Lamzede® (velmanase alfa-tycv)
(Intravenous)

Document Number: IC-0696

Last Review Date: 03/02/2023
Date of Origin: 03/02/2023
Dates Reviewed: 03/2023

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 SDV: 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**

***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]

- Patient has a confirmed negative pregnancy test in females of reproductive potential; **AND**
- 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 [1,2]

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 anaphylaxis and severe allergic or 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:

- J3590 – Unclassified biologics *(Discontinue use on 01/01/2024)*
- J0217 – Injection, velmanase alfa-tycv, 1 mg; 1 billable unit = 1 mg *(Effective 01/01/2024)*

NDC:

- Lamzede 10 mg as a lyophilized powder in a SDV for reconstitution: 10122-0180-xx

VII. References

1. Lamzede [package insert]. Cary, NC; Chiesi USA, Inc.; February 2023. Accessed February 2023.
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. GeneReviews. https://www.ncbi.nlm.nih.gov/books/NBK1396/ (Accessed on February 17, 2023).

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)

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 Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications may be covered 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
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.

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC