Worry About

- Acute airway edema resulting from laryngeal or mucous membrane swelling, which can result in definitive airway obstruction; abdominal pain from intestinal edema, which may be an associated finding on exam
- Increased infectious risk

Overview

- Hereditary angioneurotic edema is associated with a complement deficiency of the enzyme C1 esterase inhibitor. It is a rare genetic deficiency that may lead to uncontrolled production of C2, C3, and C5 complement, resulting in acute noninflammatory, painless, nonpruritc, nonpitting edema. Initial inciting events are often the result of trauma, but may even be attributed to emotional stress.
- Any component of the classical pathway, alternate pathway, or terminal common pathway may be affected.
- Virtually all deficiencies show some ↑ risk of infection and/or autoimmune disease.
- Deficiencies in other complement components, C2 and C3, have also been associated with immunocompromised pts, resulting in recurrent life-threatening infections associated with a variety of organisms.
- Increased risk of autoimmune diseases.
- Deficiency in any of the terminal components C5–C8 show selective risk of recurrent neisserial infections, which usually are not life threatening.

Etiology

- C1 complement results from a heterozygous deficiency of C1 esterase inhibitor. The mediators of the angioedema response result from coagulation, complement, and the kinin pathway. C1 esterase inhibitor is a key regulator for Hageman factor, coagulation, plasmin, and plasma kallikrein. There have been more than 100 mutations on the C1 esterase gene in pts without hereditary angioedema, and 20% of those have been new mutations with no prior history.
- All complement proteins are inherited in an autosomal fashion, with the possible exception of properdin, which appears to be X-linked.

Usual Treatment

- Treatment modalities have included stanozolol, danazol, methyltestosterone, oxymetholone, aminocaproic acid, tranexamic acid, and cinnarizine. Mechanism of action for therapeutics is increased synthesis of C1 esterase inhibitor (for the steroids) and inhibition of plasmin activation (for the antifibrinolytics).
- Acute preop prophylaxis has consisted of fresh frozen plasma and epinephrine. However, caution must be taken because plasma provides substrates that may aggravate the scenario and worsen the edema. Purified concentrates of C1 esterase inhibitor given IV have also been used outside USA.
- Antibiotic treatment dictated by specific infection.

Assessment Points

System	Effect	Test
IMMUNO	Infectious risk for all systems	CH50 screening test for complement-mediated lysis of sheep erythrocytes; tests for specific complement components available at reference labs Assess other specific organs as indicated by autoimmune disease (e.g., renal for SLE)

Key References: O'Neil KM: Complement deficiency, *Clin Rev Allergy Immunol* 19(2):83–108, 2000; Wen L, Atkinson JP, Giclas PC: Clinical and laboratory evaluation of complement deficiency, *J Allergy Clin Immunol* 113(4):585–593, 2004.

Perioperative Implications

Preoperative Preparation

- In pts with C1 deficiency, consider preop administration of 2 units of FFP or C1 concentrate with appropriate consideration to risks and benefits of therapy.
- Sterile technique strictly observed.

Monitoring

- Routine.
- Coagulation profile.
- Minimize invasive lines.

Airway

- Airway management should minimize trauma. Tracheal intubation is acceptable, but preparation for an emergency tracheostomy should be made. Laryngeal mask airway use should be tempered by the concerns for upper-airway edema and resulting ineffective ventilation. Regional anesthesia is an acceptable alternative to prevent airway manipulation.

Induction

- Routine

Maintenance

- Routine

Extubation

- Extubate and remove all lines at earliest opportunity.

Postoperative Period

- Maintain sterile techniques.

Anticipated Problems/Concerns

- If emergency intubation is required, it is recommended that an otolaryngologist or surgical personnel be present for a possible tracheostomy or cricothyroidotomy.
- Ensure meticulous sterile technique to minimize risk of infection.

