Fabrazyme® (agalsidase beta)
(Intravenous)

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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 single-dose vial: 6 vials per 14 days
- Fabrazyme 35 mg single-dose vial: 3 vials per 14 days

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

- 115 billable units every 14 days

III. Initial Approval Criteria [1]

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

Coverage is provided in the following conditions:

- Patient is at least 2 years of age; **AND**

Universal Criteria

- Must not be used in combination with migalastat or pegunigalsidase alfa-iwxj; **AND**

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

- Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
  - Deficiency in α-galactosidase A (α-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (*males only*); **OR**
  - Detection of pathogenic mutations in the *GLA* gene by molecular genetic testing; **AND**
- Patient has a baseline of one or more 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); Φ Orphan Drug

IV. Renewal Criteria [1,3,13]

Coverage may be renewed based on the following criteria:

- Patient 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, 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 [1]

Indication	Dose
Fabry Disease	Administer 1 mg/kg (based on body weight) every two weeks as an intravenous (IV) infusion.

VI. Billing Code/Availability Information

HCPCS Code:

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

NDC:

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

VII. References

1. Fabrazyme [package insert]. Cambridge, MA; Genzyme Corporation.; March 2023. Accessed January 2024.
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January 4, 2024. Available from https://www.uptodate.com/contents/fabry-disease-clinical-features-and-diagnosis.

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, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
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