



STANDARD COMMERCIAL DRUG FORMULARY  
PRIOR AUTHORIZATION GUIDELINES

CARGLUMIC ACID

Generic	Brand	HICL	GCN	Medi-Span	Exception/Other
CARGLUMIC ACID	CARBAGLU CARGLUMIC ACID	25643		GPI-10 (3090823000)	

**GUIDELINES FOR USE**

**INITIAL CRITERIA (NOTE: FOR RENEWAL CRITERIA SEE BELOW)**

1. Does the patient have a diagnosis of acute or chronic hyperammonemia (HA) due to N-acetylglutamate synthase (NAGS) deficiency **AND** meet the following criterion?
  - NAGS gene mutation is confirmed by biochemical or genetic testing

If yes, continue to #2.

If no, continue to #4.

2. Is the request for generic carglumic acid?

If yes, **approve carglumic acid (generic only) by HICL or GPI-10 as follows:**

- **Acute HA due to NAGS deficiency: approve for 7 days.**
- **Chronic HA due to NAGS deficiency: approve for 6 months.**

If no, continue to #3.

3. Is the request for brand Carbaglu **AND** the patient meets the following criterion?

- The patient had a trial of generic carglumic acid

If yes, **approve by HICL or GPI-10 as follows:**

- **Acute HA due to NAGS deficiency: approve for 7 days.**
- **Chronic HA due to NAGS deficiency: approve for 6 months.**

If no, do not approve.

**DENIAL TEXT:** See the initial denial text at the end of the guideline.

4. Does the patient have a diagnosis of acute hyperammonemia (HA) due to propionic acidemia (PA) **AND** meet following criterion?

- The diagnosis is confirmed by the presence of elevated methylcitric acid and normal methylmalonic acid OR genetic testing confirming mutation in the PCCA or PCCB gene

If yes, **approve for 7 days by HICL or GPI-10.**

If no, continue to #5.

**DENIAL TEXT:** See the initial denial text at the end of the guideline.

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INITIAL CRITERIA (CONTINUED)

5. Does the patient have a diagnosis of acute hyperammonemia (HA) due to methylmalonic acidemia (MMA) **AND** meet following criterion?
- The diagnosis is confirmed by the presence of elevated methylmalonic acid, methylcitric acid OR genetic testing confirming mutation in the MMUT, MMA, MMAB or MMADHC genes

If yes, **approve for 7 days by HICL or GPI-10.**

If no, do not approve.

**INITIAL DENIAL TEXT: \*Some terms are already pre-defined in parenthesis. Please use these definitions if the particular text you need to use does not already have definition(s) in it.**

Our guideline named **CARGLUMIC ACID (Carbaglu)** requires the following rule(s) be met for approval:

- A. You have ONE of the following diagnoses:
1. Acute or chronic hyperammonemia (HA) due to N-acetylglutamate synthase (NAGS) deficiency (short-term or long-term high ammonia blood levels due to a genetic disorder)
  2. Acute hyperammonemia (HA) due to propionic acidemia (PA) or methylmalonic acidemia (MMA) (short-term high ammonia blood levels due to a genetic disorder)
- B. **If you have acute or chronic hyperammonemia due to N-acetylglutamate synthase deficiency, approval also requires:**
1. Your N-acetylglutamate synthase gene mutation is confirmed by biochemical or genetic testing (types of lab test)
  2. Requests for brand Carbaglu requires a trial of generic carglumic acid
- C. **If you have acute hyperammonemia due to propionic acidemia, approval also requires:**
1. Your diagnosis is confirmed by the presence of elevated methylcitric acid and normal methylmalonic acid (substances that indicate presence of a disease) OR genetic testing confirming mutation in the PCCA or PCCB gene (types of abnormal genes)
- D. **If you have acute hyperammonemia due to methylmalonic acidemia, approval also requires:**
1. Your diagnosis is confirmed by the presence of elevated methylmalonic acid, methylcitric acid OR genetic testing confirming mutation in the MMUT, MMA, MMAB or MMADHC genes (types of abnormal genes)

Your doctor told us **[INSERT PT SPECIFIC INFO PROVIDED]**. We do not have information showing you **[INSERT UNMET CRITERIA]**. This is why your request is denied. Please work with your doctor to use a different medication or get us more information if it will allow us to approve this request.

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GUIDELINES FOR USE (CONTINUED)

RENEWAL CRITERIA

**NOTE:** For the diagnoses of acute hyperammonemia (HA) due to N-acetylglutamate synthase (NAGS) deficiency or acute hyperammonemia (HA) due to propionic acidemia (PA) or methylmalonic acidemia (MMA), please refer to the Initial Criteria section.

- Does the patient have a diagnosis of chronic hyperammonemia (HA) due to N- acetylglutamate synthase (NAGS) deficiency **AND** meet the following criterion?
  - The patient has clinical improvement or improved plasma (blood) ammonia levels

If yes, **approve for 12 months by HICL or GPI-10.**

If no, do not approve.

**RENEWAL DENIAL TEXT:** *\*Some terms are already pre-defined in parenthesis. Please use these definitions if the particular text you need to use does not already have definition(s) in it.*

Our guideline named **CARGLUMIC ACID (Carbaglu)** requires the following rule(s) be met for renewal:

- You have chronic hyperammonemia (HA) due to N-acetylglutamate synthase (NAGS) (long-term high ammonia blood levels due to a genetic disorder)
- You have clinical improvement or improved plasma (blood) ammonia levels

Your doctor told us **[INSERT PT SPECIFIC INFO PROVIDED]**. We do not have information showing you **[INSERT UNMET CRITERIA]**. This is why your request is denied. Please work with your doctor to use a different medication or get us more information if it will allow us to approve this request.

**RATIONALE**

For further information, please refer to the Prescribing Information and/or Drug Monograph for Carbaglu.

**REFERENCES**

- Carbaglu [Prescribing Information]. Lebanon, NJ: Recordati Rare Diseases, Inc.; August 2021.

Library	Commercial	NSA
Yes	Yes	No

Part D Effective: N/A

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