Effective Date **7/1/2024**
Coverage Policy Number **IP0062**

Inebilizumab

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Related Coverage Resources

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. 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.

Overview

This policy supports medical necessity review for inebilizumab-cdon (Uplizna®).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Inebilizumab (Uplizna) is considered medically necessary when the following are met:

Neuromyelitis Optica Spectrum Disorder (NMOSD). Individual meets ALL of the following:
A. Age 18 years of age or older
B. Diagnosis of neuromyelitis optica spectrum disorder (NMOSD) was confirmed by blood serum test for anti-aquaporin-4 antibody positive
C. Medication is being prescribed by, or in consultation with, a neurologist.

Dosing. ONE of the following:

A. 300 mg by intravenous infusion once every 2 weeks for 2 doses
B. 300 mg by intravenous infusion once every 6 months

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Continuation of inebilizumab (Uplizna™) is considered medically necessary for Neuromyelitis Optica Spectrum Disorder (NMOSD) when the above medical necessity criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval authorization is 12 months.
Reauthorization approval duration is 12 months.

Conditions Not Covered

Any other use is considered experimental, investigational or unproven, including the following (this list may not be all inclusive):

Concomitant use with a rituximab product, Soliris® (eculizumab injection), or Enspryng™ (satralizumab-mwge injection). There is no evidence to support additive efficacy of combining Uplizna with rituximab, Soliris, or Enspryng.

Coding Information

Note: 1) This list of codes may not be all-inclusive.
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

HCPCS Codes	Description
J1823	Injection, inebilizumab-cdon, 1 mg

Background

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 central nervous system inflammatory disorder that can lead to significant morbidity and mortality.[2,3] The predominant symptoms are inflammation of the optic nerve (optic neuritis) and inflammation of the spinal cord (myelitis). Optic neuritis may lead to pain inside the eye and can progress to blindness. Myelitis tends to affect some, and often all, motor, sensory, and autonomic functions (bladder and bowel). Affected patients may experience pain in the spine or limbs, mild to severe paralysis of the lower limbs, and loss of bowel and bladder control. For acute attacks, typical treatment is high-dose intravenous

corticosteroids.[2] Plasma exchange may be effective in patients who suffer acute severe attacks and who do not respond to intravenous corticosteroids. For long-term control of the disease (relapse prevention), a variety of immunosuppressive drugs are utilized as first-line therapy; most widely prescribed are corticosteroids, azathioprine, mycophenolate mofetil, and rituximab. Interleukin-6 signaling blocking agents (e.g., Enspryng® [satralizumab-mwge subcutaneous {SC} injection], Actemra [tocilizumab injection for intravenous {IV} or SC use]), Soliris® (eculizumab IV infusion), and IV immunoglobulins are also used for relapse prevention.[3] Note that of the listed agents, only Enspryng and Soliris are FDA-approved for NMOSD.[4,5]

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 July 7, 2023.
3. Chan KH, Lee CY. Treatment of neuromyelitis optica spectrum disorders. Int J Mol Sci. 2021;22(16):8638.
4. Enspryng® subcutaneous injection [prescribing information]. South San Francisco, CA: Genentech; March 2022.
5. Soliris® intravenous infusion [prescribing information]. Boston, MA: Alexion; November 2020.
6. 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