

# **Guidelines for Anesthesia in Children with Metabolic and Mitochondrial Disorders**

rev May 19, 2017

## **CASE SCHEDULING**

Elective cases should be planned well in advance by the attending service to allow adequate preparation by all involved specialties.

Cases ideally are scheduled for first case of the day to allow minimum fasting periods and maximum postoperative observation.

The primary service scheduler should specify under the Comments section of the scheduling request form "Mitochondrial Disease" or "Metabolic Disease" to flag the case for the anesthesia scheduler.

The primary service scheduler should also specify on the scheduling form the name and pager number of the geneticist or metabolic specialist who will be on-call and available on the day of anesthesia for urgent consultations.

## **PREOPERATIVE PREPARATION**

Indications for preoperative overnight hospitalization:

- Surgery anticipated to enter any body cavity.
- Bowel preparation (e.g. for colonoscopy) or need for prolonged NPO time.
- Unreliable social home environment.

Fasting should be minimized and limited to no more than 6hr for solids and milk/formula, 4hr for breast-milk, and 2hr for clear liquids. Parents should be instructed to awaken their child during the night to allow oral hydration if there is not a gastric or jejunal tube. If there is a G- or J-tube, continuous feeds with clear glucose containing liquids should continue until 2hr prior to the induction of anesthesia (see exception for children on ketogenic diets).

Children on ketogenic diets: should not receive glucose or sucrose containing liquids during the preoperative period. They should continue on their ketogenic diet until 6hr prior to anesthesia, then be offered water until 2hr prior to the induction of anesthesia.

Preoperative sedation: oral or IV midazolam in the usual doses is safe and appropriate when indicated.

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.

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Children on a ketogenic diet: ½NS + 20KCl/L at maintenance.

## GENERAL ANESTHESIA

Inhaled agents:

- All 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.
- In general, the anesthetic should be converted to a TIVA at the earliest appropriate time after induction of anesthesia.

### Intravenous hypnotics:

- Propofol inhibits mitochondrial acylcarnitine transferase and Complex III.
- Propofol as a single induction dose is probably safe except children on ketogenic diets. However, propofol infusions are not tolerated.
- Dexmedetomidine: effects are unknown but no adverse effects reported yet.
- 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:

- Remifentanyl 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.

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## **Neuromuscular blocking drugs:**

- There are no mitochondrial effects.
- However succinylcholine may cause rhabdomyolysis and hyperkalemia in children who are myopathic or debilitated, as most of these children are.

**Maintenance of anesthesia:** as for all children, maintain euthermia and hydration.

## **REGIONAL ANESTHESIA**

### **Local Anesthetics and adjuvants:**

- All local anesthetics 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 15min 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.