OLPRUVA™
(sodium phenylbutyrate)
for oral suspension

Welcome to OLPRUVA™ (sodium phenylbutyrate) for oral suspension

Within this patient guide you will find information to help you start your OLPRUVA journey—from how it works in the body to tracking daily doses, and more.

OLPRUVA is a prescription medicine used along with certain therapy, including changes in diet, for the long-term management of adults and children weighing 44 pounds (20 kg) or greater and with a body surface area (BSA) of 1.2 m^2 or greater, with urea cycle disorders (UCDs), involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC) or argininosuccinic acid synthetase (AS).

Episodes of rapid increase of ammonia in the blood (acute hyperammonemia) may happen in people during treatment with OLPRUVA. OLPRUVA is not for the treatment of acute hyperammonemia, which can be life-threatening and requires emergency medical treatment.

Please see additional Important Safety Information throughout and in full on pages 12-14 and full Prescribing Information and Patient Information, including Instructions for Use.

Convenient single-dose envelopes provide people with UCDs

AMMONIA CONTROL *ON THE GO*

Each single-dose envelope contains the premeasured OLPRUVA packet(s) and Mix-Aid packet needed to mix 1 dose.

Important Safety Information

OLPRUVA is not approved in children weighing less than 44 pounds (20 kg) or in children weighing 44 pounds (20 kg) or greater with a BSA of less than 1.2 m^2.

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OLPRUVA™ (sodium phenylbutyrate) for oral suspension is:

- **Palatable**—Dual-coating* formulation designed for palatability
- **Premeasured**—Convenient individual dose packets support dosing accuracy
- **Portable**—Discreet single-dose envelopes are easily carried when you or your loved one is on the go

*The active ingredient of OLPRUVA is covered by a seal coating and an outer polymer coating.

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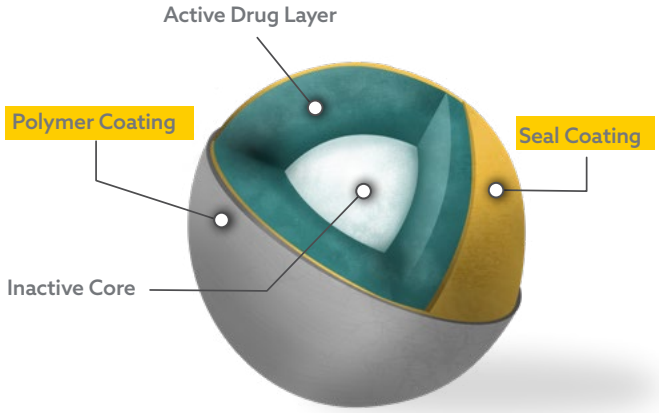
Unique formulation of sodium phenylbutyrate

Features a polymer coating designed for palatability

- Beneath the polymer coating, an additional layer of seal coating covers the active drug
- Patented dual-coating formulation should be taken within a 5-minute window to minimize dissolving of the polymer coating

OLPRUVA™ (sodium phenylbutyrate) for oral suspension

Unique, Dual-Coating Formulation



Important Safety Information

Before taking OLPRUVA, tell your or your child's healthcare provider about all your medical conditions, including if you: • have heart problems • have kidney or liver problems • are pregnant or plan to become pregnant. It is not known if OLPRUVA will harm your unborn baby. If you become pregnant during treatment with OLPRUVA, call Acer Therapeutics Inc. at 1-833-657-7882 to report the pregnancy. • are breastfeeding or plan to breastfeed. It is not known if OLPRUVA passes into your breast milk. Talk to your doctor about the best way to feed your baby if you take OLPRUVA.

OLPRUVA is designed with you or your loved one in mind

The active ingredient of OLPRUVA is sodium phenylbutyrate. Sodium phenylbutyrate powder was approved for treating UCDs in 1996 and has been foundational treatment for UCDs for over 20 years.

Therefore, the US Food and Drug Administration (FDA) approved OLPRUVA:

- Because it provides an equivalent amount of medicine in the body as sodium phenylbutyrate powder
- Based on FDA evaluation of the safety and effectiveness of sodium phenylbutyrate powder

How does OLPRUVA work?

In people with UCDs, there is a problem with the enzymes that help remove harmful ammonia from the body. **OLPRUVA helps the body get rid of excess nitrogen to help avoid the dangerous buildup of ammonia.**

What is the appropriate dose of OLPRUVA?

Your doctor will determine your or your loved one's dose based on weight and body surface area. Always be sure to prepare and take or give OLPRUVA exactly as prescribed by your doctor.

