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Eculizumab

Table of Contents

Overview .....1  
Initial Approval Criteria.....2  
Continuation of Therapy Criteria.....4  
Authorization Duration .....4  
Conditions Not Covered.....5  
Coding Information .....5  
Background.....6  
References .....8  
Revision Details .....8

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Overview

This policy supports medical necessity review for eculizumab intravenous infusion (**Bkemv, Epysqli, Soliris**).

## Initial Approval Criteria

**Eculizumab is considered medically necessary for the treatment of complement-mediated hemolytic uremic syndrome (atypical hemolytic uremic syndrome) when the individual meets ALL of the following criteria:**

1. Diagnosis of thrombocytopenic purpura (TTP) has been excluded (for example, normal ADAMTS 13 activity) OR a trial of plasma exchange did not result in clinical improvement
2. Absence of Shiga toxin-producing escherichia coli (E. coli) infection
3. Has been vaccinated against meningococcal infection (at least 2 weeks prior to treatment, if not previously vaccinated), where and when clinically appropriate
4. Medication is prescribed by, or in consultation, with a hematologist and/or a nephrologist

**Dosing.** The recommended intravenous dose for Complement-mediated hemolytic uremic syndrome (atypical hemolytic uremic syndrome) is:

1. For individuals 18 years of age or older, **ONE** of the following:
  - A. Induction: Up to 900 mg weekly for the first 4 weeks
  - B. Maintenance: Up to 1,200 mg at week 5, then up to 1,200 mg every 2 weeks thereafter
2. For individuals less than 18 years of age, **ONE** of the following:
  - A. 40 kg or more:
    - i. Induction: 900 mg weekly for 4 doses
    - ii. Maintenance: 1,200 mg at week 5, then 1,200 mg every 2 weeks
  - B. 30 kg to less than 40 kg:
    - i. Induction: 600 mg weekly for 2 doses
    - ii. Maintenance: 900 mg at week 3, then 900 mg every 2 weeks
  - C. 20 kg to less than 30 kg:
    - i. Induction: 600 mg weekly for 2 doses
    - ii. Maintenance: 600 mg at week 3, then 600 mg every 2 weeks
  - D. 10 kg to less than 20 kg:
    - i. Induction: 600 mg weekly for 1 dose
    - ii. Maintenance: 300 mg at week 2, then 300 mg every 2 weeks
  - E. 5 kg to less than 10 kg:
    - i. Induction: 300 mg weekly for 1 dose
    - ii. Maintenance: 300 mg at week 2, then 300 mg every 3 weeks

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**Eculizumab is considered medically necessary for the treatment of generalized myasthenia gravis when the individual meets ALL of the following criteria:**

1. Patient is  $\geq 6$  years of age
2. Documentation that the individual has confirmed anti-acetylcholine receptor antibody positive generalized myasthenia gravis
3. If patient is  $\geq 18$  years of age, patient meets BOTH of the following (A and B):
  - A. Myasthenia Gravis Foundation of America (MGFA) clinical classification class of II-IV (prior to starting therapy with eculizumab)
  - B. Myasthenia Gravis-Activities of Daily Living (MG-ADL) score of 6 or higher (prior to starting therapy with eculizumab)

4. Documentation of **ONE** of the following:
  - A. Is currently receiving pyridostigmine
  - B. Failure, contraindication, or intolerance to pyridostigmine
5. Documentation of **ONE** of the following:
  - A. Is currently receiving two different immunosuppressant therapies (for example, corticosteroid, azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, tacrolimus, cyclophosphamide, prednisone) for 1 year or longer
  - B. Failure, contraindication, or intolerance to two different immunosuppressant therapies
6. Has objective evidence of unresolved symptoms of generalized myasthenia gravis, such as difficulty swallowing, difficulty breathing, or a functional disability resulting in the discontinuation of physical activity (for example, double vision, talking, impairment of mobility)
7. The medication is prescribed by, or in consultation with a neurologist

