RAVICTI®▼

(glycerol phenylbutyrate) Oral Liquid

Rethink potential.

Keeping ammonia under control.

All day, every day.

Mean ammonia and glutamine levels were shown to be lower on Ravicti treatment compared to Sodium Phenylbutyrate (NaPBA) in both short and long-term prospective studies [1-7]

- Individual studies indicated non-inferiority of GPB vs NaPBA and ammonia and glutamine levels were statistically significantly lower in pooled analysis.

Patients switched from NaPBA treatment to long term Ravicti treatment showed significant improvements in executive function [6]

- In a long-term open label study in paediatric patients aged 6 – 17 years of age, effective treatment with Ravicti showed significant improvements in Behaviour Rating Inventory of Executive Function (BRIEF) scores.

-Most BRIEF subscales at baseline were consistent with borderline clinically significant dysfunction. After a year of effective Ravicti treatment all BRIEF domains including behavioural regulation, goal setting, planning and self-monitoring were significantly improved in 22 out of 26 patients.

Long term treatment has been associated with fewer hyperammonaemic crises vs NaPBA [6, 7]

Designed to facilitate easy administration and adherence to treatment:

- nearly tasteless and odourless oral liquid
- convenient low volume dosing [1,8]

Ravicti prescribing information can be found overleaf.

References: 1.Ravicti SPC 2. Ravicti EPAR 3. Smith W, et al. J Pediatr. 2013 Jun 13;162(6):1228–1234 4. Lee B, et al. Mol Genet Metab. 2010 Jul;100(3):221–228.
5. Lichter-Konecki U, et al. Mol Genet Metab. 2011 Aug;103(4):323–329 6. Diaz GA, et al. Hepatology. 2013;57(6):2171–2179 7. Berry SA et al, Mol Genet Metabol 122 (2017) 46-53
8. Nagamani SCS et al. MGM 2015;116;29-34

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Immedica pharma

Ravicti®▼(glycerol phenylbutyrate) abbreviated prescribing information

For further prescribing information, please refer to the Ravicti Summary of Product Characteristics

Presentation: Each ml of liquid contains 1.1g of glycerol phenylbutyrate. **Indications:** Ravicti is indicated for use as adjunctive therapy for chronic management of patients with urea cycle disorders including deficiencies of carbamoyl phosphate synthetase I, ornithine carbamoyltransferase, argininosuccinate synthetase, argininosuccinate lyase, arginase I and ornithine translocase deficiency hyperornithinaemia-hyperammonaemia homocitrullinuria syndrome who cannot be managed by dietary protein restriction and/or amino acid supplementation alone.Ravicti must be used with dietary protein restriction and, in some cases, dietary supplements.
Dosage and Administration: Ravicti should be prescribed by a physician experienced in the management of UCDs. Ravicti liquid should be administered orally or via gastrostomy or nasogastric tube. The daily dose should be individually adjusted according to the patient's protein tolerance and the daily dietary protein intake needed. The recommended dosages for patients naïve to phenylbutyric acid and for patients switching from sodium phenylbutyrate or from sodium phenylacetate/sodium benzoate injection to Ravicti are different. The recommended total daily dose of Ravicti is based on body surface area and ranges from 4.5 ml/m2/day to 11.2 ml/m2/day [5.3 g/m2/day to 12.4 g/m2/day). Total daily dosage is given in equally divided dosages three to six daily. See SPC for full details.
Contraindications: Hypersensitivity to the active substance and treatment of acute hyperammonaemia.
Warnings and Precautions: Whilst on treatment with glycerol phenylbutyrate, acute hyperammonaemia including hyperammonaemic encephalopathy may occur in a proportion of patients. Ammonia levels should be monitored in patients with pancreatic insufficiency or intestinal malabsorption. High levels of the active metabolite PAA should be suspected in patients with unexplained somnolence, confusion, nausea and lethargy who have normal or low ammonia. **Overdose:** Phenylacetic acid is associated with signs and symptoms of neurotoxicity and could accumulate in patients who receive an overdose.
Pregnancy and Lactation: Ravicti should not be used during pregnancy. **Undesirable Effects:** See SPC for full details. The most frequently reported adverse reactions during glycerol phenylbutyrate treatment were diarrhoea, flatulence, headache, decreased appetite, vomiting, fatigue, nausea and, skin odour abnormal. **UK List Price:** Ravicti 1.1 g/ml oral liquid. £161.00 per bottle excluding VAT. EIRE List Price: Available on request. **Legal Category:** POM. Marketing Authorisation Numbers: EU/1/15/1062/001. Further Information is available from Immedica Pharma UK Ltd, The Townhouse, 114-116 Fore Street, Hertford, Herts, SG14 1AJ. **Marketing Authorisation Holder:** Immedica Pharma AB, Norrtullsgatan 15, SE-113 29 Stockholm, Sweden **Date of Preparation:** February 2020 **Company Reference:** 001_RAV_UK_2020

▼This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Adverse reactions are to be reported according to national local regulations. Reporting forms and information can be found for UK at www.mhra.gov.uk/yellowcard and for Ireland at www.hpra.ie. Adverse reactions can also be reported to Immedica Pharma UK Ltd by email at safety@immedica.com

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