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.