Drug Coverage Policy

Effective Date 5/15/2024
Coverage Policy Number.....IP0129
Title..... Tepezza

Ophthalmology – Tepezza

- Tepezza™ (teprotumumab intravenous infusion – Horizon)

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 Healthcare Coverage Policy

Tepezza, an insulin-like growth factor-1 receptor (IGF-1R) antagonist, is indicated for the treatment of **thyroid eye disease**, regardless of thyroid eye disease activity or duration.[1]

Dosing Information
The recommended dose is 10 mg/kg administered by intravenous (IV) infusion for the initial dose, followed by 20 mg/kg administered IV once every 3 weeks for seven additional doses.[1]

Disease Overview
Thyroid eye disease, a rare, serious, debilitating and painful autoimmune disease, is also known as thyroid-associated ophthalmopathy, Graves' ophthalmopathy, or Graves' orbitopathy.[2] Thyroid eye

disease is most commonly related to Graves' disease; however, it can also develop in patients with other thyroid diseases (e.g., Hashimoto's thyroiditis). The prevalence is higher in females than males (16 per 100,000 vs. 3 per 100,000, respectively).[3] Risk factors include female gender, middle age, and smoking.[2]

Most patients with thyroid eye disease develop eye disease while being treated for hyperthyroidism under the care of an endocrinologist.[4] Thyroid eye disease is characterized by endomysial interstitial edema, expansion, and proliferation of cells within the fibrofatty compartment, resulting in clinical manifestations of periorbital edema, lid retraction, proptosis, diplopia, corneal breakdown and in rare cases, optic nerve compression. This disease is associated with major comorbidities that can lead to blindness.

Consensus Statement
The American Thyroid Association and the European Thyroid Association issued a consensus statement in 2022 for the management of thyroid eye disease.[4] The Task Force notes "active" thyroid eye disease as disease with a clinical activity score (CAS) of ≥ 3 or if the patient has history or documentation of progression of thyroid eye disease based on subjective or objective worsening of vision, soft tissue inflammation, motility, or proptosis. CAS assesses seven items (spontaneous retrobulbar pain, pain on attempted up or lateral gaze, redness of the eyelids, redness of the conjunctiva, swelling of the eyelids, inflammation of the caruncle and/or plica, and conjunctival edema); each item is given one point if present. The severity of disease is divided into three groups: mild (features of disease have a minor impact on daily life insufficient to justify treatment), moderate (patient does not have sight-threatening disease but disease has sufficient impact on daily life to justify the risks of medical or surgical intervention), or sight-threatening (patient with dysthyroid optic neuropathy and/or corneal breakdown and/or globe subluxation). Pharmacologic treatment includes oral or IV glucocorticoids; mycophenolate, rituximab, Tepezza, and Actemra (tocilizumab IV infusion). Tepezza is noted as a preferred treatment with the following goals: disease inactivation and diplopia; reduction of proptosis; and improvement of eye motility. It is an acceptable treatment for disease inactivation and reduction of soft tissue involvement.

Medical Necessity Criteria

Tepezza is considered medically necessary when the following are met:

FDA-Approved Indication

1. **Thyroid Eye Disease.** Approve for 6 months if the patient meets the following (A, B, C, and D):
   Note: Thyroid Eye Disease is also recognized as Graves' ophthalmopathy, Graves' orbitopathy, thyroid-associated ophthalmopathy, and thyroid orbitopathy.
   A) Patient is ≥ 18 years of age; AND
   B) Patient has been assessed as having at least moderate severity level of disease based on signs and symptoms, according to the prescriber; AND
   Note: Examples of signs and symptoms of disease of at least moderate severity include the following: lid retraction ≥ 2 mm, moderate or severe soft tissue involvement, proptosis ≥ 3 mm above normal for race and sex, and diplopia (Gorman score 2 to 3).
   C) Patient has not received 8 doses (total) of Tepezza; AND
   Note: The maximum recommended treatment is for 8 doses. For a patient who has started therapy but has not completed 8 doses, approve the number of doses required for the patient to receive a total of 8 doses.
   D) The medication is prescribed by or in consultation with an ophthalmologist, endocrinologist, or a physician who specializes in thyroid eye disease.

Dosing: Up to 20 mg/kg per dose administered by intravenous infusion no more frequently than every 3 weeks for 8 doses

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.

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

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven (criteria will be updated as new published data are available).

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
J3241	Injection, teprotumumab-trbw, 10 mg

References

1. Tepezza intravenous infusion [prescribing information]. Lake Forest, IL: Horizon; July 2023.
2. Horizon Therapeutics. Teprotumumab for injection. Briefing document for the Food and Drug Administration Dermatologic and Ophthalmic Drugs Advisory Committee. Meeting Date: December 13, 2019. Available at: https://www.fda.gov/advisory-committees/advisory-committee-calendar/updated-public-participation-information-december-13-2019-meeting-dermatologic-and-ophthalmic-drugs#event-information. Accessed on January 18, 2024.
3. Bartley GB, Fatourechi V, Kadrmas EF, et al. Clinical features of Graves' ophthalmopathy in an incidence cohort. *Am J Ophthalmol.* 1996;121(3):284-290.
4. Burch HB, Perros P, Bednarczuk T, et al. Management of thyroid eye disease: a consensus statement by the American Thyroid Association and the European Thyroid Association. *Thyroid*. 2022;32(12):1439-1470. doi: 10.1089/thy.2022.0251. Epub 2022 Dec

Revision Details

Type of Revision	Summary of Changes	Date
Annual Review	**Thyroid Eye Disease**: Removed if the individual is a smoker, smoking cessation has been discussed	5/1/2024

	Thyroid Eye Disease: The criterion that the patient has active disease of at least moderate severity based on signs and symptoms, according to the prescriber was changed to remove the word "active". The new criterion requires that the patient has at least moderate severity level of disease based on signs and symptoms, according to the prescriber. The Note was also revised to read: Examples of signs and symptoms of disease of at least moderate severity include the following: lid retraction ≥ 2 mm, moderate or severe soft tissue involvement, proptosis ≥ 3 mm above normal for race and sex, and diplopia (Gorman score 2 to 3).	

The policy effective date is in force until updated or retired.