reflux material help determine extent of mucosal damage and degree of esophagitis.
- Esophagitis may lead to bleeding, which may result in hematemesis, iron-deficiency anemia, and esophageal stricture. Also, pts are predisposed to Barrett esophagus.
- GER may be a cause of neonatal apnea.
- Diagnostic procedures include upper GI series, esophagoscopy, and esophageal pH probe.

Etiology

- Immature maturation of the lower esophageal sphincter
- Discoordination of swallowing mechanism in neurologically impaired pts

Usual Treatment

- Medical:
  - Infants: Thicken feeds, elevate the head of the bed maximum of 30 degrees, or place infant in prone position (although noted to be a risk for SIDS).
  - Older children: Avoid foods and beverages that exacerbate acid reflux (e.g., citrus, tomatoes, spicy/fried foods, caffeine). Place pt in upright position; sitting position worsens reflux.
  - H_2 blockers or PPIs to decrease gastric acidity.
- Surgical:
  - Indicated when medical therapy fails or in the presence of significant comorbidities (e.g., recurrent aspiration, apnea, failure to thrive, Barrett's esophagus).
  - Open or laparoscopic Nissen fundoplication has a 95% success rate in neurologically intact pts. Pts who are neurologically impaired have a greater morbidity and mortality with surgical repair.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Chronic aspiration RAD	Cough, cyanotic episodes, apnea Dyspnea, wheezing, cough	Rales, rhonchi Wheezing Decreased breath sounds, prolonged expiration	CXR, ABG (if indicated) CXR, peak flow ABG (if indicated)
HEME	Iron deficiency		Pallor	CBC
GENERAL	Malnutrition	Weight loss	Decreased SQ tissue	Serum albumin

Key References: Suwandhi E, Ton MN, Schwarz SM: Gastroesophageal reflux in infancy and childhood, *Pediatri Annals* 35(4):259–266, 2006; Hammer G, Hall S, Davis PJ: Anesthesia for general abdominal, thoracic, urologic, and bariatric surgery. In Davis PF, Cladis FP, Motoyama EK, editors: *Smith's anesthesia for infants and children,* ed 8, Philadelphia, PA, 2011, Elsevier, pp 745–785.

Perioperative Implications

Preoperative Preparation

- Assess the severity of pulm compromise.
- Optimize respiratory status: Treat pneumonia and control bronchospasm.
- Correct anemia.
- Improve nutritional status.
- Confirm availability of blood.
- Continue acid-suppressing therapy.

Monitoring

- Consider arterial line.

Induction

- For pts at risk for aspiration, utilize rapid-sequence induction with cricoid pressure.
- For pts with RAD, ensure adequate depth of anesthesia prior to instrumenting the airway.

Maintenance

- No one anesthetic is preferred.
- Avoid N_2O in laparoscopic procedure.
- Esophageal bougie may be required.
- Watch for possible pneumothorax, trauma to viscera, hemorrhage, and vena cava compression or laceration. Air or carbon dioxide embolism may occur during laparoscopic procedures.
- During laparoscopic procedures, intra-abdominal pressures of ≤12 mm Hg should be maintained.

Extubation/Postoperative Period

- May be extubated after uncomplicated surgery.
- Pts with severe respiratory compromise preop or with neurologic impairment may require a period of postop ventilation.
- Analgesic requirements will be less after laparoscopic procedures.

Surgical Procedure

- Fundus of the stomach is wrapped around the lower part of the esophagus. May be accomplished either open or laparoscopically.
- Pyloroplasty may be performed for associated delayed gastric emptying.
- Pneumoperitoneum created during laparoscopic surgery will result in increased SVR, increased CVP, increased CO, and increased BP. Intra-abdominal pressures >20 mm Hg will decrease venous return and decrease CO, but the BP will remain unchanged due to increased SVR.
- Pneumoperitoneum will also elevate the diaphragms, which will decrease lung volumes, decrease FRC, decrease pulm compliance, increase airway resistance, and increase V/Q mismatch.
- Pneumoperitoneum should not exceed 12 mm Hg. Pts are placed in the reverse Trendelenburg position. This will help ameliorate both diaphragmatic elevation and the CVP elevation.
- Pneumoperitoneum is accomplished by the insufflation of CO_2, which may necessitate increased minute ventilation.
- Laparoscopic procedures are associated with reduced rates of postop respiratory and wound complications and analgesic requirements, and shorter hospital stays.

