MEDICAL MUTUAL®

Drug Policy

Policy:	**201421**	**Initial Effective Date: 10/30/2014**
Code(s):	**J1300, J3590**	**Annual Review Date: 06/18/2024**
SUBJECT:	**Soliris® (Eculizumab)** **Bkemv Eculizumab-aeeb)**	**Last Revised Date: 06/18/2024**

☒Subject to Site of Care

Prior approval is required for some or all procedure codes listed in this Corporate Drug Policy.

Initial and renewal requests for the medication(s) listed in this policy are subject to site of care management. When billed under the medical benefit, administration of the medication will be restricted to a non-hospital facility-based location (i.e., home infusion provider, provider's office, free-standing ambulatory infusion center) unless the member meets the site of care exception criteria. To view the exception criteria and a list of medications subject to site of care management please click here.

Policy Statement
This policy involves the use of Soliris. Prior authorization is recommended for medical benefit coverage of Soliris. Approval is recommended for those who meet the conditions of coverage in the **Initial Approval and Renewal Criteria, Preferred Drug (when applicable), Dosing/Administration, Length of Authorization, and Site of Care (when applicable)** for the diagnosis provided.

RECOMMENDED AUTHORIZATION CRITERIA
Coverage of Soliris is recommended in those who meet the following criteria:

I. Length of Authorization

- PNH and aHUS: Coverage will be provided for twelve (12) months and may be renewed.
- gMG and NMOSD: Initial coverage will be provided for six (6) months and may be renewed annually thereafter.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Soliris 300 mg/30 mL single-dose vial:
  - Loading Doses: 3 vials days 1, 8, 15, & 22; then 4 vials day 29
  - Maintenance Dose: 4 vials every 14 days

Bkemv 300 mg/30 mL single-dose vial:

- Loading Doses: 3 vials days 1, 8, 15, & 22; then 4 vials day 29
- Maintenance Dose: 4 vials every 14 days

B. Max Units (per dose and over time) [HCPCS Unit]:

Indication	Loading Doses	Maintenance Dose
PNH	60 billable units (600 mg) Days 1, 8, 15, & 22; then 90 billable units (900 mg) Day 29	90 billable units (900 mg) every 14 days
aHUS, gMG, NMOSD	90 billable units (900 mg) Days 1, 8, 15, & 22; then 120 billable units (1200 mg) Day 29	120 billable units (1200 mg) every 14 days

III. Initial Approval Criteria [1,2]

Coverage is provided in the following conditions:

- Patient is at least 18 years of age (unless otherwise specified); **AND**

Universal Criteria [1,2]

- Prescriber is enrolled in the applicable Ultomiris and Soliris OR Bkemv Risk Evaluation and Mitigation Strategy (REMS) program; **AND**
- Patient must be vaccinated against meningococcal infection (serogroups A,C,W,Y and B) according to current ACIP recommendations at least two weeks prior to initiation of therapy and will continue to be revaccinated in accordance with ACIP recommendations *(Note: if urgent eculizumab therapy is indicated in a patient who is not up to date with meningococcal vaccines according to ACIP recommendations, provide the patient with antibacterial drug prophylaxis and administer the vaccines as soon as possible)*; **AND**
- Patient does not have an unresolved, serious systemic infection (e.g., *Neisseria meningitidis*, etc.); **AND**
- Will not be used in combination with other immunomodulatory biologic therapies (e.g., efgartigimod, efgartigimod-hyaluronidase, ravulizumab, pegcetacoplan, satralizumab, tocilizumab, inebilizumab, rozanolixizumab, rituximab, zilucoplan, pozelimab, etc.) *[Note: a 4-week run-in period is allowed when transitioning from eculizumab to pegcetacoplan]*; **AND**

Paroxysmal Nocturnal Hemoglobinuria (PNH) † Φ [1-7,10,17,24]

- Diagnosis must be confirmed by detection of PNH clones of at least 10% by flow cytometry diagnostic testing; **AND**

  - Patient has at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes); **AND**