**Dosing.** The recommended intravenous dose for Generalized Myasthenia Gravis meets **ONE** of the following (A or B):

- A) Adults ≥ 18 years of age: Approve the following regimen (i or ii):
  - i. Induction: Up to 900 mg weekly for the first 4 weeks
  - ii. Maintenance: Up to 1,200 mg at week 5, then up to 1,200 mg every 2 weeks thereafter
- B) Pediatric patients ≥ 6 and < 18 years of age. Approve ONE of the following regimens (based on body weight) [i, ii, iii, iv, or v]:
  - i. 40 kg and over: 900 mg weekly for the first 4 weeks; 1,200 mg at Week 5; then 1,200 mg every 2 weeks; OR
  - ii. 30 kg to < 40 kg: 600 mg for the first 2 weeks; 900 mg at Week 3; then 900 mg every 2 weeks; OR
  - iii. 20 kg to < 30 kg: 600 mg for the first 2 weeks; 600 mg at Week 3; then 600 mg every 2 weeks; OR
  - iv. 10 kg to < 20 kg: 600 mg single dose at Week 1; 300 mg at Week 2; then 300 mg every 2 weeks; OR
  - v. 5 kg to < 10 kg: 300 mg single dose at Week 1; 300 mg at Week 2; then 300 mg every 3 weeks.

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**Eculizumab is considered medically necessary for the treatment of neuromyelitis optica spectrum disorder (NMOSD) when the individual meets ALL of the following criteria:**

1. 18 years of age or older
2. Neuromyelitis optica spectrum disorder diagnosis confirmed by blood serum test for anti-aquaporin-4 antibody positive
3. Has been vaccinated against meningococcal infection (at least 2 weeks prior to treatment, if not previously vaccinated) where and when clinically appropriate
4. Medication is prescribed by, or in consultation with a neurologist

**Dosing.** The recommended intravenous dose for Neuromyelitis Optica Spectrum Disorder (NMOSD) is **ONE** of the following:

1. Induction: Up to 900 mg weekly for the first 4 weeks
2. Maintenance: Up to 1,200 mg at week 5, then up to 1,200 mg every 2 weeks thereafter

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**Eculizumab is considered medically necessary for the treatment of Paroxysmal nocturnal hemoglobinuria (PNH) when the individual meets ALL of the following criteria:**

1. 18 years of age or older

2. Flow cytometry demonstrates one of the following:
  - A. At least 10% PNH type III red cells
  - B. Greater than 50% of glycosylphosphatidylinositol-anchored proteins (GPI-AP)-deficient polymorphonuclear cells (PMNs)
3. At least one transfusion related to anemia secondary to PNH **OR** occurrence of a thromboembolic event
4. Has been vaccinated against meningococcal infection (at least 2 weeks prior to treatment, if not previously vaccinated) where and when clinically appropriate
5. Medication is prescribed by, or in consultation, with a hematologist

**Dosing.** The recommended intravenous dose for Paroxysmal nocturnal hemoglobinuria (PNH) is **ONE** of the following:

1. Induction: Up to 600 mg weekly for the first 4 weeks
2. Maintenance: Up to 900 mg at week 5, then up to 900 mg every 2 weeks thereafter

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When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Receipt of sample product does not satisfy any criteria requirements for coverage.

## Continuation of Therapy Criteria

Continuation of eculizumab is considered medically necessary for **ALL** covered diagnoses when initial criteria are met AND beneficial response is demonstrated by **ANY** of the following:

1. **Complement-mediated Hemolytic Uremic Syndrome (atypical hemolytic uremic syndrome):** Reduced hemolysis, improved thrombocytopenia or renal function
2. **Generalized Myasthenia Gravis:** Reductions in exacerbations of MG; improvements in speech, swallowing, mobility, and respiratory function, improvement in MG-ADL or QMG scores
3. **Neuromyelitis Optica Spectrum Disorder (NMOSD):** Reduction in relapse rate, reduction in symptoms (for example, pain, fatigue, motor function), or a slowing progression in symptoms
4. **Paroxysmal Nocturnal Hemoglobinuria (PNH):** Stabilization of hemoglobin levels, decreased transfusion requirements or transfusion independence, reductions in hemolysis

## Authorization Duration

Initial approval duration:

- **Complement-mediated Hemolytic Uremic Syndrome (atypical hemolytic uremic syndrome):** up to 6 months
- **Generalized Myasthenia Gravis:** up to 6 months
- **Neuromyelitis Optica Spectrum Disorder (NMOSD):** up to 12 months
- **Paroxysmal Nocturnal Hemoglobinuria (PNH):** up to 6 months

Reauthorization approval duration:

- **Complement-mediated Hemolytic Uremic Syndrome (atypical hemolytic uremic syndrome):** up to 12 months

- **Generalized Myasthenia Gravis:** up to 12 months
- **Neuromyelitis Optica Spectrum Disorder (NMOSD):** up to 12 months
- **Paroxysmal Nocturnal Hemoglobinuria (PNH):** up to 12 months

## Conditions Not Covered

Any other use is considered experimental, investigational, or unproven, including the following (this list may not be all inclusive; criteria will be updated as new published data are available):

