Pharmacy Policy Bulletin: J-1012 Enspryng (satralizumab-mwge) – Commercial and Healthcare Reform	
Number: J-1012	**Category:** Prior Authorization
Line(s) of Business: ☒ Commercial ☒ Healthcare Reform ☐ Medicare	**Benefit(s):** **Commercial:** **Prior Authorization (1.):** **1.** Miscellaneous Specialty Drugs Injectable = Yes w/ Prior Authorization **Quantity Limits (1., 2., 3., or 4.):** **1.** Quantity Limits = Safety/Specialty **2.** Quantity Limits = Safety/Specialty + Dose Opt **3.** Quantity Limits = Safety/Specialty + Dose Opt + Watchful **4.** Rx Mgmt Performance = MRXC = Yes **Healthcare Reform:** Not Applicable
Region(s): ☒ All ☐ Delaware ☐ New York ☐ Pennsylvania ☐ West Virginia	**Additional Restriction(s):** None
Version: J-1012-006	**Original Date:** 10/07/2020
Effective Date: 10/28/2024	**Review Date:** 10/02/2024

Drugs Product(s):	• Enspryng (satralizumab-mwge)
FDA-Approved Indication(s):	• Treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive

Background:	• The precise mechanism by which Enspryng exerts therapeutic effects in NMOSD is unknown but is presumed to involve inhibition of interleukin (IL)-6-mediated signaling through binding to soluble and membrane-bound IL-6 receptors. • NMOSD is a rare autoimmune disease that causes multifocal central nervous system (CNS) inflammation primarily affecting the optic nerves and spinal cord. NMOSD attacks are associated with loss of vision and paralysis separated by periods of remission. It is characterized by severe immune-mediated demyelinations and axonal damage caused by the presence of AQP4-antibodies that attack the optic nerve, spinal cord, and brainstem. The AQP4-IgG autoantibody is detectable in more than 80% of patients with NMOSD and is pathogenic, initiating the inflammatory CNS lesions and clinical manifestations of the disease. In 2023, there was an update on NMOSD diagnosis and treatment. The update stated that a diagnosis of NMOSD has at least 1 core clinical characteristic including optic neuritis, acute myelitis, area postrema syndrome, acute brain stem syndrome, symptomatic narcolepsy or acute diencephalic

	clinical syndrome with NMOSD-typical diencephalic magnetic resonance imaging (MRI) lesions, and symptomatic cerebral syndrome with NMOSD-brain lesions. • Immunosuppressives are recommended as first-line therapy for management of NMOSD. Recommended first-line treatments include azathioprine, methotrexate, and mycophenolate mofetil. Switching to a different immunosuppressive is recommended as second-line therapy. Monoclonal antibody therapy is an alternative for those who experience therapeutic failure, contraindication, or intolerance to first-line immunosuppressives. • Prescribing Considerations: ○ Enspryng is contraindicated in patients with active hepatitis B infection and active or untreated latent tuberculosis. ○ Delay Enspryng administration in patients with an active infection until the infection is resolved. Vaccination with live or live-attenuated vaccines is not recommended during treatment. ○ Hepatitis B virus, tuberculosis, and liver transaminase screening is required before the first dose. ○ Use of multiple monoclonal antibodies (e.g., Soliris, rituximab, Uplizna) for the treatment of NMOSD has not been established. ○ The recommended loading dosage of Enspryng for the first three administrations is 120 mg by subcutaneous injection at Weeks 0, 2, and 4, followed by a maintenance dosage of 120 mg every 4 weeks.

Approval Criteria

I. Initial Authorization

When a benefit, coverage of Enspryng may be approved when all of the following criteria are met **(A. through G.)**:

A. The member is 18 years of age or older.

B. The member has a diagnosis of NMOSD. (ICD-10: G36.0)

C. The prescriber attests the member is anti-aquaporin-4 (AQP4) antibody positive.

D. The member exhibits one (1) of the following core clinical characteristics of NMOSD **(1. through 6)**:

1. Optic neuritis

2. Acute myelitis

3. Area postrema syndrome

4. Acute brainstem syndrome

5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions

6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions

E. Enspryng is prescribed by or in consultation with a neurologist or other healthcare provider experienced in treating NMOSD.

