Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Enamel hypoplasia: Blisters, microstomia, ankyloglossia, supraglottic ulceration or narrowing,corneal erosion, ectropion	Delayed eruption and caries of teeth; painful perioral and intraoral lesions; hoarseness and respiratory obstruction; painful swallowing, spasm, food impaction	Poor oral hygiene and malocclusion; tongue atrophy; obliteration of vestibular sulci, stricture, webs, and vocal cord lesions	Airway assessment, endoscopy
GI	Bullae Perianal blisters, poor absorption, diarrhea	Esophageal stricture Anal pain, tenesmus, constipation	Reflux, regurgitation Anal fissure or stricture	Endoscopy
GU	Blisters	Urinary diversion	Obstruction, sepsis	Renal function
MS	Contractures, growth retardation	Movement limitations, stature	Flexion contracture, pseudosyndactyly	
DERM	Blisters	Age at onset, Hx of remissions and infections	Scars, milia, nail dystrophy, cancer	Skin biopsy

Key References: Lin YC, Golianu B: Anesthesia and pain management for pediatric patients with dystrophic epidermolysis bullosa, *J Clin Anesth* 18(4):268–271, 2006; Saraf SV, Mandawade NJ, Gore SK, et al.: Epidermolysis bullosa: careful monitoring and no touch principle for anesthetia management, *J Anaesthesiol Clin Pharmacol* 29(3):390–393, 2013.

Perioperative Implications

Preoperative Preparation

- Careful planning of monitoring, IV placement, positioning in the OR, prevention of reflux, and airway management

Monitoring

- No contraindication to pulse oximeter use.
- Protect blisters on the face with foam adhesive inverted to pad mask.
- Pad automated BP cuff heavily and limit intervals.
- Cut off adhesive from ECG leads and hold in place with defibrillator jelly pads.
- Suture invasive monitoring and IVs or wrap in place with petrolatum gauze.
- Esophageal stethoscope may damage mucosa.
- Avoid excessive heat or sweating, which increases the risk of blisters.

Induction

- Regional anesthesia encouraged; use spray antiseptics or pour prep solutions; no intradermal local anesthetics.
- No GA or muscle relaxant specifically contraindicated.

Airway

- All airway management techniques are reported successful.
- The mask (or nasal mask) should be lubricated and padded with petrolatum gauze; pad the chin under fingers; bullae occurred in 1:50.
- LMA one size too small, heavily lubricated, cuff soft with audible leak, extubated deep to prevent trauma; lingual bulla occurred in 1:57.
- Intubation is less frequent; use blind nasal, fiberoptic, and oral techniques; a heavily lubricated small tube and laryngoscope; cricoid pressure without lateral movement is permissible; 66% are class I or II view of larynx and 7–23% have difficult airway incidence; use soft lubricated gauze to prevent tube movement in the mouth; do not allow lateral forces on the corners of the mouth by the tube and do not use tape; the trachea is lined with columnar epithelium and, therefore, is less likely to blister.

Emergence

- Aim for a quiet emergence.
- No suction on intraoral mucosa.

Anticipated Problems/Concerns

- Positioning is performed by the pt if possible; lateral shear forces from lifting cause blisters.
- Corneal abrasion can occur because of poor eyelid retraction; use ointment generously and protect the eyes in while pt is in prone position.
- Treat hemorrhage with epinephrine or a thrombin-soaked sponge.
- Avoid sweating and warming devices, but if unavoidable, the device should be no warmer than skin temperature.
- Extremity tourniquets, IM or rectal medications and EMLA can be used.
- Common procedures include release of syndactyly, dressing change, squamous cell carcinoma, esophageal dilatation, and dental surgery.

Epiglottitis

Maurice S. Zwass | Jeffrey D. Roizen

Risk

- Prevalent in children 1–7 y; sometimes called supraglottitis, it does occur in adults (decreasing incidence in children >3 y related to vaccines against *Haemophilus influenzae* type B, but still found, particularly if pt is not immunized).
- Adult incidence remains constant with organisms group A *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Klebsiella pneumoniae*.

Perioperative Risks

- Acute deterioration of airway patency resulting in complete obstruction worse in children
- Difficulty in tracheal intubation due to severe edema of epiglottis and arytenoids

Worry About

- Airway compromise in children who appear toxic, with increasing distress, drooling, and hypoxemia. The acute risks of airway compromise (of concern in small children) appear to be less critical in adults, most likely because of larger airways.
- Loss of airway control and aspiration.

Overview

- An acute, potentially life-threatening cause of upper airway obstruction (etiologic agents may include bacteria other than *H. influenzae* type B).
- Produces inflammatory edema of epiglottis and other supraglottic structures.
- Onset is usually rapid; progression to severe obstruction can occur in several hours.
- High fever, sore throat, and dysphagia are frequently so severe that swallowing is inhibited and drooling results.
- Differential diagnosis also include retropharyngeal abscess (a bacterial infection), which can have the same presentation. It can be differentiated from epiglottitis by the presence of torticollis and trismus and with radiographic studies (contrast CT). Treatment is with antibiotics and surgical drainage.

