Naglazyme® (galsulfase) (Intravenous)

Effective Date: 01/01/2020  
Review Date: 12/13/2019, 1/29/2020, 5/27/2021  
Revision date: 12/13/2019, 1/29/2020  
Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

I. Length of Authorization

Coverage will be provided for 6 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:
   - Naglazyme 5 mg 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 Criteria¹,²,⁴,⁵,⁶

Coverage is provided in the following conditions:

MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

Universal Criteria

- Patient aged 5 years or older; AND

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 glycosaminoglycan (uGAG) level (i.e. dermatan sulfate or chondroitin sulfate) defined as being above the upper limit of normal by the reference laboratory; **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)

† FDA-approved indication(s)

### IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

• Patient continues to meet universal and other 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, 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; **OR**
    - Improvement in or stability of pulmonary function testing (e.g., FEV1, etc.)

### V. Dosage/Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome)</td>
<td>1 mg/kg administered as an intravenous (IV) infusion once a week</td>
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</table>

### VI. Billing Code/Availability Information

**HCPCS Code:**

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


Appendix 1 – Covered Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tr>
<td>E76.29</td>
<td>Other mucopolysaccharidoses</td>
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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 Articles may exist and comply with these policies is required where applicable. They can be found at: [http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx](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/Article): N/A

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<tr>
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