



### **Additional information on Management of Hyperammonemia In Urea Cycle Disorders**

- Stop all sources of protein, both enteral and parenteral nutrition (including albumin).
- In general, start treatment for plasma ammonia greater than 150 micromoles/L or if patient is symptomatic
- Start TFI at 1.5 times maintenance, unless exceptions are noted. TFI should take into account intralipids and medications.

Total Fluid Intake (TFI) – calculated using the 4-2-1 Rule. See table below

<b>Patient's Weight</b>	<b>Recommended Regular Maintenance Daily mL/hour</b>	<b>1.5 Maintenance mL/hour</b>
0 – 10 kg	4 x patient's weight (kg)	6 x patient's weight (kg)
11 – 20 kg	2 x (patient's weight – 10) + 40	3 x (patient's weight – 10) + 60
Greater than 20 kg	1 x (patient's weight – 20) + 60	1.5 x (patient's weight – 20) + 90

- Do not decrease dextrose fluid rate or amount and do not stop calorie delivery in the acute state for any reason as this can precipitate catabolism, which will further worsen the patient's condition. The purpose of the glucose administration is to promote endogenous hyperinsulinemia rather than to prevent hypoglycemia.
- Usual dose of fat emulsion 20% is 1-3 g/kg/day.
- Limited compatibility information exists for IV Ammonul<sup>®</sup>.
  - Ammonul<sup>®</sup> may be run at the y-site with arginine infusion.
  - There is no compatibility information of Ammonul<sup>®</sup> mixed or y-sited with NaCl.
  - D10W + NaCl 0.45% or 0.9% +/- KCL should be run via separate IV line and can be y-sited with fat emulsion (SMOFlipid or Intralipid).
- Ammonul<sup>®</sup> 10 mg/mL mixed in D10W contains approximately 5.2 mmol sodium/100 mL equivalent to approximately NaCl 0.3%
- Pharmacy to prepare Ammonul<sup>®</sup> and Arginine infusions during regular business hours. Pharmacy will return to hospital to prepare if initiation can wait ~2 hours. IV Ammonul<sup>®</sup> is in pyxis medication and can be prepared by nursing if required STAT.
- IV Ammonul<sup>®</sup> has a strong offensive smell. Avoid spilling when preparing or manipulating bag for administration.