March 2, 2017

To whom it may concern:

has a mitochondrial disease due to EARS2 mutation resulting in defect with mitochondrial transcription/protein synthesis. Her clinical presentation is characterized by CNS lesions (thalamus, brainstem, white matter), lactic acidosis, and recurrent infections.She is at risk of metabolic decompensation due to impaired energy production at the time of illness / stress or catabolism.

The following are recommendations for care in the peri-operative period.

Fasting Allowance and Fluid Support:

- **Fasting up to 4hrs is allowed without IV fluid support.**
- if fasting longer than 4hrs is needed, start D10 ½ NS @ 40 ml/hr to minimize metabolic stress. This will provide 1x maintenance IV fluid.
- **Lactated Ringer's solution is contraindicated.**

Anesthesia:

- **Avoid depolarizing neuromuscular junction blockers** (such as succinylcholine) and Propofol whenever possible as these agents have been reported to increase stress to mitochondria.

During or immediately post-anesthesia

- Please check a blood gas (capillary or venous OK), CBC, CMP, and lactic acid pre and post procedure to monitor adequacy of supportive care peri-operatively.

After Anesthesia:

- Resume home meds and diet when cleared by the anesthesiologist
- When the patient is able to tolerate PO (liquids and food), the IVF (D10) can be hep-locked.
- If laboratory results are at baseline, patient can be discharged.

Please call the Metabolic Division on call with any questions at the numbers below.
I thank you in advance for your kind attention.