

# UT Mitochondrial Center of Excellence



Mary Kay Koenig, MD, Director  
Brunessia Lewis, LMSW Social Worker

LaKeesha Minor, RN, BSN Clinic Coordinator  
Rahmat Adejumo, Research Coordinator

Phone: 713-500-7164 | Fax: 713-500-0719 | Email: [ut.mito@uth.tmc.edu](mailto:ut.mito@uth.tmc.edu) | Website: [www.utmito.org](http://www.utmito.org)

## ANESTHESIA PROTOCOL FOR PATIENTS WITH MITOCHONDRIAL DISEASE

Clinical manifestations of mitochondrial disease, including seizures, arrhythmias, cardiac dysfunction, myopathy, and endocrinopathies, can be worsened by fasting, illness, or surgical stress. To date, no controlled trials of anesthetic agents or techniques have been conducted in patients with mitochondrial disease.

Adverse effects on mitochondrial function of many agents used in anesthesia have been documented in vitro, but there are few reports of adverse events in vivo. Even propofol, for which adverse effects have been reported both in vitro and in vivo, has been used successfully in isolated cases. Thus, the theoretical effects of any agent need to be considered in the general context of any one patient's medical history. It is important to remember that **the absence of published reports of adverse effects with an agent does not mean that the agent is safe.** It more likely reflects a publication bias.

**This being said, the following anesthesia protocol should be respected when caring for these patients:**

1. **Keep NPO status to a minimum.** Mitochondrial patients should take clear fluids until two hours prior to anesthesia or should be admitted for IV hydration the night before. Fasting before surgery limits ATP production and patients with mitochondrial disorders may be unable to compensate.
2. **Ringer lactate should not be used** as it may precipitate or exacerbate lactic acidosis.
3. **Propofol should not be used.** Propofol uncouples oxidative phosphorylation, inhibits electron flow along electron transport chain, antagonizes beta receptor binding, and acts directly on Ca channel proteins to diminish contractility.
4. **Ketamine, Versed, Precedex** (Dexmedetomidine) can be used as appropriate.
5. Secondary to the risk of complex I inhibition seen with in vitro studies of sevoflurane, **Precedex is the agent of choice** in children with mitochondrial disease. **Inhalation agents**, including **Sevoflurane**, can be used at the discretion of the anesthesiologist. The attending anesthesiologist should use his or her professional judgment to weigh the risks of metabolic decompensation against the risk of using alternate anesthetic agents.
6. Although the risk of malignant hyperthermia is questionable, **Succinylcholine should not be used.**
7. MD patients have a **higher sensitivity to non-depolarizing muscle relaxants**, if needed they should be titrated carefully. Because of possible underlying myopathy, twitches should be monitored and muscle relaxants should be reversed before extubation.
8. **Perioperative monitoring** of pH, glucose, lactate, pyruvate, and electrolytes is indicated.
9. **Normothermia** should be maintained.

Mary Kay Koenig, MD  
UT Mitochondrial Clinic Director

Maria Matuszczak, MD  
Pediatric Anesthesia Division Chair