Prescribing Information – Republic of Ireland

Pheburane (483mg/g sodium phenylbutyrate)
Please refer to the full Summary of Product Characteristics (SmPC) before prescribing
Presentation: Each gram of granules contains 483 mg of sodium phenylbutyrate. Each bottle contains 174 g of granules. Excipients with known effect: Each gram of sodium phenylbutyrate contains 124 mg (5.4 mmol) of sodium and 768 mg of sucrose.
Indication: Adjunctive therapy in the chronic management of urea cycle disorders, involving deficiencies of carbamylphosphate synthetase, ornithine transcarbamylase or argininosuccinate synthetase. In all patients with neonatal-onset disease (complete enzyme deficiencies, presenting within the first 28 days of life). In patients with late-onset disease (partial enzyme deficiencies, presenting after the first month of life) who have a history of hyperammonaemic encephalopathy.
Dosage: The daily dose should be individually adjusted according to the patient's protein tolerance and the daily dietary protein intake needed to promote growth and development (see section 4.2 of full SmPC). The usual total daily dose of sodium phenylbutyrate in clinical experience is 450-600 mg/kg/day in neonates, infants and children weighing less than 20 kg; and 9.9-13.0 g/m^2/day in children weighing more than 20 kg, adolescents and adults. The safety and efficacy of doses in excess of 20 g/day have not been established. The total daily dose should be divided into equal amounts and given with each meal or feeding (e.g. 4-6 times per day in small children). A calibrated dosing spoon is provided which dispenses up to 3g of sodium phenylbutyrate by graduation of 250 mg. For oral use only. Therapeutic monitoring and nutritional management should be conducted as per section 4.2 of the full SmPC.
Contraindications: Hypersensitivity to any active substance excipients; pregnancy; breast-feeding.
Special warnings and precautions for use: Contains 124 mg (5.4 mmol) of sodium per gram of sodium phenylbutyrate. Should be used with caution in patients with congestive heart failure or severe renal insufficiency, and where there is sodium retention with oedema. Serum potassium should be monitored during therapy. Even on therapy, acute hyperammonaemic encephalopathy may occur. Not recommended for the management of acute hyperammonaemia, which is a medical emergency. Considered high in sodium. this should be taken into account for those on a low salt diet. Contains sucrose, this should be taken into account for those with diabetes mellitus. Patients with rare hereditary problems of fructose intolerance, glucose-galactose malabsorption or sucrase-isomaltase insufficiency should not take this medicinal product.
Fertility, pregnancy and lactation: Women of child-bearing potential must use effective contraception. Contraindicated in pregnancy and breast feeding.
Interactions with other medicinal products and other forms of interaction: Concurrent administration of probenecid may affect renal excretion of the conjugation product of sodium phenylbutyrate. There have been published reports of hyperammonaemia being induced by haloperidol and by valproate. Corticosteroids may cause the breakdown of body protein and thus increase plasma ammonia levels. More frequent monitoring of plasma ammonia levels is advised when these medicinal products have to be used.
Undesirable effects: Adverse reactions mainly involved the reproductive and gastrointestinal system. The SmPC should be consulted for a full list of adverse reactions. V*ery common*: amenorrhoea, irregular menstruation; *common*: anaemia, thrombocytopenia, leukopenia, leukocytosis, thrombocytosis, metabolic acidosis, alkalosis, decreased appetite, depression, irritability, syncope, headache, oedema, abdominal pain, vomiting, nausea, constipation, dysgeusia, rash, abnormal skin odour, renal tubular acidosis.Decreased blood potassium, albumin, total protein and phosphate. Increased blood alkaline phosphatase, transaminases, bilirubin, uric acid, chloride, phosphate and sodium. Increased weight.
Shelf life: 3 years. After the first opening, to be used within 45 days.
MA number: EU/1/13/822/001
Legal Category: Prescription Only Medicine (POM).
Marketing Authorisation Holder (MAH): Eurocept International BV, Trapgans 5, 1244RL Ankeveen, The Netherlands.
Date of last revision of the prescribing information: March 2022

Adverse events should be reported. Reporting forms and information can be found at: www.hpra.ie
Adverse events should also be reported to Proveca Pharma Ltd.
E-mail: medical@proveca.com Phone: +353 1 513 6233