SODIUM PHENYLBUTYRATE This drug must be guardrailed

Trade Name	Ambutyrate
Class	Ammonium Detoxicant
Mechanism of Action	Sodium Phenylbutyrate is a pro-drug, it is converted to phenylacetglutamine in the body and serves as a urea substitute.
Indications	Adjunctive therapy for maintenance management of urea cycle disorder, usually in combination with sodium benzoate.
Contraindications	Use with caution in patients with severe hypertension, congestive heart failure, oedema or renal impairment. (This medication contains significant amounts of sodium)
Supplied As	Sodium -4-Phenylbutyrate 200mg/mL
Dilution	**Drug** / **Add 5% or 10% Glucose** / **Total Volume** / **Concentration** 1g (5mL) / 45mL / 50mL / **20mg/mL** Can concentrate further to a maximum of 50mg/mL if needed
Dosage ***Must chart guardrail and use Alaris pump***	**Loading dose:** 250mg/kg over 90 minutes **IV Maintenance:** 10 -20mg/kg/hr (see infusion sheet) **Oral Maintenance:** 300-600mg/kg/day after iv stabilisation adjusted to response **Maximum dose:** 20g/day
Guardrails	Conc: Min – 20 mg/mL Max – 50 mg/mL Soft Min: 10 mg/kg/hr Hard Max: 167 mg/kg/hr Soft Max: 20 mg/kg/hr Default: 20 mg/kg/hr
Interval	**Loading Dose:** Once only, given over 90 minutes **Maintenance:** Continuous iv infusion Can be changed to oral 6-8 hourly when stable
Administration	**IV:** acute management – loading and initial maintenance **Oral**: maintenance when stable
Compatible With	Dextrose 5% and Dextrose 10% Oral doses may be mixed with fruit drinks, milk or feeds
Incompatible With	Very limited compatibility data Do not mix with any other medications

Monitoring	Plasma ammonia and glutamine concentrations, liver and renal function tests.
Stability	Single use vial, discard immediately after use
Storage	Store unopened vials at room temperature Oral powder should be stored in an air tight container.
Adverse Reactions	Nausea, vomiting, anorexia, metabolic acidosis, anaemia, rash, arrhythmias, hypoalbuminaemia, electrolyte disturbances, increased liver enzymes, increased bilirubin, offensive body odour.
Metabolism	Sodium Phenylbutyrate undergoes saturable conjugation to phenylacetglutamine (Micahelis Menton Kinetics) Half life = 0.8hours (parent compound) = 1.2hours (phenylacetglutamine) 80% of phenylacetglutamine is eliminated in urine in 24hours
Interactions	No known drug interactions Avoid mixing with acidic beverages (cola, lemonade, grape juice etc) as these may cause sodium phenylbutyrate to precipitate.
Comments	Section 29 medication. Will need funding approval for discharge Sodium content of 200mg/mL Inj = 1.1 mmol/mL
References	1. BNF for Children 2016-17 pp 572- 573. 2. Taketomo el al Paediatric Dosage Handbook 16th Edition 2009. 3. https://www.nucdf.org/documents/Consensus%20Documents/neonatal.pdf
Updated By	A Lynn, B Robertshawe, M Meeks July 2012 A Lynn B Robertshawe November 2018. A Lynn B Robertshawe February 2022 (routine review) Aug 2022 infusion sheet created