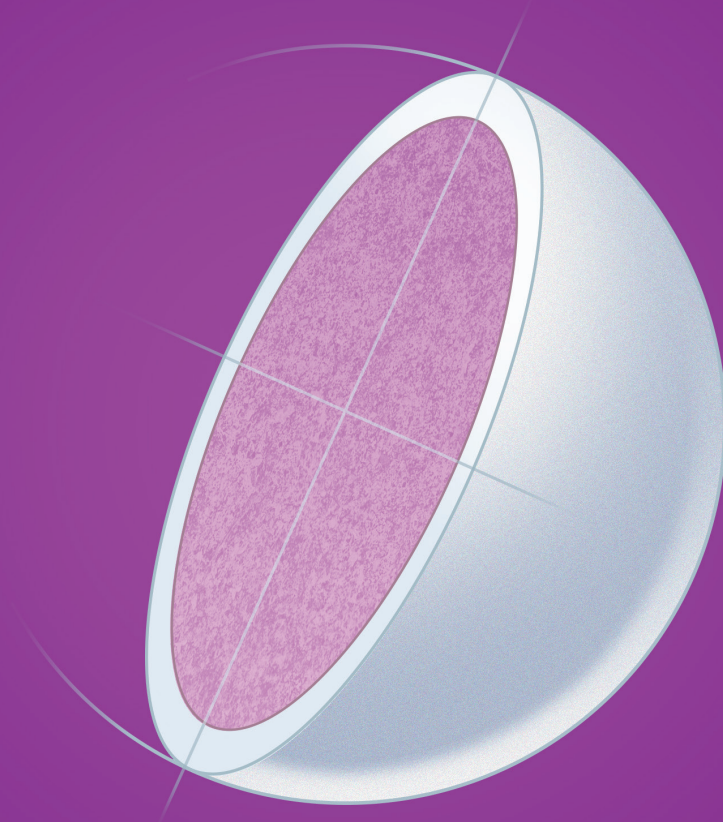


# How to treat **NEPHROPATHIC CYSTINOSIS WITH PROCYSBI**



## **What is PROCYSBI?**

PROCYSBI (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is a prescription medicine used to treat nephropathic cystinosis in adults and children 1 year of age and older. It is not known if PROCYSBI is safe and effective in children under 1 year of age.

## **Who should not take PROCYSBI?**

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

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and [click here for the Patient Package Insert](#).**



**PROCYSBI<sup>®</sup>**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules

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This guide is meant to go along with the information you receive from your healthcare team. If after consulting this guide you still have questions about PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules or how to take it, contact your doctor, pharmacist, or another healthcare team member.

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and [click here for the Patient Package Insert](#).**



# INTRODUCTION TO PROCYSBI

Your doctor has prescribed PROCYSBI—a medicine that treats nephropathic cystinosis by reducing and controlling cystine levels in adults and children 1 year of age and older.

PROCYSBI is a cystine-depleting therapy, or CDT, which means it reduces the amount of cystine in your cells. **PROCYSBI is the only FDA-approved CDT with 12-hour dosing**, which allows you to find a schedule that works for you.

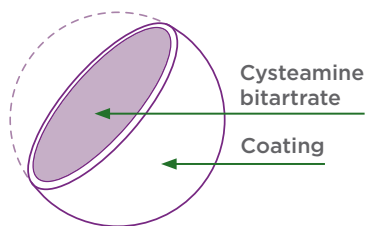
PROCYSBI gives you a choice because it is available in capsules *and* in tear-open packets. Both contain the same medicine that provides 12 hours of continuous cystine control.



Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here](#) for the Patient Package Insert.

