## Etiology

- Incompletely understood and likely multifactorial. Has been associated with several genes (connexin protein 43, lesion at 11q23.3, cardiac homeobox transcription factor NKX2) and chromosomal abnormalities (Jacobsen syndrome, Turner syndrome, trisomy 18, trisomy 13).

## Usual Treatment

- Series of palliative procedures with the goal of creating reliable systemic and pulm blood flow.
- Stage a connection of systemic venous return directly to pulm artery, dedicating the SV to systemic circulation.
- First, stable blood flow to systemic and pulm circulations are established and balanced.
  - For TA, a BT shunt is placed.
  - For HLHS, a stage I Norwood procedure is performed.
- For other SV lesions, BT shunt or PA banding as dictated by anatomy.
- Complete intracardiac mixing of blood is imperative.
- Stage I Norwood:
  - A neoaorta is created from hypoplastic aortic arch and native PA tissue, connecting the SV to systemic circulation.
  - A BT shunt provides pulm blood flow, connecting branch of neoaorta to ipsilateral pulm artery.
  - An atrial septectomy is performed to ensure complete intracardiac mixing of systemic and pulm venous blood.
- At completion of the stage I Norwood, the SV provides cardiac output to the systemic circulation via the neoaorta and the pulm circulation via the BT shunt.
- The second stage is the first of two procedures to direct systemic venous return to the pulm artery.
  - The SVC is connected to the ipsilateral PA, which remains connected to the PA confluence.
  - This procedure is referred to as a cavopulmonary connection, Bidirectional Glenn or hemi-Fontan, and is commonly performed around 6 mo of age.
  - Low PVR is necessary to promote pulm blood flow, which is passive.
- The final stage, Fontan completion, is typically done 18 mo-5 y.
  - The IVC blood is directed to the ipsilateral PA, either intracardiac via lateral tunnel or extracardiac via graft.
  - This effectively separates the circulations and reduces volume workload on SV; systemic venous return now flows passively to the PA without interposed pumping chamber.
  - A small fenestration from the IVC-PA conduit to the atrium is sometimes created. The fenestration ensures preload to the systemic circulation even when PA pressures fluctuate, maintaining cardiac output but at the expense of decreased $O_2$ sat via right-left shunt.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| CV | CHF | Dyspnea, tachypnea, feeding difficulties | $S_3$, rales, wheeze, enlarged liver, metabolic acidosis | CXR, pulse oximetry, ABG<br>ECHO |
| | Hypoxia<br>Arrhythmia | Dyspnea, tachypnea, feeding difficulties<br>cyanosis | Cyanosis | CXR, pulse oximetry, ABG<br>ECG |
| HEME | Polycythemia | See above | See above | Hgb, Hct |

**Key References:** Barron DJ, Kilby MD, Davies B, et al.: Hypoplastic left heart syndrome, *Lancet* 374(9689):551–564, 2009; Yuki K Casta A, Uezono S: Anesthetic management of noncardiac surgery for patients with single ventricle physiology, *J Anesth* 25(2):247–256, 2011.

## Perioperative Implications

### Preoperative Preparation

- Depending on the stage of the palliative process (Norwood stage I, Glenn/hemi-Fontan, completion Fontan), optimize hemodynamics.
- Cardiac catheterization is typically performed prior to Glenn/hemi-Fontan to measure PVR and coil any collateral venous vessels.
- Higher $O_2$ sat can decrease $O_2$ delivery to the tissues by facilitating overcirculation to the lungs, particularly when pulm blood flow is via BT shunt.

### Monitoring

- Arterial BP.
- CVP monitoring via IJ is controversial due to SVC thrombosis risk and implications for subsequent staging, which requires patency of these vessels.
- Consider TEE.

## Preinduction/Induction

- Dependent on exact anatomy and stage of palliation.
- Induction technique should consider impact of PVR and SVR changes on myocardial, systemic, and pulm blood flow.

### Airway

- ET intubation and PPV.
- Minimize intrathoracic pressures where possible to encourage pulm blood flow.

### Maintenance

- IV or inhalational agents are acceptable.
- Body temperature as dictated by potential use of cardiopulmonary bypass.

