

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

PATIENT MEDICATION INFORMATION

Pr **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 in adults and children 2 years of age and older 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.

RAVICTI is not to be used in children less than 2 months of age. It is not known if **RAVICTI** is safe and effective in children between the ages of 2 months and 2 years.

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:

- Children are less than 2 months of age.
- You are experiencing acute hyperammonemia.

- You are allergic to glycerol phenylbutyrate, phenylbutyric acid (PBA), phenylacetic acid (PAA), and/or phenylacetylglutamine (PAGN).
- 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 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:

Usual dose:

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

- The daily dose range of RAVICTI is 4.5 – 11.2 mL/m²/day.
- The total daily dose should be divided into equal amounts and given with each meal or feeding.
- Each dose should be rounded up to the nearest 0.5 mL.
- RAVICTI should be taken by mouth using an oral syringe that is provided to you by your pharmacist.

- You will need regular blood tests to determine the correct daily dose.
- Take RAVICTI exactly as your doctor tells you.
- Stay on the diet that your doctor gives you.

For people who have a nasogastric or gastric 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 gastric tubes.
- For patients who cannot swallow, a nasogastric or gastric tube can be used to administer RAVICTI as follows:
 - Use an oral syringe to take the prescribed dose of RAVICTI from the bottle.
 - Place the tip of the syringe into the tip of the nasogastric or gastric tube and administer RAVICTI into the tube.
 - Flush the nasogastric or gastric tube with at least 10 mL of water or formula.

Overdose:

If you think you 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.

The most common side effects include diarrhea, gas, headache, decreased appetite, vomiting, nausea, fatigue and skin odor.

Other side effects that may occur include:

- Stomach pain and discomfort, constipation, indigestion
- Dizziness
- Tremor
- Irregular menstrual bleeding
- Acne

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		Get immediate medical help
	Only if severe	In all cases	
RARE 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			√
Allergic Reaction: rash, hives, swelling of the face, lips, tongue or throat, difficulty swallowing or breathing			√

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 help improve the safe use of health products for Canadians by reporting serious and unexpected side effects to Health Canada. Your report may help to identify new side effects and change the product safety information.

3 ways to report:

- Online at [MedEffect](#);
- By calling 1-866-234-2345 (toll-free);
- By completing a Consumer Side Effect Reporting Form and sending it by:
 - Fax to 1-866-678-6789 (toll-free), or
 - Mail to: Canada Vigilance Program
Health Canada, Postal Locator 0701E
Ottawa, ON
K1A 0K9

Postage paid labels and the Consumer Side Effect Reporting Form are available at [MedEffect](#).

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 \(http://hc-sc.gc.ca/index-eng.php\)](http://hc-sc.gc.ca/index-eng.php); the manufacturer's website <http://www.RAVICTI.CA>, or by calling 1-855-823-7878.

Talk to your doctor about participating in a UCD registry. The purpose of this registry is to collect information about people with UCD to improve care.

This leaflet was prepared by Horizon Pharma Ireland Ltd.

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