Clinical Policy: Galsulfase (Naglazyme)

Reference Number: PA.CP.PHAR.161
Effective Date: 01/2018
Last Review Date: 04/2026

Description
Galsulfase (Naglazyme®) is a hydrolytic lysosomal glycosaminoglycan-specific enzyme.

FDA approved indication
Naglazyme is indicated for the treatment of patients with Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Naglazyme has been shown to improve walking and stair-climbing capacity.

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of PA Health & Wellness that Naglazyme is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Maroteaux-Lamy Syndrome (Mucopolysaccharidosis VI [MPS VI]): (must meet all):

1. Diagnosis of Maroteaux-Lamy syndrome (MPS VI) confirmed by one of the following (a or b):
   a. Enzyme assay demonstrating a deficiency in N-acetygalactosamine 4-sulfatase (arylsulfatase B) activity;
   b. DNA testing;
2. Age ≥ 3 months;
3. Documentation of member's current weight (in kg);
4. Dose does not exceed 1 mg/kg/week.

Approval duration: 12 months

B. Other diagnoses/indications: Refer to PA.CP.PMN.53

II. Continued Approval

A. Maroteaux-Lamy Syndrome (MPS VI): (must meet all)**:**

1. Currently receiving medication via PA Health & Wellness benefit or member has previously met all initial approval criteria; or the Continuity of Care policy (PA. PHARM.01) applies;
2. Member is responding positively to therapy as evidenced by improvement in the individual member's MPS VI (Maroteaux-Lamy syndrome) manifestation profile including 12-minute walking test distance, 3-minute stair climb rate, poor endurance, vision problems, respiratory infections, breathing problems, sleep apnea, high blood pressure, joint stiffness, height and weight, hepatomegaly, or splenomegaly;
3. Documentation of member's current weight (in kg);
4. If request is for a dose increase, new dose does not exceed 1 mg/kg/week.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**
1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy; or the Continuity of Care policy (PA.PHARM.01) applies; or
2. Refer to PA.CP.PMN.53

III. Appendices/General Information

Appendix A: Abbreviation/Acronym Key
FDA: Food and Drug Administration
MPS VI: Mucopolysaccharidosis VI

Appendix B: Therapeutic Alternatives
Not applicable

Appendix C: Contraindications/Boxed Warnings
- Contraindication(s): none reported
- Boxed warning(s): risk of life-threatening hypersensitivity reactions

Appendix D: General Information
The presenting symptoms and clinical course of MPS VI can vary from one individual to another. Some examples, however, of improvement in MPS VI disease as a result of Naglazyme therapy may include improvement in:
- 12-minute walking test distance;
- 3-minute stair climb rate;
- Poor endurance;
- Vision problems;
- Respiratory infections;
- Breathing problems, sleep apnea;
- High blood pressure;
- Joint stiffness;
- Hepatomegaly, splenomegaly.

IV. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
MPS VI	1 mg/kg IV once weekly	1 mg/kg/week

V. Product Availability

Vial: 5 mg/5 mL

VI. References

1. Naglazyme Prescribing Information. Novato, CA: BioMarin Pharmaceutical, Inc.; September 2024. Available at http://www.naglazyme.com. Accessed January 14, 2026.

2. Muenzer J. The mucopolysaccharidoses: a heterogeneous group of disorders with variable pediatric presentations. J Pediatr. 2004; 144(5 Suppl): S27-S34.
3. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS VI: systematic evidence- and consensus-based guidance. Orphanet J of Rare Dis, 2019;12(118)1-21.
4. Stapleton M, Hoshina H, Sawamoto K, et al. Critical review of current MPS guidelines and management. Molecular Genetics and Metabolism. 2019;126:238-45.

Coding Implications
Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J1458	Injection, galsulfase, 1 mg

Reviews, Revisions, and Approvals	Date
2Q 2018 annual review: Modified age restriction to 3 months per PI. Added prescriber requirement. Added max dose criteria. Added requirement for positive response to therapy. References reviewed and updated.	02/2018
2Q 2019 annual review: references reviewed and updated.	04/2019
2Q 2020 annual review: references reviewed and updated.	04/2020
2Q 2021 annual review: references reviewed and updated.	04/2021
2Q 2022 annual review: added requirement for documentation of member's current weight for dose calculation purposes; references reviewed and updated.	04/2022
2Q 2023 annual review: no significant changes; references reviewed and updated.	04/2023
2Q 2024 annual review: no significant changes; references reviewed and updated.	04/2024
2Q 2025 annual review: no significant changes; added new Boxed Warning per label; references reviewed and updated.	04/2025
2Q 2026 annual review: no significant changes; updated initial approval duration from 6 months to 12 months; for Continued Therapy added examples of positive treatment response that were already outlined in Appendix D and previously referred to within the criteria; references reviewed and updated.	04/2026