

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

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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.