Assessment Points

System	Effect	Assessment by Hx	Test
HEENT	Airway anomalies	Snoring Difficult breathing	
RESP	Decreased mucus	Repeated infections	
ANS	Delayed gastric emptying Hemodynamic instability	Vomiting Hypotension/bradycardia	
OPHTHAL	Decreased lacrimation	Dryness, ulceration	
METAB	Hyperpyrexia		Record/monitor temp

Key References: Zlotnik A, Natanel D, Kutz R, et al: Anesthetic management of patients with congenital insensitivity to pain with anhidrosis: a retrospective analysis of 358 procedures performed under general anesthesia, *Anesth Analg* 121(5):1316–1320, 2015; Zlotnik A, Gruenbaum SE, Rozet I, et al: Risk of aspiration during anesthesia in patients with congenital insensitivity to pain with anihidrosis: case reports and review of the literature, *J Anesth* 24(5):778–782, 2010.

Perioperative Implications

Preoperative Preparation
- Avoid anticholinergic premedication; however atropine has been used to treat bradycardia.

Monitoring
- Routine
- Temp

Airway
- Full stomach precautions.
- Awkward mask fit.
- Laryngoscopy and intubation may be difficult.

Maintenance
- Regional anesthesia may be preferable when possible.
- Humidify anesthetic gases.
- Control room temp to avoid hyperthermia.

Extubation
- Vigorous postop chest physical therapy

Adjuvants
- Protect eyes with tape and ophthalmic ointment (higher risk for corneal abrasion).

Anticipated Problems/Concerns
- Difficult airway (mask and/or intubation)
- Hyperthermia
- Postop chest infections
- Regurgitation/vomiting/aspiration
- High incidence of CV events (hypotension and bradycardia) reported

Ankylosing Spondylitis

John E. Tetzlaff

Risk
- 1:2000 incidence in Caucasians; rare in non-Caucasians
- M:F 10:1; more severe in males
- 18-50% incidence in Native Americans

Perioperative Risks
- Difficult airway and atlantoaxial instability
- "Bamboo spine" with potential for fracture during airway manipulation
- Rigid chest with difficult ventilation
- Myocarditis and myocardial conduction defects
- Increased blood loss due to abnormal chest structure or mechanics

Worry About
- Inability to intubate, spine fracture, arrhythmia, inability to ventilate, and massive blood loss
- Airway edema after extubation

Overview
- An arthritic process, seronegative for rheumatoid factor, which attacks ligamentous attachments of the spinal column
- Characterized by low-back pain, sacroiliitis, multiplane rigidity of the spine, chest stiffness, uveitis, and insidious onset at <40 y of age
- Autosomal dominant and strongly prevalent among first-degree relatives

Etiology
- Etiology unknown
- Genetic transmission led to discovery of a genetic marker, HLA-B27. Also involved are the major histocompatibility complex, numerous HLA-B27 subtypes, IL23R (also associated with ulcerative colitis), and ERAP-1.
- Infectious origin speculated; one species of *klebsiella* is reported to be associated with some cases.

Usual Treatment
- Symptomatic, with exercise and NSAIDs; Immunosuppression can be tried in severe cases.
- Wedge osteotomy is a drastic surgical intervention.
- Infliximab: monoclonal antibody specific for TNF.
- Etanercept: Anti-TNF protein.
- Adalimumab: Monoclonal antibody specific for TNF.

Assessment Points

System	Effect	Assessment By Hx	PE	Test
HEENT	Uveitis TMJ arthritis Arytenoid deviation	Visual disturbance Limited mouth opening, jaw pain, voice abnormality	Funduscopic exam Airway exam Indirect laryngoscopy	Fiberoptic nasopharyngoscopy
CV	Cardiomyopathy, conduction defects	SOB, chest pain, palpitation	Distant heart sounds, rales, arrhythmia	ECG, CXR, ECHO
RESP	Pleuritic inflammation, chest rigidity	Chest pain, limited exercise tolerance	Decreased breath sounds, chest excursion	PFTs, CXR
GI	Irritable bowel syndrome Ulcerative colitis	Abdominal pain, bowel dysfunction	Abdominal pain	
GU	Chronic prostatitis	Pain with urination	Rectal exam	
CNS	Atlantoaxial subluxation, occult spinal fracture	Long tract signs, sphincter abnormality; sometimes no symptoms	Basic neurologic exam	Cervical spine x-ray with flexion-extension, MRI
PNS	Radiculopathy	Radiating pain in extremities	ROM of the extremity	EMG (medicolegal use)
MS	Back pain, sacroiliitis, joint ankylosis, kyphosis ("chin on chest"), "bamboo spine," spondylodiscitis	Review of skeletal function	Spine, skeleton	Radiologic studies

Key References: Hu SS, Ananthakrishman D: Ankylosing spondylitis. In Rothman RA, Simeone FA, editors: *The spine,* ed 5, Philadelphia, 2011, Elsevier; Schlew BL, Vaghadia H: Ankylosing spondylitis and neuraxial anesthesia: a 10 year review, *Can J Anesth* 43(1):65–68, 1996.

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 O_2 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.