Ochsner
Children's Health Center

Anesthesia Guidelines for Patients with Mitochondrial Disorders

Mitochondrial disease (MD) are a heterogeneous group of disorders that results in patients having abonormal cellular energy metabolism. Organs with high energy consumption (e.g., brain, muscle, heart and liver), are especially at risk for complications in the perioperative period. Most of the current information regarding anesthesia and mitochondrial disorders come from in vitro trials or case reports. Currently, it is not clear whether any particular anesthetic technique is superior in patients with MD. When caring for a Mitochondrial Disorder patient under anesthesia, consider the following:

Stress in these patients can have clinical significance:

Avoid hypothermia.

Keep NPO to a minimum – try to avoid anaerobic metabolism which limits ATP production and of which lactic acid as a byproduct. Allow clears up to 2hrs prior to anesthesia or consider admission the night prior for IV hydration.

Avoid Lactated Ringers as it may precipitate acidosis. Remember that tissue oxidative capacity governs the rate of lactate removal. Use a **dextrose containing fluid** to allow these patients to better compensate during the fasting period. Set an Alaris pump to half of patient's maintenance fluid rate and run throughout the case. Use D5 ¼ NS in patients less than 1 year of age and D5 ½ NS in older patients. Discuss this with staff in case an alternate fluid is needed.

Halogenated agents can be used with caution. Some in vitro studies recommend discretion in the use of sevoflurane in patients with complex I mitochondrial disease.

Precedex is the agent of choice for these patients. It should be the mainstay anesthetic in these cases.

Do not use Propofol – Like barbiturates, Propofol uncouples oxidative phosphorylation and thus, normal mitochondrial function.

Benzodiazepines and Ketamine can be use as needed.
Literature is unclear about **Etomidate** and MD – use it with caution and only if needed.

Non depolarizing neuromuscular blocking agents should be titrated carefully as these patients are sensitive to their effects.
There is no direct relation between mitochondrial disorders and malignant hyperthermia, nevertheless avoid the use of **Succinylcholine** as with any other pediatric patient, except in emergency airway situations.

Assess pH, blood glucose, urine ketones, lactate, pyruvate and electrolytes as needed.

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