



Fabrazyme[®] (agalsidase beta) (Intravenous)

Document Number: IC-0042

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Date of Origin: 11/28/2011

Dates Reviewed: 02/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018

I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Fabrazyme 5 mg vial: 6 per 14 days
- Fabrazyme 35 mg vial: 3 per 14 days

B. Max Units (per dose and over time) [Medical Benefit]:

- 115 billable units every 14 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Fabry Disease (alpha-galactosidase A deficiency) †

- Patient is 8 years of age or older; **AND**
- 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(Gb₃/GL-3) or globotriaosylsphingosine (lyso-Gb₃); **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; **AND**
- Must not be used in combination with migalastat
- † FDA approved indication(s)

IV. Renewal Criteria

Authorizations can be renewed based on 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 the following: severe hypersensitivity reactions, severe infusion site 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	<ul style="list-style-type: none">▪ The recommended dosage of Fabrazyme is 1 mg/kg body weight infused every two weeks as an intravenous (IV) infusion.▪ Patients should receive antipyretics prior to infusion. The initial IV infusion rate should be no more than 0.25 mg/min (15 mg/hr). The infusion rate may be slowed in the event of infusion reactions. After patient tolerance to the infusion is well established, the infusion rate may be increased in increments of 0.05 to 0.08 mg/min (increments of 3 to 5 mg/hr) with each subsequent infusion. For patients weighing < 30 kg, the maximum infusion rate should remain at 0.25 mg/min (15 mg/hr). For patients weighing ≥ 30 kg, the administration duration should not be less than 1.5 hours (based on individual patient tolerability).

VI. Billing Code/Availability Information

Jcode:

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.; July 2010. Accessed August 2018.
2. Mehta A, Beck M, Eyskens F, et al. Fabry disease: a review of current management strategies. QJM. 2010 Sep; 103(9):641-59.
3. Mehta A, Hughes DA. Fabry Disease. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1292/ (Accessed on September 6, 2017).

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.
6. Laney DA, Bennett RL, Clarke V, et al. Fabry disease practice guidelines: recommendations of the National Society of Genetic Counselors. *J Genet Couns.* 2013 Oct;22(5):555-64.
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. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Drugs and Biologics (L34741). Centers for Medicare & Medicaid Services, Inc. Updated on 06/21/2017 with effective date 07/1/2017. Accessed September 2017.

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 Biologics. 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>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD):

Jurisdiction(s): 5,8	NCD/LCD Document (s): L34741
https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34741&bc=gAAAAAAAAAAAAA==	

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)

FABRAZYME® (agalsidase beta) Prior Auth Criteria

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Medicare Part B Administrative Contractor (MAC) Jurisdictions

Jurisdiction	Applicable State/US Territory	Contractor
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto Government Benefit Administrators, 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