

To whom it may concern:

PATIENT:

DATE OF BIRTH:

DIAGNOSIS:

(name) has a disorder of mitochondrial metabolism. The clinical manifestations in this patient include (EXAMPLES – write in primary symptoms): seizures, migraine, strokes, stroke-like events, myopathy, neuropathy, movement disorder, hearing loss, visual loss, cardiomyopathy, cardiac conduction defect, gastroparesis resulting in intestinal pseudo-obstruction, fasting intolerance, clinical deterioration during viral infections and dehydration, etc.)

The diagnosis of mitochondrial disease is based on the following clinical and laboratory findings: (write method of diagnosis). Individuals with such a metabolic disease have a risk of deterioration, sometimes permanent, due to the common effects of a physiologic stress such as a viral or bacterial infection, such as fever, anorexia or dehydration. There is no evidence-based medicine review to support these recommendations, but they are based on a consensus opinion of several authorities in the field.

Because of the patient's diagnosis, I have instructed the patient to carry this letter on their person and present this to any physician or emergency department that will be performing a medical evaluation. In addition to the standard evaluation and treatment, I do recommend that strong consideration be made for intravenous fluids if there is any history or anorexia, vomiting or diarrhea. Oral rehydration therapy does not seem to be as effective as it is in otherwise healthy patients. Although there is no substitute for the standard history and examination, special attention should be paid for evaluation of electrolytes, glucose, liver enzymes, CK, CBC, urine analysis (including pH, presence of ketones, and specific gravity). If blood lactate and ammonia are readily available, these can be obtained as well. In general, I recommend a bolus of normal saline if there is evidence of dehydration, followed by D5 (or D10) with $\frac{1}{4}$ to $\frac{1}{2}$ NS with KCl (if appropriate) to run in at about 1.5 times maintenance. If levocarnitine is available I would also recommend placing 1000 mg in each liter bag of

intravenous fluids, or bolus with 1000 mg if the need for IV fluids will be brief. In some circumstances, patients feel much better after the IV fluid bolus and a few hours of IV therapy, but in other situations, IV fluids should be continued for 1-3 days (an observation admission is generally acceptable). In this particular patient, the history of prior response suggests that fluids will likely be necessary for (your doctors recommendations) hours.

Any identified medical condition should be treated as you would any other patient. Some patients with mitochondrial disease are susceptible to the ototoxic effects of systemic aminoglycosides, so these are best avoided unless there is no other adequate antibiotic coverage. There is no absolute contraindication to aminoglycosides.

If there is a need for emergency surgery we do suggest avoiding Lactated Ringers solution unless the clinical situation suggests otherwise. During anesthesia and in the post-operative period, blood glucose and lactate levels should be monitored. Some individuals with mitochondrial disease are more sensitive to volatile anesthetics, and in some circumstances only 10% of the typical dose of a volatile anesthetic is necessary to obtain adequate anesthesia. If possible BIS monitoring (bispectral index) should be done during anesthesia as a much lower dose achieves a BIS<60. Short courses of propofol has been safely used in patients with this diagnosis (1-2 hours) but prolonged anesthesia should be avoided. Malignant hyperthermia is rare in patients with this diagnosis but there is a concern for rhabdomyolysis. For elective and non-urgent procedures, the patient should be at baseline health status without any dehydration or infection prior to the procedure. Serious attention with respiratory therapy measures (such as incentive spirometers) should be used in the post-operative period.

If there are any questions, please call me. My office number is (your doctors telephone number) and after hours my service can be reached at (your doctors telephone number). I would appreciate if you could give the patient a copy of all lab results and a discharge summary.

Sincerely

(your doctors signature)