NOVA SCOTIA PROVINCIAL PHARMACARE PROGRAMS

Initial Request for Insured Coverage of Ravulizumab (Ultomiris) for aHUS

PATIENT INFORMATION			
PATIENT SURNAME	**PATIENT GIVEN NAME**	**HEALTH CARD NUMBER**	**DATE OF BIRTH**
PATIENT ADDRESS			**PATIENT WEIGHT (KG)**

INITIAL REQUEST

For renewal requests, please refer to the separate renewal form.

1. For the treatment of adult and pediatric patients 1 month of age and older with atypical hemolytic uremic syndrome (aHUS) who meet all of the following criteria:

☐ **A. Confirmed diagnosis of aHUS at initial presentation, defined by presence of thrombotic microangiopathy (TMA), who meet all the following criteria:**

- ☐ **i.** A disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13 (ADAMTS-13) activity ≥ 10% on blood samples taken before plasma exchange or plasma infusion (PE/PI); **AND**
- ☐ **ii.** Shiga toxin–producing Escherichia coli (STEC) test negative in patients with a history of bloody diarrhea in the preceding 2 weeks; **AND**
- ☐ **iii.** TMA must be unexplained (not a secondary TMA)

☐ **B. Evidence of ongoing active TMA and progressing, defined by laboratory test abnormalities despite plasmapheresis, if appropriate. Patients must demonstrate:**

- ☐ **i**. Unexplained (not a secondary TMA) thrombocytopenia (platelet count < 150 × 10^9/L); and hemolysis as indicated by the documentation of 2 of the following: schistocytes on the blood film; low or absent haptoglobin; or lactate dehydrogenase (LDH) above normal. **OR**
- ☐ **ii.** Tissue biopsy confirms TMA in patients who do not have evidence of platelet consumption and hemolysis.

☐ **C. Evidence of at least 1 of the following documented clinical features of active organ damage or impairment:**

- ☐ **i. Kidney impairment, as demonstrated by one of the following:**
  - ☐ 1. A decline in estimated glomerular filtration rate (eGFR) of > 20% in a patient with pre-existing renal impairment; **AND/OR**
  - ☐ 2. Serum creatinine (SCr) > upper limit of normal (ULN) for age or GFR < 60mL/min and renal function deteriorating despite prior PE/PI in patients who have no history of preexisting renal impairment (i.e., who have no baseline eGFR measurement); **OR**
  - ☐ 3. SCr > the age-appropriate ULN in pediatric patients (as determined by or in consultation with a pediatric nephrologist)
- ☐ **ii. The onset of neurological impairment related to TMA.**
- ☐ **iii. Other TMA-related manifestations, such as cardiac ischemia, bowel ischemia, pancreatitis, and retinal vein occlusion.**

2. For transplant patients with a documented history of aHUS (i.e., history of TMA [not a secondary TMA only] with ADAMTS 13 > 10%) who meet the following criteria:

- ☐ A. Develop TMA immediately (within hours to 1 month) following a kidney transplant; **OR**
- ☐ B. Previously lost a native or transplanted kidney due to the development of TMA; **OR**
- ☐ C. Have a history of proven aHUS and require prophylaxis with ravulizumab at the time of a kidney transplant

Exclusion Criteria

Patients should not have a history of ravulizumab treatment failure (i.e., treated with ravulizumab with a previous aHUS recurrence). Treatment failure is defined as:

☐ Yes	☐ No	**A.** Dialysis-dependent at 6 months, and failed to demonstrate resolution or stabilization of neurological or extrarenal complications if these were originally present; **OR**
☐ Yes	☐ No	**B**. On dialysis for ≥ 4 of the previous 6 months while receiving ravulizumab and failed to demonstrate resolution or stabilization of neurological or extrarenal complications if these were originally present; **OR**
☐ Yes	☐ No	**C.** Worsening of kidney function with a reduction in eGFR or increase in SCr ≥ 25% from baseline.

PRESCRIBER NAME & ADDRESS:

LICENCE # | **PRESCRIBER SIGNATURE** | **DATE**

If you need assistance, please contact the Pharmacare Office at (902) 496-7001 or 1-800-305-5026

Please Return Form To: Nova Scotia Pharmacare Programs
P.O. Box 500, Halifax, NS B3J 2S1; Fax: (902) 496-4440

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