Elfabrio® (pegunigalsidase alfa-iwxj) (Intravenous)

Document Number: IC-0708

Last Review Date: 06/01/2023
Date of Origin: 06/01/2023
Dates Reviewed: 06/2023

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]:
   • Elfabrio 20 mg/10 mL single-dose vial for injection: 6 vials every 14 days

B. Max Units (per dose and over time) [HCPCS Unit]:
   • 120 mg every 14 days

III. Initial Approval Criteria ¹

Coverage is provided in the following conditions:

• Patient is at least 18 years of age; AND

Universal Criteria

• Must not be used in combination with migalastat or agalsidase beta; AND

Fabry Disease (alpha-galactosidase A deficiency) † ¹,³,⁷,¹³

• Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
  o α-galactosidase A (α-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (males only); OR
  o Detection of pathogenic mutations in the GLA gene by molecular genetic testing; AND

• Baseline value for plasma GL-3 and/or GL-3 inclusions, plasma or urinary globotriaosylceramide (Gb3/GL-3); or plasma globotriaosylphosphoglycerine (lyso-Gb3)

† FDA approved indication(s); ‡ Compendia recommended indication(s); © Orphan Drug
IV. Renewal Criteria

Coverage 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 severe hypersensitivity reactions, severe infusion-associated reactions, glomerulonephritis, etc.: AND
  - Disease response with treatment as defined by a reduction or stabilization in one or more of the following, as compared to pre-treatment baseline:
    - plasma GL-3 and/or GL-3 inclusions
    - plasma or urinary globotriaosylceramide (Gb3/GL-3)
    - plasma globotriaosylsphingosine (lyso-Gb3): OR
  - Disease response with treatment as defined by a improvement or stabilization in the rate of decline of the estimated glomerular filtration rate (eGFR)

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:
- J3590 – Unclassified biologics

NDC:
- Elfabrio 20 mg/10 mL single-dose vial for injection: 10122-0160-xx

VII. References


Appendix 1 – Covered Diagnosis Codes

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

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 Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/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/LCA): 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>
</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>
</tr>
<tr>
<td>6</td>
<td>MN, WI, IL</td>
<td>National Government Services, Inc. (NGS)</td>
</tr>
<tr>
<td>H (4 &amp; 7)</td>
<td>LA, AR, MS, TX, OK, CO, NM</td>
<td>Novitas Solutions, Inc.</td>
</tr>
<tr>
<td>8</td>
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<td>Wisconsin Physicians Service Insurance Corp (WPS)</td>
</tr>
<tr>
<td>N (9)</td>
<td>FL, PR, VI</td>
<td>First Coast Service Options, Inc.</td>
</tr>
<tr>
<td>J (10)</td>
<td>TN, GA, AL</td>
<td>Palmetto GBA, LLC</td>
</tr>
<tr>
<td>M (11)</td>
<td>NC, SC, WV, VA (excluding below)</td>
<td>Palmetto GBA, LLC</td>
</tr>
<tr>
<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>
</tr>
<tr>
<td>K (13 &amp; 14)</td>
<td>NY, CT, MA, RI, VT, ME, NH</td>
<td>National Government Services, Inc. (NGS)</td>
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<tr>
<td>15</td>
<td>KY, OH</td>
<td>CGS Administrators, LLC</td>
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