

**ACUTE ILLNESS PROTOCOL
UREA CYCLE DISORDERS
THE INFANT/CHILD WITH ARGININOSUCCINATE LYASE DEFICIENCY
(also known as argininosuccinic acidemia)**

ASSESSMENT

Assess for cardiorespiratory instability, dehydration, fever, infection or any other physical stressor (e.g. surgery), as a potential precipitant for metabolic decompensation. Assess hepatic and neurological status.

- **Blood glucose**
- **Electrolytes, CO₂ and blood gas**
- **Ammonia** (1.5 ml blood in sodium-heparin tube sent STAT to lab on ice)
- **Plasma amino acids**
- **LFTs** (AST,ALT,AlkPO₄, bilirubin)

Plasma ammonia is a direct index of toxicity, important for acute management. A level greater than 250 µg/dl (150 µmol/L), typically with the absence of metabolic acidosis (though may occur secondary to a primary respiratory alkalosis).

Plasma amino acids should be drawn first thing in the morning , calling the metabolic lab in advance for urgent samples. Glutamine acts as an ammonia buffer and reflects the direction of control of hyperammonemia. It is therefore essential that amino acids are checked daily in the acutely sick child with hyperammonemia secondary to a urea cycle defect.