

Perioperative Implications**Preoperative Preparation**

- Airway evaluation, pulm function assessment; consider positioning difficulties.
- Antisialagogue for awake intubation.
- Review MRI of the spine.

Monitoring

- ST-segment analysis; pulm artery cath if severe myocardial dysfunction
- Arterial line, central venous access for extensive osteotomy secondary to blood loss

Airway

- Difficult intubation possible, owing to cervical spine fusion or distortion; fiberoptic intubation may be necessary; cervical spine instability possible; spinal fracture possible with airway manipulation; occult spinal fracture may already be present.
- Increasing role for videolaryngoscopy.

Induction

- If general anesthesia, any approach is acceptable. If limited cardiac reserves, avoid depressants of myocardial contractility. If regional, skeletal abnormality can make the block difficult to perform, and response to injection is unpredictable. In some cases, epidural space is obliterated and cannot be completely accessed. If local anesthetic toxicity, airway management can be difficult.

Maintenance

- With positive pressure ventilation, decrease tidal volume and increase rate. Consider pressure support ventilation.
- High ventilating pressure may predict large blood loss.

Extubation

- Awake is preferable.
- Airway edema is possible after extensive anterior osteotomy, decompression, and/or fusion. Compression of the airway from retropharyngeal hematoma is possible. Consider leak test before extubation, or maintaining the pt intubated and sedation for 12-24 h postop. Consider extubation over tube exchanger.

Adjuvants

- Ischemic optic neuropathy with prolonged procedures in the prone position

Postoperative Period

- Comfortable pt position and pain control without airway obstruction

Anticipated Problems/Concerns

- Airway control
 - The extreme distortion of the spine, especially the neck, may make intubating the trachea and ventilating the pt very difficult.

- Any airway compromise or depression of ventilation can result in catastrophe.
- Depression of ventilation with opiate analgesics can be dangerous.
- Pulm function
 - Owing to abnormal mechanics of the thorax and neck, the ability to ensure normal oxygenation during surgery and in the postop period can be a potential problem.
- Regional anesthesia
 - Placement of spinal, epidural, or caudal block could be technically very difficult. Action of local anesthetics in the central axis could be unpredictable. Consider preop x-rays of the lumbar spine to facilitate access for neuraxial block.
 - Strongly consider paramedian approach to central block.
- Prolonged postop intubation
 - Substantial blood loss, fluid/blood product administration, and the prone position make airway edema likely, requiring extended postop intubation. Pt should be informed preop to avoid panic postop.

Anomalous Pulmonary Venous Drainage

Roger A. Moore

Risk

- One percent of all congenital heart defects.
- TAPVD, the severe form, or PAPVD, the less severe form, exists when pulm veins drain into the venous circulation.
- M:F 4:1 in infradiaphragmatic type.

Perioperative Risks

- Rapid CV deterioration secondary to hypercapnia and resultant acidosis
- Sudden pulm Htn and RHF during hypoventilation
- Periop mortality: 2-20% depending on preop status

Worry About

- Air bubbles entering the venous circuit
- Endocarditis risk
- Concurrent pneumonia with hypoxemia or hypercarbia

- Polycythemic hyperviscosity attack with:
 - Periop dehydration
 - Cold OR environment

Overview

- TAPVD incompatible with life unless an ASD allows adequate R-to-L shunting of blood. TAPVD pts with small ASDs are more critically ill and often require balloon septostomy as a bridge to surgery. Some cyanosis, usually with O₂ saturations of 85-95%.
- Increased flow through pulm vascular beds results in pulm Htn.
- Four types of TAPVD:
 - Supracardiac: Pulm veins connect to the left innominate vein via an anomalous "vertical vein" or connect to the right SVC via an anomalous "short connecting vein," or connect to the left SVC (45%).

- Cardiac: Pulm veins drain into the coronary sinus or directly into the right atrium (23%).
- Infracardiac: Pulm veins drain into IVC, portal veins, hepatic veins, or ductus venosus (21%).
- Mixed: Combined supracardiac, cardiac, and infracardiac connections (11%).

Etiology

- Embryologic atresia or malformation of the common pulm venous system resulting in persistence of abnormal connections

Usual Treatment

- Severe TAPVD with little systemic shunt needs immediate cardiac correction after birth. Most children with TAPVD require cardiac correction before 1 y of age.
- Cardiac correction of PAPVD may be postponed into childhood.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Hypoxemia	Snoring	Airway class	
CV	CHF Hypoxemia Monitoring problems	Decreased activity level Dyspnea Anomalous peripheral vessels	Rales Cyanosis Pulses and blood pressures in all four extremities	ECG: RVH, RAH ECHO, cath Cardiac consultation
RESP	Hypoxemia	Bronchospasm SOB Pulm edema Exertional cyanosis	Wheezing Tachypnea Clubbing	CXR Granular lung fields
HEME	Sludging DIC	Polycythemia Bleeding or bruising	Clubbing Bruises	Hgb PT PTT, bleeding time
CNS		Previous stroke	Complete neurologic evaluation	CT scan if neurologic findings
MS		Feeding difficulty Failure to thrive	Ht, wt, head circumference	Plot of growth curves

Key References: Müller M, Scholz S, Maxeiner H, et al: Efficacy of inhaled iloprost in the management of pulmonary hypertension after cardiopulmonary bypass in infants undergoing congenital heart surgery. A case series of 31 patients, *HSR Proc Intensive Care Cardiovasc Anesth* 3(2):123-130, 2011; Young TW: Anomalous pulmonary venous return. In Moodie DS, editor: *Clinical management of congenital heart disease from infancy to adulthood*, Minneapolis, 2014, Cardiotext, pp.77-92.

