

Assessment Points		
System	Physical Examination	Test
RESP	Cyanosed but more “blue” than “sick”	15-30% MetHb
HEME	RCM types I and II: Mild erythrocytosis HbM variants: Mild hemolytic anemia	CBC
CV	May be unable to meet increased metabolic demand	ECG

**Key References:** Jaffe E, Hultquist D: Cytochrome b5 reductase deficiency and enzymopenic hereditary methemoglobinemia. In Scriver C, Beaudet A, Sly W, et al., editors: *The metabolic and molecular bases of inherited disease*, ed 3, vol. 3, New York, NY, 1995, McGraw Hill, pp 3399; Guay J: Methemoglobinemia related to local anesthetics: a summary of 242 episodes, *Anesth Analg* 108(3):837–845, 2009.

**Perioperative Implications**

**Preoperative Preparation**

- Can give reducing agents to pts with RCM type I, but no data exist on whether treatment is indicated before anesthesia.

**Monitoring**

- Pulse oximeter overestimates at low SpO<sub>2</sub> and underestimates at high SpO<sub>2</sub>. In practice, it reads between 80–85%, regardless of true saturation.
- Use co-oximetry for SaO<sub>2</sub> and MetHb levels.
- Monitor ECG for ischemic changes.
- May see “chocolate brown” blood in the operative field or arterial cannula.

**Airway**

- None

**Preinduction/Induction**

- Adequate preoxygenation with 100% O<sub>2</sub> because O<sub>2</sub>-carrying capacity is already decreased.

**Maintenance**

- Prilocaine, benzocaine, and EMLA cream are contraindicated. The literature is contradictory on lidocaine. The effects are probably due to respiratory/myocardial depression in patients with low reserve, rather than an increase in MetHb.
- Nitrous oxide, propofol, and volatile agents are okay.

**Adjuvants**

- None

**Postoperative Period**

- Avoid acetanilides, paracetamol, metoclopramide, and nitrites. Narcotics may be used.

**Anticipated Problems/Concerns**

- Avoid oxidant drugs in both homozygotes and heterozygotes.
- Pulse oximetry is inaccurate; use ABG with co-oximetry.
- May require supplemental O<sub>2</sub> postop.
- May have limited cardiac and respiratory reserve.

## Congenital Pulmonary Cystic Lesions/Lobar Emphysema

Francine S. Yudkowitz

**Risk**

- Cause of cardiorespiratory compromise
- 10–15% associated with CHD

**Perioperative Risks**

- May develop worsening of cardiorespiratory status
- Contamination of unaffected lung by infected material from cyst

**Worry About**

- Associated congenital anomalies
- Tension pneumothorax
- Cardiorespiratory compromise

**Overview**

**Congenital Pulmonary Cystic Lesions (Three Types)**

- Bronchogenic: Abnormal budding and branching of tracheobronchial tree leading to resp distress, pneumonia, and atelectasis

- Dermoid: Lined with keratinized, squamous epithelium
- CPAM: Previously known as CCAM; similar to bronchioles but without alveoli, bronchial glands, and cartilage; overdistension because of gas trapping, which leads to resp distress

**Congenital Lobar Emphysema**

- Hyperinflation and air trapping result in expansion of affected lobe.
- Most commonly occurs in the left upper lobe, followed in frequency by the right middle, and then the right upper lobe.
- Preterm infants on mechanical ventilation develop emphysema in the right upper lobe.
- CXR shows emphysematous lobe crossing midline, mediastinal shift, and atelectasis in other lobes and possibly the contralateral lung. The presence of bronchovascular markings distinguishes this from pneumothorax and congenital cysts.

**Etiology**

- Congenital pulmonary cystic lesions may be bronchogenic, alveolar, or a combination of both, and anomalous development of the bronchopulmonary system occurs.
- Congenital lobar emphysema has extrinsic bronchial obstruction from abnormal vessels or enlarged lymph nodes and intrinsic bronchial obstruction from deficient bronchial cartilage, bronchial stenosis, or redundant bronchial mucosa.

