Policy Title: Soliris (eculizumab) NON HEMATOLOGY POLICY Intravenous

<table>
<thead>
<tr>
<th>Department:</th>
<th>PHA</th>
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<tr>
<td>Effective Date:</td>
<td>01/01/2020</td>
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<tr>
<td>Review Date:</td>
<td>09/18/2019, 12/20/2019, 1/22/2020, 12/2020, 5/27/2021</td>
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<tr>
<td>Revision Date:</td>
<td>09/18/2019, 1/22/2020, 12/2020</td>
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**Purpose:** To support safe, effective and appropriate use of Soliris (eculizumab).

**Scope:** Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

**Policy Statement:**
Soliris (eculizumab) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process. **For Hematology indications, please refer to the NHPRI Soliris Hematology Policy**

**Procedure:**
Coverage of Soliris (eculizumab) will be reviewed prospectively via the prior authorization process based on criteria below.

**Initial Criteria:**
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

**Neuromyelitis optica spectrum disorder (NMOSD)**
- Submission of medical records (e.g., chart notes, laboratory values, etc.) to support the diagnosis of neuromyelitis optica spectrum disorder (NMOSD) by a neurologist confirming all of the following:
  - Past medical history of one of the following:
    - Optic neuritis
    - Acute myelitis
    - Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
    - Acute brainstem syndrome
    - Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
    - Symptomatic cerebral syndrome with NMOSD-typical brain lesions; AND
  - Positive serologic test for anti-aquaporin-4 immunoglobulin G (AQP4-IgG)/NMO-IgG antibodies; AND
  - Diagnosis of multiple sclerosis or other diagnoses have been ruled out; AND
- Patient has not failed a previous course of Soliris therapy; AND
- One of the following:
  - History of at least two relapses during the previous 12 months prior to initiating Soliris; OR
  - History of at least three relapses during the previous 24 months, at least one relapse occurring within the past 12 months prior to initiating Soliris; AND
- Soliris is initiated and titrated according to the US FDA labeled dosing for NMOSD, up to a maximum of 1200 mg every 2 weeks; AND
- Prescribed by, or in consultation with, a neurologist; AND
- Patient is not receiving Soliris in combination with any of the following:
  - Disease modifying therapies for the treatment of multiple sclerosis [e.g., Gilenya (fingolimod), Tecfidera (dimethyl fumarate), Ocrevus (ocrelizumab), etc.]
  - Anti-IL6 therapy [e.g., Actemra (tocilizumab), Enspryng (satralizumab)]
  - Uplizna (inebilizumab)
  - Rituximab; AND
- Patient has experienced a failure, contraindication or intolerance to Enspryng (satralizumab)* AND Uplizna (inebilizumab)

* This requirement ONLY applies to Medicaid Members

Generalized myasthenia gravis (gMG)
- Submission of medical records (e.g., chart notes, laboratory values, etc.) to support the diagnosis of generalized myasthenia gravis (gMG) by a neurologist or in consultation with a neurologist confirming all of the following:
  - Patient has not failed a previous course of Soliris therapy; AND
  - Positive serologic test for anti-AChR antibodies; AND
  - One of the following:
    - History of abnormal neuromuscular transmission test demonstrated by single-fiber electromyography (SFEMG) or repetitive nerve stimulation
    - History of positive anticholinesterase test, e.g., edrophonium chloride test
    - Patient has demonstrated improvement in MG signs on oral cholinesterase inhibitors, as assessed by the treating neurologist; AND
  - Patient has a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of class II, III, or IV at initiation of therapy; AND
  - Patient has a Myasthenia Gravis-specific Activities of Daily Living scale (MG-ADL) total score ≥ 6 at initiation of therapy; AND
  - Both of the following:
    - History of failure of at least two immunosuppressive agents over the course of at least 12 months [e.g., azathioprine, methotrexate, cyclosporine, mycophenolate, etc.]; AND
    - Patient has received 2 or more courses of plasmapheresis/plasma exchanges and/or intravenous immune globulin for at least 12 months without symptom control; AND
  - Soliris is initiated and titrated according to the US FDA labeled dosing for gMG, up to a maximum of 1200 mg every 2 weeks; AND
• Prescribed by, or in consultation with, a neurologist

Continuation of Therapy Criteria:

• Neuromyelitis optica spectrum disorder (NMOSD)
  • Patient has previously been treated with Soliris; AND
  • Submission of medical records (e.g., chart notes, laboratory tests) to demonstrate a positive clinical response from baseline as demonstrated by at least both of the following:
    ▪ Reduction in the number and/or severity of relapses or signs and symptoms of NMOSD
    ▪ Maintenance, reduction, or discontinuation of dose(s) of any baseline immunosuppressive therapy (IST) prior to starting Soliris. Note: Add on, dose escalation of IST, or additional rescue therapy from baseline to treat NMOSD or exacerbation of symptoms while on Soliris therapy will be considered as treatment failure.
  • Soliris is dosed according to the US FDA labeled dosing for NMOSD: up to a maximum of 1200 mg every 2 weeks; AND
  • Prescribed by, or in consultation with, a neurologist; AND
  • Patient is not receiving Soliris in combination with any of the following:
    ▪ Disease modifying therapies for the treatment of multiple sclerosis [e.g., Gilenya (fingolimod), Tecfidera (dimethyl fumarate), Ocrevus (ocrelizumab), etc.]
    ▪ Anti-IL6 therapy [e.g., Actemra (tocilizumab), Enspryng (satralizumab)]
    ▪ Uplizna (inebilizumab)
    ▪ Rituximab

• Generalized myasthenia gravis (gMG)
  • Patient has previously been treated with Soliris; and
  • Submission of medical records (e.g., chart notes, laboratory tests) to demonstrate a positive clinical response from baseline as demonstrated by all of the following:
    ▪ Improvement and/or maintenance of at least a 3-point improvement (reduction in score) in the MG-ADL score from pre-treatment baseline.
    ▪ Reduction in signs and symptoms of myasthenia gravis
    ▪ Maintenance, reduction, or discontinuation of dose(s) of baseline immunosuppressive therapy (IST) prior to starting Soliris. Note: Add on, dose escalation of IST, or additional rescue therapy from baseline to treat myasthenia gravis or exacerbation of symptoms while on Soliris therapy will be considered as treatment failure; AND
  • Soliris is dosed according to the US FDA labeled dosing for gMG: up to a maximum of 1200 mg every 2 weeks; and
  • Prescribed by, or in consultation with, a neurologist.
Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

Dosage/Administration:

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<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 mg)</th>
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<tbody>
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<td>Generalized Myasthenia Gravis (gMG) or Neuromyelitis optica spectrum disorder (NMOSD)</td>
<td>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</td>
<td>Loading dose: 90 billable units Days 1, 8, 15, &amp; 22; then 120 billable units Day 29 Maintenance dose: 120 billable units every 14 days</td>
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Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT code is:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J1300</td>
<td>Injection, eculizumab, 10 mg</td>
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References: