Autoimmune Diseases, Cold

Joseph L. Seltzer | Lee A. Fleisher

Risk

- Rare
- Autoimmune hemolytic anemias occur in 1 of 80,000 persons; of these, 17.3% are due to cold antibodies.

Perioperative Risks

- Acute hemolysis due to cold
- Hemoglobinemia
- Hemoglobinuria
- Rarely, vascular occlusion

Worry About

- Cooling to 28–31° C will cause hemolysis.
- These temperatures can be reached in extremities during cardiopulmonary bypass.

Overview

- In two circumstances antibodies will react in the cold to produce hemolysis:
  - IgG antibodies associated with mononucleosis, mycoplasmal pneumonia.
  - IgM antibodies found in the idiopathic form of the disease and in lymphoproliferative disease.
- Hemolysis usually occurs at temp below 31°C.

Etiology

- Idiopathic
- Lymphoid (B-cell) malignancy
- Infections: mycoplasmal pneumonia, mononucleosis, cytomegalovirus, varicella, EBV

Usual Treatment

- Keep warm; administer folic acid.
- For severe cases, chlorambucil or cyclophosphamide.
- Plasmapheresis.
  - Rituximab.
  - Prednisone.

Assessment Points

System	Effect	PE	Test
HEME	Mild to moderate anemia		Hgb, blood bank antiglobulin tests
GU	Hemoglobinuria		
CV	Dyspnea on exertion if anemia is severe		
DERM	Agglutination of RBCs in cold	Acrocyanosis	

Key References: Young S, Haldane G: Major colorectal surgery in a patient with cold agglutinin disease, *Anaesthesia* 61(6):593–596, 2006; Bratkovic K, Fahy C: Anesthesia for off-pump coronary artery surgery in a patient with cold agglutinin disease, *J Cardiothorac Vasc Anesth* 22(3):449–452, 2008.

Perioperative Implications

Preoperative Preparation

- Determine risks of operating vs. not operating.
- Plasmapheresis—may be used, but no more than 2 d before surgery.

Monitoring

- Temp
- Urine output

Maintenance

- Keep pt warm, including extremities.
  - Consider forced-air warming.
  - Warm all fluids.
- Normothermic cardiopulmonary bypass.
- No preferred agent or technique.
- Consider hemodilutional autologous transfusion or other techniques to avoid homologous transfusion and formation of new antibody.

Postoperative Period

- Warm fluids and extremities.
- Monitor for manifestations of cold agglutinin disease.

Anticipated Problems/Concerns

- Hemolysis if temperature falls.
- Renal dysfunction due to hemoglobinuria.
  - Molting or cyanosis of the skin can occur.

Autonomic Dysreflexia

Kieran A. Slevin

Risk

- AD occurs with greatest frequency in pts with spinal cord injury at T6 or above.
- Occurs with highest frequency following urologic or lumbar and thoracic spine procedures.
- Tetraplegic pts develop AD if cystoscopy and lithotripsy are performed without anesthesia.
- The higher the injury level, the greater clinical manifestations of CV dysfunction.
- Risk of AD greater with complete (91%) versus incomplete (27%) cord transections.
- AD more often a delayed finding in chronic SCI; minor clinical evidence seen in first d/wk.

Perioperative Risks

- AD most commonly triggered by irritation and/or manipulation of urinary bladder or colon, as well as in labor.
- Severe increased BP and increased or decreased HR is associated with stimulation below level of transection.
- Objectively, increased SBP >20-30 mm Hg is considered a dysreflexic episode. However, be aware that the usual resting ABP in these pts is 15-20 mm Hg less than in non-SCI subjects.
- Awake pts may complain of HA; anxiety; sweating, piloerection; and flushing above injury level; and dry, pale skin below. In anesthetized pts, SBP rising to up to 300 mm Hg heralds onset of severe, life-threatening AD.

Worry About

- Untreated, uncontrolled hypertensive episodes, which can lead to intracranial hemorrhage, retinal detachment, seizures, and death.

Overview

- Physiologically, AD is caused by a massive sympathetic discharge triggered by a noxious or nonnoxious stimulus originating below the level of the SCI.
- Specifically, destruction of the vasomotor pathways results in a loss of inhibitory and excitatory supraspinal input to the sympathetic preganglionic neurons, thus causing labile BP.
- Also, changes in spinal sympathetic neurons and primary afferents underlie abnormal CV Δs.
- Symptoms are usually short-lived because of treatment or self-limiting nature of the episode.

Etiology

- Most common cause is traumatic interruption of the spinal cord.
- Can also occur due to infectious or oncologic processes causing destructive spinal lesions.