Ongoing support from NAVIGATOR by acertherapeutics

Your OLPRUVA Navigator Team is available for questions about your medication or financial support programs Monday-Friday, 7 AM-7 PM Central Time, at **1-833-OLPRUVA** (1-833-657-7882)

They will contact you before each refill date to help you avoid running out of medication

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How is OLPRUVA™ (sodium phenylbutyrate) for oral suspension prepared and taken?

For complete administration information, see the Instructions for Use in the Patient Information at OLPRUVA.com.

Here's what you will need to prepare and take or give 1 dose of OLPRUVA:

- 1 dosage envelope containing OLPRUVA and Mix-Aid* packets. The contents of 1 envelope equals 1 full dose
- An open drinking cup
- A spoon
- 8 ounces of water (4 ounces for mixing the OLPRUVA dose and 4 ounces for a "rinse" to ensure you get the full dose)

*Mix-Aid (100% modified corn starch) is mixed with water to thicken the water and create a "suspension" for administration of the medication.

STEP 1

Get an open drinking cup and spoon.

STEP 2

Add about 4 ounces of water to an open drinking cup.

STEP 3

Remove 1 dosage envelope from the kit.

Note: Each kit is divided into 30 individual sections. Each section contains 3 dosage envelopes, for a total of 90 dosage envelopes.

STEP 4

Open the dosage envelope and remove all packets (1 packet of Mix-Aid and 1 or 2 packets of OLPRUVA).

Note: All packets in the dosage envelope must be used for 1 full dose.

Important Safety Information

Tell your healthcare provider about all the medicines you or your child take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

STEP 5

To open the Mix-Aid packet, tear or cut with scissors, straight across from the notch.

STEP 6

Add the full contents of the Mix-Aid packet to the water (in the open drinking cup) and stir with the spoon for at least 30 seconds.

Note: The contents will not dissolve but will make the water thicker so that OLPRUVA does not sink to the bottom of the cup.

Do not drink yet.

STEP 7

To open the OLPRUVA packet(s), fold at the notch and tear or cut with scissors. Add the full contents of the OLPRUVA packet(s) to the open drinking cup containing the mixture from Step 6 (the water and Mix-Aid) and stir for 15 seconds.

Drink the entire contents of the open drinking cup within 5 minutes to help prevent the coating from dissolving.

Note: The entire contents of each packet must be used for 1 full dose.

STEP 8

To make sure that you get the full dose, pour another 4 ounces of water in the open drinking cup, stir, and drink the entire contents.

Do not take or give OLPRUVA suspension in a gastrostomy or nasogastric tube.

The mixed OLPRUVA suspension should be thrown away (discarded) after 30 minutes if not used right away.

Take your OLPRUVA dose with food. If you miss a dose of OLPRUVA, take the missed dose as soon as possible that same day.

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Dosing tracker

Keeping track of treatment

Keeping track of your or your loved one's treatment can be helpful to ensure you or they are taking OLPRUVA as prescribed. Use your smartphone or other device to set reminders, or use another app that helps you set a schedule. If you prefer to track your or your loved one's treatment by writing it down, use the tracker below to help you document a day's worth of OLPRUVA treatment. You can also make copies of this tracker.

Date:

Dose 1 OLPRUVA is taken 3-6 times a day with food	Time: Notes:
Dose 2	Time: Notes:
Dose 3	Time: Notes:

Important Safety Information

Know the medicines you take. Keep a list of them to show your or your child's healthcare provider and pharmacist when you get a new medicine. **Keep OLPRUVA and all medicines out of the reach of children**.

Date:

Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:

Dosing tracker continued on page 10.

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Dosing tracker

Date:	
Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:

Date:	
Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:
Dose ____	Time: Notes:

Important Safety Information

The most common side effects of OLPRUVA include absent or irregular menstrual periods, decreased appetite, body odor, and bad taste or avoiding foods that you ate prior to getting sick (taste aversion).

Please see additional Important Safety Information throughout and in full on pages 12-14 and full Prescribing Information and Patient Information, including Instructions for Use.

OLPRUVA™
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Summary of Important Safety Information for OLPRUVA
OLPRUVA [ol proo vah] (sodium phenylbutyrate) for oral suspension

This summary does not include all information about OLPRUVA and is not meant to take the place of discussions with your healthcare provider about your or your child's treatment. Please read this important information carefully and discuss any questions about OLPRUVA with your healthcare provider.

What is OLPRUVA?

