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 metabolic specialist** - pager and phone 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 metabolic specialist:
   - 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 Metabolic Specialist if available.
5. **Immediate** phone consultation with a metabolic specialist regarding treatment and clinical management is required.
6. In coordination with the metabolic specialist, 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
Metabolic Nurse Specialist - Julie Boyd, RN
Phone (405) 271-8001, ext. 42074

Newborn Screening Follow-up Program (405) 271-6617 or 1-800-766-2223
www.nsp.health.ok.gov

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<thead>
<tr>
<th>Metabolic Specialists</th>
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<tbody>
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<td>Klaas Wierenga, MD</td>
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