1. Acute antibody mediated rejection
2. Chronic antibody-mediated rejection in recipients with persistently high B flow crossmatch after positive crossmatch kidney transplantation
3. **Concomitant Use with Empaveli > 4 Weeks.** Concomitant use of eculizumab with Empaveli is not recommended. However, to reduce the risk of hemolysis from abrupt treatment discontinuation in a patient switching from eculizumab to Empaveli, patient should use both therapies for 4 weeks; after which, eculizumab is discontinued and patient is continued on Empaveli monotherapy.
4. **Concomitant Use with Another Complement Inhibitor Except Voydeya (danicopan tablets).** There is no evidence to support concomitant use of eculizumab with another complement inhibitor, except Voydeya.  
Note: Examples of complement inhibitors are Fabhalta (iptacopan capsules), PiaSky (crovalimab-akkz intravenous infusion or subcutaneous injection), and Ultomiris (ravulizumab-cwvz intravenous infusion).
5. **Concomitant Use with a Rituximab Product, a Neonatal Fc Receptor Blocker, or Zilbrysq (zilucoplan subcutaneous injection).** There is no evidence to support concomitant use of eculizumab with a rituximab product, a neonatal Fc receptor blocker, or Zilbrysq.  
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).
6. **Concomitant Use with Enspryng (satralizumab-mwge subcutaneous injection) or Uplizna (inebilizumab-cdon intravenous infusion).** There is no evidence to support concomitant use of eculizumab with Enspryng or Uplizna
7. Geographic atrophy in age-related macular degeneration
8. Prevention of delayed graft function
9. Systemic lupus erythematosus
10. Stem cell transplant-associated thrombotic microangiopathy
11. Typical hemolytic uremic syndrome (HUS)

## Coding Information

**Note:** 1) This list of codes may not be all-inclusive.  
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

**Considered Medically Necessary when criteria in the applicable policy statements listed above are met:**

HCPSC Codes	Description
J1299	Injection, eculizumab, 2 mg (Code effective 4/1/2025)
J1300	Injection, eculizumab, 10 mg (Code effective until 3/31/2025)

HPCS Codes	Description
J1299	Injection, eculizumab, 2 mg (Code effective 4/1/2025)
Q5151	Injection, eculizumab-aagh (epysqli), biosimilar, 2 mg (Code effective 4/1/2025)
Q5152	Injection, eculizumab-aeeb (bkemv), biosimilar, 2 mg (Code effective 4/1/2025)

## Background

### OVERVIEW

Eculizumab, a complement C5 inhibitor, is indicated for the following uses:<sup>1</sup>

- **Atypical hemolytic uremic syndrome (aHUS)**, to inhibit complement-mediated thrombotic microangiopathy.  
Limitation of Use. Eculizumab is not indicated for the treatment of patients with Shiga toxin *Escherichia coli*-related hemolytic uremic syndrome.
- **Generalized myasthenia gravis (gMG)**, in adults and pediatric patients  $\geq 6$  years of age who are anti-acetylcholine receptor (AChR) antibody-positive.
- **Neuromyelitis optica spectrum disorder (NMOSD)**, in adults who are anti-aquaporin-4 (AQP4) antibody-positive.
- **Paroxysmal nocturnal hemoglobinuria (PNH)**, to reduce hemolysis.

Eculizumab has a Boxed Warning about serious meningococcal infections.<sup>1</sup> Soliris and biosimilars are only available through a restricted access program (Risk Evaluation and Mitigation Strategy [REMS]).

The safety and effectiveness of eculizumab for the treatment of PNH or NMOSD in pediatric patients have not been established.<sup>1</sup> The safety and effectiveness of eculizumab in pediatric patients for aHUS is supported by evidence from four adequate and well-controlled clinical studies assessing the safety and effectiveness of eculizumab for the treatment of aHUS. The safety and effectiveness of eculizumab in pediatric patients for gMG is supported by evidence from an adequate and well-controlled trial in adults with additional pharmacokinetic and safety data in pediatric patients with gMG who are  $\geq 12$  years of age, and pharmacokinetic and safety data in other pediatric populations 6 to  $< 12$  years of age.