### Extubation

- Following stage I Norwood, pt requires mechanical ventilation for >2 d.
- Early extubation is recommended to facilitate pulm blood flow after stage II (Glenn or hemi-Fontan) or the completion Fontan. High intrathoracic pressure from PPV impedes venous flow to the pulm circulation, while negative intrathoracic pressure (spontaneous respiration) enhances flow.

## Anticipated Problems/Concerns

- Overcirculation.
- Hypoxemia.
- New anatomy postprocedure will necessitate a reassessment of desired PVR and SVR to optimize flow to both circulations.
- Postop low cardiac output syndrome.

# Sleep Apnea, Central and Mixed

Michael F. Roizen | Andreas M. Ostermeier

## Risk

- Incidence USA is 3–12% of middle-aged adults (which has increased fourfold in last 15 y, presumably due to increase in obesity). The M:F ratio is 2–2.5:1; obstructive or mixed.
- Risk increases with male sex, upper middle age (55–64 y), obesity, and Hx of snoring with impaired daytime performance.
- In elderly, risk is 2× higher for African Americans.

## Perioperative Risks

- Increased risk of central and mixed (central and obstructive) apnea. In mixed SAS, obstructive apnea component can mask central apnea.
- Risk for respiratory depression also in intubated, tracheotomized, and awake pts.
- Increased risk with sedative-hypnotic narcotics, postop with any form of pain relief.

## Worry About

- See medical records for previous problems.
- Look for related medical disorders (e.g., cor pulmonale, cardiac arrhythmias, erythrocytosis, disordered cognition, daytime somnolence).
- Apnea possible even several h postop, especially after epidural anesthesia.
- When administering $O_2$, think of possible dependence of ventilation on hypoxic drive.

## Overview

- Central sleep apnea implies failure of respiratory rhythmogenesis. In SAS pts, at least 30 periods of apnea, defined as cessation of airflow for ≥10 sec, are found during normal nocturnal sleep.
- Obstructive sleep apnea relates to a failed or inadequate respiratory activation of upper airway muscles, resulting in lack of airflow.
- In central apnea, hypoventilation persists despite relief of obstruction.
- Central apnea is unaccompanied by any respiratory effort, in contrast to obstructive sleep apnea.
- Related to central alveolar hypoventilation syndrome, also known as Ondine curse.

## Etiology

- Central: Familial basis is evident in some cases; possible relation to neurologic disorders (e.g., encephalitis in childhood, damaged respiratory centers, autonomic neuropathy in diabetes)

- Mixed: Has obstructive component. Upper airway narrowing superimposed on coexistent abnormality of neurologic control or function of upper airway muscle tone or ventilatory control.
- Associated with obesity and nasal obstruction (polyps, rhinitis, deviated septum, acromegaly, hypothyroidism, Htn).

## Usual Treatment

- CPAP or BiPAP; bring to hospital and OR/PACU.
- Tracheotomy and mechanical vent at night.
- Diaphragmatic pacing, especially at night.
- Surgery to remove obstruction
- For central/mixed apnea, additional medical treatment with protriptyline, progesterone.
- For mixed apnea, weight loss and physical aids.
- Avoid narcotics, benzodiazepines, and alcohol.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Obstructive apnea | Snoring, partner gives Hx of pt's awakening at night with grunts | Visualization of uvula and tonsillar pillars | |
| CV | Htn | Dyspnea at rest, DOE<br>Poor exercise tolerance, angina | Cardiomegaly<br>$S_3/S_4$ murmur | ECG, ECHO |
| RESP | Right-sided heart dysfunction, snoring, respiratory dysfunction, DOE | Awakening at night with grunts | Venous engorgement<br>Rapid respiratory rate<br>Cardiomegaly | $SaO_2$ supine<br>ECG, CXR, ABG, Hct<br>Polysomnogram, home sleep study |
| GI | Hepatic dysfunction<br>Full stomach<br>T2DM | Jaundice, bleeding disorders, ascites, heartburn, hiatus hernia, polydipsia, polyuria | Hepatomegaly, ascites, spider nevi, jaundice | LFTs, PT, PTT<br>Fasting glucose |
| ENDO | Obesity<br>Hypothyroidism<br>Acromegaly | | Mental function reflexes<br>BMI | Free $T_4$ estimate<br>TSH, GH levels |
| HEME | Polycythemia | | Plethora, clubbing, cyanosis | $O_2$ sat, Hct |
| CNS | Disturbed sleep, impaired daytime performance, morning headache, memory problems, irritability | Daytime sleepiness, complaints of disrupted sleep<br>Ask for encephalitis, autonomic neuropathy, brainstem damage | | Polysomnogram, home sleep study |

