Jefferson Health Plans

DR.009.C TEPEZZA® (teprotumumab-trbw)

Original Implementation Date : 11/1/2020
Version [C] Date : 8/21/2024
Last Reviewed Date: 8/21/2024

PRODUCT VARIATIONS

This policy applies to all Jefferson Health Plans product lines unless noted below.

POLICY STATEMENT

TEPEZZA® is considered Medically Necessary for the treatment of Thyroid Eye Disease when the criteria listed in this policy are met.

FDA APPROVED INDICATIONS

TEPEZZA® is an insulin-like growth factor-1 receptor inhibitor indicated for the treatment of Thyroid Eye Disease regardless of Thyroid Eye Disease activity or duration.

OFF-LABEL USE

Authorization for off-labeled use of medication will be evaluated on an individual basis. Review of an off-labeled request by the Medical Staff will be predicated on the appropriateness of treatment and full consideration of medical necessity.

For off-label use, Medical Directors will review scientific literature and local practice patterns.

PRIOR AUTHORIZATION CRITERIA

INITIAL CRITERIA

*AUTHORIZATION DURATION: IF **ALL CRITERIA MET**, APPROVE FOR 6 MONTHS (MAX 8 TOTAL INFUSIONS)*

1. Adults 18 years of age and older; AND
2. Patient has moderate to severe Thyroid Eye Disease confirmed by at least ONE of the following:

- Lid retraction of greater than or equal to 2 mm.
- Moderate or severe soft-tissue involvement.
- Proptosis of greater than or equal to 3 mm above the normal values for race and sex.
- Periodic or constant diplopia.

3. Patient does not have poorly controlled diabetes; AND
4. Medication is being prescribed by or in consultation with a specialist (ophthalmology).

RENEWAL CRITERIA

Authorization Duration: Coverage cannot be renewed.

DOSAGE AND ADMINISTRATION

DOSING RECOMMENDATIONS:

- Initiate dosing with 10 mg/kg for first infusion, followed by 20 mg/kg every 3 weeks for 7 additional infusions.
- Administer the diluted solution intravenously for over 90 minutes for the first two infusions. If well tolerated, the minimum time for subsequent infusions can be reduced to 60 minutes. If not well tolerated, the minimum time for subsequent infusions should remain at 90 minutes.

RISK FACTORS/SIDE EFFECTS

- **Exacerbation of Preexisting Inflammatory Bowel Disease:** TEPEZZA® may cause an exacerbation of preexisting inflammatory bowel disease (IBD). Monitor patients with IBD for flare of disease. If IBD exacerbation is suspected, consider discontinuation of TEPEZZA®.
- **Hyperglycemia:** Hyperglycemia or increased blood glucose may occur in patients treated with TEPEZZA®. In clinical trials, 10% of patients (two thirds of whom had pre-existing diabetes or impaired glucose tolerance) experienced hyperglycemia. Hyperglycemic events should be controlled with medications for glycemic control, if necessary. Monitor patients for elevated blood glucose and symptoms of hyperglycemia while on treatment with TEPEZZA®. Patients with pre-existing diabetes should be under appropriate glycemic control before and while receiving TEPEZZA®.

- **Hearing Impairment Including Hearing Loss:** TEPEZZA® may cause severe hearing impairment including hearing loss, which in some cases may be permanent. Assess patients' hearing before, during, and after treatment with TEPEZZA® and consider the benefit-risk of treatment with patients.

- **Infusion Reactions:** TEPEZZA® may cause infusion reactions. Infusion reactions have been reported in approximately 4% of patients treated with TEPEZZA®. Signs and symptoms of infusion-related reactions include transient increases in blood pressure, feeling hot, tachycardia, dyspnea, headache, and muscular pain. Infusion reactions may occur during any of the infusions or within 1.5 hours after an infusion. Reported infusion reactions are usually mild or moderate in severity and can usually be successfully managed with corticosteroids and antihistamines. In patients who experience an infusion reaction, consideration should be given to pre-medicating with an antihistamine, antipyretic, corticosteroid and/or administering all subsequent infusions at a slower infusion rate.

MONITORING

- Monitor patients with preexisting IBD for flare of disease; discontinue TEPEZZA® if IBD worsens.
- Monitor glucose levels in all patients; treat hyperglycemia with glycemic control medications.
- TEPEZZA® (teprotumumab) is contraindicated during pregnancy.

