

Carbaglu[®]
carglumic acid

How to Use and Store CARBAGLU



How to Take CARBAGLU® (carglumic acid)

CARBAGLU is a prescription drug used in all ages to help treat a rare inherited disorder called NAGS deficiency, a lack or shortage of NAGS enzyme activity, which leads to high blood ammonia levels (hyperammonemia). CARBAGLU is a white, elongated tablet that is scored and coded “C” on one side. Each tablet contains 200 mg of carglumic acid.

CARBAGLU is usually given for the first time in a hospital or emergency setting. Based on your specific symptoms and ammonia level, your initial dose may range from 100 mg/kg/day to 250 mg/kg/day. Your doctor may also recommend other therapies to help lower your ammonia levels.

After an initial phase of dose adjustment, you may be placed on a maintenance dose. This dose of CARBAGLU should be adjusted to target a normal blood ammonia level for your age.

Your total daily dose should be divided into 2 to 4 doses, rounded to the nearest 100 mg (half of a CARBAGLU tablet), and taken right before meals or feedings. You can find this and additional information in the Full Prescribing Information, including Instructions for Use. Always consult your doctor with questions about your prescribed dose of CARBAGLU.

Different Ways CARBAGLU Can Be Taken

There are three different ways that CARBAGLU can be taken, based on patient age and condition: by mouth, by mouth using an oral syringe, or through a special tube (called a nasogastric or NG tube) that has been medically inserted through the nose, past the throat, and into the stomach.

Important Information

- **CARBAGLU tablet for oral suspension (CARBAGLU tablet) must be mixed in water before taking.** CARBAGLU tablets should not be mixed in any other food or liquid.
- **Do not** swallow CARBAGLU tablets whole.
- **Do not** crush CARBAGLU tablets.
- Take CARBAGLU **right before** meals or feedings.
- The CARBAGLU tablet and water mixture has a slightly sour taste.

You may need to ask your healthcare provider or pharmacist for a medicine cup to measure the correct amount of water you will need to prepare your dose of CARBAGLU .

IMPORTANT SAFETY INFORMATION

- **Hyperammonemia:** Monitor plasma ammonia level during treatment. Prolonged exposure to elevated plasma ammonia level can result in brain injury or death. Prompt use of all therapies necessary to reduce blood ammonia level is essential.

Carbaglu[®]
carglumic acid

By Mouth Using a Cup



Children and Adults

1. Add about 2.5 mL of water into a small cup for each CARBAGLU tablet, or each $\frac{1}{2}$ CARBAGLU tablet, needed for the prescribed dose. For example, if the prescribed dose is 2 CARBAGLU tablets, add about 5 mL of water into the cup. If the prescribed dose is $2\frac{1}{2}$ CARBAGLU tablets, add about 7.5 mL of water into the cup. Ask your healthcare provider if you are not sure of how much water you should use for the prescribed dose of CARBAGLU.
2. Place the prescribed number of CARBAGLU tablets into the water in the cup.
3. Carefully stir the CARBAGLU tablet and water mixture in the cup to avoid spilling the mixture. CARBAGLU tablets do not dissolve completely in water.
4. Swallow the CARBAGLU tablet and water mixture **right away**.
5. Pieces of the tablet may remain in the cup. Add more water to the cup to rinse the cup and swallow the mixture **right away**.
6. Repeat step 5 until there are no pieces of the tablet left in the cup.

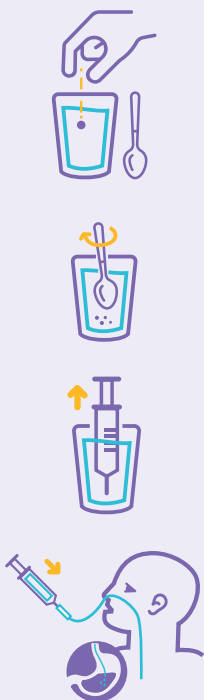
By Mouth Using an Oral Syringe



Children

1. Add about 2.5 mL of water into a small cup for each CARBAGLU tablet, or each $\frac{1}{2}$ CARBAGLU tablet, needed for the prescribed dose. For example, if the prescribed dose is 2 CARBAGLU tablets, add about 5 mL of water into the cup. If the prescribed dose is $2\frac{1}{2}$ CARBAGLU tablets, add about 7.5 mL of water into the cup. Ask your healthcare provider if you are not sure of how much water you should use for the prescribed dose of CARBAGLU.
2. Place the prescribed number of CARBAGLU tablets into the water in the cup.
3. Carefully stir the CARBAGLU tablet and water mixture in the cup to avoid spilling the mixture. CARBAGLU tablets do not dissolve completely in water.
4. Draw up all of the CARBAGLU tablet and water mixture in the cup into an oral syringe.
5. Give your child the CARBAGLU tablet and water mixture **right away** by placing the tip of the oral syringe along the inner cheek of their mouth, on either the right or left side. Slowly push all the way down on the plunger to give the medicine.
6. Pieces of the tablet may remain in the oral syringe. Refill the oral syringe with at least 1 mL to 2 mL of water, and give your child the mixture **right away**.
7. Repeat step 6 until there are no pieces of the tablet left in the oral syringe.

Through a Nasogastric (NG) Tube



Children and Adults

1. Add about 2.5 mL of water into a small cup for each CARBAGLU tablet, or each $\frac{1}{2}$ CARBAGLU tablet, needed for the prescribed dose. For example, if the prescribed dose is 2 CARBAGLU tablets, add about 5 mL of water into the cup. If the prescribed dose is $2\frac{1}{2}$ CARBAGLU tablets, add about 7.5 mL of water into the cup. Ask your healthcare provider if you are not sure of how much water you should use for the prescribed dose of CARBAGLU.
2. Place the prescribed number of CARBAGLU tablets into the water in the cup.
3. Carefully stir the CARBAGLU tablet and water mixture in the cup to avoid spilling the mixture. CARBAGLU tablets do not dissolve completely in water.
4. Draw up all of the CARBAGLU tablet and water mixture in the cup into a catheter-tip syringe.
5. Connect the catheter-tip syringe to the NG tube.
6. Give the CARBAGLU tablet and water mixture through the NG tube **right away**.
7. Pieces of the tablet may remain in the catheter-tip syringe or NG tube. Refill the catheter-tip syringe with 1 mL to 2 mL of water and flush the NG tube **right away**.
8. Repeat step 7 until there are no pieces of the tablet left in the catheter-tip syringe or NG tube.

Storing CARBAGLU® (carglumic acid)



Before opening, store CARBAGLU in a refrigerator between 36°F to 46°F (2°C to 8°C) in the container it comes in.



After opening, store CARBAGLU at room temperature between 59°F to 86°F (15°C to 30°C). **Do not store CARBAGLU in a refrigerator.**



Keep CARBAGLU tablets in a tightly closed container to protect the tablets from moisture.



Write the date the CARBAGLU tablet container is opened on the container label. Throw away any unused tablets one month after opening the tablet container.

Do not use CARBAGLU tablets after the expiration date on the tablet container.

Keep CARBAGLU and all medicines out of the reach of children.

INDICATIONS AND USAGE

CARBAGLU® (carglumic acid) is a prescription drug used in all ages to help treat a rare inherited disorder called NAGS deficiency, a lack or shortage of NAGS enzyme activity, which leads to high blood ammonia levels (hyperammonemia).

- CARBAGLU is used to supplement treatment of acute hyperammonemia due to the lack or shortage of the NAGS enzyme. Other ammonia lowering therapies such as other medications, hemodialysis and limiting protein in the diet are recommended during an acute episode.
- CARBAGLU is used to maintain normal blood ammonia levels in patients with NAGS deficiency. During maintenance therapy, the use of other ammonia lowering therapies and limiting protein may be needed based on plasma ammonia levels.

IMPORTANT SAFETY INFORMATION

- **Hyperammonemia:** Monitor plasma ammonia level during treatment. Prolonged exposure to elevated plasma ammonia level can result in brain injury or death. Prompt use of all therapies necessary to reduce blood ammonia level is essential.
- Most common side effects in >9% of patients are: vomiting, abdominal pain, fever, inflammation of the tonsils, reduced red cells in the blood, diarrhea, ear infection, infections, inflammation of the throat and nasal passages, protein molecules in red blood cells decreased, and headache.
- **To report SUSPECTED SIDE EFFECTS, contact Recordati Rare Diseases Inc. at 1-888-575-8344, or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.**

Obtaining CARBAGLU® (carglumic acid)

CARBAGLU is not available in retail pharmacies. Instead, it is available only through a specialty pharmacy called Accredo. Accredo will work with you and your insurance company and will mail your medication to your home.

Your doctor will send your CARBAGLU prescription and enrollment form to Accredo. Accredo may contact you if your insurance company needs additional information.

It is important to return all calls from Accredo so that you'll get your prescribed medication when you need it.

You can contact Accredo by calling 1-888-454-8860.

Financial Assistance Programs

For individuals experiencing financial hardship, Recordati Rare Diseases Inc. supports a Patient Assistance Program and a Co-Pay Assistance Program, administered by Accredo. For more information, call Accredo: 1-888-454-8860.

**Please see accompanying Full Prescribing Information,
including Instructions for Use.**

For more information, visit www.CARBAGLU.com