

**Treatment and Emergency Protocol Example For
Medium-Chain Acyl-CoA Dehydrogenase Deficiency (MCAD)
For Teens and Adults**

Diagnosis: Medium-Chain Acyl-CoA Dehydrogenase Deficiency (MCAD)

Clinical Presentation: Vomiting, lethargy, +/- hypoglycemia, Reye-like syndrome, carnitine deficiency

• **Routine Daily Treatment:**

Diet: High carbohydrate with Fat restriction to @ 25-30% of total Kcal - **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)

Medication: L-carnitine tablets (**Carnitor®**) **1 tablet 3-4 times per day**

Supplied: 330 mg tablets (or 100mg/ml solution for infants/children)

Carnitine total per day (for an adult) should not exceed 2 grams

(Read about Carnitor® at www.sigmaptau.com or www.carnitor.com or call 1-800-447-0169)

Avoid all Steroid medications (causes metabolic complications)

•• **Minor Acute Illness without Fever:** (i.e. ear infections, URI, etc.)

Diet: As above plus increase fluid intake – i.e. gatorade, pop, juice, jello, etc

Medication: Maintain Carnitor® dose at 1 tablet 3-4 x day

••• **Acute Febrile Illness / EMERGENCY:** (i.e. vomiting, lethargy, anorexia, diarrhea, surgery)

[Please note that **one cannot totally rely on blood sugar alone**, as an indicator of illness. **Some children/adults may present with 'normal' blood sugar but other levels (i.e. ammonia, liver enzymes, etc) may be abnormal. Changes in behavior should also be taken into consideration. Additionally, avoid ALL STEROID medications, which can cause metabolic complications.**]

Bloodwork: CBC with differential, Complete Metabolic Profile, Carnitine Levels

Treatment:

Medication: Continue 1 Carnitor® tablet 3-4 x day, if not tolerated switch to IV Carnitor®

Loading Dose: **IV Carnitor® (1000 mg/ml) solution** ~ 40 mg/kg slowly over 2-5 minutes, followed by equivalent for each subsequent 24 hour period, until patient is able to resume his/her normal PO diet

IV D10 ¼ NS @ 1¼ - 1½ maintenance to maintain blood sugar ~ 100 mg/dl

[Start IV D10 IMMEDIATELY after drawing blood for lab work. DO NOT WAIT for lab results.]

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

[This Example of an MCAD Protocol was originally written by Dr Charles Roe at the Institute of Metabolic Disease for Kevin Gould (Deb and Dan's son) and has been adjusted over the years as Kevin has grown into teen/adulthood. Please be aware that this may not be appropriate for your specific situation (i.e. different FOD, different age ~ infant versus adult, other medications, supplements, or medical complications involved etc., so work with your Drs and other specialists on an individualized Protocol for yourself or your child. You can also include emergency numbers/contacts, insurance information, other daily medications, as well as a detailed clinical description of what MCAD is and possibly what others can look for as far as 'symptoms' are concerned when your child/self does not feel well. Our website's Medical Information page has a clinical description of MCAD for Families and Professionals ~ www.fodsupport.org]