[NM-291] Anesthetic Management of a Patient with Friedrich's Ataxia with Associated Hypertrophic Cardiomyopathy for Posterior Spinal Fusion

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Background: 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.[1]

Case description:
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 epicardial enhancement of the left lateral ventricular wall. Echocardiography showed mild LV hypertrophy with mildly decreased systolic performance. Holter monitor analysis revealed multiple premature ventricular contractions (PVC's). Due to increased risk of arrhythmias and history of PVCs, beta blockade with metoprolol was continued perioperatively and defibrillator pads were placed prior to induction. The patient was premedicated with intravenous (IV) midazolam and had an IV 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: Although rare, Friedrich's ataxia is the most common hereditary ataxia characterized by 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. Its features consist of left ventricular outflow obstruction due to mitral valve systolic anterior motion and ventricular septal contact, diastolic dysfunction that is dependent on atrial systole for ventricular filling, impaired coronary vasodilator reserve, and supraventricular/ventricular tachyarrhythmias. Our anesthetic goals included preserving stroke volume by reducing contractility with perioperative beta blockade, blunting sympathetic discharge with premedication and use of intraoperative opioids, maintaining preload with judicious use of volume and afterload augmentation with phenylephrine. Other goals included avoiding tachycardia and maintaining sinus rhythm.[2] Although literature is limited, anesthetic management should focus on the pathophysiologic mechanisms that may exacerbate hypertrophic cardiomyopathy.

1. Pancaro C et al. Paediatr Anaesth. 2005.
2. Poliac LC et al. Anesthesiology. 2006.