F. The prescriber submits documentation of baseline number of relapse(s), which occurred over the last year.

G. The member has experienced therapeutic failure or intolerance to one (1) immunosuppressant (e.g., mycophenolate mofetil, azathioprine, methotrexate), or all are contraindicated.

II. Reauthorization

When a benefit, reauthorization of Enspryng may be approved when the following criterion is met **(A.)**:

A. The prescriber attests the member has experienced a decrease from baseline in the number of NMOSD relapse(s).

III. Quantity Limitations

When prior authorization is approved, Enspryng may be authorized in quantities as follows:

Diagnosis	Induction Therapy	Maintenance Therapy
NMOSD	Three (3) prefilled syringes within the first four (4) weeks of therapy	One (1) prefilled syringe every four (4) weeks

Coding of quantity level limitations is at the maintenance therapy threshold. Claims for quantities of Enspryng prefilled syringes that exceed maintenance therapy limitations will reject at point of sale. Patient Level Authorization (PLA) will be needed for authorized quantities of pre-filled syringes that exceed maintenance therapy limitations (i.e., induction therapy).

IV. An exception to some or all of the criteria above may be granted for select members and/or circumstances based on state and/or federal regulations.

Limitations of Coverage

I. Enspryng is not used in combination with another monoclonal antibody (e.g., Soliris, Uplizna) used for the treatment of NMOSD.
II. Coverage of drug(s) addressed in this policy for disease states outside of the FDA-approved indications should be denied based on the lack of clinical data to support effectiveness and safety in other conditions unless otherwise noted in the approval criteria.
III. For Commercial or HCR members with a closed formulary, a non-formulary product will only be approved if the member meets the criteria for a formulary exception in addition to the criteria outlined within this policy.

Authorization Duration

Initial Authorization –

- Commercial and HCR Plans: If approved, up to a 6 month authorization may be granted.
  - Note: For induction therapy authorization duration, refer to the Quantity Limitations tables for the respective drug and diagnosis.

Reauthorization –

- Commercial and HCR Plans: If approved, up to a 12 month authorization may be granted.

Automatic Approval Criteria

None.

References:

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2. Neuromyelitis optica spectrum disorder - symptoms, causes, treatment: NORD. National Organization for Rare Disorders. Available at: https://rarediseases.org/rare-diseases/neuromyelitis-optica/. Accessed September 4, 2024.
3. Trebst C, Jarius S, Berthele A, et al. Update on the diagnosis and treatment of neuromyelitis optica: Recommendations of the Neuromyelitis Optica Study Group (NEMOS). *J Neurol*. 2014;261(1):1-16.
4. Mealy MA, Wingerchuk DM, Palace J, et al. Comparison for relapse and treatment failure rates among patients with neuromyelitis optica. *JAMA Neurol*. 2014;71(3):324-330.
5. Kitley J, Elsone L, George J, et al. Methotrexate is an alternative to azathioprine in neuromyelitis optica spectrum disorders with aquaporin-4 antibodies. *J Neurol Neurosurg Psychiatry*. 2013 Aug;84(8):918-21.

6. Wingerchuk DM, Lucchinetti CF. Neuromyelitis Optica Spectrum Disorder. *N Engl J Med*. 2022;387:631-639.
7. Diagnosing NMOSD. Available at: https://nmosd.com/hcp/diagnosing-nmosd. Accessed August 30, 2024.
8. Jarius S, Aktas O, Ayzenberg I, Bellmann-Strobl J, et al.; Neuromyelitis Optica Study Group (NEMOS). Update on the diagnosis and treatment of neuromyelits optica spectrum disorders (NMOSD) - revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part I: Diagnosis and differential diagnosis. J Neurol. 2023 Jul;270(7):3341-3368.