EMERGENCY METABOLIC LETTER

RE: [PATIENT NAME]
DOB:

[PATIENT NAME] is followed by the Division of Medical Genetics and Metabolism for a possible diagnosis of very long chain acyl-CoA dehydrogenase (VLCAD) deficiency based upon results from newborn screening. This is a defect of fatty acid oxidation characterized by severe hypoglycemia and the possibility of sudden death (cardiac arrest) at time of fasting due to cardiac arrhythmia.

These episodes are triggered by infections, fasting, fever or vomiting, and exercise. The disease is treated with the following: frequent feedings to prevent fasting and the need for fat oxidation; a diet that contains medium chain fatty acids (that can be metabolized by patients with this disease); cornstarch supplements (if needed); and with 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.

VLCAD 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 illness [PATIENT NAME] should receive the following immediately:
- D10 one-half normal saline with 20mEq/L KCL at 1.5 maintenance, DO NOT DELAY INITIATING IV FLUIDS EVEN IF THE CHILD APPEARS WELL, please use D5 one-half NS until D10 one-half NS is available.

The following labs should also be obtained:
- Basic metabolic panel, Liver function tests, CK, Plasma ammonia, and Urine analysis

For further management and guidance for this rare condition please contact the geneticist on call at (xxx) XXX-XXXX and ask for the geneticist on call.

Thank you in advance,

Division of Medical Genetics
(xxx) xxx-xxxx