Solution for infusion

- **Potassium chloride (Non-proprietary)**

Potassium chloride 150 mg per 1 ml Potassium chloride 15% (potassium 20mmol/10ml) solution for infusion 10ml ampoules | 10 ampoule PoM £7.00–£10.00 | 20 ampoule PoM £6.50–£11.24
Potassium chloride 15% (potassium 20mmol/10ml) solution for infusion 10ml Mini-Plasco ampoules | 20 ampoule PoM £10.70
Potassium chloride 200 mg per 1 ml Potassium chloride 20% (potassium 13.3mmol/5ml) solution for infusion 5ml ampoules | 10 ampoule PoM £8.00

Oral solution

CAUTIONARY AND ADVISORY LABELS 21

- **Kay-Cee-L** (Geistlich Sons Ltd)

Potassium chloride 75 mg per 1 ml Kay-Cee-L syrup sugar-free | 500 ml P £8.77 DT = £8.77

Infusion

- **Potassium chloride (Non-proprietary)**

Potassium chloride 30 mg per 1 ml Potassium chloride 3% (potassium 40mmol/100ml) infusion 100ml bags | 1 bag PoM
Potassium chloride 3% (potassium 20mmol/50ml) infusion 50ml bags | 1 bag PoM

3 Metabolic disorders

Metabolic disorders

Use of medicines in metabolic disorders

Metabolic disorders should be managed under the guidance of a specialist. As many preparations are unlicensed and may be difficult to obtain, arrangements for continued prescribing and supply should be made in primary care.

General advice on the use of medicines in metabolic disorders can be obtained from:
Alder Hey Children's Hospital, Medicines Information Centre
(0151) 252 5381
and
Great Ormond Street Hospital for Children, pharmacy
(020) 7405 9200

Urea cycle disorders

Sodium benzoate p. 664 and sodium phenylbutyrate p. 665 are used in the management of urea cycle disorders. Both, either singly or in combination, are indicated as adjunctive therapy in all patients with neonatal-onset disease and in those with late-onset disease who have a history of hyperammonaemic encephalopathy. Sodium benzoate is also used in non-ketotic hyperglycinaemia.

The long-term management of urea cycle disorders includes oral maintenance treatment with sodium benzoate and sodium phenylbutyrate combined with a low protein diet and other drugs such as arginine p. 662 or citrulline p. 663, depending on the specific disorder.

Emergency management

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.

3.1 Acute porphyrias

Acute porphyrias

01-Oct-2017

Overview

The acute porphyrias (acute intermittent porphyria, variegate porphyria, hereditary coproporphyria, and 5-aminolaevulinic acid dehydratase deficiency porphyria) are hereditary disorders of haem biosynthesis; they have a prevalence of about 1 in 75 000 of the population.

Great care must be taken when prescribing for patients with acute porphyria, since certain drugs can induce acute porphyric crises. Since acute porphyrias are hereditary, relatives of affected individuals should be screened and advised about the potential danger of certain drugs.

EvGr Where there is no safe alternative, drug treatment for serious or life-threatening conditions should not be withheld from patients with acute porphyria. Where possible, the clinical situation should be discussed with a porphyria specialist for advice on how to proceed and monitor the patient. In the UK clinical advice can be obtained from the National Acute Porphyria Service or from the UK Porphyria Medicines Information Service (UKPMIS)—see details below. E

Haem arginate p. 653 is administered by short intravenous infusion as haem replacement in moderate, severe, or unremitting acute porphyria crises.

In the United Kingdom the National Acute Porphyria Service (NAPS) provides clinical support and treatment with haem arginate from two centres (University Hospital of Wales and King's College Hospital). To access the service telephone (029) 2074 7747 and ask for the Acute Porphyria Service.

Drugs unsafe for use in acute porphyrias

EvGr The following list contains drugs on the UK market that have been classified as 'unsafe' in porphyria because they have been shown to be porphyrinogenic in animals or in vitro, or have been associated with acute attacks in patients. Absence of a drug from the following lists does not necessarily imply that the drug is safe. For many drugs no information about porphyria is available. E

An up-to-date list of drugs considered safe in acute porphyrias is available from the UKPMIS, see *Useful resources* below.

Further information may be obtained from: porphyria.eu/ and also from:
The UK Porphyria Medicines Information Service (UKPMIS)
University Hospital of Wales
CF14 4XW
Cardiff
(029) 2074 2979/3877

Quite modest changes in chemical structure can lead to changes in porphyrinogenicity but where possible general statements have been made about groups of drugs; these should be checked first.

Unsafe Drug Groups (check first)

- Anabolic steroids
- Antidepressants, MAOIs (contact UKPMIS for advice)
- Antidepressants, Tricyclic and related (contact UKPMIS for advice)
- Barbiturates (includes primidone and thiopental)
- Contraceptives, hormonal (for detailed advice contact UKPMIS or a porphyria specialist)
- Hormone replacement therapy (for detailed advice contact UKPMIS or a porphyria specialist)
- Imidazole antifungals (applies to oral and intravenous use; topical antifungals are thought to be safe due to low systemic exposure)
- Non-nucleoside reverse transcriptase inhibitors (contact UKPMIS for advice)
- Progestogens (for detailed advice contact UKPMIS or a porphyria specialist)
- Protease inhibitors (contact UKPMIS for advice)
- Sulfonamides (includes co-trimoxazole and sulfasalazine)
- Sulfonylureas (glipizide and glimepiride are thought to be safe)
- Taxanes (contact UKPMIS for advice)
- Triazole antifungals (applies to oral and intravenous use; topical antifungals are thought to be safe due to low systemic exposure)