

PHARMACY POLICY STATEMENT

Ohio Medicaid

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| DRUG NAME | Lamzede (velmanase alfa-tycv) |
| BENEFIT TYPE | Medical |
| STATUS | Prior Authorization Required |

Lamzede, approved by the FDA in 2023, is recombinant human lysosomal alpha-mannosidase indicated for the treatment of non-central nervous system manifestations of alpha-mannosidosis in adult and pediatric patients. Alpha-mannosidosis (AM) is a rare, progressive lysosomal storage disorder caused by pathogenic variants in the MAN2B1 gene, resulting in accumulation of mannose-rich oligosaccharides.

Lamzede is an enzyme replacement therapy intended to provide alpha-mannosidase, the enzyme that is deficient in AM. It is the first approved treatment for AM but does not cross the blood-brain barrier and therefore is not expected to benefit the neurological aspects of the disease. In a Phase 3 clinical trial, 3-minute stair climbing test (3MSCT), 6-minute walking test (6MWT) and forced vital capacity (FVC) numerically favored the Lamzede group and results were supported by a reduction in serum oligosaccharide concentration.

Lamzede (velmanase alfa-tycv) will be considered for coverage when the following criteria are met:

Alpha-Mannosidosis

For **initial** authorization:

1. Medication must be prescribed by or in consultation with a metabolic or genetics specialist, or other specialist experienced with lysosomal storage disorders; AND
2. Member has a diagnosis of alpha-mannosidosis confirmed by enzyme assay showing alpha-mannosidase activity less than 10% of normal; AND
3. Member's disease is mild to moderate, without significant central nervous system (CNS) manifestations; AND
4. Member has NOT had a bone marrow transplant or HSCT.
5. **Dosage allowed/Quantity limit:** 1 mg/kg (actual body weight) once every week as IV infusion.

If all the above requirements are met, the medication will be approved for 12 months.

For **reauthorization**:

1. Chart notes must show improvement or stabilized signs and symptoms of disease demonstrated by at least one of the following:
 - a) Clinically significant reduction of serum oligosaccharide concentration from baseline
 - b) Stable or improved 3MSCT, 6MWT, or FVC

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Lamzede (velmanase alfa-tycv) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

| DATE | ACTION/DESCRIPTION |
|------------|---------------------------------|
| 04/13/2023 | New policy for Lamzede created. |

References:

1. Lamzede [prescribing information]; Chiesi USA, Inc.; 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;41(6):1215-1223. doi:10.1007/s10545-018-0185-0
3. Lund AM, Borgwardt L, Cattaneo F, et al. Comprehensive long-term efficacy and safety of recombinant human alpha-mannosidase (velmanase alfa) treatment in patients with alpha-mannosidosis. *J Inherit Metab Dis*. 2018;41(6):1225-1233. doi:10.1007/s10545-018-0175-2
4. Guffon N, Tytki-Szymanska A, Borgwardt L, et al. Recognition of alpha-mannosidosis in paediatric and adult patients: Presentation of a diagnostic algorithm from an international working group. *Mol Genet Metab*. 2019;126(4):470-474. doi:10.1016/j.ymgme.2019.01.024
5. Malm D, Nilssen Ø. Alpha-Mannosidosis. 2001 Oct 11 [Updated 2019 Jul 18]. In: Adam MP, Mirzaa GM, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2023. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK1396/>

Effective date: 01/01/2024

Revised date: 04/13/2023