UTILIZATION MANAGEMENT MEDICAL POLICY

POLICY: Neurology – Imaavy Utilization Management Medical Policy
- Imaavy™ (nipocalimab-aahu intravenous infusion – Johnson & Johnson)

REVIEW DATE: 05/13/2026; selected revision 07/08/2026 and 09/23/2026

OVERVIEW

Imaavy, a neonatal Fc receptor blocker, is indicated for the treatment of **generalized myasthenia gravis** in adults and pediatric patients ≥ 12 years of age who are anti-acetylcholine receptor (AChR) or anti-muscle-specific tyrosine kinase (MuSK) antibody positive.[1] Imaavy is also indicated for the treatment of warm autoimmune hemolytic anemia (wAIHA) in adults and pediatric patients ≥ 12 years of age currently or previously treated with corticosteroids.

Disease Overview

Myasthenia gravis is a chronic autoimmune neuromuscular disease that causes weakness in the skeletal muscles, which are responsible for breathing and moving parts of the body, including the arms and legs.[2] Myasthenia gravis is caused by the production of pathogenic immunoglobulin G (IgG) autoantibodies against neuromuscular junction components (AChR, MuSK, and low density lipoprotein receptor-related protein 4 [LRP4]).[3] Approximately 85% of patients with myasthenia gravis are anti-AChR antibody-positive and approximately 5% to 8% of patients are anti-MuSK antibody-positive.[4] The result of the antibodies at the junction is unsuccessful nerve transmission and deficiency or weakness of muscle contractions.[3] The hallmark of myasthenia gravis is muscle weakness that worsens after periods of activity and improves after periods of rest.[2] 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. MuSK is rare and frequently more severe subtype of myasthenia gravis. Symptomatic treatment with acetylcholinesterase inhibitors (pyridostigmine) is generally unsatisfactory and may be deleterious in MuSK.[10]

wAIHA

Autoimmune hemolytic anemias (AIHAs) are generally due to premature destruction of red blood cells by a self-directed immune attack.[11] wAIHA is caused by IgG autoantibodies (reacting at body temperature) and is the most frequent form of AIHA (60% to 70% of cases). It is a rare, life-threatening disease with a global prevalence of 4.6 to 17 per 100,000.[12] Patients with wAIHA are at increased risk for infections, venous thromboembolism, and major cardiovascular events. Fatigue is a prominent symptom related to anemia. Therapy for AIHAs has historically been based on interventions with few evidence-based studies (e.g., steroids, immunosuppressors, splenectomy, rituximab).

Clinical Efficacy

Myasthenia Gravis

The efficacy of Imaavy was evaluated in one randomized, double-blind, multicenter, placebo-controlled Phase III pivotal study, Vivacity-MG3 (published) [n = 196].[5] Enrolled patients were ≥ 18 years of age with symptoms of gMG categorized as Myasthenia Gravis Foundation of America (MGFA) Class II to IV at screening. In addition, patients had a suboptimal response (defined as myasthenia gravis-activities of daily living [MG-ADL] score of ≥ 6 at baseline) to their current, stable standard-of care therapy for gMG (e.g., acetylcholinesterase inhibitor, corticosteroid, immunosuppressant). Patients were randomized to receive Imaavy (at the FDA-approved regimen) or placebo in addition to their stable gMG regimen. The majority of patients were anti-AChR antibody positive (82% in the Imaavy group vs. 93% in the placebo

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group), followed by anti-MuSK antibody-positive (16% vs. 5%, respectively) and anti-lipoprotein receptor-related protein 4 (LRP4) antibody positive (3% vs. 1%, respectively). The efficacy analysis dataset included all randomly assigned patients who received at least one dose (partial or complete) of study drug in the double-blind phase and were antibody-positive for a gMG-related pathogenic antibody (anti-AChR, anti-MuSK, or anti-LRP4). The primary efficacy endpoint was the difference between Imaavy and placebo in the least-squares (LS) mean change from baseline in the MG-ADL total score averaged over Weeks 22, 23, and 24. A significantly greater reduction in the MG-ADL total score was observed in the Imaavy group compared with placebo (LS mean change from baseline was -4.70 vs. -3.25, respectively; difference of -1.45; P = 0.0024). The first key secondary endpoint was the difference in the LS mean change in the quantitative myasthenia gravis (QMG) total score from baseline over Weeks 22 and 24; the results favored Imaavy (-4.86 for Imaavy vs. -2.05 for placebo; difference of -2.81; P = 0.00012). The second key secondary endpoint was the percentage of patients with at least a 2-point improvement in the MG-ADL score over Weeks 22, 23, and 24 and significantly more patients in the Imaavy group achieved this endpoint compared with placebo (69% vs. 53%, respectively; difference of 16.2%; P = 0.021).

