Lamzede® (velmanase alfa-tycv)
(Intravenous)

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I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

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

- Lamzede 10 mg as a lyophilized powder in a single-dose vial: 11 vials per 7 days

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

- 110 billable units (110 mg) every 7 days

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Patient is at least 3 years of age; **AND**
- Documented baseline serum oligosaccharides; **AND**
- Documented baseline age-appropriate values for one or more of the following have been obtained: 6-minute walk test (6-MWT), 3-minute stair climb test (3-MSCT), pulmonary function tests (e.g., forced vital capacity), motor function [i.e., Bruininks-Oseretsky Test of Motor Proficiency (BOT-2)], etc.; **AND**
- Females of reproductive potential have a confirmed negative pregnancy test prior to initiating treatment; **AND**

***NOTE:** For very young patients in which FVC or 6-MWT are not suitable for measuring, requests will be reviewed on a case-by case basis.*

Universal Criteria [1]

- Therapy used to treat non-central nervous system manifestations of alpha mannosidosis (i.e., skeletal abnormalities, myopathy, motor function disturbances, immunodeficiency, etc.); **AND**

Alpha Mannosidosis † Φ [1-3]

- Patient has a definitive diagnosis of alpha mannosidosis as confirmed by ONE of the following:
  - Identification of deficient acid alpha-mannosidase enzyme activity in peripheral blood leukocytes or other nucleated cells such as fibroblasts of <11% of normal activity; **OR**
  - Identification of biallelic pathogenic variants in *MAN2B1* by molecular genetic testing

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

IV. Renewal Criteria [1-3]

Coverage may be renewed based on the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: hypersensitivity reactions including anaphylaxis, infusion-associated reactions, etc.; **AND**
- Patient has demonstrated a beneficial response to therapy or stabilization of disease compared to pretreatment age-appropriate baseline values in one or more of the following:
  - Stability or improvement in serum oligosaccharide concentration
  - Stability or improvement in 6-minute walking test (6-MWT)
  - Stability or improvement in 3-minute stair climbing test (3-MSCT)
  - Stability or improvement in forced vital capacity (FVC) (% predicted)
  - Stabilization or slowing in the rate of disease progression or clinical decline

V. Dosage/Administration [1]

Indication	Dose
Alpha-mannosidosis	1 mg/kg (actual body weight) administered once every week as an intravenous infusion

VI. Billing Code/Availability Information

HCPCS Code:

- J0217 – Injection, velmanase alfa-tycv, 1 mg; 1 billable unit = 1 mg

NDC:

- Lamzede 10 mg as a lyophilized powder in a single-dose vial for reconstitution: 10122-0180-xx

VII. References

1. Lamzede [package insert]. Cary, NC; Chiesi USA, Inc.; February 2023. Accessed February 2024.
2. Borgwardt L, Guffon N, Amraoui Y, et al. Efficacy and safety of Velmanase alfa in the treatment of patients with alpha-mannosidosis: results from the core and extension phase analysis of a phase III multicentre, double-blind, randomised, placebo-controlled trial. J Inherit Metab Dis. 2018 Nov;41(6):1215-1223. doi: 10.1007/s10545-018-0185-0. Epub 2018 May 30.
3. Malm D, Nilssen Ø. Alpha-Mannosidosis. Initial Posting: October 11, 2001; Last Revision: July 18, 2019. In: Adam MP, Feldman J, Mirzaa GM, et al, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2024. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1396/. Accessed on February 12, 2024.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E77.1	Defects in glycoprotein degradation

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

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

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

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
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