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

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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.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Deafness Corneal and lenticular opacities Vertigo Mucosal lesions	Difficulty hearing Light sensitivity Dizziness Nausea (vertigo)	Impaired pupillary constriction, hearing loss, oropharyngeal mucosal lesions	Audiometry, ophthalmologic exam including slit lamp, visual acuity, and fields
RESP	Obstructive ventilatory defect Exercise intolerance Airway obstruction (bronchospasm)	Dyspnea Cough	Tachypnea Wheezing	CXR Spirometry Oximetry Treadmill exercise testing
CV	LVH, RVH, aortic dilation Diastolic dysfunction LVOT obstruction Mild valvular insufficiency Coronary artery stenosis	Palpitations, angina, dyspnea	Irregular heartbeat, heart murmur	ECG ECHO Stress imaging Holter monitor
GI	Difficulty gaining weight Delayed gastric emptying Achalasia	Postprandial abdominal pain, N/V, early satiety	Smaller height and weight compared with unaffected siblings	Endoscopic or radiographic evaluations
CNS	TIA/stroke Mild dementia (late finding) Autonomic dysfunction Acroparesthesias	Pain in extremities, cold/heat intolerance, joint pain, stroke symptoms	Neurologic exam, pain inventory, hypohydrosis, orthostatic hypotension	Brain CT or MRI with T1, T2, and FLAIR images
HEME	Abnormal vascular reactivity Prothrombotic state	Angina, stroke, DVT	Thrombophlebitis	Proteins C & S, factor V Leiden, prothrombin G20210A, ATIII, lupus anticoagulant, anticardiolipin antibody
RENAL	Renal failure Proteinuria Impaired concentration ability	Fluid retention	Edema	Electrolytes, BUN, creatinine, GFR, 24-h urine for total protein/Cr
MS	Angiokeratomas, osteopenia, osteoporosis	Osteoporotic fractures	Raised skin lesions	Bone mineral density

Key References: Eng CM, Germain DP, Banikazemi M, et al.: Fabry disease: guidelines for the evaluation and management of multi-organ system involvement, *Genet Med* 8(9):539–548, 2006; Woolley J, Pichel AC: Peri-operative considerations for Anderson-Fabry disease, *Anaesthesia* 63(1):101–102, 2008.

Perioperative Implications

Preoperative Preparation

- Cardiac evaluation with ECG and echocardiogram should be obtained. Consider noninvasive cardiac stress imaging in pts older than 30 y and with concerning symptoms.
- Consider sodium citrate for gastric prophylaxis in pts with symptoms of achalasia.
- Consider preop sedation to avoid excessive activation of abnormal autonomic nervous system.
- Obtain baseline visual exam to differentiate from new deficit after surgical positioning or hemodynamic instability.
- Recurrent pain in extremities may be a relative contraindication for regional anesthesia.

Monitoring

- Consider arterial line given potential autonomic instability and cardiac history.
- Consider CVP or PA cath as indicated.
- Temperature monitoring is especially important.

Airway

- Risk for difficult direct laryngoscopy as a result of TMJ stiffness leading to limited mouth opening.
- Inspect airway for oropharyngeal lesions.
- Risk of bronchospasm.

Preinduction/Induction

- Be prepared for BP swings.
- Anticipate need for bronchodilators and avoid drugs that may cause histamine release when possible.

Maintenance

- Monitor ECG vigilantly given risk for arrhythmias and conduction abnormalities.
- Vasoactive medications should be ready to treat both hypotension and Htn.
- Avoid nephrotoxic medications and administer judicious IV fluids accounting for any renal impairment.
- Warming and cooling equipment should be available given autonomic instability.
- If taking carbamazepine for pain control, increased amounts of nondepolarizing neuromuscular blockade may be required.

Extubation

- Pts with achalasia or delayed gastric emptying should be considered at risk for aspiration.
- Ensure neuromuscular blockade not prolonged from renal insufficiency.
- Anticholinergics may exacerbate hypohidrosis.

Postoperative Period

- Analgesia plan critically important for those with chronic pain

Anticipated Problems/Concerns

- Dose medications appropriately in those with renal impairment.
- Amiodarone can exacerbate lysosomal abnormalities and should be avoided.

Factor V Leiden Mutation

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Risk

- Most common hereditary thrombophilia
- Autosomal dominant inheritance pattern
- Heterozygous form in 5% of white population in USA (up to 15% in Europe), 2% of Hispanic Americans, 1% in both African and Native Americans
- Homozygosity in white population 1:5000
- May account for 85–95% of pts with APC resistance
- Relative risk of venous thrombosis sevenfold in heterozygous and 80-fold in homozygotes

Perioperative Risks

- VTE: DVT most likely; lower risk of PE
- Risk of arterial thrombosis unknown

Worry About

- Hypercoagulability
- DVT
- Recurrent fetal loss (twofold to fivefold increased relative risk)
- Conflicting data regarding association with placental abruption, severe preeclampsia, IUGR
- Cerebral vein thrombosis
- Renal transplant rejection
- Risk of thrombosis increased by protein S deficiency, prothrombin 20210 gene mutation, hyperhomocysteinemia, OCP use, pregnancy, increasing age, immobilization, and obesity

Overview

- Factor Va is a procoagulant that is inactivated by APC, with protein S as cofactor, causing less thrombin generation during the propagation phase.