Emergency Management Protocol for C3 – Propionic and Methylmalonic Acidemia
Newborn Screening Program of the Oklahoma State Department of Health

Evaluation & Initial Management Guidelines for High Risk Propionic Acidemia and Methylmalonic Acidemia Screen Results.

1. Contact the family within **one hour** of notification. Inform family of newborn screening results and assess clinical status (poor feeding, vomiting, lethargy, tachypnea).
2. **Immediate consult with a** geneticist– pager number listed below.
3. History and Physical Exam by **on same day of notification** either in the pediatrician's office or at the local Emergency department, in consultation with a geneticist:
   - Assess specifically for signs and symptoms of Metabolic Crises:
     - Metabolic Ketoacidosis
     - Hyperammonemia
     - Poor Feeding/ loss of appetite
     - Hypoglycemia
     - Lethargy
     - Vomiting /Dehydration
     - Failure to thrive
     - Hypotonia
     - Coma
4. If patient lives in metro OKC or Tulsa initial assessment should be done with the geneticist, if available.
5. **Immediate** phone consultation with a geneticist regarding treatment and clinical management is required.
6. In coordination with the geneticist, consider **immediate transportation** to metabolic center for diagnostic work-up and medical management.

Description
These disorders are caused by the body's inability to process certain proteins and fats properly. Abnormal levels of organic acids in the blood, urine and tissue can be toxic and lead to serious health problems.

Resources
- **ACMG Newborn Screening ACT Sheets:** https://www.ncbi.nlm.nih.gov/books/NBK55827/
- **Integris Pediatric Specialty Clinic, Inborn Error of Metabolism (IEM) Clinic**
  Geneticist pager: (405) 630-3794
- **OU Children's Physicians – Genetics Clinic**
  Page Operator: (405) 271-3636
- **Newborn Screening Follow-Up Program**
  (405) 271-6617 option 2 or (800) 766-2223; www.nsp.health.ok.gov