ACUTE MANAGEMENT OF PRIMARY HYPERAMMONEMIA

Ammonia level (µmol/l)	Undiagnosed case	Known urea cycle disorders
Above upper limit of normal	• Stop protein intake • Give IV glucose at an appropriate dosages to prevent catabolism (10mg/kg/min)+/- insuline	Same
>100 but less than 250 in neonate >150 but <250)	• Start drug treatment with nitrogen scavengers (l-arginine and ammonul) • Start carnitine, biotin, vitamine B12 • Start Carbaglu® • Start lipid IV 2-3 gram/kg to give higher calories	• Start medications and nitrogen scavengers according to the protocol of each disorder
250-50	• Same as above • Prepare for hemodialysis • Begin hemodialysis if no rapid drop of ammonia within 3–6 h	Same
> 500	Start hemodialysis with above measure	Same

Recommended dosages for medications used in acute management of urea cycle disorders

	Arginine HCl		Ammonul®		N-carbamylglutamate (Carbaglu®)
Kg	<20kg	>20kg	<20 kg	>20kg	Only oral/enteral drug)
Pending diagnosis	250-400mg/kg. Up to 600mg/kg was recommended	250-400 mg/kg Up to 600mg/kg was recommended	250mg/kg	5.5gram/m^2	100mg/kg bolus per NG tube then 25–62.5mg/kg every 6h
NAGS deficiency	250mg/kg	250mg/kg	Not indicated		Same as above
CPS or OTC deficiency	250mg/kg	250mg/kg or 4000mg/m^2/day	250mg/kg Maintenance dose up to 500mg/kg/day	5.5gram/m^2	Not indicated
ASL deficiency	200-400mg/kg. Up to 600mg/kg was recommended.	200-400mg/kg or 12000mg/m^2/day	Same as above		Not indicated
Arginase deficiency	Not indicated		Same as above		Not indicated

Normal ammonia level by age

Age	Upper limit (umol/l)
0-7 day	**94**
8-30 days	**80**
1 m-15yr	**48**
>16	**26**

Secondary hyperammonemia:

Can be due to inborn errors of metabolism such as organic acidemia and fatty acids oxidation defects or drugs or other metabolites that may interfere with urea cycle function, or severe liver disease. Laboratory studies can help to distinguish the underlying primary defect and cause of hyperammonemia and guide appropriate treatment.

Caution:

False positive hyperammonemia is not uncommon, therefore, several precaution should be taken in consideration when collecting blood sample to measure ammonia:

- A free-flowing venous (or arterial) blood sample without tourniquet should be collected into green-top tube (containing lithium or heparin).
- The sample should be transported on ice to the laboratory, separated within 15 minutes of collection and analyzed immediately.