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

can prolong the QT interval. If the decision is made to stop TCAs before surgery, it must be done 2 wk before the surgery to prevent withdrawal symptoms. If TCA treatment will continue through surgery, an ECG must be obtained prior to the start of surgery.

Monitoring
- Routine.
- Anesthesia should be performed in an inpatient setting.

General Anesthesia
- Avoid if possible.
- May cause significant postop resp depression.
- Diaphragmatic pacing stimulation may improve resp compliance and stimulate respirations.
- Extubate when pt is fully awake.

Regional Anesthesia
- May be preferred compared to general anesthesia.
- Case reports have documented successful use of epidural anesthesia.
- Minimize neuraxial extent of blockade to reduce risk of resp depression.

Postoperative Period
- Anticipate prolonged postop ventilation.
- Use nonsedating medications for pain control.

Anticipated Problems/Concerns
- Anticipate hospitalization secondary to prolonged weaning from ventilator.
- Communication with ALS pts may be difficult because pts have weakened oropharyngeal muscles. Prior to anesthesia, determine the best way to communicate with pts (i.e., letter boards) and have family members available to assist.
- Close resp monitoring is essential following anesthesia. Exacerbation of apnea may result from supplemental oxygen.

Anaphylaxis

Karen Hand

Risk
- Lifetime prevalence of anaphylaxis is 0.05% to 2%, most common triggers being food, stings, and iatrogenic causes.
- Occurs in approximately 1 in 10,000 to 1 in 20,000 anesthetic procedures, and 1 in 6500 administrations of neuromuscular blocking agents (NMDAs). Causes 3% of anesthesia-related deaths.
- Females outnumber males 3:1.
- Hx of atrophy, prior anaphylaxis, and prior adverse reaction to anesthesia.

Perioperative Risks
- Significant risks of life-threatening CV collapse, airway compromise, and bronchospasm.
- Most common causes are NMDAs (60%), latex (15%), and antibiotics (15%).
- Increased risk of life-threatening reactions with beta blockers, ACEIs, asthma, and underlying cardiac disease.

Worry About
- Hx of atrophy, prior anaphylaxis, and prior adverse reaction to anesthesia.
- Timing: Most reactions occur around the time of induction or within 10 min of drug administration. May be difficult to distinguish from other drug reactions or mechanical problems.
- Rapid progression: Time to cardiac or resp arrest is within 5 min for anesthetic reactions, compared to 30 min for food and 15 min for stings.
- Diagnostic difficulty: Varied presentations, tachycardia or bradycardia, less than 50% have bronchospasm, cutaneous signs may be absent or occur later in severe reactions.
- Biphasic response: May recur from 4-24 h later

Overview
- Defined as a severe, life-threatening, generalized or systemic hypersensitivity reaction.
- Classified as:
  - Allergic reactions, usually involving IgE.
  - Nonallergic reactions, previously called anaphylactoid.
  - Itching, burning hands, feet, mouth or genitals, abdominal pain, nausea, and a feeling of doom or tunnel vision may be reported by awake pts.
- Most common initial features during anesthesia are pulselessness, desaturation, and difficult ventilation.

Etiology
- Allergic: IgE antibodies crosslink receptors on mast cells and basophils, causing degranulation, releasing many vasoactive substances, incl histamine, in an inflammatory cascade.
- Usually requires prior exposure. However, can occur with NMDAs with first exposure, thought to be due to common quaternary amine in NMBAs and chemicals (e.g., found in cleaners and cosmetics). In Europe, linked with ingredient in cough syrup, pholcodine.
- Can occur with any muscle relaxant, most commonly succinylcholine. Increasing reports with rocuronium; also reported with sugammadex.
- Risk factors for latex allergy include meningomyelocele, as well as allergy to figs, papayas, or avocados. Increased in healthcare workers
- Rarely due to opiates or local anesthetics (more likely intravascular injection or epinephrine)
- Nonallergic: Related to drug dose and speed of injection. Usually less severe than IgE-mediated reactions.

Usual Treatment
- Halt exposure to trigger
- Epinephrine: Standard treatment (e.g., no IV is 0.3 mg IM to outer thigh). Under anesthesia adjust IV dose according to severity of reaction from 10 s mcg to 100 s mcg to multiple 1 mg doses.
- Clinical Severity Scale:
  - Grade 1: Cutaneous or mucous signs.
  - Grade 2: +/– hypotension, tachycardia, dyspnea, GI disturbance.
  - Grade 3: Life-threatening CV or resp collapse.
  - Grade 4: Cardiac arrest.
- IV fluids (large bore IV); may require large volume.
- O_2 and supportive measures.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Head and neck swelling, and potential glottic edema	Will occur suddenly	Swelling	Clinically obvious
CV	Increased or decreased HR, decreased BP and SVR, increased ectopy, acute coronary events		Hypotension, CV collapse	ECG may reveal premature ventricular contractions PVCs or change in P-R interval
RESP	Bronchospasm		Wheezing	Increased peak inspiratory pressure, decreased O_2 saturation
DERM	Urticaria, erythema, hives, edema		Body rash	May be hidden under drapes or absent initially in severe reactions

Key References: Simons FE, Sheikh A: Anaphylaxis: the acute episode and beyond, *BMJ* 346:f602, 2013; Dewachter P, Mouton-Faivre C, Emala CW: Anaphylaxis and anesthesia: controversies and new insights, *Anesthesiology* 111(5):1141–1150, 2009.

Perioperative Implications

Preoperative Preparation
- Prophylactic H1/H2 blockers and steroids may attenuate the severity, although not the incidence of reactions. There is more support for their use in preventing nonallergic reactions.
- Consider administering antibiotics preop rather than at the time of induction.

Monitoring
- Standard ASA monitors are essential to rapidly identify anaphylaxis.
- Always consider anaphylaxis in CV collapse with or without bronchospasm or cutaneous manifestations during induction.
- The airway may swell, making intubation very difficult.

Induction
- Reactions usually occur during induction. Reactions to latex may occur within 30 min.

Maintenance
- Perpetuation of reaction can occur, particularly if due to latex.
- Significant cross-reactivity between NMDAs (approaching 80%).
- Avoid all muscle relaxants in pts with prior reactions.

Extubation
- Ensure stability from a cardiorespiratory viewpoint.
- Assess for airway edema.
- Beware reactions to sugammadex.