LCHAD Emergency Care Protocol

Date:

Regarding:

Date of birth:

To Whom It May Concern:

INDIVUIDUAL’S NAME 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. Episodes are triggered by infections, fasting, fever, 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 the child is unable to eat, has high fever, or is 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 PHONE NUMBER or the geneticist on call at PHONE NUMBER for further help.