Medical Policy

Healthcare Services Department

Policy Name	Policy Number	Scope
Teprotumumab-trbw (Tepezza)	MP-RX-FP-89-23	☒ MMM MA ☒ MMM Multihealth

Service Category

☐ Anesthesia
☐ Surgery
☐ Radiology Procedures
☐ Pathology and Laboratory Procedures
☐ Medicine Services and Procedures
☐ Evaluation and Management Services
☐ DME/Prosthetics or Supplies
☒ Part B DRUG

Service Description

This document addresses the use of ***Teprotumumab (Tepezza)*** , a drug approved by the Food and Drug Administration (FDA) for the treatment of Thyroid Eye Disease (TED), otherwise known as Graves' Orbitopathy or Graves' Ophthalmopathy.

Background Information

Thyroid Eye Disease is a rare vision-threatening autoimmune disease. It is associated with dry or irritated eyes, outward bulging of eyes (proptosis), double vision (diplopia), and optic nerve compression. TED is often associated with Graves' disease, the most common cause of hyperthyroidism and develops in roughly 40% of patients with Graves' disease. Therefore, classic findings would include orbitopathy in the setting of current or past Graves' hyperthyroidism (low TSH, high free thyroxine [T4] and/or triiodothyronine [T3]). However, hyperthyroidism is not directly linked to TED; and about 10% of TED patients have a normally functioning thyroid. This "euthyroid" Graves' disease is still characterized by high serum thyroid autoantibody concentrations, which contribute to the development of TED. The natural history of the disease is variable and may include a period of rapid deterioration followed by stabilization, or individuals may experience exacerbations and remissions. Most patients have self-limiting mild forms of the disease where lifestyle modifications (smoking cessation, local therapies such as artificial tears, and elevating the head of the bed to decrease swelling) are warranted.

The 2016 American Thyroid Association Guidelines for Diagnosis and Management of Hyperthyroidism and Other Causes of Thyrotoxicosis recommend that euthyroidism be achieved and maintained in hyperthyroid patients with TED or risk factors for the development of orbitopathy. Surgery and antithyroid medications are the preferred treatments for Graves' Disease; no recommendation is provided for the treatment of TED itself. The 2021 European Group on Graves' Orbitopathy (EUGOGO) Guidelines for the Management of Graves' Orbitopathy recommends high-dose intravenous glucocorticoids be considered as first-line therapy for moderate-to-severe and active GO. Second-line treatment options include a subsequent course of intravenous glucocorticoids, oral corticosteroids combined with either cyclosporine or azathioprine, orbital radiotherapy combined with oral or intravenous glucocorticoids, teprotumumab (Tepezza), rituximab, or tocilizumab. Surgical options for TED include orbital decompression and muscle surgery to correct diplopia. Tepezza has not been directly compared to corticosteroid therapy in the treatment of TED.

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Approved Indications

A. Treatment of Thyroid Eye Disease regardless of Thyroid Eye Disease activity or duration.

Other Uses

A. N/A

Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

HCPCS	Description
J3241	Injection, teprotumumab-trbw, 10 mg (Effective 10/1/2020

ICD-10	Description
E05.00	Thyrotoxicosis with diffuse goiter without thyrotoxic crisis or storm

Medical Necessity Guidelines

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

Teprotumumab-trbw (Tepezza)

A. Criteria For Initial Approval

i. Individual has a diagnosis of Thyroid Eye Disease; AND
ii. Documentation is provided that individual has symptomatic moderate to severe disease, as defined by one or more of the following:
   A. Lid retraction ≥ 2 mm; OR
   B. Moderate or severe soft tissue involvement; OR
   C. Proptosis ≥ 3 mm above normal for race and gender; OR
   D. Intermittent or constant diplopia; AND
iii. Documentation is provided that individual has a clinical activity score (CAS) greater than or equal to 4 in the more severely affected eye; AND
iv. Documentation is provided that one of the following applies:
   A. Thyroid function tests are provided and are within normal limits as defined by laboratory standard (i.e. individual is euthyroid); OR
   B. Thyroid function tests show free thyroxine (T4) and free triiodothyronine (T3) levels less than 50% above or below normal limits as defined by laboratory standard

B. Authorization Duration

i. One course of treatment; defined as a total of 8 intravenous infusions of Tepezza (teprotumumab) administered every 3 week

C. **Conditions Not Covered**

Any other use is considered experimental, investigational, or unproven, including the following (this list may not be all inclusive):

i. More than one course* of treatment; OR
ii. Individual has had prior orbital irradiation or eye surgery for TED; OR
iii. Individual has decreased best-corrected visual acuity due to optic neuropathy as defined by decrease in vision of 2 lines on the Snellen chart, new visual field defect, or color defect; OR
iv. Individual has unresponsive corneal decompensation; OR
v. When the above criteria are not met and for all other indications

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Limits or Restrictions

A. Quantity Limitations

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines. The chart below includes dosing recommendations as per the FDA-approved prescribing information.

Drug	Limit
Tepezza (teprotumumab-trbw) 500 mg vial	Initial dose: One 10 mg/kg infusion
Tepezza (teprotumumab-trbw) 500 mg vial	Subsequent doses: 20mg/kg every 3 weeks for seven infusions
Exceptions	
N/A	

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Reference Information

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2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: April 7, 2022.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically.
5. 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; 26:1343-1421. Erratum in: Thyroid. 2017 Nov;27(11):1462
6. Smith TJ, Kahaly GJ, Ezra DG, et al. Teprotumumab for Thyroid-Associated Ophthalmopathy. N Engl J Med. 2017; 376: 1748- 1761.
7. Douglas RS, Kahaly GJ, Patel A, et al. Teprotumumab for the Treatment of Active Thyroid Eye Disease. N Engl J Med. 2020; 382: 341-352.
8. Douglas RS, Kahaly GJ, Ugradar S, et al. Teprotumumab Efficacy, Safety, and Durability in Longer-Duration Thyroid Eye Disease and Re-treatment: OPTIC-X Study. Ophthalmology. 2022;129(4):438-449. doi:10.1016/j.ophtha.2021.10.017
9. Bartalena L, Kahaly G, Baldeschi L, et al. The 2021 European Group on Graves' orbitopathy (EUGOGO) clinical practice guidelines for the medical management of Graves' orbitopathy. European Journal of Endocrinology. 2021; 185(4), G43-G67

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

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Policy History

Revision Type	Summary of Changes	P&T Approval Date	MPCC Approval Date
Policy Inception	Elevance Health's Medical Policy adoption.	N/A	11/30/2023

Revised: 9/27/23