For the gMG indication, eculizumab was studied in adults with gMG with anti-AChR antibodies with a Myasthenia Gravis Foundation of America (MGFA) clinical classification class II to IV and a Myasthenia Gravis-Activities of Daily Living (MG-ADL) total score  $\geq 6$ .<sup>1</sup>

### Disease Overview

Hemolytic uremic syndrome (HUS) is defined as the triad of non-immune hemolytic anemia, thrombocytopenia, and acute renal failure, in which the underlying lesions are mediated by systemic thrombotic microangiopathy.<sup>2</sup> aHUS should be distinguished from a more common condition referred to as typical HUS.<sup>4</sup> aHUS is a sub-type of HUS in which thrombotic microangiopathy is the consequence of endothelial damage in the microvasculature of the kidneys and other organs due to a dysregulation of the activity of the complement system. The typical form is caused by infection with certain strains of *E. coli* bacteria that produce toxic substances called Shiga-like toxins; eculizumab is not indicated for the treatment of Shiga toxin *E. coli*-related hemolytic uremic syndrome.<sup>1-3</sup>

Myasthenia gravis (MG) 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.<sup>4</sup> The hallmark of MG is muscle weakness that worsens after periods of activity and improves after periods of rest. Acquired MG results from the binding of autoantibodies to components of the neuromuscular junction, most commonly the AChR.<sup>5</sup>

NMOSD is a rare, relapsing, autoimmune disorder of the brain and spinal cord with optic neuritis and/or myelitis as predominate characteristic symptoms.<sup>6,7</sup> NMOSD often causes significant, permanent damage to vision and/or spinal cord function resulting in blindness or impaired mobility. Patients may experience pain, paralysis, loss of bowel and bladder control, loss of visual acuity, uncontrolled motor functions, and complications can cause death.

PNH is a rare, genetic disorder of hematopoietic stem cells.<sup>8,9</sup> The mutation in the X-linked gene phosphatidylinositol glycan class A (PIGA) results in a deficiency in the glycosylphosphatidylinositol (GPI) protein, which is responsible for anchoring other protein moieties to the surface of the erythrocytes. Loss of anchoring of these proteins causes cells to hemolyze and leads to complications such as hemolytic anemia, thrombosis, and peripheral blood cytopenias. PNH is a clinical diagnosis that should be confirmed with peripheral blood flow cytometry to detect the absence or severe deficiency of GPI-anchored proteins on at least two cell lineages.<sup>8,10</sup> Prior to the availability of complement inhibitors, only supportive measures, in terms of managing the cytopenias and controlling thrombotic risk, were available. Supportive measures include platelet transfusion, immunosuppressive therapy for patients with bone marrow failure, use of erythropoietin for anemias, and aggressive anticoagulation.

## Recommendations

There are no formal guidelines for treatment of aHUS.

A consensus statement for the diagnosis and treatment of PNH was published in 2021.<sup>8</sup> Treatment options for PNH are supportive care, allogeneic hematopoietic stem cell transplantation, and complement blockade by the anti-C5 monoclonal antibody (eculizumab). Supportive care include use of oral iron to replace the large urinary losses; folate and vitamin B<sub>12</sub> supplementation; red blood cell transfusion when these measures do not maintain adequate hemoglobin levels; use of antibiotics to treat bacterial infections as soon as possible since infections can exacerbate hemolytic crises in patients with PNH; use of corticosteroids to reduce the severity and duration of the hemolytic crises; use of eculizumab as primary prophylaxis in patients with high PNH clone size (granulocyte clone > 50%), high level of D dimer, pregnancy, perioperative condition, and other associated thrombophilia risk factors; and use of immunosuppressives in patients with PNH and aplastic anemia and bone marrow deficiency.

An international consensus guidance for the management of MG was published in 2016.<sup>5</sup> The consensus guidance recommends pyridostigmine for the initial treatment in most patients with MG. 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 MG who have not met treatment goals after an adequate trial of pyridostigmine. Nonsteroidal immunosuppressant agents used in MG 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 this consensus guidance provides new recommendations for methotrexate, rituximab, and eculizumab.<sup>11</sup> All recommendations should be considered extensions or additions to recommendations made in the initial international consensus guidance. Oral methotrexate may be considered as a steroid-sparing agent in patients with gMG who have not tolerated or responded to steroid-sparing agents. Rituximab should be considered as an early therapeutic option in patients with anti-muscle specific kinase antibody-positive MG who have an unsatisfactory response to initial immunotherapy. Eculizumab should be considered in the treatment of severe, refractory, anti-AChR antibody-positive MG.

*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.<sup>12</sup> 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.

