ALASKA MEDICAID
Prior Authorization Criteria

Soliris®, Ultomiris® (eculizumab,ravulizumab-cwvz)

FDA INDICATIONS AND USAGE[1,2]

Soliris and Ultomiris are complement inhibitors indicated for the treatment of adult and pediatric patients one month of age and older with paroxysmal nocturnal hemoglobinuria (PNH) and the treatment of adult and pediatric patients one month of age and older with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy (TMA). Soliris is also indicated for the treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AchR) antibody positive and the treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive.

APPROVAL CRITERIA[1,2,3,4,5,6,7,8,9,10]

Atypical Hemolytic Uremic Syndrome (aHUS) (**Soliris or Ultomiris**)

1. Patient is 1 month of age or older **AND**;
2. Patient has a documented diagnosis of aHUS **AND**;
3. Diagnosis of thrombocytopenic purpura (TTP) has been ruled out (I.E., normal ADAMTS 13 activity) or a trial of plasma exchange did not result in clinical improvement **AND**;
4. Patient does not have a Shiga toxin-producing escherichia coli (E. coli) infection **AND**;
5. Documented baseline values for one or more of the following:
    a. Serum lactate dehydrogenase (LDH)
    b. Serum creatinine/eGFR
    c. Platelet count
    d. Plasma exchange/infusion requirement

Paroxysmal Nocturnal Hemoglobinuria (PNH) (**Soliris or Ultomiris**)

1. Patient is 1 month of age or older **AND**;
2. Patient has a documented diagnosis of PNH confirmed by flow cytometry diagnostic testing **AND**;
3. Patient has one of the following indications for therapy:
    a. Presence of a thrombotic event
    b. Presence of organ damage secondary to chronic hemolysis
    c. Patient is pregnant and potential benefit outweighs potential fetal risk
    d. Patient is transfusion dependent
    e. Patient has high LDH activity (defined as ≥1.5 x ULN) with clinical symptoms **AND**;
4. Patient has documented baseline values for all the following:
    a. Serum lactate dehydrogenase (LDH)
    b. Hemoglobin level
    c. Packed RBC transfusion requirement

Generalized Myasthenia Gravis (gMG) (**Soliris only**)

1. Patient is 18 years of age or older **AND**;
2. Patient has a documented diagnosis of gMG with a Myasthenia Gravis Foundation of America (MGFA)clinical classification of II,III, or IV **AND;**
3. Patient has Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) total score of ≥ 6 **AND**;
4. Patient has a positive serologic test for anti-acetylcholine receptor (AchR) antibodies **AND**;
5. Prescriber has assessed the baseline Quantitative Myasthenia Gravis (QMG) score **AND**;
6. Documentation of inadequate response to or has a labeled contraindication to TWO or more immunosuppressive drug agents used alone or in combination for at least 12 months (I.E. azathioprine, mycophenolate mofetil, cyclosporine, cyclophosphamide, methotrexate, tacrolimus, rituximab) **OR** documentation of inadequate response to or has a labeled contraindication to ONE or more immunosuppressive drug agents as monotherapy or in combination therapy and requires chronic plasma exchange, plasmapheresis or intravenous immunoglobulin therapy.

Neuromyelitis Optica Spectrum Disorder (NMOSD) (**Soliris only**)

1. Patient is 18 years of age or older **AND**;
2. Patient has a documented diagnosis of NMOSD confirmed by a positive serologic test for anti-aquaporin-4 immunoglobulin G (AQP4-IgG)/NMP-IgG antibodies **AND**;
3. Documentation of failure or inadequate response [history of at least one relapse (acute attack from neuromyelitis spectrum disorder) in the last 12 months, or two relapses in the last 2 years] to ONE of the following systemic therapies; OR a Contraindication per FDA label or significant intolerance to ALL the following systemic therapies: .
    a. Azathioprine
    b. Mycophenolate
    c. Rituximab **AND**;
4. The diagnosis of multiple sclerosis has been ruled out.

DENIAL CRITERIA [1,2]

1. Failure to meet approval criteria **OR**;
2. Patient has not had a vaccination against Neisseria meningitides at least 2 weeks prior to initiation **OR**;
3. Not being prescribed by or in consultation with a hematologist, nephrologist, or neurologist **OR**;
4. Prescriber is not enrolled in the REMS program.

CAUTIONS [1,2]

- See package insert for cautions:
    - Soliris: https://solirispro.com/pdf/Soliris_USPI.pdf
    - Ultomiris: https://alexion.com/Documents/Ultomiris_USPI.pdf

DURATION OF APPROVAL

- Initial Approval: up to 4 months
- Reauthorization Approval: up to 12 months if the prescriber documents the patient has disease improvement

QUANTITY LIMIT[1,2]

- Soliris max dose = 1200mg
- Soliris J1300 = max HCPCS 120
- Ultomiris max dose = 3600mg
- Ultomiris J1303 = max HCPCS 360

REFERENCES / FOOTNOTES:

1. Soliris ® [package insert]. Boston, MA: Alexion Pharmaceuticals, Inc. November 2020.
2. Ultomiris ® [package insert]. Boston, MA: Alexion Pharmaceuticals, Inc. January 2022.
3. Kielstein JT, Beutel G, Fleig S, et al. Best supportive care and therapeutic plasma exchange with or without eculizumab in Shiga-toxin-producing E. coli O104:H4 induced haemolytic-uraemic syndrome: an analysis of the German STEC-HUS registry. Nephrol Dial Transplant. 2012 Oct;27(10):3807-15.
4. Campistol JM, Arias M, Ariceta G, et al. An update for atypical haemolytic uraemic syndrome: diagnosis and treatment. A consensus document. Nefrologia 2013;33:27–45.
5. Franchini M. Atypical hemolytic uremic syndrome: from diagnosis to treatment. Clin Chem Lab Med. 2015 Oct;53(11):1679- 88.
6. Sutherland DR, Keeney M, Illingworth A. Practical guidelines for the high-sensitivity detection and monitoring of paroxysmal nocturnal hemoglobinuria clones by flow cytometry. Cytometry B Clin Cytom. 2012 Jul;82(4):195-208.
7. Howard JF Jr, Utsugisawa K, Benatar M, et al. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody positive refractory generalised myasthenia gravis (REGAIN): a phase 3, randomised, double-blind, placebo-controlled, multicentre study. Lancet Neurol. 2017 Dec;16(12):976-986.
8. Howard JF Jr, Barohn RJ, Cutter GR, et al. A randomized, double-blind, placebo-controlled phase II study of eculizumab in patients with refractory generalized myasthenia gravis. Muscle Nerve. 2013 Jul;48(1):76-84.
9. Pittock SJ, Berthele A, Fujihara K, et al. Eculizumab in Aquaporin-4-Positive Neuromyelitis Optica Spectrum Disorder. N Engl J Med. 2019 May 3.
10. Nikoo Z, Badihian S, Shaygannejad V, et al. Comparison of the efficacy of azathioprine and rituximab in neuromyelitis optica spectrum disorder: a randomized clinical trial. J Neurol. 2017 Sep;264(9):2003-2009.