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Section:	Prescription Drugs	**Effective Date:**	April 1, 2025
Subsection:	Endocrine and Metabolic Drugs	**Original Policy Date:**	March 10, 2023
Subject:	Lamzede	**Page:**	1 of 4

Last Review Date: March 7, 2025

Lamzede

Description

Lamzede (velmanase alfa-tycv)

Background
Alpha-mannosidosis is a lysosomal storage disease that results from reduced activity of the enzyme alpha-mannosidase, caused by gene variants in Mannosidase Alpha Class 2B Member 1. Alpha-mannosidase catalyzes the degradation of accumulated mannose-containing oligosaccharides. The deficiency of alpha-mannosidase causes an intra-lysosomal accumulation of mannose-rich oligosaccharides in various tissues. Lamzede provides an exogenous source of alpha-mannosidase. Lamzede is internalized via binding to the mannose-6-phosphate receptor on the cell surface and transported into lysosomes where it is thought to exert enzyme activity (1).

Regulatory Status
FDA-approved indication: Lamzede is indicated for the treatment of non-central nervous system manifestations of alpha-mannosidosis in adult and pediatric patients (1).

Lamzede has a boxed warning regarding the risk of hypersensitivity reactions including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Lamzede administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue Lamzede immediately and initiate appropriate medical treatment. In patients with severe hypersensitivity reaction, a desensitization procedure to Lamzede may be considered (1).

Prior to Lamzede administration, consider pre-treating with antihistamines, antipyretics, and/or corticosteroids (1).

Lamzede may cause embryo-fetal harm when administered to a pregnant female. For females of reproductive potential, verify that the patient is not pregnant prior to initiating treatment with Lamzede. Advise female of reproductive potential to use effective contraception during treatment with Lamzede and for 14 days after the last dose if Lamzede is discontinued (1).

The safety and effectiveness of Lamzede has been established in pediatric patients (1).

Related policies

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Lamzede may be considered **medically necessary** if the conditions indicated below are met.

Lamzede may be considered **investigational** for all other indications.

Prior-Approval Requirements

Diagnosis

Patient must have the following:

Alpha-mannosidosis

AND ALL of the following:

1. Alpha-mannosidase? activity below 11% of normal
2. Prescriber agrees to monitor patient for infusion-related reactions, such as anaphylaxis, and provide appropriate medical treatment if necessary
3. Females of reproductive potential **only:** patient will be advised to use effective contraception during treatment with Lamzede and for 14 days after the last dose

Prior–Approval *Renewal* Requirements

Diagnosis

Patient must have the following:

Alpha-mannosidosis

AND ALL of the following:

1. Prescriber agrees to monitor patient for infusion-related reactions, such as anaphylaxis, and provide appropriate medical treatment if necessary
2. Females of reproductive potential **only:** patient will be advised to use effective contraception during treatment with Lamzede and for 14 days after the last dose

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 2 years

Prior–Approval *Renewal* Limits

Same as above

Rationale

Summary

Alpha-mannosidosis is a lysosomal storage disease that results from reduced activity of the enzyme alpha-mannosidase. The deficiency of alpha-mannosidase causes an intra-lysosomal accumulation of mannose-rich oligosaccharides in various tissues. Lamzede provides an exogenous source of alpha-mannosidase. Lamzede has a boxed warning regarding severe hypersensitivity reactions and prescribers should be prepared to manage severe reactions, including anaphylaxis (1).

Prior approval is required to ensure the safe, clinically appropriate, and cost-effective use of Lamzede while maintaining optimal therapeutic outcomes.

References

1. Lamzede [package insert]. Cary, NC: Chiesi USA, Inc.; February 2023.

Policy History

Date	Action
March 2023	Addition to PA
June 2023	Annual review
June 2024	Annual review
March 2025	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 7, 2025 and is effective on April 1, 2025.