

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Intravascular fat Hypoperfusion Pulm Htn RV failure	Fever Syncope Dyspnea	Hypotension Tachycardia Oliguria Vasoconstriction Mental status changes	Fat staining of blood Bronchoalveolar lavage, macrophage staining TTE/ TEE, CVP, PA cath Lactic acidosis
RESP	ARDS Hypoxemia	Dyspnea	Tachypnea Cyanosis Rales	Pulse oximetry CXR, ABGs Pulm compliance (on mechanical ventilation)
HEME	Thrombocytopenia DIC Anemia	Bleeding	Bleeding (rare)	CBC Plts PT, PTT D dimer Fibrinogen
DERM	Capillary fat embolism		Petechiae (60%) Axilla, chest Base of neck Conjunctiva Oral mucous membranes	
CNS	Neurologic injury Cerebral edema	Mental status changes	Delirium Confusion, agitation Focal deficits (rare) Seizure (rare) Coma (rare)	MRI

Key References: Akhtar S: Fat embolism, *Anesthesiol Clin* 27(3):533–550, 2009; Kwiatt ME, Seamon MJ: Fat embolism syndrome, *Int J Crit Ill Inj Sci* 3(1):64-68, 2013.

Perioperative Implications

Preoperative Preparation

- Avoid sedatives and/or narcotics if hypoxemic and not mechanically ventilated or with obtundation.

Monitoring

- Arterial cath
- TTE/TEE, CVP, and PA cath to diagnose and manage RV failure and/or pulm Htn

Airway

- May have ARDS; decreased FRC and O₂ reserve and limited tolerance for apnea.
- May already be intubated and ventilated in severe cases.

Induction

- Minimize myocardial depression.

- Avoid increases in PA pressures (hypoxemia, hypercarbia, acidosis).

Maintenance

- CV: Anticipate decrease in BP with femoral reaming/cementing; anesthetic reduction, fluid, vasopressors; pts with RV dysfunction may require longer-term inotropic support.
- Resp: Pts with ARDS may require increased FIO₂ and PEEP; use lung protective strategy, ARDSnet protocol.
- Heme: Factor replacement for coagulopathy with bleeding.

Extubation

- Maintain intubation and mechanical ventilation in hemodynamically unstable pts and those requiring increased FIO₂, PEEP or with reduced compliance.

- Pts with CNS involvement may have a prolonged or exaggerated response to anesthetics and narcotics and may require intubation postop for airway protection/patency.

Anticipated Problems/Concerns

- Embolism during femoral reaming, prosthesis cementing.
- FES may be delayed by up to 72 h following fat embolism.
- Pts with ARDS may be difficult to ventilate and oxygenate.
- Hypotension is due to RV dysfunction and pulm Htn.

Foreign Body Aspiration

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Risk

- Most prevalent in children ages ≤3 y.
- In adults, elderly are most susceptible to FB aspiration. Risk factors include Alzheimer disease or dementia, stroke, loss of consciousness due to trauma, alcohol intoxication, or drug overdose.
- Foods are most commonly aspirated foreign objects.

Perioperative Risks

- Hypoxemia due to FB obstruction
- Fragmentation of the FB and distal dislodgement during retrieval

- Severe inflammation due to presence of high oil contents in a FB, leading to bulky granulation resulting in bronchial stenosis, bronchiectasis, pneumonia, and lung abscess

Worry About

- Exacerbation of hypoxemia due to ineffective ventilation during either diagnosis or treatment secondary to sedation or prolonged periods of apnea during extraction of FB
- Prolonged extraction of FB, airway swelling, and bleeding, which may lead to further respiratory compromise

Overview

- Pts with FB aspiration may present with cough, stridor, wheezing, throat pain, drooling, dysphagia, respiratory distress, and hypoxemia (oxygen saturation <90%).
- Subglottic FBs are often aspirated into the right main bronchus, likely due to the less acute angles of the right bronchus.

Usual Treatment

- Flexible and rigid bronchoscopy.
- Bronchotomy or lung resection may be performed in rare occasions, especially when the FB aspiration is diagnosed late (> 1 wk).

Assessment Points

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RESP	Hypoxemia	Choking, coughing	Dyspnea, dysphagia, respiratory distress	Pulse oximetry, radiographic imaging, bronchoscopy

Key References: Won C, Michaud G, Kryger MH: Upper airway obstruction in adults. In Grippi MA, Elias JA, Fishman JA, et al., editors: *Fishman's pulmonary diseases and disorders*, ed 5, New York, NY, 2015, McGraw-Hill Education; Duan L, Chen X, Wang H, et al.: Surgical treatment of late-diagnosed bronchial foreign body aspiration: a report of 23 cases, *Clin Respir J* 8(3):269–273, 2014.

