

nipocalimab-aahu (Imaavy™)

Policy # 00942

Original Effective Date: 11/01/2025

Current Effective Date: 08/01/2026

Applies to all products administered or underwritten by Blue Cross and Blue Shield of Louisiana and its subsidiary, HMO Louisiana, Inc. (collectively referred to as the "Company"), unless otherwise provided in the applicable contract. Medical technology is constantly evolving, and we reserve the right to review and update Medical Policy periodically.

Investigational or experimental services are not covered. This includes any drug, device, procedure, or service provided under the investigational arm of a clinical trial or clinical study. These services are excluded from coverage under benefits.

When Services May Be Eligible for Coverage

Coverage for eligible medical treatments or procedures, drugs, devices or biological products may be provided only if:

- *Benefits are available in the member's contract/certificate, and*
- *Medical necessity criteria and guidelines are met.*

Based on review of available data, the Company may consider nipocalimab-aahu (Imaavy™)† for the treatment of myasthenia gravis to be **eligible for coverage**.**

Patient Selection Criteria

Coverage eligibility for nipocalimab-aahu (Imaavy) will be considered when the following criteria are met:

- **Initial (6 months)**
 - Patient is greater than or equal to 12 years of age; AND
 - Patient has a diagnosis of generalized myasthenia gravis; AND
 - Patient has an anti-acetylcholine receptor autoantibody positive serologic test OR an anti-muscle-specific tyrosine kinase autoantibody positive serologic test; AND
 - Patient has a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV; AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
 - Patient has a Myasthenia Gravis Activities of Daily Living (MG-ADL) total score of greater than or equal to 6; AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
 - Patient has received or is currently receiving pyridostigmine or corticosteroids unless there is clinical evidence or patient history that suggests the use of pyridostigmine or corticosteroids will cause an adverse effect or inadequate response to the patient; AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*

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- Patient has received or is currently receiving at least one nonsteroidal immunosuppressive therapy (NSIST) for at least 6 months unless there is clinical evidence or patient history that suggests NSISTs will be ineffective or cause an adverse reaction to the patient. Examples of NSISTs include azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, tacrolimus, and cyclophosphamide; AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
- Patient has evidence of unresolved symptoms of myasthenia gravis, resulting in functional impairment (e.g., difficulty swallowing, difficulty breathing, double vision, impaired speech, or impairment of mobility); AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
- The requested dose does not exceed 30 mg/kg mg at week 0, followed by 15 mg/kg at week 2 and then 15 mg/kg every 2 weeks thereafter.
- **Continuation (12 months)**
 - Patient has received an initial authorization for Imaavy; AND
 - Patient has experienced improvement on therapy as evidenced by at least ONE of the following:
 - Improvement in the Myasthenia Gravis Activities of Daily Living (MG-ADL) total score; OR
 - Improvement in the Quantitative Myasthenia Gravis (QMG) total score; AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
 - The requested dose does not exceed 15mg/kg every 2 weeks.

When Services Are Considered Not Medically Necessary

Based on review of available data, the Company considers the use of nipocalimab-aahu (Imaavy) when any of the following criteria are NOT met to be **not medically necessary.****

- For initial requests:
 - Patient has a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
 - Patient has a Myasthenia Gravis Activities of Daily Living (MG-ADL) total score of greater than or equal to 6
 - Patient has received or is currently receiving pyridostigmine
 - Patient has received or is currently receiving at least one nonsteroidal immunosuppressive therapy (NSIST) for at least 6 months
 - Patient has evidence of unresolved symptoms of myasthenia gravis, resulting in functional impairment (e.g., difficulty swallowing, difficulty breathing, double vision, impaired speech, or impairment of mobility)

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- For continuation requests:
 - Patient has experienced improvement on therapy as evidenced by at least ONE of the following:
 - Improvement in the Myasthenia Gravis Activities of Daily Living (MG-ADL) total score; OR
 - Improvement in Quantitative Myasthenia Gravis (QMG) total score

When Services Are Considered Investigational

Coverage is not available for investigational medical treatments or procedures, drugs, devices or biological products.

Based on review of available data, the Company considers the use of nipocalimab-aahu (Imaavy) when the patient selection criteria are not met (with the exception of those denoted above as **not medically necessary****) to be **investigational**.*

Policy Guidelines

Myasthenia Gravis Foundation of America (MGFA) Clinical Classification

Class	Description
I	Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal
IIa	Mild weakness affecting muscles other than ocular muscles. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles
IIb	Mild weakness affecting muscles other than ocular muscles. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
IIIa	Moderate weakness affecting muscles other than ocular muscles. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
IIIb	Moderate weakness affecting muscles other than ocular muscles. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
IVa	Severe weakness affecting muscles other than ocular muscles. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
IVb	Severe weakness affecting muscles other than ocular muscles. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
V	Intubation with or without mechanical ventilation except when employed during routine postoperative management.

