Anesthesia for Patients with Mitochondrial Disease

Elliot Krane, M.D.

Julie Williamson, D.O.
Mitochondrial Disease

Estimated to have a lifetime prevalence of 1 in 5,000, mitochondrial disease is the most common group of neuro-metabolic diseases \(^1\). Typically, multiple organ systems are involved, and deterioration is progressive. Children with suspected mitochondrial disease often present for anesthesia for MRI, audiology testing, muscle biopsy, or central line or gastric tube placement.

On arrival to preoperative area:

Prior to induction of anesthesia an IV should be started (with the aid of topical anesthesia with a Synera patch or lidocaine cream).

Children NOT on a ketogenic diet: D5-½NS + 20KCl/L at maintenance.
Children on a ketogenic diet: ½NS + 20KCl/L at maintenance.

General Anesthesia:

Inhaled agents:
- Children with mitochondrial disease are not at increased risk of malignant hyperthermia (ref).
- Volatile anesthetics inhibit mitochondrial Complex I.
- **Sevoflurane** may be the preferred agent.
  
  - Increased sensitivity to volatile anesthetics should be anticipated, including both a reduction of MAC and also enhanced vasodilatation and myocardial depression.
  
  - Inhalation induction with sevoflurane is appropriate in those cases in which IV access could not be obtained preoperatively.
  
  - When using sevoflurane for maintenance consider using remifentanil or dexmedetomidine as a volatile sparing agent.

Intravenous hypnotics:
- Propofol inhibits mitochondrial acylcarnitine transferase and Complex III.
- Propofol as a single induction dose is probably safe **except children on ketogenic diets**. Propofol infusions, however, are not tolerated.
  
  - Dexmedetomidine: effects are unknown but no adverse mitochondrial effects reported yet. Use with extreme caution in patients with arrhythmia or heart block. Bradycardia is a known risk with dexmedetomidine and asystole can occur. **Dexmedetomidine and remifentanil** have been used as TIVA for patients with mitochondrial disease. Remifentanil infusions carry the risk of chest wall rigidity. Use with caution in an uncontrolled airway.
  
  - Ketamine is a known inhibitor of mitochondrial Complex I and uncouples Complex II.
  
  - Etomidate is a known inhibitor of mitochondrial Complex I, III, and uncouples Complex II.
  
  - Barbiturates inhibit mitochondrial Complex I.

Opioids
- **Remifentanil** has no effect on mitochondrial energetics and is preferred over fentanyl, which has mild effects, and which is preferred over morphine due to known mitochondrial effects.
- Neuraxial morphine or hydromorphone: no contraindication due to the small doses employed.

Neuromuscular blocking drugs:
- There are no mitochondrial effects.
- Succinylcholine may cause rhabdomyolysis and hyperkalemia in children who are myopathic or debilitated, as most of these children are. Generally avoided in these patients.

Maintenance of anesthesia: as for all children, maintain euthermia and hydration.
Regional Anesthesia:
• All inhibit acylcarnitine transferase and are mild inhibitors of Complex I.
• Lidocaine is preferred over ropivacaine, which is preferred over bupivacaine or etidocaine.
• Clonidine: there are no known effects on mitochondrial pathways.

Postoperative care:
In addition to standard PACU monitoring, blood glucose should be measured within 15 minutes of admission to PACU.

Indications for same day surgery discharge:
• Fully returned to baseline neurologic status.
• Glucose in acceptable range.
• Tolerating PO or G- or J-tube feedings with usual food or formula.

Indications for postoperative hospital admission:
• Surgery in any body cavity.
• Inability to return to full p.o. intake.
• Decrement in neurologic function compared with baseline status.

References:


© 2013 by Elliot Krane and Julie Williamson. This may not be reproduced in whole or part without permission from the author.