**Key References:** Ostermeier AM, Roizen MF, Hautkappe M, et al.: Three sudden postoperative arrests associated with epidural opioids in patients with sleep apnea, *Anesth Analg* 85(2):452–460, 1997; Somers VK, White DP, Amin R, et al.: Sleep apnea and cardiovascular disease: an American Heart Association/American College of Cardiology Foundation Scientific Statement from the American Heart Association Council for High Blood Pressure Research Professional Education Committee, Council on Clinical Cardiology, Stroke Council, and Council on Cardiovascular Nursing. In collaboration with the National Heart, Lung, and Blood Institute National Center on Sleep Disorders Research, *Circulation* 118(10):1080–1111, 2008; American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea: Practice guidelines for the perioperative management of patients with obstructive sleep apnea: an updated report by the American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea, *Anesthesiology* 120(2):268–286, 2014.

## Perioperative Implications

### Preoperative Preparation

- Take sleep Hx, if possible, from bed partner.
- If a question of sleep apnea, use home sleep apnea tests (helmet or wrist; several distinct types now exist) as a screen. Sleep lab unnecessary for this screen. If positive, refer to sleep lab preop.
- Avoid preop sedation with benzodiazepines and narcotics.
- Examine airway carefully.
- Consider metoclopramide 10 mg, cimetidine 300 mg PO the night before and IV preop.
- Assess myocardial and volume status.
- Initiate CPAP or BiPAP therapy over periop period, and in recovery room.

### Monitoring

- Routine; consider arterial line.
- UO, possible CVP or PA catheter, if volume status likely to be significantly altered.

### Airway

- Airway control necessary if prominent central component and sedation mandatory.
- Awake, sitting, fiberoptic intubation may be indicated if difficulty anticipated.

### Induction

- Pt may need to remain semisitting if $SaO_2$ drops when supine. Preoxygenation should be complete throughout lungs.

### Maintenance

- Oxygenation may deteriorate with upper abdominal surgery or increased intraabdominal pressure.
- Consider the use of short-acting substances (e.g., propofol, remifentanil).
- Minimize postop sedation.

### Extubation

- Extubate as soon as pt maintains normocapnia and responds to command.
- Consider close monitoring after extubation.

### Adjuvants

- Initial dose of induction agent and narcotics calculated on a mg/kg basis, and muscle relaxants calculated on estimated lean body mass.
- Subsequent doses of sedatives, hypnotics, relaxants, and narcotics calculated on estimated lean body mass.
- RA if physically possible and if pt can use accessory muscles to help with breathing.

### Postoperative Period

- Pain control with opioids only when NSAIDs and/or RA is contraindicated and/or insufficient, as (sudden) complete pain relief may increase risk of respiratory arrest.
- Some think epidural or narcotics are indicated, and others think these are relatively contraindicated.
- Extended respiratory monitoring.
- Stabilize ABG to adequate levels.
- Pain control necessary; PCA acceptable in sleep apnea, but not in continuous mode.
- Consider DVT prophylaxis if pt is overweight.

## Anticipated Problems/Concerns

- Respiratory insufficiency and pneumonia postop; use devices and/or CPAP in immediate and long-term preop and postop periods.
- Postop thromboembolic phenomena.
- If problems occur, inform pt before discharge with written instructions, especially for further anesthetic interventions.

# Sleep Apnea, Obstructive

Michael F. Roizen | Charles Ahere | Claude Brunson

## Risk

- Incidence in USA is 3–15% of the whole population (increased fourfold in last 15 y, presumably due to increase in obesity).
- M:F ratio: 2.5:1.
- Race with highest prevalence: Unknown.

## Perioperative Risks

- Increased risk of pulm Htn, RV failure, and systemic Htn.
- Some pts may be polycythemic and have an increased risk of CVA.
- Complications associated with obesity and craniofacial and upper airway soft tissue abnormality.
- Increased risk in supine position of sudden arrest postop.