Medicare Advantage Medical Benefit Drug Policy

BlueCross BlueShield Nebraska

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Effective Date: 04/01/2023

Tepezza™ (teprotumumab-trbw)

HCPCS: J3241

Policy:

Requests must be supported by submission of chart notes and patient specific documentation.

A. Coverage of the requested drug is provided when all the following are met:
   a. FDA approved indication
   b. FDA approved age
   c. Prescribed by or in consultation with an endocrinologist or ophthalmologist
   d. Clinical activity score (CAS) greater than or equal to 4 in the most severely affected eye
      OR
   e. Moderate to severe active thyroid eye disease defined as one or more of the following:
      i. Decrease in color vision
      ii. Moderate or severe soft tissue involvement
      iii. Exophthalmos greater than or equal to 3 mm above normal for race and gender
      iv. Inconstant or constant diplopia
   f. Must have treated thyroid disease defined as:
      i. Euthyroid function with free triiodothyronine (T3) and thyroxine (T4) within the normal limits for the range of the laboratory
         OR
      ii. Thyroid function is normalizing with both T3 and T4 levels being less than 50% above or 50% below the normal limits for the range of the laboratory
   g. Treatment with an adequate course of oral or intravenous (IV) corticosteroids (for example 30 mg/day prednisone for 4 weeks) has been ineffective, not tolerated, or is contraindicated
   h. Physician attestation a discussion has been had with the member to stop smoking if they are a current smoker
   i. Trial and failure, contraindication, OR intolerance to the preferred drugs as listed in the BCBSNE MA Part B drugs prior authorization list

B. Quantity Limitations, Authorization Period and Renewal Criteria
   a. Quantity Limits: Align with FDA recommended dosing
   b. Authorization Period: 6 months
   c. Renewal Criteria: Not applicable as no further authorization will be provided.

***Note: Coverage may differ for Medicare Part B members based on any applicable criteria outlined in Local Coverage Determinations (LCD) or National Coverage Determinations (NCD) as determined by Center for Medicare and Medicaid Services (CMS). See the CMS website at http://www.cms.hhs.gov/. Determination of coverage of Part B drugs is based on medically accepted indications which have supported citations included or approved for inclusion determined by CMS approved compendia.

Background Information

- Thyroid eye disease is a complex orbital inflammatory disease, which can be sight threatening, debilitating, and disfiguring. It is caused by antibodies directed against receptors in the thyroid cells and also on the surface of the cells behind the eyes. Muscles and fatty tissues behind the eye become inflamed leading to proptosis (eye bulging), strabismus (misalignment of the eyes), and diplopia (double vision) and in some cases can lead to blindness. Risk factors for the disease include female gender, middle age, and smoking. Active TED lasts for up to three years with the disease only responding to pharmacotherapy while it is active and inflammation is ongoing.

- Tepezza is an insulin-like growth factor-1 receptor antagonist indicated for the treatment of thyroid eye disease (TED).

- Goals of treatment in thyroid disease consists of achieving a euthyroid state and symptom management. The majority of patients with thyroid eye disease have mild to moderate disease and require primarily supportive care with ocular lubrication, topical cyclosporine, and lifestyle modification, such as, smoking cessation, sodium restriction, and sunglasses.

- The current mainstay of treatment for moderate to severe thyroid eye disease is oral or intravenous corticosteroids. Treatment can be initiated at doses of oral prednisone 30 mg daily for four weeks or methylprednisolone 500 mg once weekly for weeks 1 to 6, then 250 mg once weekly for weeks 7 to 12 with cumulative dose 4.5 to 5 g over 12 weeks. The European Thyroid Association recommends initial treatment with IV glucocorticoids for moderate-to-severe, active orbitopathy, citing several studies that suggest it is more efficacious and associated with fewer side effects than oral therapy. Similar trials have not been performed in the United States, where initiation with oral glucocorticoids remains the most common first-line treatment. Treatment for thyroid eye disease should start in the early months of the active inflammatory phase, as treatment becomes less effective as the disease progresses.

- There are no clinical head to head studies that compare steroids and Tepezza for the treatment of moderate to severe TED. The 2021 European Group on Graves' orbitopathy (EUGOGO) clinical practice guidelines for the medical management of Graves' orbitopathy state Tepezza's incorporation into routine clinical practice is currently limited by the lack of comprehensive long-term efficacy and safety data and the absence of head-to-head comparison trials with intravenous glucocorticoids.

- Safety and efficacy were evaluated in the OPTIC trials, two multicenter, randomized, double-masked, placebo-controlled trials of 171 patients with thyroid eye disease. Patients were required to have active, moderate to severe thyroid eye disease with significant symptoms, including 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, and periodic or constant diplopia. Patients were also required to have a clinical activity score (CAS) greater than or equal to 4 and symptoms less than 9 months from the onset of thyroid eye disease. All patients were euthyroid or with mild hypothyroidism or hyperthyroidism, defined as free thyroxine (T4) and free triiodothyronine (T3) levels less than 50% above or below the normal limits for the testing laboratory. Patients with previous orbital irradiation or surgery for thyroid eye disease were not allowed. The primary endpoint in the first trial was a composite endpoint of reduction of greater than or equal to 2 points in the CAS and a reduction of greater than or equal to 2 mm in proptosis. The primary endpoint in the second trial was a reduction in proptosis of greater than or equal to 2 mm. In both trials, significantly more patients treated with teprotumumab demonstrated less symptoms of thyroid eye disease than patients treated with placebo.

References:

1. Tepezza [prescribing information]. Lake Forest, IL: Horizon Therapeutics USA, Inc.; October 2021.
2. 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 - 1421.
3. Bartalena L, Baldeschi L, Boboridis K, et al. The 2016 european thyroid association/european group on graves' orbitopathy guidelines for the management of graves' orbitopathy. Eur Thyroid J. 2016 Mar; 5 (1): 9 - 26.
4. Smith TJ, Kahaly GJ, Ezra DG, et al. Teprotomumab for thyroid-associated opthalmopathy. NEJM. 2017 May 4; 376 (18): 1748 – 61.
5. 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: 183- 90.
6. Douglas RS, Kahaly GJ, Patel A, et al. Teprotumumab for the treatment of active thyroid eye disease. NEJM. 2020 Jan 23; 382: 341 – 52.
7. McAlinden C. An overview of thyroid eye disease. Eye and Vision. 2014; 1: 9 – 12.
8. Bartalena L, Kahaly GJ, Baldeschi L, at al. 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.

Policy History		
#	Date	Change Description
1.0	Effective Date: 04/01/2023	New policy

** The prescribing information for a drug is subject to change. To ensure you are reading the most current information it is advised that you reference the most updated prescribing information by visiting the drug or manufacturer website or http://dailymed.nlm.nih.gov/dailymed/index.cfm.*