Nexviazyme™ (avalglucosidase alfa-ngpt)
(Intravenous)

Document Number: SHP-0615

Last Review Date: 04/01/2022
Date of Origin: 09/01/2021
Dates Reviewed: 09/2021, 02/2022, 04/2022

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 powder for inj.: 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]

Coverage is provided in the following conditions:

> 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.

- 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); **AND**

- Patient has not experienced a severe hypersensitivity reaction including anaphylaxis to alglucosidase alfa *(Note: exception to this criterion can be made when Nexviazyme is used as part of a desensitization procedure);* **AND**
- Patient is not susceptible to fluid volume overload and does not have an acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated; **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: disease 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	**Patients weighing ≥30 kg:** Administer 20 mg/kg (of actual body weight) every two weeks as an intravenous infusion. **Patients weighing <30 kg:** Administer 40 mg/kg (of actual body weight) every two weeks as an intravenous infusion.

VI. Billing Code/Availability Information

HCPCS Code:

- J3590 – Unclassified Biologics *(Discontinue use on 04/01/2022)*
- C9085 – Injection, avalglucosidase alfa-ngpt, 4 mg; 1 billable unit = 4 mg *(Discontinue use on 04/01/2022)*
- J0219 – Injection, avalglucosidase alfa-ngpt, 4 mg; 1 billable unit = 4 mg *(Effective 04/01/2022)*

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.; August 2021. Accessed January 2022.
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: May 11, 2017. Accessed on January 20, 2022.
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. Genzyme. A Phase 3 Randomized, Multicenter, Multinational, Double-blinded Study Comparing the Efficacy and Safety of Repeated Biweekly Infusions of Avalglucosidase Alfa (neoGAA, GZ402666) and Alglucosidase Alfa in Treatment naïve Patients With Late-onset Pompe Disease **(COMET).** Available from: https://clinicaltrials.gov/ct2/show/NCT02782741?term=NCT02782741&draw=2&rank=1. NLM identifier: NCT02782741. Accessed August 9, 2021.

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), 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: https://www.cms.gov/medicare-coverage-database/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