Perioperative Implications**Preoperative Preparation**

- Provide supplemental oxygen and monitor oxygenation levels (pulse oximetry).
- Avoid manipulation of airway to prevent further dislodgement of FB.
- Discuss with surgeon plan of anesthetic (MAC with topical anesthesia vs. GA) and agree on ventilation plan (spontaneous, controlled, or jet ventilation).
- Topical anesthesia with aerosolized 4% lidocaine may be beneficial, especially if using spontaneous ventilation technique.

Intraoperative Management

- Preoxygenate.

- Rapid sequence induction is likely necessary; however consider effect of fasciculation on dislodgement of FB if using succinylcholine.
- Literature is indecisive whether controlled ventilation is preferred over spontaneous ventilation.
- If pt is a child, consider use of inhalational induction using sevoflurane and O₂.
- Ensure availability of surgical airway kit in cases of aspiration of large supraglottic FB.
- Consider use of passive oxygenation via bronchoscope, especially during periods of inadequate ventilation or apnea when a controlled ventilation mode is chosen.

Monitoring

- Standard monitoring especially using pulse oximetry
- ETCO₂ waveform

Postoperative Management

- Ensure hemostasis is reached if bleeding occurs during extraction of FB.
- Ensure return of safe cognitive function, muscle strength, and ventilatory function before extubation.
- Consider the need to monitor oxygenation levels after PACU discharge.

Anticipated Problems/Concerns

- Fragmentation of FB
- Pneumonia and lung abscess
- Atelectasis
- Hemoptysis

Friedreich Ataxia

Mark Helfaer | Lee A. Fleisher

Risk

- Prevalence: 2:100,000; 80–90% have cardiac involvement.

Worry About

- Cardiac involvement, which does not correlate with neurologic involvement.
- Electrophysiologic disturbances.
- Cardiac dysfunction and failure.

Overview

- Progressive degeneration of posterior columns and corticospinal and posterior spinocerebellar tracts.
- Muscle weakness.

- General anesthesia can lead to postop respiratory disorders caused by thoracic kyphoscoliosis, which is associated with restrictive respiratory function.
- Abnormal glucose homeostasis.
- Most individuals have onset of symptoms of FA between the ages of 5–18 y.
- Proprioceptive sensory loss, areflexia, ataxia of limbs, Babinski sign.
- Pes cavus and scoliosis.
- Cardiomyopathy.

Etiology

- Inherited: Usually autosomal recessive but occasionally dominant
- Mutations or DNA changes in the *FXN* gene

- Frataxin (mitochondrial iron content protein) deficiency

Usual Treatment

- Usually untreatable and progressive
- Medical management of cardiac abnormalities
- Scoliosis repair
- Can be mistaken for metabolic disorders (hexosaminidase A deficiency, adrenomyeloneuropathy, vitamin E deficiency)
- Clinical trials of coenzyme Q10 (CoQ10)/vitamin E ongoing.

Assessment Points

System	Effect	Assessment by Hx	Test
CV	Left ventricular hypokinesia Concentric and asymmetric hypertrophy Cardiomyopathy	Severities of heart and neurologic manifestations are not proportional	ECG ECHO Endomyocardial biopsy
RESP	Severe scoliosis Neuromuscular impairment	Noncardiac dyspnea	Lung function
MS	Pes cavus Scoliosis Respiratory muscle weakness Unpredictable and variable response to muscle relaxants	Ability to walk without assistance	

Key References: Pancaro C, Renz D: Anesthetic management in Friedreich's ataxia, *Paediatr Anaesth* 15(5):433–434, 2005; Huercio I, Guasch E, Brogly N, Gilsanz F: Anaesthesia for orphan disease: combined spinal–epidural anaesthesia in a patient with Friedreich's ataxia, *Eur J Anaesthesiol* 31(6):340–341, 2014.

Perioperative Implications**Preoperative Preparation**

- Usual premedication

Monitoring

- Train of four to monitor effects of neuromuscular blocking agent with unpredictable response due to neuromuscular disease

Airway

- None

Preinduction/Induction

- Case report of sensitivity to curare (0.06 mg/kg caused 90 min of apnea)

- Possibility of hyperkalemia and cardiac arrhythmias after succinylcholine

Maintenance

- Case reports of successful spinal and epidural anesthesia
- Case reports of spotty lumbar epidural block
- Case reports of successful GA with cautious use of nondepolarizing agents
- Case report of successful use of hypotensive anesthesia with isoflurane
- Case report of marked decrease in cardiac output and supraventricular tachycardia with nitroprusside for hypotensive anesthesia
- Case report of successful use of epidural narcotic

Extubation

- If adequate strength from neuromuscular blocker and adequate pulm function, extubation is appropriate.

Adjuvants

- See Maintenance.