Anesthetic Management of a Patient with Friedrich’s Ataxia with Associated Cardiomyopathy for Posterior Spinal Fusion

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Introduction

Friedrich’s ataxia is a rare, hereditary, neurodegenerative disorder characterized by progressive limb and gait ataxia. Its associated cardiomyopathy, impaired glucose tolerance, and neuromuscular dysfunction has implications in anesthetic management.

Case Presentation

A 20 year-old male with Friedrich’s ataxia, secondary hypertrophic cardiomyopathy and kyphoscoliosis presented for posterior spinal fusion. He was wheelchair-bound and recently diagnosed with hypertrophic cardiomyopathy, medically managed with enalapril and metoprolol. Preoperative work up included a cardiac MRI, echocardiogram, and Holter monitor analysis. Cardiac MRI revealed global hypokinesis, mildly reduced left ventricular (LV) function, and delayed gadolinium epicardial enhancement of the left lateral ventricular wall consistent with Friedrich’s ataxia. Echocardiography showed mild LV hypertrophy with mildly decreased systolic performance. Holter monitor analysis revealed multiple premature ventricular contractions (PVC’s).

Due to the increased risk of arrhythmias and history of PVC’s, beta blockade with metoprolol was continued perioperatively, and defibrillator pads were placed prior to induction. The patient was premedicated with intravenous midazolam and had an intravenous induction with propofol, lidocaine, and fentanyl. After induction, a radial arterial line was placed. To facilitate motor and sensory evoked potential monitoring, neuromuscular blockers were avoided and anesthesia was maintained with a combination of sevoflurane and infusions of propofol and fentanyl. A phenylephrine infusion was initiated due to intermittent hypotension. Total volume administered included 1.7 liters of crystalloid, 1 liter of albumin, and 540 ml of salvaged autologous blood. The case proceeded for 7 hours with an estimated liter of blood loss. The patient was awakened at the end of surgery and transferred to the pediatric intensive care unit for recovery and postoperative care.

Discussion

- Friedrich’s ataxia is the most common hereditary ataxia
- Characterized by the degeneration of the posterior spinal columns, corticospinal tract, and spinocerebellar tract.
- Clinical presentation: lower limb ataxia, dysarthria, nystagmus, skeletal muscle weakness, scoliosis, and pulmonary complications
- Cardiac disease is present in 90% of cases, with hypertrophic cardiomyopathy being the most common
- Left ventricular outflow obstruction due to mitral valve systolic anterior motion and ventricular septal contact
- Diastolic dysfunction dependent on atrial systole for ventricular filling
- Impaired coronary vasodilator reserve
- Supraventricular and ventricular tachyarrhythmias
- Adverse perioperative events: congestive heart failure, myocardial ischemia, systemic hypotension, and supraventricular or ventricular arrhythmias
- Anesthetic goals:
  - Preserve stroke volume by reducing contractility with perioperative beta blockade
  - Blunt sympathetic discharge with midazolam for premedication and use of intraoperative opioids
  - Maintain preload and hemodynamic stability with judicious use of volume
  - Augment afterload
  - Avoid tachycardia and arrhythmias while maintaining sinus rhythm with defibrillator pads
- Although Friedrich’s ataxia is rare and literature is limited, anesthetic management should focus on the pathophysiological mechanisms that may exacerbate hypertrophic cardiomyopathy.

References