Medical Drug Clinical Criteria

Subject:	Lamzede (velmanase alfa-tycv)		
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Table of Contents

Overview

This document addresses the use of Lamzede (velmanase alfa-tycv), an enzyme replacement therapy approved by the Food and Drug Administration (FDA) for the treatment of non-central nervous system manifestations of alpha-mannosidosis in adult and pediatric individuals.

Alpha-mannosidosis is a rare genetic lysosomal storage disorder caused by a deficiency of the enzyme alpha-mannosidase due to pathogenic variants in the *MAN2B1* gene. The enzyme deficiency leads to progressive accumulation of oligosaccharides with characteristic disease features including intellectual disability, ataxia, coarse face, hearing loss, skeletal dysplasia and immunodeficiency. Accurate diagnosis is important to differentiate alpha-mannosidosis from other lysosomal storage disorders and provide appropriate enzyme replacement therapy. Diagnosis is confirmed through enzymatic activity measurement or genetic testing.

Lamzede has a black box warning for severe hypersensitivity reactions including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Lamzede administration. If a severe hypersensitivity reaction occurs, Lamzede should be discontinued immediately, and appropriate medical treatment should be initiated. In individuals with severe hypersensitivity reaction, a desensitization procedure to Lamzede may be considered.

Clinical Criteria

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.

Lamzede (velmanase alfa-tycv)

Initial requests for Lamzede (velmanase alfa-tycv) may be approved if the following criteria are met:

I. Individual has a diagnosis of alpha-mannosidosis; **AND**
II. Documentation is provided that diagnosis is demonstrated by one of the following (Malm 2019):
    A. Deficiency in alpha-mannosidase enzyme activity as measured in fibroblasts or leukocytes; **OR**
    B. *MAN2B1* gene mutation; **AND**
III. Individual is using for the treatment of non-central nervous system disease manifestations.

Continuation requests for Lamzede (velmanase alfa-tycv) may be approved if the following criterion is met:

I. There is clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to improvement in motor function, improvement in pulmonary function, reduction in serum oligosaccharides) compared to the predicted natural history trajectory of disease.

Lamzede (velmanase alfa-tycv) may not be approved when the above criteria are not met and for all other indications.

Quantity Limits

Lamzede (velmanase alfa-tycv) Quantity Limit

Drug	Limit
Lamzede (velmanase alfa-tycv) 10 mg vial	1 mg/kg once per week

Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

HCPCS	
J3490	Unclassified drugs (when specified as [Lamzede] (velmanase alfa-tycv))
C9399	Unclassified drugs or biologicals
ICD-10 Diagnosis	
	All diagnoses pend

Document History

New: 02/24/2023
Document History:

- 02/24/2023 – Annual Review: New clinical criteria and quantity limit for Lamzede. Coding Review: Added HCPCS J3490, C9399. All diagnoses pend.

References

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2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
4. Malm D, Nilssen Ø. Alpha-Mannosidosis. 2001 Oct 11 [updated 2019 Jul 18]. In: Adam MP, Everman DB, Mirzaa GM, Pagon RA, Wallace SE, Bean LJH, Gripp KW, Amemiya A, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2023. PMID: 20301570. Available at: https://www.ncbi.nlm.nih.gov/books/NBK1396/ Accessed on: February 18, 2023.

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.