- Patient has laboratory evidence of significant intravascular hemolysis (i.e., LDH ≥1.5 x ULN) with symptomatic disease and at least one other indication for therapy from the following (regardless of transfusion dependence):
  - Patient has symptomatic anemia (i.e., hemoglobin < 7 g/dL or hemoglobin < 10 g/dL, in at least two independent measurements in a patient with cardiac symptoms)
  - Presence of a thrombotic event related to PNH
  - Presence of organ damage secondary to chronic hemolysis (i.e., renal insufficiency, pulmonary insufficiency/hypertension)
  - Patient is pregnant and potential benefit outweighs potential fetal risk
  - Patient has disabling fatigue
  - Patient has abdominal pain (requiring admission or opioid analgesia), dysphagia, or erectile dysfunction; **AND**
- Documented baseline values for one or more of the following (necessary for renewal): serum lactate dehydrogenase (LDH), hemoglobin level, packed RBC transfusion requirement, and history of thrombotic events; **AND**
- Patient had an inadequate response, contraindication, or intolerance to a trial of ravulizumab (Ultomiris®)

Atypical Hemolytic Uremic Syndrome (aHUS) † Φ [1,8,9,11,18,27]

- Patient is at least 2 months of age; **AND**
- Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.); **AND**
- Thrombotic Thrombocytopenic Purpura (TTP) has been ruled out by evaluating ADAMTS-13 level (i.e., ADAMTS-13 activity level ≥ 10%); **AND**
- Shiga toxin *E. coli* related hemolytic uremic syndrome (STEC-HUS) has been ruled out; **AND**
- Other causes have been ruled out such as coexisting diseases or conditions (e.g., bone marrow transplantation, solid organ transplantation, malignancy, autoimmune disorder, drug-induced, malignant hypertension, HIV infection, Streptococcus pneumoniae sepsis or known genetic defect in cobalamin C metabolism, etc); **AND**
- Documented baseline values for one or more of the following (necessary for renewal): serum lactate dehydrogenase (LDH), serum creatinine/eGFR, platelet count, and plasma exchange/infusion requirement; **AND**
- Patient had an inadequate response, contraindication, or intolerance to a trial of ravulizumab (Ultomiris®)

Generalized Myasthenia Gravis (gMG) † Φ [1,12,13,19-23]

- **Patient had an inadequate response, contraindication, or intolerance to a trial of efgartimod alfa-fcab (Vyvgart®), efgartimod alfa-fcab and hyaluronidase-qvfc (Vyvgart Hytrulo®), or rozanolixizumab-noli (Rystiggo®); AND**
- Patient has Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of Class II to IV disease **§**; **AND**
- Patient has a positive serologic test for anti-acetylcholine receptor (AChR) antibodies; **AND**
- Patient has had a thymectomy (*Note: Applicable only to patients with thymomas OR non-thymomatous patients who are 50 years of age or younger)*; **AND**
- Physician has assessed objective signs of neurological weakness and fatiguability on a baseline neurological examination [e.g., including, but not limited to, the Quantitative Myasthenia Gravis (QMG) score, etc.]; **AND**
- Patient has a baseline MG-Activities of Daily Living (MG-ADL) total score of ≥6; **AND**
    - Patient had an inadequate response after a minimum one-year trial of concurrent use with two (2) or more immunosuppressive therapies (e.g., corticosteroids plus an immunosuppressant such as azathioprine, cyclosporine, mycophenolate, etc.); **OR**
    - Patient required chronic treatment with plasmapheresis or plasma exchange (PE) or intravenous immunoglobulin (IVIG) in addition to immunosuppressant therapy; **AND**
- Patient will avoid or use with caution medications known to worsen or exacerbate symptoms of MG (e.g., certain antibiotics, beta-blockers, botulinum toxins, hydroxychloroquine, etc.); **AND**
- Patient had an inadequate response, contraindication, or intolerance to a trial of ravulizumab (Ultomiris®)

§ Myasthenia Gravis Foundation of America (MGFA) Disease Clinical Classification [20]:

– **Class I**: Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal.
– **Class II**: Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
- **IIa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.

- **IIb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

– **Class III**: Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IIIa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IIIb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

– **Class IV**: Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IVa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IVb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

– **Class V**: Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVb.

