

## ACUTE ILLNESS PROTOCOLS

### 3-HYDROXYMETHYLGLUTARYL-CoA (3-HMG CoA) LYASE DEFICIENCY

#### *THERAPY*

#### TREATMENT

**1. INDICATION FOR IV (NEVER less than 10% dextrose infusion)**  
(One or more indication is sufficient for IV)

- Vomiting
- Hypoglycemia
- Poor PO intake
- Dehydration Do not rely on ketones as indicating dehydration!
- Decreased alertness
- Metabolic acidosis

Start 10% glucose continuous infusion at 1.5x maintenance to provide 7-8mg/kg/min

#### **2. DO NOT ADMINISTER LIPIDS IN ANY FORM**

#### **3. HYPOGLYCEMIA**

Push 25% dextrose 2ml/kg and follow with a continuous 10% dextrose infusion at 1.5x maintenance to provide 7-8mg/kg/min glucose

#### **4. METABOLIC ACIDOSIS (Bicarbonate level<16)**

Must be treated aggressively with IV Sodium bicarbonate (1 mEq/kg). Treating conservatively in the expectation of a re-equilibration of acid/base balance as other biochemical/clinical parameters are normalized can lead to tragic consequences.

#### **5. CARNITINE**

Should be provided PO (100-200 mg/kg/day divided TID) or IV (30-50 mg/kg/day).

#### **6. PRECIPITATING FACTORS**

Should be treated aggressively to help minimize further catabolism

#### **7. APPARENTLY WELL**

If drinking oral fluids well and none of the above factors present, there is no need for emergent IVI. But history of earlier vomiting, pyrexia, or other stressor should be taken seriously and a period of observation

undertaken to ensure that PO fluids are taken frequently and well tolerated, with glucose status monitored periodically.

**In conjunction with this protocol, please call or have paged the genetics metabolism fellow on call, or failing this, the metabolic attending on call at your hospital or nearest pediatric tertiary care center**