Clinical Pediatric Data: Use in pediatric patients ≥ 12 years of age for this indication is supported by evidence from an adequate and well-controlled trial in adults and additional pharmacokinetic and safety data in pediatric patients who are ≥ 12 years of age.[1] There is an ongoing Phase II/III open-label study to evaluate the efficacy of Imaavy in pediatric patients 2 to < 18 years of age with gMG who are anti-AChR or anti-MuSK antibody positive.[6] Data are not yet available.

wAIHA
The efficacy of Imaavy was established in one phase 2/3, randomized, 24-week, double-blind, placebo-controlled study called ENERGY (n = 77).[12] Patients (≥ 18 years of age) were randomized 1:1:1 to Imaavy 30 mg/kg intravenously (IV) every 4 weeks, Imaavy 15 mg/kg IV every 2 weeks, or placebo IV every 2 weeks. Patients had a confirmed diagnosis of wAIHA for at least 3 months and were either receiving or had previously received treatment for the condition.[1] In addition, patients had a Hgb level < 10 g/dL and evidence of hemolysis and positive monospecific direct antiglobulin test. Exclusion criteria included transfusion ≤ 30 days prior to randomization, rituximab ≤ 3 months prior to screening, intravenous immunoglobulin ≤ 6 weeks prior to screening, other types of AIHA (cold antibody AIHA, cold agglutin syndrome, mixed-type AIHA) or paroxysmal cold hemoglobinuria.[12] The primary endpoint was the proportion of patients who achieved Hgb response (defined as Hgb ≥ 10 g/dL and ≥ 2 g/dL increase from baseline for 3 consecutive visits spanning ≥ 28 days starting by Week 16 and without the need for rescue therapy or dose modifications of ongoing background treatment). The primary endpoint was met by 22.4% (17/76) in the combined Imaavy group (both doses of Imaavy) [1-sided P = 0.017] vs. 7.7% (3/39) with placebo. The primary endpoint was met by 23.7% (9/38) with Imaavy 30 mg/kg IV every 4 weeks (1-sided P = 0.015) and 21.8 (8/38) with 15 mg/kg IV every 2 weeks (1-sided P = 0.044; not significant).

Clinical Pediatric Data: Use in pediatric patients ≥ 12 years of age for this indication is supported by evidence from an adequate and well-controlled trial in adults and additional pharmacokinetic and safety data in pediatric patients who are ≥ 12 years of age.[1]

Dosing Information
Myasthenia Gravis
The initial dose of Imaavy is 30 mg/kg administered once via IV infusion over at least 30 minutes.[1] Two weeks after the initial dose, administer a maintenance dose of 15 mg/kg via IV infusion over at least 15 minutes. Continue the maintenance dose every 2 weeks thereafter.

wAIHA

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The initial dose of Imaavy is 30 mg/kg administered once via IV infusion over at least 30 minutes.[1] Four weeks after the initial dose, administer a subsequent dose of 30 mg/kg via IV infusion over at least 15 minutes. Continue the subsequent dose every 4 weeks thereafter.

Guidelines

Myasthenia Gravis

An international consensus guidance for the management of myasthenia gravis was published in 2016.[7] The guidelines recommend pyridostigmine for the initial treatment in most patients with myasthenia gravis. The ability to discontinue pyridostigmine can indicate that the patient has met treatment goals and may guide the tapering of other therapies. Corticosteroids or immunosuppressant therapy should be used in all patients with myasthenia gravis who have not met treatment goals after an adequate trial of pyridostigmine. Nonsteroidal immunosuppressant agents include azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, and tacrolimus. It is usually necessary to maintain some immunosuppression for many years, sometimes for life. Plasma exchange and intravenous immunoglobulin can be used as short-term treatments in certain patients. A 2020 update to these guidelines provides new recommendations for methotrexate, rituximab, and eculizumab intravenous infusion (Soliris®, biosimilars).[8] All recommendations should be considered extensions or additions to recommendations made in the initial international consensus guidance (2016). Oral methotrexate may be considered as a steroid-sparing agent in patients with generalized myasthenia gravis who have not tolerated or responded to steroid-sparing agents. Rituximab should be considered as an early therapeutic option in patients with anti-MuSK antibody-positive myasthenia gravis who have an unsatisfactory response to initial immunotherapy. Eculizumab should be considered in the treatment of severe, refractory, anti-AChR antibody-positive generalized myasthenia gravis.

