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# 5.30.077

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<b>Section:</b>	Prescription Drugs	<b>Effective Date:</b>	April 1, 2026
<b>Subsection:</b>	Endocrine and Metabolic Agents	<b>Original Policy Date:</b>	August 27, 2021
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**Last Review Date:** March 6, 2026

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## Nexviazyme

### Description

#### Nexviazyme (avalglucosidase alfa-ngpt)

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#### Background

Nexviazyme (avalglucosidase alfa-ngpt) is indicated for late-onset Pompe disease, a rare genetic disorder. In Pompe disease, a gene mutation prevents the body from making an enzyme or making enough of the enzyme called acid alpha-glucosidase (GAA), necessary for proper muscle function. GAA is used by the heart and muscle cells to convert stored glycogen into energy. Without sufficient enzyme action, glycogen builds up in the cells, ultimately weakening the heart and other muscles. Infusion of Nexviazyme replaces the deficient GAA, reducing the accumulated glycogen in the body (1).

#### Regulatory Status

FDA-approved indication: 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) (1).

Nexviazyme has a boxed warning for severe hypersensitivity reactions, including anaphylaxis, infusion-associated reactions, and risk of cardiorespiratory failure in susceptible patients. In severe hypersensitivity reactions (e.g., anaphylaxis), Nexviazyme should be discontinued immediately and appropriate treatment initiated. Infusion-associated reactions (IARs) have also been reported to occur at any time during infusion, up to a few hours after the infusion has concluded. In severe IARs, immediate discontinuation of Nexviazyme and appropriate care should be provided. In mild to moderate IARs, rechallenge with slower infusion rates or lower doses have been shown to reduce symptoms. Patients with advanced Pompe disease may

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have compromised heart and lung function and are at serious risk of decompensation during Nexviazyme infusion. Vitals should be monitored more frequently in this population, and some patients may require prolonged observation times (1).

Patients with Pompe disease may have compromised cardiac function, including issues such as cardiomyopathy, heart failure, and arrhythmia. Prior to Nexviazyme infusion, an echocardiogram may be helpful to assess for cardiomyopathy or cardiac hypertrophy and a 12 lead EKG can assist in excluding arrhythmia (2).

The safety and effectiveness of Nexviazyme in pediatric patients less than 1 year of age have not been established (1).

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## Related policies

Lumizyme

[Policy](#)

*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.*

Nexviazyme may be considered **medically necessary** if the conditions indicated below are met.

Nexviazyme may be considered **investigational** for all other indications.

## Prior-Approval Requirements

**Age** 1 year of age and older

### Diagnosis

Patient must have **ALL** of the following:

1. Late-onset Pompe disease (acid alpha-glucosidase (GAA) deficiency)
  - a. Prescriber agrees to monitor for hypersensitivity reactions and infusion-associated reactions and to initiate treatment as needed
  - b. Prescriber agrees to frequently monitor the vitals of patients at risk for fluid volume overload during medication infusion

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- c. Prescriber agrees to assess for cardiac issues such as cardiomyopathy, cardiac hypertrophy and arrhythmia using an echocardiogram and a 12 lead EKG prior to initiating therapy with Nexviazyme

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## Prior-Approval *Renewal* Requirements

Same as above

### Policy Guidelines

## Pre-PA Allowance

None

## Prior-Approval Limits

**Duration** 2 years

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## Prior-Approval *Renewal* Limits

Same as above

### Rationale

#### Summary

Nexviazyme (avalglucosidase alfa-ngpt) is a lysosomal glycogen-specific enzyme indicated for patients 1 year of age and older with late-onset Pompe disease (acid  $\alpha$ -glucosidase (GAA) deficiency). Nexviazyme has a boxed warning that hypersensitivity reactions, infusion-associated reactions and cardiorespiratory failure in susceptible patients has been observed during and after infusions. Appropriate medical support should be available during infusion and some patients may require prolonged observation time after infusion has concluded. The safety and effectiveness of Nexviazyme in pediatric patients less than 1 year of age have not been established (1).

Prior approval is required to ensure the safe, clinically appropriate, and cost-effective use of Nexviazyme while maintaining optimal therapeutic outcomes.

#### References

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1. Nexviazyme [package insert]. Cambridge, MA: Genzyme Corporation; September 2023.
2. Kishnani, P. S., Steiner, R. D., Bali, D., Berger, K., Byrne, B. J., Case, L. E., Crowley, J. F., Downs, S., Howell, R. R., Kravitz, R. M., Mackey, J., Marsden, D., Martins, A. M., Millington, D. S., Nicolino, M., O'Grady, G., Patterson, M. C., Rapoport, D. M., Slonim, A., Watson, M. S. (2006, May). *Pompe disease diagnosis and management guideline*. Genetics in medicine : official journal of the American College of Medical Genetics. Retrieved from <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3110959/>.

## Policy History

Date	Action
December 2021	Addition to PA. Annual review. Per SME, added requirement: prescriber agrees to assess for cardiac issues such as cardiomyopathy, cardiac hypertrophy and arrhythmia using an echocardiogram and a 12 lead EKG prior to initiating therapy with Nexviazyme.
September 2022	Annual review
March 2023	Annual review
March 2024	Annual review and reference update
March 2025	Annual review
March 2026	Annual review

## Keywords

**This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 6, 2026 and is effective on April 1, 2026.**