

Soliris® (eculizumab)



When requesting Soliris® (eculizumab), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved Indications

Soliris is a complement inhibitor indicated for:

- The treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis.
- The treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy.
- The treatment of adult patients with generalized myasthenia gravis (gMG) who are anti-acetylcholine receptor (AChR) antibody positive.
- The treatment of neuromyelitis optica spectrum disorder (NMOSD) in adults who are anti-aquaporin-4 (AQP4) antibody positive.

Coverage Guidelines

Atypical Hemolytic Uremic Syndrome (aHUS)

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

- The individual does not have Shiga toxin *Escherichia coli* related hemolytic uremic syndrome;
- Soliris 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:

- The diagnosis of PNH has been confirmed by peripheral blood flow cytometry results showing the absence or deficiency of glycosylphosphatidylinositol (GPI)-anchored proteins on at least 2 cell lineages;
- The individual is 18 years of age or older;
- Soliris is prescribed by or in consultation with a hematologist.

For re-authorization, the individual must meet **all** of the following criteria:

- Is 18 years of age or older;
- The individual is continuing to receive benefit from Soliris therapy (e.g., stabilization of hemoglobin levels, decreased transfusion requirement or transfusion independence, reduction in hemolysis);
- Soliris 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:

- Has a confirmed diagnosis of anti-acetylcholine receptor (AChR) antibody positive generalized myasthenia gravis;
- Is 18 years of age or older;
- Meets both of the following:
 - Myasthenia Gravis Foundation of America classification of II to IV;
 - Myasthenia Gravis Activities of Daily Living (MG-ADL) score of 5 or higher;
- Meets one of the following:
 - Is currently receiving pyridostigmine; OR
 - Has had inadequate efficacy, a contraindication, or significant intolerance to pyridostigmine;
- Meets one of the following:
 - Has received or is currently receiving two different immunosuppressive therapies for at least 1 year; OR
 - Has had inadequate efficacy, a contraindication, or significant intolerance to two different immunosuppressive therapies (e.g., azathioprine, cyclosporine, mycophenolate mofetil, methotrexate, tacrolimus, cyclophosphamide);
- Has evidence of unresolved symptoms of generalized myasthenia gravis, such as difficulty swallowing, difficulty breathing, or a functional disability (e.g., double vision, impairment of mobility);
- Soliris is prescribed by or in consultation with a neurologist.

For re-authorization, the individual must meet **all** of the following criteria:

- Is 18 years of age or older;
- The individual is continuing to receive benefits from Soliris therapy (e.g., reductions in exacerbations of myasthenia gravis; improvement in speech, swallowing, mobility, or respiratory functions);
- Soliris is prescribed by or in consultation with a neurologist.

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

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Neuromyelitis Optica Spectrum Disorder (NMOSD)

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

- Diagnosis has been confirmed by blood serum test for anti-aquaporin-4 antibody positive NMOSD;
- Is 18 years of age or older;
- Meets one of the following:
 - Has tried either Enspryng (satralizumab-mwge) or Uplizna (inebilizumab-cdon); OR
 - Has previously tried or is currently taking two of the following therapies: azathioprine, corticosteroid, mycophenolate mofetil, or rituximab;
- The patient has a history of at least 1 relapse in the last 12 months or two relapses in the last 2 years;
- Soliris is prescribed by or in consultation with a neurologist.

For re-authorization, the individual must meet **all** of the following criteria:

- Diagnosis has been confirmed by blood serum test for anti-aquaporin-4 antibody positive NMOSD;
- Is 18 years of age or older;
- The individual is continuing to receive benefit from Soliris therapy (e.g., reduction in relapse rates, reduction in symptoms [pain, fatigue, motor function], or a slowing progression in symptoms);
- Soliris is prescribed by or in consultation with a neurologist.

Approval duration (initial and renewal): 12 months

Dosing Recommendations

Soliris is only administered as an intravenous infusion.

Paroxysmal nocturnal hemoglobinuria (PHN)

- 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) *(for patients 18 years of age and older)*

- 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.

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Atypical hemolytic uremia syndrome (aHUS) (for patients less than 18 years of age)

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

Generalized myasthenia gravis (gMG)

- 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.

Neuromyelitis optica spectrum disorder (NMOSD)

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

References

1. Soliris® intravenous infusion [prescribing information]. New Haven, CT: Alexion; April 2021.
2. Campistol JM, Arias M, Ariceta G, et al. An update for atypical haemolytic uraemic syndrome: diagnosis and treatment. A consensus document. *Nefrologia*. 2015;35:421–447.
3. Genetics Home Reference. Atypical hemolytic-uremic syndrome. National Institutes of Health (NIH). Available at: <https://ghr.nlm.nih.gov/condition/atypical-hemolytic-uremic-syndrome#sourcesforpage>. Accessed on April 29, 2022.
4. National Institute of Neurological Disorders and Stroke (NINDS). Myasthenia Gravis Fact Sheet. National Institutes of Health (NIH) Publication No. 17-768. Publication last updated: November 15, 2021. Available at: <https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Myasthenia-Gravis-Fact-Sheet>. Accessed on April 29, 2022.
5. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis. *Neurology*. 2016;87:419–425.
6. National Organization for Rare Disorders. Neuromyelitis Optica Spectrum Disorder. Available at: <https://rarediseases.org/rare-diseases/neuromyelitis-optica/>. Accessed April 29, 2022.
7. National Institute of Health, U.S. National Library of Medicine. Genetics Home Reference. Neuromyelitis optica. Available at: <https://ghr.nlm.nih.gov/condition/neuromyelitis-optica#genes>. Accessed April 29, 2022.
8. Enspryng™ subcutaneous injection [prescribing information]. South San Francisco, CA: Genentech; August 2020.
9. Uplizna™ intravenous infusion [prescribing information]. Gaithersburg, MD: Viela Bio; July 2021.
10. Bradshaw MJ, Kimbrough D. Neuromyelitis Optica Spectrum Disorders. *Practical Neurology*. 2019;76-84.
11. Siegel Rare Neuroimmune Association. Neuromyelitis Optica Spectrum Disorders. https://wearesma.org/wp-content/uploads/2018/06/About_NMOSD_2018.pdf. Accessed on April 29, 2022.
12. Hillmen P, Szer J, Weitz I, et al. Pegcetacoplan versus eculizumab in paroxysmal nocturnal hemoglobinuria. *N Engl J Med*. 2021;384(11):1028-1037.
13. Parker CJ. Update on the diagnosis and management of paroxysmal nocturnal hemoglobinuria. *Hematology Am Soc Hematol Edu Program*. 2016;2016(1):208-216.
14. Roth A, Maciejewski J, Nishinura JI, et al. Screening and diagnostic clinical algorithm for paroxysmal nocturnal hemoglobinuria: Expert consensus. *Eur J Haematol*. 2018;101(1):3-11.
15. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthenia Gravis: 2020 Update. *Neurology*. 2021 Jan 19;96(3):114-122.