

Sample Emergency/Illness Precautions Letter

To Whom It May Concern:

X has a disorder of mitochondrial metabolism. Individuals with such a metabolic disease are more sensitive to routine illness and simple fasting than others and can have a rapid decompensation. These individuals need an urgent evaluation since they can transition quickly from well-appearing to fatally ill. Some individuals can progress to a Reye-syndrome like illness and permanent liver failure.

If X is unable to maintain oral intake with illness, he should be evaluated by a physician, with a low threshold to admit him for a short hospital stay for intravenous fluids with dextrose.

The admission should not occur exclusively for dehydration - but for any metabolic stressor that may lead to dehydration or catabolism. The goal is to admit the patient and treat them him/her prior to any dehydration or catabolism occurring.

The treatment for acute metabolic decompensation in these disorders includes:

1. Hydration with dextrose containing IV fluids. D10 should be used with insulin piggy-backed to control hyperglycemia. Insulin is a potent anabolic hormone, promoting protein and lipid synthesis. Give fluids at 1.25-1.5X times the maintenance rate. IV fluids should never contain lactated ringers.
2. Correct any biochemical abnormalities. Routine chemistries, CBC, liver function (synthetic and cellular), ammonia, glucose, ketosis and lactic acidosis should be monitored and any derangements corrected.

Hypoglycemia - if hypoglycemic, administer 1-2 g/kg of glucose IV STAT; follow with (at least) a 10% glucose solution

Metabolic acidosis - administer NaHCO_3 as a bolus (1 mEq/kg) if acutely acidotic with pH < 7.22 or bicarb level < 14, followed by a continuous infusion.

Hyperammonemia - the elevated ammonia reflects a secondary inhibition of the urea cycle. As treatment for the metabolic decompensation proceeds, the ammonia level should diminish. A level > 200 may require treatment.

3. Eliminate toxic metabolites by making the patient NPO for 24-72 hours and by giving levo-carnitine via an IV, at a dose of at least 100 mg/kg/day divided tid. If the patient is on a higher oral dose, please use that dose for the IV treatment. Any other supplements being given should be continued by mouth if possible. Once the patient's laboratories begin to normalize, restarting the patient on their home-based diet is advised.

4. Treat any underlying infection and fever.

Medications that should generally be avoided in individuals with mitochondrial disease include valproic acid, statins, aminoglycoside antibiotics, erythromycin, and propofol. There are no absolute contraindications and these medications can be given if an alternative drug is not available or appropriate.

Some individuals with mitochondrial diseases are more sensitive to volatile anesthetics and need a much lower dose to achieve a bispectral index of <60. Sevoflurane is tolerated better than isoflurane and halothane.

Should there be any questions or concerns please contact my office at X