Naglazyme® (galsulfase)
(Intravenous)

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I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

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

- 115 billable units every 7 days

III. Initial Approval Criteria [1]

Submission of supporting clinical documentation (including but not limited to medical records, chart notes, lab results, and confirmatory diagnostics) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission as part of the evaluation of this request. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e., genetic, and mutational testing) supporting initiation when applicable. Please provide documentation via direct upload through the PA web portal or by fax. Failure to submit the medical records may result in the denial of the request due to inability to establish medical necessity in accordance with policy guidelines.

Coverage is provided in the following conditions:

Patient is required to meet Site of Service specialty infusion program requirements (refer to the Dean Health Plan Site of Service Policy).

- Patient 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 patient does not have severe, irreversible cognitive impairment; **AND**

NOTE: *Requests for continued therapy in patients 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]

- Patient has a definitive diagnosis of MPS VI as confirmed by the following:
  - Detection of pathogenic mutations 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**
    - Patient has normal enzyme activity of a different sulfatase (excluding patients with Multiple Sulfatase Deficiency [MSD]); **AND**
    - Patient 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); Φ Orphan Drug

IV. Renewal Criteria [1,4,5]

Coverage can be renewed based on the following criteria:

- Patient 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**
- Patient has a documented reduction in uGAG levels compared to pretreatment baseline; **AND**
- Patient 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 2025.
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.
4. Vairo F, Federhen A, Baldo G, et al. Diagnostic and treatment strategies in mucopolysaccharidosis VI. Appl Clin Genet. 2015 Oct 30;8:245-55.
5. Valaannopoulos V, Nicely H, Harmatz P, et al. Mucopolysaccharidosis VI. Orphanet J Rare Dis. 2010; 5: 5.
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/irdr.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 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