

**ACUTE ILLNESS PROTOCOL
FATTY ACID OXIDATION DISORDERS
LONG CHAIN HYDROXY Acyl-CoA DEHYDROGENASE DEFICIENCY
(LCHADD)**

TREATMENT

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

- Vomiting
- Hypoglycemia
- lactic acidosis
- Poor PO intake
- Dehydration do not rely on urinary ketones as indicating dehydration!
- Decreased alertness
- Metabolic Acidosis
- Cardiac decompensation

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

2. HYPOGLYCEMIA

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

3. METABOLIC ACIDOSIS (Bicarbonate level <16mEq/L)

Must be treated aggressively with IV sodium bicarbonate (1mEq/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.

4. PRECIPITATING FACTORS

Should be treated aggressively to help minimize further catabolism

5. CARDIAC CONSIDERATIONS

A cardiology assessment is necessary to properly evaluate a child with acute symptomatic LCHADD (specifically for heart failure or pericardial effusion).

5. 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.

POST EMERGENCY MANAGEMENT

1. Child unable to take/maintain PO intake

- Start, or continue, 10% glucose continuous infusion at 1.5x maintenance.
- Blood glucose and acid/base status should be monitored regularly. If the child is physically stressed keep the blood sugar levels elevated (glucose levels should be kept between 120-170 mg/dl)

2. Carnitine

The use of carnitine in FAODs is controversial and there are concerns that excessive long chain acylcarnitines which may be produced may induce arrhythmias. Consult with the metabolic physician for guidance regarding this in each individual case.

3. **DO NOT ADMINISTER LIPIDS IN ANY FORM**

4. Avoidance of fasting when stop IVI

this may include complex carbohydrate in the form of cornstarch supplementation to get through the night as the child gets older; and a high carbohydrate/low fat diet.

LCHADD chronic management is complicated as many children take a significant amount of time (days to weeks) to improve clinically even once their biochemical parameters have normalized. Particular problems include improvement in mental status, hypotonia, hepatomegaly and cardiomyopathy. It is important to be aware that despite therapy children with LCHADD have died or been left with chronic neurologic, cardiac and hepatic problems. Though the long-term prognosis for children with LCHADD is unclear treatment can be optimized by:-

- avoidance of fasting (this may include complex carbohydrate in the form of cornstarch supplementation to get through the night as the child gets older)
- high carbohydrate/low fat intake
- Early detection of physiologic stresses inc. infection, surgery with especial attention to REGULAR feedings/source of glucose AROUND the clock.
- Regular review by cardiology and ophthalmologic services.

Note that the pregnant mother carrying a fetus with LCHADD is at risk for the HELLP syndrome (hemolysis, elevated liver enzymes and low platelets). She should be closely followed up for counseling and antenatal care for future pregnancies as there will be a 25% risk of each future pregnancy having an affected LCHADD fetus.

Any questions about the patient or 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.

Additional information may be obtained via OMIM at <http://www.ncbi.nlm.nih.gov/htbin-post/Omim/dispim?600890#TEXT>