Anticipated Problems/Concerns

- Respiratory system may be compromised.
- Pts are unable to vomit postop and up to 3 mo after surgery. Therefore, intestinal obstruction in the postop period should be treated as a dire emergency.

Gaucher Disease

Sydney E. Rose

Risk

- General population: 1:50,000 to 1:100,000.
- Inheritance follows an autosomal recessive pattern.
- Type 1 (nonneuropathic) most common and represents 99% of cases.
  - Population with highest prevalence is Ashkenazi Jewish.
  - 1:18 carrier rate
  - 3:1000 of the Ashkenazi Jewish population have type 1 disease.
- Types 2 (infantile) and 3 (juvenile) are exceedingly rare
  - No specific populations are at elevated risk.

Perioperative Risks

- Upper airway obstruction
- Coagulopathies
- GERD
- Insulin resistance

Worry About

- Glycosphingolipid deposits in head and neck
- Chronic aspiration
- Interstitial lung disease
- Cardiovascular calcifications
- Hepatosplenomegaly
- Osteonecrosis
- Coagulopathies
- Hematologic malignancies
- Polyneuropathies
- Parkinson disease

Overview

- Inborn error of metabolism.
- Lysosomal storage disorder due to defect or deficiency of the enzyme glucocerebrosidase.
- Nondegraded glucocerebroside and other glycolipids build up in macrophage and other peripheral leucocyte lysosomes.
- Lipid-laden macrophages then deposit in the spleen, liver, bone marrow, and other visceral organs, setting off an inflammatory and hyperplastic cellular response.
- Disease severity ranges from fatal in the perinatal period to completely asymptomatic and incidentally detected.
- Presenting features are variable and may occur at any age.
- Typically, visceral organs, bone marrow, and bones are involved in all three types.
- Type 1 can be differentiated from types 2 and 3 by lack of CNS involvement.
- Types 2 and 3 are both considered neuropathic:
  - Type 2, or "infantile GD," is acute in nature with onset occurring typically within the first year.
  - Type 3, or "juvenile GD," is more subacute, and disease progression is generally slower.
- Splenomegaly is typically the initial presenting sign.

Etiology

- Autosomal recessive trait with variable penetrance.
- Caused by mutations in the GBA gene located on chromosome 1q21.
- Greater than 200 mutations have been reported.
- 80% of mutations are caused by single nucleotide substitutions.
- A parent with GD has a 100% chance of passing on the gene.
- If the other parent is heterozygous for a mutated GBA gene, the child has a 50% chance of getting the disease.
- If parents are both heterozygous carriers, there is a 25% chance a child will be born with GD.
- Disease severity prediction based on genotypes is difficult due to the range of phenotypic variability.

Usual Treatment

- Treatment depends on severity of the disease.
- ERT with recombinant glucocerebrosidase in pts with GD1.
- Substrate reduction therapy reduces glycolipid buildup by decreasing the amount of glucocerebroside synthesized in patients unable to get ERT.

Assessment Points

System	Effect	Assessment by Hx and PE	Test
NEURO*	Apnea/asphyxia Spasticity Seizures Ataxia	Progressive brainstem degeneration Hyporeflexia Jerking movements Chorea Hypertonia/myoclonus Severe developmental delay Postop delirium	Reflex hammer Age-appropriate cognitive testing MRI brain
HEENT	Glycosphingolipid accumulation in head and neck	Ophthalmoplegia Strabismusa* Hx of upper airway obstruction Difficult airway Small mouth Trismusa*arching of neck	Vision testing Airway exam ENT consult
CV	Infiltration of myocardium by Gaucher cells Increase in LV mass Septal muscular prominence Apical akinesis structural pericardial changes Aortic and mitral valve pathologies Constrictive/hemorrhagic pericarditis	Arrhythmias Decreased exercise tolerance Cardiomyopathy Calcifications in ascending aorta, mitral valve, aortic valve CHF	ECG ECHO (TEE) Cardiac cath
ENDO	Glucocerebroside accumulation in lysosomes Peripheral insulin resistance Increase basal glucose production	Hypoglycemia	glucocerebroside and Phosphatase activity assays Blood glucose/HbA_{1c} Serum insulin assay
RESP	Post-extubation laryngospasm and/or respiratory failure Pulm Htn Cor pulmonale Intrapulmonary shunting Restrictive lung disease	History of choking/aspiration, may be chronic Chronic cough Pneumonia Glycosphingolipid deposits in lungs Kyphoscoliosis	CXR ABG PFTs LFTs Bronchoscopy ECHO
GI	GERD Portal Htn Splenic infarct	Epigastric pain GERD Weight loss Malnutrition	Barium swallow EGD Serum albumin assay Serum transferrin assay
HEME	Anemia Thrombocytopenia Acquired coagulation deficiencies in factors IX, XI, and vWF	Bleeding diathesis	CBC PT/PTT/INR BMP Glucocerebrosidase assay
MS	Pathologic osseous fractures Kyphoscoliosis Avascular necrosis of femoral head	Painful bone crises Fevers of unknown origin Decreased bone density in long bones Vertebral collapse Pes cavus	Bone density scan Bone marrow biopsy X-rays CT scan when necessary
IMMUNE	Infiltration of lymph nodes and lymphoid tissue Lymphoid tumors	Enlargement of lymphoid tissue lymph nodes, thymus, and tonsils Presence of peyer patches	US neck FNA lymphoid tumor
DERM	Pigmentation changes	Yellowish brown skin discoloration on face and/or lower extremities	Skin exam

