Policy and Procedure	
PHARMACY PRIOR AUTHORIZATION AND STEP THERAPY POLICY AND CRITERIA ORPTCEND080.0425	**ENDOCRINE AND METABOLIC DRUGS** **TEPEZZA®** (teprotumumab-trbw vial)
Effective Date: 6/1/2025	**Review/Revised Date**: 08/23, 03/24, 06/24, 02/25 (KN)
Original Effective Date: 06/23	**P&T Committee Meeting Date**: 04/23, 08/23, 04/24, 08/24, 04/25
Approved by: Oregon Region Pharmacy and Therapeutics Committee	

SCOPE:
Providence Health Plan and Providence Health Assurance as applicable (referred to individually as "Company" and collectively as "Companies").

APPLIES TO:
Medicare Part B

POLICY CRITERIA:

COVERED USES:
All Food and Drug Administration (FDA) approved indications not otherwise excluded from the benefit.

REQUIRED MEDICAL INFORMATION:
1. For initiation of therapy (new starts), must meet all the following criteria:
    a. Confirmed diagnosis of moderate-to-severe thyroid eye disease/Grave's Orbitopathy, as defined as eye disease that significantly impacts quality of life and at least one of the following:
        i. Lid retraction of at least 2 mm
        ii. Moderate or severe soft-tissue involvement (such as swelling or redness of the eyes)
        iii. Proptosis of at least 3 mm above normal for race or sex
        iv. Inconstant diplopia (diplopia at extremes of gaze) or constant diplopia (continuous diplopia in primary or reading position)
    b. Laboratory evidence of euthyroid state or T4 (thyroxine) and free T3 (triiodothyronine) levels less than 50% above or below normal limits
    c. For patients without diplopia or significant proptosis: Inadequate response to at least two weeks of therapy with high-dose intravenous (IV) glucocorticoid therapy (equivalent to methylprednisolone 0.5 g once weekly) unless there is a contraindication or intolerance
    d. Dosing is within the Food and Drug Administration approved label dose

2. For patients established on the requested therapy (within the previous year): Documentation that the member has not received 8 doses of Tepezza®

EXCLUSION CRITERIA: N/A

AGE RESTRICTIONS:
Age must be appropriate based on FDA-approved indication

PRESCRIBER RESTRICTIONS:
Must be prescribed by, or in consultation with, an ophthalmologist

COVERAGE DURATION:
Authorization will be approved for six months for a total of up to eight infusions

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Requests for indications that were approved by the FDA within the previous six (6) months may not have been reviewed by the health plan for safety and effectiveness and inclusion on this policy document. These requests will be reviewed using the New Drug and or Indication Awaiting P&T Review; Prior Authorization Request ORPTCOPS047.

Requests for a non-FDA approved (off-label) indication requires the proposed indication be listed in either the American Hospital Formulary System (AHFS), Drugdex, or the National Comprehensive Cancer Network (NCCN) and is considered subject to evaluation of the prescriber's medical rationale, formulary alternatives, the available published evidence-based research and whether the proposed use is determined to be experimental/investigational.

Coverage decisions are made on the basis of individualized determinations of medical necessity and the experimental or investigational character of the treatment in the individual case.

INTRODUCTION:
Teprotumumab-trbw is the first and only FDA approved drug for the treatment of thyroid eye disease. Teprotumumab-trbw is a monoclonal antibody of the insulin-like growth factor type-1 receptor.

FDA APPROVED INDICATIONS:
Thyroid Eye Disease regardless of Thyroid Eye Disease activity or duration

POSITION STATEMENT:
Thyroid eye disease (TED), or Grave's orbitopathy (GO), is one of the main extra thyroidal manifestations of Graves' disease (been noted to develop in approximately 40% of patients). TED/GO causes inflammation and tissue expansion behind the eye which leads to proptosis (abnormal protrusion) and is often accompanied by diplopia (double vision) and pain. Severe cases of TED can cause blindness.

There is a lack of treatment options for thyroid eye disease currently available, other than glucocorticoids. Teprotumumab represents the first in class for treatment of thyroid eye disease. This agent is a monoclonal antibody that blocks the activation and signaling of insulin-like growth factor-1, which is thought to play a role in thyroid eye disease via immunoglobulin signaling.

This drug carries warnings for:
1. Exacerbation of preexisting inflammatory bowel disease (IBD) – consider discontinuation if IBD exacerbation suspected, and
2. Hyperglycemia - 10% of patients (two thirds of whom had pre-existing diabetes or impaired glucose tolerance) experienced hyperglycemia

The 2021 European Group on Graves' orbitopathy (EUGOGO) clinical practice guidelines recommend that high-dose systemic glucocorticoids (GCs) be used as first-line treatment in combination with mycophenolate for moderate to severe disease.

The American Thyroid Association and European Thyroid Association joined forces to develop a consensus statement (2022). Intravenous glucocorticoid (IVGC) therapy is a preferred treatment for active moderate-to-severe thyroid eye disease when disease activity is the prominent feature in the absence of either significant proptosis or diplopia. Teprotumumab is a preferred therapy, if available, in patients with active moderate-to-severe thyroid eye disease with significant proptosis and/or diplopia. The consensus statement defines moderate to severe disease as disease that significantly impacts the activities of daily life without threat to the patient's sight. These patients typically have at least one of the following: lid retraction of at least 2 mm, moderate or severe soft tissue involvement, proptosis of at least 3 mm above normal for race and sex, or diplopia (inconstant or constant).

Treatment with IVGC is recommended for sight-threatening disease. While there are reports showing efficacy of teprotumumab in sight-threatening disease, additional clinical trials are currently lacking.

REFERENCE/RESOURCES:

1. Tepezza Package insert. Lake Forest, IL. Horizon Therapeutics USA, Inc. December 2022.
2. Teprotumumab-trbw In: DRUGDEX® System [Internet database]. Greenwood Village, Colo: Thomson Reuters (Healthcare) Inc. Updated periodically. Accessed February 3, 2025.
3. Teprotumumab-trbw In: Lexi-Drugs Online [Internet database]. Hudson, OH: Lexi-Comp, Inc. Updated periodically. Accessed February 3, 2025.

4. Bartalena L, Kahaly GJ, Baldeschi L, et al. EUGOGO 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.
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. *Thyroid*. Dec 2022.1439-1470.

BILLING GUIDELINES AND CODING:

DRUG CODE*	
J3241	Injection, teprotumumab-trbw, 10 mg
RELATED ADMINISTRATION CODES*	
96365	Ther/proph/diag iv inf init
96366	Ther/proph/diag iv inf addon
96413	Chemo iv infusion 1 hr
96415	Chemo iv infusion addl hr

*Coding Notes:
• The above code list is provided as a courtesy and may not be all-inclusive. Inclusion or omission of a code from this policy neither implies nor guarantees reimbursement or coverage. Some codes may not require routine review for medical necessity, but they are subject to provider contracts, as well as member benefits, eligibility and potential utilization audit.
• HCPCS/CPT code(s) may be subject to National Correct Coding Initiative (NCCI) procedure-to-procedure (PTP) bundling edits and daily maximum edits known as "medically unlikely edits" (MUEs) published by the Centers for Medicare and Medicaid Services (CMS). This policy does not take precedence over NCCI edits or MUEs. Please refer to the CMS website for coding guidelines and applicable code combinations.