CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Indiana Medicaid

DRUG NAME	Soliris (eculizumab)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Soliris is a C5 Complement inhibitor initially approved by the FDA in 2007. It is approved for the treatment of paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis, atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy, generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AchR) antibody positive, and neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive.
PNH is a rare hematopoietic stem cell disorder in which activation of the complement system destroys red blood cells.
aHUS is a type of thrombotic microangiopathy (TMA), a group of syndromes defined by the presence of hemolytic anemia, low platelets and organ damage due to microscopic blood clots in the capillaries. Unlike typical HUS, aHUS is usually genetic. The three main signs of aHUS are hemolytic anemia, thrombocytopenia, and acute kidney failure. Of note, the other type of TMA is called thrombotic thrombocytopenic purpura (TTP); Soliris is not used to treat TTP.

Soliris (eculizumab) will be considered for coverage when the following criteria are met:

Paroxysmal Nocturnal Hemoglobinuria (PNH)

For **initial** authorization:
1. Member is at least 18 years of age; AND
2. Medication is prescribed by or in consultation with a hematologist; AND
3. Member has a diagnosis of PNH as confirmed by flow cytometry; AND
4. Member has a lactate dehydrogenase (LDH) level >1.5x upper limit of normal (ULN); AND
5. Member has at least one PNH-related sign/symptom e.g., fatigue, hemoglobin <10 g/dL, thrombosis, pRBC transfusion, shortness of breath; AND
6. Member has tried and failed or is unable to try Ultomiris; AND
7. Member has received meningococcal vaccine.
8. **Dosage allowed/Quantity limit:** 600mg IV weekly x 4 weeks, then 900mg 1 week later, then 900mg every 2 weeks thereafter.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Clinical evidence of positive response to therapy such as increased hemoglobin level, decreased need for transfusions, normalized LDH levels, improved fatigue.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Atypical Hemolytic Uremic Syndrome (aHUS)

For **initial** authorization:

1. Medication is prescribed by or in consultation with a hematologist or nephrologist; AND
2. Member has a diagnosis of aHUS supported by ALL of the following:
   a) Thrombocytopenia (platelet count < 150 x 10^9/L),
   b) Evidence of microangiopathic hemolytic anemia (MAHA) e.g., hemoglobin < 10 g/dL, elevated lactate dehydrogenase (LDH), low haptoglobin, presence of fragmented red blood cells or schistocytes on blood smear
   c) Evidence of renal impairment (e.g., raised SCr or low eGFR); AND
3. Shiga toxin-producing E. coli related HUS (STEC-HUS) has been ruled out; AND
4. ADAMTS13 activity level is > 10% (to rule out TTP); AND
5. Member has tried and failed or is unable to try Ultomiris; AND
6. Member has received meningococcal vaccine.
7. **Dosage allowed/Quantity limit:**
   Pediatrics: See weight-based dosing in package insert.
   Adults: 900mg IV weekly x 4 weeks, then 1200mg 1 week later, then 1200mg every 2 weeks thereafter.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must demonstrate hematologic normalization as evidenced by increased platelet count or LDH maintained below upper limit of normal; AND
2. Improved or preserved kidney function.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Generalized Myasthenia Gravis (gMG)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication is prescribed by or in consultation with a neurologist; AND
3. Member has a documented diagnosis of MGFA class II-IV myasthenia gravis (see Appendix); AND
4. Lab result in chart notes shows the member is seropositive for AChR antibodies; AND
5. Member has tried and failed at least 1 conventional therapy:
   a) Pyridostigmine
   b) Corticosteroid for at least 3 months
   c) Non-steroid immunosuppressant (e.g., azathioprine) for at least 6 months; AND
6. Member has tried and failed or is unable to try Ultomiris (requires trial of IV Vyvgart); AND
7. Member has received meningococcal vaccine.
8. **Dosage allowed/Quantity limit:** 900 mg IV weekly for the first 4 weeks, followed by 1200 mg for the fifth dose 1 week later, then 1200 mg every 2 weeks thereafter.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must demonstrate improvement in activities of daily living, muscle strength, and/or health related quality of life; fewer exacerbations or hospitalizations, or reduced use of rescue medication.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Neuromyelitis Optica Spectrum Disorder (NMOSD)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a neurologist; AND
3. Member has a documented diagnosis of NMOSD and is seropositive for aquaporin-4 (AQP4) IgG antibodies; AND
4. Member had had 1 or more relapses within the past year; AND
5. Member has tried and failed at least one of the following for 6 months or longer: azathioprine, mycophenolate, rituximab (requires prior auth); AND
6. Member has received meningococcal vaccine.
7. **Dosage allowed/Quantity limit:** 900 mg IV weekly for the first 4 weeks, followed by 1200 mg for the fifth dose 1 week later, then 1200 mg every 2 weeks thereafter.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must document disease stabilization, symptom improvement, and/or reduced frequency of relapses.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Soliris (eculizumab) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
11/14/2017	New policy for Soliris created.
10/26/2019	New diagnosis of Neuromyelitis optica spectrum disorder (NMOSD) added.
10/15/2020	Revised criteria for NMOSD to align with other products. Only require at least 1 relapse in past year. Added trial of a standard therapy. Added trial of Enspryng. Reworded the criteria for meningitis vaccine. Removed the part about stable immunosuppressive therapy (just assessed for study purpose). Removed restrictions on prior Rituxan, mitoxantrone, IVIG (only applicable to the study design). Changed initial auth duration to 6 months. Edited the renewal criteria to be more appropriate. Also corrected the dose information error. Changed to nonpreferred drug status.
02/08/2021	gMG: Updated references. Added specialist requirement. Removed MG-ADL score. Amended prerequisite drugs to more closely match guidelines and literature. Removed clinical trial exclusion criteria. Reduced initial auth duration to 6 months. Revised renewal criteria
06/02/2021	aHUS: Updated references. Added specialist requirement. Revised diagnostic parameters. Removed list of restrictions from clinical trials. Stated Ultomiris as preferred. Amended dosing information. Revised renewal criteria. PNH: Updated references. Added age limit. Removed nephrology as specialist. Removed transfusion and organ damage requirements. Preference for Ultomiris. Amended dosing information. Reduced initial auth duration from 12 months to 6 months. Revised renewal criteria.
07/25/2023	Transferred to new template. PNH: Updated references. Added that they must be symptomatic.

	aHUS: Updated and added references. Corrected ADAMTS13 level cutoff. Changed "evidence of hemolysis" to evidence of MAHA. NMOSD: Added references. Removed requirement for trial of Enspryng. MG: Added reference. Removed "severe, refractory" and added "MGFA class II-IV." Added MGFA appendix. Added trial of Ultomiris. Shortened and simplified list of conventional therapy trials.

APPENDIX:

MG Foundation of America (MGFA) Clinical Classification	
Class I	any ocular weakness; all other muscle strength is normal
Class II	mild weakness affecting other than ocular muscles; may also have ocular weakness at any level
Class III	moderate weakness affecting other than ocular muscles; may also have ocular weakness at any level
Class IV	severe weakness affecting other than ocular muscles; may also have ocular weakness at any level
Class V	defined by intubation, with or without mechanical ventilation

References:

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