

Instructions for Use
PROCYSBI® (Pro-CIS-bee)
(cysteamine bitartrate) delayed-release capsules

PROCYSBI capsules should be swallowed whole. If you cannot swallow the capsule whole, you can open it and take the capsule contents with certain foods and juices. PROCYSBI can also be given through a gastrostomy tube (G-tube).

Opening PROCYSBI capsules:

- Do not pinch the PROCYSBI capsule in the center.
- Do not crush the PROCYSBI capsule.
- Use both hands to open the PROCYSBI capsule.
- Hold each end of the PROCYSBI capsule with your thumb and index (pointer) fingers and gently twist the two ends in opposite directions to open.

Taking PROCYSBI with applesauce or berry jelly:

Do not take PROCYSBI with any food other than applesauce or berry jelly.

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

Taking PROCYSBI with fruit juice:

Do not take PROCYSBI with grapefruit juice.

- Step 1: Pour about ½ cup (4 ounces) of fruit juice into a clean cup.
- Step 2: Open the PROCYSBI capsule. See “Opening PROCYSBI capsules” above. You may need to use more than 1 PROCYSBI capsule for the dose prescribed by your doctor.
- Step 3: Sprinkle the granules that are inside the capsule or capsules into ½ cup (4 ounces) of fruit juice.
- Step 4: Stir gently until mixed.
- Step 5: Drink all of the fruit juice and granules mixture within 30 minutes of mixing. Do not chew the granules. Do not save the fruit juice and granules for later use.

Giving PROCYSBI through a gastrostomy tube (G-tube):

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

For people who have a gastrostomy tube (G-tube) that is **size 14 French or larger**, PROCYSBI may be given as follows:

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 starting PROCYSBI at a dose of 1 or 2 capsules.
- Step 3: Open the PROCYSBI capsule. See “Opening PROCYSBI capsules” above. You may need to use more than 1 PROCYSBI capsule for the dose prescribed by your doctor.
- 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 granules 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 granules mixture.
- Step 7: Hold the feeding tube in a horizontal (straight across) position. Give the applesauce and granules 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 granules mixture is given. **Give all of the applesauce and granules mixture through the gastrostomy tube within 30 minutes of mixing.** Do not save the applesauce and granules mixture for later use.
- Step 9: Draw up at least 10 mL of fruit juice into another catheter tip syringe. **Do not use grapefruit juice.** Gently swirl the syringe. Flush the gastrostomy tube with the fruit juice. Use enough fruit juice to flush the gastrostomy tube so that there is no applesauce and granules mixture left in the gastrostomy tube.

How should I store PROCYSBI?

- Store PROCYSBI at room temperature, 20°C to 25°C (68°F to 77°F).
- Dispose of by date on bottle.
- Discard expired capsules per your local or state regulations.
- Store PROCYSBI in a dry place away from light.
- Keep PROCYSBI tightly closed in the original bottle.
- The PROCYSBI bottle (25 mg) contains a desiccant canister and one oxygen absorber canister.
- The PROCYSBI bottle (75 mg) contains a desiccant canister and two oxygen absorber canisters.
- Do not eat or throw away the desiccant canister or oxygen absorber canister(s).

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

This Instructions for Use has been approved by the U.S. Food and Drug Administration.

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

Issued 3/2017