

PROTOCOL FOR SURGERY REQUIRING SEDATION OR ANESTHESIA WHEN EATING OR THE GUT IS DISRUPTED

(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, procedures that impact the gut (at any level) can potentially destabilize these patients.

Mitochondrial disease is often associated with some degree of abnormal gut motility associated with uncoordinated movement or dysmotility; regions of the gut can be affected to different degrees. This can result in problems that can include any of the following: swallowing incoordination, gagging or choking, gastroesophageal reflux, vomiting, delayed gastric emptying, bloating, abdominal pain, constipation and/or incomplete evacuation. Infectious illnesses, surgical manipulation/disruption of the GI tract, and anesthesia can reduce motility further, usually transiently.

In the post-operative period, oral intake of food and liquid diminishes and these patients are at risk for becoming dehydrated. This can occur for several reasons – pain at the surgical site (e.g., as with tonsillectomy), fatigue from the surgical procedure and/or anesthesia, or worsening gut motility with increased gastro-esophageal reflux and/or delayed gastric emptying. Refusal to eat or drink and poor calorie intake can exacerbate fatigue and result in a prolonged recovery and occasionally readmission.

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;
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. IV fluids should be continued until the patient is able to tolerate adequate volumes of fluids/food PO/enterally without vomiting;
7. If this patient demonstrates a refusal to eat or drink after 24 hours post-surgery, consideration should be given to providing 1-2 days of parenteral nutrition which can improve calorie intake, reduce fatigue and related symptoms, and shorten the stay in hospital;
8. If the patient takes any vitamins as part of his/her mitochondrial management, these can be provided once PO/enteral fluids are tolerated;
9. For patients with respiratory insufficiency due to trunk muscle weakness, the use of a cough assist to expand their lung capacity has been found to be beneficial.

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.