VENTURA COUNTY
HEALTH CARE PLAN

PRIOR AUTHORIZATION POLICY

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

DATE REVIEWED: 01/29/2020

OVERVIEW

Tepezza, an insulin-like growth factor-1 receptor (IGF-1R) antagonist antibody, is indicated for the treatment of patients with thyroid eye disease (TED).[1] The safety and efficacy have not been established in patients who are pregnant or pediatric patients. Tepezza is a fully human immunoglobulin G monoclonal antibody that binds IGF-1R, a tyrosine kinase cell surface receptor that is overexpressed in the orbital fibroblasts of TED patients.[2,3] Tepezza targets and binds to IGF-1R, inhibits IGF-1R autophosphorylation, decreases cell surface expression of IGF-1R and prevents downstream signaling. Based on the mechanism of action, Tepezza is theorized to decrease inflammation and tissue growth, thus reducing the signs and symptoms of TED. The recommended dose is 10 mg/kg administered by intravenous (IV) infusion for the initial dose, followed by 20 mg/kg by IV infusion administered once every 3 weeks for seven additional doses.

Disease Overview

TED is a progressive, vision-threatening autoimmune inflammatory disease of the eye and orbital tissues with predominant features of fibrosis and adipogenesis.[4] It is also recognized in literature as Graves' ophthalmopathy, Graves' orbitopathy, thyroid-associated ophthalmopathy, and thyroid orbitopathy. TED is most commonly related with Graves' disease, it can also develop in patients with other thyroid diseases (e.g., Hashimoto's thyroiditis) and has a higher prevalence in women than men (16 per 100,000 vs. 3 per 100,000, respectively).[5] Orbital fibroblasts appear responsible for soft tissue enlargement by expressing potential pathogenic autoantigens, such as thyrotropin receptor and IGF-1R.[6] Activation of orbital fibroblasts leads to increased hyaluronic acid production, proinflammatory cytokine synthesis, and enhanced differentiation into either myofibroblasts or adipocytes. These processes result in inflammation, enlargement of extraocular muscles and expansion of orbital tissue and fat, which in turn cause forward displacement of the eye, resulting in proptosis and inflammation.[4] The degree of severity can be staged as mild, moderate-to-severe, or sight-threatening, following quantitative assessment of lid aperture width, proptosis measurement, diplopia score, degrees of abduction in eye muscle movement, examination of the cornea for evidence of exposure keratitis or ulceration, and assessment of optic nerve function.

POLICY STATEMENT

Prior authorization is recommended for prescription benefit coverage of Tepezza. All approvals are provided for the duration noted below. 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 Indications

1. **Thyroid Eye Disease.** (Note: Thyroid Eye Disease is also recognized as Graves' ophthalmopathy, Graves' orbitopathy, thyroid-associated ophthalmopathy, and thyroid orbitopathy.) Approve for 6 months if the patient meets the following criteria (A, B, and C):
   - **A)** The patient is ≥ 18 years of age; AND
   - **B)** According to the prescriber, the patient has been assessed as having active disease of at least moderate severity based on signs and symptoms (e.g., the degree of inflammation, degree of proptosis, presentation of diplopia, etc.); AND
   - **C)** Teppeza is prescribed by or in consultation with an ophthalmologist, endocrinologist, or a physician who specializes in thyroid eye disease.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Tepezza not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. Rationale for non-coverage for these specific conditions is provided below. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval.)

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 injection [prescribing information]. Lake Forest, IL: Horizon Therapeutics; January 2020.
2. Wang Y, Smith TJ. Current concepts in the molecular pathogenesis of thyroid-associated ophthalmopathy. IOVS. 2014;55(3):1735-1748.
3. Douglas RS. Teprotumumab, an insulin-like growth factor-1 receptor antagonist antibody, in the treatment of active thyroid eye disease: a focus on proptosis. Eye. 2019;33(2):183-90.
4. Horizon. Teprotumumab for injection. Breifing 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 7, 2020.
5. Bartley GB, Fatourechi V, Kadrmas EF, Jacobsen SJ, Ilstrup DM, Garrity JA, Gorman CA. Clinical features of Graves' ophthalmopathy in an incidence cohort. Am J Ophthalmol 1996;121(3):284-290.
6. Shan S, Douglas R. The pathophysiology of thyroid eye disease. J Neuroophthalmol. 2014 Jun;34(2):177-85.