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Myasthenia Gravis Activities of Daily Living (MG-ADL) profile

<i>Grade</i>	<i>0</i>	<i>1</i>	<i>2</i>	<i>3</i>	<i>Score</i>
1. Talking	Normal	Intermittent slurring or nasal speech	Constant slurring or nasal, but can be understood	Difficult to understand speech	
2. Chewing	Normal	Fatigue with solid food	Fatigue with soft food	Gastric tube	
3. Swallowing	Normal	Rare episode of choking	Frequent choking necessitating changes in diet	Gastric tube	
4. Breathing	Normal	Shortness of breath with exertion	Shortness of breath at rest	Ventilator dependence	
5. Impairment of ability to brush teeth or comb hair	None	Extra effort, but no rest periods needed	Rest periods needed	Cannot do one of these functions	
6. Impairment of ability to arise from a chair	None	Mild, sometimes uses arms	Moderate, always uses arms	Severe, requires assistance	
7. Double vision	None	Occurs, but not daily	Daily, but not constant	Constant	
8. Eyelid droop	None	Occurs, but not daily	Daily, but not constant	Constant	
				MG-ADL score total (items 1-8)=	

Quantitative Myasthenia Gravis (QMG) Score

<i>Test Item</i>	<i>None</i>	<i>Mild</i>	<i>Moderate</i>	<i>Severe</i>	<i>Score</i>
<i>Grade</i>	<i>0</i>	<i>1</i>	<i>2</i>	<i>3</i>	
Double vision on lateral gaze (secs)	61	11 - 60	1 - 10	Spontaneous	
Ptosis (upward gaze)	61	11 - 60	1 - 10	Spontaneous	
Facial muscles	Normal lid closure	Complete, weak, some resistance	Complete without resistance	Incomplete	
Swallowing 4 oz water	Normal	Minimal coughing or throat clearing	Severe coughing/choking or nasal congestion	Cannot swallow (test not attempted)	

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Speech after counting aloud from 1 to 50 (onset of dysarthria)	None at 50	Dysarthria at 30 - 49	Dysarthria at 10 - 29	Dysarthria at 9	
Right arm outstretched (90 degrees sitting), seconds	240	90 - 239	10 - 89	0 - 9	
Left arm outstretched (90 degrees sitting), seconds	240	90 - 239	10 - 89	0 - 9	
Forced Vital Capacity	≥ 80	65 - 79	50 - 64	≤ 50	
Rt-hand grip, kg Men Women	≥ 45 ≥ 30	15 - 44 10 - 29	5 - 14 5 - 9	0 - 4 0 - 4	
Lt-hand grip, kg Men Women	≥ 35 ≥ 25	15 - 34 10 - 24	5 - 14 5 - 9	0 - 4 0 - 4	
Head lifted (45 degrees supine), seconds	120	30 - 119	1 - 29	0	
Right leg outstretched (45 degrees supine), seconds	100	31 - 99	1 - 30	0	
Left leg outstretched (45 degrees supine), seconds	100	31 - 99	1 - 30	0	
				Total QMG Score:	

Background/Overview

Imaavy is a recombinant human immunoglobulin G1 (IgG1) monoclonal antibody indicated for the treatment of generalized myasthenia gravis (gMG) in adult and pediatric patients 12 years of age and older who are anti-acetylcholine receptor (AChR) or anti-muscle-specific tyrosine kinase (MuSK) antibody positive. It works by binding to the neonatal Fc receptor, resulting in the reduction of circulating IgG. Because it causes this reduction in IgG levels, immunization with live-attenuated or live vaccines is not recommended during treatment with Imaavy. If indicated, these vaccines should be administered before initiation of treatment with Imaavy. The recommended dosage of Imaavy is 30 mg/kg administered once via intravenous infusion over at least 30 minutes. Two weeks after the initial dosage, a maintenance dosage of 15 mg/kg via intravenous infusion should be administered over at least 15 minutes. The maintenance dosage should be continued every two weeks thereafter.

