Washington State Health Care Authority

Endocrine and Metabolic Agents: Teprotumumab (Tepezza)

Medical policy no. 30.19.20.AA-1 Effective Date: 8/1/2024

Note: *New-to-market drugs included in this class based on the Apple Health Preferred Drug List are non-preferred and subject to this prior authorization (PA) criteria. Non-preferred agents in this class require an inadequate response or documented intolerance due to severe adverse reaction or contraindication to at least TWO preferred agents. If there is only one preferred agent in the class documentation of inadequate response to ONE preferred agent is needed. If a drug within this policy receives a new indication approved by the Food and Drug Administration (FDA), medical necessity for the new indication will be determined on a case-by-case basis following FDA labeling.*

To see the list of the current Apple Health Preferred Drug List (AHPDL), please visit: https://www.hca.wa.gov/assets/billers-and-providers/apple-health-preferred-drug-list.xlsx

Medical necessity

Drug	Medical Necessity
teprotumumab (Tepezza)	**teprotumumab (Tepezza)** may be considered medically necessary in patients who meet the criteria described in the clinical policy below. If all criteria are not met, the clinical reviewer may determine there is a medically necessary need and approve on a case-by-case basis. The clinical reviewer may choose to use the reauthorization criteria when a patient has been previously established on therapy and is new to Apple Health. **NOTE:** The approved dose will be rounded down to the nearest 500 mg vial if the dose reduction is 10% or less of the calculated dose based on body weight.

Clinical policy:

Clinical Criteria	
Thyroid eye disease (TED) teprotumumab (Tepezza)	Teprotumumab (Tepezza) may be approved when all the following documented criteria are met: 1. Patient is 18 years of age or older, **AND** 2. Prescribed by, or in consultation with, a specialist in ophthalmology or endocrinology; **AND** 3. Patient's thyroid levels are being controlled prior to starting therapy with Tepezza, defined as: a. Thyroxine [T4] and triiodothyronine [T3] are within normal limits per laboratory specifications; **OR**

	b. T4 and T3 levels are within 50% of normal limits per laboratory specifications; **AND** 4. Patient has a diagnosis of Thyroid eye disease (TED) that is related to Graves' Disease (i.e. Graves' orbitopathy); **AND** 5. Patient has a TED clinical activity score (CAS) of 4 or greater in at least one eye; **AND** 6. Patient meets ONE of the following criteria: a. Presence of diplopia**; OR** b. Provider attests there is significant proptosis defined as: i. Proptosis is 3mm or greater than the upper level of normal; **OR** ii. Proptosis significantly affects daily life (e.g. difficulty blinking, dry eyes, decreased vision); **OR** c. Patient has had an inadequate response, intolerance, or contraindication to intravenous glucocorticoids (IVGC); i. An inadequate response is defined as still meeting criteria 5 after at least a 6-week trial of IVGC at recommended dosing ii. Recommended dosing with IVGC consists of intravenous methylprednisolone (IVMP) at cumulative doses of 4.5g over 3 months (0.5 g weekly × 6 weeks followed by 0.25 g weekly for an additional 6 weeks). If ALL criteria are met, the request will be authorized for a maximum of 8 doses that must be used within **12 month**s after approval.
	Criteria (Reauthorization) Teprotumumab (Tepezza) cannot be renewed even if not all doses are administered within the initial 12-month approval period. Teprotumumab may only be authorized once per lifetime.

Dosage and quantity limits

Drug	Indication	FDA Approved Dosing	Dosage Form and Quantity Limit
Teprotumumab (Tepezza)	Thyroid eye disease	Initiate dosing with 10 mg/kg for the first infusion, followed by 20 mg/kg every 3 weeks for 7 additional infusions (8 infusions total)	500 mg single-dose vial for injection: 3 vials for the initial dose followed by 5 vials for each of 7 additional doses (115 billable units initially followed by 230 billable units every 3 weeks thereafter for a total of 8 doses) **NOTE:** The approved dose will be rounded down to the nearest 500 mg vial if the dose reduction is 10% or less of the calculated dose based on body weight.

Coding:

HCPCS Code	Description
J3241	Injection, teprotumumab-trbw, 10 mg: 1 billable unit = 10 mg

Background:

Thyroid eye disease (also called Graves' orbitopathy or ophthalmopathy) is an autoimmune disease of the orbit and retro-ocular tissues occurring most commonly in patients with Graves' disease. Patients with thyroid eye disease may have no ocular symptoms or could present with the following common symptoms: gritty or foreign object sensation in the eyes, excessive tearing, eye discomfort or pain, blurry vision, and diplopia.[1] In severe cases, loss of vision can occur. Upon examination, the characteristic signs of thyroid eye disease are proptosis, conjunctival inflammation, and periorbital edema. For patients with moderate-to-severe thyroid eye disease, the initial treatment for many years has been glucocorticoids.[1] Their use is supported by a long history of clinical experience with a known risk-to-benefit ratio. With the entrance of teprotumumab (Tepezza), there may be increased interest in use as an initial therapy in select patients (marked proptosis and diplopia); however, there are no head-to-head comparisons establishing the superiority of teprotumumab (Tepezza) compared to glucocorticoids. Further, the evidence supporting the durability of response and relapse rates following treatment with teprotumumab (Tepezza) are still immature.

Teprotumumab (Tepezza) was evaluated in two double-blind, randomized, placebo-controlled clinical trials totaling 177 members. Each participant had a diagnosis of thyroid eye disease (TED), a Clinical Activity Score (CAS) of 4 or greater and was either euthyroid or had thyroid levels within 50% of normal.[2,3] At 24 weeks 71% (Study 1) and 83% (Study 2) of participants in the teprotumumab group achieved a greater than 2mm reduction in proptosis compared to 20% and 10%, respectively in the placebo group. Over half of participants in the treatment group maintained proptosis response at 51 weeks. Additionally, in study 2, 59% of those taking temprotumumab achieved a CAS of 0 or 1 at week 24 compared to 21% of the placebo group. The most common adverse reactions included muscle spasms, alopecia, nausea, and fatigue.[2,3]

References

1. Bartalena L, Kahaly GJ, Baldeschi L, Dayan CM, Eckstein A, Marcocci C, Marinò M, Vaidya B, Wiersinga WM; EUGOGO. The 2021 European Group on Graves' orbitopathy (EUGOGO) clinical practice guidelines for the medical management of Graves' orbitopathy. Eur J Endocrinol. 2021 Aug 27;185(4):G43-G67. doi: 10.1530/EJE-21-0479. PMID: 34297684.
2. Smith TJ, Kahaly GJ, Ezra DG, et al. Teprotumumab for Thyroid-Associated Ophthalmopathy. N Engl J Med 2017; 376:1748-1761. DOI: 10.1056/NEJMoa1614949
3. Douglas RS, Kahaly GJ, Patel A, et al. Teprotumumab for the treatment of active thyroid eye disease. N Engl J Med. 2020;382(4):341-352.
4. Tepezza [Prescribing Information]. Dublin, Ireland: Horizon Therapeutics Ireland DAC. December 2022
5. 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. Eur Thyroid J. 2022;11(6):e220189.

History

Approved Date	Effective Date	Version	Action and Summary of Changes
02/28/2024	08/01/2024	30.19.20.AA-1	New policy created. Approved by DUR Board