Lumizyme® (alglucosidase alfa) (Intravenous)

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]:
   - Lumizyme 50 mg vial: 46 vials every 14 days
B. Max Units (per dose and over time) [HCPCS Unit]:
   - 230 billable units every 14 days

III. Initial Approval Criteria
Coverage is provided in the following conditions:

Universal Criteria

Pompe disease (Acid alpha-glucoSIDase (GAA) deficiency) †
- Diagnosis has been confirmed by one of the following:
  - Deficiency of acid alpha-glucosidase (GAA) enzyme activity; OR
  - Detection of biallelic pathogenic variants in the GAA gene by molecular genetic testing; AND
- Documented baseline values for one or more of the following:
  - Infantile-onset disease: muscle weakness, motor function, respiratory function, cardiac involvement, percent predicted forced vital capacity (FVC), and/or 6 minute walk test (6MWT); OR
  - Late-onset (non-infantile) disease: FVC and/or 6MWT

† FDA approved indication(s)
IV. Renewal Criteria\textsuperscript{1,4,7,8}

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 cutaneous reactions, systemic immune-mediated reactions, acute cardiorespiratory failure, cardiac arrhythmia and sudden cardiac death during general anesthesia, etc.; AND
- No evidence that patient has developed IgG antibodies to alglucosidase alfa at a sustained titer level of $\geq 12,800$; AND
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
  - Infantile-onset disease: stabilization or improvement in muscle weakness, motor function, respiratory function, cardiac involvement, FVC, and/or 6MWT
  - Late-onset (non-infantile) disease: stabilization or improvement in FVC and/or 6MWT

V. Dosage/Administration\textsuperscript{1,7,8}

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Pompe disease</td>
<td>20 mg/kg administered as an intravenous (IV) infusion every 2 weeks</td>
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</tbody>
</table>

VI. Billing Code/Availability Information

HCPCS Code:

- J0221 – Injection, alglucosidase alfa, (Lumizyme), 10 mg; 1 billable unit = 10 mg

NDC:

- Lumizyme 50 mg single-use vial for injection: 58468-0160-xx

VII. References


**Appendix 1 – Covered Diagnosis Codes**

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tbody>
<tr>
<td>E74.02</td>
<td>Pompe disease</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 compliance 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/Articles): N/A

### Medicare Part B Administrative Contractor (MAC) Jurisdictions

<table>
<thead>
<tr>
<th>Jurisdiction</th>
<th>Applicable State/US Territory</th>
<th>Contractor</th>
</tr>
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<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>
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<td>5</td>
<td>KS, NE, IA, MO</td>
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<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>
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<td>8</td>
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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>L (12)</td>
<td>DE, MD, PA, NJ, DC (includes Arlington &amp; Fairfax counties and the city of Alexandria in VA)</td>
<td>Novitas Solutions, Inc.</td>
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## Medicare Part B Administrative Contractor (MAC) Jurisdictions

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<td>15</td>
<td>KY, OH</td>
<td>CGS Administrators, LLC</td>
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