

- Coronary vasospasm.
- Inadequacy of myocardial O₂ supply secondary to increased demand from ventricular hypertrophy.
- Endothelial cell-mediated vasoconstriction.
- Thrombosis overlying unstable plaque can lead to unstable angina/MI.

Etiology

- Acquired disease with genetic predisposition.
- Pts with diabetes have higher incidence of CAD, which is frequently silent.
- Other risk factors include Htn, hyperlipidemia, advanced age, tobacco use, and homocysteinemia.

Usual Treatment

- Medical therapy: β -adrenergic receptor antagonist, Ca²⁺-channel antagonists, nitrates, aspirin, P2Y₁₂ inhibitors, folate, lipid-reducing agents, and combination agents
- Percutaneous coronary interventions with stent placement
- CABG

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Myocardial ischemia LV dysfunction	Angina Sx Angina-equivalent Sx Dyspnea Exercise tolerance	Displaced posterior maximal impulse S ₃	ECG Exercise ECG Exercise radionuclide scintigraphy Pharmacologic stress testing ECHO Coronary angiography Coronary CT
RESP	CHF	Dyspnea Nighttime cough Orthopnea Chest tightness	S ₃ Rales Wheezing	CXR
GI		Angina-equivalent Sx LUQ pain Nausea, indigestion		See CV
RENAL	Decreased renal perfusion	Increased UO at night		Cr
CNS	Syncope	Syncope with chest pain		Exercise stress test
MS	Angina-equivalent Sx Arm pain/neck pain			See CV

Key References: Fihn SD, Blankenship JC, Alexander KP, et al.: 2014 ACC/AHA/AATS/PCNA/SCAI/STS focused update of the guideline for the diagnosis and management of patients with stable ischemic heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons, *J Am Coll Cardiol* 64(18):1929–1949, 2014; Fleisher LA, Fleischmann KE, Auerbach AD, et al.: 2014 ACC/AHA guideline on perioperative cardiovascular evaluation and management of patients undergoing noncardiac surgery: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, *Circulation* 130(24):2215–2245, 2014.

Perioperative Implications

Preoperative Preparation

- Continuation of chronic antianginal medications associated with a lower incidence of myocardial ischemia/infarction, especially beta blockers, statins, and antiplatelet agents.
- No RCT has definitively demonstrated improved outcome acutely starting any medications.

Monitoring

- ST-segment analysis.
- PA cath for large fluid shift operations or pts with signs of LV dysfunction, although RCT unable to document benefits of routine monitoring.
- TEE is most sensitive, but technical issues of real-time interpretation may occur.

Airway

- None

Preinduction/Induction

- May develop reduced CO and hypotension with ischemia.
- Avoid tachycardia, hypotension.

Maintenance

- Myocardial ischemia may manifest as
 - CV instability.
 - Intraop myocardial ischemia.
 - Reduced CO, increased PCWP.
 - Regional wall motion abnormalities.
- No one agent or technique has been shown superior.
- Maintain normothermia and adequate hematocrit ($\geq 28\%$).

Extubation

- Period at greatest risk for developing ischemia.

Postoperative Period

- Pain management may be critical.

- Consider monitoring troponin if there are any signs/symptoms of myocardial ischemia. Some authors suggest routine monitoring of troponin in this population.

Adjuvants

- β -adrenergic receptor antagonist, nitroglycerin, Ca²⁺-channel blockers

Anticipated Problems/Concerns

- Pts with angina who develop dyspnea on exertion are at greatest risk for developing periop cardiac complications.
- Exercise tolerance may be the best predictor of periop risk. Pts with a good exercise tolerance may not require further evaluation for less-invasive procedures.
- Pts who develop periop MI are at increased risk of periop death and long-term morbidity/mortality. Elevated troponin also associated with worse long-term outcomes.

Anhidrosis (Congenital Anhidrotic Ectodermal Dysplasia)

Srijaya K. Reddy | Raafat S. Hannallah

Risk

- Rare: 1:125,000,000
- Clusters in Japan and Israel

Perioperative Risks

- Impaired thermoregulation (risk of fatal hyperpyrexia in infants)
- Postop chest infections

Worry About

- Absence of sweat leads to impaired thermoregulation.
- Insensitivity to superficial and deep painful stimuli with intact tactile perception; still require

considerable amounts of inhalational or IV anesthetics to maintain hemodynamic stability and obtund stress response to airway manipulation.

Overview

- Innervation of the eccrine sweat glands is lacking; heat loss by evaporation is impaired.
- Absent mucous glands from resp tract and esophagus; frequent resp infections.
- Partial or complete absence of teeth.
- Hypotrichosis (absent hair).
- Self-mutilating behavior and mental retardation.
- Characteristic facies: Prominent supraorbital ridges, depressed bridge and root of nose, large

deformed ears, thick lips, underdeveloped maxilla and mandible.

Etiology

- Sex-linked autosomal recessive disorder.
- Human *TRKA* (*NTRK1*) encodes the RTKs for NGF and is the gene responsible.
- Full expression only in males; carrier females may be mildly affected.

Usual Treatment

- Protect from risks of hyperpyrexia due to infection, hot weather, vigorous exercise

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 anhidrosis: 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, multi-plane 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, Ananthakrishnan 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.