- Pressure gradients may be low despite severe AS in pts with reduced EF (low-flow/low-gradient AS) or small ventricular volumes (low-flow/low-gradient AS with preserved EF).

## Etiology

- Congenital bicuspid (and rarely unicuspid) aortic valve
- Rheumatic aortic stenosis
- Calcific degenerative disease

## Usual Treatment

- Early in the disease process, medical therapy is indicated, including lifestyle modification (e.g., smoking cessation, exercise), and judicious use of antihypertensives.
- In severe AS, medical therapy does not prolong life, and AVR is the only effective treatment.
- AVR is a Class I indication in pts with
  - Symptomatic severe AS.
  - Asymptomatic severe AS with a LVEF <50%.
  - Asymptomatic severe AS in pts undergoing other cardiac surgery.
- AVR is a Class IIa indication in pts with:
  - Asymptomatic severe AS and low surgical risk.
  - Asymptomatic severe AS and decreased exercise tolerance or fall in BP with exercise.
  - Symptomatic low-flow/low-gradient severe AS.
  - Moderate AS in pts undergoing other cardiac surgery.
- TAVR is now a commonly used alternative to open AVR in pts who pose a high surgical risk due to age, comorbidities, and/or previous cardiac surgery.
- Balloon valvuloplasty may be used as a bridge to definitive surgical treatment with open AVR or TAVR.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| CV | Severe aortic stenosis<br>Myocardial ischemia<br>Diastolic dysfunction<br>Arrhythmias | Angina, dyspnea, syncope<br>Angina<br>Dyspnea<br>Palpitations, syncope | Systolic murmur<br>Rales, edema, wheeze<br>Rales, edema, JV distention<br>Irregular pulse on exam | ECHO, cardiac cath, and dobutamine stress ECHO in suspected low-gradient AS<br>ECG, ECHO, coronary angiography<br>CXR, ECHO<br>ECG, Holter |
| CNS | Syncope | Syncope | | ECG, Holter, ECHO |

**Key References:** Rashedi N, Otto C: Aortic stenosis: changing disease concepts, *J Cardiovasc Ultrasound* 23(2):59–69, 2015; Cook D, Housmans P, Rehfeldt K: Valvular heart disease: replacement and repair. In Kaplan J, editor: *Kaplan's cardiac anesthesia: the echo era,* ed 6. St. Louis, 2011, Elsevier, pp 570–584.

## Preoperative Preparation

- Adequate premedication to reduce preop tachycardia due to anxiety.
- Ensure adequate preload but beware of administering large amounts of volume rapidly due to diastolic dysfunction.
- Pts with severe symptomatic AS may benefit from postponement of elective surgery until after AVR is performed.

**Monitoring**

- ECG for ST segment analysis.
- Preinduction invasive arterial pressure monitoring.
- Pulm artery cath may be useful in major surgery with large fluid shifts to better assess LVED pressure and volume status.
- Transesophageal ECHO warranted when blood loss or volume shifts are anticipated and an experienced echocardiographer is available.

**Airway**

- None

**Preinduction/Induction**

- Narcotic heavy induction is beneficial due to bradycardia, maintenance of SVR, and blunting of sympathetic response to laryngoscopy.
- Alpha agonist like phenylephrine should be used to treat hypotension with induction.
- Judicious use of propofol (and co-administration of an alpha agonist) is warranted to limit drop in SVR. Etomidate may also be useful for this reason.
- Laryngoscopy only after sufficient sympathetic attenuation.

**Maintenance**

- Intraop fluid management should aim for maintaining already elevated left-sided filling pressures with adequate replacement of blood loss and insensible losses.
- Balanced anesthetic using narcotics, muscle relaxant, and a lower dose of volatile agent is preferred.
- Higher doses of volatile agents may depress cardiac function, increase risk of arrhythmia-induced hypotension, and drop SVR, leading to hypotension and myocardial ischemia.
- Caution with agents that decrease preload and afterload (e.g., nitroglycerin, nitroprusside), or any agent with significant histamine release.
- Caution with agents that directly or indirectly increase heart rate (e.g., pancuronium, atropine).
- Early electrical cardioversion for intraop atrial fibrillation.
- Generally, avoid neuraxial anesthesia (especially spinal) due to hypotension from sympathectomy. Epidural anesthesia may be used with extreme caution in laboring pts and other cases where its benefit is strong, but must be carefully dosed for a gradual onset of block with minimal drop in SVR, while simultaneously augmenting preload and administering vasoconstrictors when needed.

**Extubation**

- Minimize sympathetic stimulation and tachycardia.

**Postoperative Period**

- Aggressive pain control
- Maintenance fluids as appropriate to maintain adequate preload

## Anticipated Problems/Concerns

- Myocardial ischemia with intraop hypotension.
- Diastolic dysfunction.
- Dysrhythmias can lead to precipitous hypotension and ischemia and should be treated aggressively until a return to sinus rhythm is achieved or hemodynamics stabilize.

# Apert Syndrome (Acrocephalosyndactyly Type 1 and 2)

Andrea Johnson

## Risk

- 15:1,000,000 live births
- Equal M:F ratio

## Perioperative Risk

- Aspiration
- Bronchospasm
- Resp depression
- Airway obstruction

## Worry About

- Difficult mask, airway, or IV access
- Elevated intracranial pressure, temperature dysregulation, and seizures
- Corneal abrasions (due to exophthalmos)
- PACU and perioperative monitoring for apnea
- Cardiac anomalies (10% of cases)
- Anatomic anomalies (regional/neuraxial anesthesia)

## Overview

- Apert syndrome is a disorder identified by synostoses of the cranium, vertebral bodies, and digits. It is caused by a mutation in the FGFR-2 gene.
- Two major manifestations are bicoronal synostosis and maxillary hypoplasia. High-arched V-shaped palates and cleft palates are common.
- Strabismus, syndactyly, and conductive and neuronal hearing loss are common manifestations.
- Cognitive delay (IQ <70) see in 67% of pts.

