Title: Anesthesia for Spine Surgery in a Child with Mitochondrial Disease: A Case Report

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**Background:** A child with mitochondrial disease often requires anesthesia for a large variety of surgical procedures. There is no ideal anesthetic recipe and commonly used anesthetic agents are controversial. We report the anesthetic management of a pediatric patient with mitochondrial disease undergoing posterior spinal fusion, with a discussion of the anesthetic implications of these disorders and this surgical procedure.

**Case presentation:** We describe the anesthetic management of a 10 year old boy, 25.9 kg, with a history of mitochondrial disease, respiratory transport chain deficiency secondary to pyruvate dehydrogenase deficiency. Clinical manifestations of this disease include scoliosis, greater than 70 by Cobb angle, hypotonia, static encephalopathy with developmental delay, and optic nerve atrophy. He has chronic respiratory failure which he is being ventilated during the night via tracheostomy tube and requires Bipap 14/5 during the day. In addition, he has diastolic dysfunction secondary to mild hypertrophic cardiomyopathy and a history of supraventricular tachycardia and possible WPW. The patient had posterior spinal fusion with instrumentation and allograph bone graft, from T2-pelvis. Anesthesia was induced with sevoflurane/O2 via existing tracheostomy for approximately 40 minutes until the appropriate lines were placed and patient moved to prone position. Anesthesia was maintained with an oxygen/air mixture for FIO2 of 27%, and continuous infusions of dexametomidine, sufentanil, midazolam, and cisatracurium. In addition, the patient received boluses of opioids, and midazolam for episodes of increased blood pressure. Since the patient did not take his atenolol by mouth for his history of supraventricular tachycardia, titrated intravenous doses of propanolol and esmolol were supplemented. These medications maintained the mean arterial pressure to 65mmHg. Intraoperatively, we monitored plasma lactate, hemoglobin, glucose, acid base and amount of crystalloids to decrease risk of congestive heart failure. The lowest hematocrit was 32 from 40. He was transfused early, along with continuous amicar infusion. At the end of surgery, his hematocrit was 36 and the patient did not require any blood transfusion in the post operative period. The patient’s last arterial lactate level was 28 prior to surgery. During the intraoperative period, the arterial lactate level varied between 0.7-0.9. Also, the patient received D5.2NS at a rate of 10ml/hr. Blood glucose was monitored frequently, with the highest level of 138. No hypoglycemia was noted after the postoperative period. The surgery lasted 8 hours, with an estimated blood loss of 2000ml. Fluid management consisted of 1200ml plasmalyte, 4U of packed red blood cells, 118 ml of cell saver, 262 ml of fresh frozen plasma, 269 ml of platelets, 720 ml 5% albumin. Total opioids used was fentanyl 30mcg/kg (1000mcg) and sufentanyl 14 mcg/kg (375mcg). The child was transferred, ventilated, stable to the pediatric intensive care unit. Perioperative course was uneventful and despite potential life-threatening complications, the patient made good recovery and he was discharged six days after surgery.

**Discussion:** Posterior spine fusion in this child with mitochondrial disease present with many challenging anesthetic issues. Our anesthetic management was planned to avoid metabolic stressors such as; hypotension, hypothermia, hypoglycemia and metabolic acidosis which can precipitate ketosis. Lactate free intravenous fluids, esmolol and propanolol maintained the MAP at 65mmHg, warming blankets and warm fluids kept the temperature at 37.5 celcius. Since preoperative fasting shifts metabolism towards fat utilization which these patients are limited in their ability to metabolize fat, we used a dextrose infusion, with frequent blood glucose monitoring. It was important to decrease blood loss. Therefore, we kept this child’s hematocrit close to his baseline by blood transfusions and amicar infusion. This was also helpful to prevent postoperative visual loss which prevalence is high after spine surgery. (7) In addition, this child had diastolic dysfunction secondary to cardiomyopathy, supraventricular tachycardia and possible WPW. These patients are very sensitive to peripheral vasodilations and reductions in circulatory volume. It was important to maintain a supranormal left atrial pressure for an adequate stroke volume. (1) Our CVP varied between 7-12. It was important to maintain and restore sinus rhythm. For this reason, we administered propanolol 2mg and esmolol 10-20mg IV in titrated doses. Few retrospective studies indicated rare events with general anesthesia after minor surgery such as muscle biopsy (6). Sevoflurane and propofol have been reported to have successful outcomes for only short procedures,(3) (4). Recent literature indicated no apparent relation between MH and mitochondrial myopathy. (2) There was no contraindication to opioids, midazolam, and atracurium. (5) Even though, we discussed our anesthetic plan to the family of this child, the mother was very knowledgeable about the complications associated with this surgery and anesthesia.

**Conclusion:** We successfully managed a child with mitochondrial disease undergoing a major surgery associated with significant blood loss that did not develop any perioperative metabolic, respiratory or cardiac complications.

**References:**