

Naglazyme® (galsulfase) (Intravenous)

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Date of Origin: 11/28/2011

Dates Reviewed: 12/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018

I. Length of Authorization

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

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Naglazyme 5 mg vial: 23 vials per 7 days

B. Max Units (per dose and over time) [Medical Benefit]:

- 115 billable units every 7 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome) †

- Patient has a definitive diagnosis of MPS VI 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 GAG level as defined as being above the upper limit of normal by the reference laboratory; **AND**
- Patient aged 5 years or older; **AND**
- Documented baseline 12-minute walk test (12-MWT), 3-minute stair climb test, and/or pulmonary function tests (e.g., FEV₁, etc.); **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG)

IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: anaphylaxis and allergic reactions, immune-mediated reactions, acute respiratory complications, acute cardiorespiratory reactions, 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**
 - 12-minute walk test (12-MWT); **OR**
 - 3-minute stair climb test compared to pre-treatment baseline; **OR**
 - Pulmonary function testing (e.g., FEV1, etc.)

V. Dosage/Administration

Indication	Dose
Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome)	1 mg/kg of body weight administered as an intravenous infusion over no less than 4 hours once a week

VI. Billing Code/Availability Information

Jcode:

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

NDC:

Naglazyme 5 mg per 5 mL solution; single-use vial: 68135-0020-xx

VII. References

1. Naglazyme [package insert]. Novato, CA; BioMarin Pharmaceutical Inc.; March 2013. Accessed August 2018.
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.

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) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-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): 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 Government Benefit Administrators, 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