Congenital Methemoglobinemia

Bronwyn R. Rae

Risk

- Navajo Indians, Alaskan Indians, and people of Puerto Rican and Cuban ancestry
- Normal life span (except for RCM type II)

Perioperative Risks

- Oxidizing agents may increase MetHb to dangerous levels.
- Mild respiratory/cardiac depression may adversely affect pts with already limited reserve.
- Pregnancies not compromised.

Worry About

- Measurement of SpO_2
- Oxidant drugs (e.g., prilocaine, benzocaine, nitroglycerin, sulfonamides, phenacetin, and nitric oxide), which are contraindicated
- Myocardial ischemia due to decreased O_2 delivery
- Blood loss/anemia due to O_2-carrying capacity already reduced

Overview

- Enzyme deficiency: Shift of O_2 dissociation curve to the left leads to mild erythrocytosis (normal RBC life span).
- RCM type I defect restricted to red cell soluble cytochrome b5 reductase only. Cyanosis is sole clinical symptom. Homozygotes have compensatory increase in RBC mass. Heterozygotes may develop acute symptomatic methemoglobinemia after exposure to exogenous MetHb-inducing agents. Defect may be the cause of unexplained periop cyanosis.
- RCM type II: Defect occurs in all cells and involves both soluble and microsomal forms of cytochrome b5 reductase. Results include mental retardation, spasticity, opisthotonos, microcephaly, growth retardation, and death by 2–3 y of age.
- RCM type III: Nonerythroid enzyme deficiency, but CNS spared.
- HbM variations: Alpha chain variants affected from birth, and beta chain variants by 3–6 mo of age. Patients develop mild hemolytic anemia.

Etiology

- RCM types I, II, and III: Occur by autosomal recessive inheritance because of deficient reducing capacity of oxidized heme caused by NADH cytochrome b5 reductase (diaphorase) deficiency.
- HbM variants: Occur by autosomal dominant inheritance due to structural abnormality in globin moiety; amino acid substitutions create abnormal environment for heme residues, displacing the equilibrium toward the ferric state.

Usual Treatment

- RCM types I, II and III: Reducing agents (e.g., riboflavin 20–60 mg orally, methylene blue 1–2 mg/kg IV; the effect lasting 10–14 d) and ascorbic acid used for chronic management
- HbM variants: No available treatment. (In an emergency, hyperbaric O_2 therapy and exchange transfusion may be used.)

Assessment Points

System	Physical Examination	Test
RESP	Cyanosed but more "blue" than "sick"	15-30% MetHb
HEME	RCM types I and II: Mild erythrocytosis HbM variants: Mild hemolytic anemia	CBC
CV	May be unable to meet increased metabolic demand	ECG

Key References: Jaffe E, Hultquist D: Cytochrome b5 reductase deficiency and enzymopenic hereditary methemoglobinemia. In Scriver C, Beaudet A, Sly W, et al., editors: *The metabolic and molecular bases of inherited disease,* ed 3, vol. 3, New York, NY, 1995, McGraw Hill, pp 3399; Guay J: Methemoglobinemia related to local anesthetics: a summary of 242 episodes, *Anesth Analg* 108(3):837–845, 2009.

Perioperative Implications

Preoperative Preparation

- Can give reducing agents to pts with RCM type I, but no data exist on whether treatment is indicated before anesthesia.

Monitoring

- Pulse oximeter overestimates at low SpO_2 and underestimates at high SpO_2. In practice, it reads between 80–85%, regardless of true saturation.
- Use co-oximetry for SaO_2 and MetHb levels.
- Monitor ECG for ischemic changes.
- May see "chocolate brown" blood in the operative field or arterial cannula.

Airway

- None

Preinduction/Induction

- Adequate preoxygenation with 100% O_2 because O_2-carrying capacity is already decreased.

Maintenance

- Prilocaine, benzocaine, and EMLA cream are contraindicated. The literature is contradictory on lidocaine. The effects are probably due to respiratory/myocardial depression in patients with low reserve, rather than an increase in MetHb.
- Nitrous oxide, propofol, and volatile agents are okay.

Adjuvants

- None

Postoperative Period

- Avoid acetanilides, paracetamol, metoclopramide, and nitrites. Narcotics may be used.