Myasthenia gravis is a chronic autoimmune neuromuscular disease that causes weakness in the skeletal muscles. The hallmark of the condition is muscle weakness that worsens after periods of activity and improves after periods of rest. Certain muscles such as those that control eye and eyelid movement, facial expression, chewing, talking, and swallowing are often involved in the disorder, however, the muscles that control breathing and neck and limb movements may also be affected. Acquired myasthenia gravis results from the binding of autoantibodies to the components of the neuromuscular junction, most commonly the acetylcholine receptor (AChR). However, antibodies to other proteins, such as the muscle-specific tyrosine kinase (MuSK) protein, can also lead to impaired transmission at the neuromuscular junction. Myasthenia gravis most commonly occurs in young adult women (< 40 years of age) and older men (> 60 years of age), but it can occur at any age, including childhood. The incidence ranges from 0.3 to 2.8 per 100,000 and it is estimated to affect more than 700,000 people worldwide. Various clinical scoring systems are available to assess the severity of disease and include the Myasthenia Gravis Foundation of America (MGFA) clinical classification system, Myasthenia Gravis Activities of Daily Living (MG-ADL), and Quantitative Myasthenia Gravis (QMG) test.

Medications to treat myasthenia gravis include anticholinesterase agents (e.g., pyridostigmine), which slow the breakdown of acetylcholine at the neuromuscular junction and thereby improve neuromuscular transmission and increase muscle strength. Immunosuppressive drugs improve muscle strength by suppressing the production of abnormal antibodies and may include prednisone, azathioprine, mycophenolate mofetil, tacrolimus, and rituximab. Plasmapheresis and intravenous immunoglobulin (IVIG) may be options in severe cases to remove the destructive antibodies; however, their effectiveness frequently only lasts a few weeks to months. Additionally, the Food and Drug Administration (FDA) has approved eculizumab (Soliris®)†, ravulizumab (Ultomiris™)‡, and zilucoplan (Zilbrysq®)‡, all complement inhibitors, as well as rozanolixizumab (Rystiggo®)‡ and efgartigimod alfa products (Vyvgart®, Vyvgart® Hytrulo)‡ which contain an IgG monoclonal antibody that binds to the neonatal Fc receptor for the treatment of generalized myasthenia gravis.

Although Soliris, Ultomiris, Vyvgart, Vyvgart Hytrulo, Rystiggo, Zilbrysq, and Imaavy are the only agents with FDA approval for the condition, the other agents have been used off-label and are still recommended as first-line therapy in clinical practice guidelines. Available guidelines have not been updated to address these newer treatments.

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FDA or Other Governmental Regulatory Approval

U.S. Food and Drug Administration (FDA)

Imaavy was approved in April 2025 for the treatment of generalized myasthenia gravis (gMG) in adult and pediatric patients 12 years of age and older who are anti-acetylcholine receptor (AChR) or anti-muscle-specific tyrosine kinase (MuSK) antibody positive.

Rationale/Source

This medical policy was developed through consideration of peer-reviewed medical literature generally recognized by the relevant medical community, U.S. Food and Drug Administration approval status, nationally accepted standards of medical practice and accepted standards of medical practice in this community, technology evaluation centers, reference to regulations, other plan medical policies, and accredited national guidelines.

The efficacy of Imaavy for the treatment of gMG in adults who are anti-AChR or anti-MuSK antibody positive was established in a 24-week, multicenter, randomized, double-blind, placebo-controlled study. Patients were treated with Imaavy with the recommended dosage regimen.

The study enrolled patients with gMG who met the following criteria:

- Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
- Myasthenia Gravis-Activities of Daily Living (MG-ADL) total score of at least 6
- On stable dose of standard of care MG therapy prior to baseline that included acetylcholinesterase (AChE) inhibitors, steroids or non-steroidal immunosuppressive therapies (NSISTs), either in combination or alone.

In the study, a total of 196 patients were randomized 1:1 to receive Imaavy (n = 98) or placebo (n = 98). Baseline characteristics were similar between treatment groups. Eighty-eight percent (n = 134) of patients were positive for AChR antibodies and 10% (n = 16) were positive for MuSK antibodies. At baseline, in each group, 85% of patients received AChE inhibitors, 66% of patients received steroids, and 54% of patients received NSISTs at stable doses.

The efficacy of Imaavy was measured using the MG-ADL scale, which assesses the impact of gMG on daily functions of 8 signs and symptoms that are typically affected in gMG. Each item is assessed on a 4-point scale, where a score of 0 represents normal function and a score of 3 represents loss of ability to perform that function. A total score ranges from 0 to 24, with the higher scores indicating more impairment.

