Medical Drug Clinical Criteria

Subject:	Imaavy (nipocalimab-aahu)		
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Overview

This document addresses the use of Imaavy (nipocalimab-aahu), a neonatal Fc receptor inhibitor (FcRn) approved for the treatment of generalized myasthenia gravis (gMG) in adult and pediatric members 12 years of age and older who are anti-acetylcholine receptor (AChR) or anti-muscle-specific tyrosine kinase (MuSK) antibody positive. Imaavy is intravenously administered through an infusion pump by a healthcare professional once every 2 weeks.

Generalized myasthenia gravis (gMG) is an autoimmune neuromuscular disorder characterized by fluctuating motor weakness causing dyspnea, dysphagia, diplopia, dysarthria, and ptosis. Generalized myasthenia gravis is commonly mediated by IgG autoantibodies directed against the neuromuscular junction. Approximately 95% of patients with generalized myasthenia gravis test positive for antibodies (about 85% antiacetylcholine receptor [AChR]-positive, about 8% anti-muscle-specific tyrosine kinase [MuSK]-positive, and about 1–2% anti-low-density lipoprotein receptor-related protein 4 [LRP4]-positive). Treatment strategies include symptomatic therapy (with anticholinesterase agents such as pyridostigmine), chronic immunotherapy with steroids or other immunosuppressive drugs (such as azathioprine, cyclosporine, or methotrexate), rapid immunotherapy (with plasmapheresis or IV immune globulin), and/or surgical treatment. Eculizumab, ravulizumab, and zilucoplan are immunotherapies which block complement activation triggered by acetylcholine receptor antibodies at the neuromuscular junction. Rozanolixizumab and efgartigimod alfa reduce autoantibodies by binding to the neonatal Fc receptor (FcRn), but differ in product administration, frequency, and population. Imaavy is the first FcRn inhibitor approved for gMG in the pediatric population; and the first FcRn inhibitor administered continuously every 2 weeks. Myasthenia Gravis Foundation of America (MGFA) international consensus guidelines, published prior to the approval of FcRn inhibitors, recommend immunosuppressive drugs and/or corticosteroids for individuals who have not met treatment goals after an adequate trial of pyridostigmine.

Current published evidence for Imaavy includes a phase 3, multicenter, randomized, placebo-controlled trial that enrolled individuals with antibody-positive gMG who had an inadequate response (defined as a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class IIa/b to Iva/b disease and a Myasthenia Gravis Activities of Daily Living (MG-ADL) score of ≥6) to their current, stable standard-of-care therapy for gMG (cholinesterase inhibitors, corticosteroids, or non-steroidal immunosuppressants) prior to screening and throughout the study. Participants in the trial were treated with nipocalimab 30 mg/kg initial dose followed by 15 mg/kg or placebo by intravenous infusion at study sites every 2 weeks for a 24-week duration. Nipocalimab or placebo was administered as an add-on treatment to background standard-of-care therapies, with no changes permitted during the double-blind phase. The primary efficacy endpoint was the difference in the least-squares mean change from baseline in the MG-ADL total score between the nipocalimab and placebo groups, averaged over weeks 22, 23, and 24. This averaging aimed to demonstrate sustained efficacy over the final two weeks. Secondary endpoints included the average change from baseline in the Quantitative Myasthenia Gravis (QMG) score over weeks 22 and 24 among other measures. A clinically meaningful improvement in the MG-ADL response was defined as a reduction of 2 or more points. The trial showed statistically significant improvements in the MG-ADL total score for the nipocalimab group compared to placebo over weeks 22, 23, and 24, meeting the primary efficacy endpoint. Additionally, there was a significantly greater reduction from baseline in the QMG total score observed over weeks 22 and 23, achieving the first key secondary endpoint.

Clinical Criteria

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Imaavy (nipocalimab-aahu)

Initial requests for Imaavy (nipocalimab-aahu) may be approved if the following criteria are met:

I. Individual is 12 years of age or older; **AND**
II. Individual has a diagnosis of generalized myasthenia gravis (gMG); **AND**
III. Documentation is provided that İndividual has one of the following:
    A. A positive serologic test for the presence of anti-acetylcholine receptor antibodies (AchR); **OR**
    B. A positive serologic test for the presence of anti-muscle-specific tyrosine kinase (MuSK) antibodies;
AND
IV. Individual has Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV disease (Antozzi 2025); **AND**
V. Documentation is provided that individual has a Myasthenia Gravis Activities of Daily Living (MG-ADL) score of at least 6 or higher (Antozzi 2025); **AND**
VI. Individual meets both of the following (A and B):
    A. Individual has had a trial and inadequate response or intolerance to an acetylcholinesterase inhibitor; **OR**
        1. Individual is on a stable dose of an acetylcholinesterase inhibitor; **OR**
        2. Individual has a contraindication to acetylcholinesterase inhibitors;
    AND
    B. Individual has had a trial and inadequate response or intolerance to one or more immunosuppressive agents (including but not limited to systemic corticosteroids or non-steroidal immunosuppressants); **OR**
        1. Individual is on a stable dose of one or more immunosuppressive agents (including but not limited to systemic corticosteroids or non-steroidal immunosuppressants); **OR**
        2. Individual has a contraindication to systemic corticosteroids and non-steroidal immunosuppressants

Initial Approval Duration: 26 weeks

Requests for continued use of Imaavy (nipocalimab-aahu) may be approved if the following criteria are met:

I. Individual has experienced a prior clinical response to Imaavy (nipocalimab-aahu) treatment as defined by the following:
    A. Reduction in signs or symptoms that impact daily function; **AND**
    B. Documentation is provided that there is at least a 2-point reduction in MG-ADL total score from pre-treatment baseline

Requests for Imaavy (nipocalimab-aahu) may not be approved for the following:

I. Individual is using in combination with maintenance immunoglobulin treatment, eculizumab, ravulizumab, efgartigimod-alfa, rozanolixizumab, zilucoplan, or rituximab; **OR**
II. If the above criteria are not met and for all other indications.

Continuation Approval Duration: 1 year

Quantity Limits

Imaavy (nipocalimab-aahu) Quantity Limits

Drug	Limit
Imaavy (nipocalimab-aahu) 300 mg/1.62 mL (185 mg/mL) single dose vial; 1200 mg/6.5 mL (185 mg/mL) single dose vial	**Initial dose**: One 30 mg/kg infusion **Subsequent doses**: 15 mg/kg every 2 weeks

Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

HCPCS

C9399	Unclassified drugs or biologicals [when specified as Imaavy (nipocalimab-aahu)]
J3590	Unclassified biologics [when specified as Imaavy (nipocalimab-aahu)]

ICD-10 Diagnosis

All diagnosis pend

Document History

New: 5/16/2025
Document History:

- 5/16/2025 – Select Review: Add new clinical criteria and quantity limit for Imaavy. Administrative update to add documentation. Coding Reviewed: Added HCPCS NOC C9399, J3590, and all diagnosis pend for Imaavy.

References

1. Antozzi C, Vu T, Ramchandren S, et al. Safety and efficacy of nipocalimab in adults with generalised myasthenia gravis (Vivacity-MG3): a phase 3, randomised, double-blind, placebo-controlled study. *Lancet Neurol*. 2025;24(2):105-116. doi:10.1016/S1474-4422(24)00498-8
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
5. Narayanaswami P, Sanders DB, Wolfe G, et al for the Task Force of the Myasthenia Gravis Foundation of America (MGFA). International consensus guidance for management of myasthenia gravis 2020 update. Neurology 2021; 96:114-122

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.