NEONATAL

Hyperammonaemia Medications

This document should be read in conjunction with this **DISCLAIMER**

Highly Restricted: Requires Neonatologist/Metabolic Physician approval before commencing

⚠ **ALL 3 medications are combined in ONE syringe in a final volume of 50 mL, and then administered to the patient by intravenous infusion.**
Plasma ammonia levels to be monitored regularly and at the discretion of the treating neonatologist/metabolic physician**.**

Presentation	**L-Arginine** 50% (5g/10mL) ampoule = 500mg/mL (Armagine®) **Sodium Benzoate** 2g/10mL ampoule = 200mg/mL (Amzoate®) **Sodium Phenylbutyrate** 2g/10mL ampoule = 200mg/mL (Amybutyrate®) KEMH: Hyperammonaemia kit kept in SCN3 Medication Imprest Cupboard PCH: Medications are kept in the ADM as virtual kit: **'Hyperammonaemia kit'**
Classification	Ammonia Scavengers
Indication	Acute Hyperammonaemia – Suspected Urea Cycle Disorders
Dose	**Dosage is determined by the Metabolic Physician. The following regimen represents maximum dosages for each medication.** **Loading Dose** is administered over **2** hours. **Maintenance Dose** is administered over **24** hours or as directed by the Metabolic Physician. See **Appendix 1** for prescribing on Neonatal Variable Rate Infusion Chart MR725.01 (KEMH)/ MR828.02(PCH).

Medication	Loading Dose	Maintenance Dose
Arginine	250mg/kg (0.5mL/kg)	250mg to 600mg/kg (0.5mL to 1.2mL/kg)
Sodium benzoate	250mg/kg (1.25mL/kg)	250mg/kg (1.25mL/kg)
Sodium phenylbutyrate	250mg/kg (1.25mL/kg)	250mg/kg (1.25mL/kg)

Administration	Administer by IV infusion via a large vein or central line where possible. **Loading Dose** is administered over **2** hours. **Maintenance Dose** is administered over **24** hours or as directed by the Metabolic Physician. Follow with the chaser to ensure that the full dose is administered.	
Monitoring	**Note: During the LOADING DOSE, there is no need to give maintenance fluids. See 'Notes' below.** • Monitor plasma ammonia levels 1-2 hourly as ordered • Monitor blood sugar and blood gases every 2 to 4 hours • Monitor electrolytes – sodium may increase due to sodium content in sodium benzoate and sodium phenylbutyrate • Monitor plasma potassium concentration as hypokalaemia is common. Potassium should be added once urine flow is normal and the plasma potassium concentration is known.	
Dose Adjustment	Adjust dose according to response, as advised by the Metabolic Physician.	
Guidelines & Resources	Hyperammonaemia	
Compatible Fluids	Glucose 5% (preferred), Glucose 10%, Sodium Chloride 0.9%	
Preparation	Prepare Loading and Maintenance doses separately and clearly mark each syringe as Loading or Maintenance "Ammonia scavengers" All three medications are to be combined together in 5% glucose, in a 50mL syringe. Draw up the required doses of arginine, sodium phenylbutyrate and sodium benzoate and make up to 50mL in the syringe with 5% glucose.	
Adverse Reactions	**Common**	Extravasation risk, hypokalaemia , electrolyte imbalance
Storage	Store at room temperature **KEMH:** Hyperammonaemia kit kept in SCN3 Medication Imprest Cupboard **PCH:** Medications are kept in the ADM as virtual kit: **'Hyperammonaemia kit'**	

