

## Cysteamine (systemic): Drug information

### Brand Names: US

- Cystagon;
- Procysbi

### Brand Names: Canada

- Procysbi

### Pharmacologic Category

- Anticystine Agent;
- Urinary Tract Product

### Mechanism of Action

Reacts with cystine within the lysosome to convert it to cysteine and to a cysteine-cysteamine mixed disulfide, both of which can then exit the lysosome in patients with cystinosis, an inherited defect of lysosomal transport

### Pricing: (US)

#### **Capsule, delayed release (Procysbi Oral)**

25 mg (per each): \$110.48; 75 mg (per each): \$110.48

#### **Capsules (Cystagon Oral)**

50 mg (per each): \$0.44; 150 mg (per each): \$1.28

### Dosage Forms

Capsule, Oral: Cystagon: 50 mg, 150 mg

Capsule Delayed Release, Oral: Procysbi: 25 mg, 75 mg

**Generic Equivalent Available (US):** No

**Use: Nephropathic cystinosis:** Management of nephropathic cystinosis

### Dosing: Adult

**Nephropathic cystinosis:** Oral: **Note:** Begin therapy as soon as the diagnosis of nephropathic cystinosis has been confirmed.

**Immediate release (Cystagon):**

*Initial:* Initiate therapy with  $\frac{1}{6}$  to  $\frac{1}{4}$  of maintenance dose; titrate slowly upward over 4 to 6 weeks.

*Maintenance dose:* 500 mg 4 times daily; maximum dose: 1.95 g/m<sup>2</sup>/day or 90 mg/kg/day (off-label dose) (Gahl 2002)

***Delayed release (Procysbi):***

*Initial:* 0.2 to 0.3 g/m<sup>2</sup>/day divided every 12 hours; titrate slowly upward over 4 to 6 weeks to target maintenance dose

*Maintenance:* 1.3 g/m<sup>2</sup>/day divided every 12 hours; may increase as needed in 10% increments; maximum daily dose: 1.95 g/m<sup>2</sup>/day

**Dosing: Renal Impairment: Adult**

There are no dosage adjustments provided in the manufacturer's labeling.

**Dosing: Hepatic Impairment: Adult**

There are no dosage adjustments provided in the manufacturer's labeling.

**Contraindications**

Hypersensitivity to cysteamine, penicillamine, or any component of the formulation

**Food Interactions**

Concurrent ingestion of food and the delayed release formulation of cysteamine can reduce the systemic exposure of cysteamine. Management: Administer  $\geq 30$  minutes before and  $\geq 2$  hours after meals; if necessary, patients may eat only a small amount (~4 ounces or  $\frac{1}{2}$  cup) of food between 1 hour before and 1 hour after administration.

Delayed release: Do not administer with grapefruit juice. Take  $\geq 30$  minutes before and  $\geq 2$  hours after meals; if necessary, may eat only a small amount (~4 ounces or  $\frac{1}{2}$  cup) of food between 1 hour before and 1 hour after administration. Avoid high fat food close to time of dose. Avoid alcohol intake during therapy.