Tepezza® (teprotumumab-trbw)
(Intravenous)

Document Number: IC-0525

Last Review Date: 01/04/2022
Date of Origin: 02/04/2020
Dates Reviewed: 02/2020, 10/2020, 01/2021, 02/2021, 01/2022

I. Length of Authorization

Coverage will be provided for 6 months (max total of 8 infusions) and may NOT be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Tepezza 500 mg single-dose vial for injection: 3 vials for initial dose followed by 5 vials for each of 7 additional doses

B. Max Units (per dose and over time) [HCPCS Unit]:

- 115 billable units initially followed by 230 billable units every 3 weeks thereafter for a total of 8 doses

III. Initial Approval Criteria [1-8,10]

Coverage is provided in the following conditions:

- Patient is at least 18 years of age; **AND**

Universal Criteria

- Must be prescribed by, or in consultation with, a specialist in ophthalmology, endocrinology, oculoplastic surgery or neuro-ophthalmology; **AND**
- Patient has not had a decrease in best corrected visual acuity (BVCA) due to optic neuropathy within the previous six months (i.e., decrease in vision of 2 lines on the Snellen chart, new visual field defect, or color defect secondary to optic nerve involvement); **AND**
- Patient is euthyroid [Note: mild hypo- or hyperthyroidism is permitted which is defined as free thyroxine (FT4) and free triiodothyronine (FT3) levels less than 50% above or below the normal limits (every effort should be made to correct the mild hypo- or hyperthyroidism promptly)]; **AND**
- Patient does not have corneal decompensation that is unresponsive to medical management; **AND**
- Patient does not have uncontrolled diabetes; **AND**
- Used as single agent therapy; **AND**

Thyroid Eye Disease (TED) † Φ

- Patient has a clinical diagnosis of TED that is related to Graves' Disease (i.e., Graves' orbitopathy); **AND**
- Patient has a baseline clinical activity score (CAS) of at least 4 §; **AND**
- Patient has active phase TED that is non-sight threatening but has a significant impact on daily living (e.g., lid retraction ≥ 2 mm, moderate or severe soft tissue involvement, exophthalmos ≥ 3 mm above normal for race and gender, and/or inconstant or constant diplopia); **AND**
- Patient must have active disease (this may include, but is not limited to, the following: onset of TED symptoms within the previous 9 months); **AND**
- Patient had an inadequate response, or there is a contraindication or intolerance, to high-dose intravenous glucocorticoids

§ Assessment of Thyroid Eye Disease (TED): Clinical Activity Score (CAS) Elements [11]
– Spontaneous retrobulbar pain – Pain on attempted upward or downward gaze – Redness of eyelids – Redness of conjunctiva – Swelling of caruncle or plica – Swelling of eyelids – Swelling of conjunctiva (chemosis) – Increase in exophthalmos of ≥2 mm* – Decrease in eye motility of ≥8°* – Decrease in visual acuity in the last 1–3 months*
*Note: Each element is assigned a score of one. Elements denoted with a * can be used when a previous assessment is available. A seven-point scale is used when prior assessment is not available.*

† FDA Approved Indication(s); ‡ Compendium Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria

Coverage cannot be renewed.

V. Dosage/Administration

Indication	Dose
Thyroid Eye Disease	Administer 10 mg/kg intravenously initially, then 20 mg/kg intravenously every three weeks for 7 additional infusions (8 infusions total). Administer the diluted solution intravenously 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.

VI. Billing Code/Availability Information

HCPCS code:

- J3241 – Injection, teprotumumab-trbw, 10 mg: 1 billable unit = 10 mg

NDC:

- Tepezza 500 mg single-dose vial for injection: 75987-0130-xx

VII. References

1. Tepezza [package insert]. Dublin, Ireland; Horizon Therapeutics Ireland, DAC; October 2021. Accessed December 2021.
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, Sile S, Thompson EHZ, et al. Teprotumumab Treatment Effect on Proptosis in Patients With Active Thyroid Eye Disease; Results From a Phase 3, Randomized, Double-Masked, Placebo-Controlled, Parallel-Group, Multicenter Study. Amer Assoc of Clin Endo. Los Angeles: Endocrine Practice; 2019.
4. Patel A, Yang H, Douglas RS. Perspective: A New Era in the Treatment of Thyroid Eye Disease. Am J Ophthalmol 2019;208:281–288.
5. Ross DS, Burch HB, Cooper DS, et al. 2016 . 2016;26(10):1343.
6. Mourits MP, Koornneef L, Wiersinga WM, et al. Clinical criteria for the assessment of disease activity in Graves' ophthalmopathy: a novel approach. Br J Ophthalmol. 1989 Aug; 73(8): 639–644.
7. Mourits MP, Prummel MF, Wiersinga WM, et al. Clinical activity score as a guide in the management of patients with Graves' ophthalmopathy. Clin Endocrinol (Oxf). 1997 Jul;47(1):9-14.
8. 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.
9. Ye X, Bo X, Hu X, et al. Efficacy and safety of mycophenolate mofetil in patients with active moderate-to-severe Graves' orbitopathy. Clin Endocrinol (Oxf). 2017;86(2):247.
10. Zang S, Ponto KA, Kahaly GJ. Intravenous Glucocorticoids for Graves' Orbitopathy: Efficacy and Morbidity. J Clin Endocrinol Metab. 2011 Feb;96(2):320-32.
11. 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. 27 Aug 2021. https://doi.org/10.1530/EJE-21-0479

Appendix 1 – Covered Diagnosis Codes

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

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Articles (LCAs) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC