



Bonus Monographs



Cysteamine bitartrate (SIS-tee-a h-meen)

PREGNANCY CATEGORY: C CLASSIFICATION(S):

Urinary tract drug
Rx: Cystagon

ACTION/KINETICS

Lowers cystine levels of cells in cystinosis, which is an inherited defect of lysosomal transport. In those with cystinosis, cystine transport out of lysosomes is abnormal, resulting in the formation of crystals which damage the kidney. Other tissues are damaged as well, including the retina, muscles, and CNS. Acts within the cell to convert cystine into both cysteine and cysteine-cysteamine mixed disulfide, which can leave the lysosome in those with cystinosis.

USES

Management of nephropathic cystinosis in adults and children.

CONTRAINDICATIONS

Hypersensitivity to cysteamine or penicillamine. Use during lactation.

SIDE EFFECTS

CNS: Lethargy, somnolence, depression, encephalopathy, **seizures**, headache, ataxia, confusion, tremor, hyperkinesia, dizziness, jitteriness, nervousness, abnormal thinking, emotional lability, hallucinations, nightmares.

GI: N&V, anorexia, abdominal pain (may be severe), diarrhea, bad breath, dyspepsia, constipation, gastroenteritis, duodenitis, duodenal ulceration.

Hematologic: Reversible leukopenia, anemia. **Miscellaneous;** Decreased

hearing, fever, rash, dehydration, hypertension, urticaria.

LABORATORY TEST CONSIDERATIONS

Abnormal LFTs.

OD OVERDOSE MANAGEMENT

Symptoms: Extension of side effects, respiratory symptoms. *Treatment:* Support the cardiovascular and respiratory systems. Hemodialysis may be effective in removing the drug from the body.

HOW SUPPLIED

Capsule: 50 mg, 150 mg

DOSAGE

• CAPSULES

Nephropathic cystinosis.

Initial: New clients should be started on one-fourth to one-sixth of the maintenance dose. The dose is then raised gradually over 4–6 weeks to avoid intolerance. **Maintenance, children up to age 12 years:** 1.3 g/m²/day (of the free base) given in four divided doses. **Maintenance, children over 12 years and over 110 lb:** 2 g/day in four divided doses.

NURSING CONSIDERATIONS

ADMINISTRATION/STORAGE

1. Initiate therapy in children and adults promptly after the diagnosis has been confirmed by increased white cell cystine levels.
2. Do not give intact cysteamine capsules to children under 6 y.o. due to the possibility of aspiration. Sprinkle contents of the capsule over food.
3. The goal is to keep leukocyte cystine levels less than 1 nmol/½ cystine/mg protein 5–6 hr following administration of cysteamine. Those with intolerance to cysteamine can

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still get a beneficial effect if cystine levels are less than 2 nmol/½ cystine/mg protein. To achieve this level, the dose of cysteamine may be increased to a maximum of 1.95 g/m²/day.

4. Cystinotic clients taking cysteamine HCl or phosphocysteamine solutions may be transferred to equimolar doses of cysteamine bitartrate capsules. Clients being transferred should have their white cell cystine levels measured in 2 weeks and every 3 months thereafter.

ASSESSMENT

Obtain baseline CBC, renal and LFTs, C_{CR} and white cell cystine levels; repeat levels 5–6 hr after therapy.

CLIENT/FAMILY TEACHING

1. Take only as directed. May be given with electrolyte and mineral replace-

ments, vitamin D, and thyroid hormone to manage adverse renal effects.

2. Report rash; provider will withhold drug until cleared and then reinstitute at a lower dose with gradual increases to therapeutic dose. This may also have to be done if CNS or GI side effects occur.

3. Obtain labs to assess for leukocyte cystine levels, abnormal liver function, or reversible leukopenia. Report any unusual or adverse side effects.

OUTCOMES/EVALUATE

- Prevention of organ damage R/T cystine accumulation
- White cell cystine levels of <1 nmol/½ cystine/mg protein