Ultomiris® (ravulizumab-cwvz)
(Intravenous/Subcutaneous)

Document Number: IH-0427

Last Review Date: 05/02/2024
Date of Origin: 02/04/2019
Dates Reviewed: 02/2019, 10/2019, 12/2019, 11/2020, 07/2021, 10/2021, 06/2022, 09/2022, 9/2023, 05/2024

I. Length of Authorization

Coverage will be provided for twelve (12) months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Ultomiris 10 mg/mL** – 30 mL SDV: 10 vials on day zero followed by 13 vials starting on day 14 and every 8 weeks thereafter
- Ultomiris 100 mg/mL – 3 mL SDV: 10 vials on day zero followed by 13 vials starting on day 14 and every 8 weeks thereafter
- Ultomiris 100 mg/mL – 11 mL SDV: 3 vials on day zero followed by 3 vials starting on day 14 and every 8 weeks thereafter
- Ultomiris 245 mg/3.5 mL single-dose cartridge on-body delivery system: 2 on-body delivery systems weekly

B. Max Units (per dose and over time) [HCPCS Unit]:

- Ultomiris IV
  - PNH/aHUS/gMG/NMOSD: 300 units on Day 0 followed by 360 units on Day 14 and every 8 weeks thereafter
- Ultomiris SQ
  - PNH/aHUS: 49 units weekly

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Patient is at least 18 years of age *(unless otherwise specified)*; AND
- Confirmation that patient does not have an unresolved serious Neisseria meningitidis infection prior to initiating therapy; AND

Universal Criteria [1]

- Prescriber is enrolled in the Ultomiris and Soliris Risk Evaluation and Mitigation Strategy (REMS) program; **AND**
- Patient must be vaccinated against meningococcal infection (serogroups A, C, W, Y and B) according to current ACIP recommendations at least two weeks prior to initiation of therapy and will continue to be revaccinated in accordance with ACIP recommendations *(Note: If urgent Ultomiris therapy is indicated in a patient who is not up to date with meningococcal vaccines according to ACIP recommendations, provide the patient with antibacterial drug prophylaxis and administer these vaccines as soon as possible.)*; **AND**
- Will not be used in combination with other immunomodulatory biologic therapies (e.g., efgartigimod, efgartigimod-hyaluronidase, eculizumab, pegcetacoplan, satralizumab, tocilizumab, inebilizumab, rozanolixizumab, rituximab, zilucoplan, pozelimab, etc.); **AND**

Paroxysmal Nocturnal Hemoglobinuria (PNH) † Φ [1,4,8,9,18]

- Patient is at least 1 month of age; **AND**
    - Used as switch therapy; **AND**
        - Patient is currently receiving treatment with eculizumab and has shown a beneficial disease response and absence of unacceptable toxicity while on therapy; **OR**
    - Patient is complement inhibitor treatment-naïve; **AND**
        - Diagnosis must be confirmed by detection of PNH clones of at least 5% by flow cytometry testing; **AND**
        - Patient has at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes); **AND**
        - Patient has laboratory evidence of significant intravascular hemolysis (i.e., LDH ≥1.5 x ULN) with symptomatic disease and at least one other indication for therapy from the following (regardless of transfusion dependence):
            - Patient has symptomatic anemia (i.e., hemoglobin < 7 g/dL or hemoglobin < 10 g/dL, in at least two independent measurements in a patient with cardiac symptoms)
            - Presence of a thrombotic event related to PNH
            - Presence of organ damage secondary to chronic hemolysis (i.e., renal insufficiency, pulmonary insufficiency/hypertension)
            - Patient is pregnant and potential benefit outweighs potential fetal risk
            - Patient has disabling fatigue

- Patient has abdominal pain (requiring admission or opioid analgesia), dysphagia, or erectile dysfunction; **AND**

- Documented baseline values for one or more of the following (necessary for renewal): serum lactate dehydrogenase (LDH), hemoglobin level, packed RBC transfusion requirement, and history of thrombotic events

Atypical Hemolytic Uremic Syndrome (aHUS) † Φ [1,5,7,19-21,26]

- Patient is at least 1 month of age; **AND**
  - Used as switch therapy; **AND**
    - Patient is currently receiving treatment with eculizumab and has shown a beneficial disease response and absence of unacceptable toxicity while on therapy; **OR**
  - Patient is complement inhibitor treatment-naïve; **AND**
    - Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.); **AND**
    - Thrombotic Thrombocytopenic Purpura (TTP) has been ruled out by evaluating ADAMTS13 level (ADAMTS13 activity level ≥ 10%); **AND**
    - Shiga toxin *E. coli* related hemolytic uremic syndrome (STEC-HUS) has been ruled out; **AND**
    - Other causes have been ruled out such as coexisting diseases or conditions (e.g., bone marrow transplantation, solid organ transplantation, malignancy, autoimmune disorder, drug-induced, malignant hypertension, HIV infection, Streptococcus pneumoniae sepsis or known genetic defect in cobalamin C metabolism, etc.); **AND**
    - Documented baseline values for one or more of the following (necessary for renewal): serum lactate dehydrogenase (LDH), serum creatinine/eGFR, platelet count, and plasma exchange/infusion requirement

