Date:

RE:

DOB:

To Whom It May Concern:

This child has a diagnosis of long chain 3-hydroxy acyl-CoA dehydrogenase (LCHAD)/trifunctional protein (TFP) deficiency. This is a defect of fatty acid oxidation characterized by liver damage, hypoglycemia, and the possibility of sudden death (cardiac arrest) at time of fasting/fever due to cardiac arrhythmia. These episodes are triggered by infections, fasting, fever or vomiting, and exercise.

The disease is treated with frequent feedings to prevent fasting; a diet low in fat in which most are medium chain fatty acids (that can be normally metabolized by patients with this disease); cornstarch supplements (if needed); and low-level carnitine supplementation (25 mg/kg/day).

The parents have been instructed to bring the child to the Emergency Room if unable to eat or if the child has high fever/vomiting. LCHAD deficiency can cause sudden cardiac arrest and sudden death. This child needs to be seen by the ER physician as soon as possible.

In case of acute attack the child should receive the following immediately:

* D10 one half Normal Saline 20 mEq/L KCl at 1.5 maintenance
* D-sticks or blood glucose should be monitored immediately and as the therapy progresses.

The following additional labs should also be obtained:

* Basic metabolic panel, Liver function tests, CK, Plasma ammonia, Plasma acylcarnitine profile, and Urine analysis.

The child should not be discharged home until able to eat adequately by mouth.

Please contact our office at 801 585 2457 or the geneticist on call at 801-581-2121 (or 801-662-1000 for the Primary Children’s Hospital operator) for further help.