*Types 2 and 3 only.

Key References: Kita T, Kitamura S, Takeda K, Fukumitsu K, Kinouchi K: Anesthetic management involving difficult intubation in a child with Gaucher disease, *Masui* 47(1):69–73, 1998; Dell'Oste C, Vincenti F: Anaesthetic management of children with type II and III Gaucher disease, *Minerva Pediatr* 49(10):495–498, 1997.

Perioperative Implications

Preoperative Preparation

- Evaluate for signs of neurologic, pulmonary, cardiac, hepatic, and bone marrow dysfunction.
- Warm the room.
- CBC, BMP.
- ST segment analysis in pts with signs or history of CAD.
- Consider TEE +/– PA catheter in surgeries expected to have large fluid shifts or in pts with infiltration of myocardium with Gaucher cells.

Monitoring

- A-line if comorbidities dictate.
- CVP +/– PA cath if pulm Htn or cardiac comorbidities dictate.
- Routine monitors.

Airway

- Thorough airway history and examination; discuss previous airway manipulation and evaluate oropharynx for deformities, fullness, and trismus.
- Prepare for possible difficult airway; glycosphingolipid deposits can narrow the upper airway yet be difficult to identify on PE.
- Smaller ETTs readily available.
- Have multiple airway devices available for multimodal airway management (LMA, video laryngoscope).
- Consider awake fiberoptic intubation.
- If warranted, have surgeon available at bedside for possible tracheostomy.

Positioning

- Bone fragility may be present; double-check pressure points and padding are adequate.

Induction

- Pts with swallowing difficulties are at risk for aspiration; consider rapid sequence.

Intubation
- Trismus resolves with muscle relaxant.
- If cardiac involvement, avoid agents that depress cardiac contractility.

Maintenance
- Choose drugs based on hemodynamic status.
- If hepatosplenomegaly or liver disease is present, avoid muscle relaxants dependent on hepatic metabolism.
- Regional anesthetics have been used and may be beneficial in minimizing airway manipulation and aiding in postoperative pain control. Bleeding dyscrasias must be ruled out prior to having a regional anesthetic.

Emergence/Extubation
- Fully awake and supine pts are ideal.
- Check for cuff leak if airway felt "tight" on intubation.
- Potential for prolonged ventilation, especially chronic aspirators.
- Rapid emergence agents (propofol, remifentanil, desflurane) will help facilitate process especially if neurologic component.
- Avoid hypoxia and hypercarbia.

Regional Anesthesia
- Thrombocytopenia and/or factor deficiencies may preclude regional anesthesia.

Adjuvants
- Dexmedetomidine 0.5 mcg/kg can be administered for postop delirium or agitation, especially in children.
- IV acetaminophen 1 g (adults) for pain control if no hepatic involvement; children may receive up to 15 mg/kg.
- H_2 blocker or PPI IV if symptomatic GERD preop.

Anticipated Problems/Concerns
- Airway difficulty during induction and intubation, especially if Hx of dysphagia or upper airway obstruction.
- If present preop, aspiration risk may still continue postop; consider leaving an NG or OG tube.
- Postop airway obstruction due to swelling of lymphoid tissue in head and neck area.
- Postop respiratory failure.
- Postop muscle spasms.
- Postop bleeding, especially if known bleeding dyscrasia preop.

