

Naglazyme® (galsulfase) (Intravenous)

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I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 12 months (365 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

II. Dosing Limits

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

- 115 billable units every 7 days

III. Initial Approval Criteria ¹

Prior authorization validity is provided in the following conditions:

- Member is at least 5 years of age; **AND**
- Documented baseline 12-minute walk test (12-MWT), 3-minute stair climb test (3-MSCT), and/or pulmonary function tests (e.g., FEV1, etc.); **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG); **AND**
- Therapy is being used to treat non-central nervous system manifestations of the disease and member does not have severe, irreversible cognitive impairment; **AND**

NOTE: Requests for continued therapy in members with severe, irreversible cognitive impairment will be reviewed on a case-by-case basis.

Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome) † Φ ^{1,4,5,7}

- Member has a definitive diagnosis of MPS VI as confirmed by the following:
 - Detection of pathogenic (or likely pathogenic) variants in the *ARSB* gene by molecular genetic testing; **OR**
 - Arylsulfatase B (ASB) enzyme activity of <10% of the lower limit of normal in cultured fibroblasts or isolated leukocytes; **AND**

- Member has normal enzyme activity of a different sulfatase (excluding members with Multiple Sulfatase Deficiency [MSD]); **AND**
- Member has an elevated urinary glycosaminoglycan (uGAG) level (i.e. dermatan sulfate or chondroitin sulfate) defined as being above the upper limit of normal by the reference laboratory

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

IV. Renewal Criteria ^{1,4,5}

Prior authorization validity may be renewed based on the following criteria:

- Member continues to meet 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 hypersensitivity reactions, immune-mediated reactions, acute respiratory complications associated with administration, acute cardiorespiratory failure, severe infusion reactions, spinal or cervical cord compression, etc.; **AND**
- Member has a documented reduction in uGAG levels compared to pretreatment baseline; **AND**
- Member has demonstrated a beneficial disease response to therapy compared to pre-treatment baseline values as defined in one or more of the following:
 - Improvement in or stability of 12-minute walk test (12-MWT); **OR**
 - Improvement in or stability of 3-minute stair climb test (3-MSCT); **OR**
 - Improvement in or stability of pulmonary function testing (e.g., FEV1, etc.)

V. Dosage/Administration ¹

Indication	Dose
Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy Syndrome)	1 mg/kg of body weight administered as an intravenous (IV) infusion once a week

VI. Billing Code/Availability Information

HCPCS Code:

- J1458 – Injection, galsulfase, 1 mg; 1 billable unit = 1 mg

NDC:

- Naglazyme 5 mg/5 mL solution; single-dose vial: 68135-0020-xx

VII. References

1. Naglazyme [package insert]. Novato, CA; BioMarin Pharmaceutical Inc.; September 2024. Accessed March 2026.
2. Giugliani R, Harmatz P, Wraith JE. Management guidelines for mucopolysaccharidosis VI. *Pediatrics*. 2007 Aug;120(2):405-18.
3. Giugliani R, Federhen A, Rojas MV, et al. Mucopolysaccharidosis I, II, and VI: Brief review and guidelines for treatment. *Genet Mol Biol*. 2010 Oct;33(4):589-604. Epub 2010 Dec 1.
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6. Harmatz P, Giugliani R, Schwartz I, et al. Enzyme replacement therapy for mucopolysaccharidosis VI: a phase 3, randomized, double-blind, placebo-controlled, multinational study of recombinant human N-acetylgalactosamine 4-sulfatase (recombinant human arylsulfatase B or rhASB) and follow-on, open-label extension study. *J Pediatr*. 2006 Apr;148(4):533-539.
7. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS VI: systematic evidence- and consensus-based guidance. *Orphanet J Rare Dis*. 2019 May 29;14(1):118. doi: 10.1186/s13023-019-1080-y.
8. Zhou J, Lin J, Leung WT, Wang L. A basic understanding of mucopolysaccharidosis: Incidence, clinical features, diagnosis, and management. *Intractable Rare Dis Res*. 2020 Feb;9(1):1-9. doi: 10.5582/iridr.2020.01011.
9. Shapiro EG, Eisengart JB. The natural history of neurocognition in MPS disorders: A review. *Mol Genet Metab*. 2021 May;133(1):8-34. doi: 10.1016/j.ymgme.2021.03.002. Epub 2021 Mar 11. PMID: 33741271.

Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	Yes: Consider for PA

Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E76.29	Other mucopolysaccharidoses

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
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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