## Etiology

- Autosomal dominate disorder; however, most cases are sporadic mutations of the FGFR-2 gene.
- Sporadic mutations are associated with paternal age >40 y.

## Usual Treatment

- Craniosynostosis release: Frontoorbital advancement usually around 6 to 8 mo of age
- Midface advancement: Correction of brachycephaly, orbital dystopia, or midface hypoplasia
- Correction of hypertelorism: Interorbital bone resection
- Mandibular and maxillary advancement and orthodontics: Usually conducted after cranial maturation to enhance cosmetic appearance

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| CNS | CNS malformations<br>Ventriculomegaly<br>Hydrocephaly<br>Elevated intracranial pressure<br>Thermoregulatory disorders<br>Cognitive delay | Developmental delay, nausea/vomiting, headache | Papilledema | CT/MRI |
| HEENT | Craniosynostosis, midface hypoplasia, nasopharyngeal, and palatal anomalies | Dyspnea<br>Difficulty phonating | Early fusion of cranial sutures<br>Hypertelorism<br>Cleft or V-shaped palate | Radiologic studies |
| CV | Atrial septal defect/ventricular septal defect<br>Patent foramen ovale<br>Overriding aorta | Dyspnea<br>Lethargy | Heart murmur | ECHO |
| RESP | Central/obstructive sleep apnea<br>Aspiration<br>Bronchospasm<br>Increased airway secretions | Daytime somnolence<br>Snoring<br>Witnessed apnea<br>Coughing<br>Wheezing | Wheezing<br>Course breath sounds<br>Increased oral secretions | Sleep study |
| MS | Cervical spine abnormalities (usually fusion at C5-C6)<br>Syndactyly | Limb abnormalities | Decreased cervical ROM | Radiographic studies |

**Key References:** Niraj K, Shubhangi A, Ashish B, et al.: Anesthetic management of craniosynostosis repair in patient with Apert syndrome, *Saudi J Anaesth* 8(3):399–401, 2014; Losee JE, Gimbel ML, Rubin J, Plastic and reconstructive surgery. In Brunicardi F, Andersen DK, Billiar TR, editors: *Schwartz's principles of surgery*, ed 10, New York, 2014, McGraw-Hill.

## Perioperative Management

### Preoperative Consideration

- Review imaging to assess for increased ICP and cardiac and airway anomalies.
- Preoperative warming measures.
- Minimize risk of bronchospasm; consider antisialagogue and beta-2 agonist.
- Anticipate difficult IV scenario.
- Discuss with surgical team the risk for admission, especially with a history of sleep apnea.

### Monitoring

- Standard ASA monitors
- Arterial line if indicated
- Core temperature when possible

### General Anesthesia

- Maintain normothermia.
- Multimodal approach to pain management to minimize opiates.
- Consider intermittent gentle suctioning through ETT to prevent mucus plugs.

### Regional Anesthesia

- Due to anatomic anomalies with bone and soft tissues, consider performing all regional techniques under ultrasound.
- Exercise caution with regional techniques known to interrupt innervation of the diaphragm or cough reflexes.

### Postoperative Period

- Anticipate prolonged postop ventilation.
- Exercise caution with sedating medications for pain control.
- Monitor and treat for bronchospasm or laryngospasm in PACU.

## Anticipated Problems/Concerns

- Anticipate prolonged monitoring or hospitalization for pts with history of sleep apnea.
- Anticipate bronchospasm, especially during periods of light of anesthesia.
- Suction airway and ETT judiciously before extubation and weigh risk/benefit of deep extubation.

# Apnea of the Newborn

Shanique Brown Kilgallon | Alan Jay Schwartz

## Risk

- Full-term infants with an underlying pathology (i.e., neurologic disorders, metabolic derangements)
- Premature infants, with or without an underlying pathology
- Infants less than 60 wk post conceptual age
- Underweight infants <1000 g
- Anemia

## Perioperative Risks

- More prone to apnea during local or neuraxial anesthesia or when additionally administered IV sedative
- More prone to apnea after general anesthesia

## Worry About

- Unexpected apnea in recovery room
- Unexpected apnea in hours after outpatient procedures
- Unexpected apnea on ward hours after inpatient procedures

## Overview

- Apnea is defined as pauses that last >20 sec without physiologic derangement or that last >10 sec with physiologic derangement (i.e., bradycardia, oxygen desaturation).
- Apnea in term infants is never physiologic.
- Apnea in preterm infants may signal CNS disorder or developmental immaturity.
- Sudden onset of apnea in any infant may also reflect a new-onset sepsis or hypoglycemia.
- Utility of pneumogram screening controversial.
- Indications for home apnea monitoring controversial.

## Etiology

- Term or preterm infants:
  - CNS disorders (seizures, bleeds, and structural changes)
  - Systemic disorders (hypoglycemia, sepsis, and GE reflux)
- Preterm infants:
  - Same as term infants
  - If full evaluation is negative, physiologic apnea of prematurity diagnosed

## Usual Treatment

- Theophylline or caffeine
- $O_2$
- Transfusion
- CPAP