MASSACHUSETTS

Blue Cross Blue Shield of Massachusetts is an Independent Licensee of the Blue Cross and Blue Shield Association

Pharmacy Medical Policy

Soliris, Ultomiris, and Neuromyelitis Optica Policy

Table of Contents

Policy Number: 093
BCBSA Reference Number: None

Related Policies

- Quality Care Dosing guidelines apply to the following medications and can be found in Medical Policy #**621A**

Policy

Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity

Note: All requests for outpatient retail pharmacy for indications listed and not listed on the medical policy guidelines may be submitted to BCBSMA Clinical Pharmacy Operations by completing the Prior Authorization Form on the last page of this document. Physicians may also call BCBSMA Pharmacy Operations department at (800)366-7778 to request a prior authorization/formulary exception verbally. Patients must have pharmacy benefits under their subscriber certificates.

Please refer to the chart below for the formulary and step status of the medications affected by this policy.

Standard Formulary	
Drug	**Formulary Status**
Empaveli ™ (pegcetacoplan)	PA Required
Enspryng ™ (satralizumab)	PA Required
Soliris ®** (eculizumab)	PA Required
Ultomiris ®** (ravulizumab)	PA Required
Uplizna ™** (Inebilizumab)	PA Required

** - This Drug is part of Medications covered only under the pharmacy benefit only program. This program does not apply when the medication is administered: in the emergency room, as an inpatient, at a surgical day care facility, in an ambulatory surgery-center, or through home infusion therapy or dialysis.

We may cover Empaveli™ (pegcetacoplan) for the treatment of Paroxysmal nocturnal hemoglobinuria (PNH) when **all** of the following criteria are met:

- Confirmed diagnosis of Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
- 18 years of age or older, **AND**
- Documented baseline value for serum lactate dehydrogenase (LDH) is ≥1.5 times the upper limit of normal, **AND**
- **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), Empaveli™ (pegcetacoplan), or with Soliris® (eculizumab), **AND**
- Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]) [1]
- Prescribing physician is enrolled in Empaveli REMS program

*It is also recommended that patients receive vaccinations for pneumococcus and haemophilus but will not be required for approval.

1 - For patients switching from Soliris ® (eculizumab), dual coverage with Empaveli™ is permitted for a 4-week period. After 4 weeks, Soliris ® (eculizumab) should be discontinued before continuing monotherapy with Empaveli™.

Prior – Approval *Continuation* Requirements for Empaveli

1. Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
2. Confirmed diagnosis of Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
3. 18 years of age or older, **AND**
4. Decrease in serum LDH from pretreatment baseline, **AND**
5. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), Empaveli™ (pegcetacoplan), or with Soliris® (eculizumab), **AND**
6. Prescribing physician is enrolled in Empaveli REMS program, **AND**
7. Absence of unacceptable toxicity from the drug

We may cover Soliris®* (eculizumab) **OR we may cover** Ultomiris®* (ravulizumab) when **all** of the following criteria are met:

1. Paroxysmal nocturnal hemoglobinuria (PNH)
   a. 18 years of age or older or for Ultomiris the age can include one month of age or older.
   b. Documented baseline value for serum lactate dehydrogenase (LDH) is ≥1.5 times the upper limit of normal.
   c. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab) in combination with Soliris® (eculizumab)
   d. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed])
   e. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program

   *It is also recommended that patients receive vaccinations for pneumococcus and haemophilus but will not be required for approval.

2. Atypical hemolytic uremic syndrome (aHUS)
    a. Documented baseline value for serum lactate dehydrogenase (LDH) ≥ Upper Level of Normal (ULN) unless the patient has been receiving plasma exchange
    b. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab) in combination with Soliris® (eculizumab)
    c. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]
    d. Does **NOT** have Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS)
    e. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program

    *It is also recommended that patients receive vaccinations for pneumococcus and haemophilus but will not be required for approval.

We may cover Soliris®* (eculizumab) when **all** of the following criteria are met:

I. Myasthenia Gravis (gMG)
    a. 18 years of age or older
    b. Positive serologic test for anti-AChR antibodies
    c. Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
    d. Documented baseline MG-Activities of Daily Living (MG-ADL) total score ≥ 6 (*https://info.soliris.net/wp-content/uploads/2017/12/Assessment_Tool_Booklet.pdf*)
    e. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]
    f. Prescribing physician is enrolled in Soliris REMS program.

    *It is also recommended that patients receive vaccinations for pneumococcus and haemophilus but will not be required for approval.