Pediatric patients with generalized myasthenia gravis. Cholinesterase inhibitors are used first-line for the symptomatic treatment of juvenile myasthenia gravis (JMG); pyridostigmine is the most widely used cholinesterase inhibitor for JMG.[8] There are no formal guidelines for the use of immunosuppressive therapy in JMG and current practice has been taken from adult guidelines and expert opinions based on individual experience. Prednisolone is accepted as the first-line immunosuppressive therapy in JMG. Second-line therapies or steroid-sparing agents include, but are not limited to, azathioprine, mycophenolate mofetil, tacrolimus, rituximab, cyclosporine, and cyclophosphamide.

wAIHA

International recommendations for the diagnosis and treatment of AIHA in adults (2020) recommends corticosteroids and “sometimes” rituximab as first-line therapy.[13] Approximately 80% of patients respond to daily dose equivalent to prednisone 60 mg to 100 mg. It is noted that a prednisone-rituximab combination may be considered in selected patients with severe disease or as a corticosteroid-sparing strategy. The second-line treatment recommended is rituximab and third-line therapy options include splenectomy or an alternative immunosuppressant (e.g., azathioprine, cyclosporine, mycophenolate). Immunoglobulins and plasma exchange can be used for rescue therapy and should be regarded as bridging therapy.

POLICY STATEMENT

Prior Authorization is recommended for medical benefit coverage of Imaavy. Approval is recommended for those who meet the **Criteria** and **Dosing** for the listed indication. Extended approvals are allowed if the patient continues to meet the Criteria and Dosing. Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). All approvals are provided for the duration noted below. In cases where the approval is authorized in months, 1 month is equal to 30 days. Because of the specialized skills required for evaluation and diagnosis of patients treated with Imaavy as well as the monitoring required for adverse events and

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long-term efficacy, approval requires Imaavy to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Imaavy is recommended in those who meet the following criteria:

FDA-Approved Indication

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1. **Generalized Myasthenia Gravis.** Approve for the duration noted if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve for 6 months if the patient meets ALL of the following (i, ii, iii, iv, v, and vi):
     - **i.** Patient is ≥ 12 years of age; AND
     - **ii.** If patient is ≥ 18 years of age, patient meets BOTH of the following (a and b):
       - **a)** Myasthenia Gravis Foundation of America classification of II to IV; AND
       - **b)** Myasthenia Gravis Activities of Daily Living (MG-ADL) score of ≥ 6; AND
     - **iii.** Patient meets ONE of the following (a or b):
       - **a)** Patient has confirmed anti-acetylcholine receptor antibody-positive generalized myasthenia gravis; OR
       - **b)** Patient has confirmed anti-muscle-specific tyrosine kinase (MuSK) antibody-positive generalized myasthenia gravis; AND
     - **iv.** Patient meets ONE of the following (a, b, or c):
       - **a)** Patient has received or is currently receiving pyridostigmine; OR
       - **b)** Patient has a contraindication to pyridostigmine; OR
       - **c)** Patient has anti-MuSK antibody-positive generalized myasthenia gravis; AND
     - **v.** Patient has evidence of unresolved symptoms of generalized myasthenia gravis; AND
       Note: Examples of unresolved symptoms include difficulty swallowing, difficulty breathing, or a functional disability resulting in the discontinuation of physical activity (e.g., double vision, talking, impairment of mobility).
     - **vi.** The medication is being prescribed by or in consultation with a neurologist; OR
   - **B)** Patient is Currently Receiving Imaavy. Approve for 1 year if the patient meets ALL of the following (i, ii, and iii):
     - **i.** Patient is ≥ 12 years of age; AND
     - **ii.** According to the prescriber, patient is continuing to derive benefit from Imaavy; AND
       Note: Examples of derived benefit include reductions in exacerbations of myasthenia gravis; improvements in speech, swallowing, mobility, and respiratory function.
     - **iii.** The medication is being prescribed by or in consultation with a neurologist.

   Dosing. Approve the following (A or B):
   - **A)** Initial Therapy. Approve initial dose of 30 mg/kg administered once via intravenous infusion; followed by maintenance dose of 15 mg/kg administered via intravenous infusion (starting 2 weeks after the initial dose and administer every 2 weeks thereafter); AND
   - **B)** Patient is Currently Receiving Imaavy. Approve maintenance dose of 15 mg/kg administered via intravenous infusion every 2 weeks.

