

Sample Care Plans and Protocols

Wisconsin Regional Centers – toolkit

<http://wimedicalhometoolkit.aap.org/careplans/index.cfm>

AAP (American Academy of Pediatrics) Care Plans, Toolkit

<http://www.medicalhomeinfo.org/tools/assess.html>

Illinois Medical Home Model for Physicians

<http://internet.dccc.uic.edu/medhome/mdprimer/MHPhysicianPrimer.asp#coordinated>

Center for Medical Home Improvement

<http://www.medicalhomeinfo.org/tools/CarePlans/ComprehensiveCarePlanningII.pdf>

Indiana CDM Program

<http://www.indianacdmprogram.com/Collaborative/PDF/Expectations%20of%20Care%20in%20Chronic%20Care%20Model.pdf>

National Academy for State Health Policy

http://www.partnershipforsolutions.org/DMS/files/Care_coordination.pdf

Emergency Management Protocol
Medium-Chain Acyl-Coenzyme A Dehydrogenase (MCAD) Deficiency

Patient Name:

DOB:

Disorder: MCAD Deficiency is an inborn error of metabolism affecting the degradation of medium-chain fatty acids by mitochondrial β -oxidation. Since energy production via β -oxidation of fatty acids is impaired, once glycogen stores become depleted, hypoglycemia will develop. Infections or any other catabolic condition (fasting) may lead to hypoketotic hypoglycemia, metabolic acidosis, coma or death. Immediate medical intervention is imperative for intercurrent illness and subsequent catabolic state.

Rationale for Rx: Treatment consists primarily in avoiding fasting and catabolism by frequent feedings relatively high in carbohydrates.

Acute Symptoms: Even for mild common infections, evaluation by a physician is needed.

- Increase sugar-containing clear liquids (pedialyte, apple juice, popsicles, powerade, Gatorade).
- Treat bacterial infections promptly with antibiotics (OM, etc).
- Continue oral carnitine at 50-100mg/kg/day.
- In case of vomiting, diarrhea, or decreased oral intake, admit for IV fluids containing glucose. Monitor glucose levels. Do not delay IV placement and starting IV fluids with glucose. **Hypoglycemia can develop suddenly and can be severe.**

Emergency Room Protocol*:

- 1. Please contact the on-call metabolic specialist by dialing 313-745-0203, followed by pager # 96025 and then enter your call back number including area code.**
- 2. IV Treatment:**
 - An IV should be started immediately with D10 with age appropriate electrolytes at 1.5 X maintenance even if the initial blood glucometer check shows normal results.
 - Do **not** wait for results of laboratory evaluation before starting IV fluids with glucose.
 - Monitor blood glucose levels.
- 3. Labs:**
 - Glucose level and electrolytes (Na, K, Cl, HCO₃)
 - Liver function tests (AST, ALT, Total and direct bilirubin)
 - CBC+diff (add blood cultures if febrile)
 - Plasma total and free carnitine levels
 - Urinalysis
- 4. Continue oral Carnitine. If not tolerated, give IV carnitine 100mg/kg/dose every 6 hours.**

***Medical therapy for underlying intercurrent illness should be instituted concurrent to the treatment of the inborn error of metabolism.**

Metabolic Clinic
313-745-4513

Maple Syrup Urine Disease Emergency Care Sick Plan

Prepared by: Children's Hospital of Michigan Metabolic Clinic (CHMMC)
Division of Genetic and Metabolic Disorders

Date of Plan: 1/25/2008

Patient Name:

DOB:

CHM Medical Record Number:

PARENT:

- Home monitoring includes the measurement of α -ketoacids in the urine using DNPH. Cloudiness of the urine (1:1 mixture of urine:DNPH) indicates the presence of α -ketoacids. Bring your DNPH kit with you to the emergency room.
- Bring your MSUD formula, supplemental valine and isoleucine and newborn screening cards for dried blood spot collection with you to the emergency room.
- If you are going to a hospital, bring your prepared MSUD formula with you to the emergency room.

EMERGENCY ROOM PHYSICIAN:

- The purpose of this MSUD Emergency Care Sick Plan is to provide you with a treatment plan for acute management to help prevent metabolic decompensation. Left untreated, the child can go into a coma and die. Precipitating factors which cause symptoms to develop include any common illness, such as fever, gastroenteritis, viral or bacterial infections or starvation. Symptoms of excessive leucine in the blood (Leucinosi) include CNS depression (irritability, poor feeding, vomiting/diarrhea, lethargy, seizures, ataxia, altered mental status, slurred speech) and cerebral edema (hallucinations, unusual breathing, coma, increased intracranial pressure).

