

## A Quality Improvement Initiative

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### Introduction

- Mitochondrial disorders (MD) comprise a diverse set of diseases and result from dysfunction of the mitochondria, the energy producers of the cell.
- Often, patients with MD require anesthesia for a variety of procedures, such as muscle biopsy, MRI, lumbar puncture, and gastrostomy tube placement.
- These patients have unique anesthetic considerations that must be taken into account when formulating an anesthetic plan.
- Currently at our institution, there is no anesthetic protocol for patients with MD.

### Purpose

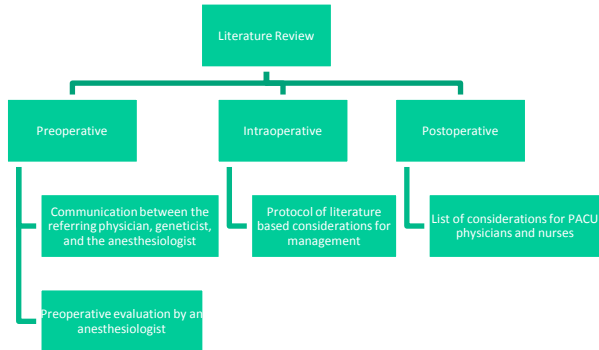
- To review the current process for caring for patients with mitochondrial disorders in the perioperative period
- To develop a protocol to standardize care

### Goals

- To reduce risk for anesthetic complications
- To ensure that our practice adheres to the most current literature

### Initiative

- Develop a literature-based, standardized method of anesthetic care for patients with mitochondrial disorders
- Impact all phases of the perioperative period



**•Preoperative Considerations**

- Metabolic:** increased risk of fasting intolerance/ development of lactic acidosis
  - Obtain IV access
  - Begin glucose containing solution at maintenance rate preoperatively
  - Avoid Lactate Ringers
  - Minimize fasting time by scheduling as first case
  - Check preoperative glucose if concerned
  - Know patient's baseline lactate level
- Cardiac:** conduction abnormalities and cardiomyopathy
  - ECG
  - Echocardiogram if indicated clinically
- Respiratory:** insufficiency
- Renal/Hepatic:** insufficiency; check labs
- Endocrine:** diabetes
- Neurologic:** seizure disorder, stroke, ataxia
- Musculoskeletal:** weakness, easy fatigue
- GI:** reflux and delayed gastric emptying

**•Intraoperative Considerations**

- Consider metabolic stressors that can lead to decompensation
  - Hyper- or hypoglycemia: monitor intraoperative glucose
  - Hypotension: monitor volume status, maintain normotension
  - Hyper- or hypothermia: maintain normothermia
  - PONV: prophylaxis
- Malignant hyperthermia (MH) precautions not necessary for mitochondrial disorders
  - If cause of myotonia unknown, MH precautions may be necessary
  - Discuss with geneticist and primary care physician
- Rapid sequence induction/intubation may be necessary depending on GI involvement
- Possible induction agents
  - Ketamine, etomidate, opioids
  - Propofol: use with caution, avoid if possible
    - Can be used safely as induction bolus
- Maintenance
  - Inhalational maintenance acceptable
    - Avoid nitrous oxide which may exacerbate complex I inhibition
    - Sensitivity may be increased; consider use of BIS monitor
  - Drugs to consider for IV maintenance
    - Ketamine, remifentanyl, dexmedetomidine
    - Propofol infusion relatively contraindicated
- Postoperative Considerations**
  - Standard monitoring
  - Monitor closely for arrhythmias and respiratory insufficiency
  - Low threshold for overnight observation if any sign of metabolic decompensation or if not tolerating PO
  - Consider multi-organ system involvement as evaluated preoperatively
  - Check postoperative glucose
  - Continue glucose containing solutions until tolerating PO
  - Maintain normothermia
  - Treat PONV

Figure 1. Excerpt from protocol.

### Protocol

- This protocol is available to all of our perioperative care providers.
- Additional information is given to parents regarding their child's anesthetic.

### Discussion

- By developing an evidence-based anesthetic protocol for patients with MD, our institution can more confidently provide safe, effective care for this patient population.
- This protocol ensures that our anesthetic providers are following the best practices identified in literature.
- Additionally, we now have a more effective system to identify critical issues prior to the day of surgery.
- Through the enactment of this system, we can:
  - Improve communications between the anesthesiologist and the primary care physicians.
  - Ease the concerns of nervous parents as their children undergo anesthesia.
  - Provide patients with the highest current standard of care.

### References

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