Emergency Department Guideline
Metabolic Emergency

Objective: To evaluate and rapidly begin treatment for children presenting to the ED with Inborn Errors of Metabolism (IBEM).

Background:
1. Minnesota has an extensive newborn screening program, to identify at-risk infants for therapy and close clinical monitoring.
2. Many IBEM patients present to the ED with a known diagnosis and treatment plan, and have a readily available Emergency Information Form (EIF) on the MEMSCIS (www.memscis.org) network, including subspecialist contact numbers, medications, allergies, treatments, and other clinical details. Use it!

Indication:
1. The undiagnosed IBEM patient may clinically present with:
   a. Recurrent episodes of illness, or may be surprisingly sick from an apparently minor illness
   b. Family History of infant or childhood death, especially siblings. (Most conditions are autosomal-recessive so detailed FH may not be informative, BUT consanguinity?)
   c. History of developmental delay
   d. History of a significant neurologic event
   e. History of abnormal feeding pattern/cyclic vomiting
   f. Uncommon (but suspicious) historical findings
      i. Strong history of food aversion
      ii. Distinctive unusual odor
         Sweaty feet
         Maple syrup
      iii. Maternal history of liver disease in pregnancy (HELLP, AFLP)
2. Remember not all happen in babies - milder forms of most IBEM occur!
3. Common clinical presentations may include:
   a. Hypoglycemia
   b. Respiratory distress (tachypnea)
   c. Acidosis
   d. Vomiting
   e. Hepatic dysfunction, with bleeding, hepatomegaly, or jaundice

This UMHC! Guideline addresses only key points of care for the specific population; it is not intended to be a comprehensive practice guideline. These recommendations result from review of literature and practices current at the time of their formulation. This Guideline does not preclude using care modalities proven efficacious in studies published subsequent to the current revision of this document. This document is not intended to impose standards of care presenting selective variances from the recommendations to meet the specific and unique requirements of individual patients. Adherence to this Statement is voluntary. The clinician in light of the individual circumstances presented by the patient must make the ultimate judgments regarding the priority of any specific procedure or course of action.
f. Heart failure

g. Muscle dysfunction, with pain, rhabdomyolysis, or weakness

h. Seizures

i. Signs and symptoms of infection or sepsis

j. Altered sensorium/mental status

**Evaluation/Intervention:**

1. Check vital signs, oral or tympanic temperature, and pulse oximetry.
2. For temperature >38.5, give acetaminophen 15mg/kg PO/PR, or ibuprofen 10mg/kg PO, if none previously given, or if >4 hours since last dose.
3. Assign nurse and room for patient, notify Attending ED physician.
4. IV access
   a. Initial lab studies:
      i. *Get an i-Stat 7 for the unstable or seriously ill patient!*
      ii. CBC, Chem-20, VBG or CBG, UA, Coagulation panel
      iii. Ammonia, lactate/pyruvate, CK
   b. Follow-up labs:
      i. Uric acid
      ii. Plasma Amino Acids
      iii. Urine Organic Acids
      iv. Plasma acylcarnitine profile
      v. Plasma carnitine
      vi. Urine acylglycine

5. Treatment:
   a. Address ABC’s!
   b. Dextrose and IV fluid infusion
      i. For weight <15 kg, start IV infusion of D10-0.2NS at 1.5x maintenance rate
      ii. For weight ≥15 kg, start IV infusion of D10-0.45NS at 1.5x maintenance rate
      iii. (Use D5-0.2NS or D5-0.45NS at 2x maintenance rate if D10 solution is not ready from pharmacy, and switch to D10 solution at 1.5x maintenance when it becomes available)
      iv. Continue to monitor hourly serum glucose levels
         May require an insulin drip for hyperglycemia (*Do not reduce the Dextrose infusion rate!*) starting at 0.05 units/kg/hour IV
      v. Goal is serum glucose of 100-150 mg/dl
   c. Rare IBEM diagnoses may worsen with Dextrose infusion!
      i. Pyruvate dehydrogenase complex deficiency
      ii. A primary lactic acidosis
      iii. X-linked disorder (but both M-F affected)
      iv. The infusion of glucose will cause:
         Acute lactic acidosis
         Profound weakness
      v. *This will be quickly evident!!!*
         Treatment: Change the Dextrose infusion to an IV NS infusion with Intralipid 2 g/kg IV
Documentation:
1. Triage vital signs, weight, oximetry.
2. Document time of each intervention, and repeat vital signs.