

- **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**
 - ▶ With oral use Oral dose may be mixed with fruit drinks, milk, or feeds. Granules should be mixed with food before taking. *Pheburane*® granules must not be administered by nasogastric or gastrostomy tubes.
 - ▶ With intravenous use For *intravenous infusion*, dilute to a maximum concentration of 50 mg/mL with Glucose 10%.
- **PATIENT AND CARER ADVICE** Medicines for Children leaflet: Sodium phenylbutyrate for urea cycle disorders www.medicinesforchildren.org.uk/sodium-phenylbutyrate-urea-cycle-disorders

- **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, solution for infusion

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

3.14 Wilson's disease

Other drugs used for Wilson's disease Penicillamine, below

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**
- ▶ Child 1-5 years: 25 mg twice daily
- ▶ Child 6-15 years (body-weight up to 57 kg): 25 mg 3 times a day
- ▶ Child 6-15 years (body-weight 57 kg and above): 50 mg 3 times a day
- ▶ Child 16-17 years: 50 mg 3 times a day

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)
- **INTERACTIONS** → Appendix 1: zinc
- **SIDE-EFFECTS**
 - ▶ **Common or very common** Epigastric discomfort (usually transient)
 - ▶ **Uncommon** Leucopenia · sideroblastic anaemia
 - ▶ **Frequency not known** Condition aggravated
- **SIDE-EFFECTS, FURTHER INFORMATION** Transient gastric irritation may be reduced if first dose is taken mid-morning or with a little protein.
- **PREGNANCY**
 - ▶ **Dose adjustments** Usual dose 25 mg 3 times daily adjusted according to plasma-copper concentration and urinary copper excretion.
- **BREAST FEEDING** Manufacturer advises avoid; present in milk—may cause zinc-induced copper deficiency in infant.
- **MONITORING REQUIREMENTS** Monitor full blood count and serum cholesterol.
- **DIRECTIONS FOR ADMINISTRATION** Capsules may be opened and the contents mixed with water.

- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug.

Capsule**CAUTIONARY AND ADVISORY LABELS 23**

- ▶ **Wilzin** (Recordati Rare Diseases UK Ltd)
Zinc (as Zinc acetate) 25 mg Wilzin 25mg capsules | 250 capsule [PoM](#) £132.00 DT = £132.00
- ▶ **Zinc (as Zinc acetate) 50 mg** Wilzin 50mg capsules | 250 capsule [PoM](#) £242.00 DT = £242.00

ANTIDOTES AND CHELATORS > COPPER CHELATORS

Penicillamine

- **DRUG ACTION** Penicillamine aids the elimination of copper ions in Wilson's disease (hepatolenticular degeneration).

INDICATIONS AND DOSE**Wilson's disease**

- ▶ **BY MOUTH**
- ▶ Child 1 month-11 years: 20 mg/kg daily in 2-3 divided doses, to be taken 1 hour before food; maximum 2 g per day
- ▶ Child 12-17 years: Initially 20 mg/kg daily in 2-3 divided doses, maintenance 0.75-1 g daily, to be taken 1 hour before food; maximum 2 g per day

Cystinuria

- ▶ **BY MOUTH**
- ▶ Child: 20-30 mg/kg daily in 2-3 divided doses, lower doses may be used initially and increased gradually, doses to be adjusted to maintain 24-hour urinary cystine below 1 mmol/litre, maintain adequate fluid intake, to be taken 1 hour before food; maximum 3 g per day

- **CONTRA-INDICATIONS** Lupus erythematosus
- **CAUTIONS** Neurological involvement in Wilson's disease
- **INTERACTIONS** → Appendix 1: penicillamine
- **SIDE-EFFECTS**
 - ▶ **Common or very common** Proteinuria · thrombocytopenia
 - ▶ **Rare or very rare** Alopecia · breast enlargement (males and females) · connective tissue disorders · haematuria (discontinue immediately if cause unknown) · hypersensitivity · oral disorders · skin reactions
 - ▶ **Frequency not known** Agranulocytosis · aplastic anaemia · appetite decreased · fever · glomerulonephritis · Goodpasture's syndrome · haemolytic anaemia · increased risk of infection · jaundice cholestatic · leucopenia · lupus-