Policy Title:	Soliris (eculizumab) **NON HEMATOLOGY POLICY** Intravenous		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	09/18/2019, 12/20/2019, 1/22/20, 12/2020		
Revision Date:	09/18/2019, 1/22/20, 12/2020		

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 agent over the course of at least 12 months [e.g., azathioprine, methotrexate, cyclosporine, mycophenylate, 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 at least 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:

Indication	Dose	Maximum dose (1 billable unit = 10 mg)
Generalized Myasthenia Gravis (gMG) or 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	Loading dose: 90 billable units Days 1, 8, 15, & 22; then 120 billable units Day 29 Maintenance dose: 120 billable units every 14 days

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:

HCPCS/CPT Code	Description
J1300	Injection, eculizumab, 10 mg

References:

1. Soliris [package insert]. New Haven, CT: Alexion Pharmaceuticals, Inc.; October 2017.
2. Sanders D, Wolfe G, Benatar M et al. International consensus guidance for management of myasthenia gravis. *Neurology*. 2016; 87 (4):419-425.
3. Jaretzki A, Barohn RJ, Ernstoff RM et al. Myasthenia Gravis: Recommendations for Clinical Research Standards. *Ann Thorac Surg*. 2000;70: 327-34.
4. Howard JF, Utsugisawa K, Benatar M. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalized myasthenia gravis (REGAIN); a phase 3, randomized, double-blind, placebo-controlled, multicenter study. *Lancet Neurol*. 2017 Oct 20. http://dx.doi.org/10.1016/S1474-4422(17)30369-1Ingenix HCPCS Level II, Expert 2011