Updated: 06/2021
PARP Approved: 06/2021

Gateway Health
Prior Authorization Criteria

C5b Complement inhibitors

All requests for C5b Complement inhibitors require a prior authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

C5b Complement inhibitors Prior Authorization Criteria:

C5b Complement inhibitors include Soliris (eculizumab), and Ultomiris (ravulizumab-cwvz). New products with this classification will require the same documentation.

For all requests for C5b Complement inhibitors all of the following criteria must be met:

- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines
- Is age-appropriate according to FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature

Coverage may be provided with a diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) and the following criteria are met:

- Medication is prescribed by, or in consultation with, a hematologist, oncologist, immunologist, or genetic specialist
- Member has a diagnosis of PNH confirmed by flow cytometry testing. Flow Cytometry pathology report must be supplied and demonstrate at least 2 different GPI protein deficiencies within 2 different cell lines from granulocytes, monocytes, or erythrocytes
- Member has one of the following:
    - Member's hemoglobin is less than or equal to 7 g/dL
    - Member has symptoms of anemia and the hemoglobin is less than or equal to 9 g/dL **(Soliris only)**
    - Member has symptoms of anemia and the hemoglobin is less than or equal to 10 g/dL **(Ultomiris only)**
- Must have a Lactate dehydrogenase (LDH) level at least 1.5 times the upper limit of the normal range (laboratory results with reference range must be submitted)
- If requesting Soliris, must have documentation of inadequate response, contraindication or intolerance to Ultomiris.
- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
    - Documentation of each of the following:
        - Documentation of a recent (within 3 months) LDH level that shows a reduction from baseline
        - Documentation that hemoglobin has not dropped by more than 2 g/dL from baseline.
            - If baseline hemoglobin was less than 9g/dL, then the most recent hemoglobin must remain above 7g/dL
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided with a diagnosis of atypical hemolytic uremic syndrome (aHUS) and the following criteria are met:

- Member weights at least 5 kilograms
- Medication is prescribed by, or in consultation with, a hematologist, oncologist, immunologist, genetic specialist, or nephrologist
- Must provide documentation of hemolysis such as an elevation in serum LDH and serum creatinine above the upper limits of normal or required dialysis.
- The diagnosis of aHUS is supported by the absence of Shiga toxin-producing *E.coli* infection
- Must provide documentation member does not have a disintegrin and metalloproteinase with thrombospondin type 1 motif member 13 (ADAMTS13) deficiency
- If requesting Soliris, must have documentation of inadequate response, contraindication or intolerance to Ultomiris.
- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
    - Documentation from the provider that the member had a positive clinical response as evidenced by any of the following:
        - An increase in platelet count from baseline
        - Maintenance of normal platelet counts and LDH levels for at least four weeks
        - A 25% reduction in serum creatinine for a minimum of four weeks
        - The member has not experienced one of the following for at least 12 weeks after initiation of treatment:
            - A decrease in platelet count of >25% from baseline
            - Plasma exchange or plasma infusion
            - New dialysis requirement
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided with a diagnosis of generalized Myasthenia Gravis (gMG) **(Soliris (eculizumab) ONLY)** and the following criteria are met:

- Medication is prescribed by, or in consultation with a neurologist
- Documentation of a positive serologic test for anti-acetylcholine antibodies
- Documentation the member meets the Myasthenia Gravis Foundation of America Clinical Classification II to IV generalized myasthenia gravis
- Documentation the member has a Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) total score ≥6
- Documentation of at least one of the following:
    - Failed treatment over 1 year or more with 2 or more immunosuppressive therapies either in combination or as monotherapy (e.g. azathioprine, cyclophosphamide, methotrexate)
    - Failed treatment over 1 year or more with at least 1 immunosuppressive therapy while on chronic plasmapheresis or plasma exchange (PE)
- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
    - First reauthorization criteria (member on therapy for 0 to 6 months)
        - Documentation from the provider that the member had a positive clinical response and tolerates Soliris therapy supported by at least one of the following:

    - A 3 point improvement in the member's total MG-ADL score
    - A 5 point improvement in QMG total score
  - Subsequent reauthorization criteria (member on therapy ≥ 6 months)
    - Documentation from the prescriber indicating stabilization or improvement in condition.
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided with a diagnosis of Neuromyelitis Optica Spectrum Disorder (NMOSD) **(Soliris (eculizumab) ONLY)** and the following criteria are met:

