**POLICY:** Ophthalmology – Tepezza™ (teprotumumab injection for intravenous use – Horizon Therapeutics)

**EFFECTIVE DATE:** 8/1/2020
**LAST REVISION DATE:** 09/16/2024

**COVERAGE CRITERIA FOR:** All UCare Plans

## OVERVIEW

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 $\geq 3$ 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:

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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.

**POLICY STATEMENT**

Prior Authorization is recommended for medical benefit coverage of Tepezza. Approval is recommended for those who meet the **Criteria** and **Dosing** for the listed indication. Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). All approvals are provided for the duration noted below. In cases where the approval is authorized in months, 1 month is equal to 30 days. Because of the specialized skills required for evaluation and diagnosis of patients treated with Tepezza as well as the monitoring required for adverse events and long-term efficacy, approval requires Tepezza to be prescribed by or in consultation with a physician who specializes in the condition being treated.

**Automation:** None.

**RECOMMENDED AUTHORIZATION CRITERIA**

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

**FDA-Approved Indication**

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**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 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.

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**D)** The medication is prescribed by or in consultation with an ophthalmologist, endocrinologist, or a physician who specializes in thyroid eye disease.

**Dosing.** Approve up to 20 mg/kg per dose administered by intravenous infusion no more frequently than every 3 weeks for 8 doses.

## CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Tepezza is not recommended in the following situations:

**1.** 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. 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 8.

## HISTORY

| Type of Revision | Summary of Changes | Review Date |
|---|---|---|
| Annual Revision | No criteria changes. | 01/18/2023 |
| Update | 05/24/2023: Tepezza prescribing information was revised in April 2023. FDA-approved indication was revised from "Treatment of thyroid eye disease" to "Treatment of thyroid eye disease, regardless of thyroid eye disease or duration". Criteria were not changed. | -- |

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| Annual Revision | **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 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). | 02/07/2024 |
| UCare P&T Review | Policy reviewed and approved by UCare P&T committee. Annual review process | 09/16/2024 |

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