Neuromyelitis Optica Spectrum Disorder (NMOSD) † Φ [1,14-16,25,26]

- Patient has a confirmed diagnosis based on the following:
  - Patient was found to be seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**
  - Patient has at least one core clinical characteristic **§** *(*Note: some core clinical characteristics require both clinical and typical MRI findings)*; **AND**
  - Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.]; **AND**
- Patient has a history of at least 2 relapses in the last 12 months OR 3 relapses in the last 24 months, with at least 1 relapse in the last 12 months; **AND**
- Patient has an Expanded Disability Status Score (EDSS) of ≤ 7.0; **AND**
- Patients who are receiving concurrent corticosteroid therapy are on ≤20 mg per day and those receiving immunosuppressive therapy (e.g. azathioprine, glucocorticoids, mycophenolate, etc.) are on a stable dose regimen; **AND**
- Patient has not received therapy with rituximab or mitoxantrone in the last 3 months; **AND**
- Patient has not received intravenous immune globulin (IVIG) in the last 3 weeks; **AND**

- Patient had an inadequate response, or has a contraindication or intolerance, to rituximab OR inebilizumab (Uplizna); **AND**
- **Patient had an inadequate response, contraindication, or intolerance to a trial of ravulizumab (Ultomiris®)**

§ Core Clinical Characteristics of NMOSD [16,25]
▪ Acute optic neuritis ▪ Acute myelitis ▪ Acute area postrema syndrome (APS): episode of otherwise unexplained hiccups and/or nausea and vomiting (lasting for at least 48 hours or with MRI evidence of a dorsal brainstem lesion) ▪ Acute brainstem syndrome other than APS ▪ Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on MRI ¥ ▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI ψ
¥ Diencephalic syndrome: Periependymal lesion (3rd ventricle) OR hypothalamic/thalamic lesion ψ Cerebral syndrome: Extensive periependymal lesion (lateral ventricle; often with Gd) OR long (> 1⁄2 length), diffuse, heterogeneous or edematous corpus callosum lesion OR long corticospinal tract lesion (unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle) OR large, confluent (unilateral or bilateral) subcortical or deep white matter lesion

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); **Φ** Orphan Drug

IV. Renewal Criteria [1-7,24]

Coverage may be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: serious meningococcal infections (septicemia and/or meningitis), infusion reactions, serious infections, etc.; **AND**
- **Paroxysmal Nocturnal Hemoglobinuria (PNH)**
  - **Patient had an inadequate response, contraindication, or intolerance to ravulizumab (Ultomiris®) [Note: Exceptions to the trial of ravulizumab will be made for patients that have are attempting pregnancy or are currently pregnant or breastfeeding]**
  - Patient has not developed severe bone marrow failure syndrome (i.e., aplastic anemia or myelodysplastic syndrome) OR experienced a spontaneous disease remission OR received curative allogeneic stem cell transplant; **AND**

- Disease response compared to pre-treatment baseline as indicated by one or more of the following:
  - Decrease in serum LDH
  - Stabilization/improvement in hemoglobin level
  - Decrease in packed RBC transfusion requirement (i.e., reduction of at least 30%)
  - Reduction in thromboembolic events

- **Atypical Hemolytic Uremic Syndrome (aHUS)**
  - **Patient had an inadequate response, contraindication, or intolerance to ravulizumab (Ultomiris®) [Note: Exceptions to the trial of ravulizumab will be made for patients that have are attempting pregnancy, or are currently pregnant or breastfeeding]; AND**
  - Disease response compared to pre-treatment baseline indicated by one or more of the following**:**
  - Decrease in serum LDH
  - Stabilization/improvement in serum creatinine/eGFR from pretreatment baseline
  - Increase in platelet count
  - Decrease in plasma exchange/infusion requirement

- **Generalized Myasthenia Gravis (gMG)**
  - **Patient had an inadequate response, or has a contraindication or intolerance to ravulizumab (Ultomiris®); [Note: Exceptions to the trial of ravulizumab will be made for patients that have are attempting pregnancy, or are currently pregnant or breastfeeding; and for patients who have experienced severe events due to their gMG (i.e., hospitalization or respiratory support) and are currently well controlled with eculizumab]; AND**
    - Patient has had an improvement (i.e., reduction) of at least 1-point from baseline in the Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score **Δ**; **AND**
    - Improvement in muscle strength testing with fatigue maneuvers as evidenced on neurologic examination when compared to baseline
  - *[Δ May substitute an improvement of at least 1-point from baseline in the Quantitative Myasthenia Gravis (QMG) total score, if available]*