# HOW PROCYSBI IS DESIGNED TO WORK

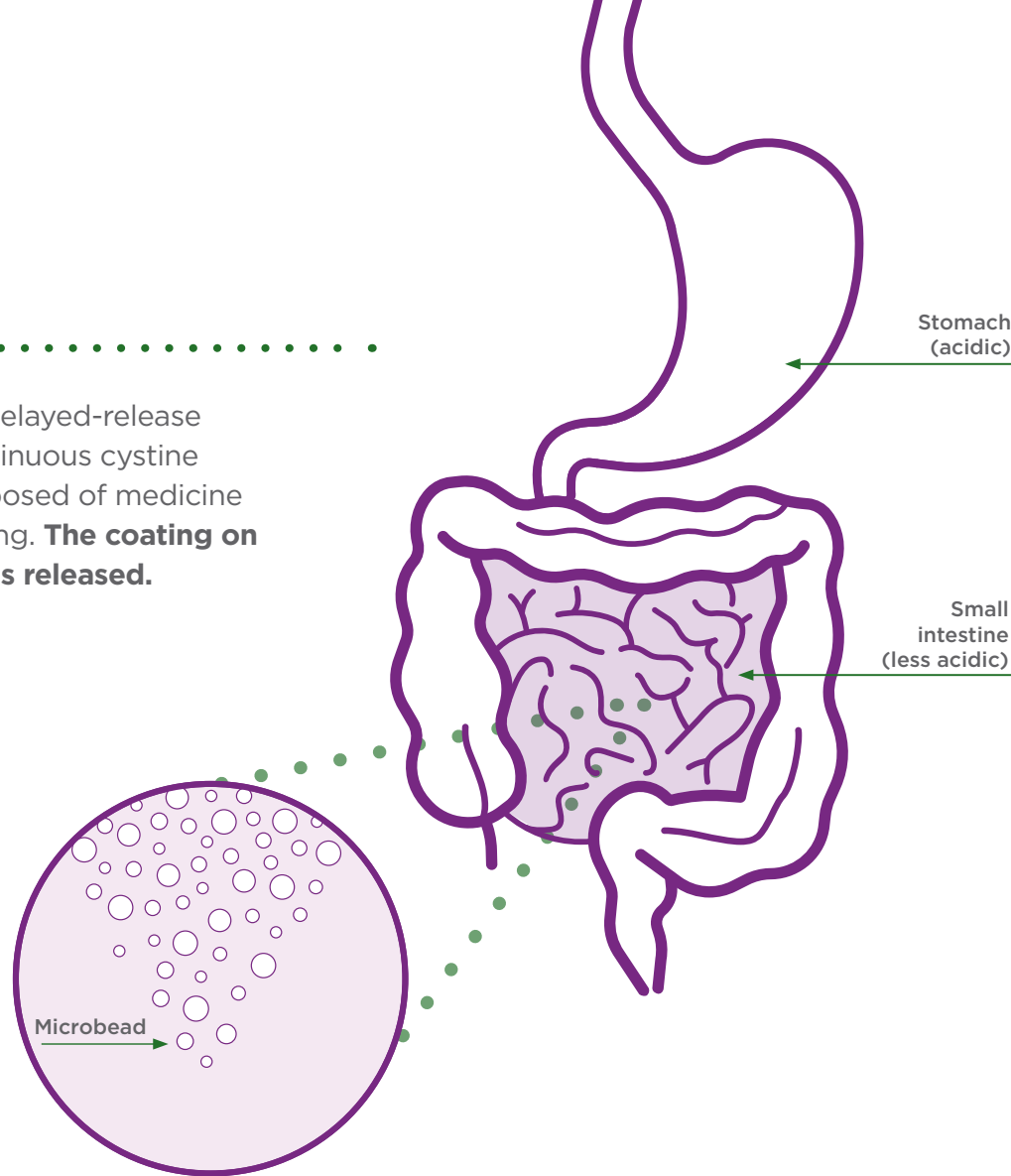
A protective coating helps PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules provide 12-hour continuous cystine control. PROCYSBI granules, also called “microbeads,” are composed of medicine called cysteamine bitartrate surrounded by this protective coating. **The coating on the microbeads helps to control where and how the medicine is released.**



For PROCYSBI to work properly, the microbeads must release medicine *only* in your small intestine. The coated microbeads allow the medicine to pass through the stomach and into the small intestine without dissolving. Once in the small intestine, the coating begins to dissolve and the microbeads release medicine gradually, which allows PROCYSBI to control cystine continuously over 12 hours.

**The most common side effects of PROCYSBI include:** vomiting, nausea, stomach (abdominal) pain, pink eye, diarrhea, cold, tiredness, flu, headache, problems with body salts or electrolytes, infection of ear, nose or throat, joint pain.

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



 **PROCYSBI®**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules

# MAINTAINING THE RIGHT STOMACH ACID LEVELS

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For PROCYSBI microbeads to pass through your stomach and dissolve in your small intestine, the acid in your stomach must be at the right levels. If acid levels are too low, the coating on the microbeads may start dissolving in the stomach, which could change the way PROCYSBI works in the body and may also cause side effects such as stomach pain, nausea, and vomiting.