- Medication is prescribed by, or in consultation with a neurologist
- Documentation of a positive test for AQP4-IgG antibodies
- Documentation of at least 2 relapses in the last 12 months or 3 relapses in the last 24 months with at least 1 relapse in the last 12 months
- Documentation of an Expanded Disability Status Scale (EDSS) score of ≤ 7
- If using concurrent corticosteroids, dose is less than or equal to the equivalent of prednisone 20 mg per day
- Must have documentation of inadequate response, contraindication or intolerance to rituximab or any of its biosimilars.
- **Initial Duration of Approval:** 12 months
- **Reauthorization criteria**
  - Documentation from the prescriber indicating stabilization or improvement in condition.
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

Attachment 1. MG Activities of Daily Living (MG-ADL) profile

Grade	0	1	2	3	Score
Talking	Normal	Intermittent slurring or nasal speech	Constant slurring or nasal, but can be understood	Difficult to understand speech	
Chewing	Normal	Fatigue with solid food	Fatigue with soft food	Gastric tube	
Swallowing	Normal	Rare episode of choking	Frequent choking necessitating changes in diet	Gastric tube	
Breathing	Normal	Shortness of breath with exertion	Shortness of breath at rest	Ventilator dependence	
Impairment of ability to brush teeth or comb hair	None	Extra effort, but no rest periods needed	Rest periods needed	Cannot do one of these functions	
Impairment of ability to arise from a chair	None	Mild, sometimes uses arms	Moderate, always uses arms	Severe, requires assistance	
Double vision	None	Occurs, but not daily	Daily, but not constant	Constant	
Eyelid droop	None	Occurs, but not daily	Daily, but not constant	Constant	

Total score _____

Attachment 2. Quantitative Myasthenia gravis scale

Test Items Weakness	None	Mild	Moderate	Severe
Double vision on lateral gaze **right** or **left** (circle one)	61	11–60	1–10	spontaneous
Ptosis (upward gaze)	61	11–60	1–10	spontaneous
Facial muscles	normal lid	complete, weak, some resistance	complete, without resistance	incomplete
Swallowing 4 oz. water (½ cup)	normal	Minimal coughing or throat clearing	severe coughing/ choking or nasal regurgitation	cannot swallow (test not attempted)
Speech following counting aloud from 1 to 50 (onset of dysarthria)	none at #50	dysarthria at #30–49	dysarthria at #10–29	dysarthria at #9
Right arm outstretched (90° sitting)	240	90–239	10–89	0–9
Left arm outstretched (90° sitting)	240	90–239	10–89	0–9
Vital capacity (% predicted)	≥80%	65–79%	50–64%	<50%
Right hand grip (kg)				
male	≥45	15–44	5–14	0–4
female	≥30	10–29	5–9	0–4
Left hand grip (kg)				
male	≥35	15–34	5–14	0–4
female	≥25	10–24	5–9	0–4
Head lifted (45° supine)	120	30–119	1–29	0
Right leg outstretched (45° supine)	100	31–99	1–30	0
Left leg outstretched (45° supine)	100	31–99	1–30	0

[a]Total QMG score range 0–39.

Attachment 3. Myasthenia Gravis Foundation of America Clinical Classification

Class	Clinical symptoms
I	Any ocular weakness
II	Mild Weakness. May also have ocular muscle weakness of any severity
II A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
II B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
III	Moderate weakness affecting other than ocular muscles. May also have ocular muscle weakness of any severity
III A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
III B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
IV	Severe weakness affecting other than ocular muscles. May also have ocular muscle weakness of any severity
IV A	Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal, respiratory muscles or both
IV B	Predominantly affecting orорpharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles or both
V	Defined by intubation, with or without mechanical ventilation, except when employed during routine postoperative management

Attachment 4. Expanded Disability Status Scale (EDSS)

Score	Description
1.0	No disability, minimal signs in one functional system (FS)
1.5	No disability, minimal signs in more than one FS
2.0	Minimal disability in one FS
2.5	Mild disability in one FS or minimal disability in two FS
3.0	Moderate disability in one FS, or mild disability in three or four FS. No impairment to walking
3.5	Moderate disability in one FS and more than minimal disability in several others. No impairment to walking
4.0	Significant disability but self-sufficient and up and about some 12 hours a day. Able to walk without aid or rest for 500m
4.5	Significant disability but up and about much of the day, able to work a full day, may otherwise have some limitation of full activity or require minimal assistance. Able to walk without aid or rest for 300m
5.0	Disability severe enough to impair full daily activities and ability to work a full day without special provisions. Able to walk without aid or rest for 200m
5.5	Disability severe enough to preclude full daily activities. Able to walk without aid or rest for 100m
6.0	Requires a walking aid – cane, crutch, etc. – to walk about 100m with or without resting
6.5	Requires two walking aids – pair of canes, crutches, etc. – to walk about 20m without resting
7.0	Unable to walk beyond approximately 5m even with aid. Essentially restricted to wheelchair; though wheels self in standard wheelchair and transfers alone. Up and about in wheelchair some 12 hours a day
7.5	Unable to take more than a few steps. Restricted to wheelchair and may need aid in transfering. Can wheel self but cannot carry on in standard wheelchair for a full day and may require a motorised wheelchair
8.0	Essentially restricted to bed or chair or pushed in wheelchair. May be out of bed itself much of the day. Retains many self-care functions. Generally has effective use of arms
8.5	Essentially restricted to bed much of day. Has some effective use of arms retains some self-care functions
9.0	Confined to bed. Can still communicate and eat
9.5	Confined to bed and totally dependent. Unable to communicate effectively or eat/swallow
10.0	Death due to MS

