

## Elfabrio® (pegunigalsidase alfa-iwxj) (Intravenous)

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### I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 12 months (365 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

### II. Dosing Limits

**Max Units (per dose and over time) [HCPCS Unit]:**

- 115 billable units (115 mg) every 14 days

### III. Initial Approval Criteria <sup>1</sup>

Prior authorization validity is provided in the following conditions:

- Member 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) † <sup>1,3,7,13</sup>**

- Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
  - Deficiency in  $\alpha$ -galactosidase A ( $\alpha$ -Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (*males only*); **OR**
  - Detection of pathogenic (or likely pathogenic) variants in the *GLA* gene by molecular genetic testing; **AND**
- Member has a baseline value for at least one of the following:
  - Tissue globotriaosylceramide (Gb3/GL-3) inclusions
  - Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lyso-Gb3)

- Clinical signs and/or symptoms of disease (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations)

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Ⓢ Orphan Drug

#### IV. Renewal Criteria <sup>1,3,13</sup>

Prior authorization validity may be renewed based on the following criteria:

- Member continues to meet the 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:
    - Tissue globotriaosylceramide (Gb3/GL-3) inclusions
    - Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lyso-Gb3); **OR**
  - Disease response with treatment as defined by an improvement or stabilization of clinical signs and/or symptoms (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations)

#### V. Dosage/Administration <sup>1</sup>

| Indication    | Dose                                                                                           |
|---------------|------------------------------------------------------------------------------------------------|
| Fabry Disease | Administer 1 mg/kg (based on actual body weight) by intravenous (IV) infusion every two weeks. |

#### VI. Billing Code/Availability Information

HCPSC Code:

- J2508 – Injection, pegunigalsidase alfa-iwxj, 1 mg; 1 billable unit = 1 mg

NDC:

- Elfabrio 5 mg/2.5 mL single-dose vial for injection: 10122-0165-xx
- Elfabrio 20 mg/10 mL single-dose vial for injection: 10122-0160-xx

## VII. References

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## Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

| Factor                     | Conclusion            |
|----------------------------|-----------------------|
| Indication                 | Yes: Consider for PA  |
| Safety and efficacy        | Yes: Consider for PA  |
| Potential for misuse/abuse | No: PA not a priority |
| Cost of drug               | Yes: Consider for PA  |

## Appendix 1 – Covered Diagnosis Codes

| ICD-10 | ICD-10 Description        |
|--------|---------------------------|
| E75.21 | Fabry (-Anderson) disease |

## Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications, including any preceding information, may be applied 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 |                                                                                             |                                                   |
|---------------------------------------------------------------|---------------------------------------------------------------------------------------------|---------------------------------------------------|
| Jurisdiction                                                  | Applicable State/US Territory                                                               | Contractor                                        |
| E (1)                                                         | CA, HI, NV, AS, GU, CNMI                                                                    | Noridian Healthcare Solutions, LLC                |
| F (2 & 3)                                                     | AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ                                                      | Noridian Healthcare Solutions, LLC                |
| 5                                                             | KS, NE, IA, MO                                                                              | Wisconsin Physicians Service Insurance Corp (WPS) |
| 6                                                             | MN, WI, IL                                                                                  | National Government Services, Inc. (NGS)          |
| H (4 & 7)                                                     | LA, AR, MS, TX, OK, CO, NM                                                                  | Novitas Solutions, Inc.                           |
| 8                                                             | MI, IN                                                                                      | Wisconsin Physicians Service Insurance Corp (WPS) |
| N (9)                                                         | FL, PR, VI                                                                                  | First Coast Service Options, Inc.                 |
| J (10)                                                        | TN, GA, AL                                                                                  | Palmetto GBA                                      |
| M (11)                                                        | NC, SC, WV, VA (excluding below)                                                            | Palmetto GBA                                      |
| L (12)                                                        | DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA) | Novitas Solutions, Inc.                           |
| K (13 & 14)                                                   | NY, CT, MA, RI, VT, ME, NH                                                                  | National Government Services, Inc. (NGS)          |
| 15                                                            | KY, OH                                                                                      | CGS Administrators, LLC                           |