**What you eat and drink can affect acid levels in your stomach. To maintain the right stomach acid levels:**

- Take PROCYSBI **at least 1 hour before or 1 hour after** you take medicines that contain bicarbonate or carbonate
- Do not eat for at least **2 hours before** taking PROCYSBI and at least **30 minutes after** you take it
  - If you can't take PROCYSBI without eating, take it with no more than 1/2 cup (4 oz) of food **up to 1 hour before or after** you take PROCYSBI
- **Avoid high-fat foods** (such as avocados, cheese, and nuts) right before and after dosing
- **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

**Review approved foods and liquids with which to take PROCYSBI on pages 8 and 9.**

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



# TWO OPTIONS FOR PROCYSBI

PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is available in capsules *and* in tear-open packets. **Both contain the same PROCYSBI microbeads that provide you with 12 hours of continuous cystine control.**

## Capsules



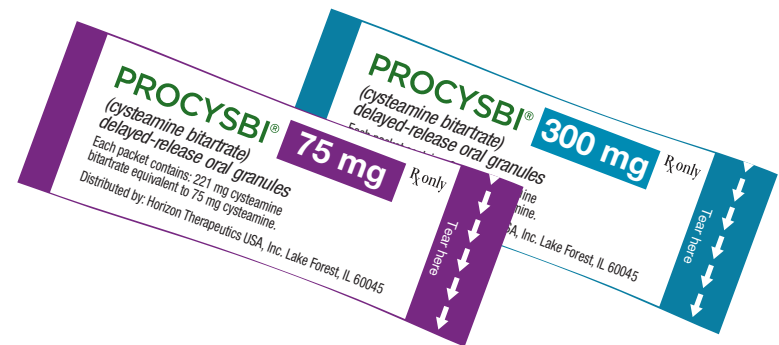
Capsules may be swallowed whole with select liquids, or opened and the microbeads mixed with select foods or liquids.

**PROCYSBI in capsules may be a good option for people who prefer to swallow whole capsules.**

### Who should not take PROCYSBI?

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

## Packets



Packets should be opened and the microbeads mixed with select foods or liquids.

**PROCYSBI in packets may be a good option for people who:**

- Have trouble swallowing
- Take PROCYSBI via a gastrostomy tube (G-tube)
- Are taking the time to open individual capsules at every dose

**Talk to your doctor about whether capsules or packets are best for you. With PROCYSBI, there's a choice!**

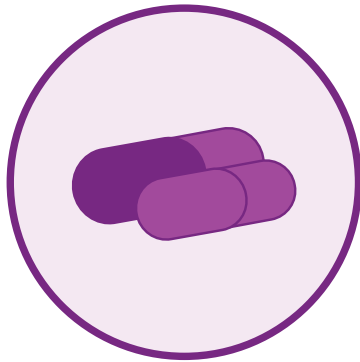
**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



# WAYS TO TAKE PROCYSBI

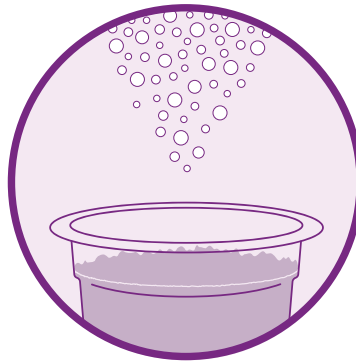
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## Whole capsules



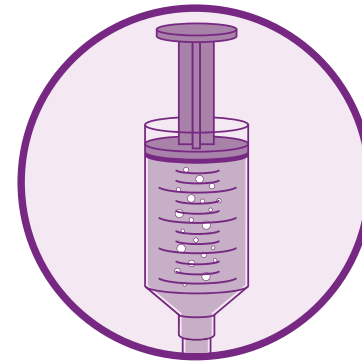
Capsules can be swallowed whole with water or fruit juice (**except grapefruit juice**).

## Mix microbeads with select foods or liquids



Capsules or packets can be opened and the microbeads mixed with select foods or liquids.

## Delivered through G-tube



Microbeads can be mixed with select foods or liquids and delivered through a G-tube that is **size 14 French or larger**.

**You should take PROCYSBI the same way each time.**

Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and [click here for the Patient Package Insert](#).

 **PROCYSBI<sup>®</sup>**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules

# HOW TO TAKE PROCYSBI BY MOUTH

.....

**Capsules** may be swallowed whole with select liquids, or opened and the microbeads mixed with select foods or liquids. **Packets** should be opened and the microbeads mixed with select foods or liquids.

