

PATIENT MEDICATION INFORMATION

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

RAVICTI®

Glycerol Phenylbutyrate Oral Liquid

Read this carefully before you start taking **RAVICTI** and each time you get a refill. This leaflet is a summary and will not tell you everything about this drug. Talk to your healthcare professional about your medical condition and treatment and ask if there is any new information about **RAVICTI**.

What is RAVICTI used for?

- RAVICTI (rah-VIK-tee) is a prescription medicine used for long-term management of high blood levels of ammonia (hyperammonemia) caused by a condition called 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 should only be prescribed by a healthcare professional experienced in the treatment of UCDs.
- RAVICTI is not to be used to treat acute (severe) high blood levels of ammonia in patients with UCDs.
- It is not known if RAVICTI is safe and effective for the treatment of *N*-acetylglutamate synthase (NAGS) deficiency.

How does RAVICTI work?

Patients with UCD are unable to get rid of ammonia that is normally produced in the body. RAVICTI works by helping the body to remove excess ammonia.

What are the ingredients in RAVICTI?

Medicinal ingredients: glycerol phenylbutyrate

Non-medicinal ingredients: none

RAVICTI comes in the following dosage forms:

Oral liquid, 1.1 g/mL

Do not use RAVICTI if:

- You are experiencing acute hyperammonemia.
- You are allergic to glycerol phenylbutyrate, phenylbutyric acid (PBA), phenylacetic acid (PAA), and/or phenylacetylglutamine (PAGN), and/or any component of the container
- You are breastfeeding.

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take RAVICTI. Talk about any health conditions or problems you may have, including if you:

- have liver or kidney problems
- have heart 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.

While taking RAVICTI it is still possible to develop an acute episode of excess ammonia in your blood. This is a medical emergency, and medical assistance should be sought immediately. Symptoms may include nausea, vomiting, confusion, combativeness, slurred speech, difficulty walking, and even loss of consciousness. An infection can cause an episode of excess ammonia; therefore, if you develop a fever you should seek prompt medical assistance.

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

The following medicines may change the effect of RAVICTI and you may need more frequent blood tests:

- Midazolam, corticosteroids, barbiturates, topiramate, carbamazepine, and some immunosuppressive and anti-cancer drugs.
- Probenecid: May interfere with the removal of RAVICTI from the body.
- Corticosteroids: Use of corticosteroids may cause the breakdown of body protein and increase ammonia levels in your blood.
- Valproic Acid and Haloperidol: May cause high blood ammonia.

How to take RAVICTI:

- Take RAVICTI exactly as your healthcare professional tells you. Check with your healthcare professional if you are not sure.
- Your healthcare professional will tell you how much RAVICTI to take and when to take it.
- Your healthcare professional may change your dose if needed.
- RAVICTI is an oral liquid that is taken by mouth using an oral syringe. Ask your healthcare professional for an oral syringe if you do not have one.
- Take RAVICTI with food or formula.
- Stay on the diet that your healthcare professional gives you.
- For infants: In an infant who is breastfeeding, give RAVICTI just before breastfeeding.

Patients are to use the RAVICTI bottle and oral syringe as follows:

- Use a new reclosable bottle cap adapter with each new bottle that is opened.
- Open the RAVICTI bottle and twist on the new reclosable bottle cap adapter.
- Use a new and dry oral syringe to withdraw each dose of RAVICTI.
- Throw away the oral syringe after each dose.
- Tightly close the tethered tab on the reclosable bottle cap adapter after each use.
- Do not rinse the reclosable bottle cap adapter.

If water or moisture enters the RAVICTI bottle, the contents will become cloudy. If the contents of the bottle look cloudy at any time, do not use the remaining RAVICTI in the bottle. Return it to the pharmacy or patient program to be thrown away.

For people who have a nasogastric or gastrostomy tube in place, RAVICTI should be given as follows:

- It is recommended that all patients who can swallow take RAVICTI orally, even those with nasogastric and/or gastrostomy tubes.
- For patients who cannot swallow, a nasogastric or gastrostomy tube can be used to give RAVICTI as follows:

- Use a new dry oral syringe to withdraw each prescribed dose of RAVICTI from the bottle.
- Place the tip of the syringe into the tip of the nasogastric or gastrostomy tube. Push the plunger of the syringe to give RAVICTI into the tube.
- Use a separate syringe to flush the tube. Add 10 mL of water or formula to the syringe and push the plunger of the syringe to flush any remaining medicine from the tube into the stomach.
- If needed, flush the tube again with 10 mL of water or formula to clear the tube.

