PRIOR AUTHORIZATION POLICY

POLICY: Enspryng Prior Authorization Policy
- Enspryng® (satralizumab-mwge subcutaneous injection – Genentech)

REVIEW DATE: 09/20/2023; selected revision 03/20/2024

INSTRUCTIONS FOR USE

THE FOLLOWING COVERAGE POLICY APPLIES TO HEALTH BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. CERTAIN CIGNA COMPANIES AND/OR LINES OF BUSINESS ONLY PROVIDE UTILIZATION REVIEW SERVICES TO CLIENTS AND DO NOT MAKE COVERAGE DETERMINATIONS. REFERENCES TO STANDARD BENEFIT PLAN LANGUAGE AND COVERAGE DETERMINATIONS DO NOT APPLY TO THOSE CLIENTS. COVERAGE POLICIES ARE INTENDED TO PROVIDE GUIDANCE IN INTERPRETING CERTAIN STANDARD BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. PLEASE NOTE, THE TERMS OF A CUSTOMER'S PARTICULAR BENEFIT PLAN DOCUMENT [GROUP SERVICE AGREEMENT, EVIDENCE OF COVERAGE, CERTIFICATE OF COVERAGE, SUMMARY PLAN DESCRIPTION (SPD) OR SIMILAR PLAN DOCUMENT] MAY DIFFER SIGNIFICANTLY FROM THE STANDARD BENEFIT PLANS UPON WHICH THESE COVERAGE POLICIES ARE BASED. FOR EXAMPLE, A CUSTOMER'S BENEFIT PLAN DOCUMENT MAY CONTAIN A SPECIFIC EXCLUSION RELATED TO A TOPIC ADDRESSED IN A COVERAGE POLICY. IN THE EVENT OF A CONFLICT, A CUSTOMER'S BENEFIT PLAN DOCUMENT ALWAYS SUPERSEDES THE INFORMATION IN THE COVERAGE POLICIES. IN THE ABSENCE OF A CONTROLLING FEDERAL OR STATE COVERAGE MANDATE, BENEFITS ARE ULTIMATELY DETERMINED BY THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT. COVERAGE DETERMINATIONS IN EACH SPECIFIC INSTANCE REQUIRE CONSIDERATION OF 1) THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT IN EFFECT ON THE DATE OF SERVICE; 2) ANY APPLICABLE LAWS/REGULATIONS; 3) ANY RELEVANT COLLATERAL SOURCE MATERIALS INCLUDING COVERAGE POLICIES AND; 4) THE SPECIFIC FACTS OF THE PARTICULAR SITUATION. EACH COVERAGE REQUEST SHOULD BE REVIEWED ON ITS OWN MERITS. MEDICAL DIRECTORS ARE EXPECTED TO EXERCISE CLINICAL JUDGMENT AND HAVE DISCRETION IN MAKING INDIVIDUAL COVERAGE DETERMINATIONS. COVERAGE POLICIES RELATE EXCLUSIVELY TO THE ADMINISTRATION OF HEALTH BENEFIT PLANS. COVERAGE POLICIES ARE NOT RECOMMENDATIONS FOR TREATMENT AND SHOULD NEVER BE USED AS TREATMENT GUIDELINES. IN CERTAIN MARKETS, DELEGATED VENDOR GUIDELINES MAY BE USED TO SUPPORT MEDICAL NECESSITY AND OTHER COVERAGE DETERMINATIONS.

CIGNA NATIONAL FORMULARY COVERAGE:

OVERVIEW

Enspryng, an interleukin-6 receptor antagonist, 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 Enspryng, Soliris® (eculizumab intravenous infusion), Ultomiris® (ravulizumab-cwyz intravenous infusion and subcutaneous injection) [awaiting FDA approval], Uplizna® (inebilizumab-cdon 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 Enspryng. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Enspryng as well as the monitoring required for adverse events and long-term efficacy, approval requires Enspryng to be prescribed by or in consultation with a physician who specializes in the condition being treated.

- **Enspryng® (satralizumab-mwge subcutaneous injection – Genentech)**

is(are) covered as medically necessary when the following criteria is(are) met for FDA-approved indication(s) or other uses with supportive evidence (if applicable):

FDA-Approved Indication

1. **Neuromyelitis Optica Spectrum Disorder**. Approve for the duration noted 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 was confirmed by a positive blood serum test for anti-aquaporin-4 antibody; AND
     - **iii.** The medication is being prescribed by or in consultation with a neurologist.
   - **B)** Patient is Currently Receiving Enspryng. 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 was confirmed by a positive blood serum test for anti-aquaporin-4 antibody; AND
     - **iii.** According to the prescriber, patient has had clinical benefit from the use of Enspryng; AND

Note: Examples of clinical benefit include reduction in relapse rate, reduction in symptoms (e.g., pain, fatigue, motor function), and a slowing in progression of symptoms.

iv. The medication is being prescribed by or in consultation with a neurologist.

CONDITIONS NOT COVERED

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is(are) considered experimental, investigational, or unproven for ANY other use(s) including the following (this list may not be all inclusive; criteria will be updated as new published data are available):

1. **Concomitant Use with a Rituximab Product, Soliris (eculizumab intravenous infusion), or Uplizna (inebilizumab-cdon intravenous infusion).** There is no evidence to support concomitant use of Enspryng with a rituximab product, Soliris or Uplizna.

REFERENCES

1. Enspryng® subcutaneous injection [prescribing information]. South San Francisco, CA: Genentech; March 2022.
2. National Organization for Rare Disorders. Neuromyelitis Optica Spectrum Disorder. Updated July 2022. Available at: https://rarediseases.org/rare-diseases/neuromyelitis-optica/. Accessed September 18, 2023.
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. Kűmpfel 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.

HISTORY

Type of Revision	Summary of Changes	Review Date
Annual Revision	No criteria change.	08/31/2022
Annual Revision	No criteria change.	09/20/2023
Selected Revision	**Neuromyelitis Optica Spectrum Disorder – Initial Therapy:** Removed criterion that required prior use of two systemic therapies and criterion that patient has had a history of at least one relapse in the last 12 months or two relapses in the last 2 years. Enspryng is listed as a first-line treatment option in the Neuromyelitis Optica Study Group (NEMOS) recommendations for the treatment of Neuromyelitis Optica Spectrum Disorder (2024).	03/20/2024