

## **Illness Protocol Example For Long-Chain Hydroxy-Acyl-CoA Dehydrogenase (LCHAD) Deficiency**

### **Clinical Presentation:**

Rhabdomyolysis, Anorexia, Vomiting, Lethargy, +/- Hypoglycemia, +/- Cardiomyopathy, +/- Retinopathy

### **Routine Treatment:**

**Diet:** Portagen for infants or MCT oil supplements for older children and adults. High Carbohydrate, restricted fat 25-30% of total calories. Avoid Fasting longer than 6-8 hours ~ eating often throughout the day is important for any child/adult with an FOD

**Medication:** Carnitor® (100 mg/ml) solution give 50 mg/kg/day, divided in 3-4 doses

### **Minor Acute Illness without Fever (Ear infections, URI, etc...):**

**Diet:** As above plus, increase fluid intake - Gatorade, Powerade, Pedialyte, Popsicles, Jello, Apple juice

**Medication:** Carnitor® (100 mg/ml) solution give 50 mg/kg/day, divided in 3-4 doses

### **Acute Febrile Illness associated with significant anorexia, +/- vomiting, diarrhea:**

**Bloodwork:** CBC with differential  
Complete Metabolic profile  
CPK  
Dry bloodspot card for Acylcarnitine profile  
Urine OA

### **Treatment:**

**Medications:** Carnitor® (100 mg/ml) solution give 100 mg/kg/day, divided in 3-4 doses. If not tolerated switch to IV Carnitor®

**Loading dose: IV Carnitor®** (1000 mg/5 ml), 50 mg/kg slowly over 2-5 minutes, followed by 50 mg/kg over each subsequent 24 hour period, until the patient is able to resume their normal PO diet

**IV D10 1/4 NS @ 1 ¼ - 1 ½** maintenance to maintain blood sugar ~100 mg/dl

**\*\* Intralipids are CONTRAINDICATED in patients with Fatty Acid Oxidation Disorders**

### **Acute Rhabdomyolysis:**

If CPK  $\geq$  25,000 and rising, recommend ICU admission for combined IV Glucose and Insulin drip to stop further lipolysis. Monitor blood CPK and Glucose levels.

**\*\*\* [www.fodsupport.org](http://www.fodsupport.org) has more information on LCHAD and other FODs**