

PRIOR AUTHORIZATION POLICY

POLICY: Uplizna Prior Authorization Policy

- Uplizna® (inebilizumab-cdon intravenous infusion – Horizon Therapeutics)

REVIEW DATE: 04/10/2024

OVERVIEW

Uplizna, a CD19-directed cytolytic antibody, is indicated for the treatment of **neuromyelitis optica spectrum disorder** (NMOSD) in adults who are anti-aquaporin-4 antibody-positive.¹

Disease Overview

NMOSD is a rare, relapsing, autoimmune disorder of the brain and spinal cord with optic neuritis and/or myelitis as predominant characteristic symptoms.² NMOSD often causes significant, permanent damage to vision and/or spinal cord function resulting in blindness or impaired mobility.³ Patients may experience pain, paralysis, loss of bowel and bladder control, loss of visual acuity, and uncontrolled motor functions. Complications can lead to death.

Recommendations

The Neuromyelitis Optica Study Group (NEMOS) published revised recommendations for the treatment of NMOSD in 2024.⁴ The standard of care for the treatment of NMOSD attacks (for both AQP4-IgG-positive and double-negative cases) are high-dose glucocorticoids and/or apheresis therapy. Long-term immunotherapy is recommended for patients with AQP4-IgG-positive NMOSD. NEMOS notes the first-choice therapies for the treatment of AQP4-IgG-positive NMOSD are Uplizna, Enspryng® (satralizumab-mwge subcutaneous injection), Soliris® (eculizumab intravenous infusion), Ultomiris® (ravulizumab-cwyz intravenous infusion), and rituximab. The order of preference for these therapies is unclear and further comparative trials and real-world data are needed. The choice of treatment is dependent on several factors, including disease activity and severity, mode and onset of action, possibility to combine it with immunosuppressive drugs, effect on autoimmune and other comorbidities, gender (family planning issues), frequency and route of administration, side effect profile as well as patient and physician preference. In general, if a patient fails a first-choice treatment, another first-choice treatment should be tried; other options include use of a second-choice treatment (azathioprine, mycophenolate mofetil, low-dose oral glucocorticoids) or the addition of a second-choice treatment to the regimen.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Uplizna. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Uplizna as well as the monitoring required for adverse events and long-term efficacy, approval requires Uplizna to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Uplizna is recommended in those who meet the following criteria:

FDA-Approved Indication

1. **Neuromyelitis Optica Spectrum Disorder.** Approve if the patient meets ONE of the following (A or B):
 - A) **Initial Therapy.** Approve for 1 year if the patient meets ALL of the following (i, ii, and iii):
 - i. Patient is ≥ 18 years of age; AND
 - ii. Diagnosis of neuromyelitis optica spectrum disorder was confirmed by blood serum test for anti-aquaporin-4 antibody positive disease; AND
 - iii. The medication is being prescribed by or in consultation with a neurologist.
 - B) **Patient is Currently Receiving Uplizna.** Approve for 1 year if the patient meets ALL of the following (i, ii, iii, and iv):
 - i. Patient is ≥ 18 years of age; AND
 - ii. Diagnosis of neuromyelitis optica spectrum disorder was confirmed by blood serum test for anti-aquaporin-4 antibody-positive disease; AND
 - iii. According to the prescriber, patient has had clinical benefit from the use of Uplizna; AND
Note: Examples of clinical benefit include reduction in relapse rate, reduction in symptoms (e.g., pain, fatigue, motor function), and a slowing progression in symptoms.
 - iv. The medication is being prescribed by or in consultation with a neurologist.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Uplizna is not recommended in the following situations:

1. **Concomitant Use With a Rituximab Product, Enspryng (satralizumab-mwge subcutaneous injection, Soliris (eculizumab intravenous infusion), or Ultomiris (ravulizumab-cwyz intravenous infusion).** There is no evidence to support additive efficacy of combining Uplizna with rituximab, Enspryng, Soliris, or Ultomiris Intravenous.
2. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Uplizna® intravenous infusion [prescribing information]. Deerfield, IL: Horizon Therapeutics; July 2021.
2. National Organization for Rare Disorders. Neuromyelitis Optica Spectrum Disorder. Last updated July 27, 2022. Available at: <https://rarediseases.org/rare-diseases/neuromyelitis-optica/>. Accessed on April 5, 2024.
3. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015;85(2):177-189.
4. Kämpfel T, Giglhuber K, Aktas O, et al. Update on the diagnosis and treatment of neuromyelitis optica spectrum disorders (NMOSD) – revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part II: Attack therapy and long-term management. *J Neurol*. 2024;271:141-176.

