

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

PATIENT MEDICATION INFORMATION

PROCYSBI™

Cysteamine delayed-release capsules

Read this carefully before you start taking **PROCYSBI** and each time you get a refill. This leaflet is a summary and will not tell you everything about this drug. Talk to your healthcare professional about your medical condition and treatment and ask if there is any new information about **PROCYSBI**.

What is PROCYSBI used for?

PROCYSBI is used for treatment of nephropathic cystinosis.

How does PROCYSBI work?

Nephropathic cystinosis is a rare disease where the amino acid cystine builds up in organs and tissues, causing damage. PROCYSBI changes cystine so that it does not build up in the organs and tissues.

What are the ingredients in PROCYSBI?

Medicinal ingredient: Cysteamine bitartrate (also called mercaptamine bitartrate)

Non-medicinal ingredients:

Capsule Contents: Hypromellose, methacrylic acid copolymer, microcrystalline cellulose, purified water, sodium lauryl sulfate, talc and triethyl citrate.

Capsule Shell Ingredients: FD&C Blue#2, gelatin and titanium dioxide.

PROCYSBI comes in the following dosage forms:

Delayed-release capsules, 25 mg and 75 mg.

Do not use PROCYSBI if you:

- are allergic to cysteamine bitartrate or to any of the ingredients in PROCYSBI.
- are allergic to penicillamine.

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take PROCYSBI.

Talk about any health conditions or problems you may have, including if you:

- have skin or bone problems including rashes, stretch marks, fractures, painful joints.
- have a serious skin rash including severe skin peeling especially mouth and eyes, red or purple rash, flu-like symptoms.
- have or have had stomach or bowel (intestinal) problems including ulcers or bleeding or changes in stomach or bowel problems.
- have a history of seizures, lack of energy, unusual sleepiness, depression, ringing in the ear, double vision, loss of vision, pain behind the eye or pain with eye movement 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. PROCYSBI might harm your unborn baby. Tell your healthcare professional right away if you think that you are pregnant. Talk with your healthcare professional about the benefits and risks of taking PROCYSBI during pregnancy. You should also discuss the importance of using birth control during your treatment with PROCYSBI. Your healthcare professional can tell you which birth control options are best for you.
 - are breastfeeding or plan to breastfeed. You should not breastfeed during treatment with PROCYSBI. Talk with your healthcare professional about the best way to feed your baby if you take PROCYSBI.

Other warnings you should know about:

Driving and using machines

Do not drive or operate heavy machinery until you know how PROCYSBI affects you. PROCYSBI can make you sleepy or less alert than normal.

If you are currently taking cysteamine eye drops, do not stop taking them without talking to your healthcare professional since PROCYSBI does not prevent deposits of cystine crystals in the eye.

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

The following may interact with PROCYSBI:

- Bicarbonates and proton pump inhibitors used to reduce stomach acid.
- Do not take with alcohol.

How to take PROCYSBI:

- PROCYSBI should be taken exactly as you are told by your healthcare professional.
- Your healthcare professional will do blood tests before you start treatment with PROCYSBI to decide on the dose that is best for you. You will also have blood tests regularly while you are taking PROCYSBI.
- Your healthcare professional 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 you are told to by your healthcare professional.
- PROCYSBI should be taken without food. Do not eat for at least 2 hours before taking PROCYSBI and 2 hours after. If this is not possible, food can be eaten 30 minutes after taking PROCYSBI.
- Avoid eating foods that are high in fat and protein (e.g. dairy) close to the time that you will take a dose of PROCYSBI.
- Swallow PROCYSBI capsules whole with orange juice. Do not crush or chew

PROCYSBI or the capsule contents.

- For children who are at risk of choking on the capsules (approximately 6 years of age and younger) and for adults who cannot swallow the capsules whole, the capsules can be opened and the capsule contents taken with applesauce (see instructions below). PROCYSBI can also be given through a gastrostomy tube, size 14 French or larger (see instructions below).

Taking PROCYSBI with applesauce:

Do not take PROCYSBI with any food other than applesauce.

- Step 1: Place about ½ cup (4 ounces) of applesauce into a clean container. Do not use any other food.
- Step 2: Open the PROCYSBI capsule. You may need to use more than 1 PROCYSBI capsule for the dose prescribed by your healthcare professional.
- Step 3: Sprinkle the granules that are inside of the capsule or capsules onto the applesauce.
- Step 4: Mix the granules with the applesauce.
- Step 5: Swallow the applesauce and granule mixture within 30 minutes of mixing. Do not chew the granules. Do not save the applesauce and granules for later use.

Giving PROCYSBI through a gastrostomy tube (G-tube) size 14 French or larger:

- It is best to use a straight (bolus) feeding tube.

Use only strained applesauce with no chunks when giving PROCYSBI through a gastrostomy tube (G-tube).

