Soliris (eculizumab)

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year except as noted within the criteria below

Medications	Quantity Limit
Soliris (eculizumab) 300 mg/30 mL vial*	8 vials per 28 days

*Initiation of therapy for Atypical Hemolytic Uremic Syndrome (aHUS), generalized Myasthenia Gravis (MG), or neuromyelitis optica spectrum disorder (NMOSD): May approve 4 (four) additional vials (300 mg/mL) in the first 28 days (4 weeks) of treatment.

If individual receives plasma exchange [PE], plasmapheresis [PP], or fresh frozen plasma infusion during therapy, supplemental doses of Soliris (up to 600 mg following each PE or PP intervention or up to 300 mg following fresh frozen plasma) may be approved.

APPROVAL CRITERIA

Requests for initiation of therapy with Soliris (eculizumab) in **paroxysmal nocturnal hemoglobinuria** (PNH) may be approved if the following criteria are met:

I. Individual is 18 years of age or older; **AND**
II. Individual has PNH as verified by flow cytometry, including the presence of (Parker 2005):
    A. PNH type III red cell clone or measurable granulocyte or monocyte clone; **OR**
    B. Glycosylphosphatidylinositol-anchored proteins (GPI-AP)-deficient polymorphonuclear cells (PMNs);

AND

III. Individual has completed or updated meningococcal vaccination at least 2 weeks prior to administration of the first dose of Soliris (eculizumab), unless that the risks of delaying Soliris (eculizumab) outweigh the risk of meningococcal infection;

AND

IV. Individual has (Hillmen 2006):
    A. Lactate dehydrogenase is greater than 1.5 times the upper limit of normal, and documentation is provided; **AND**
    B. One or more PNH-related sign or symptom (such as but not limited to anemia or history of a major adverse vascular event from thromboembolism), or history of transfusion due to PNH).

Initial Approval Duration: 6 months

Requests for continued use of Soliris (eculizumab) in PNH may be approved if the following criteria are met:

I. Documentation is provided that individual has experienced a clinical response as shown by one of the following:
   A. Stabilization of hemoglobin levels; **OR**
   B. Reduction in number of transfusions required; **OR**
   C. Improvement in hemolysis (for example, normalization or decrease of LDH levels).

Note: If Soliris therapy is discontinued, individuals should be closely monitored for at least 8 weeks after cessation to detect hemolysis.

Requests for initiation of therapy with Soliris (eculizumab) in **neuromyelitis optica spectrum disorder** (NMOSD) may be approved if the following criteria are met:

I. Individual is 18 years of age or older with NMOSD; **AND**
II. Documentation is provided that NMOSD is seropositive as verified by the presence of anti-aquaporin-4 (AQP4) antibodies;
AND
III. Documentation is provided that individual has a history of at least 2 acute attacks or relapses in the last 12 months prior to initiation of therapy;
OR
IV. Documentation is provided that individual has a history of at least 3 acute attacks or relapses in the last 24 months AND at least 1 relapse in the 12 months prior to initiation of therapy;
AND
V. Individual has completed or updated meningococcal vaccination at least 2 weeks prior to
administration of the first dose of Soliris (eculizumab), unless the risks of delaying Soliris (eculizumab) outweigh the risk of meningococcal infection.

Requests for continued use of Soliris (eculizumab) in NMOSD may be approved if the following criteria are met:

I. Documentation is provided that individual has experienced a clinical response (for example, a reduction in the frequency of relapse).

Requests for initiation of therapy with Soliris (eculizumab) in **atypical hemolytic uremic syndrome** (aHUS) may be approved if the following criteria are met:

I. Individual is 2 months of age or older with a diagnosis of aHUS;
AND
II. The diagnosis of aHUS is supported by the absence of Shiga toxin-producing E. coli infection;
AND
III. Thrombotic thrombocytopenic purpura has been ruled out [for example, normal ADAMTS 13 activity and no evidence of an ADAMTS 13 inhibitor (Loirat 2011, 2016)],

or if thrombotic thrombocytopenic purpura cannot be ruled out by laboratory and clinical evaluation, a trial of plasma exchange did not result in clinical improvement;

AND

IV. Individual has completed or updated meningococcal vaccination at least 2 weeks prior to administration of the first dose of Soliris (eculizumab) unless the risks of delaying Soliris (eculizumab) outweigh the risk of meningococcal infection.

Initial Approval Duration: 12 weeks.

