

ACUTE ILLNESS PROTOCOL ORGANIC ACID DISORDERS

ISOVALERIC ACIDEMIA

DIAGNOSIS

ASSESSMENT

- STAT chemstrip to check for hypoglycemia
- vital signs, cardiovascular stability
- hydration status
- presence of fever; signs of infection
- hepatomegaly
- neurologic status; evidence of increased intracranial pressure

LABS

Blood

- (arterial) blood gas
- electrolytes, measured CO₂, glucose
- ammonia (in ice STAT to lab)
- AST, ALT, AlkPO₄, PT, PTT
- plasma amino acids
- serum carnitine
- CBC, differential WBC count, platelets

Urine

- urinalysis for specific gravity and ketones
- urine for organic acids
- as needed, cultures of blood, urine and throat

NOTE, organic acids and ammonia are toxic to the brain and accumulations of these may result in cerebral edema. Caution should be exercised when considering the need for a lumbar puncture.