CLINICAL EVIDENCE

Teprotumumab (an insulin-like growth factor 1 [IGF-1] receptor inhibitor) was approved for the treatment of Graves' orbitopathy by the US Food and Drug Administration (FDA) in 2020, based on the findings from two 24-week trials comparing teprotumumab with placebo in 171 patients with active, moderate-to-severe orbitopathy. In each trial, a greater proportion of patients in the teprotumumab group had a reduction in clinical activity score and degree of proptosis (69 versus 20 percent with placebo and 78 versus 7 percent with placebo, respectively). The durability of efficacy requires confirmation with long-term follow-up studies. Eye symptoms in the patients in the trial had to have begun within nine months of trial entry, and it is unclear whether the drug would be as effective in patients whose disease was of longer duration. In addition, there was no comparison with the effectiveness of glucocorticoids, the standard therapy for patients with moderate-to-severe orbitopathy.

CODING

Note*:* The Current Procedural Terminology (CPT®), Healthcare Common Procedure Coding System (HCPCS), and the 10th revision of the International Statistical Classification of Diseases and Related Health Problems (ICD-10) codes that *may* be listed in this policy are for reference purposes only. Listing a code in this policy does not imply that the service is covered and is not a guarantee of

payment. Other policies and coverage guidelines may apply. When reporting services, providers/facilities should code to the highest level of specificity using the code that was in effect on the date the service was rendered. This list may not be all inclusive.

CPT® is a registered trademark of the American Medical Association.

CPT Code	Description
N/A	

HCPCS Code	Description
J3241	Injection, teprotumumab-trbw, 10 mg
S9338	Home infusion therapy, immunotherapy, administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem

ICD 10 Codes	Description
E05.00	Thyrotoxicosis with diffuse goiter without thyrotoxic crisis or storm
E05.01	Thyrotoxicosis with diffuse goiter with thyrotoxic crisis or storm
H05.89	Other disorders of orbit

DISCLAIMER

Approval or denial of payment does not constitute medical advice and is neither intended to guide nor influence medical decision making.

Policy Bulletins are developed by Jefferson Health Plans to assist in administering plan benefits and constitute neither offers of coverage nor medical advice.

This Policy Bulletin may be updated and therefore is subject to change.

For HealthChoices (Medicaid) and Children's Health Insurance Program (CHIP) products: Any requests for services that do not meet criteria set in PARP will be evaluated on a case-by-case basis.

POLICY HISTORY

This section provides a high-level summary of changes to the policy since the previous version.

Summary	Version	Version Date
2024 annual review. Risk factors/Side effects section was updated.	C	8/21/2024
2023 Annual policy review. Reference section was updated. FDA Approved Indications updated	B	9/1/2023
2022 Annual policy review. Reference section was updated.	A	11/1/2020
2021 Annual policy review. Code J3241 was added to the coding. Codes C9061, J3490, J3590 were removed.	A	11/1/2020
New Drug Policy	A	11/1/2020

REFERENCES

1. Tepezza® [package insert]. Dublin, Ireland; Horizon Therapeutics Ireland, DAC, January 2020. Updated April 2023. Accessed June 2023

2. Smith TJ, Kahaly GJ, Ezra DG, Fleming JC, Dailey RA, Tang RA, Harris GJ, Antonelli A, Salvi M, Goldberg RA, Gigantelli JW, Couch SM, Shriver EM, Hayek BR, Hink EM, Woodward RM, Gabriel K, Magni G, Douglas RS. Teprotumumab for Thyroid-Associated Ophthalmopathy. N Engl J Med. 2017 May 4;376(18):1748-1761. doi: 10.1056/NEJMoa1614949.

3. Douglas RS, Kahaly GJ, Patel A, Sile S, Thompson EHZ, Perdok R, Fleming JC, Fowler BT, Marcocci C, Marinò M, Antonelli A, Dailey R, Harris GJ, Eckstein A, Schiffman J, Tang R, Nelson C, Salvi M, Wester S, Sherman JW, Vescio T, Holt RJ, Smith TJ. Teprotumumab for the Treatment of Active Thyroid Eye Disease. N Engl J Med. 2020 Jan 23;382(4):341-352. doi: 10.1056/NEJMoa1910434.

4. Ross DS, Burch HB, Cooper DS, et al. 2016 American Thyroid Association Guidelines for Diagnosis and Management of Hyperthyroidism and Other Causes of Thyrotoxicosis. Thyroid. 2016;26(10):1343.

5. Terry F. Davies, Henry B. Barch , UpToDate , last update Jan.03,2023 Clinical features and diagnosis of Graves' orbithopthy (ophthalmopathy).

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