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2. **Warm autoimmune hemolytic anemia (wAIHA).** Approve for the duration noted if the patient meets ONE of the following (A or B):

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A) Initial Therapy. Approve for 6 months if the patient meets ALL of the following (i, ii, and iii):
- **i.** Patient is ≥ 12 years of age; AND
- **ii.** Patient has tried, or is currently receiving, a systemic corticosteroid, unless contraindicated; AND
  Note: Examples of systemic corticosteroids are prednisone, prednisolone, methylprednisolone.
- **iii.** The medication is being prescribed by or in consultation with a hematologist, oncologist, rheumatologist, or immunologist.

B) Patient is Currently Receiving Imaavy. Approve for 1 year if the patient meets ALL of the following (i, ii, and iii):
- **i.** Patient is ≥ 12 years of age; AND
- **ii.** According to the prescriber, patient is continuing to derive benefit from Imaavy; AND
  Note: Examples include increased or stabilized hemoglobulin level, decreased reticulocytosis, decreased fatigue, decreased or stabilized number of relapses.
- **iii.** The medication is being prescribed by or in consultation with a hematologist, oncologist, rheumatologist, or immunologist.

Dosing. Approve the following (A or B):

A) Initial Therapy. Approve initial dose of 30 mg/kg administered once via intravenous infusion; followed by a subsequent dose of 30 mg/kg administered via intravenous infusion (starting 4 weeks after the initial dose and administer every 4 weeks thereafter); OR

B) Patient is Currently Receiving Imaavy. Approve subsequent doses of 30 mg/kg administered via intravenous infusion every 4 weeks.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Imaavy is not recommended in the following situations:

1. **Concomitant Use with Another Neonatal Fc Receptor Blocker, a Complement Inhibitor, a Rituximab Product, or Uplizna® (inebilizumab-cdon intravenous infusion).** There is no evidence to support concomitant use of Imaavy with another neonatal Fc receptor blocker, a complement inhibitor, a rituximab product, or Uplizna.
   Note: Examples of neonatal Fc receptor blockers are Rystiggo (rozanolixizumab-noli subcutaneous infusion), Vyvgart (efgartigimod alfa-fcab intravenous infusion) and Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc subcutaneous injection).
   Note: Examples of complement inhibitors are eculizumab intravenous infusion (Soliris, biosimilars), Ultomiris (ravulizumab-cwvz intravenous infusion), and Zilbrysq (zilucoplan subcutaneous injection).

2. **Other Types of Autoimmune Hemolytic Anemia (AIHA), including Cold Antibody AIHA, Cold Agglutin Syndrome, Mixed-Type AIHA, or Paroxysmal Cold Hemoglobinuria.** Imaavy is FDA-approved for wAIHA. There is no evidence to support the use of Imaavy in these conditions.

3. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

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HISTORY

Type of Revision	Summary of Changes	Review Date
New Policy	--	05/14/2025
Selected Revision	**Conditions Not Recommended for Approval**, the condition "Concomitant Use with Another Neonatal Fc Receptor Blocker, a Complement Inhibitor, a Rituximab Product" was revised to "Concomitant Use with Another Neonatal Fc Receptor Blocker, a Complement Inhibitor, a Rituximab Product, or Uplizna® (inebilizumab-cdon intravenous infusion)".	02/18/2026
Annual Revision	No criteria changes.	05/13/2026
Selected Revision	**Generalized Myasthenia Gravis**. Updated wording from "Patient previously received or is currently receiving pyridostigmine" to "Patient has received or is currently receiving pyridostigmine." Updated wording from "Patient has had inadequate efficacy, a contraindication, or significant intolerance to pyridostigmine" to "Patient has a contraindication to pyridostigmine." Added a criterion to bypass the trial of pyridostigmine if the patient has anti-muscle-specific tyrosine kinase (MuSK) antibody-positive generalized myasthenia gravis.	07/08/2026
Selected Revision	**Warm Autoimmune Hemolytic Anemia (wAIHA)**. This condition of approval was added to the policy. **Conditions Not Recommended for Approval**. The conditions of 'other types of autoimmune hemolytic anemia (AIHA)', including cold antibody AIHA, cold agglutin syndrome, mixed-type AIHA or paroxysmal cold hemoglobinuria were added as conditions not recommended for approval.	09/23/2026

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