**Keep instructions at your fingertips by printing pages 20 and 21**

**Important notes about taking PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules:**

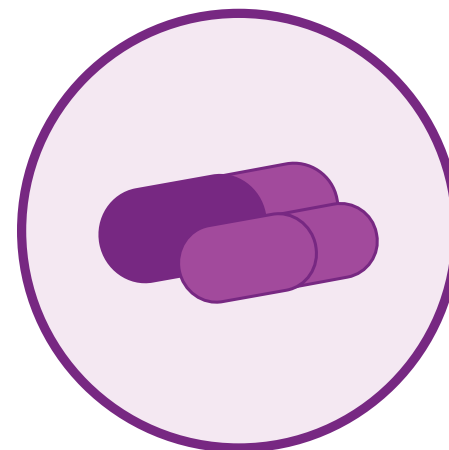
- Take PROCYSBI **the same way each time**
- Do not eat for at least **2 hours before** and **30 minutes after** taking PROCYSBI
  - If you can't take PROCYSBI without eating, take it with no more than 1/2 cup (4 oz) of food **up to 1 hour before or after** you take PROCYSBI
- Take PROCYSBI at least **1 hour before or 1 hour after** you take medicines that contain bicarbonate or carbonate

**Keep PROCYSBI and all medicines out of the reach of children.**

If you have questions about how to take PROCYSBI, contact your doctor or pharmacist, or review the Patient Package Insert located in the pocket of this brochure.

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**

Whole capsules



## STEPS REQUIRED

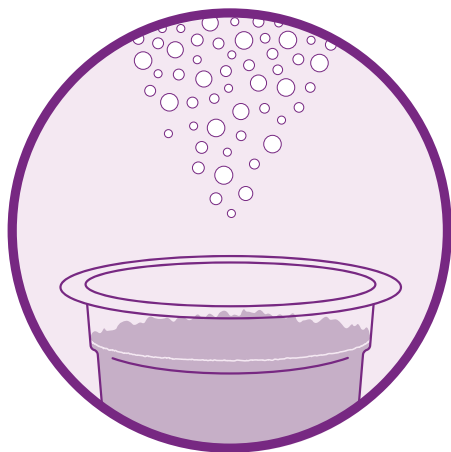
- 1. Swallow** with water or fruit juice **(except grapefruit juice)**. Do not crush or chew the capsules.

 **PROCYSBI®**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules



# WAYS TO TAKE PROCYSBI (CONTINUED)

## Mix microbeads with select foods



### STEPS REQUIRED

1. **Open** the capsule(s) or packet(s). See instructions on page 11.
2. Gently **mix** the microbeads with about 1/2 cup (4 oz) of applesauce or berry jelly. Do not crush or chew the microbeads. **Do not mix with any food other than applesauce or berry jelly.**
3. **Eat** the applesauce or berry jelly mixture within 30 minutes of mixing. Do not save the applesauce or berry jelly mixture for later use.

## Mix microbeads with select liquids



### STEPS REQUIRED

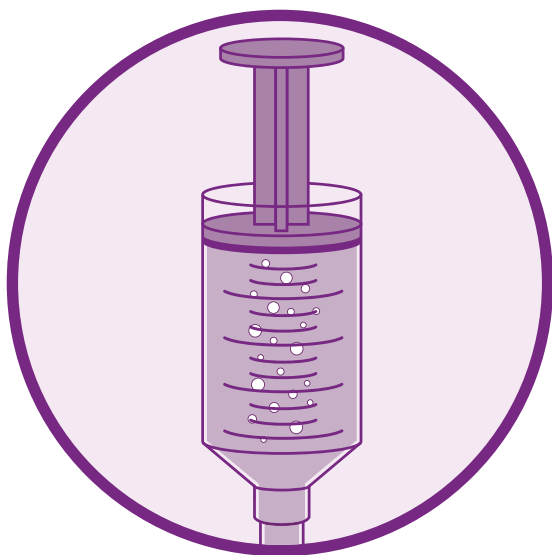
1. **Open** the capsule(s) or packet(s). See instructions on page 11.
2. Gently **mix** the microbeads with about 1/2 cup (4 oz) of any fruit juice (**except grapefruit juice**). Do not crush or chew the microbeads.
3. **Drink all of the fruit juice mixture within 30 minutes of mixing.** Do not save the fruit juice mixture for later use.

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here](#) for the Patient Package Insert.

 **PROCYSBI**<sup>®</sup>  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules

# HOW TO TAKE PROCYSBI VIA G-TUBE

PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules may be given through a G-tube that is **size 14 French or larger** using a straight (bolus) feeding tube. **Capsules or packets** should be opened and the microbeads **mixed with strained applesauce (with no chunks)**.



