

**Clinical Policy: Avalglucosidase Alfa-ngpt (Nexviazyme)**

Reference Number: LA.PHAR.521

Effective Date: 09.29.23Last Review Date: 05.01.23 04.05.24

Line of Business: Medicaid

[Coding Implications](#)[Revision Log](#)

See [Important Reminder](#) at the end of this policy for important regulatory and legal information.

**\*\*Please note:** This policy is for medical benefit\*\*

**Description**

Avalglucosidase alfa-ngpt (Nexviazyme™) is a hydrolytic lysosomal glycogen-specific enzyme.

**FDA Approved Indication(s)**

Nexviazyme is indicated for the treatment of patients 1 year of age and older with late-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency).

**Policy/Criteria**

*Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.*

It is the policy of Louisiana Healthcare Connections® that Nexviazyme is **medically necessary** when the following criteria are met:

**I. Initial Approval Criteria****A. Pompe Disease** (must meet all):

1. Diagnosis of late-onset Pompe disease confirmed by one of the following (a or b):
  - a. Enzyme assay confirming low GAA activity;
  - b. DNA testing;
2. Age ≥ 1 year;
3. Nexviazyme is not prescribed concurrently with Lumizyme®;
4. Dose does not exceed any of the following (a or b):
  - a. Members weighing ≥ 30 kg: 20 mg/kg every 2 weeks;
  - b. Members weighing < 30 kg: 40 mg/kg every 2 weeks.

**Approval duration:** 6 months

**B. Other diagnoses/indications** (must meet 1 or 2):

1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255
2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy for the relevant line of business: LA.PMN.53 for Medicaid.

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#### II. Continued Therapy

##### A. Pompe Disease (must meet all):

1. Currently receiving medication via Louisiana Healthcare Connections benefit or member has previously met initial approval criteria;
2. Member is responding positively to therapy as evidenced by improvement in the individual member's Pompe disease manifestation profile (*see Appendix D for examples*);
3. Nexviazyme is not prescribed concurrently with Lumizyme;
4. If request is for a dose increase, new dose does not exceed any of the following (a or b):
  - a. Members weighing  $\geq 30$  kg: 20 mg/kg every 2 weeks;
  - b. Members weighing  $< 30$  kg: 40 mg/kg every 2 weeks.

**Approval duration:** 12 months

##### B. Other diagnoses/indications (must meet 1 or 2):

1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255
2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy for the relevant line of business: LA.PMN.53 for Medicaid.

#### III. Diagnoses/Indications for which coverage is NOT authorized:

- ##### A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policies – LA.PMN.53 for Medicaid, or evidence of coverage documents.

#### IV. Appendices/General Information

*Appendix A: Abbreviation/Acronym Key*

6MWT: 6 minute walk test

GAA: acid alpha-glucosidase

FDA: Food and Drug Administration

*Appendix B: Therapeutic Alternatives*

Not applicable

*Appendix C: Contraindications/Boxed Warnings*

- Contraindication(s): none reported
- Boxed warning(s): severe hypersensitivity reactions; infusion-associated reactions; risk of acute cardiorespiratory failure in susceptible patients

*Appendix D: Measures of Therapeutic Response*

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- Pompe disease manifests as a clinical spectrum that varies with respect to age at onset\*, rate of disease progression, and extent of organ involvement. Patients can present with a variety of signs and symptoms, which can include cardiomegaly, cardiomyopathy, hypotonia, muscle weakness, respiratory distress (eventually requiring assisted ventilation), and skeletal muscle dysfunction.
- While there is not one generally applicable set of clinical criteria that can be used to determine appropriateness of continued therapy, clinical parameters that can indicate therapeutic response to Nexviazyme include improved or maintained forced vital capacity, improved or maintained 6 minute walk test (6MWT) distance.

*\*Although infantile-onset disease typically presents in the first year of life, age of onset alone does not necessarily distinguish between infantile- and late-onset disease since juvenile-onset disease can present prior to 12 months of age.*

#### V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
Pompe disease	For patients weighing $\geq 30$ kg: 20 mg/kg every 2 weeks For patients weighing $< 30$ kg: 40 mg/kg every 2 weeks	40 mg/kg/2 weeks

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#### VI. Product Availability

Lyophilized powder in a single-dose vial: 100 mg

#### VII. References

- Nexviazyme Prescribing Information. Cambridge, MA: Genzyme Corporation; August 2021. Available at: [https://www.accessdata.fda.gov/drugsatfda\\_docs/label/2021/761194s0001b1.pdf](https://www.accessdata.fda.gov/drugsatfda_docs/label/2021/761194s0001b1.pdf). [https://www.accessdata.fda.gov/drugsatfda\\_docs/label/2021/761194s0001b1.pdf](https://www.accessdata.fda.gov/drugsatfda_docs/label/2021/761194s0001b1.pdf). Accessed February 14, 2022, 2023.
- Pena LDM, Barohn RJ, Byrne BJ, et al. Safety, tolerability, pharmacokinetics, pharmacodynamics, and exploratory efficacy of the novel enzyme replacement therapy avalglucosidase alfa (neoGAA) in treatment-naïve and alglucosidase alfa-treated patients with late-onset Pompe disease: A phase 1, open-label, multicenter, multinational, ascending dose study. *Neuromuscular Disorders* 2019;29:167-86.
- Cupler EJ, Berger KI, Leshner RT, et al. *Consensus treatment recommendations for late-onset Pompe disease*. *Muscle Nerve* 2012;45:319-33.

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#### Coding Implications

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPSC Codes	Description
<del>C9085</del> J0219	Injection, avalglucosidase alfa-ngpt, 4 mg

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Reviews, Revisions, and Approvals	Date	LDH Approval Date
Policy created	05.01.23	<u>08.28.23</u>
<u>Annual review: no significant changes; HCPCS coding update; references reviewed and updated.</u>	<u>04.05.24</u>	

#### **Important Reminder**

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. LHCC makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions, and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable LHCC administrative policies and procedures.

This clinical policy is effective as of the date determined by LHCC. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. LHCC retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment, or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

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