PROTOCOL FOR PROCEDURES OR SURGERY REQUIRING SEDATION OR ANESTHESIA

(Date)

Re: (NAME)
D.O.B.

(NAME) is a patient with mitochondrial disease with symptoms that include: _____.

Patients with mitochondrial disease can tolerate surgery and anesthesia safely. However, precautions to reduce further the risk from the procedure include:

1. Elective procedures should be postponed if the patient develops any signs of infectious illness around the time of the procedure date;
2. Minimize the time necessary for fasting. The patient should be encouraged to take some fluids (orally or enterally) just before becoming NPO;
3. An intravenous line should be placed pre-operatively and fluids provided until the patient is eating/drinking well, or able to tolerate fluids through a g-tube if present. If the procedure is delayed, IV fluids can keep the patient stable;
4. IV fluids should contain dextrose and electrolytes; do not administer Ringer's Lactate since patients with mitochondrial disease may have disturbed lactate metabolism;
5. For patients with a history of fasting intolerance and/or documented hypoglycemia, or if there is secondary disturbance in fatty acid oxidation, IV fluids should contain 10% dextrose with electrolytes to run at 1.25x maintenance or higher. The higher glucose solution is necessary to minimize catabolism and flux through an impaired fatty acid oxidation pathway. 10% dextrose is more effective than 5% dextrose in accomplishing this goal;
6. If the patient takes any vitamins as part of his/her mitochondrial management, these can be provided once PO/enteral fluids are tolerated;
7. If the patient has a problem with vomiting post-operatively, s/he should be admitted and continued on intravenous fluids until able to tolerate fluids/food.

If there are any questions regarding the safety of induction agents or other anesthetic medications, please consult the following references:

A. ***Anesthesia and Mitochondrial Cytopathies,*** by Cohen, Shoffner, and DeBoer. It can be downloaded from the United Mitochondrial Disease Foundation web-site: http://www.umdf.org/library/pdfarticles.aspx.

B.

Please call with any questions.