Package leaflet: Information for the user

PROCYSBI 25 mg gastro-resistant hard capsules
PROCYSBI 75 mg gastro-resistant hard capsules
Cysteamine (mercaptamine bitartrate)

Read all of this leaflet carefully before you start using this medicine because it contains important information for you.

- Keep this leaflet. You may need to read it again.
- If you have further questions, ask your doctor or pharmacist.
- This medicine has been prescribed for you only. Do not pass it on to others. It may harm them, even if their signs of illness are the same as yours.
- If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed in this leaflet. See section 4.

What is in this leaflet

1. What PROCYSBI is and what it is used for

PROCYSBI contains the active substance cysteamine (also known as mercaptamine) and is taken for the treatment of nephropathic cystinosis in children and adults. Cystinosis is a disease affecting how the body functions, with an abnormal accumulation of the amino acid cystine in various organs of the body such as the kidney, eye, muscle, pancreas, and brain. Cystine build-up causes kidney damage and excretion of excess amounts of glucose, proteins, and electrolytes. Different organs are affected at different ages.

PROCYSBI is a medicine that reacts with cystine to decrease its level within the cells. Cysteamine therapy should be initiated promptly after confirmation of the diagnosis of cystinosis to achieve maximum benefit.

2. What you need to know before you take PROCYSBI

Do not take PROCYSBI:

- If you are allergic to cysteamine (also known as mercaptamine) or any of the other ingredients of this medicine (listed in section 6).
- If you are allergic to penicillamine.
- If you are breast-feeding.

Warnings and precautions

Talk to your doctor or pharmacist before taking PROCYSBI.

- Since oral cysteamine doesn't prevent deposits of cystine crystals in the eye, you should continue taking cysteamine eye drops as prescribed by your doctor.
- Whole cysteamine capsules should not be given to children under the age of 6 years due to the risk of choking (refer to section 3 How to take PROCYSBI – Method of administration).
- Serious skin lesions can occur in patients treated with high doses of cysteamine. Your doctor will routinely monitor your skin and bones and reduce or stop your treatment if needed (see section 4).
- Stomach and intestinal ulcers and bleeding can occur in patients receiving cysteamine (see section 4).
- Other intestinal symptoms including nausea, vomiting, anorexia and stomach ache can occur with cysteamine. Your doctor may interrupt and change your dose if these occur.
- Talk to your doctor if you have any unusual stomach symptoms or changes in stomach symptoms.
- Symptoms such as seizures, tiredness, sleepiness, depression, and brain disorders (encephalopathy) can occur with cysteamine. If such symptoms develop, tell your doctor who will adjust your dose.
- Abnormal liver function or reduced white blood cell count (leukopenia) can occur with use of cysteamine. Your doctor will routinely monitor your blood counts and liver function.
- Your doctor will monitor you for benign intracranial hypertension (or pseudotumor cerebri (PTC)) and/or swelling of the optic nerve (papilledema) associated with cysteamine treatment. You will receive regular eye examinations to identify this condition as early treatment can prevent vision loss.

Other medicines and PROCYSBI

Tell your doctor or pharmacist if you are taking, have recently taken, or might take any other medicines. If your doctor prescribes bicarbonate, do not take it at the same time as PROCYSBI; take bicarbonate at least one hour before or at least one hour after the medicine.

PROCYSBI with food and drink

For at least 1 hour before and 1 hour after taking PROCYSBI try to avoid meals, which are rich in fat or proteins as well as any food or liquid that could decrease the acidity in your stomach, like milk or yogurt. If this is not possible, you can eat a small amount (about 100 grams) of food (preferably carbohydrates e.g. bread, pasta, fruits) during the hour before and after taking PROCYSBI.
Take the capsule with an acidic drink (such as orange juice or any acidic juice) or water. For children and patients who have problems to swallow, please refer to section 3 How to take PROCYSBI – Method of administration.

Pregnancy and breastfeeding

If you are pregnant or breast-feeding, think you may be pregnant or are planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.

