Pharmacy Medical Policy

**Soliris, Ultomiris, Myasthenia Gravis, and Neuromyelitis Optica Policy**

**Table of Contents**
- Policy: Commercial
- Policy: Medicare
- Coding Information
- Policy History
- Information Pertaining to All Policies
- References
- Endnotes
- Forms

**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.
Prior Authorization Information

☒ Prior Authorization
☐ Step Therapy
☒ Quality Care Dosing

Pharmacy (Rx) or Medical (MED) benefit coverage
☒ Rx
☒ MED

Policy applies to Commercial Members:
- Managed Care (HMO and POS),
- PPO and Indemnity
- MEDEX with Rx plan
- Managed Major Medical with Custom BCBSMA Formulary
- Comprehensive Managed Major Medical with Custom BCBSMA Formulary
- Managed Blue for Seniors with Custom BCBSMA Formulary

Pharmacy Operations:
Tel: 1-800-366-7778
Fax: 1-800-583-6289

Policy last updated 9/2023

To request for coverage: Providers may call, fax, or mail the attached form (Formulary Exception/Prior Authorization form) to the address below.

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department
25 Technology Place
Hingham, MA 02043

Individual Consideration: Policy for requests that do not meet clinical criteria of this policy, see section labeled Individual Consideration

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

<table>
<thead>
<tr>
<th>Drug</th>
<th>Formulary Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Empaveli ™ (pegcetacoplan)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Enspryng ™** (satralizumab)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Soliris ® (eculizumab)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Ultomiris ® (ravulizumab)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Uplizna ™ (Inebilizumab)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Vyvgart ® (efgartigimod alfa)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Vyvgart ® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)</td>
<td>PA Required</td>
</tr>
</tbody>
</table>

** - 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

3. 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
   e. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]
   f. Prescribing physician is enrolled in Soliris REMS program and or the Ultomiris REMS program
g. Previous treatment with or contraindication to Vyvgart ® (efgartigimod alfa) or Vyvgart ®

Hytrulo (efgartigimod alfa and hyaluronidase-qvfc).

*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. 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

We may cover Vyvgart® (efgartigimod alfa) or Vyvgart® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) for the treatment of generalized myasthenia gravis (gMG) when all of the following criteria are met:

- 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 ≥ 5

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:

<table>
<thead>
<tr>
<th>HCPCS codes:</th>
<th>Code Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>J1823</td>
<td>Injection, inebilizumab-cdon(UPLIZNA), 1 mg</td>
</tr>
<tr>
<td>J1300</td>
<td>Injection, eculizumab, 10 mg (Soliris)</td>
</tr>
<tr>
<td>J1303</td>
<td>Injection, ravulizumab-cwzw, 10 mg (Ultomiris)</td>
</tr>
<tr>
<td>J3590, C9399</td>
<td>Not otherwise Classified Code (Empaveli)</td>
</tr>
</tbody>
</table>

Individual Consideration

Our medical policies are written for most people with a given condition. Each policy is based on peer reviewed clinical evidence. We also take into consideration the needs of atypical patient populations and diagnoses.

If the coverage criteria outlined is unlikely to be clinically effective for the prescribed purpose, the health care provider may request an exception to cover the requested medication based on an individual’s unique clinical circumstances. This is also referred to as "individual consideration" or an "exception request."

Some reasons why you may need us to make an exception include: therapeutic contraindications; history of adverse effects; expected to be ineffective or likely to cause harm (physical, mental, or adverse reaction).

To facilitate a thorough and prompt review of an exception request, we encourage the provider to include additional supporting clinical documentation with their request. This may include:

- Clinical notes or supporting clinical statements.
• The name and strength of formulary alternatives tried and failed (if alternatives were tried) and specifics regarding the treatment failure, if applicable.
• Clinical literature from reputable peer reviewed journals.
• References from nationally recognized and approved drug compendia such as American Hospital Formulary Service® Drug Information (AHFS-DI), Lexi-Drug, Clinical Pharmacology, Micromedex or Drugdex®, and
• References from consensus documents and/or nationally sanctioned guidelines.

Providers may call, fax or mail relevant clinical information, including clinical references for individual patient consideration, to:

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

We may also use prescription claims records to establish prior use of formulary alternatives or to show if step therapy criteria has been met. We will require the provider to share additional information when prescription claims data is either not available or the medication fill history fails to establish use of preferred formulary medications or that step therapy criteria has been met.

Policy History

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>9/2023</td>
<td>Updated to add Vyvgart ® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) to the policy and updated IC to align with 118E MGL § 51A.</td>
</tr>
<tr>
<td>7/2023</td>
<td>Reformatted Policy.</td>
</tr>
<tr>
<td>8/2022</td>
<td>Updated to add new gMG indication for Ultomiris ®.</td>
</tr>
<tr>
<td>4/2022</td>
<td>Updated to add Vyvgart ™ (efgartigimod alfa) to the policy.</td>
</tr>
<tr>
<td>11/2021</td>
<td>Updated to clarify Empaveli ™ criteria.</td>
</tr>
<tr>
<td>8/2021</td>
<td>Updated to include age update for Ultomiris ® and to add Empaveli ™ to the policy.</td>
</tr>
<tr>
<td>2/2021</td>
<td>Updated vaccination requirements for Soliris ®.</td>
</tr>
<tr>
<td>1/2021</td>
<td>Updated to include Enspryng™ and to change the policy name again.</td>
</tr>
<tr>
<td>9/2020</td>
<td>Updated to include Uplizna™ and to change the policy name.</td>
</tr>
<tr>
<td>7/2020</td>
<td>Implement a policy for both Ultomiris ® &amp; Soliris ®</td>
</tr>
</tbody>
</table>

References

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