

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

PROCYSBI is available as:

- Capsules in bottles (see “**Taking PROCYSBI Capsules**”)
- Oral granules in packets (see “**Taking PROCYSBI Oral Granules**”)

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

Taking PROCYSBI capsules

PROCYSBI capsules should be swallowed whole with fruit juice (except grapefruit juice) or water. If you cannot swallow the capsule whole, you can open each capsule and take the capsule contents with certain foods and juices, or the capsule contents can be given through a gastrostomy tube (G-tube).

Opening PROCYSBI capsules:

- Do not pinch the PROCYSBI capsule in the center.
- Do not crush or chew 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. If needed, use a smaller amount you can finish in one feeding. 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 onto the applesauce or berry jelly.
- Step 4: Mix the granules with the applesauce or berry jelly. Do not crush the granules.
- 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 all the granules that are inside the capsule into ½ cup (4 ounces) of fruit juice.
- Step 4: Stir gently until mixed. Do not crush the granules.
- 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 or water and granules mixture 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. Children who weigh 55 pounds (25 kilograms) or less can take PROCYSBI with at least 1/8 cup (1 ounce) of applesauce.
- 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 on the applesauce. Gently mix the granules with the applesauce. Do not crush the granules.
- 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 Step 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 or water into another catheter tip syringe. **Do not use grapefruit juice.** Gently swirl the syringe. Flush the gastrostomy tube with the fruit juice or water. Use enough fruit juice or water to flush the gastrostomy tube so that there is no applesauce and granules mixture left in the gastrostomy tube.

Taking PROCYSBI oral granules

You must mix the PROCYSBI oral granules in packets with certain foods and juices to take your PROCYSBI dose. PROCYSBI oral granules can also be given through a **gastrostomy tube (G-tube)**.

Opening PROCYSBI oral granules in packets:

- Tear open the packet along the dotted (perforated) line.
- Do not store granules in opened packets for your next dose.

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 packet of PROCYSBI oral granules. See “**Opening PROCYSBI oral granules in packets**” above. You may need to use more than 1 packet of PROCYSBI oral granules for the dose prescribed by your doctor.
- Step 3: Sprinkle all the granules that are inside the packet onto the applesauce or berry jelly.
- Step 4: Mix the granules with the applesauce or berry jelly. Do not crush the granules.
- 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 packet. See “**Opening PROCYSBI oral granules in packets**” above. You may need to use more than 1 packet of PROCYSBI oral granules for the dose prescribed by your doctor.
- Step 3: Sprinkle all the granules that are inside the packet into ½ cup (4 ounces) of fruit juice.
- Step 4: Stir gently until mixed. Do not crush the granules.
- 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. Children who weigh 55 pounds (25 kilograms) or less can take PROCYSBI with at least 1/8 cup (1 ounce) of applesauce.
- Step 3: Open the packet of PROCYSBI oral granules. See “**Opening PROCYSBI oral granules in packets**” above. You may need to use more than 1 packet of PROCYSBI oral granules for the dose prescribed by your doctor.
- Step 4: Sprinkle all the granules that are inside the packet on the applesauce. Gently mix the granules with the applesauce. Do not crush the granules.
- 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 packets.
- 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 Step 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 capsules and PROCYSBI oral granules at room temperature between 68°F to 77°F (20°C to 25°C).
- Keep PROCYSBI in the original packaging. Do not put PROCYSBI in another container (repackage).
- Keep PROCYSBI in a dry place to protect from moisture.
- The PROCYSBI bottle contains a desiccant to help keep the medicine dry (protect it from moisture). Do not remove the desiccant from the bottle.
- Protect from light.
- Do not store opened packets of PROCYSBI oral granules for later use.
- Throw away (dispose of) any unused PROCYSBI by the “discard after” date on the bottle or carton.

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.

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