

Lamzede® (velmanase alfa-tycv intravenous infusion)

When requesting Lamzede® (velmanase alfa-tycv), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved Indication

Lamzede (velmanase alfa-tycv) is indicated for the treatment of non-central nervous system manifestations of alpha-mannosidosis in adult and pediatric patients.

Coverage Guidelines

Alpha-mannosidosis

The individual must meet **all** of the following criteria for approval:

- Has a confirmed diagnosis of alpha-mannosidosis, defined as alpha-mannosidase activity less than 10% of normal activity in blood leukocytes;
- Has non-central nervous system manifestations (examples of non-central nervous system manifestations include progressive motor function disturbances, physical disability, hearing and speech impairment, skeletal abnormalities, and immune deficiency);
- Has biallelic pathogenic variants in Mannosidase Alpha Class 2B Member 1 (*MAN2B1*) as confirmed by mutation testing;
- Lamzede is prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Approval duration: 12 months

Dosing Recommendation

The recommended dosage of Lamzede is 1 mg/kg (actual body weight) administered once every week as an intravenous infusion.

References

1. Lamzede® intravenous infusion [prescribing information]. Cary, NC: Chiesi USA; 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;41(6):1215-1223.
3. Guffon N, Konstantopoulou V, Hennermann JB, et al. Long-term safety and efficacy of velmanase alpha (VA) treatment in children under 6 years of age with alpha-mannosidosis (AM). Presented at: 14th International Congress of Inborn Errors of Metabolism (ICIM 2021); Sydney, Australia; November 21-23, 2021.

4. Lund A, 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;41:1225-1233.
5. Guffon N, Tylki-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.