VANCOUVER GENERAL HOSPITAL
CSU PHARMACEUTICAL SCIENCES
SPECIAL ACCESS PROGRAM (SAP) DRUG DATA SHEET

DRUG NAME
Sodium Phenylacetate / Sodium Benzoate (SA/SB)

ALTERNATE NAME
$C_8H_7NaO_2$ / $C_7H_5NaO_2$

MANUFACTURER
Ucyclyd Pharma

STRENGTH
Sodium Phenylacetate 100mg/mL and
Sodium Benzoate 100mg/mL in 50mL vials

DOSAGE FORM
Injection

INDICATIONS
- Treatment of hyperammonemia in patients with urea cycle enzyme deficiencies.

DOSAGE AND ADMINISTRATION
For intercurrent hyperammonemic encephalopathy in patients with confirmed diagnosis of urea cycle disorder under management with sodium phenylbutyrate:

Carbamyl Phosphate Synthetase (CPS) or Ornithine Transcarbamylase (OTC) Deficiency
- Priming infusion: 0.25g/kg/dose or 5.5g/M^2/dose SA/SB (with 0.20g/kg/dose or 4g/M^2/dose of arginine) in 400 to 600mL/M^2 of D10W infused over 90 minutes
- Sustaining infusion: 0.25g/kg/day or 5.5g/M^2/day SA/SB (with 0.20g/kg/day or 4g/M^2/day of arginine) in 400 to 600mL/M^2 of D10W infused over 24 hours

Argininosuccinic Acid Synthetase Deficiency
- Priming infusion: 0.25g/kg/dose or 5.5g/M^2/dose SA/SB (with 0.60g/kg/dose or 12g/M^2/dose of arginine) in 400 to 600mL/M^2 of D10W infused over 90 minutes
- Sustaining infusion: 0.25g/kg/day or 5.5g/M^2/day SA/SB (with 0.60g/kg/day or 12g/M^2/day of arginine) in 400 to 600mL/M^2 of D10W infused over 24 hours

KNOWN SIDE EFFECTS*
- Nausea, vomiting, increased anion gap, mild hyperventilation, mild respiratory alkalosis, exacerbation of peptic ulcers, edema (in view of the sodium content).
- Signs of toxicity include confusion, lethargy, vomiting, obtundation in the absence of hyperammonemia, hyperventilation, hypernatremia, hyperkalemia, mixed respiratory alkalosis/metabolic acidosis, hyperosmolar coma, cerebral edema, cardiovascular collapse, death.

SPECIAL PRECAUTIONS
- Sodium phenylacetate / sodium benzoate for injection must be further diluted prior to administration. Diluted solutions are stable at for 24 hours at room temperature.
- Punctured vials are stable for up to 3 days when refrigerated.
- Compatible with arginine in D10W. No other compatibility information is available.
- If the plasma ammonium level does not decrease significantly within 8 hours, hemodialysis should be started as an emergency procedure. Repeat priming doses are NOT recommended.
- Hypokalemia may occur secondary to urine and potassium loss; monitor plasma potassium levels.
- Some antibiotics such as penicillin may affect the overall disposition of, probenecid may affect the renal excretion of, and valproic acid may antagonise the efficacy of sodium phenylacetate / sodium benzoate.

*** REPORT ANY ADVERSE DRUG REACTIONS TO CSU PHARMACEUTICAL SCIENCES**