RX.PA.054.MPC Ultomiris

PURPOSE

The purpose of this policy is to define the prior authorization process for Ultomiris (ravulizumab-cwvz).

Ultomiris (ravulizumab-cwvz) is indicated for the following:

- Treatment of adult and pediatric patients one month of age and older with paroxysmal nocturnal hemoglobinuria (PNH)
- 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)
- Treatment of adult patients with generalized myasthenia gravis (gMG) who are anti-acetylcholine receptor (AChR) antibody-positive
- Treatment of Neuromyelitis optica spectrum disorder (NMOSD)

DEFINITIONS

Atypical Hemolytic Uremic Syndrome (aHUS) – a rare autoimmune disorder that results in low red blood cell counts, low platelet counts, and acute renal failure

Paroxysmal Nocturnal Hemoglobinuria (PNH) – a rare disorder where the immune system attacks red blood cells, resulting in anemia and thrombosis

PROCEDURE

A. <u>Initial Authorization Criteria:</u>

Must meet all of the criteria listed under the respective diagnosis:

1. Paroxysmal Nocturnal Hemoglobinuria (PNH)

- Must be prescribed by or in consultation with a hematologist, oncologist, immunologist or genetic specialist
- Must have a laboratory confirmed diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) as evidenced by having detectable GPI-deficient hematopoietic clones (Type III PNH RBC) via Flow Cytometry. Documentation of Flow Cytometry pathology report support must indicate presence of PNH-type RBC (red blood cell) and must be submitted.

- Must have an LDH level of 1.5 times the upper limit of the normal range

(laboratory result with reference range must be submitted)

- Must provide documentation that a meningococcal vaccine was given at least two (2) weeks prior to the administration of the first dose of Ultomiris
- Must have documentation of an adequate trial of at least 3 months with Soliris and experienced an inadequate response/significant side effects/toxicity or have a contraindication to this treatment
- Ultomiris is not prescribed concurrently with Empaveli or Soliris, unless the member is in a 4-week cross-titration between Soliris and Empaveli
- Verification prescriber and patient are enrolled in Ultomiris REMS Program

2. Atypical Hemolytic Uremic Syndrome (aHUS)

- Must be prescribed by or in consultation with a nephrologist, hematologist, oncologist, immunologist or genetic specialist
- Must have a diagnosis of atypical hemolytic uremic syndrome
- Patient must weigh ≥ 5kg
- ADAMTS 13 activity level above 5%
- Absence of Shiga toxin
- Must have documentation of an adequate trial of at least 3 months with Soliris and experienced an inadequate response/significant side effects/toxicity or have a contraindication to this treatment
- Ultomiris is not prescribed concurrently with Soliris
- Must provide documentation that a meningococcal vaccine was given at least two (2) weeks prior to the administration of the first dose of Ultomiris
- Verification prescriber and patient are enrolled in Ultomiris REMS Program

3. Generalized Myasthenia Gravis (gMG)

- Must be prescribed by or in consultation with a neurologist
- Must have a diagnosis of Myasthenia Gravis
- Member must be 18 years of age or older
- Must be anti-acetylcholine receptor (AChR) antibody positive
- Myasthenia Gravis Foundation of America (MGFA) clinical classification II to IV
- Myasthenia Gravis-Activities of Daily Living (MG-ADL) score greater than or equal to 6 at baseline
- Documented intolerance, contraindication, or failed treatment to at least two immunosuppressive therapies listed below:
    - Azathioprine
    - Cyclosporine
    - Mycophenolate mofetil
    - Tacrolimus
    - Methotrexate
    - Cyclophosphamide
- Must have documentation of an adequate trial of at least 3 months with Soliris and experienced an inadequate response/significant side effects/toxicity or have a contraindication to this treatment
- Ultomiris is not prescribed concurrently with Soliris

- Must provide documentation that a meningococcal vaccine was given at least two (2) weeks prior to the administration of the first dose of Ultomiris
- Verification prescriber and patient are enrolled in Ultomiris REMS Program

4. Neuromyelitis Optica Spectrum Disorder (NMOSD)

- Must be prescribed by or in consultation with a neurologist
- Must have a diagnosis of neuromyelitis optica spectrum disorder
- Member must be 18 years of age or older
- Must be anti-aquaporin-4 (AQP4) antibody positive
- Member exhibits one of the following core clinical characteristics of NMOSD:
    - Optic neuritis
    - Acute myelitis
    - Area postrema syndrome
    - Acute brainstem syndrome
    - Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
    - Symptomatic cerebral syndrome with NMOSD-typical brain lesions
- Must provide documentation that a meningococcal vaccine was given at least two (2) weeks prior to the administration of the first dose of Ultomiris
- Member must not be receiving the requested drug concomitantly with other biologics indicated for NMOSD
- Verification prescriber and patient are enrolled in Ultomiris REMS Program

B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.

C. Ultomiris will be considered investigational or experimental for any other use and will not be covered.

D. <u>Reauthorization Criteria:</u>

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon:

<u>MPC Renewal:</u>

- Chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.
- For Generalized myasthenia gravis (gMG) and Neuromyelitis optica spectrum disorder (NMOSD):
    - Must be prescribed by or in consultation with a neurologist
- For Paroxysmal Nocturnal Hemoglobinuria (PNH):
    - Must be prescribed by or in consultation with a hematologist, oncologist, immunologist or genetic specialist

- For Atypical Hemolytic Uremic Syndrome (aHUS):
    - Must be prescribed by or in consultation with a nephrologist, hematologist, oncologist, immunologist or genetic specialist

Renewal from Previous Insurer:

- Members who have received prior approval (from insurer other than MPC), or have been receiving medication samples, should be considered under criterion A (Initial Authorization Criteria)
- Provider has documented clinical response of the member's condition which has stabilized or improved based upon the prescriber's assessment

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 3 months
Reauthorization	Up to 1 year

If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.

APPLICABLE CODES:	
CODE	**DESCRIPTION**
J1303	Injection, ravulizumab-cwvz, 10mg
C9052	Injection, ravulizumab-cwvz, 10mg

REFERENCES

1. Ultomiris Prescribing Information. Boston, MA: Alexion Pharmaceuticals, Inc.; October 2019. Available at: www.ultomiris.com. Accessed October 18, 2022.

2. Parker C, Omine M, Richards S, et al. Diagnosis and management of paroxysmal nocturnal hemoglobinuria. Blood 2005; 106(12):3699-3709. doi:10.1182/blood-2005-04-1717.

3. Loirat C, Fakhouri F, Ariceta G, et al. An international consensus approach to the management of atypical hemolytic uremic syndrome in children. Pediatr Nephrol. 2016; 31: 15-39

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review	*02/2026*

Annual Review	*02/2025*
Selected Review *Addition of criteria requirements for Neuromyelitis optica spectrum disorder (NMOSD)*	*11/2024*
Annual Review *Change in Non-MPC renewal to renewal from previous insurer*	*02/2024*
Selected Review *Addition of trial and failure with Soliris for all indications*	*12/2023*
Annual Review	*02/2023*
New Policy	*10/2022*