

Nexviazyme (avalglucosidase alfa-ngpt)

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Dosing Limit
Nexviazyme (avalglucosidase alfa-ngpt) 100 mg vial	20 mg/kg every 2 weeks*

*Nexviazyme (avalglucosidase alfa-ngpt) may be approved 40 mg/kg every 2 weeks for individuals <30 kg.

APPROVAL CRITERIA

Initial requests for Nexviazyme (avalglucosidase alfa-ngpt) may be approved if the following criteria are met:

- I. Individual has a diagnosis of non-infantile onset (late-onset) Pompe disease as confirmed by all of following (ACMG 2006):
 - A. Documentation is provided for GAA enzyme assay which shows reduced enzyme activity less than 40% of the lab specific normal mean value; **AND**
 - B. Documentation is provided for a second GAA enzyme activity assay in a separate sample (from purified lymphocytes, fibroblasts or muscle) or by GAA gene sequencing (AANEM 2009); **AND**
 - C. Forced vital capacity (FVC) 30 -79% of predicted value, and documentation is provided; **AND**
 - D. Ability to walk 40 meters on a 6-minute walk test (assistive devices permitted), and documentation is provided;

Continuation requests for Nexviazyme (avalglucosidase alfa-ngpt) may be approved if the following criteria are met:

- I. Individuals with non-infantile onset (late-onset) Pompe disease are responding to therapy (including improvement, stabilization, or slowing of disease progression)

Requests for Nexviazyme (avalglucosidase alfa-ngpt) may not be approved for the following:

- I. In combination with Lumizyme (alglucosidase alfa); **OR**
- II. In combination with Pombiliti; **OR**
- III. When the above criteria are not met and for all other indications.

Key References:

1. American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM). Pompe. Available at: <https://www.aanem.org/Patients/Disorders/Pompe>. Accessed on August 5, 2022.
2. American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) Consensus Committee on Late-onset Pompe Disease. Consensus treatment recommendations for late-onset Pompe disease. Muscle Nerve. 2012 Mar;45(3):319-33. Accessed: August 3, 2022.
3. American College of Medical Genetics (ACMG) Work Group on Management of Pompe Disease. Pompe disease diagnosis and management guideline. Genetics in Med. 2006; 8(5):267-288.
4. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2022. URL: <http://www.clinicalpharmacology.com>. Updated periodically.
5. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. <http://dailymed.nlm.nih.gov/dailymed/about.cfm>. Accessed: August 5, 2022
6. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
7. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically.
8. Lumizyme [Package insert], Cambridge, MA. Genzyme Corporation; 2022.
9. Nexviazyme [Package insert], Cambridge, MA. Genzyme Corporation; 2021.

Federal and state laws or requirements, contract language, and Plan utilization management programs or policies may take precedence over the application of this clinical criteria.

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