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-like syndrome · myasthenia gravis · nausea · nephrotic syndrome · neurological deterioration in Wilson's Disease · neutropenia · pancreatitis · polymyositis · pulmonary haemorrhage · rash (consider dose reduction) · respiratory disorders · Stevens-Johnson syndrome · taste loss (mineral supplements not recommended) · vomiting · yellow nail syndrome
- **ALLERGY AND CROSS-SENSITIVITY** Patients who are hypersensitive to penicillin may react rarely to penicillamine.
- **PREGNANCY** Fetal abnormalities reported rarely; avoid if possible.
- **BREAST FEEDING** Manufacturer advises avoid unless potential benefit outweighs risk—no information available.
- **RENAL IMPAIRMENT**
Dose adjustments Reduce dose and monitor renal function or avoid (consult product literature).
- **MONITORING REQUIREMENTS**
- Consider withdrawal if platelet count falls below 120 000/mm^3 or white blood cells below 2500/mm^3 or if 3 successive falls within reference range (can restart at reduced dose when counts return to within reference range but permanent withdrawal necessary if recurrence of leucopenia or thrombocytopenia).
- Monitor urine for proteinuria.
- Monitor blood and platelet count regularly.
- **PATIENT AND CARER ADVICE** Counselling on the symptoms of blood disorders is advised. Warn patient and carers to tell doctor immediately if sore throat, fever, infection, non-specific illness, unexplained bleeding and bruising, purpura, mouth ulcers, or rashes develop.

- **MEDICINAL FORMS** There can be variation in the licensing of different medicines containing the same drug. Forms available from special-order manufacturers include: oral solution

Tablet

CAUTIONARY AND ADVISORY LABELS 6, 22

- **Penicillamine (Non-proprietary)**
Penicillamine 125 mg Penicillamine 125mg tablets | 56 tablet PoM £54.00 DT = £45.00
Penicillamine 250 mg Penicillamine 250mg tablets | 56 tablet PoM £88.77 DT = £88.77

Trientine dihydrochloride

- **INDICATIONS AND DOSE**

Wilson's disease in patients intolerant of penicillamine

- BY MOUTH
- Child 2-11 years: Initially 0.6–1.5 g daily in 2–4 divided doses, adjusted according to response, to be taken before food
- Child 12-17 years: 1.2–2.4 g daily in 2–4 divided doses, adjusted according to response, to be taken before food

- **INTERACTIONS** → Appendix 1: trientine
- **SIDE-EFFECTS**
- **Rare or very rare** Anaemia
- **Frequency not known** Gastrointestinal disorders · nausea · neurological deterioration in Wilson's Disease · rash
- **PREGNANCY** Teratogenic in *animal* studies—use only if benefit outweighs risk.
Monitoring Monitor maternal and neonatal serum-copper concentrations.
- **PRESCRIBING AND DISPENSING INFORMATION** Trientine is **not** an alternative to penicillamine for rheumatoid arthritis or cystinuria. Penicillamine-induced systemic lupus erythematosus may not resolve on transfer to trientine.

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

Capsule

CAUTIONARY AND ADVISORY LABELS 6, 22

- **Trientine dihydrochloride (Non-proprietary)**
Trientine dihydrochloride 300 mg Trientine dihydrochloride 300mg capsules | 100 capsule PoM £3,090.00 DT = £3,090.00

4 Mineral and trace elements deficiencies

4.1 Zinc deficiency

Zinc deficiency

Overview

Zinc supplements should not be given unless there is good evidence of deficiency (hypoproteinaemia spuriously lowers plasma-zinc concentration) or in zinc-losing conditions. Zinc deficiency can occur as a result of inadequate diet or malabsorption; excessive loss of zinc can occur in trauma, burns, and protein-losing conditions. A zinc supplement is given until clinical improvement occurs, but it may need to be continued in severe malabsorption, metabolic disease, or in zinc-losing states.

Zinc is used in the treatment of Wilson's disease and acrodermatitis enteropathica, a rare inherited abnormality of zinc absorption.