## STEPS REQUIRED

1. **Flush** the G-tube button with 5 mL of water to clear the button.
2. **Open** the PROCYSBI capsule(s) or packet(s). See instructions on page 11.
3. In a clean container, **sprinkle** the microbeads on about 1/2 cup (4 oz) of strained applesauce with no chunks and mix gently.\*  
\*Children who weigh 55 lb (25 kg) or less can take PROCYSBI with at least 1/8 cup (1 oz) of applesauce.
4. **Place** the tip of a catheter tip syringe at the bottom of the container of the 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.
5. **Place** the tip of the catheter tip syringe into a feeding tube and fill it with the mixture.

Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and [click here for the Patient Package Insert](#).

 **PROCYSBI®**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules

# HOW TO TAKE PROCYSBI VIA G-TUBE (CONTINUED)

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6. Holding the feeding tube in a horizontal position, **push** the mixture through the G-tube at a quick and steady rate of 10 mL over 10 seconds. **Repeat** until all the mixture is given.
7. **Be sure to push all of the mixture through the G-tube within 30 minutes of mixing.** Do not save the mixture for later use.
8. **Draw up** at least 10 mL of fruit juice (**except grapefruit juice**) or water into another catheter tip syringe. Gently swirl the syringe. Flush the G-tube with the fruit juice or water until there is no mixture left in the G-tube.

For questions about how to take PROCYSBI, contact your doctor or pharmacist, or [click here for the Patient Package Insert](#).

## To open PROCYSBI capsules:

- Use dry hands to hold each end of the capsule
- Gently twist the 2 ends in opposite directions to open
- Do not pinch the capsule in the center
- Do not crush or chew the capsule

## To open PROCYSBI packets:

- Tear the packet open along the perforated line

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here for the Patient Package Insert](#).



# HOW YOUR DOCTOR DETERMINES YOUR DOSE OF PROCYSBI

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PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is available in capsules and packets.

- **Capsules** come in 25 mg and 75 mg strengths
- **Packets** come in 75 mg and 300 mg strengths

**Your doctor will determine the appropriate dose for you, which means you may take multiple capsules OR multiple packets every time you take a dose.** Each time you take your medicine, it's important to take the complete dose prescribed by your doctor. Do not change your dose of PROCYSBI unless your doctor tells you to.

## Starting PROCYSBI

When you start on PROCYSBI, your dose may be different than the dose you will take long term (also known as your “maintenance dose”). Your doctor will adjust your dose slowly over time until your cystine levels reach their target.

## Switching between capsules and packets

Capsules and packets have the same weight-based dosing. In some instances, there may not be an exact dose and your doctor may need to make an adjustment. He or she will prescribe a dose that uses entire contents of the packets.

**Your doctor should use cystine level tests to determine the dose of PROCYSBI that is appropriate for you. See pages 14 and 15 for more information about cystine level tests.**

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



# TAKING PROCYSBI

## TWICE A DAY

For PROCYSBI to continuously control your cystine levels, it must be taken **once every 12 hours—2 times a day**.

Example daily dosing schedule for 12-hour PROCYSBI:



Your doctor will tell you the number of capsules or packets you need to take for each dose. If you have any questions, talk to your doctor.

If you take too much PROCYSBI, call your doctor or go to the nearest hospital emergency room right away.

### Missing a dose of PROCYSBI

- If you realize you missed a dose of PROCYSBI **within 8 hours** of when you were scheduled to take it, then **take the dose as soon as possible**
- If you realize you missed a dose of PROCYSBI when your next dose is **due in less than 4 hours**, **wait and take the next dose at your regularly scheduled time**
- Do not take 2 doses at one time to make up for a missed dose

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here for the Patient Package Insert](#).



# THE ROLE OF CYSTINE LEVEL TESTING

Your doctor will regularly order cystine level tests to measure the amount of cystine in your white blood cells (WBCs).

**Cystine level tests help your doctor determine:**

- The dose of PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules that is right for you
- How well the medicine is working to manage your cystine levels
- When and by how much the dose should be increased in growing children

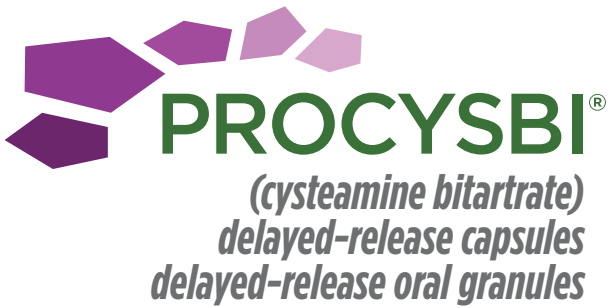
## Types of cystine level tests

Your cystine levels can be measured with regularly scheduled granulocytes or mixed leukocytes tests. Granulocytes and mixed leukocytes are both types of white blood cells. **Your target cystine level is different depending on the type of test, as follows:**