Usual Treatment

- Stop initiating stimulus as first-line therapy when possible.
- Can decrease or prevent AD by use of neuraxial blockade (spinal >>epidural).
- When signs of AD are evident, administer ganglionic blockers (trimethaphan), direct vasodilators (nitroprusside) or α-antagonists (phentolamine), GA, or spinal anesthesia.
- Level 1 evidence that intrasphincteric anal block with lidocaine limits the AD response in pts undergoing anorectal procedures; level 1 evidence that topical lidocaine does not.
- Level 1 evidence that prazosin is superior to placebo in prophylactic management of AD.
- Level 2 evidence that nifedipine can prevent BPΔs during cystoscopy in SCI pts with AD.
- Level 4 evidence that epidural anesthesia may be effective in pts with AD during labor and delivery.
- Centrally acting hypotensive agents (e.g., clonidine) are NOT effective in treating AD.
- Treat tachyarrhythmias with β-blockers in combination with antihypertensives.
- Nicardipine may be preferable in a pt with an upper spinal cord injury undergoing operation in the paralyzed area.
- Magnesium sulfate has significant beneficial effects on AD in labor in a pt with a high spinal cord injury.
- Complete bladder deafferentation does not abolish AD during bladder urodynamic studies.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway	C-spine trauma/surgery H/O; difficult intubation	↓ C-spine ROM ↓ Mouth opening	Airway exam
CV	Orthostatic hypotension Baseline relative hypotension (15–20 mm Hg)	H/O dizziness when going from supine to upright position	↓ BP, orthostasis, tachycardia, bradycardia, AFIB	Orthostatic BPs ECG
RESP	Decreased resp volumes, atelectasis, pneumonia, hypoxemia Impaired cough reflex	SOB Difficulty w/secretions	Tachypnea Cyanosis Decreased/unequal BS	CXR ABG evaluation
GI	Full stomach status due to GI atonicity	Complaints of reflux		
RENAL	UTI, renal stone disease, renal failure	Flank pain	Chronic Foley catheter	UA and BUN/Cr
CNS	Bowel and bladder dysfunction Chronic and central pain states Altered MS (if severe head trauma)	Incontinence Chronic opioid therapy Adjuvant pain meds	Hyperreflexic below level of transection Babinski sign positive	Hyperalgesia Allodynia
PNS	Insensate below level of transection Pain at level of transection	Skin-color changes	Flushing/piloerection above Dry, pale skin below	
MS	Paralysis, muscular atrophy below Sacral decubiti	Paraplegia or quadriplegia	Muscle atrophy Sacral decubiti	

Key References: Krassioukov A: A systematic review of the management of autonomic dysreflexia after spinal cord injury, *Arch Phys Med Rehabil* 90:682–695, 2009; Liu N: Iatrogenic urological triggers of autonomic dysreflexia: a systematic review, *Spinal Cord* 53(7):500–509, 2015.

Perioperative Implications

Preoperative Preparation

- Nifedipine can be used for prophylaxis; given 30 min before procedure, likely to trigger AH.
- Attention to CV and pulm function, volume status, and airway exam.

Monitoring

- Consider preinduction invasive monitoring (arterial and CVP/PA catheters) if volume changes are expected and in setting of poor cardiac reserve (high lesions) and renal insufficiency.

Airway

- Be prepared for fiberoptic intubation.

Induction

- Use nondepolarizing muscle blockers when relaxation is necessary.
- IV nicardipine can be used to treat AD.
- Succinylcholine can cause severe K^+ release and hyperkalemia in chronic lesions.
- Consider nitroprusside before induction.

Maintenance

- GA with volatile agent superior to nitrous-narcotic technique for prevention/treatment of AD.

Regional Anesthesia

- Anesthetic technique of choice when possible.
- Spinal anesthesia highly effective in preventing AD precipitated by surgery.
- Ensure careful assessment of level of spinal blockade in SCI pts due to sensory deficits below injury: avoid unnecessarily high or inadequate blocks.
- Epidural anesthesia effective in preventing AD in laboring pts.

Extubation

- May be difficult due to resp insufficiency in pts with high-level spinal lesions

Adjuvants

- Muscle relaxants required in abdominal surgery due to diffuse increase in muscle tone

Postoperative Period

- AD can occur postop in setting of unrecognized or untreated distended bladder or rectum.
- Consider intracerebral hemorrhage protocol in the setting of unexplained delayed emergence with increased BP.

Becker Disease

Pikulkaew Dachsangvorn

Risk

- Prevalence is approximately 1:50,000

Perioperative Risks

- Myotonia

Worry About

- Myotonic episode leading to a difficult to ventilate/intubate situation

Overview

- Genetic disease that results in muscle membrane hyperexcitability and delayed relaxation
- Recessively inherited form of MC
- Initial symptoms start around 4–12 y of age, with generalized myotonia and moderate to pronounced muscular hypertrophy from chronically increased muscle activity
- Signs include muscle stiffness after voluntary contraction that improves with repetitive movement ("warm-up" phenomenon) and worsens after prolonged rest
- Many experience transient weakness (<1 min) upon initiating movement; history of clumsiness, dropping objects, impaired postural control, or uncontrolled falling upon standing
- Rarely, can have atrophy in the forearms and painful muscle cramps
- Most have normal life expectancy without significant handicap
- Aggravating factors: dietary insufficiencies, sleep deprivation, prolonged physical activity, and emotional stress
- Menstruation, pregnancy, and hypothyroidism may alleviate or worsen symptoms in some individuals
- No involvement in smooth and cardiac muscles, no extramuscular manifestations
- It is important to differentiate this from myotonia with dystrophy, which is a multisystem disorder
- Diagnosis:
  - Characteristic symptoms (described previously)
  - "Percussion myotonia": reflex hammer produces obvious dimpling or fasciculation in prominent muscles, such as thenar eminence or thighs, that lingers for several seconds
  - Objective evidence: electromyography
  - Molecular genetic testing is commercially available, although not sensitive for less common mutations

Etiology

- Impaired functioning of skeletal muscle ClC-1
- Skeletal muscle chloride channels serve to stabilize membrane potential at the resting level; impaired ClC-1 leads to sarcolemmal excitability and delayed muscle relaxation
- More than 120 mutations have been described; most mutations are unique to individual families or isolated cases

Usual Treatment

- Most pts prefer to minimize their symptoms by avoiding triggers.
- Pharmacologic therapies include quinidine and quinine, which are effective and well tolerated in low-dose, short-term use; however, continued administrations can lead to toxicity affecting vision, hearing, gastrointestinal, central nervous systems, and possibly causing death.
- Other drugs with variable success include procaine, tocainide, mexiletine, carbamazepine, and phenytoin, by use-dependent blockade of voltage-gated sodium channels.