Assessment Points

System	Effect	Assessment by Hx	PE	Test
PRENATAL				Amniocentesis Chorionic villus sampling
HEENT	Sinusitis	Sinus congestion		X-ray or CT scan
RESP	Emphysema Recurrent infection		Barrel chest Limited chest excursion Reduced air entry	CXR CT scan Spirometry (especially decreased FEV_1) ABGs Gas transfer
CV	Sinus tachycardia Cor pulmonale			
GI	Chronic hepatitis Liver cirrhosis Inflammatory bowel disease Reflux	Unintended weight loss		LFTs
HEME	Polycythemia			Hb, Hct
METAB	Fatigue			6-min walk test Serum AAT Genotyping Immunoelectrophoresis Radial immunodiffusion Nephelometry Thin-layer isoelectric focusing

Key References: American Thoracic Society, European Respiratory Society: American Thoracic Society/European Respiratory Society Statement: standards for the diagnosis and management of individuals with alpha-1 antitrypsin deficiency, *Am J Respir Crit Care Med* 168(7):818–900, 2003; Myles P, Weeks AM: Alpha 1-antitrypsin deficiency: circulatory arrest following induction of anaesthesia, *Anaesth Intensive Care* 20(3):358–362, 1992.

Perioperative Implications

Preoperative Preparation

- Thorough pulm evaluation.
- General anesthesia can be tolerated in most pts with an FEV_1 of ≥1.0 L.
- Consider local, regional, or neuraxial anesthesia.

Monitoring

- Routine, but consider arterial line in case of major surgery (to monitor hyperinflation-induced hypotension).
- Lung ultrasound can be used to rule out pneumothorax.

Induction

- Adjust positive pressure ventilation to prolong expiration time, with an I:E ratio ≥3.
- Avoid high PEEP.

Maintenance

- Permissive hypercapnia

Postoperative Period

- Opioid-sparing analgesic regimen.
- Avoid hepatotoxins; use caution with acetaminophen.
- Titrated oxygen therapy to avoid hyperoxic hypoventilation.
- High-dependency or critical care admission.
- Monitor for concomitant liver and kidney dysfunction.
- Consider AAT augmentation therapy.

Amniotic Fluid Embolism

Ryan Palacio | Mohammed M. Minhaj

Risk

- Risk factors include: advanced maternal age (>35 y); cesarean delivery; placenta previa; meconium; intrauterine fetal demise; placental abruption; meconium staining of the amniotic fluid; chorioamnionitis; and macrosomia.
- True incidence is unknown but estimated to occur in 2 to 8 per 100,000 deliveries.

Perioperative Risks

- Amniotic fluid embolism accounts for approx 6% of maternal deaths in USA.
- Mortality was once as high as 61% to 86%, but more recent registries have reported mortality between 11% and 44% of pts.
- Morbidity is also high as it is suggested that up to 60% of pts have persisting neurologic deficits.

Worry About

- Hypoxia.
- Hypotension/cardiopulmonary collapse.
- Heart failure (can have both right and left ventricular failure).
- DIC: Occurs in nearly all survivors of the initial catastrophic event.
- Hemorrhage: 40% of amniotic fluid embolism-associated deaths are due to hemorrhage.
- Altered mental status.
- Seizures.
- ARDS.
- Acute pulm Htn.

Overview

- Amniotic fluid going to central circulation.
- There are three necessary conditions:
  - Amniotomy (breach in the barrier between the intact fetal membranes that isolate amniotic fluid from the maternal circulation).
  - Laceration of endocervical or uterine vessels or site of placental attachment.
  - Traditionally it was thought that a pressure gradient (intrauterine pressure > CVP or uterine venous pressure) was needed, but the presence of an electrochemical gradient can provide the means for mediators of AFE to inflict damage.
- Immunologic factors also likely to be involved, and complement activation may play a role in the pathophysiology of AFE (e.g., SIRS).

Etiology

- Postulated mechanism of action: Powerful contractions force amniotic fluid into the maternal circulation through a defect in the fetal membranes, placenta, or elsewhere.
- Diagnosis based on clinical symptoms and diagnosis of exclusion of other potential etiologies. No uniform diagnostic criteria exist nor are there specific laboratory findings pathognomonic for AFE. Fetal cells in maternal pulm circulation are not a reliable marker.
- AFE can also occur up to 48 h postpartum and in rare cases following intrauterine surgery or blunt abdominal trauma.

