LCHAD EMERGENCY CARE PROTOCOL

NOTE OF CAUTION: Please check with your healthcare professionals regarding this protocol sheet.

To Whom It May Concern:

_____ has **long chain 3-hydroxy acyl-Coenzyme A-dehydrogenase (LCHAD) deficiency, a disorder in the beta oxidation of fatty acids**. She presented at 2 days of age with a cardiorespiratory arrest and as a result also has a seizure disorder. Because of this enzymatic deficiency, _____ is unable to metabolize certain fatty acids efficiently and has a reduced ability to fast. Her disease is routinely managed by limiting her intake of long chain fatty acids, providing her with frequent feedings/meals, and supplementing her with medium chain fats (MCT) for energy. If _____ becomes acutely ill, she may develop vomiting, hypotonia, lethargy, metabolic acidosis, hypoglycemia, myoglobinuria, and severe myalgia. If untreated she could progress to cardiomyopathy, hepatomegaly, kidney failure, coma, and/or cardiorespiratory arrest as a result of her altered metabolism. At times of illness-induced fasting, or after more prolonged periods of increased activity and reduced caloric intake, _____ may require hospitalization for intravenous glucose therapy to end the fasting state and limit metabolic intoxication. If she presents with an acute illness and any of the above symptoms, the following management plan should be provided to suppress lipolysis and resume metabolic control. **Genetics service should be immediately notified of her presence in the ER.**

1) IV therapy with balanced glucose and electrolyte solution should be started promptly at 1-1/2 to twice maintenance for correction of hypoglycemia and acidosis. Blood glucose concentration should be kept above 80 mg/dl. Use a 10% dextrose IV solution. IV infiltrations or other IV interruptions must be remedied promptly. Even one hour without IV dextrose may result in neurologic sequelae. Metabolic acidosis may require treatment with boluses of sodium bicarbonate at 1 - 2 mEq/kg IV.

2) The need to supply caloric requirements must take precedence over fluid balance. Diuretics should be used, as long as she is not anuric, in lieu of reducing the IV rate if fluid overload is a concern. The IV rate should not be lowered because of mild to moderate hyperglycemia. 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 0.02 to 0.05 units/kg/hr should be started and adjusted to maintain blood glucose between 100 and 150 mg/dl.

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3) **Do not administer IV lipids. Intralipid is contraindicated.**

4) Labs should be sent for glucose, electrolytes, blood ammonia, CBC, BUN, creatinine, uric acid, LFTs, CPK, blood gas for pH, and urine for myoglobin. Heparinized plasma for carnitine (1 ml) and urine organic acids (15 - 20cc) should be collected, frozen, and sent STAT on dry ice to a laboratory.

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5) A chest x-ray and Doppler echocardiography should be considered since cardiomyopathy is associated with this condition.

6) Treatment for any triggering infection should be started as appropriate.

7) Because of potentially severe metabolic stress caused by fever, temperatures in excess of 37.8^{o} C should be treated aggressively with acetaminophen, and if necessary, ibuprofen. Use a cooling blanket if necessary.

8) Carnitine should be supplemented at her usual dose orally if tolerated. Do not give excess carnitine without consulting with the Metabolic/Genetics physician.

9) When enteral feeds are started they should be ordered every 3 - 4 hours. Her diet limits the use of long chain fats to 15 - 20% of total calories and recommends that MCT comprise 15 - 20 % of the calories. Daily calorie counts should be conducted to closely monitor her intake.

10) _____ should be continued on hydration therapy (1 ½ times maintenance) until her CPK level begins to decrease significantly, creatinine normalizes, and gross hematuria has resolved. She should also be clinically improved without significant myalgia. Please consult with genetics or nephrology service prior to hospital discharge.