Generalized Myasthenia Gravis (gMG) † Φ [1,11,12-17]

- Patient has tried and failed to respond to or tolerate treatment with Vyvgart™ (efgartigimod alfa-fcab) unless a contraindication exists; **AND**

- Used as switch therapy; **AND**
  - Patient is currently receiving treatment with eculizumab and has shown a beneficial disease response and absence of unacceptable toxicity while on therapy; **OR**
- Patient is complement inhibitor treatment-naïve; **AND**
  - Patient has Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of Class II to IV disease§; **AND**

- Patient has a positive serologic test for anti-acetylcholine receptor (AChR) antibodies; **AND**
- Patient has had a thymectomy (*Note: Applicable only to patients with thymomas OR non-thymomatous patients who are 50 years of age or younger)*; **AND**
- Physician has assessed objective signs of neurological weakness and fatiguability on a baseline neurological examination (e.g., including, but not limited to, the Quantitative Myasthenia Gravis [QMG] score, etc.); **AND**
- Patient has a baseline MG-Activities of Daily Living (MG-ADL) total score of ≥6; **AND**
  - Patient has had an inadequate response after a minimum one-year trial of concurrent use with two (2) or more immunosuppressive therapies (e.g., corticosteroids plus an immunosuppressant such as azathioprine, cyclosporine, mycophenolate, etc.); **OR**
  - Patient required chronic treatment with plasmapheresis or plasma exchange (PE) or intravenous immunoglobulin (IVIG) in addition to immunosuppressant therapy; **AND**
- Patient will avoid or use with caution medications known to worsen or exacerbate symptoms of MG (e.g., certain antibiotics, beta-blockers, botulinum toxins, hydroxychloroquine, etc.)

§Myasthenia Gravis Foundation of America (MGFA) Disease Clinical Classification [14]:

– **Class I**: Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal.
– **Class II**: Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IIa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IIb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
– **Class III**: Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IIIa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IIIb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
– **Class IV**: Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IVa**. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IVb**. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
– **Class V**: Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVb.

Neuromyelitis Optica Spectrum Disorder (NMOSD) † Φ [1,22-25]

- Used as switch therapy; **AND**

- Patient is currently receiving treatment with eculizumab and has shown a beneficial disease response and absence of unacceptable toxicity while on therapy; **OR**

- Patient is complement inhibitor treatment-naïve; **AND**
  - Patient has a confirmed diagnosis based on the following:
    - Patient was found to be seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**
    - Patient has at least one core clinical characteristic § *(*Note: some core clinical characteristics require both clinical and typical MRI findings)*; **AND**
    - Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.]; **AND**
  - Patient has a history of at least 1 relapse in the last 12 months; **AND**
  - Patient has an Expanded Disability Status Score (EDSS) of ≤ 7.0; **AND**
  - Patients who are receiving concurrent immunosuppressive therapy (e.g., corticosteroids, azathioprine, mycophenolate mofetil, methotrexate, tacrolimus, etc.) are on a stable dose regimen; **AND**
  - Patient has not received therapy with rituximab or mitoxantrone in the last 3 months; **AND**
  - Patient has not received intravenous immune globulin (IVIG) in the last 3 weeks

§ Core Clinical Characteristics of NMOSD [22,23]
▪ Acute optic neuritis ▪ Acute myelitis ▪ Acute area postrema syndrome (APS): episode of otherwise unexplained hiccups and/or nausea and vomiting (lasting for at least 48 hours or with MRI evidence of a dorsal brainstem lesion) ▪ Acute brainstem syndrome other than APS ▪ Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on MRI ¥ ▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI ψ
¥ Diencephalic syndrome: Periependymal lesion (3rd ventricle) OR hypothalamic/thalamic lesion ψ Cerebral syndrome: Extensive periependymal lesion (lateral ventricle; often with Gd) OR long (> 1⁄2 length), diffuse, heterogeneous or edematous corpus callosum lesion OR long corticospinal tract lesion (unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle) OR large, confluent (unilateral or bilateral) subcortical or deep white matter lesion

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria [1]

Coverage may be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: serious meningococcal infections (septicemia and/or meningitis), infusion-related reactions, other serious infections, etc.; **AND**

Paroxysmal Nocturnal Hemoglobinuria (PNH) [1,4,8,18]

- Patient has not developed severe bone marrow failure syndrome (i.e., aplastic anemia or myelodysplastic syndrome) OR experienced a spontaneous disease remission OR received curative allogeneic stem cell transplant; **AND**
- Disease response compared to pretreatment baseline as indicated by one or more of the following:
    - Decrease in serum LDH
    - Stabilization/improvement in hemoglobin level
    - Decrease in packed RBC transfusion requirement (i.e., reduction of at least 30%)
    - Reduction in thromboembolic events

Atypical Hemolytic Uremic Syndrome (aHUS) [1,5,7]

- Disease response compared to pretreatment baseline indicated by one or more of the following:
    - Decrease in serum LDH
    - Stabilization/improvement in serum creatinine/eGFR
    - Increase in platelet count
    - Decrease in plasma exchange/infusion requirement

