**Fabrazyme® (agalsidase beta)**
*(Intravenous)*

**Last Review Date:** 02/04/2020  
**Date of Origin:** 11/28/2011  
**Dates Reviewed:** 02/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018, 02/2019, 2/2020

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]:**
      - Fabrazyme 5 mg vial: 6 per 14 days
      - Fabrazyme 35 mg vial: 3 per 14 days

   B. **Max Units (per dose and over time) [HCPCS Unit]:**
      - 115 billable units every 14 days

III. **Initial Approval Criteria**

Site of care specialty infusion program requirements are met (refer to Moda Site of Care Policy).

Coverage is provided in the following conditions:

**Universal Criteria**

- Patient is 8 years of age or older: **AND**
- Must not be used in combination with migalastat: **AND**

**Fabry Disease (alpha-galactosidase A deficiency) †**

- Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
  - α-galactosidase A (α-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (males only): **OR**
  - Plasma or urinary globotriaosylceramide(Gb₃/GL–3) or globotriaosylsphingosine (lyso-Gb₃): **OR**
  - Detection of pathogenic mutations in the GALA/GLA gene by molecular genetic testing: **AND**

- Baseline value for plasma GL-3 and/or GL-3 inclusions: **AND**

† 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 the following: severe hypersensitivity reactions, severe infusion site reactions, compromised cardiac function, etc.: AND
- Disease response with treatment as defined by a reduction in plasma GL-3 and/or GL-3 inclusions compared to pre-treatment baseline

V. Dosage/Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fabry Disease</td>
<td>1 mg/kg of body weight infused every two weeks as an intravenous (IV) infusion.</td>
</tr>
</tbody>
</table>

VI. Billing Code/Availability Information

HCPCS code:

- J0180 – Injection, agalsidase beta, 1 mg; 1 billable unit = 1 mg

NDC:

- Fabrazyme 5 mg single-use vial for injection: 54868-0041-xx
- Fabrazyme 35 mg single-use vial for injection: 54868-0040-xx

Appendix 1 – Covered Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
</tr>
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<tbody>
<tr>
<td>E75.21</td>
<td>Fabry (-Anderson) disease</td>
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</table>

VII. References


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

<table>
<thead>
<tr>
<th>Jurisdiction</th>
<th>Applicable State/US Territory</th>
<th>Contractor</th>
</tr>
</thead>
<tbody>
<tr>
<td>E (1)</td>
<td>CA, HI, NV, AS, GU, CNMI</td>
<td>Noridian Healthcare Solutions, LLC</td>
</tr>
<tr>
<td>F (2 &amp; 3)</td>
<td>AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ</td>
<td>Noridian Healthcare Solutions, LLC</td>
</tr>
<tr>
<td>5</td>
<td>KS, NE, IA, MO</td>
<td>Wisconsin Physicians Service Insurance Corp (WPS)</td>
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<tr>
<td>6</td>
<td>MN, WI, IL</td>
<td>National Government Services, Inc. (NGS)</td>
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<td>H (4 &amp; 7)</td>
<td>LA, AR, MS, TX, OK, CO, NM</td>
<td>Novitas Solutions, Inc.</td>
</tr>
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<td>First Coast Service Options, Inc.</td>
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<td>Palmetto GBA, LLC</td>
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<tr>
<td>M (11)</td>
<td>NC, SC, WV, VA (excluding below)</td>
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<td>Novitas Solutions, Inc.</td>
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<tr>
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<td>CGS Administrators, LLC</td>
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