TYPE OF TEST	TESTING INSTITUTION	TEST-SPECIFIC TARGET CYSTINE LEVEL
Granulocytes	University of California San Diego	Less than <b>1.9</b> nmol ½ cystine/mg protein
Mixed leukocytes	Baylor Genetics	Less than <b>1.0</b> nmol ½ cystine/mg protein

**You should have the same cystine level test every time.** Talk with your doctor about which test is right for you.

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here for the Patient Package Insert.](#)



# HOW OFTEN TO HAVE CYSTINE LEVEL TESTS

	AGE	FREQUENCY OF CYSTINE LEVEL TESTING
IF PROCYSBI IS YOUR FIRST CDT	Children aged 1 year to younger than 6 years	<ul style="list-style-type: none"> <li>• 2 weeks after starting PROCYSBI</li> <li>• Continually as long as dose adjustments are needed</li> <li>• After target cystine levels are reached, at least once a month for 3 months, then every 3 months for a year, and then at least 2 times a year</li> </ul>
	Children and adults aged 6 years and older	<ul style="list-style-type: none"> <li>• 2 weeks after reaching the recommended long-term dose</li> <li>• After target cystine levels are reached, at least once a month for 3 months, then every 3 months for a year, and then at least 2 times a year</li> </ul>
IF YOU'RE SWITCHING TO PROCYSBI FROM IMMEDIATE-RELEASE CDT	Children and adults aged 1 year and older	<ul style="list-style-type: none"> <li>• 2 weeks after starting PROCYSBI</li> <li>• Continually as long as dose adjustments are needed</li> <li>• After that, every 3 months for half a year and then at least 2 times a year</li> </ul>

Work with your healthcare team to create a schedule for cystine level tests based on your treatment history and response to PROCYSBI.

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here for the Patient Package Insert](#).



# HOW TO FILL A PRESCRIPTION FOR PROCYSBI

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When your doctor prescribes PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules, you can't fill the prescription at your local retail pharmacy. **A specialty pharmacy fills PROCYSBI prescriptions** and sends them to your home through a package delivery service.

## Filling your first prescription

It's important to begin taking CDT, or cystine-depleting therapy, as soon as possible after being diagnosed with nephropathic cystinosis.

Before you start taking PROCYSBI, your doctor will submit a form to initiate your enrollment in Horizon By Your Side, a patient support program. A Horizon By Your Side representative will help you connect with the specialty pharmacy.

**If you have questions, call the Horizon By Your Side team at 1-855-888-4004.**

## Ordering refills

When to reorder PROCYSBI:

- Before you run out
- Before the discard date written by your pharmacist

When you fill your prescription for PROCYSBI, there should be exactly enough capsules in the bottle or packets in the box to last until your next refill arrives. Do not use expired PROCYSBI.

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



**To avoid a gap in treatment, plan ahead. Mark your calendar to contact the specialty pharmacy to schedule a timely delivery of your refill.**

 **PROCYSBI®**  
(cysteamine bitartrate)  
delayed-release capsules  
delayed-release oral granules



# HOW TO STORE AND DISPOSE OF PROCYSBI

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## Storing PROCYSBI

Keep PROCYSBI and all medicines **out of the reach of children.**

### Capsules

- Keep capsules tightly closed in the original bottle
- Store at room temperature, 68°F-77°F (20°C-25°C), in a dry place away from light
- To keep the medicine dry, capsules are bottled with a small canister that absorbs moisture. Keep the canister in the bottle with the capsules. Do not eat or throw it away

### Packets

- Keep packets closed in the original box
- Store at room temperature, 68°F-77°F (20°C-25°C), in a dry place away from light

## Disposing of unused or expired PROCYSBI

- Discard opened packets after taking your dose. **Do not store or save opened packets of PROCYSBI for your next dose**
- PROCYSBI may not work as intended after the discard date written by your pharmacist. Dispose of any unused PROCYSBI after the expiration date on the bottle or box
  - Ask your pharmacist or contact the US Drug Enforcement Administration at [deaddiversion.usdoj.gov/drug\\_disposal/index.html](https://deaddiversion.usdoj.gov/drug_disposal/index.html) or 1-800-882-9539 to find out how to properly dispose of PROCYSBI in your area

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here](#) for the Patient Package Insert.



# USE AND IMPORTANT SAFETY INFORMATION

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## What is PROCYSBI?

