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.

Perioperative Implications

Preoperative Preparation
- Optimize treatment of heart failure.
- Avoid dehydration (renal failure).
- Care with positioning and taping (skin fragility).

Monitoring
- Consider of TEE or PA cath for large fluid shift operations or pts with severe LV dysfunction.

Airway
- Macroglossia or tracheal stenosis
- Increased risk of bleeding into airway from capillary fragility and possible coagulopathy

Preinduction/Induction
- May develop reduced CO and hypotension.
- Coagulopathy may contraindicate regional anesthesia.

Maintenance
- No agent or technique shown superior.
- Maintain adequate urine output.

Extubation
- Pt fully awake to minimize risk of reintubation.
- Use caution with nasal airway as it may cause hemorrhage.

Postoperative Period
- Close monitoring of CV and renal status.
- Consider ICU setting for postop care.

Adjuvants
- Avoid digoxin: Not usually helpful in treating amyloid CHF, associated with increased arrhythmias.

Anticipated Problems/Concerns
- Difficult airway
- CHF
- Hypotension
- Renal failure
- Easy bruising; increased risk of bleeding

Amyotrophic Lateral Sclerosis

Alan David Kaye | Charles Fox III | Elyse M. Cornett

Risk
- Estimated incidence of 1-3:100,000.
- Mean age of onset is in the 60s, but ALS can occur as early as the 20s.
- Disease duration is approximately 3 y from the time of diagnosis to death.
- While there is slight male predominance of sporadic spinal ALS, slight female predominance is found in bulbar ALS
- Most cases are sporadic but 5% to 10% are familial.
- Risk of anesthesia increases as the FVC falls below 50%, ALS pts can be stratified as low risk if the FVC >50%, moderate risk if the FVC is 30% to 50%, and high risk if the FVC <30%.

Perioperative Risks
- Aspiration.
- Respiratory depression.
- Inability of pt to communicate secondary to bulbar weakness.

Worry About
- Succinylcholine-induced hyperkalemia.
- Prolonged resp depression with inability to extubate, even without use of muscle relaxants.
- Hypersensitivity to nondepolarizing neuromuscular blockers.
- Disease exacerbation with use of regional anesthesia.

Overview
- Disease of unclear etiology that leads to progressive degeneration of the upper and lower motor neurons causing amyotrophy (muscle wasting) and lateral sclerosis (gliosis of the corticospinal tracts).
- Located in the motor cortex (upper motor neurons) and anterior horn (lower motor neurons) of the spinal cord.
- ALS has a relenting course that leads to weakness of all skeletal muscles in the body.
- Typically, ALS is asymmetric involving the distal extremities first followed by bulbar muscle weakness as the disease progresses.
- After diagnosis in an adult, pts are usually wheelchair bound by 18 mo and die after 3-5 y from resp suppression.
- Juvenile forms of ALS do exist, present early in life, and are rare.
- Upper motor neuron signs include spasticity, hyperactive reflexes, and upgoing plantar response; lower motor neuron signs include muscle atrophy and fasciculations.
- Disease does not affect ocular muscles, bladder, bowel, and sensation.
- ALS variants include:
  - Primary lateral sclerosis: Progressive degeneration of upper motor neurons;
  - Progressive muscular atrophy: Progressive degeneration of lower motor neurons;
  - Progressive bulbar palsy: Progressive motoneuron loss from lower cranial nerve nuclei and cervical spine.

Etiology
- Familial ALS caused by gene mutations: 14 mutations described. Most studied mutation occurs in the gene encoding superoxide dismutase and forms aggregates leading to mitochondria and muscle complex dysfunction.
- Etiology of sporadic ALS remains uncertain; however, autoimmune, viral, and neurotoxic mechanisms likely contribute.
- Interaction between a genetic susceptibility and environmental factors likely leads to the disease.

Usual Treatment
- Care is mainly supportive, consisting of psychological therapy, symptom management, physical therapy, and palliative care.
- Care in a multidisciplinary clinic is associated with prolonged survival and improved quality of life.
- Riluzole, which inhibits glutamate release, is the only drug shown to improve survival. On average, pts live 2 to 3 mo longer on riluzole versus placebo.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Dysarthria, dysphagia, sialorrhea	Slurred speech, coughing with eating, drooling	Decreased gag reflex	Swallow study
CV	Reduced sympathetic tone Vagal dysfunction	Syncope Cardiac arrest		Prolonged QTc Tachycardia
PULM	Aspiration; nocturnal apnea; weak cough	Recurrent pneumonia, nighttime arousals, lethargy	Decreased breath sounds; coarse breath sounds	PFTs Nocturnal oximetry CXR ABG
GI	Malnourished	Caloric intake Food journal	BMI	Albumin
CNS	Motor neuron loss in spinal cord and brain	Weakness Pseudobulbar affect	Weakness Fasciculations Atrophy	EMG/NCS

Key References: Mancuso R, Navarro X: Amyotrophic lateral sclerosis: current perspectives from basic research to the clinic, *Prog Neurobiol* 133:1–26, 2015; Turakhia P, Barrick B, Berman J: Patients with neuromuscular disorder, *Med Clin North Am* 97(6):1015–1032, 2013.

Perioperative Implications

Preinduction/Induction/Maintenance
- Succinylcholine is contraindicated as it can cause hyperkalemia.
- Nondepolarizing agents may be used, but anticipate prolonged weakness.
- Short-acting muscle relaxants should be used when necessary.

Preoperative Considerations
- Preop pulmonary function tests may help to predict anesthetic risk and include FVC and nocturnal oximetry.
- Consider aspiration prophylaxis.
- Avoid opioids and benzodiazepines if possible.
- Bulbar dysfunctions occur in up to 25% of pts. Look for weight loss, dysarthria, and difficulty whistling or using a straw.
- Resp failure is the main cause of death in ALS pts.
- Depression and emotional lability are common in ALS pts and are typically treated with TCAs which