# Gastrinoma

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## Risk

- Occurs in about 0.1–3 cases per million, usually in pts aged 20–50 y; male/female predominance about equal.
- 65–80% of gastrinomas are sporadic and have high (40–85%) malignant potential.
- 20–35% of cases occur with MEN1, with low (7–12%) malignant potential.
- Less than 0.1% of all PUD is caused by gastrinomas.

## Perioperative Risks

- Risks associated with PUD
- Associated abnormalities of MEN1
- Risks associated with metastatic disease (regional lymph nodes, liver, bone)

## Worry About

- Large gastric fluid volume
- Esophageal reflux (common)
- Lyte imbalance and volume depletion secondary to secretory diarrhea
- Coagulopathy due to liver metastases/resection

## Overview

- Gastrinoma is a gastrin-secreting neuroendocrine tumor (non-beta islet cell tumor), occurring most commonly in duodenum and/or pancreas.
- Gastrin release stimulates gastric acid hypersecretion, causing symptoms of abdominal pain (due to refractory peptic ulcer disease), diarrhea, and reflux.
- Diagnosis often delayed several years from onset of symptoms because of difficulties distinguishing gastrinoma from other causes of PUD.
- Symptom control with PPIs also may delay diagnosis.
- May occur with other functional pNETs (e.g., carcinoid, insulinoma, parathyroid hormone-related peptide secreting tumor, Cushing).
- See also Multiple Endocrine Neoplasia Type I and II, Cushing Syndrome, Carcinoid Syndrome, Insulinoma, and Hyperparathyroidism for more information.

## Usual Treatment

- Control gastric acid hypersecretion with PPIs and $H_2$ blockers.
- Chemotherapy, mTOR or tyrosine-kinase inhibitors, and/or somatostatin analogues for metastatic disease.
- Surgical exploration and resection.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| CV | Hypovolemia<br>Right-sided valvular disease*<br>Tachyarrhythmias* | Weakness, dizziness<br>Dyspnea, edema, ascites, palpitations | Vital signs<br>Cardiac murmur<br>Cardiac exam | Orthostatics<br>ECHO<br>ECG |
| RESP | Wheezing* | Dyspnea | Pulmonary exam | CXR |
| GI | Gastric acid hypersecretion | Abd pain, reflux, diarrhea | Abdominal exam | Fasting gastrin, secretin stim |
| METAB | Hypokalemia | Weakness, muscle cramps | | Lytes, ECG |
| ENDO | Hyperparathyroidism*<br>Hypoglycemia* | Confusion, nausea/vomiting, abdominal pain, nephrolithiasis<br>Dizziness, confusion | Mental status exam, abdominal exam | Serum parathyroid hormone, serum calcium<br>CBG |
| RENAL | Nephrolithiasis* | Flank pain, hematuria | CVA tenderness | Urinalysis |
| CNS | Pituitary adenoma* | Headaches, visual changes | Visual field exam | MRI, prolactin level |
| MS | Weakness, arthralgias* | Proximal muscle weakness | Strength, reflexes | Serum calcium |
| HEME | Coagulation disorder | Bleeding abnormalities | | INR/PT, PTT |

*If gastrinoma presents as component of MEN I or other functional pNETs.

**Key References:** Ito T, Igarashi H, Robert J: Zollinger-Ellison syndrome: recent advances and controversies, *Curr Opin Gastroeterol* 29(6):650–661, 2013; Perry RR, Feliberti E, Vinik A: Gastrinoma Zollinger-Ellison-syndrome. In De Groot LJ, Beck-Peccoz P, Chrousos G, et al., editors: *Endotext* [Internet], South Dartmouth, MA, 2000, MDText.com, Inc., updated 2013.

## Perioperative Implications

### Preoperative Preparation

- Ensure adequate treatment of gastric hypersecretion.
- Evaluate for other endocrinopathies of MEN I syndrome.
- Evaluate for other functional pNETs; plan intraop management accordingly.
- Assess volume status.
- Check lytes and coagulation tests.
- Consider epidural catheter for intraop/postop pain control if no contraindications.
- Consider preop NG tube placement.

### Monitoring

- May require central venous pressure monitoring and arterial line, depending on associated symptoms and comorbidities. Measure urine output with a bladder cath.

### Induction

- Treat as full stomach due to increased gastric acid volumes and increased risk for aspiration.
- Use rapid-sequence induction with cricoid pressure.

### Extubation

- Ensure adequate respiratory function and neuromuscular blockade recovery before extubation.

### Postoperative Period

- Decreased vital capacity and functional residual capacity due to pain and ileus.

## Anticipated Problems/Concerns

- Continued symptoms may occur if surgical resection is not curative.
- There are lower cure rates after resection for pts with gastrinomas associated with MEN1 or in the presence of metastatic liver disease.

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# Gastroesophageal Reflux in Children

Francine S. Yudkowitz

## Risk

- Physiologic GER usually resolves by 12–15 mo of age.
- 10% of pyloric stenosis pts.
- After diaphragmatic hernia, tracheoesophageal fistula, and esophageal atresia repairs
- Neurologically impaired, developmentally delayed, trisomy syndromes, and hiatal hernia.

## Perioperative Risks

- Aspiration during induction of anesthesia
- Severe bronchospasm in pts with RAD
- Decreased pulm reserve secondary to chronic aspiration and pneumonitis

## Worry About

- Pulm complications from aspiration pneumonitis and RAD
- Anemia and malnutrition

## Overview

- Lower esophageal sphincter function matures by 6 wk postnatal age.
- GER is defined as regurgitation without pathologic consequences. GERD is defined as regurgitation resulting in esophagitis, nutritional compromise, and/or respiratory complications.
- Presence of a hiatal hernia does not necessarily mean pt will have GER.
- Sandifer syndrome: Opisthotonos or other abnormal head movements.
- Older children may complain of heartburn, dysphagia, and chest and abdominal pain.
- Degree of reflux, duration of acid exposure in the esophagus, and ability of the esophagus to clear the

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<br>RAD | Cough, cyanotic episodes, apnea<br>Dyspnea, wheezing, cough | Rales, rhonchi<br>Wheezing<br>Decreased breath sounds, prolonged expiration | CXR, ABG (if indicated)<br>CXR, peak flow<br>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.