Glaucoma, Closed-Angle

Steven Gayer

Risk
- According to the WHO, glaucoma is the second most common cause of blindness worldwide.
- Risk factors include hyperopia (far-sightedness), age >60, female gender, and family Hx.

Perioperative Risks
- Postop vision loss
- Inducing acute ACG

Worry About
- Causing sustained, marked elevations in IOP
- A chronic, narrowed angle becoming acutely closed periop

Overview
- There are a number of variants of glaucoma, and terminology can be confusing. These include acquired versus congenital, high IOP versus normal pressure, acute versus chronic, and open angle versus closed angle.
- ACG is categorized as either acute or chronic. Acute ACG is an urgent condition; chronic ACG is far more common and often asymptomatic.

Etiology
- ACG occurs when the distance at the outer periphery of the globe between the iris and cornea diminishes. Some individuals are born with narrowed angles and as they age and the lens thickens, further compromising the space.
- In predisposed individuals, chronic narrow-angle glaucoma may acutely progress to full-angle closure. Acute-angle closure occurs when the iris moves into direct contact with the cornea, physically blocking the natural egress of aqueous fluid.

Usual Treatment
- Acute ACG:
  - Administer topical beta-blocker, α2-agonist, pilocarpine 2% or 4% (pilocarpine is effective in inducing miosis only when iris ischemia is relieved, i.e., when IOP falls to <50 mm Hg).
  - Administer IV/oral acetazolamide 5–10 mg/kg (alternatives include hyperosmotic agents, e.g., IV 20% mannitol 1–2 g/kg, oral 50% glycerol 1–1.5 g/kg [contraindicated in diabetics], oral isosorbide 1.5–2.0 g/kg).
  - Topical steroids.
  - Place pts in the supine position (to allow lens-iris diaphragm to move posteriorly).
  - Analgesia and antiemetics.
  - Laser iridotomy.
- Chronic ACG:
  - Reduce IOP with prostaglandins (latanoprost, bimatoprost, travoprost).
  - Other surgical options include trabeculectomy, gonioplasty, or lens extraction.
  - Trabeculectomy, the gold standard procedure, involves creating a small hole in the sclera to allow freer drainage of aqueous humor.
  - Drainage device implants involve insertion of a tube shunt into the anterior chamber.
  - Cataract surgery may relieve a narrowed drainage angle.

Assessment Points

System	Effect	Hx	PE	Test
HEENT	Acute ACG	Sudden unilateral painful eye Blurred vision Photophobia Colored halos around lights Headache N/V	Ocular injection Hazy cornea Mid-dilated pupil	Penlight Gonioscope Slit-lamp Ultrasound biomicroscopy
	Subacute ACG	Headaches (often mistaken for migraine) or asymptomatic		
	Chronic ACG	Generally asymptomatic		

Key References: Gayer S: Prone to blindness: answers to postoperative visual loss, *Anesth Analg* 112(1):11–12, 2011; Gayer S, Gedde SJ: Intraoperative management of increased intraocular pressure in a patient with glaucoma undergoing robotic prostatectomy in Trendelenburg position, *Anesth Analg Case Rep* 6(2):19–21, 2015.

Perioperative Implications

Preoperative Preparation
- Consider preop consultation with pt's ophthalmologist if planned procedure involves prolonged steep Trendelenburg or prone position.
- Avoid mydriasis, either due to stress, dim lighting, or drugs (particularly topical sympatholytic or parasympathomimetic agents).
- Consider checking lytes if pt is on a diuretic.
- Preop antisialagogues or scopolamine are microfractionally absorbed into the globe and thus are considered generally safe to administer parenterally. Antimuscarinic ophthalmic drops (atropine, scopolamine) can induce acute ACG.
- Glaucoma surgery is generally considered to be low risk for sight-threatening bleeding. The consensus of studies exploring this controversial issue suggests that surgery can be safely performed under regional anesthesia without the need to discontinue antithrombotic agents.
- Phospholine iodide (echothiophate) should be discontinued 4 to 6 wk prior to surgery. Systemic absorption can inhibit plasma cholinesterase, causing prolonged muscle paralysis after succinylcholine, as well as inhibit metabolism of ester local anesthetics, predisposing a pt to toxicity.
- Regional anesthesia:
  - Needle-based block: Intraconal (retrobulbar) or extraconal (peribulbar)
  - Cannula-based block: Sub-Tenon's