Perioperative Implications**Preoperative Preparation**

- Desired hemodynamics: Preload, normal (CVP 10-12 mm Hg); afterload, low; PVR, normal; HR, normal to high; contractility, normal
- Liberal oral fluids preop
- Avoid premedication causing hypoventilation
- Subacute bacterial endocarditis prophylaxis

Monitoring

- Absolute air bubble precaution
- Arterial cath
- CVP cath; know specific anatomy, incl SVC variations
- TEE
- Others as per ASA routine

Airway

- Associated congenital syndromes with airway anomalies
- Cricoid ring limiting airway diameter
- Primary need to maintain airway and avoid increased PaCO₂
- PEEP, with pulm edema or elevated pulm blood flow

Induction

- If IV in place, use fentanyl or ketamine with pancuronium, vecuronium, or rocuronium.
- If no IV:
 - If unstable, ketamine IM.
 - If stable, slow inhalational induction with sevoflurane (avoid high sevoflurane levels until IV placed).
- Actively avoid hypoventilation and agents that produce myocardial depression.

Maintenance

- Use fluids judiciously to avoid RV overload.
- Positive pressure ventilation usually improves oxygenation.
- Use narcotics in conjunction with inhalational agents as tolerated.
- Avoid nitrous oxide.
- Use high FiO₂
- Capnographic ET/CO₂ will not accurately reflect PaCO₂
- Prepare for hypothermic cardiac arrest during TAPVR repair.
- Avoid hypothermia before and after bypass.

Extubation

- Do not attempt deep or early extubation.
- Before extubation, assess adequacy of ventilation (with insp pressures of at least -20 mm Hg) and tidal volumes.

Postoperative Period

- Close monitoring of ventilation and pulse oximetry.
- Active warming with avoidance of shivering.
- Be prepared for immediate reintubation.

Adjuvants

- Inotropic support with dopamine or epinephrine

Anticipated Problems/Concerns

- If pulm hypertensive crisis occurs:
 - Hyperventilate.
 - 100% inspired O₂
 - Consider iloprost, prostaglandin E₁, tolazoline, amrinone, isoproterenol, or nitric oxide.

Anorexia Nervosa

Russell T. Wall III

Risk

- Primarily in white adolescent females from middle- or upper-class families; 4% to 10% males.
- More common in models, ballet students, and professions demanding high achievement.
- Occurs in 5-10 per 100,000 population; mortality rate 5-10%.
- Bimodal peak age of onset: 14 and 18 y.

Perioperative Risks

- Predisposing conditions include:
 - CV dysfunction (bradycardia, hypotension, and dysrhythmias).
 - Acid-base abnormalities (both metabolic acidosis and alkalosis are possible), lyte abnormalities (decreased K, Mg, NA, and P)
 - Hematologic abnormalities (decreased Hgb, WBC, fibrinogen, and plt).
 - Hypothermia, delayed gastric emptying, and renal dysfunction (prerenal azotemia).
- Lyte/nutrient abnormalities associated with refeeding: most dangerous is hypophosphatasia (but also thiamine deficiency and decreased K, Mg, NA, and P).

Worry About

- Degree and duration of malnutrition (excess protein depletion = impaired cellular function)

- Degree of organ dysfunction
- Greater weight loss = greater risk
- Refeeding syndrome (severe hypophosphatasia occurred in 0.5% in largest modern study)

Overview

- Anorexia nervosa
 - Obsessive fear of obesity; pursuit of thinness
 - Dramatic decrease in food intake and excessive physical activity
 - Refusal to maintain weight above 85% IBW
 - Distorted body image
 - Amenorrhea for >3 mo
 - Radical restriction of caloric intake
 - Appears cachectic
 - Risk of death high if weight loss >40% of IBW
 - Of patients, 40% to 50% recover with treatment; 20% to 30% improve with treatment
- Bulimia
 - Means "ox hunger" or voracious appetite
 - Obsessive fear of obesity; overconcern with body shape and weight
 - Appears well nourished
 - Averages two binge-eating episodes each wk for at least 3 mo
 - Irresistible urge to overeat; loss of control in desire to eat

- Wt control by self-induced vomiting, diuretic and laxative use, strict dieting/fasting, vigorous exercise
- Greater percent of alcohol use, illicit drug use, stealing, self-mutilation, and suicide attempts than with anorexia
- Of patients, 30% to 60% recover with treatment

Etiology

- Unknown, but possibly hypothalamic dysfunction or psychiatric cause

Usual Treatment

- No specific/definitive treatment
- Therapies offered:
 - Psychotherapy (individual, group, and family)
 - Behavior modification
 - Antidepressants (TCAs, MAO inhibitors, serotonin-uptake inhibitors) often prescribed but not consistently effective
 - Nutrition counseling (1500 to 2500 calories/d, metoclopramide or bethanechol for gastric emptying, benzodiazepine before meals)
 - Relaxation exercises
- If severe: Hospitalization stressing weight gain, with tube feedings or hyperalimentation as last resort