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Updated: 06/2021
PARP Approved: 06/2021

C5B COMPLEMENT INHIBITORS (SOLIRIS AND ULTOMIRIS) PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Gateway Health℠ Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative.
PHONE: (800) 392-1147 Monday through Friday 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	NPI:
Provider Specialty:	Office Contact:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:
Gateway ID:	Member weight: _____pounds or _____kg

REQUESTED DRUG INFORMATION

Medication:	Strength:
Frequency:	Duration:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:
Is this medication being used for a chronic or long-term condition for which the medication may be necessary for the life of the patient? ☐ Yes ☐ No	

Billing Information

This medication will be billed: ☐ at a pharmacy **OR**
☐ medically (if medically please provide a JCODE:_____

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

REFERENCE VALUES

Lab	Initial (Pre-Treatment) Value	Reference Range	Date	Post-Therapy Value (Reauthorization only)	Reference Range	Date
Hemoglobin (Hgb)						
Lactate dehydrogenase (LDH)						
Platelet count						
Serum Creatinine						
Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) total score		**N/A**			**N/A**	
Quantitative Myasthenia Gravis (QMG) total score		**N/A**			**N/A**	

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C5B COMPLEMENT INHIBITORS (SOLIRIS AND ULTOMIRIS)
PRIOR AUTHORIZATION FORM (CONTINUED) – PAGE 2 OF 2

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Gateway Health℠ Pharmacy Services. **FAX:** (888) 245-2049
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PHONE: (800) 392-1147 Monday through Friday 8:30am to 5:00pm

MEMBER INFORMATION

Member Name:	DOB:
Gateway ID:	Member weight: _____pounds or _____kg

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis: _____

For Paroxysmal Nocturnal Hemoglobinuria (PNH) only:
Does the member's flow cytometry pathology report demonstrate at least 2 different GPI protein deficiencies within 2 different cell lines from granulocytes, monocytes, or erythrocytes? Please include a copy ☐ Yes ☐ No
Has the patient had a blood transfusion within the last 12 months? ☐ Yes ☐ No
Does the patient have symptoms of anemia? ☐ Yes ☐ No

For Atypical Hemolytic Uremic Syndrome only:
Has the absence of Shiga toxin-producing *E.coli* been confirmed? ☐ Yes ☐ No
Does the member have an ADAMTS13 deficiency? ☐ Yes ☐ No Is the member currently on dialysis? ☐ Yes ☐ No

For generalized Myasthenia Gravis only:
Does the patient have anti-acetylcholine antibodies? ☐ Yes ☐ No
What is the member's Myasthenia Gravis Foundation of America Clinical Classification? _____

For Neuromyelitis Optica Spectrum Disorder:
Is documentation of a positive test for AQP4-IgG antibodies provided? ☐ Yes ☐ No
What is the member's Expanded Disability Status Scale (EDSS) score? _____
Has the member had at least 2 relapses in the last 12 months or 3 relapses in the last 24 months with at least 1 relapse in the last 12 months? ☐ Yes ☐ No

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why / Current)

REAUTHORIZATION

Has the member experienced a significant improvement with treatment? ☐ Yes ☐ No If **Yes,** please include documentation

For Paroxysmal Nocturnal Hemoglobinuria (PNH) only:
Has the patient had a blood transfusion since taking Soliris? ☐ Yes ☐ No

For Atypical Hemolytic Uremic Syndrome only:
Has the patient been able to maintain a normal platelet or LDH level for at least four weeks? ☐ Yes ☐ No
Has the patient experienced a 25% serum creatinine reduction for at least four weeks? ☐ Yes ☐ No
In the past 12 weeks, has the patient had any of the following?
A decrease in platelet count of >25% from baseline ☐ Yes ☐ No
Increased need for plasma exchange or plasma infusion ☐ Yes ☐ No
New dialysis requirement ☐ Yes ☐ No

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date