

## Risk

- 1:15,000-50,000 live births
- Most common human chromosomal syndrome

## Perioperative Risks

- Difficult airway management due to anatomic abnormalities of the airways.
- 30% can present with congenital heart disease.
- Aspiration risk.
- May require temp control.
- Cardiac events (pts may have cardiac abnormalities).

## Worry About

- Difficult mask ventilation; airway obstruction secondary to hypotonia

- Difficult intubation
- Temperature regulation

## Overview

- Microcephaly with profound mental retardation and hypotonia.
- Characteristic facies with micrognathia, low-set ears, facial asymmetry.
- Characteristic high-pitched cry may be due to laryngeal abnormalities (narrow diamond-shaped larynx, long floppy epiglottis) or neurogenic defect.
- CHD: Persistent ductus arteriosus, septal defects, or pulmonic stenosis.

- Behavioral features include repetition of movements, aggression, self-injury, and hypersensitivity to sound, among others.
- Repetitive respiratory infections are common.

## Etiology

- Deletion of variable size of the short arm of chromosome 5 (5p-).
- Loss of the critical 5p15.2 region is responsible for most of the features.
- Most cases occur by spontaneous gene mutation (90%).
- 10% arise by unbalanced translocations.

## Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Micrognathia	Respiratory distress in neonatal period; inspiratory stridor	Receding mandible	
CV	ASD, VSD, PDA, PS	SOB, cyanosis	Murmur, gallop	ECHO
RESP/GI	Pneumonia, chronic aspiration		Dyspnea, rales, rhonchi, wheezing	CXR
MS	Scoliosis			
CNS	Mental retardation, seizures		Hypotonia in infancy, hypertonia later	

**Key References:** Rodríguez-Caballero A, Torres-Lagares D, Rodríguez-Pérez A, et al.: Cri du chat syndrome: a critical review, *Med Oral Patol Oral Cir Bucal* 15(3):e473–e478, 2010; Santos KM, Rezende DC, Borges ZD: Anesthetic management of a patient with cri du chat syndrome. Case report, *Rev Bras Anestesiol* 60(6):630–633, 2010.

## Perioperative Implications

### Preoperative Preparation

- Difficult airway management

### Monitoring

- Routine
- Pay particular attention to temp and neuromuscular blockade (fast-acting preferred).

### Airway

- Laryngeal mask airway must be available.
- Fiberoptic bronchoscope ready to use before induction.

- Wide assortment of laryngoscope blades and ETT must be available.

### Preinduction/Induction

- Presence of primary caregiver for uncooperative pts
- Sedation in monitored setting
- Warm OR for temp regulation

### Maintenance

- Measures to actively warm the pt (forced-air warming blankets, warm IV fluids).
- Monitor neuromuscular blockade.

### Extubation

- Preferably extubate awake.

## Anticipated Problems/Concerns

- CHD may be present: Refer to cardiac assessment before anesthesia.
- Airway management may be difficult.
- Extubation may be difficult; pt may have airway obstruction postoperatively.

# Crohn Disease

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## Risk

- Incidence of 3.1–14.6 cases per 100,000 person-years; prevalence of 26–201 cases per 100,000 persons.
- Incidence and prevalence are increasing worldwide; incidence is highest in North America and Northern Europe.
- Race: White >African American >Hispanic and Asian populations for risk.
- 3–4 times more common in ethnic Jews than non-Jewish whites.
- More likely to occur in those with a strong family history.
- Affects males and females equally.
- Peak occurrence between ages 15 and 25 y, with a second smaller peak between ages 60 and 80 y.

## Perioperative Risks

- Aspiration
- Arrhythmias due to lyte disorders

## Worry About

- Intravascular fluid volume and lyte imbalances.
- Chronic steroid use and need for perioperative supplementation.

- Nutritional status, chronic weight loss, and malnutrition.
- Difficult IV access due to chronic illness and frequent venipunctures.
- Higher risk of systemic thromboembolic events in comparison with control populations.
- Psychological mindset of the patient due to chronicity of the disease and relatively young age of pts. Depression is common.

## Overview

- Chronic inflammatory disease of GI tract that can give rise to strictures, inflammatory masses, fistulas, abscesses, and hemorrhage
- Idiopathic, chronic relapsing immune-mediated disease
- May affect any portion of the GI tract from mouth to anus, most commonly affects small bowel and colon
- Pts often present with abdominal pain, persistent diarrhea, and weight loss
- Pt may develop bowel obstruction and perforation
- Pt may develop rectocutaneous fistulas, rectal fissures, and perirectal abscesses
- Pt may have anemia from several causes, including chronic disease, chronic blood loss, and folate and vitamin B<sub>12</sub> deficiency

- Chronic malnutrition and weight loss
- Extraintestinal manifestations occur in approximately 25–30% of pts. These manifestations include uveitis and episcleritis, erythema nodosum and pyoderma gangrenosum, ankylosing spondylitis, and primary sclerosing cholangitis. When present, the extraintestinal manifestations can be more serious than the primary intestinal disease. These extraintestinal manifestations may precede, occur with, or manifest independently of the underlying bowel disease

## Etiology

- Pathogenesis incompletely understood.
- Thought to arise from environmental priming and triggering events in a genetically susceptible pt.
- Smoking is a risk factor for Crohn disease and worsens the course of Crohn disease.