You should not use this medicine if you are pregnant, particularly during the first trimester. If you are a woman planning a pregnancy or become pregnant, seek immediate advice from your doctor about stopping therapy with this medicine as continued treatment may be harmful to the unborn baby.

Do not use this medicine if you are breastfeeding (see section 2 under “Do not take PROCYSBI”).

Driving and using machines

This medicine may cause some drowsiness. When starting therapy, you should not drive, use machines, or engage in other dangerous activities until you know how the medicine affects you.

PROCYSBI contains sodium

This medicine contains less than 1 mmol sodium (23 mg) per dose, that is to say essentially “sodium-free.”

3. How to take PROCYSBI

Always take this medicine exactly as your doctor or pharmacist has told you. Check with your doctor or pharmacist if you are not sure.

The recommended dose for you or your child will depend on your or your child's age and weight. The targeted maintenance dose is 1.3 g/m2/day.

Dosing schedule

Take this medicine two times a day, every 12 hours. To get the most benefit from this medicine, try to avoid meals and dairy products for at least 1 hour before and 1 hour after PROCYSBI dosing. If this is not possible, you can eat a small amount (about 100 grams) of food (preferably carbohydrates e.g. bread, pasta, fruits) during the hour before and after PROCYSBI administration.

It is important to take PROCYSBI in a consistent way over time.

Do not increase or decrease the amount of medicine without your doctor's approval.

The total usual dose should not exceed 1.95 g/m2/day.

Duration of treatment

Treatment with PROCYSBI should continue life-long, as instructed by your doctor.

Method of administration

You should take this medicine only by mouth.

In order for this medicine to work correctly, you must do the following:

- Swallow the whole capsule with an acidic drink (such as orange juice or any acidic juice) or water. Do not crush or chew capsules or capsule contents. Do not give gastro-resistant hard capsules to children under 6 years of age because they may not be able to swallow them and they may choke. For patients who cannot swallow the whole capsule, the gastro-resistant hard capsule may be opened and the contents may be sprinkled on food (such as apple sauce or berry jelly) and eaten or administered through feeding tubes or mixed in with an acidic drink (such as orange juice or any acidic juice) or water. Consult your child's doctor for complete directions.
- Your medical treatment may include, in addition to cysteamine, one or more supplements to replace

important electrolytes lost through the kidneys. It is important to take these supplements exactly as instructed. If several doses of the supplements are missed or weakness or drowsiness develops, call your doctor for instructions.

- Regular blood tests to measure the amount of cystine inside white blood cells and/or the concentration of cysteamine in the blood are necessary to help determine the correct dose of PROCYSBI. You or your doctor will arrange for these blood tests to be performed. These tests must be obtained 12.5 hours after the evening dose the day before, and therefore 30 minutes after the following morning dose is given. Regular blood and urine tests to measure the levels of the body's important electrolytes are also necessary to help you or your doctor correctly adjust the doses of these supplements.

If you take more PROCYSBI than you should

You should contact your doctor or the hospital emergency department immediately if you have taken more PROCYSBI than you should. You may become drowsy.

If you forget to take PROCYSBI

If you missed a dose of medicine, you should take it as soon as possible. However, if it is within 4 hours of the next dose, skip the missed dose and go back to the regular dosing schedule.

Do not take a double dose to make up for a forgotten dose.

If you have any further questions on the use of this medicine, ask your doctor or pharmacist.

4. Possible side effects

Like all medicines, this medicine can cause side effects, although not everybody gets them.

Tell your doctor or nurse straight away if you notice any of the following side effects – you may need urgent medical treatment:

- Severe allergic reaction (seen uncommonly): Get emergency medical help if you have any of these signs of an allergic reaction: hives; difficulty breathing; swelling of face, lips, tongue, or throat.

If any of the following side effects occur, please contact your doctor immediately. Since some of these side effects are serious, ask your doctor to explain their warning signs.

Common side effects (may affect up to 1 in 10 people):

- Skin rash: Tell the doctor right away if you get a skin rash. PROCYSBI may need to be temporarily stopped until the rash goes away. If the rash is severe, your doctor may discontinue cysteamine treatment.
- Abnormal liver function on blood tests. Your doctor will monitor you for this.