Etiology

- *H. influenzae* type B is most often traditional associated pathogen, although this can be caused by β-hemolytic streptococci, group A *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Klebsiella pneumonia*.

Usual Treatment

- Antibiotic therapy against bacterium (usually *H. influenzae*) and airway support, which generally requires tracheal intubation.
- Because of high incidence of ampicillin-resistant strains, administer ampicillin plus a β-lactamase inhibitor (such as sulbactam) and/or chloramphenicol, cefuroxime, ceftazidime, or another penicillinase-resistant antibiotic as indicated by blood and epiglottis culture results.
- Tracheal intubation is classically performed in OR in a controlled fashion with surgical support for possible tracheotomy or cricothyrotomy present and gowned.

Assessment Points

Differentiation Between Epiglottitis and Croup (Laryngotracheobronchitis)

	Croup	Epiglottitis
Age	3 mo–3 y	1–7 y
Onset	Gradual	More rapid (usually <24 h)
Fever	Low-grade	High-grade
Cough	Characteristic barking	None
Sore throat	Occasional	Frequently severe
Posture	Any	Frequently sitting forward, mouth open, drooling
Airway sound	Inspir stridor	Inspiratory stridor
Voice	Normal	Muffled
Appearance	Nontoxic	Toxic
Seasonality	Peak winter, epidemic	Year-round

Radiographic studies may be helpful, because AP view of trachea appears normal but lateral neck view usually shows a markedly swollen, edematous epiglottis ("thumb-printing").

Key References: Jenkins I, Saunders M: Infections of the airway, *Paediatr Anaesth* 19(Suppl 1):118–130, 2009; Tibballs J, Watson T: Symptoms and signs differentiating croup and epiglottis, *J Paediatr Child Health* 47:77–82, 2011.

Perioperative Implications

Preoperative Preparation

- With suspected epiglottitis, other personnel on pt care team can set up care (e.g., OR, ICU). Radiographs can be obtained, but a team member capable of monitoring and securing the airway should be present.
- Allow pt to remain in a position of comfort (often sitting with parent). Direct exam of oropharynx is generally avoided, as are attempts to secure vascular access, because these may cause agitation leading to acute tracheal obstruction.
- Humidified O_2 should be delivered as tolerated.
- Aerosol therapy with racemic epinephrine may provide slight improvement of symptoms, but not definitive. If Dx is confirmed, pt is taken to the location for intubation (most commonly OR).

Airway Management

- For anesthesia, sevoflurane or halothane and O_2, maintaining spontaneous ventilation.
- IV cath should be placed after induction of anesthesia, followed by direct laryngoscopy.
- Large, swollen epiglottis can make identification of airway structures difficult, but once the epiglottis is identified, arytenoids and larynx are immediately below and tracheal tube can be inserted.
- Because of upper airway swelling, a tracheal tube 0.5–1 mm smaller in diameter may be needed (tracheal tube of adequate length can be made available).
- Rarely is emergency tracheotomy necessary, but surgeons are "gloved" until airway is secured.
- Frequently the orotracheal tube is changed to a nasotracheal tube for ease of securing and pt comfort.

Post-Airway-Management Plans

- Once the airway secured, cultures of blood and epiglottis are obtained, antibiotic therapy is initiated, and sedation plans are instituted.

Anticipated Problems/Concerns

- Respiratory support often for 24–72 h until swollen epiglottis returns to normal.
- Pts usually require sedative management to facilitate tolerating mechanical ventilation.
- Many pts (~25%) have assoc pneumonia that requires treatment.

Fabry Disease

Rohesh J. Fernando

Risk

- Genetic disease with reported annual incidence of approximately 1:100,000, but rarity may lead to underestimation of true prevalence
- Panethnic

Perioperative Risks

- Autonomic instability, particularly with neuraxial anesthesia
- Cardiovascular instability from conduction abnormalities and structural heart disease
- Periop stroke in those with cerebrovascular disease

Worry About

- Poor respiratory function, particularly in smokers
- Renal impairment
- Sudden swings in blood pressure
- Pain control in pts with chronic pain
- Abnormal temperature regulation

Overview

- Second most common lysosomal storage disorder.
- Disease process begins as early as fetal development but clinical symptoms are often not evident until after age 3 y, occurring a few years later in girls versus boys.
- Classically, homozygous young males are severely affected.
- Disease affects multiple organ systems but most notably affects the cardiac, cerebrovascular, renal, respiratory, and peripheral nervous systems.
- Risk of end-organ damage increases with age.

Etiology

- X-linked inherited lysosome storage disorder due to deficiency of lysosomal alpha-galactosidase A, leading to accumulation of glycosphingolipids throughout the body

Usual Treatment

- Enzyme replacement therapy can reduce symptoms and complications.
- Conventional medical treatment for disease-related morbidities.
- Transplantation, particularly renal and cardiac, may be necessary.