Generalized Myasthenia Gravis (gMG) [1,11-17]

- Patient has had an improvement (i.e., reduction) of at least 1-point from baseline in the Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score **Δ**; **AND**
- Improvement in muscle strength testing with fatigue maneuvers as evidenced on neurologic examination when compared to baseline

*[**Δ** May substitute an improvement of at least 1-point from baseline in the Quantitative Myasthenia Gravis (QMG) total score, if available]*

NMOSD [1,24]

- Disease response as indicated by stabilization/improvement in one or more of the following:
    - Neurologic symptoms as evidenced by a decrease in acute relapses, improvement of stability, or improvement in EDSS
    - Reduced hospitalizations
    - Reduction/discontinuation in plasma exchange treatments

Switch Therapy From Eculizumab to Ravulizumab

- Refer to Section III for criteria

V. Dosage/Administration [1]

<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td>All Indications</td><td>
<u>IV Dosing for Complement-Inhibitor Therapy Naïve*</u>
Administer the INTRAVENOUS doses based on the patient's body weight. Starting 2 weeks after the loading dose, begin maintenance doses once every 4 weeks or every 8 weeks (depending on body weight)
<table>
<tr><th>Indications</th><th>Body Weight Range</th><th>Loading Dose (mg)</th><th>Maintenance Dose (mg)</th><th>Dosing Interval</th></tr>
<tr><td rowspan="4">PNH, aHUS</td><td>5 kg to <10 kg</td><td>600</td><td>300</td><td rowspan="2">Every 4 weeks</td></tr>
<tr><td>10 kg to <20 kg</td><td>600</td><td>600</td></tr>
<tr><td>20 kg to <30 kg</td><td>900</td><td>2,100</td><td rowspan="2">Every 8 weeks</td></tr>
<tr><td>30 kg to <40 kg</td><td>1,200</td><td>2,700</td></tr>
<tr><td rowspan="3">PNH, aHUS, gMG, or NMOSD</td><td>40 kg to <60 kg</td><td>2,400</td><td>3,000</td><td rowspan="3">Every 8 weeks</td></tr>
<tr><td>60 kg to <100 kg</td><td>2,700</td><td>3,300</td></tr>
<tr><td>100 kg or greater</td><td>3,000</td><td>3,600</td></tr>
</table>
<u>IV Dosing for Switch Therapy from Eculizumab OR Ravulizumab SQ to Ravulizumab IV*</u>
<table>
<tr><th>Population</th><th>Weight-based Ravulizumab IV Loading Dose</th><th>Time of First Ravulizumab IV Weight-based Maintenance Dose</th></tr>
<tr><td>Currently treated with eculizumab</td><td>At time of next scheduled eculizumab dose</td><td>2 weeks after ravulizumab IV loading dose</td></tr>
<tr><td>Currently treated with ravulizumab SQ on-body delivery system§</td><td>Not applicable</td><td>1 week after last ravulizumab SQ maintenance dose</td></tr>
</table>
<u>SQ Dosing for Complement-Inhibitor Therapy Naïve §</u>
PNH & aHUS (adult patients weighing ≥40 kg ONLY): 490 mg SQ via on-body injector once weekly starting 2 weeks after the initial IV weight-based loading dose (see IV weight-based dosing table above)

<u>SQ Dosing for Switch Therapy from Eculizumab OR Ravulizumab IV to Ravulizumab SQ §</u>
<table>
<tr><th>Population</th><th>Weight-based Ravulizumab IV Loading Dose</th><th>Time of First Ravulizumab SQ Maintenance Dose</th></tr>
<tr><td>Currently treated with eculizumab</td><td>At time of next scheduled eculizumab dose</td><td>2 weeks after ravulizumab IV loading dose</td></tr>
</table>
</td></tr>
</table>

	Currently treated with ravulizumab IV	Not applicable	8 weeks after last ravulizumab IV maintenance dose

*****Note: For Supplemental Dose Therapy after plasma exchange (PE), plasmapheresis (PP), and intravenous immunoglobulin (IVIg), please refer to the ravulizumab package insert for appropriate dosing.*

***§** Adult patients with PNH and aHUS only*

VI. Billing Code/Availability Information

HCPCS Code:

- J1303 – Injection, ravulizumab-cwvz, 10 mg; 1 billable unit = 10 mg

NDC(s):

- Ultomiris 300 mg/3 mL single-dose vial for injection: 25682-0025-xx
- Ultomiris 300 mg/30 mL single-dose vial for injection: 25682-0022-xx**
- Ultomiris 1,100 mg/11 mL single-dose vial for injection: 25682-0028-xx
- Ultomiris 245 mg/3.5 mL single-dose cartridge on-body subcutaneous delivery system: 25682-0031-xx

***Note: This NDC has been discontinued as of 06/11/2021.*

VII. References

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Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D59.32	Hereditary hemolytic-uremic syndrome
D59.39	Other hemolytic-uremic syndrome
D59.5	Paroxysmal nocturnal hemoglobinuria [Marchiafava-Micheli]
G36.0	Neuromyelitis optica [Devic]
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims

payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC