

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

**DRUG NAME**

Sodium Phenylacetate / Sodium Benzoate (SA/SB)

**ALTERNATE NAME**

C<sub>8</sub>H<sub>7</sub>NaO<sub>2</sub> / C<sub>7</sub>H<sub>5</sub>NaO<sub>2</sub>

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**MANUFACTURER**

Ucyclyd Pharma

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**STRENGTH**

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

**DOSAGE FORM**

Injection

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**INDICATIONS**

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

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**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<sup>2</sup>/dose SA/SB (with 0.20g/kg/dose or 4g/M<sup>2</sup>/dose of arginine) in 400 to 600mL/M<sup>2</sup> of D10W infused over 90 minutes
- Sustaining infusion: 0.25g/kg/day or 5.5g/M<sup>2</sup>/day SA/SB (with 0.20g/kg/day or 4g/M<sup>2</sup>/day of arginine) in 400 to 600mL/M<sup>2</sup> of D10W infused over 24 hours

*Argininosuccinic Acid Synthetase Deficiency*

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

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**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.

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**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.

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

Sodium Phenylacetate Sodium Benzoate  
July2007