

Eculizumab Products (Soliris[®], Bkemy[™], Epysqli[®])

When requesting an eculizumab product (Soliris[®], Bekemy[™], Epysqli[®]), the individual requiring treatment must be diagnosed with one of the following FDA-approved indications and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-Approved Indications

Paroxysmal Nocturnal Hemoglobinuria (PNH)

Eculizumab is indicated for the treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis.

Atypical Hemolytic Uremic Syndrome (aHUS)

Eculizumab is indicated for the treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy.

Generalized Myasthenia Gravis (gMG)

Eculizumab is indicated for the treatment of generalized myasthenia gravis (gMG) in adult and pediatric patients six years of age and older who are anti-acetylcholine receptor (AChR) antibody positive.

Neuromyelitis Optica Spectrum Disorder (NMOSD)

Eculizumab is indicated for the treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive.

Coverage Guidelines

Atypical Hemolytic Uremic Syndrome (aHUS)

The individual must meet **both** of the following criteria for authorization:

- Does not have Shiga toxin *Escherichia coli*-related hemolytic uremic syndrome;
- Eculizumab is prescribed by or in consultation with a nephrologist.

Approval duration: 12 months

Paroxysmal Nocturnal Hemoglobinuria (PNH)

The individual must meet **all** of the following criteria for initial authorization:

- Is 18 years of age or older;
- Diagnosis of PNH was confirmed by peripheral blood flow cytometry results showing the absence or deficiency of glycosylphosphatidylinositol (GPI)-anchored proteins on at least two cell lineages;
- Eculizumab is prescribed by or in consultation with a hematologist.

The individual must meet **all** of the following criteria for re-authorization:

- Is 18 years of age or older;
- Is continuing to receive benefit from eculizumab therapy (e.g., stabilization of hemoglobin levels, decreased transfusion requirements or transfusion independence, reduction in hemolysis, improvement in Functional Assessment of Chronic Illness Therapy [FACIT]-Fatigue score);
- Eculizumab is prescribed by or in consultation with a hematologist.

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

Generalized Myasthenia Gravis (gMG)

The individual must meet **all** of the following criteria for initial authorization:

- Is 6 years of age or older;
- For 18 years of age or older, must meet both of the following:
 - o Has Myasthenia Gravis Foundation of America classification of II to IV;
 - o Has Myasthenia Gravis Activities of Daily Living (MG-ADL) score of 6 or higher;
- Has a confirmed anti-acetylcholine receptor (AChR) antibody positive generalized myasthenia gravis;
- Has received or is currently receiving pyridostigmine unless failed, contraindicated or intolerant to pyridostigmine;
- Has received or is currently receiving two different immunosuppressive therapies for at least one year unless failed, contraindicated or intolerant to two different immunosuppressive therapies (e.g., azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, tacrolimus, cyclophosphamide);
- Has evidence of unresolved symptoms of generalized myasthenia gravis (e.g., difficulty swallowing, difficulty breathing, or a functional disability [e.g., double vision, speech, impairment of mobility]);
- Eculizumab is prescribed by or in consultation with a neurologist.

The individual must meet **all** of the following criteria for re-authorization:

- Is 6 years of age or older;
- Is continuing to receive benefits from eculizumab therapy (e.g., reductions in exacerbations of myasthenia gravis; improvement in speech, swallowing, mobility, or respiratory functions);
- Eculizumab is prescribed by or in consultation with a neurologist.

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

Neuromyelitis Optica Spectrum Disorder (NMOSD)

The individual must meet **all** of the following criteria for initial authorization:

- Is 18 years of age or older;

- Neuromyelitis optica spectrum disorder diagnosis was confirmed by a positive blood serum test for anti-aquaporin-4 antibody;
- Eculizumab is prescribed by or in consultation with a neurologist.

The individual must meet **all** of the following criteria for re-authorization:

- Is 18 years of age or older;
- Neuromyelitis optica spectrum disorder diagnosis was confirmed by a positive blood serum test for anti-aquaporin-4 antibody;
- Is continuing to receive benefit from eculizumab therapy (e.g., reduction in relapse rates, reduction in symptoms [e.g., pain, fatigue, motor function], or a slowing progression in symptoms);
- Eculizumab is prescribed by or in consultation with a neurologist.

Approval duration (initial and renewal): 12 months

Dosing Recommendations

Eculizumab is only administered as an intravenous infusion.

For patients 18 years of age and older

Paroxysmal Nocturnal Hemoglobinuria (PNH)

- 600 mg weekly for the first 4 weeks, followed by
- 900 mg for the fifth dose 1 week later, then
- 900 mg every 2 weeks thereafter.

Atypical Hemolytic Uremia Syndrome (aHUS), Generalized Myasthenia Gravis (gMG) and Neuromyelitis Optica Spectrum Disorder (NMOSD)

- 900 mg weekly for the first 4 weeks, followed by
- 1200 mg for the fifth dose 1 week later, then
- 1200 mg every 2 weeks thereafter.

For patients less than 18 years of age

Atypical Hemolytic Uremia Syndrome (aHUS) and Generalized Myasthenia Gravis (gMG)

Body weight	Induction	Maintenance
40 kg and over	900 mg weekly x 4 doses	1200 mg at week 5; then 1200 mg every 2 weeks
30 kg to less than 40 kg	600 mg weekly x 2 doses	900 mg at week 3; then 900 mg every 2 weeks
20 kg to less than 30 kg	600 mg weekly x 2 doses	600 mg at week 3; then 600 mg every 2 weeks

10 kg to less than 20 kg	600 mg weekly x 1 dose	300 mg at week 2; then 300 mg every 2 weeks
5 kg to less than 10 kg	300 mg weekly x 1 dose	300 mg at week 2; then 300 mg every 3 weeks

References

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3. Epysqli [prescribing information]. Yeonsu-gu, Incheon: Samsung Bioepis Co., Ltd. November 2024.
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