PROCYSBI (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is a prescription medicine used to treat nephropathic cystinosis in adults and children 1 year of age and older. It is not known if PROCYSBI is safe and effective in children under 1 year of age.

## IMPORTANT SAFETY INFORMATION

### What is the most important safety information I should know about 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, such as 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. 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. **Tell your doctor right away if you get a skin rash.**
- **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.

#### Who should not take PROCYSBI?

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

# IMPORTANT SAFETY INFORMATION (CONTINUED)

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## What should I tell my doctor before taking PROCYSBI?

**Tell your doctor if you have any other medical conditions, including 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.
- 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.

## 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?

- See “What is the most important information I should know about PROCYSBI?”

### The most common side effects of PROCYSBI include:

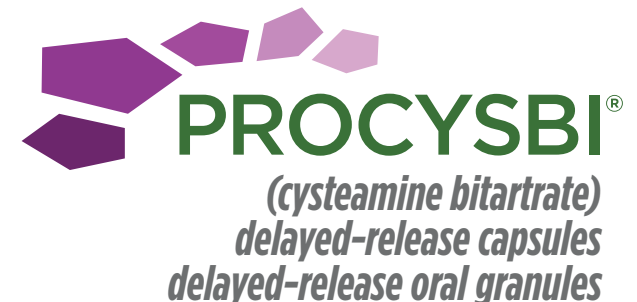
vomiting, nausea, stomach (abdominal) pain, pink eye, diarrhea, cold, tiredness, flu, headache, problems with body salts or electrolytes, infection of ear, nose or throat, joint pain.

These are not all the possible side effects of PROCYSBI. Call your doctor for medical information about side effects.

**You are encouraged to report negative side effects of prescription drugs to the FDA. Visit**

**<http://www.fda.gov/medwatch> or call 1-800-FDA-1088.**

**For additional important safety information, click here for the Patient Package Insert and discuss with your doctor.**



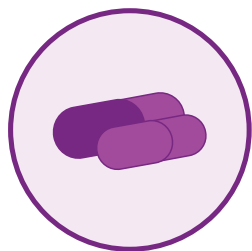
## HOW TO TAKE PROCYSBI

### HOW TO TAKE PROCYSBI BY MOUTH

PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is available in capsules and packets. **Capsules** may be swallowed whole with select liquids, or opened and the microbeads mixed with select foods or liquids. **Packets** should be opened and the microbeads mixed with select foods or liquids.

#### Important notes about taking PROCYSBI:

- Take PROCYSBI **the same way each time**
- Do not eat for at least **2 hours before** taking PROCYSBI and at least **30 minutes after** you take it
  - If you can't take PROCYSBI without eating, take it with no more than 1/2 cup (4 oz) of food **up to 1 hour before or after** you take PROCYSBI
- Take PROCYSBI at least **1 hour before or 1 hour after** you take medicines that contain bicarbonate or carbonate



Whole capsules

#### STEPS REQUIRED

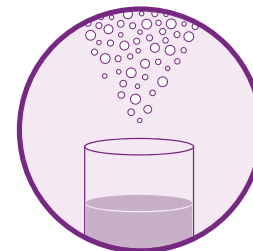
1. **Swallow** with water or fruit juice (**except grapefruit juice**). Do not crush or chew the capsules.



Mix microbeads  
with select foods

#### STEPS REQUIRED

1. **Open** the capsule(s) or packet(s). See instructions on reverse side.
2. Gently **mix** the microbeads with about 1/2 cup (4 oz) of applesauce or berry jelly. Do not crush or chew the microbeads. **Do not mix with any food other than applesauce or berry jelly.**
3. **Eat** the applesauce or berry jelly mixture within 30 minutes of mixing. Do not save the applesauce or berry jelly mixture for later use.



Mix microbeads  
with select liquids

#### STEPS REQUIRED

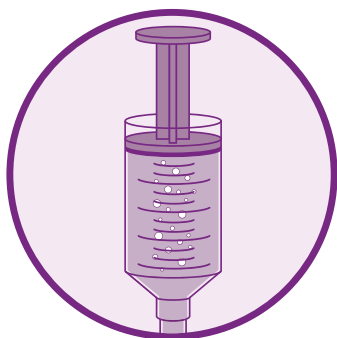
1. **Open** the capsule(s) or packet(s). See instructions on reverse side.
2. Gently **mix** the microbeads with about 1/2 cup (4 oz) of any fruit juice (**except grapefruit juice**). Do not crush or chew the microbeads.
3. **Drink all of the fruit juice mixture within 30 minutes of mixing.** Do not save the fruit juice mixture for later use.

