I. Length of Authorization

Initial coverage will be provided for 6 months and may be renewed annually thereafter.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:
   - Kanuma 20 mg/10 mL single-use vials: 68 vials per 28 day supply

B. Max Units (per dose and over time) [Medical Benefit]:
   - 340 billable units once weekly

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Lysosomal Acid Lipase (LAL) deficiency †

- Diagnosis has been confirmed by either biallelic pathogenic variants in LIPA or deficient LAL enzyme activity in peripheral blood leukocytes, fibroblasts, or dried blood spots: AND
- Patient is at least 1 month old

† FDA Approved Indication(s)

IV. Renewal Criteria

Coverage can be renewed based upon 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: hypersensitivity reactions (anaphylaxis, abdominal pain, fever, chills, pruritus, rash, vomiting), etc.: AND
- Treatment has resulted in clinical benefit; for example:
  - Improvement in weight-for-age z-scores for patients exhibiting growth failure
  - Improvement in LDL
V. **Dosage/Administration**

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>LAL deficiency</td>
<td>Pediatric &amp; Adult patients:</td>
</tr>
<tr>
<td></td>
<td>• 1 mg/kg administered once every other week as an IV infusion</td>
</tr>
<tr>
<td></td>
<td>Rapidly progressive disease presenting within the first 6 months of life:</td>
</tr>
<tr>
<td></td>
<td>• 1 mg/kg administered once weekly as an IV infusion</td>
</tr>
<tr>
<td></td>
<td>• May increase to 3 mg/kg once weekly for patients who do not achieve an optimal clinical response</td>
</tr>
</tbody>
</table>

VI. **Billing Code/Availability Information**

**Jcode:**

J2840 - Injection, sebelipase alfa, 1 mg

**NDC(s):**

Kanuma 20 mg/10 mL single-use vials: 25682-0007-01

VII. **References**


**Appendix 1 – Covered Diagnosis Codes**

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>E75.5</td>
<td>Other lipid storage disorders</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) 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](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>
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<td>Noridian Healthcare Solutions, LLC</td>
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<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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<tr>
<td>5</td>
<td>KS, NE, IA, MO</td>
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<tr>
<td>6</td>
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<tr>
<td>8</td>
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<td>Wisconsin Physicians Service Insurance Corporation (WPS)</td>
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<td>First Coast Service Options, Inc.</td>
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<tr>
<td>M (11)</td>
<td>NC, SC, WV, VA (excluding below)</td>
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</tr>
<tr>
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<td>Novitas Solutions, Inc.</td>
</tr>
<tr>
<td>K (13 &amp; 14)</td>
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<td>CGS Administrators, LLC</td>
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