

## 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

#### A. Quantity Limit (max daily dose) [NDC Unit]:

- Naglazyme 5 mg/5 mL vial: 23 vials per 7 days

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

- 115 billable units every 7 days

### III. Initial Approval Criteria <sup>1</sup>

Coverage is provided in the following conditions:

- 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**

**Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome) † Φ <sup>1,4,5</sup>**

- 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); Ⓢ Orphan Drug

#### IV. Renewal Criteria <sup>1,4,5</sup>

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**
- Disease response with treatment as defined by improvement or stability from pre-treatment baseline by the following:
  - Reduction in uGAG levels; **AND**
    - Improvement in or stability of 12-minute walk test compared (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 <sup>1</sup>

Indication	Dose
Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy Syndrome)	1 mg/kg 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.; December 2019. Accessed December 2022.
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.

## Appendix 1 – Covered Diagnosis Codes

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

## Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

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 Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered 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, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
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

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- Qualified sign language interpreters
- Written information in other formats (large print, audio, accessible electronic formats, other formats)

Provides free language services to people whose primary language is not English, such as:

- Qualified interpreters
- Information written in other languages

If you need these services, contact a Grievance Specialist.

If you believe that PCHP has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, or sex, you can file a grievance with:

Grievance Specialist  
PreferredOne Community Health Plan  
PO Box 59052  
Minneapolis, MN 55459-0052  
Phone: 1.800.940.5049 (TTY: 763.847.4013)  
Fax: 763.847.4010  
[customerservice@preferredone.com](mailto:customerservice@preferredone.com)

You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, a Grievance Specialist is available to help you.

You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at <https://ocrportal.hhs.gov/ocr/portal/lobby.jsf>, or by mail or phone at:

U.S. Department of Health and Human Services  
200 Independence Avenue, SW  
Room 509F, HHH Building  
Washington, D.C. 20201  
1-800-368-1019, 800-537-7697 (TDD)

Complaint forms are available at <http://www.hhs.gov/ocr/office/file/index.html>.

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LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1.800.940.5049 (TTY: 763.847.4013).

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Fax: 763.847.4010  
[customerservice@preferredone.com](mailto:customerservice@preferredone.com)

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Washington, D.C. 20201  
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