- OLPRUVA is a prescription medicine used along with certain therapy, including changes in diet, for the long-term management of adults and children weighing 44 pounds (20 kg) or greater and with a body surface area (BSA) of 1.2 m^2 or greater, with urea cycle disorders (UCDs), involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC) or argininosuccinic acid synthetase (AS).
- Episodes of rapid increase of ammonia in the blood (acute hyperammonemia) may happen in people during treatment with OLPRUVA. OLPRUVA is not for the treatment of acute hyperammonemia, which can be life-threatening and requires emergency medical treatment.
- OLPRUVA is not approved in children weighing less than 44 pounds (20 kg) or in children weighing 44 pounds (20 kg) or greater with a BSA of less than 1.2 m^2.

Before taking OLPRUVA, tell your or your child's healthcare provider about all your medical conditions, including if you:

- have heart problems
- have kidney or liver problems
- are pregnant or plan to become pregnant. It is not known if OLPRUVA will harm your unborn baby. If you become pregnant during treatment with OLPRUVA, call Acer Therapeutics Inc. at 1-833-657-7882 to report the pregnancy.
- are breastfeeding or plan to breastfeed. It is not known if OLPRUVA passes into your breast milk. Talk to your doctor about the best way to feed your baby if you take OLPRUVA.

Tell your healthcare provider about all the medicines you or your child take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

Certain medicines may increase the level of ammonia in your blood or cause serious side effects when taken during treatment with OLPRUVA. Especially tell your healthcare provider if you or your child take:

- corticosteroids
- valproic acid
- haloperidol
- probenecid

Know the medicines you take. Keep a list of them to show your or your child's healthcare provider and pharmacist when you get a new medicine. **Keep OLPRUVA and all medicines out of the reach of children**.

How should I or my child take OLPRUVA?

Read the detailed Instructions for Use that comes with OLPRUVA for information about the right way to prepare and take a dose of OLPRUVA.

- Take OLPRUVA exactly as prescribed by your healthcare provider.
- Your healthcare provider may change your dose if needed. Do not change your dose unless your healthcare provider tells you to.
- Your healthcare provider will prescribe OLPRUVA based on your or your child's weight.
- Take your OLPRUVA dose with food.
- If you miss a dose of OLPRUVA, take it as soon as possible that same day.
- **Do not** give or take OLPRUVA through a gastrostomy or nasogastric tube.
- If you take too much OLPRUVA, call your healthcare provider or go to the nearest hospital emergency room right away.

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Summary of Important Safety Information (continued)

What are the possible side effects of OLPRUVA?

OLPRUVA can cause serious side effects, including:

- **Nervous system problems (neurotoxicity).** Call your healthcare provider right away if you or your child get any of the following symptoms during treatment with OLPRUVA:
  - sleepiness
  - tiredness
  - lightheadedness
  - vomiting
  - nausea
  - headache
  - confusion
- **Low potassium levels in your blood (hypokalemia).** Your healthcare provider will monitor your blood potassium levels during treatment with OLPRUVA and treat if needed.
- **Conditions related to swelling (edema).** OLPRUVA contains salt (sodium), which can cause swelling from salt and water retention. Your healthcare provider will decide if OLPRUVA is right for you if you have certain medical conditions that cause edema, such as heart failure, liver problems or kidney problems.

The most common side effects of OLPRUVA include:

- absent or irregular menstrual periods
- decreased appetite
- body odor
- bad taste or avoiding foods that you ate prior to getting sick (taste aversion)

Your healthcare provider may do certain blood tests to check you or your child for side effects during treatment with OLPRUVA.

These are not all of the possible side effects of OLPRUVA. Call your doctor for medical advice about side effects. You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/MedWatch or call 1-800-FDA-1088.

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For more information, support, and resources,
visit OLPRUVA.com

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Sources: 1. OLPRUVA™ (sodium phenylbutyrate) for oral suspension. Prescribing information. Newton, MA: Acer Therapeutics Inc. **2.** Appel LE, Shockey JR, Schelling DC, inventors; Acer Therapeutics Inc, assignee. Palatable compositions including sodium phenylbutyrate and uses thereof. US patent 11,154,521 (B2). October 26, 2021. **3.** Keating AV, Soto J, Tuleu C, Forbes C, Zhao M, Craig DQM. Solid state characterisation and taste masking efficiency evaluation of polymer based extrudates of isoniazid for paediatric administration. *Int J Pharm.* 2018;536(2):536-546. **4.** US Department of Health of and Human Services, Food and Drug Administration, Center for Drug Evaluation and Research (CDER). *Determining Whether to Submit an ANDA or a 505(b)(2) Application: Guidance for Industry.* Food and Drug Administration; May 2019. Accessed April 23, 2023. https://www.fda.gov/media/124848/download

Visit OLPRUVA.com for more information, support and resources, and more.

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