# VLCAD Emergency Protocol Example

_____ carries the diagnosis of Very long-chain acyl-CoA dehydrogenase (VLCAD) deficiency, a genetic disorder of mitochondrial fatty acid beta-oxidation. Because of this enzymatic deficiency, she is unable to metabolize fatty acids efficiently and has a severely reduced ability to fast. She has the additional clinical problem of mild gross and fine motor disability, but no active or past history of hepatic or cardiac disease, which occur in some patients with VLCAD deficiency. _____ also has multiple food allergies, for which she requires a special hypoallergenic diet.

At times of illness-induced fasting or after more prolonged periods of reduced caloric intake, she may require hospitalization for intravenous glucose therapy to terminate fasting and limit metabolic intoxication. Following are the principles of her management.

## Nutrition and Complications of Fasting

Because of her limited ability to metabolize long chain fatty acids, she maintains a diet with fat reduced to approximately 20% of calories with a supplement of 15-30 cc/d (150- 300 kcal) of MCT oil. Because her metabolic intoxication is induced by fasting, she should not be allowed to fast for more than 12-15 hours, the period after which blood free fatty acid levels begin to rise rapidly and beta-oxidation is activated.

The early signs of metabolic decompensation are lethargy, irritability, and vomiting. Any of these signs requires the prompt administration of at least maintenance calories orally or intravenously.

**Her parents are the best judges of when she is beginning to deteriorate metabolically.** Classical signs of a metabolic disease, such as metabolic acidosis, hyperammonemia, or even hypoglycemia may not be present when _____ is seriously ill. An increased blood level of uric acid can be an early sign of metabolic decompensation, but is not completely reliable. Therefore, clinical signs and symptoms and her parents should be relied upon as the major criteria for hospitalization. However under no circumstances should _____'s blood glucose concentration be allowed to fall below 75 mg/dl without starting intravenous therapy.

## Treatment of Metabolic Intoxication

For mild illnesses, _____ should receive the equivalent 2 ounces of juice (or other high-carbohydrate liquid) every hour. Pedialyte and its equivalents are NOT adequate sources of calories for this purpose. If _____ is hospitalized for a more severe illness, 10% glucose at 1.5 times fluid maintenance (1.5 X 70 = 105 cc/hr for 20 kg) should be administered with appropriate amounts of sodium

chloride and, if required, potassium chloride. IV infiltrations or other IV interruptions must be remedied promptly.

Even one hour without IV glucose may lead to serious problems when she is acutely ill. If _____ is recovering but still in a marginal nutritional state, do not wait until morning to restart an IV that infiltrates at night. Unnecessary deterioration and neurological injury may otherwise occur. In addition, the need to meet _____'s full caloric requirements must take precedence over any need to restrict fluid intake. If fluid overload is a concern, diuretics should be used in lieu of reducing the IV rate, or the percentage of glucose in the fluids should be increased. Neither should the IV rate be lowered because of mild to moderate hyperglycemia, because elevations of blood glucose may occur as a physiological response to relative intracellular hypoglycemia, a common phenomenon in metabolic diseases.

If there is a need to control hyperglycemia, insulin at 0.01 to 0.02 units/kg/hr should be started and adjusted to maintain blood glucose between 100 and 150 mg/dl. In addition, if _____ is seriously ill, insulin should be started promptly to block lipolysis and thereby reverse metabolic intoxication more rapidly. Dopamine and related pressors should be used cautiously because of their lipolytic effect. If she receives intravenous fluids for more than 8 hours, serum phosphorus should be monitored carefully and corrected intravenously as needed. In addition, if _____ must remain NPO for more than 24 hours, intravenous amino acids @ 1 g/kg/d should be given. Because of its effect to raise plasma free fatty acids levels, agitation should be treated with sedatives.

Studies to be obtained on admission to assess _____'s metabolic state should include blood for ammonia, chemistry panel, amylase, lipase, electrolytes, CPK, and uric acid and urine for organic acid gas chromatography. The most sensitive routine laboratory marker of _____'s metabolic state is the serum uric acid level, which increases directly with metabolic intoxication. Plasma should also be obtained on admission for measurement of free fatty acid levels by gas chromatography (Dr_____).

As _____'s appetite returns, the IV glucose infusion can be tapered in the morning hours to heparin-lock and restarted in the afternoon or evening if her caloric or fluid intake fails to exceed maintenance levels. Recovery of appetite may be slow. Follow the recommendations of _____'s parents regarding the best fluids and foods to offer when she resumes oral intake. After initial stabilization in the hospital and during the IV weaning process, frequent glucose monitoring is not necessary as long as _____ is known to be receiving an appropriate number of calories (IV or PO).

**Fever**

Because of the additional metabolic stress caused by fevers, temperatures in excess of 38°C should be treated aggressively with 10-15 mg/kg of acetaminophen per dose. If there is breakthrough fever, reduce the time interval between doses, increase the dose of acetaminophen, or add ibuprofen. Aspirin and other salicylates should not be given because of their potential to cause hepatotoxicity or Reye syndrome. In addition, orders for antipyretics should not be written as "PRN fever." Instead, antipyretics should be continued for at least 24 hours before withholding them in the morning and checking for persistence of fever.

**Medications:** Pantothenic acid – 25 mg TID

**Allergies:** Sulfa drugs and multiple food allergies. Consult with parents for specific restrictions.

Carnitine - causes rhabdomyolysis and cardiac arrhythmias

**IN CASE OF ILLNESS OR EMERGENCY, CONTACT ONE OF THE FOLLOWING PHYSICIANS FOR CONSULTATION OR ADDITIONAL MEDICAL INFORMATION:** (specialists' names and phone numbers)

**(Submitted by Dawn, VLCAD Parent. **Please remember that this is just an EXAMPLE of an Emergency Protocol.** Discuss your/your child's specific situation/condition with the Drs to individualize the Protocol.)