



DEPARTMENT OF THE ARMY
U.S. ARMY MEDICAL DEPARTMENT ACTIVITY
7500 COCHRANE CIRCLE
FORT CARSON, CO 80913-4604

REPLY TO
ATTENTION OF

MCXE-PEDS

MEMORANDUM THRU Chief, Pediatrics, USA MEDDAC, Ft Carson, CO 80913

REGARDING:

To: Whom it may concern

1. _____ presented on _____ with _____ .
(S)He is being evaluated for a possible inborn error of metabolism. While we are not yet certain whether an underlying metabolic disorder is present, we strongly suggest that (s)he be treated as if an inborn error is present. When fasting (that is, when using body stores for fuel) individuals with inborn errors partially metabolize substrates to toxic compounds which cause neurologic abnormalities and secondary biochemical abnormalities. Therefore, there is risk of metabolic decompensation during intercurrent illness. Persistent vomiting and irritability or lethargy are usually the first symptoms of inborn errors and, if untreated, may progress to coma and possible death. Hypoglycemia, acidosis, and liver dysfunction including hyperammonemia are variable.

2. Should _____ show any of the above symptoms, or should there be any persistent decreased oral intake and/or vomiting for any reason, and if home management with sugar and fluid has not reversed the symptoms, we suggest immediate evaluation and then immediate IV glucose. Management includes the following:

- Immediate placement of an IV with glucose (at least 6-8 mg/kg/min), and appropriate fluids.
- Baseline blood glucose and electrolyte levels, and subsequent monitoring.
- Care of the precipitating event (intercurrent illness, vomiting, diarrhea, etc.)
- If there is an altered mental status, liver function and ammonia should also be measured and followed.

For diagnostic purposes, since diagnosis is still not established in this patient, we request that blood for amino acids and urine for amino acids and organic acids and (LFT's, acyl carnitine, ammonia, spare tubes, etc) be obtained while he is acutely ill.

3. _____ may present with symptoms of acute metabolic decompensation while glucose and hydration status are still normal. At these times, it is important to immediately administer IV glucose and fluids to prevent the catabolism that results from intercurrent illness. By the time glucose is low or there is evidence of dehydration, there may already be irreversible damage.

4. We would also recommend that our office be contacted prior to any scheduled surgery so that diagnosis and management issues can be reviewed.
5. In the event of acute problems, care providers should always attempt to contact me or the on-call pediatrician for Evans Army Community Hospital (pager number can be obtained from the Emergency Department, (719) 526-7964).
6. Should care providers have any questions during normal office hours, we can be contacted at Evans Army Pediatric Clinic, (719) 526-7653. You can also contact the Metabolic Program during normal office hours or through the metabolic pediatrician on-call. They can be reached by calling (303) 861-8888 or One-Call at The Children's Hospital, 1-800-332-2082. We discussed with them at the time of this letter.

cc:family