# Soliris (eculizumab)

## NOTICE

This policy contains information which is clinical in nature. The policy is not medical advice. The information in this policy is used by Wellmark to make determinations whether medical treatment is covered under the terms of a Wellmark member's health benefit plan. Physicians and other health care providers are responsible for medical advice and treatment. If you have specific health care needs, you should consult an appropriate health care professional. If you would like to request an accessible version of this document, please contact customer service at 800-524-9242.

## BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

## DESCRIPTION

The intent of the Soliris (eculizumab) drug policy is to ensure appropriate selection of patients for therapy based on product labeling, clinical guidelines, and clinical studies. The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

A. Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis
B. Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy
C. Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AchR) antibody positive
D. Neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive

Limitations of Use

Soliris is not indicated for the treatment of patients with Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS).

## POLICY

Required Documentation

Submission of the following information is necessary to initiate the prior authorization review for new requests for treatment of:

A. For initial requests:

1. Atypical hemolytic uremic syndrome: ADAMTS 13 level
2. Paroxysmal nocturnal hemoglobinuria: flow cytometry used to show results of GPI-APs deficiency
3. Generalized myasthenia gravis: anti-acetylcholine receptor (AchR) antibody positive, clinical classification of myasthenia gravis score, MG activities of daily living score, use of IVIG and rituximab, use of two immunosuppressive therapies
4. Neuromyelitis optica spectrum disorder: immunoassay used to confirm anti-aquaporin-4 (AQP4) antibody is present

B. For continuation requests: Chart notes or medical record documentation supporting positive clinical response.

Criteria for Initial Approval

**A. Atypical hemolytic uremic syndrome**

Authorization of 6 months may be granted for treatment of atypical hemolytic uremic syndrome not caused by Shiga toxin when all of the following criteria are met:

1. ADAMTS 13 activity level above 5%
2. Absence of Shiga toxin

**B. Paroxysmal nocturnal hemoglobinuria**

Authorization of 6 months may be granted for treatment of paroxysmal nocturnal hemoglobinuria (PNH) when all of the following criteria are met:

1. The diagnosis of PNH was confirmed by detecting a deficiency of glycosylphosphatidylinositol-anchored proteins (GPI-APs) as demonstrated by either of the following:
   a.) At least 5% PNH cells
   b.) At least 51% of GPI-anchored protein deficient poly-morphonuclear cells
2. Flow cytometry is used to demonstrate GPI-anchored proteins deficiency

**C. Generalized myasthenia gravis (gMG)**

Authorization of 6 months may be granted for treatment of generalized myasthenia gravis (gMG) when all of the following criteria are met:

1. Anti-acetylcholine receptor (AchR) antibody positive
2. Myasthenia Gravis Foundation of America (MGFA) clinical classification II to IV
3. MG activities of daily living (MG-ADL) total score ≥6
4. Meets both of the following:
   a). Member has had an inadequate response to at least two immunosuppressive therapies listed below:
      i. azathioprine
      ii. cyclosporine
      iii. mycophenolate mofetil
      iv. tacrolimus
      v. methotrexate
      vi. cyclophosphamide
      vii. rituximab
   b). Member has inadequate response to chronic IVIG

**D. Neuromyelitis Optica Spectrum Disorder (NMOSD)**

Authorization of 6 months may be granted for treatment of neuromyelitis optica spectrum disorder (NMOSD) in adults when all of the following criteria are met:

1. Anti-aquaporin-4 (AQP4) antibody positive

2. The medication is being prescribed by, or in consultation with, a neurologist
3. Member exhibits one of the following core clinical characteristics of NMOSD:
   a). Optic neuritis
   b). Acute myelitis
   c). Area postrema syndrome (episode of otherwise unexplained hiccups or nausea and vomiting)
   d). Acute brainstem syndrome
   e). Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
   f). Symptomatic cerebral syndrome with NMOSD-typical brain lesions
4. Member has a history of one of the following:
   a). Two or more relapses that required rescue therapy in the last 12 months prior to initiating therapy
   b). Three relapses in the past 24 months that required rescue therapy, with at least 1 relapse in the 12 months prior to initiating therapy
5. Member has had an inadequate response, intolerable adverse event, or documented contraindication to rituximab therapy AND Enspryng (satralizumab-mwge) therapy
6. The member will not receive the requested drug concomitantly with any of the following:
   a). Other complement-inhibitors (i.e., ravulizumab)
   b). Anti-CD20 therapy (i.e., rituximab)
   c). Anti-CD19 antibody (i.e., inebilizumab-cdon)
   d). IL-6 receptor antagonist (i.e., satralizumab-mwge)

Continuation of Therapy

**A. Atypical hemolytic uremic syndrome**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen and they demonstrate a positive response to therapy (e.g., normalization of lactate dehydrogenase (LDH) levels, platelet counts).

**B. Paroxysmal nocturnal hemoglobinuria**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen and they demonstrate a positive response to therapy (e.g., improvement in hemoglobin levels, normalization of lactate dehydrogenase [LDH] levels).

**C. Generalized myasthenia gravis (gMG)**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen and they demonstrate a positive response to therapy (e.g., improvement in MG-ADL score, changes compared to baseline in Quantitative Myasthenia Gravis (QMG) total score).

**D. Neuromyelitis optica spectrum disorder (NMOSD)**

Authorization of 12 months may be granted for continued treatment of neuromyelitis optica spectrum disorder (NMOSD) in anti-aquaporin-4 (AQP4) antibody positive adults requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen, they demonstrate a positive clinical response to therapy from baseline as demonstrated by a reduction in the number and/or severity of relapses, and the medication is being prescribed by, or in consultation with, a neurologist.

Soliris is considered **not medically necessary** for members who do not meet the criteria set forth above.

Dosage and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

## PROCEDURES AND BILLING CODES

**To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnostic codes.**

- J1300 Injection, eculizumab, (Soliris), 10 mg

## REFERENCES


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- Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders*. Neurology.* 2015; 85:177-189.


## POLICY HISTORY

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