Requests for continued use of Soliris (eculizumab) in aHUS may be approved if the following criteria are met:

I. There is clinical improvement after the initial trial (for example, increased platelet count or laboratory evidence of reduced hemolysis) until an individual becomes a candidate for physician-directed cessation as evidenced by the following (Merrill 2017):
   A. Complete clinical remission has been achieved (that is, resolution of thrombocytopenia and mechanical hemolysis, and normalization or new baseline plateau of renal function) *and* improvement of precipitating illness is clinically apparent; **AND**
   B. Duration of clinical remission has been stable for 2 months.

Note: Close monitoring after cessation is essential (for example: regular laboratory monitoring including complete blood count, peripheral smear, lactate dehydrogenase, renal function, and urine protein beginning the week of the held dose and weekly for 4 weeks, every 2 weeks for 1 month, and then monthly for 3 months at the discretion of the treating clinician).

Requests for resumption of Soliris (eculizumab) in aHUS may be approved if the following criteria are met (Fakhouri 2017):

I. Documentation is provided that individual experienced a relapse after discontinuation of therapy as defined by:
   A. Reduction in platelet count to less than 150,000/mm^3 or greater than 25% from baseline; **OR**
   B. Mechanical hemolysis (having 2 or more features of hemoglobin less than 10 g/dL, lactate dehydrogenase greater than 2 times upper limit of normal, undetectable haptoglobin, or presence of schistocytes on smear); **OR**
   C. Acute kidney injury with serum creatinine increase greater than 15% from baseline levels.

Requests for initiation of therapy with Soliris (eculizumab) in **generalized myasthenia gravis** (gMG) may be approved if the following criteria are met:

I. Individual is 18 years of age or older with the gMG;

AND

II. Documentation is provided that individual has a positive serologic test for binding anti-acetylcholine receptor antibodies (AChR-ab);

AND

III. Individual has Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV disease;

AND

IV. Documentation is provided that individual has a Myasthenia Gravis Activities of Daily Living (MG-ADL) score of at least 6 or higher;

AND

V. Documentation is provided that individual meets both of the following (A and B):
   A. Individual has had a trial and inadequate response or intolerance to an acetylcholinesterase inhibitor; **OR**
      1. Individual is on a stable dose of an acetylcholinesterase inhibitor; **OR**
      2. Individual has a contraindication to acetylcholinesterase inhibitors;

   AND

   B. Individual has had a trial and inadequate response or intolerance to one or more immunosuppressive agents (including but not limited to systemic corticosteroids or non-steroidal immunosuppressants); **OR**
      1. Individual is on a stable dose of one or more immunosuppressive agents (including but not limited to systemic corticosteroids or non-steroidal immunosuppressants); **OR**
      2. Individual has a contraindication to systemic corticosteroids and non-steroidal immunosuppressants;

AND

VI. Individual completed or updated meningococcal vaccination at least 2 weeks prior to administration of the first dose of Soliris (eculizumab), unless the risks of delaying Soliris (eculizumab) outweigh the risk of meningococcal infection.

Initial Approval Duration: 26 weeks

Requests for continued use of Soliris (eculizumab) in gMG may be approved if the following criteria are met:

I. Individual has experienced a clinical response as evidenced by both of the following:
   A. Reduction in signs or symptoms that impact daily function; **AND**
   B. Documentation is provided to show at least a 2-point reduction in MG-ADL total score from baseline.

Requests for Soliris (eculizumab) may not be approved for the following:

I. Individual is using in combination with efgartigimod alfa, inebilizumab, ravulizumab, rozanolixizumab-noli, rituximab, or satralizumab; **OR**
II. Individual is using in combination with pegcetacoplan for more than 4 weeks for PNH; **OR**
III. Individual has evidence of an active meningococcal infection; **OR**
IV. When the above criteria are not met and for all other indications.

Note:
Soliris (eculizumab) has a black box warning for serious meningococcal infections. Life-threatening and fatal meningococcal infections have occurred in patients treated with Soliris (eculizumab) and meningococcal infection may become rapidly life-threating or fatal if not recognized and treated early. Individuals should be immunized with meningococcal vaccines at least 2 weeks prior to initiating therapy unless the risks of delaying therapy outweigh the risk of developing a meningococcal infection. The FDA has required the manufacturer to develop a comprehensive risk management program that includes the enrollment of prescribers in the Soliris REMS Program. Additional information and forms for individuals, prescribers, and pharmacists may be found on the manufacturer's website: http://www.solirisrems.com.

Key References:

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