Usual dose:

The daily dose of RAVICTI will be based on your body surface area. The dose should be adjusted based on your protein tolerance and diet.

- The daily dose range of RAVICTI is 4.5 – 11.2 mL/m²/day.
- Patients 2 years of age and older: The total daily dose should be divided into 3 equal amounts and given with each meal or feeding. Each dose should be rounded up to the nearest 0.5 mL.
Patients less than 2 years: The total daily dose should be divided into 3 or more equal amounts and given with each meal or feeding. Each dose should be rounded up to the nearest 0.1 mL.
- You will need regular blood tests to determine the correct daily dose.

Overdose:

If you think you, or a person you are caring for, have taken too much RAVICTI, contact your healthcare professional, hospital emergency department, or regional Poison Control Centre immediately, even if there are no symptoms.

Missed Dose:

If you missed a dose of this medication, take it as soon as you remember. But if it is almost time for your next dose, skip the missed dose and continue with your next scheduled dose. Go back to the regular dosing schedule. Do not take two doses at the same time.

What are possible side effects from using RAVICTI?

These are not all the possible side effects you may feel when taking RAVICTI. If you experience any side effects not listed here, contact your healthcare professional.

- Diarrhea, gas, stomach pain and discomfort, constipation, indigestion, heart burn
- Decreased appetite
- Vomiting, nausea
- Fatigue
- Headache
- Fever
- Dizziness
- Muscle or body shaking
- Irregular menstrual bleeding
- Skin odor
- Acne
- Cough

- Dehydration
- Irritability, anxiety

RAVICTI can cause abnormal blood test results. Your healthcare professional will decide when to perform blood tests.

Serious side effects and what to do about them			
Symptom / effect	Talk to your healthcare professional		Stop taking drug and get immediate medical help
	Only if severe	In all cases	
VERY COMMON			
Acidosis (high level of acid in the blood): Weight loss, fatigue, malaise, abdominal pain, unusual muscle pain, feeling dizzy or lightheaded, fast or irregular heartbeat, shortness of breath, feeling unusually cold, especially in arms and legs,		√	
Blood disorders: bruising or bleeding for longer than usual if you hurt yourself, fatigue and weakness, low energy, infections, fever, aches, pains and flu-like symptoms, irregular heartbeats, pale complexion, shortness of breath		√	
Urinary tract infection (infection in urinary system including kidneys, ureters, bladder and urethra): Pain or burning sensation while urinating, frequent urination, blood in urine, pain in the pelvis, strong smelling urine, cloudy urine		√	
RARE			
Allergic Reaction: rash, hives, swelling of the face, lips, tongue or throat, difficulty swallowing or breathing			√
Neurotoxicity (nervous system side effects): Sleepiness, weakness, lightheadedness, change in taste, problems with hearing, confusion, problems with memory, worsening neuropathy (numbness, tingling, or burning in your hands or feet), headache			√

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, talk to your healthcare professional.

Reporting Side Effects

You can report any suspected side effects associated with the use of health products to Health Canada by:

- Visiting the Web page on Adverse Reaction Reporting (<https://www.canada.ca/en/health-canada/services/drugs-health-products/medeffect-canada.html>) for information on how to report online, by mail or by fax; or
- Calling toll-free at 1-866-234-2345.

NOTE: Contact your health professional if you need information about how to manage your side effects. The Canada Vigilance Program does not provide medical advice.

Storage:

Store RAVICTI between 15-30°C.

Keep in original packaging to protect from light.

Use the contents of the bottle within 90 days after opening.

Keep out of reach and sight of children.

If you want more information about RAVICTI:

- Talk to your healthcare professional
- Find the full product monograph that is prepared for healthcare professionals and includes this Patient Medication Information by visiting the Health Canada website (<https://www.canada.ca/en/health-canada/services/drugs-health-products/drug-products/drug-product-database.html>); the manufacturer's website www.amgen.ca, or by calling 1-866-502- 6436.

This leaflet was prepared by Amgen Canada for Horizon Therapeutics Ireland DAC

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