



Nexviazyme® (avalglucosidase alfa-ngpt) (Intravenous)

Document Number: SHP-0615

Last Review Date: 02/01/2024

Date of Origin: 09/01/2021

Dates Reviewed: 09/2021, 02/2022, 04/2022, 02/2023, 02/2024

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]:

- Nexviazyme 100 mg single-dose vial: 23 vials every 14 days

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

- 575 billable units (2300 mg) every 14 days

III. Initial Approval Criteria ^{1,4}

Submission of medical records (chart notes) related to the medical necessity criteria is **REQUIRED** on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Medical records may be submitted via direct upload through the PA web portal or by fax.

Coverage is provided in the following conditions:

- Patient age is at least 1 year of age; **AND**
- Patient has documented baseline values for percent predicted forced vital capacity (FVC) and/or 6-minute walk test (6MWT); **AND**

****NOTE:** For very young patients in which FVC or 6-MWT are not suitable for measuring, requests will be reviewed on a case-by case basis.

Universal Criteria ¹

- Will not be used in combination with other enzyme replacement therapies (i.e., alglucosidase-alfa, cipaglucosidase alfa, etc.); **AND**

- Patients susceptible to fluid volume overload or those with an acute underlying respiratory illness or compromised cardiac or respiratory function, will be closely monitored for exacerbation of their cardiac or respiratory status during infusion; **AND**

Pompe Disease (Acid Alpha-Glucosidase (GAA) deficiency) † Φ ^{1,4}

- Diagnosis has been confirmed by one of the following:
 - Deficiency of acid alpha-glucosidase (GAA) enzyme activity; **OR**
 - Detection of biallelic pathogenic variants in the *GAA* gene by molecular genetic testing; **AND**
- Patient has a diagnosis of late-onset (non-infantile) disease

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

IV. Renewal Criteria ^{1,4}

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: anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, acute cardiorespiratory failure, etc.; **AND**
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following: stabilization or improvement in FVC and/or 6-MWT; **AND**
- Patient is being monitored for antibody formation (including neutralizing antibodies)

V. Dosage/Administration ¹

Indication	Dose
Pompe Disease	<p>Patients weighing ≥30 kg: Administer 20 mg/kg (of actual body weight) every two weeks as an intravenous infusion.</p> <p>Patients weighing <30 kg: Administer 40 mg/kg (of actual body weight) every two weeks as an intravenous infusion.</p>

VI. Billing Code/Availability Information

HCPCS Code:

- J0219 – Injection, avalglucosidase alfa-ngpt, 4 mg; 1 billable unit = 4 mg

NDC:

- Nexviazyme 100 mg single-dose vial as a powder for injection: 58468-0426-xx

VII. References

1. Nexviazyme [package insert]. Cambridge, MA; Genzyme Corporation.; September 2023. Accessed January 2024.
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/. Initial Posting: August 31, 2007; Last Update: November 2, 20237. Accessed on January 14, 2024.
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.
7. Maner J, Kishani P, Kushlaf H, et al. Safety and efficacy of avalglucosidase alfa versus alglucosidase alfa in patients with late-onset Pompe disease (COMET): a phase 3, randomised, multicentre trial. *Lancet Neurol*. 2021 Dec;20(12):1012-1026. doi: 10.1016/S1474-4422(21)00241-6.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E74.02	Pompe 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