AETNA BETTER HEALTH®
Coverage Policy/Guideline

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| Name: | Soliris | Page: | 1 of 4 |
| Effective Date: | 2/26/2025 | Last Review Date: | 1/30/2025 |
| Applies to: | ☒New Jersey | | |

## Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Soliris under the patient's prescription drug benefit.

## Description:

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. *(Reference NJ State Protocol for Paroxysmal Nocturnal Hemoglobinuria (PNH) Products).*
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).*

All other indications are considered experimental/investigational and not medically necessary.

## Applicable Drug List:

Bkemv
Epysqli
Soliris

## Policy/Guideline:

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

A. Initial requests:
    1. Atypical hemolytic uremic syndrome: ADAMTS 13 level
    2. Generalized myasthenia gravis: anti-acetylcholine receptor (AchR) antibody positive, clinical classification of myasthenia gravis score, MG activities of daily living score, use of IVIG, use of two immunosuppressive therapies
    3. Neuromyelitis optica spectrum disorder: immunoassay used to confirm anti-aquaporin-4 (AQP4) antibody is present

B. 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 the following criteria are met:
1. ADAMTS 13 activity level above 5%
2. Absence of Shiga toxin
3. The requested medication will not be used in combination with another complement inhibitor (e.g., Ultomiris) for the treatment of aHUS.

**B. 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 of greater than or equal to 5
4. Meets ONE of the following:
   i. Member has had an inadequate response or intolerable adverse event to at least TWO immunosuppressive therapies over the course of at least 12 months (e.g., azathioprine, corticosteroids, cyclosporine, methotrexate, mycophenolate, tacrolimus)
   ii. Member has had an inadequate response or intolerable adverse event to at least ONE immunosuppressive therapy and intravenous immunoglobulin (IVIG) over the course of at least 12 months
   iii. Member has a documented clinical reason to avoid therapy with immunosuppressive agents and IVIG
5. The requested medication will not be used in combination with another complement inhibitor (e.g., Ultomiris, Zilbrysq) or neonatal Fc receptor blocker (e.g., Vyvgart, Vyvgart Hytrulo, Rystiggo).

**C. Neuromyelitis Optica Spectrum Disorder (NMOSD)**
Authorization of 6 months may be granted for treatment of neuromyelitis optica spectrum disorder (NMOSD) when ALL the following criteria are met:
1. Anti-aquaporin-4 (AQP4) antibody positive
2. 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

3. The member will not receive the requested drug concomitantly with other biologics for the treatment of NMOSD.

**Continuation of Therapy:**

**A. Atypical hemolytic uremic syndrome**
Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when all of the following criteria are met:
1. There is no evidence of unacceptable toxicity or disease progression while on the current regimen.
2. The member demonstrates a positive response to therapy (e.g., normalization of lactate dehydrogenase (LDH) levels, platelet counts).
3. The requested medication will not be used in combination with another complement inhibitor (e.g., Ultomiris) for the treatment of aHUS.

**B. Generalized myasthenia gravis (gMG)**
Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when ALL the following criteria are met:
1. There is no evidence of unacceptable toxicity or disease progression while on the current regimen.
2. The member demonstrates a positive response to therapy (e.g., improvement in MG-ADL score, MG Manual Muscle Test (MMT), MG Composite).
3. The requested medication will not be used in combination with another complement inhibitor (e.g., Ultomiris, Zilbrysq) or neonatal Fc receptor blocker (e.g., Vyvgart, Vyvgart Hytrulo, Rystiggo).

**C. Neuromyelitis optica spectrum disorder (NMOSD)**
Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when ALL the following criteria are met:
1. There is no evidence of unacceptable toxicity or disease progression while on the current regimen.
2. The member demonstrates a positive response to therapy (e.g., reduction in number of relapses).
3. The member will not receive the requested drug concomitantly with other biologics for the treatment of NMOSD.

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

## Approval Duration and Quantity Restrictions:

**Approval:**

- Initial Requests: 6 months
- Renewals: 12 months

## References:

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13. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015; 85:177-189.
14. Parker CJ. Update on the diagnosis and management of paroxysmal nocturnal hemoglobinuria. Hematology Am Soc Hematol Educ Program. 2016;2016(1):208-216.
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