

## Sodium Phenylbutyrate Granules



### Product Name

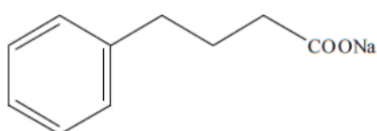
Generic name: Sodium Phenylbutyrate Granules

### Ingredient

The main ingredient of this product is sodium phenylbutyrate.

Chemical name: sodium 4-phenylbutyrate

Chemical Structure:



Molecular formula:  $C_{10}H_{11}NaO_2$

Molecular weight: 186.18

Excipients: silicon dioxide, magnesium stearate

### Description

It occurs as a white to off-white granule.

### Indication

Adjunctive therapy of urea cycle disorders, involving deficiencies of carbamylphosphate synthetase, ornithine transcarbamylase or argininosuccinate synthetase.

This product is suitable for all patients with complete enzyme deficiency in the neonatal period (within 28 days of birth), and also for patients with late-onset (partial enzyme deficiency, occurring 1 month after birth) with a history of hyperammonemic encephalopathy.

### Strength

150g/vial (It contains 0.94g phenylbutyric acid per 1g)

## **Dosage and administration**

This product should be used under the supervision of a physician experienced in the treatment of urea cycle abnormalities. It should be administered orally or via a gastrostomy or nasogastric tube. It is important to identify diagnosis early and start treatment immediately to improve survival. Any episode of acute hyperammonemia should be considered a life-threatening emergency.

### **1. The recommended dose**

The total daily dose of sodium phenylbutyrate in clinical experience is usually:

- 450- 600mg/ kg/day (newborns, infants and children weighing <20 kg)
- 9.9-13.0g/m<sup>2</sup>/day (children, adolescents and adults weighing >20kg)

The safety and efficacy of sodium phenylbutyrate at a daily dose of greater than 20g have not been established.