Usual Treatment

- Usually supportive to maintain oxygenation, circulatory support, and correct coagulopathy
- Delivery of fetus as soon as is practical; may require operative or cesarean delivery
- Employ left uterine displacement to prevent aortocaval compression
- Cardiopulmonary resuscitation, often requiring intubation with 100% O_2/PEEP. Case reports of successful outcomes with employment of inhaled nitric oxide, CPB and/or ECMO have been reported in the literature.
- Risks/benefits of uterotonic agents should take into account clinical picture of hypotension and/or hemorrhage considerations.
- Pressors and inotropes will often be required.
- Replacement of clotting factors if pt develops DIC. Clotting factors can also be replaced with recombinant factors in addition to traditional blood product transfusion.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Tachycardia Hypotension			HR BP
RESP	Hypoxia Pulm edema	Dyspnea	Tachypnea Cyanosis Frothy pink sputum	Pulse oximetry Aspirate blood from pulmonary artery or renal artery Stain buffy coat for cells and mucin
GI		Nausea	Vomiting	
HEME	DIC		Excessive bleeding Thrombolysis (bleeding from IV sites)	PT, PTT, plt, fibrinogen, FSP
CNS		Anxiety	Convulsions Shivering Sweating	

Key References: Rath WH, Hofer S, Sinicina I: Amniotic fluid embolism: an interdisciplinary challenge, *Dtsch Arztebl Int* 111(8):126–132, 2014; McDonnell NJ, Percival V, Paech MJ: Amniotic fluid embolism: a leading cause of maternal death yet still a medical conundrum, *Int J Obstet Anesth* 22(4):329–336, 2013.

Perioperative Implications

- Most common presentation is hemodynamic collapse.

Preoperative Preparation

- Maximize maternal oxygen delivery.
- Place several large-bore IVs; consider central access for inotrope administration and fluid resuscitation.
- Notify blood bank of anticipated coagulopathy and cross-match for several units of packed RBCs, FFP, platelets, and cryoprecipitate.
- Consider preparing for CPB/ECMO if an option.

Monitoring

- If amniotic fluid embolism is suspected, consider PA catheter or cardiac ultrasound (TTE/TEE) for hemodynamic management.

Maintenance

- Usually resuscitative with support of breathing and circulation.
- Case reports of use of CPB, ECMO, inhaled nitric oxide, ventricular assist devices.

Extubation

- If the pt survives, keep intubated until hemodynamically stable.

Anticipated Problems/Concerns

- Even with early and aggressive intervention, AFE can result in maternal and fetal mortality. Given that an AFE can occur unpredictably and then has a high risk for morbidity and mortality, it can be devastating for the pt's family and healthcare providers. Psychological counseling for all parties involved should be considered to deal with any posttraumatic stress.

Amyloidosis

Toby N. Weingarten

Risk

- Incidence in USA: 1:100,000
- Race with highest prevalence: Unknown

Perioperative Risks

- Increased risk of periop renal failure, cardiomyopathy (arrhythmias and ventricular dysfunction), bleeding from coagulopathy
- Autonomic neuropathy

Worry About

- Signs of CHF
- Dysrhythmias
- Decreasing urine output

Overview

- Extracellular deposition of amyloid-type proteins.
- Congo-red stain of tissue reveals green birefringence in a polarizing microscope.
- Associated end-stage renal, myocardial, and neuropathic disease.
- Best diagnosed by subcutaneous abdominal fat pad aspirate or rectal biopsy.

Etiology

- Both acquired and hereditary forms exist.
- Acquired forms are categorized as primary (AL), associated with plasma cell disorders (i.e., multiple myeloma), and secondary (AA), associated with inflammatory and infectious diseases (e.g., osteomyelitis, rheumatoid arthritis).
- Hereditary forms very rare.

Usual Treatment

- Acquired: Treatment of primary (AL) amyloidosis is directed at the underlying plasma cell disorder (e.g., chemotherapy, stem cell transplant). Treatment of secondary (AA) amyloidosis is directed at underlying infection/inflammation.
- Hereditary: Colchicine, liver transplantation.
- Treatments to clear amyloid deposits are being developed.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Macroglossia Tracheal stenosis	Enlarged tongue Dyspnea	Macroglossia Stridor	CT scan Flow-volume loop
CV	Restrictive myopathy LV and RV dysfunction Conduction abnormalities	Exercise tolerance Dyspnea Syncope	Increased JVP S_3 Bradycardia	ECHO ECG
RESP	CHF Lung nodules	Cough Chest wall pain	Rales	CXR
GI	Autonomic dysfunction Hepatomegaly	Malabsorption Diarrhea Bleeding	Hepatomegaly Ascites	Biopsy LFTs
HEME	Factor X deficiency Capillary fragility	Bruising Purpura	Periorbital bruises ("raccoon eyes")	Factor X assay
RENAL	Decreased renal perfusion Nephrotic syndrome			BUN/Cr urine
CNS	Autonomic neuropathy Cardioembolic strokes	Inability to sweat; hoarseness; early satiety; postural dizziness	Orthostasis	Biopsy

Key References: Noguchi T, Minami K, Iwagaki T, et al: Anesthetic management of a patient with laryngeal amyloidosis, *J Clin Anesth* 11:339–341, 1999; Thompson CA, Kyle R, Gertz M, et al: Systemic AL amyloidosis with acquired factor X deficiency: a study of perioperative bleeding risk and treatment outcomes in 60 patients, *Am J Hematol* 85:171–173, 2010.