**Usual Treatment**

- Surgical removal

**Assessment Points**

System	Effect	Assessment by Hx	PE	Test
RESP	Decreased lung volume	Cyanosis, dyspnea, grunting, coughing	Tachypnea, retractions, wheezing, decreased BS, asymmetric chest expansion	CXR CT scan
CV	Mediastinal shift, decreased CO, VSD, PDA	Irritability, poor feeding	Decreased heart sounds Murmur	CXR, ECG, ECHO

**Key References:** Hammer G, Hall S, Davis PJ: Anesthesia for general abdominal, thoracic, urologic, and bariatric surgery. In: Davis PJ, Cladis FP, Motoyama EK, editors: *Smith's anesthesia for infants and children*, ed 8, Philadelphia, PA, 2011, Elsevier; Guruswamy V, Roberts S, Arnold P, Potter F: Anaesthetic management of a neonate with congenital cyst adenoid malformation, *Br J Anaesth* 95(2):240–242, 2005.

**Perioperative Implications**

**Preoperative Preparation**

- Assess the severity of cardiopulmonary compromise.
- Identify associated congenital anomalies.
- Optimize resp infection if pt is stable.
- Aspirate cyst before induction if there is cardiac compromise or airway obstruction.

**Monitoring**

- Arterial line for BP monitoring and blood gas analysis

**Induction**

- Avoid positive pressure ventilation until thorax is opened to avoid expansion of cyst or lobe.
- Avoid N<sub>2</sub>O, which will expand the lobe or cyst.
- Inhalation induction with 100% O<sub>2</sub>.
- Intubate without the use of muscle relaxants.
- Affected lung may need to be isolated. In small infants and children, this may be accomplished by using a bronchial blocker or doing a mainstem intubation.

- Surgeon should be available to open the chest immediately if deterioration should occur during induction of anesthesia.

**Maintenance**

- No one anesthetic preferred.
- Maintain spontaneous ventilation or assist with low airway pressures until thorax is opened.
- Once the pathology is removed, N<sub>2</sub>O may be used.
- If Hx of repeated lung infections (cysts), there may be large blood losses.

**Extubation/Postoperative Period**

- Pt may be extubated after uncomplicated surgery and when cardiopulmonary function is adequate.
- Consider regional anesthesia (intercostal or epidural) for management of postop pain to decrease splinting and opioid use.

**Anticipated Problems/Concern**

- Pts with altered cardiopulmonary reserve before surgery may require postop intubation and ventilation.
- If pneumonectomy performed, there will be overinflation of the remaining lung, with a decrease in vital

capacity. These children may have significant exercise intolerance for a prolonged period after surgery.

- Important that pt avoid postop atelectasis, coughing, and early ambulation or increase in activity.
- Altered pulm mechanics (decreased forced vital capacity and delayed forced expiration) may be present throughout childhood.

**Congestive Heart Failure**

Miklos D. Kertai

**Risk**

- Heart failure is a syndrome, not a disease.
- Incidence in USA: About 5.1 million, with more than 650,000 new cases diagnosed annually. Primary discharge diagnosis made in more than 1 million pts.
- 1-y and 5-y survival rates are 57% and 25% in men and 64% and 38% in women. Median survival after onset is 1.7 y in men and 3.2 y in women.

**Perioperative Risks**

- Heart failure occurs in 1–6% of pts after major surgery, and between 6% and 25%, in pts with existing cardiac conditions.
- EF <35% associated with increased operative risk.
- Single greatest risk factor for cardiac surgery. Use congestive heart failure score (CASS): Hx of CHF = 1; Rx digitalis = 1; Rales = 1; overt symptoms after treatment = 1; total 0–4. If score = 4, operative risk is 8× greater.

**Worry About**

- Ventricular dysfunction preop, which is associated with increased operative mortality.
- Pt with diastolic dysfunction may be asymptomatic at rest but sensitive to increases in heart rate, which may result in flash pulm edema.
- Dysrhythmias due to cardiac ischemia (sudden cardiac death).
- Associated acute or chronic mitral insufficiency.
- Volume status.
- Prolonged effect of ACE inhibitors.