Anticipated Problems/Concerns

- Avoid oxidant drugs in both homozygotes and heterozygotes.
- Pulse oximetry is inaccurate; use ABG with co-oximetry.
- May require supplemental O_2 postop.
- May have limited cardiac and respiratory reserve.

Congenital Pulmonary Cystic Lesions/Lobar Emphysema

Francine S. Yudkowitz

Risk

- Cause of cardiorespiratory compromise
- 10–15% associated with CHD

Perioperative Risks

- May develop worsening of cardiorespiratory status
- Contamination of unaffected lung by infected material from cyst

Worry About

- Associated congenital anomalies
- Tension pneumothorax
- Cardiorespiratory compromise

Overview

Congenital Pulmonary Cystic Lesions (Three Types)

- Bronchogenic: Abnormal budding and branching of tracheobronchial tree leading to resp distress, pneumonia, and atelectasis
- Dermoid: Lined with keratinized, squamous epithelium
- CPAM: Previously known as CCAM; similar to bronchioles but without alveoli, bronchial glands, and cartilage; overdistension because of gas trapping, which leads to resp distress

Congenital Lobar Emphysema

- Hyperinflation and air trapping result in expansion of affected lobe.
- Most commonly occurs in the left upper lobe, followed in frequency by the right middle, and then the right upper lobe.
- Preterm infants on mechanical ventilation develop emphysema in the right upper lobe.
- CXR shows emphysematous lobe crossing midline, mediastinal shift, and atelectasis in other lobes and possibly the contralateral lung. The presence of bronchovascular markings distinguishes this from pneumothorax and congenital cysts.

Etiology

- Congenital pulmonary cystic lesions may be bronchogenic, alveolar, or a combination of both, and anomalous development of the bronchopulmonary system occurs.
- Congenital lobar emphysema has extrinsic bronchial obstruction from abnormal vessels or enlarged lymph nodes and intrinsic bronchial obstruction from deficient bronchial cartilage, bronchial stenosis, or redundant bronchial mucosa.

Usual Treatment

- Surgical removal

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Decreased lung volume	Cyanosis, dyspnea, grunting, coughing	Tachypnea, retractions, wheezing, decreased BS, asymmetric chest expansion	CXR CT scan
CV	Mediastinal shift, decreased CO, VSD, PDA	Irritability, poor feeding	Decreased heart sounds Murmur	CXR, ECG, ECHO

Key References: Hammer G, Hall S, Davis PJ: Anesthesia for general abdominal, thoracic, urologic, and bariatric surgery. In: Davis PJ, Cladis FP, Motoyama EK, editors: *Smith's anesthesia for infants and children,* ed 8, Philadelphia, PA, 2011, Elsevier; Guruswamy V, Roberts S, Arnold P, Potter F: Anaesthetic management of a neonate with congenital cyst adenoid malformation, *Br J Anaesth* 95(2):240–242, 2005.

Perioperative Implications

Preoperative Preparation

- Assess the severity of cardiopulmonary compromise.
- Identify associated congenital anomalies.
- Optimize resp infection if pt is stable.
- Aspirate cyst before induction if there is cardiac compromise or airway obstruction.

Monitoring

- Arterial line for BP monitoring and blood gas analysis

Induction

- Avoid positive pressure ventilation until thorax is opened to avoid expansion of cyst or lobe.
- Avoid N_2O, which will expand the lobe or cyst.
- Inhalation induction with 100% O_2.
- Intubate without the use of muscle relaxants.
- Affected lung may need to be isolated. In small infants and children, this may be accomplished by using a bronchial blocker or doing a mainstem intubation.
- Surgeon should be available to open the chest immediately if deterioration should occur during induction of anesthesia.

Maintenance

- No one anesthetic preferred.
- Maintain spontaneous ventilation or assist with low airway pressures until thorax is opened.
- Once the pathology is removed, N_2O may be used.
- If Hx of repeated lung infections (cysts), there may be large blood losses.