

Illness Protocol Example For Medium-Chain Acyl-CoA Dehydrogenase (MCAD) Deficiency For Infants and Young Children

Clinical Presentation:

Anorexia, Vomiting, Lethargy, +/- Hypoglycemia, Reye-like syndrome

Routine Treatment:

Diet: High Carbohydrate, Fat-restriction 25-30% of total calories, avoid fasting longer than 6-8 hours ~ eating often throughout the day, and a snack before bed, is important for any child/adult with an FOD. 1-1½ grams/kg of raw Cornstarch at bedtime may be considered for some infants (Note: Cornstarch cannot be metabolized until the pancreatic Amylase enzyme becomes active ~ usually in infants older than 6 months) and young children who experience symptomatic hypoglycemia (low blood sugars). It should be mixed in cool, sugar-free liquids to ensure proper absorption. Cornstarch is most useful in younger patients because they lack Glycogen stores and hormonal control of blood sugar that are intact in the older children and adults. Be aware that too much cornstarch may cause constipation.

Medication: Carnitor® (100 mg/ml) solution give 50 mg/kg/day, divided in 3-4 doses. (Also available in 330 mg tablets)

[**Please note: adolescents and adults are dosed differently than infants** – their total Carnitor® intake should not be greater than 2 grams total per day. Please refer to www.carnitor.com or www.sigmatou.com for further information or call 1-800-447-0169]

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

Diet: As above plus, increase fluid intake – i.e. Gatorade, Powerade, Pedialyte, Popsicles, Jello, Apple juice. 1-1½ grams/kg of raw Cornstarch at bedtime may be considered for infants and young children who experience symptomatic hypoglycemia (low blood sugars). It should be mixed in cool, sugar-free liquids to ensure proper absorption.

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
Carnitine Levels
Dry bloodspot card for Acylcarnitine profile
Urine OA & Acylglycines

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

*** www.fodsupport.org has more information on MCAD and other FODs