BACKGROUND INFORMATION:

- Maple Syrup Urine Disease (MSUD) is an inherited disorder in the body's ability to use three of the *essential* amino acids in protein. These three amino acids – leucine, isoleucine and valine – are often called the *branched-chain amino acids* (BCAAs). The disorder is caused by deficient activity of the enzyme (called branched chain α -ketoacid dehydrogenase [BCKDH]) necessary to break down these 3 amino acids and their byproducts (called *ketoacids*). The result of this deficient enzyme activity leads to elevations of these amino acids and their byproducts. It is these elevations which cause an infant or child with MSUD to become ill. This disorder is diagnosed at birth through the Michigan Newborn Screening Program and children with MSUD are placed on a special diet which restricts their daily intake of isoleucine, leucine and valine. The management of this diet is through the Children's Hospital of Michigan Metabolic Clinic.

EMERGENCY ROOM PROTOCOL:

- Emergency Room Labs:
 - CBC with differential
 - Electrolytes, glucose, bicarb
 - Plasma ammonia
 - Liver function tests
 - Plasma amino acids or dried blood spot (DBS) sent overnight to the following address:
ATTN: Matt Sprague or Dr. Bob Grier
DMC University Laboratories, UHC Bldg, 3C-6
Detroit, MI 48201-2153
 - Urine DNPB to check for α -ketoacids. Cloudiness of the urine (1:1 mixture of urine:DNPB in a clear test tube) indicates the presence of α -ketoacids and is evidence of metabolic decompensation.
- Begin IV fluids: D10/0.5NS @ 1½ maintenance. **DO NOT FLUID BOLUS, AS CHILD IS AT RISK FOR CEREBRAL EDEMA. DO NOT FLUID OVERLOAD!!** Decrease fluid rate to maintenance after 3 hours or earlier if recommended by the on-call metabolic physician.
- The metabolic staff at CHMMC includes the following physicians: Dr. Gerald Feldman (program director), Dr. Ayesha Ahmad (clinic director) and Dr. Era Bawle (Division Director). The on-call physician can be reached by any of the following means:
 - Detroit Medical Center Page System (metabolic on-call physician): **313-745-0203, pager 96025. Follow the recorded instructions.**
 - Daytime (8:30AM – 5PM Monday-Friday) clinic phone number: 313-745-4513. The clinic is closed weekends and holidays but the answering machine will refer you to the page operator.
 - If unable to reach the on-call metabolic physician, page the program director, Dr. Gerald Feldman, at 800-820-5435.
- Transferring patient to Children's Hospital of Michigan is recommended if hospitalization is required.
- The main treatment to correct the metabolic imbalance is to provide adequate protein intake (while restricting the branched chain amino acids) to decrease catabolism. This is accomplished by providing the daily amount of special MSUD formula, using an NG tube if necessary, over 24 hours. The goal is to create an anabolic state through adequate caloric/fluid intake.
- Isoleucine/Leucine supplementation: this is necessary, as isoleucine and valine can drop precipitously low as the leucine levels drop. Supplementation with home supplies is usually 100-150 mg of isoleucine and valine each day. The home supply is 10mg/ml, so provide 10-15 cc of each.
- Watch for evidence of fluid overload and hypernatremia. Maintain normal Na concentration.
- Treat any evidence of infection with appropriate antibiotics

IF ADMISSION TO HOSPITAL:

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 - Page (metabolic on-call physician): 313-745-0203, pager 96025

- Daytime (8:30AM – 5PM Monday-Friday) clinic phone number: 313-745-4513. The clinic is closed weekends and holidays but the answering machine will direct you to the page operator.
- If unable to reach the on-call metabolic physician, page the program director, Dr. Gerald Feldman, at 800-820-5435.
- After consultation with CHMMC, determine whether child is to be started on formula or special TPN. If BCAA-free TPN is to be used, it must be ordered specially, in consultation with CHMMC. **DO NOT ORDER TPN WITHOUT CONSULTATION WITH CHMMC. Specific instructions will be provided as needed at that time.**
- **Watch fluid intake carefully.** Preventing catabolism without fluid overload is the main goal. As the leucine level decreases, you will see an improvement of mental status. **DO NOT FLUID BOLUS, AS CHILD IS AT RISK FOR CEREBRAL EDEMA. DO NOT FLUID OVERLOAD.**
- Continue IV fluids as needed. If tolerating NG or PO feeds, decrease IV rate to KVO. Intralipids can be added for caloric intake.
- Continue supplemental isoleucine and valine at 40-80 mg/kg/day added to the formula. This will be supplied by the parents (10mg/ml).
- Daily weights, electrolytes, urine DNPH (Urine DNPH to check for α -ketoacids. Cloudiness of the urine (1:1 mixture of urine:DNPH in a clear test tube) indicates the presence of α -ketoacids and is evidence of metabolic decompensation.
- Plasma amino acids daily, to be sent to the DMC-UL address listed above
- Change continuous NG feeds to bolus feeds after 24 hours. If tolerating formula, change to PO intake and discontinue NG feeds.

10/19/2004