II. Neuromyelitis optica spectrum disorder (NMOSD)
    a. 18 years of age or older
    b. Anti-aquaporin-4 (AQP4) antibody positive
    c. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]
    d. Prescribing physician is enrolled in Soliris REMS program.

    *It is also recommended that patients receive vaccinations for pneumococcus and haemophilus but will not be required for approval.

Prior – Approval *Continuation* Requirements
Diagnoses

1. Paroxysmal nocturnal hemoglobinuria (PNH)
    a. 18 years of age or older
    b. Decrease in serum LDH from pretreatment baseline
    c. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), Empaveli™ (pegcetacoplan), or with Soliris® (eculizumab)
    d. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program
    e. Absence of unacceptable toxicity from the drug

2. Atypical hemolytic uremic syndrome (aHUS)
    a. Decrease in serum LDH from pretreatment baseline
    b. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab) in combination with Soliris® (eculizumab)
    c. Does **NOT** have Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS)

d. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program
e. Absence of unacceptable toxicity from the drug

3. Myasthenia Gravis (gMG)
   a. 18 years of age or older
   b. Decrease of (MG-ADL) total score from baseline (*https://info.soliris.net/wp-content/uploads/2017/12/Assessment_Tool_Booklet.pdf*)
   c. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program
   d. Absence of unacceptable toxicity from the drug

4. Neuromyelitis optica spectrum disorder (NMOSD)
   a. 18 years of age or older
   b. Patient has had fewer relapses while on Soliris therapy
   c. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program
   d. Absence of unacceptable toxicity from the drug

We may Uplizna™ (inebilizumab) or Enspryng™ (satralizumab) for the treatment of neuromyelitis optica spectrum disorder (NMOSD) when **all** of the following criteria are met:

- Confirmed diagnosis of Neuromyelitis optica spectrum disorder (NMOSD), **AND**
- 18 years of age or older, **AND**
- Anti-aquaporin-4 (AQP4) antibody positive

For non-formulary/non-covered medications, requests must meet criteria above and the member must have had a previous treatment failure with or a contraindication to two covered formulary alternatives when available.

We do not cover the medications listed above for other conditions not listed above.

CPT Codes / HCPCS Codes / ICD-9 Codes

The following codes are included below for informational purposes. Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.

Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.

The above medical necessity criteria MUST be met for the following codes to be covered for Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity:

HCPCS Codes

HCPCS codes:	Code Description
J1823	Injection, inebilizumab-cdon(UPLIZNA), 1 mg
J1300	Injection, eculizumab, 10 mg (Soliris)
J1303	Injection, ravulizumab-cwvz, 10 mg (Ultomiris)
J3590, C9399	Not otherwise Classified Code (Empaveli)

Individual Consideration

All our medical policies are written for the majority of people with a given condition. Each policy is based on medical science. For many of our medical policies, each individual's unique clinical circumstances may be considered in light of current scientific literature. Physicians may send relevant clinical information for individual patients for consideration to:

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department
25 Technology Place
Hingham, MA 02043
Tel: 1-800-366-7778
Fax: 1-800-583-6289

Prior Authorization Information

Outpatient

For services described in this policy, see below for products where prior authorization **IS REQUIRED** if the procedure is performed **outpatient**.

	Outpatient
Commercial Managed Care (HMO and POS)	Prior authorization is **required**.
Commercial PPO	Prior authorization is **required**.

Policy History

Date	Action
11/2021	Updated to clarify Empaveli ™ criteria.
8/2021	Updated to include age update for Ultomiris ® and to add Empaveli ™ to the policy.
2/2021	Updated vaccination requirements for Soliris ®.
1/2021	Updated to include Enspryng™ and to change the policy name again.
9/2020	Updated to include Uplizna™ and to change the policy name.
7/2020	Implement a policy for both Ultomiris ® & Soliris ®

References

1. Soliris ® [package insert]. Boston, MA: Alexion Pharmaceuticals, Inc.: 7/2019.
2. Ultomiris ® [package insert]. Boston, MA: Alexion Pharmaceuticals, Inc.: 10/2019.
3. Uplizna ™ [package insert]. Gaithersburg, MD: Viela Bio, Inc.: 6/2020.

4. Enspryng ™ [package insert]. South San Francisco, CA: Genentech, Inc.: 8/2020.
5. Empaveli ™ [package insert]. Waltham, MA: Apellis Pharmaceuticals, Inc.: 8/2021.

To request prior authorization using the Massachusetts Standard Form for Medication Prior Authorization Requests (eForm), click the link below:

http://www.bluecrossma.org/medical-policies/sites/g/files/csphws2091/files/acquiadam-assets/023%20E%20Form%20medication%20prior%20auth%20instruction%20prn.pdf