

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

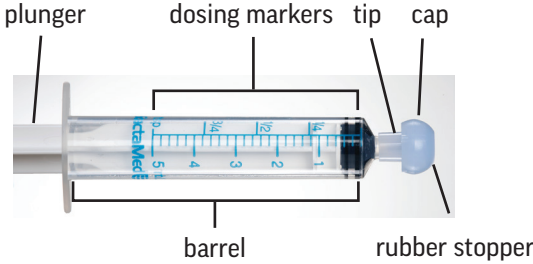
Items in your RAVICTI shipment



Bottle of RAVICTI



Blue AdaptaCap®
Bottle Adapter



Oral dosing 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 the blue AdaptaCap Bottle 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 the oral dosing syringe into the AdaptaCap Bottle Adapter.



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

APPROVED USES and IMPORTANT SAFETY INFORMATION for RAVICTI

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 to treat extremely high levels of ammonia in the blood (hyperammonemic crisis) in people with UCDs.

It is not known if RAVICTI is safe and effective for the treatment of N-acetylglutamate synthase (NAGS) deficiency.

IMPORTANT SAFETY INFORMATION

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

RAVICTI may cause serious side effects, including:

Nervous system side effects (Neurotoxicity). The breakdown of RAVICTI produces the byproduct phenylacetate (PAA), which 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

Please see additional Important Safety Information for RAVICTI on page 3.


RAVICTI
(glycerol phenylbutyrate) Oral Liquid



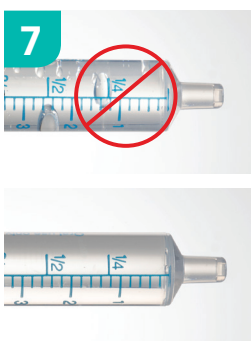
5 Get the medicine into the oral dosing syringe by pulling the plunger back. Pull until the first line of the black rubber stopper is even with the dosing mark. The dosing marks are on the barrel of the oral dosing syringe. Match the dosing mark to the amount of medicine prescribed by your doctor.



6 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 dosing syringe.

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



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



8 Place the oral dosing 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 dosing syringe. Put the cap on the oral dosing syringe.



9 After taking RAVICTI, it is suggested to drink liquid or eat some food to help make sure all of the medicine is swallowed.



10 Close the tab on the AdaptaCap Bottle Adapter. Make sure it is closed tightly.

IMPORTANT TO REMEMBER:

- Take the exact amount of RAVICTI as instructed by your doctor.
- RAVICTI is taken 3 or more times per day in equally divided doses and should be taken with food or formula.
- In an infant who is breastfeeding, give RAVICTI just prior to breastfeeding.
- When dosing RAVICTI, be sure to use the provided oral dosing syringe.

NOTE: You can use 1 oral dosing syringe and 1 blue AdaptaCap Bottle Adapter for all doses each day.

- You can also choose to use a new oral dosing syringe with each dose.
- Do not rinse the AdaptaCap Bottle Adapter or oral dosing syringe between doses.
- After the last dose of the day, dispose of the oral dosing syringe.

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 dosing syringe, please contact your doctor for more information.

IMPORTANT SAFETY INFORMATION (cont.)

Who should not take RAVICTI?

Do not take RAVICTI if you are allergic to phenylbutyrate. Call your doctor or go to the nearest hospital emergency room if you experience any of the following symptoms of an allergic reaction while taking RAVICTI:

- wheezing
- shortness of breath
- cough
- low blood pressure
- flushing
- nausea
- skin rash

What should I tell my doctor before taking RAVICTI?

Tell your doctor about any medical conditions and 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.
 - o **Pregnancy Registry:** There is a Pregnancy Registry for women who take RAVICTI just before becoming pregnant or who become pregnant during treatment with RAVICTI. The purpose of this registry is to collect information about the health of you and your baby. Talk to your doctor about how you can join the Pregnancy Registry. For more information about this registry, call 1-855-823-2595 or visit www.ucdregistry.com.
- 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
- 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.

For additional Important Safety Information, please see the Full Prescribing Information and Medication Guide available at RAVICTI.com and discuss with your doctor.

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.