BlueCross BlueShield
Federal Employee Program.

ULTOMIRIS
(ravulizumab-cwvz)

Pre - PA Allowance

None

Prior-Approval Requirements

Age 1 month of age or older

Diagnoses

Patient must have **ONE** of the following:

1. Paroxysmal nocturnal hemoglobinuria (PNH)
    a. 1 month of age or older
    b. Documented baseline value for serum lactate dehydrogenase (LDH)
    c. NO dual therapy with another Prior Authorization (PA) medication for PNH (see Appendix 1)

2. Atypical hemolytic uremic syndrome (aHUS)
    a. 1 month of age or older
    b. Documented baseline value for serum lactate dehydrogenase (LDH)
    c. Patient does **NOT** have Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS)
    d. **NO** dual therapy with another Prior Authorization (PA) medication for aHUS (see Appendix 2)

3. Generalized myasthenia gravis (gMG)
    a. 18 years of age or older
    b. Positive serologic test for anti-AChR antibodies
    c. Myasthenia Gravis Foundation of America (MGFA) clinical classification class II to IV
    d. Documented baseline Myasthenia Gravis-Activities of Daily Living (MG-ADL) total score ≥ 6 (*http://c.peerview.com/inReview/programs/150204324/downloads/PVI_practiceaids_RMU.pdf*)
    e. Patient has had an inadequate treatment response, intolerance, or contraindication to an acetylcholinesterase inhibitor and at least **ONE** immunosuppressive therapy either in combination or as monotherapy, such as:
        i. azathioprine

ii. cyclosporine
iii. mycophenolate mofetil
iv. tacrolimus
v. methotrexate
vi. cyclophosphamide

f. **NO** dual therapy with another Prior Authorization (PA) C5 complement inhibitor for gMG (see Appendix 3)

4. Neuromyelitis optica spectrum disorder (NMOSD)
   a. 18 years of age or older
   b. Anti-aquaporin-4 (AQP4) antibody positive
   c. **NO** dual therapy with another Prior Authorization (PA) C5 complement inhibitor for NMOSD (see Appendix 4)

AND ALL of the following:

a. Vaccination against Neisseria meningitidis at least 2 weeks prior to initiation [unless Ultomiris (ravulizumab-cwvz) treatment cannot be delayed]
b. Prescriber is enrolled in Ultomiris REMS program

Prior - Approval Limits

Duration 6 months

Prior – Approval *Renewal* Requirements

Age 1 month of age or older

Diagnoses

Patient must have **ONE** of the following:

1. Paroxysmal nocturnal hemoglobinuria (PNH)
   a. 1 month of age or older
   b. Decrease in serum LDH from pretreatment baseline
   c. **NO** dual therapy with another Prior Authorization (PA) medication for PNH (see Appendix 1)

2. Atypical hemolytic uremic syndrome (aHUS)
   a. 1 month of age or older
   b. Decrease in serum LDH from pretreatment baseline
   c. Patient does **NOT** have Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS)

d. **NO** dual therapy with another Prior Authorization (PA) medication for aHUS (see Appendix 2)

3. Generalized myasthenia gravis (gMG)
    a. 18 years of age or older
    b. Decrease of MG-ADL total score from baseline of ≥ 2 points (*http://c.peerview.com/inReview/programs/150204324/downloads/PVI_practiceaids_RMU.pdf*)
    c. **NO** dual therapy with another Prior Authorization (PA) C5 complement inhibitor for gMG (see Appendix 3)
4. Neuromyelitis optica spectrum disorder (NMOSD)
    a. 18 years of age or older
    b. Patient has had fewer relapses while on Ultomiris therapy
    c. **NO** dual therapy with another Prior Authorization (PA) C5 complement inhibitor for NMOSD (see Appendix 4)

AND ALL of the following:

a. Absence of unacceptable toxicity from the drug
b. Prescriber is enrolled in Ultomiris REMS program

Prior - Approval *Renewal* Limits

Duration 12 months

ULTOMIRIS
(ravulizumab-cwvz)

Appendix 1 - List of PA Medications for PNH

Generic Name	Brand Name
eculizumab	Soliris
pegcetacoplan	Empaveli
ravulizumab-cwvz	Ultomiris

Appendix 2 - List of PA Medications for aHUS

Generic Name	Brand Name
eculizumab	Soliris
ravulizumab-cwvz	Ultomiris

Appendix 3 - List of PA C5 complement inhibitors for gMG

Generic Name	Brand Name
eculizumab	Soliris
ravulizumab-cwvz	Ultomiris

Appendix 4 - List of PA C5 complement inhibitors for NMOSD

Generic Name	Brand Name
eculizumab	Soliris
ravulizumab-cwvz	Ultomiris