

Dosing CARBAGLU: Adjustments for renal impairment

Mild renal impairment (eGFR 60-89 mL/min/1.73 m²)

Patient Type	Recommended Dose
All	No dosage adjustment is warranted. Use guides on page 5.

Moderate Renal Impairment (eGFR 30-59 mL/min/1.73 m²)

Patient Type	Recommended Dose	Frequency & Timing
Acute Hyperammonemia due to NAGS Deficiency	50 mg/kg/day to 125 mg/kg/day	Divide the daily dosage into 2 to 4 doses. Round dose to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet).
Chronic Hyperammonemia due to NAGS Deficiency	5 mg/kg/day to 50 mg/kg/day	Divide the daily dosage into 2 to 4 doses. Round dose to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet).
Acute Hyperammonemia due to PA or MMA	<ul style="list-style-type: none"> 75 mg/kg/day for patients weighing ≤ 15 kg 1.7 g/m²/day for patients weighing > 15 kg 	Divide the daily dosage into 2 equal doses. Round up dose to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet). Administer each dose 12 hours apart.

Severe Renal Impairment (eGFR ≤29 mL/min/1.73 m²)

Patient Type	Recommended Dose	Frequency & Timing
Acute Hyperammonemia due to NAGS Deficiency	15 mg/kg/day to 60 mg/kg/day	Divide the daily dosage into 2 to 4 doses. Round dose to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet).
Chronic Hyperammonemia due to NAGS Deficiency	2 mg/kg/day to 25 mg/kg/day	Divide the daily dose into 2 to 4 doses. Round to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet).
Acute Hyperammonemia due to PA or MMA	<ul style="list-style-type: none"> 25 mg/kg/day for patients weighing ≤ 15 kg 0.55 g/m²/day for patients weighing > 15 kg 	Divide the daily dosage into 2 equal doses. Round up dose to the nearest 50 mg (i.e., one-quarter of a CARBAGLU tablet). Administer each dose 12 hours apart.

IMPORTANT SAFETY INFORMATION

- **Contraindications:** None.
- **NAGS deficiency:** Most common adverse reactions (≥13%) are: vomiting, abdominal pain, pyrexia, tonsillitis, anemia, diarrhea, ear infection, infections, nasopharyngitis, hemoglobin decreased, and headache.
- **PA and MMA:** Most common adverse reactions (≥5%) are neutropenia, anemia, vomiting, electrolyte imbalance, decreased appetite, hypoglycemia, lethargy/stupor, encephalopathy and pancreatitis/lipase increased.