Drug and Biologic Coverage Policy

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Tetrabenazine

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INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This policy supports medical necessity review for tetrabenazine (Xenazine®).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Tetrabenazine (Xenazine) is considered medically necessary when the following are met:

1. **Chorea Associated with Huntington's Disease.** Individual meets **ALL** of the following criteria (A, B, C and D):
   A. Individual is 18 years of age or older
   B. Diagnosis of Huntington's disease is confirmed by genetic testing (for example, an expanded HTT CAG repeat sequence of at least 36)
   C. Medication is being prescribed by, or in consultation with, a neurologist
   D. Individual meets the preferred covered alternative(s) criteria as indicated in the table below

2. **Hyperkinetic Dystonia.** Individual meets **ALL** of the following criteria (A, B, C and D):
   A. Individual is 18 years of age or older
   B. Documented diagnosis of hyperkinetic dystonia
   C. Medication is being prescribed by, or in consultation with, a neurologist
   D. Individual meets the preferred covered alternative(s) criteria as indicated in the table below

3. **Tardive Dyskinesia.** Individual meets **ALL** of the following criteria (A, B, C, D and E):
   A. Individual is 18 years of age or older
   B. Documented diagnosis of tardive dyskinesia
   C. Individual has a history of treatment with a dopamine receptor blocking agent (for example, antipsychotics, metoclopramide, prochlorperazine)
   D. Medication is being prescribed by, or in consultation with, a neurologist or psychiatrist
   E. Individual meets the preferred covered alternative(s) criteria as indicated in the table below

4. **Tourette Syndrome and Related Tic Disorders.** Individual meets **ALL** of the following criteria (A, B, C and D):
   A. Individual is 18 years of age or older
   B. Documented diagnosis of Tourette Syndrome or related Tic Disorder
   C. Medication is being prescribed by, or in consultation with, a neurologist or psychiatrist
   D. Individual meets the preferred covered alternative(s) criteria as indicated in the table below

Coverage varies across plans and requires the use of preferred products. Refer to the customer's benefit plan document for coverage details.

Employer Group Non-Covered Products and the Preferred Covered Alternatives:
Individual and Family Plan Non-Covered Products and Covered Alternative(s

Non-Covered Product	Criteria
Xenazine (tetrabenazine)	The individual has tried **tetrabenazine** (the bioequivalent generic product) [prior authorization is required] AND cannot take due to a formulation difference in the inactive ingredient(s) which would result in a significant allergy or serious adverse reaction.

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Tetrabenazine (Xenazine) is considered medically necessary for continued use when initial criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial and reauthorization approval duration: up to 12 months

Conditions Not Covered

Tetrabenazine (Xenazine) is considered experimental, investigational or unproven for ANY other use.

Background

OVERVIEW

Tetrabenazine, a vesicular monoamine transporter type 2 inhibitor, is indicated for the treatment of **chorea associated with Huntington's disease** in adults.[1]

Clinical Efficacy

There are several published studies which have assessed the efficacy and safety of tetrabenazine for the treatment of other hyperkinetic movement disorders (e.g., tics in Tourette syndrome and tardive dyskinesia).[2-4] While most of the data for treatment of Tourette syndrome indicate that antipsychotic medications, both typical and atypical, are most effective, other medications (including tetrabenazine) may be used first to avoid the potential side effects of dopamine blockade.[5]

Guidelines

The American Academy of Neurology (AAN) evidence-based guidelines on pharmacologic treatment of chorea in Huntington's disease (2012) state that if chorea in Huntington's disease requires treatment, clinicians should prescribe tetrabenazine, amantadine, or Rilutek® (riluzole tablets) [Level B].[6]

The AAN published an evidence-based guideline for the treatment of tardive syndromes (2013).[7] The authors found that tetrabenazine possibly reduces tardive syndrome symptoms (based on two consistent Class III studies). Therefore, tetrabenazine may be considered in treating tardive syndromes (Level C).

The AAN published practice guideline recommendations for the treatment of tics in patients with Tourette syndrome and chronic tic disorders (2019).[8] The guidelines state that the dopamine depleters, tetrabenazine, deutetrabenazine, and valbenazine, are lacking published, randomized, controlled trials in the treatment of tics but note that these drugs are increasingly used off-label. When appropriately dosed, these drugs are generally well-tolerated but may be associated with drowsiness, depression, and parkinsonism.

References

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8. Pringsheim T, Okun MS, Müller-Vahl K, et al. Practice guideline recommendations summary: treatment of tics in people with Tourette syndrome and chronic tic disorders. *Neurology*. 2019;92:896-906.

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