

Patient Information
PROCYSBI® (Pro-CIS-bee)
(cysteamine bitartrate) delayed-release capsules

What is PROCYSBI?

PROCYSBI is a prescription medicine used to treat nephropathic cystinosis in adults and children 2 years of age and older.

It is not known if PROCYSBI is safe and effective in children under 2 years of age.

Who should not take PROCYSBI?

Do not take PROCYSBI if you are allergic to penicillamine or cysteamine.

What should I tell my doctor before taking PROCYSBI?

Before you take PROCYSBI, tell your doctor if you:

- drink alcohol
- have a skin rash or bone problems
- have or have had stomach or bowel (intestinal) problems including ulcers or bleeding
- have a history of seizures, lack of energy, unusual sleepiness, depression, or changes in your ability to think clearly
- have liver or blood problems
- have any other medical conditions
- are pregnant or plan to become pregnant. It is not known if PROCYSBI will harm your unborn baby. Tell your doctor right away if you think that you are pregnant. Talk with your doctor about the benefits and risks of taking PROCYSBI during pregnancy.
- are breastfeeding or plan to breastfeed. You should not breastfeed during treatment with PROCYSBI. Talk with your doctor about the best way to feed your baby if you take PROCYSBI.

Tell your doctor about all medicines that you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

Know the medicines you take. Keep a list of them to show your doctor and pharmacist when you get a new medicine.

How should I take PROCYSBI?

- Take PROCYSBI exactly as your doctor tells you to.
- Your doctor may start you on a low dose of PROCYSBI and slowly increase your dose to help avoid side effects, especially if you have not taken a medicine that contains cysteamine bitartrate before.
- Do not change your dose of PROCYSBI unless your doctor tells you to.
- PROCYSBI is taken 2 times each day, every 12 hours.
- Take PROCYSBI the same way each time, either without eating or with a small amount of food, as follows:
 - **If you take PROCYSBI without eating**, do not eat for at least 2 hours before taking PROCYSBI and at least 30 minutes after taking PROCYSBI.
 - **If you are not able to take PROCYSBI without eating**, you can eat a small amount of food (½ cup) within 1 hour before you take PROCYSBI through 1 hour after you take it.
 - Avoid eating foods that are high in fat close to the time that you will take a dose of PROCYSBI.
- Talk to your doctor or pharmacist if you have questions about how to take PROCYSBI.
- Swallow PROCYSBI capsules whole.
- Take PROCYSBI with fruit juice, except for grapefruit juice, if you swallow the capsule whole.
- Do not crush or chew PROCYSBI or the capsule contents.
- If you cannot swallow the capsule whole, you can open it and take the capsule contents with certain foods and juices. Read the **“Instructions for Use”** in the package that comes with your PROCYSBI for information about the right way to use PROCYSBI if you cannot swallow the capsules whole or if you have a gastrostomy tube (G-tube).
- Take PROCYSBI at least 1 hour before or 1 hour after you take medicines that contain bicarbonate or carbonate.
- If you miss a dose, take it as soon as possible, up to 8 hours after the scheduled time of the missed dose. If it is less than 4 hours of the time the next dose is due, skip the missed dose. Take the next dose at your regularly scheduled time. Do not take 2 doses at one time to make up for a missed dose.
- If you take too much PROCYSBI, call your doctor or go to the nearest hospital emergency room right away.
- Your doctor should do blood tests to measure the amount of cystine inside your white blood cells or to measure the amount of PROCYSBI in your blood before you start taking it and regularly during treatment with PROCYSBI. This will help your doctor to prescribe the dose of PROCYSBI that is right for you.

- Take supplements if prescribed by your doctor, such as salt or mineral replacement supplements, vitamin D, or thyroid hormone.

What should I avoid while taking PROCYSBI?

- Do not drive or operate machinery until you know how PROCYSBI affects you. PROCYSBI can make you sleepy or less alert than normal.
- Do not drink alcohol if you take PROCYSBI. Drinking alcohol while taking PROCYSBI may change how PROCYSBI works and may cause an increase in the amount of PROCYSBI in your blood that may cause serious side effects.

What are the possible side effects of PROCYSBI?

PROCYSBI can cause serious side effects, including:

- **Skin, bone, and joint problems.** People treated with high doses of cysteamine bitartrate may develop abnormal changes of their skin and bones. These changes may include stretch marks, bone injuries (such as fractures), bone deformities, and joint problems. Check your skin while taking PROCYSBI. Tell your doctor if you notice any skin changes or problems with your bones or joints. Your doctor will check you for these problems.
- **Skin rash.** Skin rash is common with cysteamine bitartrate and may sometimes be severe. **Tell your doctor right away if you get a skin rash.** Your dose of PROCYSBI may need to be decreased until the rash goes away. If the rash is severe, your doctor may tell you to stop taking PROCYSBI.
- **Stomach and bowel (intestinal) problems.** Some people who take other medicines that contain cysteamine bitartrate develop ulcers and bleeding in their stomach or bowel. **Tell your doctor right away** if you get stomach-area pain, nausea, vomiting, loss of appetite, or vomit blood.
- **Central nervous system symptoms.** Some people who take other medicines that contain cysteamine bitartrate develop seizures, depression, and become very sleepy. The medicine may affect how your brain is working (encephalopathy). Tell your doctor right away if you develop any of these symptoms.
- **Low white blood cell count and certain abnormal liver function blood tests.** Your doctor should check you for these problems.
- **Benign intracranial hypertension (pseudotumor cerebri)** has happened in some people who take immediate-release cysteamine bitartrate. This is a condition where there is high pressure in the fluid around the brain. Your doctor should do eye examinations to find and treat this problem early.
Tell your doctor right away if you develop any of the following symptoms while taking PROCYSBI: headache, buzzing or "whooshing" sound in the ear, dizziness, nausea, double vision, blurry vision, loss of vision, pain behind the eye, or pain with eye movement.

The most common side effects with PROCYSBI include:

- vomiting
- nausea
- stomach (abdominal) pain
- breath odor
- diarrhea
- skin odor
- tiredness
- skin rash
- headache

These are not all of the possible side effects of PROCYSBI. Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

General information about PROCYSBI

Medicines are sometimes prescribed for purposes other than those listed in a Patient Information leaflet. Do not use PROCYSBI for a condition for which it was not prescribed. Do not give PROCYSBI to other people, even if they have the same symptoms that you have. It may harm them. You can ask your doctor or pharmacist for information about PROCYSBI that is written for health professionals.

What are the ingredients in PROCYSBI?

Active ingredients: cysteamine bitartrate

Inactive ingredients: microcrystalline cellulose, Eudragit® L 30 D-55, hypromellose, talc, triethyl citrate, sodium lauryl sulfate, and purified water

Capsule shell ingredients: gelatin, titanium dioxide, blue ink and white ink

Distributed by: Horizon Pharma USA, Inc., Lake Forest, IL 60045

For more information go to www.PROCYSBI.com or call 1-855-888-4004.

This Patient Information has been approved by the U.S. Food and Drug Administration.

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