Uncommon side effects (may affect up to 1 in 100 people):

- Skin lesions, bone lesions, and joint problems: Treatment with high doses of cysteamine can cause skin lesions to develop. These include skin striae (which are like stretch marks), bone injuries (such as fractures), bone deformities, and joint problems. Examine your skin while taking this medicine. Report any changes to your doctor. Your doctor will monitor you for these problems.
- Low white blood cell count. Your doctor will monitor you for this.
- Central nervous system symptoms: Some patients taking cysteamine have developed seizures, depression, and become too sleepy (excessive sleepiness). Tell your doctor if you have these symptoms.
- Stomach and intestinal (gastrointestinal) problems: Patients taking cysteamine have developed ulcers and bleeding. Tell your doctor right away if you get stomach ache, nausea, vomiting, loss of appetite, or throw up blood.
- Benign intracranial hypertension, also called pseudotumor cerebri, has been reported with cysteamine use. This is a condition where there is high pressure in the fluid around the brain. 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. Your doctor will monitor you with eye examinations to find and treat this problem early. This will help lessen the chance of loss of eyesight.

The other side effects listed below are given with an estimation of the frequency with which they may occur with PROCYSBI.

Very common side effects (may affect more than 1 in 10 people):

- diarrhoea
- fever
- sensation of sleep

Common side effects:

- unpleasant breath and body odour
- heartburn
- tiredness

Uncommon side effects:

- leg pain
- scoliosis (deviation of the vertebral column)
- bone fragility
- hair discolouration
- fits
- nervousness
- hallucination
- effect on the kidney manifested by swelling of the extremities and weight gain

Reporting of side effects

If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed in this leaflet. You can also report side effects directly via:
United Kingdom: Yellow Card Scheme, Website: www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store
Ireland: HPRA Pharmacovigilance, Website: www.hpra.ie
By reporting side effects you can help provide more information on the safety of this medicine.

5. How to store PROCYSBI

Keep this medicine out of the sight and reach of children.

Do not use this medicine after the expiry date, which is stated on the carton and the bottle label after EXP. The expiry date refers to the last day of that month.

Do not take this medicine if the foil seal has been open for more than 30 days. Discard the open bottle and use a new bottle.

Store in a refrigerator (2°C-8°C). Do not freeze. After opening do not store above 25°C. Keep the container tightly closed in order to protect from light and moisture.

Do not throw away any medicines via wastewater. Ask your pharmacist how to throw away the medicines you no longer use. These measures will help protect the environment.

6. Contents of the pack and other information

What PROCYSBI contains

- The active substance is cysteamine (as mercaptamine bitartrate). Each gastro-resistant hard capsule contains 25 mg or 75 mg of cysteamine.
- The other ingredients are:
  - In the capsules: microcrystalline cellulose, methacrylic acid - ethyl acrylate copolymer (1:1), hypromellose, talc, triethyl citrate, sodium lauryl sulfate.
  - In the capsule shell: gelatin, titanium dioxide (E171), indigo carmine (E132).
  - In the printing ink: shellac, povidone (K-17), titanium dioxide (E171).

What PROCYSBI looks like and contents of the pack

- PROCYSBI 25 mg is presented as blue gastro-resistant hard capsules. The light blue cap is imprinted with “PRO” in white ink and the light blue body is imprinted with “25 mg” in white ink. A white plastic bottle contains 60 capsules. The cap is child resistant and has a foil seal. Each bottle contains two plastic cylinders used for additional moisture and air protection
- PROCYSBI 75 mg is presented as blue gastro-resistant hard capsules. The dark blue cap is imprinted with “PRO” in white ink and the light blue body is imprinted with “75 mg” in white ink. A white plastic bottle contains 250 capsules. The cap is child resistant and has a foil seal. Each bottle contains three plastic cylinders used for additional moisture and air protection.
- Please keep the cylinders in each bottle during the use of the bottle. The cylinders may be discarded with the bottle after use.

Marketing Authorisation Holder

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Detailed information on this medicine is available on the website of the European Medicines Agency http://www.ema.europa.eu.

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