

Example of VLCAD Protocol

To Whom It May Concern:

Re: (Name of child)

Date of Birth:

_____ is a ___ year old boy with **very long chain acyl-CoA dehydrogenase (VLCAD) deficiency**, an inborn error of mitochondrial fatty acid oxidation. He was discharged from the hospital on (date) in stable condition.

Currently his problems include the following:

- Cardiomyopathy, which has resolved
- Skeletal muscle weakness, which is improving markedly. He is currently meeting his developmental milestones
- Dietary therapy with Portagen, a formula rich in medium-chain triglycerides; diet restricted in long chain fatty acids. Feeding is *per os* with supplementation via gastrostomy

Because of his inability to utilize long chain fatty acids for energy production, prolonged fasting (more than 10 to 12 hours), intercurrent illness, and excessive physical activity make him very vulnerable to acute episodes of metabolic compensation, hypoglycemia, coma, liver dysfunction, congestive heart failure, and rhabdomyolysis. The latter may lead to macroscopic myoglobinuria and acute renal failure.

We have instructed his parents to bring him to the nearest Emergency Room whenever he cannot or does not eat regularly or at any time if symptoms like persistent muscle pain and weakness, fever, irritability, lethargy, and vomiting are present.

In the Emergency Room, prompt administration of IV fluids is most important [10% dextrose at a rate of 10 mg of glucose/kg/minute for the first 6 hours, followed by a rate of infusion equivalent to 1 ½ maintenance over 24 hours], to be started immediately, even before laboratory results become available.

Of course, this regimen is recommended only if the associated myoglobinuria did not cause acute renal failure with oliguria/anuria. If he has symptoms of cardiomyopathy, then he may need diuretic therapy and additional pharmacologic treatment as needed.

Samples should be sent **STAT** for blood glucose, creatinine phosphokinase, LFT's, creatinine and BUN, and urine myoglobin.

If you have any further questions in this matter, please call (your Dr's Emergency number). In addition, the (Dr's office) can be reached during the day at (phone number). There is always a physician on call and available by pager (pager number).

Mr. and Mrs. _____ are very knowledgeable about their son's condition and can provide accurate information. Thank you for your consideration.

Signed: Dr's name

Address, phone

Any other pertinent names/numbers

[This protocol example was submitted by Gina R., VLCAD Parent. Please be sure to consult with your child's physician to plan an individualized emergency protocol.]