- Step 1: Flush the gastrostomy tube button with 5 mL of water to clear the button.
- Step 2: Place about ½ cup (4 ounces) of applesauce into a clean container. Use at least 1/8 cup (1 ounce) of applesauce for children 25 kg or less starting PROCYSBI at a dose of 1 or 2 capsules.
- Step 3: Open the PROCYSBI capsule. You may need to use more than 1 PROCYSBI capsule for the dose prescribed by your healthcare professional.
- Step 4: Sprinkle the granules that are inside the capsule or capsules on the applesauce. Gently mix the granules with the applesauce.
- Step 5: Place the tip of a catheter tip syringe at the bottom of the container of applesauce and granule mixture. For an adult dose, draw up about 40 mL of the mixture. When giving to a child, draw up at least 10 mL of the mixture for doses of 1 or 2 capsules.
- Step 6: Place the tip of the catheter tip syringe into the feeding tube that will be connected to the gastrostomy tube. Fill the feeding tube with the applesauce and granule mixture.
- Step 7: Hold the feeding tube in a horizontal (straight across) position. Give the applesauce and granule mixture through the gastrostomy tube at a quick and steady rate of 10 mL over 10 seconds.
- Step 8: Repeat Steps 5 through Step 7 until all of the applesauce and granule mixture is given. **Give all of the applesauce and granule mixture through the gastrostomy tube within 30 minutes of mixing.** Do not save the applesauce and granule mixture for later use.
- Step 9: Draw up at least 10 mL of orange juice into another catheter tip syringe. Gently swirl the syringe. Flush the gastrostomy tube with the orange juice. Use enough orange juice to

flush the gastrostomy tube so that there is no applesauce and granule mixture left in the gastrostomy tube.

Usual dose:

Your healthcare professional will tell you how many PROCYSBI capsules to take. PROCYSBI is taken 2 times each day, every 12 hours.

Overdose:

If you think you have taken too much PROCYSBI, contact your healthcare professional, hospital emergency department or regional Poison Control Centre immediately, even if there are no symptoms.

Missed Dose:

If you miss a dose, take it as soon as possible. If it is within 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.

What are possible side effects from using PROCYSBI?

These are not all the possible side effects you may feel when taking PROCYSBI. If you experience any side effects not listed here, contact your healthcare professional.

The most common side effects with PROCYSBI include:

- vomiting
- nausea
- stomach (abdominal) pain and discomfort
- loss of appetite
- breath odour
- diarrhea
- skin odour
- tiredness
- skin rash
- headache
- dizziness
- flushing

PROCYSBI can cause abnormal blood test results. Your healthcare professional will decide when to perform blood tests and will interpret the results.

The following serious side effects, as presented in the table, have been observed with cysteamine (as an immediate release formulation or PROCYSBI).

Serious side effects and what to do about them			
Symptom/effect	Talk to your healthcare professional		Stop taking drug and get immediate medical help
	Only if severe	In all cases	
COMMON Stomach and bowel (intestinal) problems: flu-like symptoms like fever, vomiting and diarrhea.		✓	
UNCOMMON Central nervous system symptoms: seizures, depression, becoming very sleepy, headache, ringing in the ears, dizziness, double vision, loss of vision, pain behind the eye or pain with eye movement.		✓	
Ehlers-Danlos-like Syndrome: Purplish marks on the skin, streaking of the skin, bone problems (including thinning of the bones, spine fractures, curvature of the spine, and "knock knees"), leg pain, and hyperextension of the joints.		✓	
Hypersensitivity reactions: hives, difficulty breathing, swelling of face, lips, tongue, and/or throat.			✓
Kidney problems: swollen hands or legs, or unusual weight gain, decreased urination.		✓	
Stomach and bowel (intestinal) problems: vomiting blood or blood in the stool (ulcers).			✓
VERY RARE Serious skin reactions: any combination of red itchy rash with blisters and peeling of the skin and/or of the lips, eyes, mouth, nasal passages or genitals (Stevens-Johnson Syndrome, Toxic Epidermal Necrolysis, hypersensitivity Syndrome). It often goes with fever, chills, headache, cough, body aches or joint pain. You may have dark urine, yellow skin or eyes.			✓

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, talk to your healthcare professional.

Reporting Side Effects

You can report any suspected side effects associated with the use of health products to Health Canada by:

- Visiting the Web page on Adverse Reaction Reporting (<http://www.hc-sc.gc.ca/dhp-mps/medeff/report-declaration/index-eng.php>) for information on how to report online, by mail or by fax; or
- Calling toll-free at 1-866-234-2345.

NOTE: Contact your healthcare professional if you need information about how to manage your side effects. The Canada Vigilance Program does not provide medical advice.

Storage:

- Store PROCYSBI at room temperature between 20 °C to 25 °C and in a dry place away from light.
- Dispose of capsules by the “Use by” date entered by the pharmacist on bottle.
- Discard expired capsules per your local or provincial regulations.
- Keep PROCYSBI tightly closed in the original bottle.

The 25 mg PROCYSBI bottle contains one desiccant canister and one oxygen absorber canister. The 75 mg PROCYSBI bottle contains one desiccant canister and two oxygen absorber canisters. Do not eat or throw away the desiccant canister or oxygen absorber canister(s).

Keep out of reach and sight of children.

If you want more information about PROCYSBI:

- Talk to your healthcare professional
- Find the full product monograph that is prepared for healthcare professionals and includes this Patient Medication Information by visiting the Health Canada website <http://www.hc-sc.gc.ca>, the manufacturer’s website <http://www.horizontherapeutics.ca>, or by calling 1-844-380-7850.

This leaflet was prepared by Horizon Pharma Ireland Ltd.

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