Children's National Medical Center

Emergency Department Pathway

Metabolic Pathway

Revised 8-9-04

INCLUSION CRITERIA

All patients with Urea Cycle Disorders (OTC, CPS, ASA, Citrullinemia), Maple Syrup Urine Disease (MSUD), Proprionic Acidemia (PA), Methylmalonic Acidemia (MMA), Isovalerice Aciduria, Glutaric aciduria, MCAD, SCAD, LCHAD, VLCAD, CPTII/CACT, or Possible Mitochondiral Disorders EXCEPT for Pyruvatedehydrogenase Deficiency (PDHD) presenting to the Emergency Department for care.

TRIAGE RN

1. **Expedite Triage process and facilitate placement in ED room**
2. **Notify Charge Nurse and Attending Physician of patient arrival, identify team member to contact Metabolism Attending Physician on call and notify of patient's arrival**

NURSE/TECH

1. **Place patient on C-R monitor, ask for patient's emergency letter, & initiate Metabolic Flowsheet**
2. **Draw blood for the following when inserting saline lock** *(large bore if possible)*
   - POCT **PCx Glucose** and ISTAT venous **blood gas**
   - **BMP**
   - **Ammonia** (green top on ice) if urea cycle disorder or unknown metabolic disorder *Ammonia specimen must be free-flowing and cannot be hemolyzed. Draw through as large bore IV as possible. If unable to obtain free-flowing specimen via IV placement, contact attending physician to perform arterial puncture for specimen.
   - **CBC** (If possible. Do not send if adequate blood is not available for other studies)
   - Draw and hold green top if adequate blood sample available for other studies
3. **Mark lab specimens "METABOLIC SUPERSTAT"** and notify Specimen Receiving *(ext. 5352 & 5354)*
4. **Obtain urine specimen for POCT dipstick & urinalysis.** If adequate sample, hold remaining urine for other study
5. Maintain PO status per neurologic status
6. **Document reassessments at the following intervals** *(technician may assist with vital signs)*
   - Q 15-30 minutes during 1st hour: HR, RR, B/P, and level of consciousness
   - Advance to Q 1 hr unless unstable: HR, RR, B/P, and level of consciousness
   - Q 4 hrs: Temperature

PHYSICIAN

1. Evaluate patient and review I-Stat labs
2. **Order maintenance fluid and dextrose resuscitation** on metabolic flowsheet – **10% Dextrose/0.33% Sodium Chloride with 10mEq Potassium Chloride/500 mL at 1 ½ X maintenance rate**
3. If hypotension, order Normal Saline bolus(es) (20mL/kg) per usual treatment of hypovolemia/shock
4. Contact metabolism attending physician through hospital operator
5. Order metabolic medications.

RN

1. Review management plan with physician. Administer replacement fluids and medications per order (see Metabolic Flowsheet)
2. Report decreasing level of consciousness or tachycardia/hypotension to Staff Physician immediately
3. Notify pharmacy of need for liter bag of maintenance IV fluid if patient >40kg
4. Repeat labs per orders on metabolic flowsheet

PHYSICIAN

1. Document frequent reassessments
2. Review lab results and vital signs
3. Consult with Metabolism Attending Physician regarding patient disposition:

Consider discharge (after discussing with metabolism attending MD) if:

- Normal mental status
- Baseline feeding habits
- No acidosis
- No urine ketones
- Ammonia 65 or less

Consult PICU if:

- Suspected cerebral edema
- Altered LOC
- Poorly responsive to therapy
- Persistent shock