Common) Ventricle

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Risk

- HLHS is the most common SV congenital cardiac malformation.
- HLHS accounts for 7.5% of newborns with CHD.
- Male predominance for HLHS.

Perioperative Risks

- Paradoxical emboli.
- Complications of chronic hypoxemia: Hyperviscosity, decreased coagulation factors and platelets
- Surgical shunts (narrowing of vessels anastomosed, obstructed shunts)
- Hypovolemia-induced poor pulm blood flow or shunt occlusion.
- Additional risks specific to anatomy and planned procedure.

Worry About

- Effect of changes in PVR, SVR, and cardiac function on blood flow, cardiac output, and O_2 saturation.
- Diastolic pressure and coronary perfusion.
- AV valve regurgitation.
- Systolic and diastolic dysfunction.
- Associated anomalies.
- Increasingly common to care for CHD and SV pts having noncardiac surgery who may be at various stages in the palliation repair process and may have comorbidities including protein losing enteropathies, plastic bronchitis, ventricular dysfunction, and arrhythmias.

Overview

- A wide variety of lesions are usually associated with atresia of the ipsilateral AV or semilunar valve resulting in SV physiology:
  - TA is the prototypic single left ventricle (see Tricuspid Atresia).
  - HLHS with mitral and aortic stenosis/atresia is the prototypic single right ventricle.
- Other anatomies include unbalanced AV canal, some double inlet or double outlet ventricles, and some heterotaxies.
- Initial lesion requires mixing of systemic and pulm venous return at ASD or VSD level. The SV output is divided between pulm and systemic circulations.
- SV anatomy may be associated with hypoplasia of a great vessel (pulm artery or aorta) and prior to initial palliation; systemic or pulm blood flow may be dependent on ductus arteriosus patency.
- Balance of blood flow in each Qp:Qs is governed by the relative resistance to flow as determined by both anatomic and vascular resistance considerations.
- Goal throughout all stages is to balance the Qp:Qs at 1:1.
  - With complete mixing, Qp:Qs at 1:1 results in sat of 75–80% at FiO_2 0.21.
- FiO_2, CO_2, and pH management can be used to manipulate the Qp:Qs.
- Qp:Qs > > 1 results in pulm overcirculation/pulm vascular congestion and potentially hypoperfusion to end organs.
- Qp:Qs < < 1 results in hypoxemia.