

Directions for taking RAVICTI® (glycerol phenylbutyrate) Oral Liquid by mouth

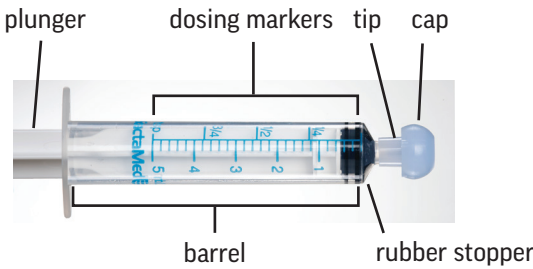
Items in your RAVICTI shipment



Bottle of RAVICTI



Reclosable bottle cap adapter



Oral syringe

Attention: If you can swallow liquids, it is recommended you take RAVICTI by mouth.



1 Open the bottle of RAVICTI. To open, push down on the cap while twisting it to the left.



2 Twist a new reclosable bottle cap adapter onto the bottle. You do this by turning it to the right. Make sure it is tightly on the bottle.



3 Place the tip of a new dry oral syringe into the reclosable bottle cap adapter.



4 Turn the bottle upside down with the oral syringe still inserted.

IMPORTANT TO REMEMBER:

- Use a new reclosable bottle cap adapter with each new bottle of RAVICTI that is opened.
- Use a new dry oral syringe to withdraw each prescribed dose of RAVICTI. Discard the oral syringe after each use.

USE and IMPORTANT SAFETY INFORMATION

What is the most important information I should know about RAVICTI?

RAVICTI may cause serious side effects, including:

Nervous system side effects (Neurotoxicity) Phenylacetate (PAA), a breakdown product of RAVICTI, may cause nervous system side effects. Call your doctor or get medical help right away if you have any of these symptoms while taking RAVICTI:

- sleepiness
- lightheadedness
- change in taste
- problems with hearing
- confusion
- problems with memory
- worsening of numbness, tingling, or burning in your hands or feet
- headache
- feeling very tired (fatigue)
- nausea
- vomiting

Your doctor may do blood tests to measure the amount of PAA in your blood during your treatment with RAVICTI.

Please see additional Important Safety Information for RAVICTI on page 3 and the Medication Guide available at RAVICTI.com.

RAVICTI[®]
(glycerol phenylbutyrate) Oral Liquid



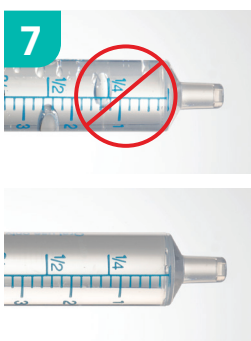
Withdraw the prescribed dose of RAVICTI with the dry oral syringe by pulling the plunger back. Pull until the first line of the black rubber stopper is even with the dosing mark. The dosing marks are on the barrel of the oral syringe. Match the dosing mark to the amount of medicine prescribed by your doctor.



Turn the bottle back to its upright position.

EXAMPLE: If your prescribed dose is 3 mL, the first line of the rubber stopper should be even with the 3-mL dosing mark on the barrel of the oral syringe.

NOTE: Pull the plunger slowly. This will help keep large air bubbles from forming in the medicine.



If there are large air bubbles, push the medicine back into the bottle. Do this slowly. Draw up the medicine again (repeat steps 3 through 6).



Place the oral syringe into the mouth. Push the plunger to squirt the medicine into the mouth. Make sure all of the medicine is pushed out of the oral syringe. Discard the oral syringe.



After taking RAVICTI (glycerol phenylbutyrate) Oral Liquid, it is suggested to drink liquid or eat some food to help make sure all of the medicine is swallowed.



Tightly close the tethered tab on the reclosable bottle cap adapter after each use. Do not rinse the reclosable bottle cap adapter.

NOTE: If RAVICTI is in contact with water, it will become cloudy in appearance. If the contents are in contact with water or cloudy, then do not use the remaining RAVICTI and contact the prescribing physician or pharmacy.

Discard the RAVICTI bottle and any remaining contents 28 days after opening.

