Ravulizumab (Ultomiris) – PA Criteria

HCPC: J1303

Ultomiris is a C5 Complement Inhibitor indicated for the treatment of anti-ACHR+ generalized myasthenia gravis (gMG), atypical hemolytic uremic syndrome (aHUS), paroxysmal nocturnal hemoglobinuria (PNH) and neuromyelitis optica spectrum disorder (NMOSD). It is covered by South Dakota Medicaid following prior authorization when the patient meets the following criteria:

- **Initial Therapy (must meet all):**
  - Therapy meets the following criteria as specified per indication
    - For gMG **(must meet all)**:
      - Therapy is requested by or in consultation with a neurologist
      - Individual is ≥18 years of age
      - Individual has a documented diagnosis of gMG with labs confirming presence of anti-ACHR antibodies
      - Individual has a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of Class II-IV disease
      - Documentation is submitted indicating the patient's Baseline Quantitative Myasthenia Gravis (QMG) score
      - MG-Activities of Daily Living (MG-ADL) total score of ≥6
      - Documentation is provided indicating inadequate response or contraindication to pyridostigmine and corticosteroids
      - Individual has failed treatment with at least 2 immunosuppressive therapies (ex., azathioprine, cyclosporine, mycophenolate, cyclophosphamide, methotrexate, tacrolimus etc.) over the course of the last 12 months OR has failed at least 1 immunosuppressive therapy and required chronic plasmapheresis, plasma exchange (PE) or intravenous immunoglobulin (IVIG)
      - Documentation is provided indicating previous failure, contraindication or intolerance to efgartigimod alfa (Vyvgart)
      - Therapy is not prescribed in combination with other biologics for gMG (e.g. Soliris, Rystiggo, Vyvgart, etc.)
      - Approval duration: 6 months
    - For aHUS **(must meet all)**:
    - ***Retrospective authorization will be accepted for this medication when used for aHUS (when all below criteria are met) in efforts to prevent delay of care***
      - Therapy is requested by or in consultation with a hematologist or nephrologist
      - Individual has a documented diagnosis of aHUS that is not Shiga toxin E. coli related
      - Individual is ≥ 1 month of age
      - Documentation is provided indicating baseline values for the following:
        - Serum lactate dehydrogenase (LDH)
        - Serum creatinine/eGFR
        - Platelet count
        - Frequency of plasma exchange/infusion requirement
      - Therapy is not prescribed in combination with other biologics for aHUS (e.g.Soliris)
      - Approval duration: 6 months
    - For PNH **(must meet all)**:
      - Therapy is requested by or in consultation with a hematologist, oncologist, or immunologist

- Individual is ≥ 1 month of age
- Individual has a documented diagnosis of PNH confirmed by flow cytometry
- Individual is transfusion dependent (requiring ≥ 4 transfusions in past 12 months) as a result of PNH and documentation is provided indicating the frequency of transfusions
- Documentation is provided indicating baseline values for hemoglobin and lactate dehydrogenase (LDH)
- Documentation of one or more of the following indicating systemic complications: abdominal pain, dysphagia/odynophagia, shortness of breath, chest pain/pressure, hemoglobinuria, end organ damage, thrombosis, etc.
- Therapy is not prescribed in combination with other biologics for PNH (e.g. Ultomiris, Piasky, Fabhalta, Empaveli)
- Approval duration: 6 months

▪ For NMOSD **(must meet all)**:

- Therapy is requested by or in consultation with a neurologist
- Individual is ≥18 years of age
- Individual has a documented diagnosis of NMOSD with presence of seropositive aquaporin-4 (AQP4) antibodies
- Individual has symptoms consistent with **at least one** core clinical characteristic below:
  - Optic neuritis
  - Acute myelitis
  - Area postrema syndrome or episode of otherwise unexplained hiccups or nausea and vomiting
  - Acute brainstem syndrome
  - Symptomatic narcolepsy
  - Acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
  - Symptomatic cerebral syndrome with NMOSD-typical brain lesions
- Documentation is provided indicating previous failure, contraindication or intolerance to **at least two** of the following (with at least one being a C5 complement inhibitor):
  - Rituximab
  - Tocilizumab
  - Inebilizumab (Uplizna)
  - Satralizumab-mwge (Enspryng)
  - Eculizumab (Soliris)
- Documentation is provided indicating at least one relapse in the last 12 months or two relapses in the last 2 years
- Therapy is not being used in combination with other biologics for NMOSD (e.g. eculizumab, rituximab, satralizumab, tocilizumab, etc.)
- Individual has an Expanded Disability Status Score (EDSS) score documented at baseline
- Approval duration: 12 months

- **Continuation of Therapy (must meet all):**
  - Therapy is not prescribed in combination with other biologics for the requested indication
  - Therapy meets the following criteria as specified per indication
    - For gMG renewal (must meet **at least one** of the following):
      - Improvement (reduction in score) in the Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score from pretreatment baseline
      - Improvement in the Quantitative Myasthenia Gravis (QMG) total score
      - Approval duration: 1 year

- For aHUS renewal:
  - Individual has had a positive clinical response as indicated by **one or more** of the following:
    - Decrease in serum LDH from pretreatment baseline
    - Stabilization/improvement in renal function (serum creatinine/eGFR) from pretreatment baseline
    - Increase in platelet count from pretreatment baseline
    - Decrease in plasma exchange/infusion requirement from pretreatment baseline
  - Approval duration: 1 year
- For PNH renewal (must meet all):
  - Individual has had a positive clinical response as indicated by **one or more** of the following:
    - Stabilization or decrease in serum LDH from pretreatment baseline
    - Stabilization/improvement in hemoglobin level from pretreatment baseline
    - Decrease in packed RBC transfusion requirement from pretreatment baseline
    - Approval duration: 1 year
- For NMOSD renewal (must meet all):
  - Individual has had a positive clinical response with reduction in number and/or severity of relapses or signs and symptoms of NMOSD
  - Documentation is provided indicating recent (within 2 months) EDSS
  - Approval duration: 1 year