Notes	The LOADING DOSE gives the approximate equivalent of 20mL/kg of a Normal Saline bolus. The accompanying glucose and fluid quantities of the LOADING DOSE means there is no need to give maintenance fluids. However, maintenance glucose, fluids and electrolytes will need to be added during the MAINTENANCE DOSE. SAS Category A Form to be completed (online submission at PCH)
References	AMH Children's Dosing Companion (online). Adelaide: Australian MedicinesHandbook Pty Ltd; 2017 January. British Inherited Metabolic Disease Group – Emergency Intravenous Treatment for Urea Cycle Disorders. BIMDG_Adult_UCD_Emergency-Therapy_2012. Summar, M. Current Strategies for the management of urea cycle disorders. The Journal of Pediatrics. Volume 138,1. doi:10.1067/mpd.2001.111834 Truven Health Analytics. Arginine, Sodium Benzoate, Sodium Phenylbutyrate. In: NeoFax [Internet]. Greenwood Village (CO): Truven Health Analytics; 2019 [cited 2019 Jan 12]. Available from: https://neofax.micromedexsolutions.com/ Baxter K. BNF for Children January 2017. London: BMJ Group, Pharmaceutical Press and RCPCH Publications Ltd; 2017. King Edward Memorial Hospital - NCCU Clinical Guidelines, Metabolic Management. Neonatal Hyperammonaemia. http://www.kemh.health.wa.gov.au/services/nccu/guidelines/documents/8378. Pdf PMH ED Guidelines: Intravenous Arginine, Carnitine, Sodium Benzoate and Sodium Phenylbutyrate- Preparation of. 2014 May. University College London Hospitals Foundation NHS Trust. UCL Hospital Injectable Medicines Administration Guide [online]. Chichester: John Wiley & Sons; 2010. Paediatric Injectable Guidelines 4th Edition, Royal Children's Hospital 2016 (online). L-Arginine Hydrochloride (Amargine®) , Sodium Benzoate (Amzoate®), Sodium Phenybutyrate (Ambutyrate®) product information.

Appendix 1 : Sample Infusion Chart Order

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DO NOT WRITE IN BINDING MARGIN

Women and Newborn Health Service
Neonatology Directorate

NEONATAL VARIABLE RATE INFUSION CHART

Year: 20_19 APPENDIX 1 SAMPLE

ALLERGIES & ADVERSE DRUG REACTIONS
☐ Nil Known ☐ Unknown ☐ Yes – refer to NIMC (Tick appropriate box)

Patient Name: Baby's Name	Date: 09/02/19
Gest Age 38+1	CGA 38+5
BW	Working Wt 3.8kg

Med Rec. No: .....
Surname: ..... UMRN Sticker
Forename: ..... AFFIX LABEL HERE
Gender: ..... D.O.B. .....

MEDICATION ORDER		ADMINISTRATION OF ALL 3 MEDICATIONS COMBINED			RATE CHANGE					
Date: 09/02/19	Medication: Arginine	Date:	09/02/19	09/02/19						
Route: IV		Time:	0900	1100						
Dose in Infusion: 950mg	Dose/kg/time (at 1mL/hr):	Rate (mL/hr):	25mL/hr	2mL/hr						
		Doctor:	Dr's Signature	Dr's Signature						
Diluent: Glucose 5%	Dose Calculation: 250mg x weight (3.8kg)	Nurse:	Nurse 1 / Nurse 2	Nurse 1 / Nurse 2						
Final Volume:	Doctor name: Dr's name Signature: Dr's Signature	Volume Discarded:								
Date: 09/02/19	Medication: Sodium Benzaote	Date:	Direction: Combine ALL 3 medications in one syringe to a final volume of 50mL as per the 'Hyperammonaemia Guideline"							
Route: IV		Time:								
Dose in Infusion: 950mg	Dose/kg/time (at 1mL/hr):	Rate (mL/hr):								
		Doctor:								
Diluent: Glucose 5%	Dose Calculation: 250mg x weight (3.8kg)	Nurse:								
Final Volume:	Doctor name: Dr's Name Signature: Dr's Signature	Volume Discarded:								
Date: 09/02/19	Medication: Sodium Phenylbutyrate	Date:								
Route: IV		Time:								
Dose in Infusion: 950mg	Dose/kg/time (at 1mL/hr):	Rate (mL/hr):								
		Doctor:								
Diluent: Glucose 5%	Dose Calculation: 250mg x weight (3.8kg)	Nurse:								
Final Volume:	Doctor name: Dr's Name Signature: Dr's Signature	Volume Discarded:								

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MR725.01 (KEMH) or MR828.02 (PMH/PCH) NEONATAL VARIABLE RATE INFUSION CHART

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