Perioperative Anesthesia Optimization Can Be Life Saving In Mitochondrial Disease: A Case Report

Christian Schroeder , DO; Ethan Mosley, DO; Austin McCrea, DO; Saptarshi Biswas, MD

Background

Mitochondrial diseases are exceedingly rare among adults. Typically seen in children with limited lifespans, they are becoming more prevalent in adult populations due to medical advancements. As this population ages and ultimately undergo surgery and anesthesia, there is still much unknown regarding the effects of anesthesia and mitochondrial function. This case report details a 23-year-old male with known mitochondrial disease within complex II and III of the electron transport chain, who initially presented with a small bowel obstruction ultimately requiring multiple surgeries.

Presentation

History and Physical

- 23 year old male with history of mitochondrial disease, malrotation, gastroparesis, seizure disorder presented with a 2 day history of low grade fever and abdominal distention.
- Denies nausea, vomiting, diarrhea, chills
- Review of systems otherwise negative

Family History

- No reported family history of medical disease

Past Surgical History

- G-tube and J-tube placement in early childhood
- Nissen during early childhood

Social History

- No reported social history of smoking, alcohol, or illicit drug use

Medications

- Atomoxetine, levetiracetam, gabapentin, levocarnitine, clonazepam,

Physical Exam

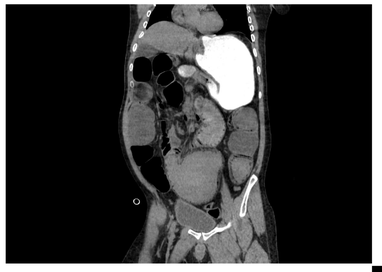
- Vitals: afebrile, HR 109, BP 104/69, sp02 98% on room air
- Exam: distended, nontender abdomen; G-tube and J-tube in position

Laboratory Data

- CBC: no leukocytosis, erythrocytosis (19.3)
- BMP: hypokalemia (3.2) lactic acidosis (9.0) anion Gap (31.0), creatinine (2.90), BUN (39)
- LFTs: total bilirubin (1.1), AST (58), ALT (58), Lipase (<50)

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

Labs and Imaging



- Figure 1 (Left): Initial CT w/contrast coronal section.
- Figure 2 (Below): Initial CT w/contrast axial section

- Impression: Multiple loops of severely dilated small bowel. Markedly dilated loop of jejunum measuring up to 12 cm in caliber. Worrisome for jejunal volvulus.

Anesthetic and Surgical Intervention

- Patient underwent exploratory laparotomy due to concern for small bowel ischemia secondary to jejunal volvulus. During the operation, patient was found to have an internal hernia. The internal hernia was reduced and an ABTera was replaced. Two days later, the patient returned to the operating room for abdominal reexploration and closure.

- Preoperative
  - Lines: LUE arterial line, 18g L hand
  - Monitoring: 5 lead ECG, R arm NIBP, EtCO2, Pulse oximetry L hand, upper warming blanket, peripheral nerve stimulator
  - Antibiotics: 1g Ancef
  - Premedication: 2mg midazolam
  - Position: Supine, left and right armboard, eyes tape

- Intraoperative
  - Induction: 100mg propofol, 100mg lidocaine, 50mcg fentanyl, 10mg rocuronium
  - Rapid sequence intubation with video laryngoscopy
  - Maintenance: 2% sevoflurane
  - Mild pressure support with intermittent pushes of neosynephrine

- Postoperative
  - Remained intubated following first procedure and transported to ICU
  - Following extubation after second procedure, patient experience respiratory arrest with sp02 in the low 70s. He was reintubated and transported to the ICU.
  - ABG acquired at that time and was found to be in respiratory acidosis

Discussion

- From an anesthesiologist's point of view, the primary complications of mitochondrial myopathies include respiratory failure, cardiac depression, conduction defects, and dysphagia[2]
- Mitochondrial dysfunction can be broken down into two categories: respiratory chain and fatty acid metabolism defects[1]
- Because of this, special considerations should be made to prevent adverse effects and postoperative complications.
- Mitochondrial patients often require smaller doses of general anesthetics, local anesthetics, analgesics and neuromuscular blockers[3]
- The most notable anesthetics that depress mitochondrial function are volatile anesthetics
- Sevoflurane has the least deleterious effects on patients with mitochondrial disorders
- Propofol has been shown to uncouple oxidative phosphorylation and inhibit complexes I, II, and IV. The strongest effect of propofol is that it inhibits the transport of long-chain acylcarnitine esters via an inhibition of acylcarnitine transferase[4]
- Optimal management: preoperative fasting minimized to 2 hours if possible, opioid-sparing techniques, avoidance of nondepolarizing neuromuscular blockers, avoid lactated ringers solution[4]

Conclusion

This case highlights the importance of understanding the effects of anesthetics in patients with altered physiology. Minimal propofol was utilized for induction to avoid propofol-infusion syndrome, a known risk in patients with mitochondrial deficiency that impairs fatty-acid oxidation. Volatile anesthetics are known to inhibit complex I; however, given our patient has a known complex II and III deficiency, sevoflurane was utilized. Patients with mitochondrial disease are prone to cardiac and respiratory depression postoperatively as they have limited physiological reserve, therefore narcotics and benzodiazepines should be administered judiciously. As demonstrated in our case, patients should be closely monitored in the perioperative period.

References

[1]Falk MJ, Sondheimer N. Mitochondrial genetic diseases. Curr Opin Pediatr. 2010;22(6):711-716

[2]Niezgoda, J., Morgan, P.G., & Brandom, B. (2013, September). Anesthetic considerations in patients with mitochondrial defects. *Pediatric Anesthesia, 23*(9), 785-793. https://doi.org/10.1111/pan.12158.

[3]Rigoulet M, Devin A, Averet N *et al*. Mechanisms of inhibition and uncoupling of respiration in isolated rat liver mitochondria by the general anesthetic 2,6-diisopropylphenol. *Eur J Biochem* 1996; **241**: 280–285

[4]Wolf A, Weir P, Segar P, Stone J, Shield J. Impaired fatty acid oxidation in propofol infusion syndrome. Lancet. 2001; 357(9256):606-607