

Lumizyme[®] (alglucosidase alfa) (Intravenous)

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Date of Origin: 11/28/2011

Dates Reviewed: 12/2011, 02/2013, 02/2014, 08/2014, 07/2015, 10/2016, 10/2017, 10/2018, 02/2019

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]:

- Lumizyme 50 mg vial: 46 vials every 14 days

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

- 230 billable units every 14 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Pompe disease (Acid alpha-glucosidase (GAA) deficiency) †

- Diagnosis has been confirmed by one of the following:
 - Deficiency of acid alpha-glucosidase (GAA) enzyme activity; OR
 - Detection of pathogenic variants in the GAA gene by molecular genetic testing;
AND
- Documented baseline values for one or more of the following:
 - Infantile-onset disease: muscle weakness, motor function, respiratory function, cardiac involvement, percent predicted forced vital capacity (FVC), and/or 6 minute walk test (6MWT); OR
 - Late-onset (non-infantile) disease: FVC and/or 6MWT

† 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 allergic and anaphylactic reactions, severe cutaneous and systemic immune-mediated reactions, acute cardiorespiratory failure, cardiac arrhythmia and sudden cardiac death during general anesthesia, etc.; AND
- No evidence that patient has developed IgG antibodies to alglucosidase alfa at a sustained titer level of $\geq 12,800$; AND
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
 - Infantile-onset disease: stabilization or improvement in muscle weakness, motor function, respiratory function, cardiac involvement, FVC, and/or 6MWT
 - Late-onset (non-infantile) disease: stabilization or improvement in FVC and/or 6MWT

V. Dosage/Administration

Indication	Dose
Pompe disease	20 mg/kg body weight as an intravenous (IV) infusion every 2 weeks

VI. Billing Code/Availability Information

Jcode:

- J0221 – Injection, alglucosidase alfa, (Lumizyme), 10 mg; 1 billable unit = 10 mg

NDC:

- Lumizyme 50 mg single-use vial for injection: 58468-0160-xx

VII. References

1. Lumizyme [package insert]. Cambridge, MA; Genzyme Corporation.; August 2014. Accessed January 2019.
2. Cupler EJ, Berger KI, Leshner RT, et al. Consensus treatment recommendations for late-onset Pompe disease. Muscle Nerve. 2012 Mar; 45(3):319-33. doi: 10.1002/mus.22329. Epub 2011 Dec 15.
3. Kishnani PS, Steiner RD, Bali D, et al. Pompe disease diagnosis and management guidelines. Genet Med 2006; 8:267-88.

4. Nancy L, Bailey L. Pompe Disease. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1261/ (Accessed on August 11, 2018).
5. Tarnopolsky M, Katzberg H, Petrof BJ, et al. Pompe Disease: Diagnosis and Management. Evidence-Based Guidelines from a Canadian Expert Panel. Can J Neurol Sci. 2016 Jul;43(4):472-85.
6. Kishnani PS, Hwu WL, et al. Introduction to the Newborn Screening, Diagnosis, and Treatment for Pompe Disease Guidance Supplement. Pediatrics 2017 Jul;(1):S1-S3

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E74.02	Pompe 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) 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): 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