**Overview**

- Different types of failure (left vs. right; acute vs. chronic; systolic vs. diastolic; low output vs. high output)
- Reduced contractility, decreased stroke volume, increased heart rate, and hypertrophy and ventricular dilatation
- Acute ischemia, which can lead to global diastolic dysfunction and CHF
- Papillary muscle ischemia, which may lead to severe mitral regurgitation and pulm congestion
- New York Heart Association classification: I, no limitation; II, slight limitation; III, marked limitation; IV, inability to carry out any physical activity; overall 1-year mortality for classes III and IV: 34–58%

**Etiology**

- Acquired, acute or chronic: CHD and MI; cardiomyopathy (idiopathic, hypertrophic, hypertrophic obstructive, congestive, and alcoholic). Valvular heart disease: Arrhythmias and severe hypertension.
- Congenital: Congenital heart disease, left-to-right shunts; intracardiac (ASD, VSD, and AV canal), and extracardiac (PDA and anomalous pulm venous connection). Obstructive (coarctation of the aorta and aortic stenosis). Complex (Ebstein anomaly).
- Multiple precipitating causes: Noncompliance with medications (digitalis and diuretics), excessive Na<sup>+</sup>; excessive IV fluids; drugs (doxorubicin, corticosteroids, disopyramide, nortriptyline, NSAIDs, thiazolidinediones, metformin, cilostazol, PDE-5

inhibitors [sildenafil, vardenafil] androgens, and estrogens). Pulm embolism: High-output states (pregnancy, fever, hyperthyroidism, sepsis, AV fistula, and anemia).

**Usual Treatment**

- Chronic.
- Physical activity encouraged.
- Restriction of sodium intake.
- Chronic, well-titrated beta-blockade may lead to substantial clinical benefit (carvedilol and metoprolol).
- Inhibit RAAS (ACE inhibitors, angiotensin receptor blockers, and aldosterone inhibitors).
- Improvement in systolic heart failure (digitalis).
- Diuretics (hydrochlorothiazide, furosemide, and spironolactone).
- Vasodilators.
- Acute.
- Optimize preload and afterload before starting inotropes and vasodilators.
- Inotropes (dobutamine, epinephrine, milrinone, and amrinone).
- Vasodilators (nitroglycerin, nitroprusside, and nesiritide).
- Maintenance of beta-blocker therapy in acute exacerbation of systolic heart failure.
- Special measures
- Stimulation therapy (biventricular pacing + ICD)
- Surgical correction (CABG, CHD, valvular surgery, cardiomyoplasty, and cardiac transplantation)
- Assist devices (IABP, LV assist, and artificial heart)

**Assessment Points**

System	Effect	Assessment by Hx	Physical Examination	Test
CV	Inadequate cardiac output, congestion	Tachycardia, arrhythmias	Peripheral edema Facial edema (infants/young children), cardiomegaly, pulsus alternans, distended neck veins, Kussmaul sign, abdominojugular reflex	Exercise testing ECG, CXR Circulation time
RESP	Pulm congestion; decreased lung compliance, VC, TLC, pulm diffusion capacity	Breathlessness (exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea) Frequent resp infections	Rales and wheezes Pleural effusions Expectoration: Frothy blood-tinged sputum	PFT ABG CXR
GI	Hepatic and intestinal congestion	Nausea, bloating, fullness	Congestive hepatomegaly, ascites, icterus, cachexia	Liver enzymes
RENAL	Decreased GFR, activation RAAS	Nocturia, oliguria	Ankle edema	BUN/Cr, K <sup>+</sup> , Na <sup>+</sup> , proteinuria, specific gravity
CNS	Hypoperfusion	Confusion and impairment of memory	Mental status exam	
PNS	Increased sympathetic tone	Cool extremities	Peripheral vasoconstriction, pallor, diaphoresis, tachycardia, clubbing	

**Key Reference:** Hammill BG, Curtis LH, Bennett-Guerrero E, et al.: Impact of heart failure on patients undergoing major noncardiac surgery, *Anesthesiology* 108(4):559–567, 2008.

**Perioperative Implications****Preoperative Preparation**

- Stabilize pt by treating CHF before surgery.
- Continue inotropic support.
- Continue cardiac medications (ACE inhibitors may cause hypotension on induction).

**Monitoring**

- Consider arterial line.
- Consider CVP, PA cath, or TEE.
- CVP may be inaccurate in assessing volume.

**Airway**

- Frothy secretions may lead to difficult visualization.

**Induction**

- Preop therapeutic regimen (diuretics) causes hypovolemia, hypokalemia, and hyponatremia, which are potential problems before surgery.
- Replace volume judiciously (avoid dehydration and overhydration).