Please see complete **IMPORTANT SAFETY INFORMATION** on pages 18 and 19, and [click here](#) for the Patient Package Insert.

## HOW TO TAKE PROCYSBI

### HOW TO TAKE PROCYSBI VIA G-TUBE

PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules may be given through a gastrostomy tube (G-tube) that is **size 14 French or larger** using a straight (bolus) feeding tube. **Capsules or packets** should be opened and the microbeads **mixed with strained applesauce (with no chunks)**.



#### To open PROCYSBI capsules:

- Use dry hands to hold each end of the capsule
- Gently twist the 2 ends in opposite directions to open
- Do not pinch the capsule in the center
- Do not crush or chew the capsule

#### To open PROCYSBI packets:

- Tear the packet open along the perforated line

#### STEPS REQUIRED

1. **Flush** the G-tube button with 5 mL of water to clear the button.
2. **Open** the PROCYSBI capsule(s) or packet(s). See instructions to the left.
3. In a clean container, **sprinkle** the microbeads on about 1/2 cup (4 oz) of strained applesauce with no chunks and mix gently.\*  
\*Children who weigh 55 lb (25 kg) or less can take PROCYSBI with at least 1/8 cup (1 oz) of applesauce.
4. **Place** the tip of a catheter tip syringe at the bottom of the container of the 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.
5. **Place** the tip of the catheter tip syringe into a feeding tube and fill it with the mixture.
6. Holding the feeding tube in a horizontal position, **push** the mixture through the G-tube at a quick and steady rate of 10 mL over 10 seconds. **Repeat** until all the mixture is given.
7. **Be sure to push all of the mixture through the G-tube within 30 minutes of mixing.** Do not save the mixture for later use.
8. **Draw up** at least 10 mL of fruit juice (**except grapefruit juice**) or water into another catheter tip syringe. Gently swirl the syringe. Flush the G-tube with the fruit juice or water until there is no mixture left in the G-tube.

Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.

## WORK WITH YOUR DOCTOR

# YOUR PROCYSBI TREATMENT: WORKING WITH YOUR DOCTOR AND HEALTHCARE TEAM

Before each appointment with the doctor and healthcare team, consider how you're feeling about your treatment with PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules. Fill out the information below, then write down any questions you'd like to ask.

### Notes related to treatment

Current weight: \_\_\_\_\_

Current dose: \_\_\_\_\_

I take PROCYSBI in: ☐ Capsules ☐ Packets

Recently I have missed a dose, skipped a dose, or taken a dose late: ☐ Yes ☐ No

Side effect(s) I am experiencing (if any): \_\_\_\_\_

On a scale of 1 to 5 (1 is not at all bothersome and 5 is extremely bothersome), I find these side effects to be at a level \_\_\_\_\_.

Date of my last cystine level test: \_\_\_\_\_

I receive the following cystine level test (check one):

☐ UCSD granulocytes test (target: less than 1.9 nmol ½ cystine/mg protein)

☐ Baylor Genetics mixed leukocytes test (target: less than 1.0 nmol ½ cystine/mg protein)

My cystine level at my last test: \_\_\_\_\_

Questions I'd like to ask my doctor at my next visit:

**Did you know? PROCYSBI comes in capsules *and* in tear-open packets. See the next page to learn more!**

**Please see complete IMPORTANT SAFETY INFORMATION on pages 18 and 19, and click here for the Patient Package Insert.**



## WORK WITH YOUR DOCTOR

### PROCYSBI IN CAPSULES AND PACKETS

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PROCYSBI® (cysteamine bitartrate) delayed-release capsules and delayed-release oral granules is available in capsules *and* in tear-open packets. **Both contain the same medicine that provides you with 12-hour dosing.**

**Capsules** may be a good option for people who prefer to swallow whole capsules.

**Packets** may be a good option for people who have trouble swallowing, take their medicine via a gastrostomy tube (G-tube), or take the time to open individual capsules at every dose.

#### Switching between capsules and packets

Fill out the information below and talk to your doctor if you're interested in making the switch.

**Administration type (check one):** ☐ Swallow capsules whole ☐ Sprinkle microbeads into foods or liquids ☐ Via G-tube

**On a scale of 1 to 5, rate your satisfaction with this method (1 is not satisfied and 5 is extremely satisfied):** \_\_\_\_\_

**If dissatisfied, explain why:**

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#### Here are some questions you might want to ask your doctor:

- Am I a good candidate for capsules or packets? Why?
- What would switching involve?
- What steps do I need to go through?

#### Who should not take PROCYSBI?

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

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