The primary efficacy endpoint was the comparison of the mean change from baseline to Weeks 22, 23, and 24 between treatment groups in the MG-ADL total score. A statistically significant difference favoring Imaavy was observed in MG-ADL total score change from baseline [-4.7 points in the Imaavy-treated group vs. -3.3 points in the placebo-treated group (p = 0.002)].

References

1. Imaavy [package insert]. Janssen Biotech, Inc. Horsham, PA. April 2025.

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Policy History

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10/02/2025 Medical Policy Committee review

10/08/2025 Medical Policy Implementation Committee approval. New policy.

01/01/2026 Coding update

07/21/2026 Pre Pharmacy & Therapeutics Committee review

07/22/2026 Pharmacy & Therapeutics Committee review approval. Updated trial period of a nonsteroidal immunosuppressive therapy (NSIST) from 1 year to 6 months. Clarified criterion describing evidence of unresolved symptoms. In addition, authorization approval lengths were added reflecting a 6 month approval for initial requests and a 12 month approval for continuation requests.

Next Scheduled Review Date: 07/2027

Coding

The five character codes included in the Louisiana Blue Medical Policy Coverage Guidelines are obtained from Current Procedural Terminology (CPT®)†, copyright 2025 by the American Medical Association (AMA). CPT is developed by the AMA as a listing of descriptive terms and five character identifying codes and modifiers for reporting medical services and procedures performed by physician.

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CPT is a registered trademark of the American Medical Association.

Codes used to identify services associated with this policy may include (but may not be limited to) the following:

Code Type	Code
CPT	NA
HCPCS	J9256 Delete code effective 01/01/2026: C9305 Delete codes effective 08/01/2026: C9399, J3590
ICD-10 Diagnosis	All related diagnoses

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*Investigational – A medical treatment, procedure, drug, device, or biological product is Investigational if the effectiveness has not been clearly tested and it has not been incorporated into standard medical practice. Any determination we make that a medical treatment, procedure, drug, device, or biological product is Investigational will be based on a consideration of the following:

- A. Whether the medical treatment, procedure, drug, device, or biological product can be lawfully marketed without approval of the U.S. Food and Drug Administration (FDA) and whether such approval has been granted at the time the medical treatment, procedure, drug, device, or biological product is sought to be furnished; or
- B. Whether the medical treatment, procedure, drug, device, or biological product requires further studies or clinical trials to determine its maximum tolerated dose, toxicity, safety, effectiveness, or effectiveness as compared with the standard means of treatment or diagnosis, must improve health outcomes, according to the consensus of opinion among experts as shown by reliable evidence, including:
 1. Consultation with technology evaluation center(s);
 2. Credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community; or
 3. Reference to federal regulations.

**Medically Necessary (or “Medical Necessity”) - Health care services, treatment, procedures, equipment, drugs, devices, items or supplies that a Provider, exercising prudent clinical judgment, would provide to a patient for the purpose of preventing, evaluating, diagnosing or treating an illness, injury, disease or its symptoms, and that are:

- A. In accordance with nationally accepted standards of medical practice;
- B. Clinically appropriate, in terms of type, frequency, extent, level of care, site and duration, and considered effective for the patient's illness, injury or disease; and
- C. Not primarily for the personal comfort or convenience of the patient, physician or other health care provider, and not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of that patient's illness, injury or disease.

For these purposes, “nationally accepted standards of medical practice” means standards that are based on credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community, Physician Specialty Society recommendations and the views of Physicians practicing in relevant clinical areas and any other relevant factors.

‡ Indicated trademarks are the registered trademarks of their respective owners.

NOTICE: If the Patient’s health insurance contract contains language that differs from the BCBSLA Medical Policy definition noted above, the definition in the health insurance contract will be relied upon for specific coverage determinations.

NOTICE: Medical Policies are scientific based opinions, provided solely for coverage and informational purposes. Medical Policies should not be construed to suggest that the Company recommends, advocates, requires, encourages, or discourages any particular treatment, procedure, or service, or any particular course of treatment, procedure, or service.

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NOTICE: Federal and State law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage.

NOTICE: If an authorization for an ongoing course of treatment has been provided to a member and the member changes from one health plan to another health plan (e.g., a member moves from carrier A to Louisiana Blue), Louisiana Blue may honor the previous health plan's authorization for the same service under the same type of in-network benefit for a 90-day transition period. Documentation of the authorization for the ongoing course of treatment from the previous health plan must be provided to us by the member or their provider and the services provided for the course of treatment must otherwise be a covered service under the Louisiana Blue health plan. This provision does not apply to medications covered under the plan's pharmacy benefit.