MASSACHUSETTS

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Pharmacy Medical Policy

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

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Policy Number: 093
BCBSA Reference Number: None

Related Policies

1. 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 Operations:** Tel: 1-800-366-7778 Fax: 1-800-583-6289
		Policy last updated **5/2024**
Pharmacy (Rx) or Medical (MED) benefit coverage	☒ **Rx** ☒ **MED**	**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
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		

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
Fabhalta ® (iptacopan)	PA Required
Rystiggo ® (rozanolixizumab)	PA Required
Soliris ® (eculizumab)	PA Required
Ultomiris ® (ravulizumab)	PA Required
Uplizna ™ (Inebilizumab)	PA Required
Vyvgart ® **(**efgartigimod alfa**)**	PA Required
Vyvgart ® **Hytrulo** (efgartigimod alfa and hyaluronidase-qvfc)	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:

a. Confirmed diagnosis of Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
b. 18 years of age or older, **AND**
c. Documented baseline value for serum lactate dehydrogenase (LDH) is ≥1.5 times the upper limit of normal, **AND**

d. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), **Empaveli** ™ (pegcetacoplan), or with **Soliris** ® (eculizumab), **AND**
e. Vaccination against Neisseria meningitides prior to initiation [unless treatment cannot be delayed]) [1*]
f. 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. Confirmed diagnosis of Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
2. 18 years of age or older, **AND**
3. Decrease in serum LDH from pretreatment baseline, **AND**
4. **NO** dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), **Empaveli** ™ (pegcetacoplan), or with **Soliris** ® (eculizumab), **AND**
5. Prescribing physician is enrolled in Empaveli REMS program, **AND**
6. Absence of unacceptable toxicity from the drug

We may cover **Fabhalta** ® (iptacopan) for the treatment of Paroxysmal nocturnal hemoglobinuria (PNH) when all of the following criteria are met:

a. Documented diagnosis of Paroxysmal nocturnal hemoglobinuria (PNH), **AND**
b. 18 years of age or older, **AND**
c. Prescribing physician is enrolled in FABHALTA REMS program, **AND**
d. NO dual therapy with another terminal complement inhibitor such as Ultomiris® (ravulizumab), Empaveli ™ (pegcetacoplan), or with Soliris ® (eculizumab), **AND**
e. Vaccination against Streptococcus pneumoniae and Neisseria meningitidis prior to initiation [unless treatment cannot be delayed])

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.
   f. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

   *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.
f. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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 *(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 and or the Ultomiris REMS program
g. Previous treatment with or contraindication to **Vyvgart** ® (efgartigimod alfa) or **Vyvgart** ® **Hytrulo** (efgartigimod alfa and hyaluronidase-qvfc).
h. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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

4. 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.
e. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

*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
f. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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
f. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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
e. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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
e. Documented Dose and Frequency must be submitted and must be within the FDA approved_ Dosing and Frequency.

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

1. Myasthenia Gravis (gMG)
a. 18 years of age or older
b. Positive serologic test for anti-AChR antibodies **OR** Positive serologic test for MuSK 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. Previous treatment with or contraindication to **Vyvgart** ® (efgartigimod alfa) or **Vyvgart** ® For anti-AChR antibodies members whereas, MuSK positive members do not need to meet this requirement.

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

a. Confirmed diagnosis of Neuromyelitis optica spectrum disorder (NMOSD), **AND**
b. 18 years of age or older, **AND**
c. 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:

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)
J9333	Injection, rozanolixizumab-noli, 1 mg (Rystiggo)
J9334	Injection, efgartigimod alfa, 2 mg and hyaluronidase-qvfc (Vyvgart)

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

Date	Action
5/2024	Updated to add Fabhalta to the policy.
3/2024	Updated to require dose and frequency for Soliris ® and Ultomiris ® to coincide with the Medical claim edits and coding clarified.
9/2023	Updated to add Vyvgart ® Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) to the policy and updated IC to align with 118E MGL § 51A.
7/2023	Reformatted Policy.
8/2022	Updated to add new gMG indication for Ultomiris ®.
4/2022	Updated to add Vyvgart ™ (efgartigimod alfa) to the policy.
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.
6. Vyvgart ™ [package insert]. Boston, MA: argenx US, Inc.: 2/2022.
7. Fabhalta ® [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corporation: 3/2024.

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