The Neuromyelitis Optica Study Group (NEMOS) published revised recommendations for the treatment of NMOSD in 2024.<sup>13</sup> The standard of care for the treatment of NMOSD attacks (for both AQP4-IgG-positive and double-negative cases) are high-dose glucocorticoids and/or apheresis therapy. Long term immunotherapy is recommended for patients with AQP4-IgG-positive NMOSD. NEMOS notes the first-choice therapies for the treatment of AQP4-IgG-positive NMOSD are eculizumab, Ultomiris® (ravulizumab-cwyz intravenous infusion), Enspryng® (satralizumab-mwge subcutaneous injection), Uplizna® (inebilizumab-cdon intravenous infusion), and rituximab. The order of preference for these therapies is unclear and further comparative trials and real-world data are needed. The choice of treatment is dependent on several factors, including disease activity and severity, mode



and onset of action, possibility to combine it with immunosuppressive drugs, effect on autoimmune and other comorbidities, gender (family planning issues), frequency and route of administration, side effect profile, as well as patient and physician preference. In general, if a patient fails a first-choice treatment, another first-choice treatment should be tried; other options include use of a second-choice treatment (azathioprine, mycophenolate mofetil, low-dose oral glucocorticoids) or the addition of a second-choice treatment to the regimen.

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Revision Details

Type of Revision	Summary of Changes	Date
Selected Revision	<b>Conditions Not Covered:</b> <b>Updated from</b> "Concomitant Use with a Rituximab Product, a Neonatal Fc Receptor Blocker, Enspryng (satralizumab-mwge subcutaneous injection), Fabhalta (iptacopan capsule), Ultomiris (ravulizumab-cwvz intravenous infusion or subcutaneous injection), Uplizna (inebilizumab-cdon intravenous infusion), or Zilbrysq (zilucoplan subcutaneous injection). There is no evidence to support concomitant use of Soliris with a rituximab product, a neonatal Fc receptor blocker, Enspryng, Fabhalta, Ultomiris, Uplizna, or Zilbrysq. Examples	12/1/2024



	<p>of Neonatal Fc receptor blockers are: Vyvgart (efgartigimod alfa-fcab IV infusion), Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc SC injection), and Rystiggo (rozanolixizumab-noli SC infusion).” <b>to “Concomitant Use with Empaveli &gt; 4 Weeks.</b> Concomitant use of Soliris with Empaveli is not recommended. However, to reduce the risk of hemolysis from abrupt treatment discontinuation in a patient switching from Soliris to Empaveli, patient should use both therapies for 4 weeks; after which, Soliris is discontinued and patient is continued on Empaveli monotherapy; <b>Concomitant Use with Another Complement Inhibitor Except Voydeya (danicopan tablets).</b> There is no evidence to support concomitant use of Soliris with another complement inhibitor, except Voydeya. <u>Note:</u> Examples of complement inhibitors are Fabhalta (iptacopan capsules), PiaSky (crovalimab-akkz intravenous infusion or subcutaneous injection), and Ultomiris (ravulizumab-cwzy intravenous infusion); <b>Concomitant Use with a Rituximab Product, a Neonatal Fc Receptor Blocker, or Zilbrysq (zilucoplan subcutaneous injection).</b> There is no evidence to support concomitant use of Soliris with a rituximab product, a neonatal Fc receptor blocker, or Zilbrysq. <u>Note:</u> 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); <b>Concomitant Use with Enspryng (satralizumab-mwge subcutaneous injection) or Uplizna (inebilizumab-cdon intravenous infusion).</b> There is no evidence to support concomitant use of Soliris with Enspryng or Uplizna”</p>	
Selected Revision	<p><b>Updated HCPCS Coding</b>  <b>Added</b> new code J1299 that will be effective on 4/1/2025  <b>Added</b> that J1300 will be effective until 3/31/2025</p>	3/15/2025
Selected Revision	<p><b>Bkemv, Epysqli (biosimilars to Soliris):</b> These agents were added to the policy; the same criteria apply as that for Soliris.</p> <p><b>Generalized Myasthenia Gravis:</b></p> <ul style="list-style-type: none"> <li>– Age requirement was changed to “≥ 6 years of age”; previously it was “≥18 years of age”.</li> <li>– Criterion that addresses the Myasthenia Gravis Foundation of America classification and Myasthenia Gravis Activities of Daily Living score was changed such that this requirement only applies to patients ≥ 18 years of age.</li> <li>– Corticosteroid was added to the Note of examples of immunosuppressant therapies.</li> </ul> <p><b>Updated HCPCS Coding</b>  <b>Added:</b> Q5151 &amp; Q5152 (Codes effective 4/1/2025)</p>	5/15/2025

The policy effective date is in force until updated or retired.

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