or squash puree and used within 2 hours. For administration advice via nasogastric or gastrostomy tube—consult product literature.

- **HANDLING AND STORAGE** Manufacturer advises discard contents of bottle 14 days after opening.
- **PATIENT AND CARER ADVICE**
  Driving and skilled tasks Manufacturer advises patients and carers should be counselled on the effects on driving and performance of skilled tasks—increased risk of dizziness.
- **NATIONAL FUNDING/ACCESS DECISIONS**
  Scottish Medicines Consortium (SMC) decisions
  SMC No. 1342/18
  The *Scottish Medicines Consortium* has advised (August 2018) that glycerol phenylbutyrate (*Ravicti*®) is accepted for use within NHS Scotland as an adjunctive therapy for chronic management of adult and paediatric patients aged 2 months and older with urea cycle disorders who cannot be managed by dietary protein restriction and/or amino acid supplementation alone. Glycerol phenylbutyrate must be used with dietary protein restriction and, in some cases, dietary supplements. This advice is contingent upon the continuing availability of the patient access scheme in NHS Scotland or a list price that is equivalent or lower.
  All Wales Medicines Strategy Group (AWMSG) decisions
  AWMSG No. 2127
  The *All Wales Medicines Strategy Group* has advised (December 2019) that glycerol phenylbutyrate (*Ravicti*®) is recommended as an option for use within NHS Wales for use as an adjunctive therapy for chronic management of patients with urea cycle disorders 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. This recommendation applies only in circumstances where the approved Wales Patient Access Scheme (WPAS) is utilised or where the list/contract price is equivalent or lower than the PAS/WPAS price.
- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug.
  Liquid
  - Ravicti (Immedica Pharma AB) ▼
    Glycerol phenylbutyrate 1.1 gram per 1 ml Ravicti 1.1g/ml oral liquid | 25 ml PoM £161.00

Sodium phenylbutyrate

29-Apr-2020

- **INDICATIONS AND DOSE**
  Long-term treatment of urea cycle disorders (as adjunctive therapy in all patients with neonatal-onset disease and in those with late-onset disease who have a history of hyperammonaemic encephalopathy) (under expert supervision)
  - BY MOUTH
  - Adult: 9.9–13 g/m^2 daily in divided doses, with meals; maximum 20 g per day

IMPORTANT SAFETY INFORMATION

EMERGENCY MANAGEMENT OF UREA CYCLE DISORDERS

For further information on the emergency management of urea cycle disorders consult the British Inherited Metabolic Disease Group (BIMDG) website at www.bimdg.org.uk.

- **CAUTIONS** Conditions involving sodium retention with oedema (preparations contain significant amounts of sodium) · congestive heart failure (preparations contain significant amounts of sodium)
- **INTERACTIONS** → Appendix 1: sodium phenylbutyrate
- **SIDE-EFFECTS**
  - **Common or very common** Abdominal pain · anaemia · appetite decreased · constipation · depression · headache · irritability · leucocytosis · leucopenia · menstrual cycle irregularities · metabolic acidosis · metabolic alkalosis · nausea · oedema · renal tubular acidosis · skin reactions · syncope · taste altered · thrombocytopenia · thrombocytosis · vomiting · weight increased
  - **Uncommon** Anorectal haemorrhage · aplastic anaemia · arrhythmia · gastrointestinal disorders · pancreatitis
- **CONCEPTION AND CONTRACEPTION** Manufacturer advises adequate contraception during administration in women of child-bearing potential.
- **PREGNANCY** Avoid—toxicity in *animal* studies.
- **BREAST FEEDING** Manufacturer advises avoid—no information available.
- **HEPATIC IMPAIRMENT** Manufacturer advises caution.
- **RENAL IMPAIRMENT** Manufacturer advises use with caution (preparations contain significant amounts of sodium).
- **DIRECTIONS FOR ADMINISTRATION** Granules should be mixed with food before taking. *Pheburane*® granules must not be administered by nasogastric or gastrostromy tubes.
- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug. Forms available from special-order manufacturers include: capsule, oral suspension, oral solution
  Granules
  - Ammonaps (Immedica Pharma AB)
    Sodium phenylbutyrate 940 mg per 1 gram Ammonaps 940mg/g granules sugar-free | 266 gram PoM £860.00 DT = £860.00
  - Pheburane (Eurocept International bv)
    Sodium phenylbutyrate 483 mg per 1 gram Pheburane 483mg/g granules | 174 gram PoM £331.00

  Tablet
  - Ammonaps (Immedica Pharma AB)
    Sodium phenylbutyrate 500 mg Ammonaps 500mg tablets | 250 tablet PoM £493.00 DT = £493.00

2.13 Wilson's disease

Other drugs used for Wilson's disease Penicillamine, p. 1158

ANTIDOTES AND CHELATORS › COPPER ABSORPTION INHIBITORS

Zinc acetate

- **DRUG ACTION** Zinc prevents the absorption of copper in Wilson's disease.
- **INDICATIONS AND DOSE**
  Wilson's disease (initiated under specialist supervision)
  - BY MOUTH
  - Adult: 50 mg 3 times a day (max. per dose 50 mg 5 times a day), adjusted according to response

  DOSE EQUIVALENCE AND CONVERSION
  - Doses expressed as elemental zinc.

  PHARMACOKINETICS
  - Symptomatic Wilson's disease patients should be treated initially with a chelating agent because zinc has a slow onset of action. When transferring from chelating treatment to zinc maintenance therapy, chelating treatment should be co-administered for 2–3 weeks until zinc produces its maximal effect.
- **CAUTIONS** Portal hypertension (risk of hepatic decompensation when switching from chelating agent)