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 vial: 6 per 14 days
- Fabrazyme 35 mg vial: 3 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; **AND**

Fabry Disease (alpha-galactosidase A deficiency) † Φ [1-6]

- Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
  - α-galactosidase A (α-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (males only); **OR**
  - Plasma or urinary globotriaosylceramide(Gb_3/GL-3) or globotriaosylsphingosine (lyso-Gb_3); **OR**

- Detection of pathogenic mutations in the *GALA/GLA* gene by molecular genetic testing; **AND**

- Baseline value for plasma GL-3 and/or GL-3 inclusions

† FDA approved indication(s); Φ Orphan Drug

IV. Renewal Criteria [1]

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 the following: anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, compromised cardiac function, etc.; **AND**
- Disease response with treatment as defined by a reduction in plasma GL-3 and/or GL-3 inclusions compared to pre-treatment baseline

V. Dosage/Administration

Indication	Dose
Fabry Disease	1 mg/kg of body weight infused 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-use vial for injection: 54868-0041-xx
- Fabrazyme 35 mg single-use vial for injection: 54868-0040-xx

VII. References

1. Fabrazyme [package insert]. Cambridge, MA; Genzyme Corporation.; March 2021. Accessed March 2021.
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4. Biegstraaten M, Arngrímsson R, Barbey F, et al. Recommendations for initiation and cessation of enzyme replacement therapy in patients with Fabry disease: the European Fabry Working Group consensus document. Orphanet J Rare Dis. 2015 Mar 27;10:36.
5. Hopkin RJ, Jefferies JL, Laney DA, et al. The management and treatment of children with Fabry disease: A United States-based perspective. Mol Genet Metab. 2016 Feb;117(2):104-13.
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7. Kes VB, Cesarik M, Zavoreo I, et al. Guidelines for diagnosis, therapy and follow up of Anderson-Fabry disease. Acta Clin Croat. 2013 Sep;52(3):395-405.
8. Branton MH, Schiffmann R, Sabnis SG, et al. Natural history of Fabry renal disease: influence of alpha-galactosidase A activity and genetic mutations on clinical course. Medicine (Baltimore). 2002 Mar;81(2):122-38.
9. Banikazemi M, Bultas J, Waldek S, et al. Agalsidase-beta therapy for advanced Fabry disease: a randomized trial. 2007 Jan 16;146(2):77-86. doi: 10.7326/0003-4819-146-2-200701160-00148. Epub 2006 Dec 18.
10. Wraith E, Tylki-Szymanska A, Guffon N, et al. Safety and efficacy of enzyme replacement therapy with agalsidase beta: an international, open-label study in pediatric patients with Fabry disease. J Pediatr. 2008 Apr;152(4):563-70, 570.e1. doi: 10.1016/j.jpeds.2007.09.007. Epub 2007 Dec 3.
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Appendix 1 – Covered Diagnosis Codes

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

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: 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/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