

Clinical Policy: Avalglucosidase Alfa-ngpt (Nexviazyme)

Reference Number: PA.CP.PHAR.521

Effective Date: 10/2021

Last Review Date: 01/2026

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 PA Health & Wellness® 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;
 - c. Increased lysosomal glycogen;
2. Nexviazyme is not prescribed concurrently with Lumizyme® or the combination of Pombiliti™ with Opfolda™;
3. 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: 12 months

B. Other diagnoses/indications

1. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

II. Continued Therapy

A. Pompe Disease (must meet all):

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.PHARM.01) applies ;
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 or the combination of Pombiliti™ with Opfolda™;

4. If request is for a dose increase, new 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: 12 months

B. Other diagnoses/indications

1. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

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 – PA.CP.PMN.53

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

- 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 ≥ 30 kg: 20 mg/kg every 2 weeks	40 mg/kg/2 weeks
	For patients weighing < 30 kg: 40 mg/kg every 2 weeks	

VI. Product Availability

Lyophilized powder in a single-dose vial: 100 mg

VII. References

1. Nexvzyme Prescribing Information. Cambridge, MA: Genzyme Corporation; September 2023. Available at: www.nexvzyme.com. Accessed November 6, 2025.
2. 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.
3. Cupler EJ, Berger KI, Leshner RT, et al. Consensus treatment recommendations for late-onset Pompe disease. *Muscle Nerve* 2012;45:319-33.
4. Stevens D, Milani-Nejad S, Mozaffar T. Pompe disease: a clinical, diagnostic, and therapeutic overview. *Curr Treat Options Neurol.* 2022 November;24(11):573-88. doi:10.1007/s11940-022-00736-1.

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.

HCPCS Codes	Description
J0219	Injection, avalglucosidase alfa-ngpt, 4 mg

Reviews, Revisions, and Approvals	Date
Policy created	10/2021
2Q 2022 annual review: references reviewed and updated.	04/2022
2Q 2023 annual review: no significant changes; references reviewed and updated.	04/2023
2Q 2024 annual review: added exclusion for concomitant use with Pombiliti+Opfolda to align with the Pombiliti criteria; references reviewed and updated.	04/2024
1Q 2025 annual review: moving forward to 1Q annual review cycle to consolidate with the Opfolda+Pombiliti annual review cycle; added increased lysosomal glycogen as an additional option for confirming a Pompe disease diagnosis; references reviewed and updated.	01/2025
1Q 2026 annual review: no significant changes; updated initial auth duration from 6 months to 12 months; references reviewed and updated.	01/2026