IMPORTANT TO REMEMBER:

- Take the exact amount of RAVICTI (glycerol phenylbutyrate) Oral Liquid as instructed by your doctor.
- Your doctor may change your dose if needed.
- Take RAVICTI with food or formula.
- In an infant who is breastfeeding, give RAVICTI just prior to breastfeeding.
- Use a new reclosable bottle cap adapter with each new bottle of RAVICTI that is opened.
- Use a new dry oral syringe to withdraw each prescribed dose of RAVICTI.
- If RAVICTI is in contact with water, it will become cloudy in appearance. If the contents are in contact with water or cloudy, then do not use the remaining RAVICTI and contact your prescribing doctor or pharmacy.
- Discard bottle and any remaining contents 28 days after opening.

- Ask your pharmacist for oral syringes and a reclosable bottle cap adapter for each bottle you receive if you do not have them.

If you take too much RAVICTI, call your doctor or the Poison Control Center at 1-800-222-1222 or go to the nearest hospital emergency room right away.

Do not stop taking RAVICTI unless your doctor tells you to stop.

- **If you miss a dose of RAVICTI, take it as soon as possible.**
- **Do not take 2 doses of RAVICTI at the same time.**
- **If you miss more than 1 dose and are not feeling well, call your doctor or nurse right away.**

If you still have questions about how to take RAVICTI with an oral syringe, please contact your doctor for more information.

Please see additional Important Safety Information for RAVICTI on page 3 and the Medication Guide available at RAVICTI.com.

RAVICTI[®]
(glycerol phenylbutyrate) Oral Liquid

USE and IMPORTANT SAFETY INFORMATION (continued)

What is RAVICTI?

- RAVICTI (glycerol phenylbutyrate) Oral Liquid is a prescription medicine used for long-term management of high blood levels of ammonia (hyperammonemia) caused by a condition called a urea cycle disorder (UCD). RAVICTI should be used if the UCD cannot be managed with a low-protein diet and dietary supplements alone. RAVICTI must be used along with a low-protein diet and in some cases dietary supplements.
- RAVICTI is not used for the acute treatment of hyperammonemia in people with UCD.
- It is not known if RAVICTI is safe and effective for the treatment of N-acetylglutamate synthase (NAGS) deficiency.

Do not take RAVICTI if you are allergic to phenylbutyrate. Call your doctor or go to the nearest hospital emergency room if you have wheezing, shortness of breath, cough, low blood pressure, flushing, nausea or a rash while taking RAVICTI.

Before taking RAVICTI, tell your doctor about all of your medical conditions, including if you:

- have liver or kidney problems.
- have pancreas or bowel (intestine) problems
- are pregnant or plan to become pregnant. It is not known if RAVICTI will harm your unborn baby. If you become pregnant during treatment with RAVICTI, call Horizon at 1-866-479-6742 to report the pregnancy.
- are breastfeeding or plan to breastfeed. It is not known if RAVICTI passes into your breast milk. Breastfeeding is not recommended during treatment with RAVICTI. Talk to your doctor about the best way to feed your baby if you take RAVICTI.

What are possible side effects of RAVICTI?

RAVICTI may cause serious side effects, including:

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

The most common side effects of RAVICTI in adults include:

- diarrhea
- gas
- headache
- abdomen (stomach) pain
- vomiting
- tiredness
- decreased appetite
- indigestion or heartburn

The most common side effects of RAVICTI in children 2 years to 17 years of age include:

- upper abdomen (stomach) pain
- rash
- nausea
- vomiting
- diarrhea
- decreased appetite
- headache

The most common side effects of RAVICTI in children 2 months to less than 2 years of age include:

- low white blood cell count (neutropenia)
- vomiting
- constipation
- diarrhea
- fever
- reduced food intake
- cough
- stuffy nose
- runny nose
- skin rash
- small round bumps on the skin

The most common side effects of RAVICTI in children less than 2 months of age include:

- vomiting
- rash
- gastroesophageal reflux
- increased levels of liver enzymes in the blood
- decreased appetite and reduced food intake
- low red blood cell count (anemia)
- cough
- loss of too much body fluid (dehydration)
- too much acid in the blood (acidosis)
- high blood platelet count (thrombocytosis)
- low blood platelet count (thrombocytopenia)
- low blood neutrophil count (type of white blood cell) (neutropenia)
- high white blood cell count (lymphocytosis)
- diarrhea
- gas
- constipation
- fever
- drowsiness (lethargy)
- irritability
- agitation

These are not all of the possible side effects of RAVICTI. Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

For additional Important Safety Information, see the accompanying Medication Guide available at [RAVICTI.com](https://www.ravicti.com) and discuss with your doctor.