- **Neuromyelitis Optica Spectrum Disorder (NMOSD)**
  - **Patient had an inadequate response, or has a contraindication or intolerance to ravulizumab (Ultomiris®); [Note: Exceptions to the trial of ravulizumab will be made for patients that have are attempting pregnancy, or are currently pregnant or breastfeeding; and for patients who have**

experienced severe events due to their NMOSD and are currently well controlled with eculizumab]; AND

- Disease response as indicated by stabilization and/or improvement in one or more of the following:
  - Neurologic symptoms as evidenced by a decrease in acute relapses, improvement of stability, or improvement in EDSS
  - Reduced hospitalizations
  - Reduction/discontinuation in plasma exchange treatments

V. Dosage/Administration

Indication	Dose*
Paroxysmal nocturnal hemoglobinuria (PNH)	Loading dose: – 600 mg intravenously every 7 days for the first 4 weeks, followed by 900 mg intravenously for the fifth dose 7 days later Maintenance dose: – 900 mg intravenously every 14 days
Atypical hemolytic uremic syndrome (aHUS)	**Adults** Loading dose: – 900 mg intravenously every 7 days for the first 4 weeks, followed by 1,200 mg intravenously for the fifth dose 7 days later Maintenance dose: – 1200 mg intravenously every 14 days **Patients < 18 years** 5 kg - <10 kg: – 300 mg weekly x 1 dose, 300 mg at week 2, then 300 mg every 3 weeks 10 kg - <20 kg: – 600 mg weekly x 1 dose, 300 mg at week 2, then 300 mg every 2 weeks 20 kg - <30 kg: – 600 mg weekly x 2 doses, 600 mg at week 3, then 600 mg every 2 weeks 30 kg - <40 kg: – 600 mg weekly x 2 doses, 900 mg at week 3, then 900 mg every 2 weeks ≥ 40 kg: – 900 mg weekly x 4 doses, 1200 mg at week 5, then 1200 mg every 2 weeks

Generalized Myasthenia Gravis (gMG) and Neuromyelitis Optica Spectrum Disorder (NMOSD)	Loading dose: – 900 mg intravenously every 7 days for the first 4 weeks, followed by 1,200 mg intravenously for the fifth dose 7 days later Maintenance dose: – 1200 mg intravenously every 14 days

**Doses should be administered at the above intervals, or within two days of these time points.*

VI. Billing Code/Availability Information

HCPCS Code(s):

- J1300 – Injection, eculizumab, 10 mg; 1 billable unit = 10 mg *(Soliris ONLY)*
- J3590 – Unclassified biologics *(Bkemv ONLY)*

NDC(s):

- Soliris 300 mg/30 mL single-dose vial for injection: 25682-0001-xx
- Bkemv 300 mg/30 mL single dose vial for injection: 55513-0180-xx

VII. References

1. Soliris [package insert]. Boston, MA; Alexion Pharmaceuticals, Inc.; March 2024. Accessed April 2024.
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Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D59.32	Hereditary hemolytic-uremic syndrome
D59.39	Other hemolytic-uremic syndrome
D59.5	Paroxysmal nocturnal hemoglobinuria [Marchiafava-Micheli]
G36.0	Neuromyelitis optica [Devic]
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes		
Jurisdiction	**NCD/LCA/LCD Document (s)**	**Contractor**
6, K	A54548	National Government Services, Inc

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdictio	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT,	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in	Novitas Solutions, Inc.
K (13 &	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC

Prior approval is required for HCPCS Code J1300.

Edits and Denials:

Prior Approval: Prior approval is required for Soliris **(HCPCS Code J1300)**. Requests for prior approval will be authorized by a nurse reviewer if submitted documentation meets criteria outlined within the Corporate Medical Policy.

Requests for prior approval will be forwarded to a qualified physician reviewer if submitted documentation does not meet criteria outlined within the Corporate Medical Policy.

TOPPS: Claims received with **HCPCS Code J1300** will edit with **Remark Code M3M or M4M** and will be adjudicated in accordance with the Corporate Medical Policy.

Liability: A participating provider will be required to write off charges denied as not medically necessary. A provider may bill a member for charges denied as investigational.