## Usual Treatment

- Pharmacologic: Aminosalicylates; steroids; immunomodulating agents, such as azathioprine, 6-mercaptopurine, and methotrexate; antitumor necrosis factor therapy with infliximab, adalimumab, or certolizumab pegol.

- Surgical: Indications for surgery are failure of medical management, intestinal obstruction, intra-abdominal abscess, fistulas, fulminant colitis, toxic megacolon, massive hemorrhage, cancer, and growth retardation; 70–90% of Crohn disease patients will need surgical intervention at some point.
- Surgical procedures may include stricturoplasty, bowel resection, and abscess drainage.
- Main surgical principle is to preserve bowel length to avoid short bowel syndrome.
- Both medical and surgical management of Crohn disease are aimed at providing long-lasting symptomatic relief while avoiding excessive morbidity.

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Hypovolemia	Bowel prep, wt loss, diarrhea	Hypotension, tachycardia	Lytes, Hct
GI	Bowel perforation Malabsorption	Abdominal pain Diarrhea, weight loss	Abdominal tenderness, fever Cachexia	WBCs Albumin
MS	Ankylosing arthritis	Joint mobility	Decreased ROM of joints	

**Key References:** Baumgart DC, Sandborn WJ: Crohn's disease, *Lancet* 380(9853):1590-1605, 2012; Patel S, Lutz JM, Panchagnula U, et al.: Anesthesia and perioperative management of colorectal surgical patients—a clinical review (Part 1), *J Anaesthesiol Clin Pharmacol* 28(2):162–171, 2012.

### Perioperative Implications

#### Preoperative Preparation

- Optimization of nutritional status preop can improve operative outcomes.
- Ensure volume status and lytes are normalized.
- If pt is on hyperalimentation preop, continue it during the case; monitor glucose.
- Assess current or recent steroid use and need for periop supplementation.
- Discontinue methotrexate at least 1 wk before surgery because it has been shown to decrease wound healing; resume after wound healing occurs.
- Pts with significant anemia should be transfused preop.
- Careful pt positioning and padding of extremities.

#### Monitoring

- Standard monitoring.
- Large-bore PIV access.
- Consider CVL if pt has difficult IV access or if patient is hypovolemic or large fluid shifts are anticipated.

- Consider arterial line if significant comorbidities exist.

- Foley catheter to monitor urine output.

#### Airway

- Aspiration risk if bowel obstruction present

#### Induction

- Rapid sequence induction in patients with gastric outlet or bowel obstruction.
- Consider preinduction placement of NG tube to suction gastric contents.

#### Maintenance

- Avoid nitrous oxide if bowel obstruction present.
- Abdominal relaxation with nondepolarizing muscle relaxants usually needed. If liver disease is present, avoid muscle relaxants dependent on hepatic metabolism.
- Check glucose regularly if on hyperalimentation.
- Consider need for significant fluid administration (open abdomen, long case).

- Maintain normothermia; fluid warmer and forced air warming device.

#### Extubation

- Awake extubation

#### Postoperative Period

- Consider epidural analgesia or IV PCA for pain control.
- Monitor fluid status carefully in the postop period.

#### Anticipated Problems/Concerns

- Possibly long surgery due to adhesions and multiple strictures
- May need aggressive fluid replacement due to hypovolemia and anemia worsened by third space losses
- May have severe nutritional deficiency, especially with short bowel syndrome from extensive resection
- Need for stress dose steroids if patient treated with steroids for medical management

## Croup (Laryngotracheobronchitis)

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### Risk

- Children between 6 mo–6 y are at risk (6 mo–3 y at greatest risk).
- Children with underlying airway abnormalities (e.g., subglottic stenosis) or difficult intubations (e.g., micrognathia) and symptoms are at increased risk and require particular planning.

### Perioperative Risks

- Difficulty with intubation because of very narrowed subglottic region
- Obstruction of the small tracheal tube because of airway secretions.

### Worry About

- Risk of rebound tracheal edema several hours after racemic epinephrine treatment.
- Cardiorespiratory crisis in progressive or severe Sx, agitation, younger pts, difficulties with oxygenation or ventilation, failure to oxygenate.
- Bacterial superinfection of airway.

### Overview

- Common childhood ailment with prodromal illness accompanied by a characteristic cough (which often sounds like seal barking).
- Sx and respiratory compromise from progressive swelling of subglottic region tracheal mucosa.
- Frequently present when inspiratory stridor and respiratory distress develop.
- Radiographs of the neck often demonstrate gradual progressive tracheal narrowing; most narrow just below level of vocal cords (referred to as steeple sign). Upper glottis on a lateral neck radiograph is normal.
- When obtained, evaluation of CBC is consistent with viral illness.

### Etiology

- Viral agents are typical etiologies and include parainfluenza viruses (most common). Adenoviruses, influenza virus, RSV, and measles virus also associated.

### Usual Treatment

- Cool mist often greatly improves Sx; supplemental O<sub>2</sub>.
- If symptoms more severe, aerosolized racemic epinephrine can dramatically reduce airway swelling (rebound tracheal edema risk several hours after administration necessitates observation in hospital).
- Steroid administration controversial; may decrease severity of disease and decrease need for tracheal intubation or hasten improvement in first 24 h of illness.
- Small percentage of pts with this disease require tracheal intubation.
- Parenteral steroids (dexamethasone) and inhaled steroids (budesonide) have been used.
- Breathing helium-oxygen mixtures has been reported as helpful in some cases (lower density and viscosity).