BIMDG
British Inherited Metabolic Disease Group

INTRAVENOUS DRUG CALCULATORS FOR THE EMERGENCY TREATMENT OF HYPERAMMONAEMIA

- **Please read these instructions carefully**
- **These calculators have been checked but the BIMDG is not liable for any errors. Do not modify the spreadsheets**

1. Enter child's weight in the yellow box in kilograms and press return. If the child weights more than 40kg it may be appropriate to use the adult protocol
2. For the first 24 hours, use the standard loading given over 1.5 hours (if appropriate) and the maintenance dose over 24 hours. A loading dose is not always necessary in patients with known diagnoses - if in doubt discuss with the regional metabolic unit
3. If there are problems, discuss the dose with the regional metabolic unit. If not using the standard dose enter it in mg/kg per 24 hours in the green box
4. For arginine, enter the concentration in the ampoule in the orange box, then proceed as above

IMPORTANT NOTES: For convenience and consistency, the solutions should be made up at a concentration of 50mg/ml and delivered via:

- Either a syringe drive (maximum concentration 2.5g in 50ml syringe)
- Or an infusion pump (maximum concentration 25g in 500ml bag)
- The concentration reaching the patient should be more than 25mg/ml, which is most easily achieved by piggy-backing all the drug infusions into the maintenance fluid infusion.

WARNING: The doses of sodium benzoate and phenylbutyrate are based on standard ampoules of 1g in 5ml except AMMONUL.

For OTC, CPS and NAGS deficiencies – CLICK HERE

For Citrullinaemia and Arginosuccinic Aciduria – CLICK HERE

For HHH/LPI and Arginase deficiency – CLICK HERE

If using Sodium Benzoate for Proprionic Acidaemia or Methylmalonic Acidaemia – CLICK HERE

For an undiagnosed patient with hyperammonaemia – CLICK HERE

If using the combined sodium benzoate/sodium phenylbutyrate preparation AMMONUL – CLICK HERE