Avalglucosidase alfa-ngpt (Nexviazyme®)	Place of Service Office Administration Infusion Center Administration Home Infusion Administration Outpatient Facility Administration* [*Prior authorization required – see section (1)] HCPCS: J0219 per 4 mg

Condition listed in policy (*see criteria for details*)

- Late-onset Pompe disease

AHFS therapeutic class: Enzyme

Mechanism of action: Hydrolytic lysosomal glycogen-specific enzyme

(1) Special Instructions and pertinent Information

Covered under the medical benefit, please submit clinical information for prior authorization review via fax.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****

AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

Members with the following plans: **PPO, Direct Contract HMO, and when applicable, Medi-Cal, ASO/Shared Advantage/HMO (non-direct contract),** may be required to have their medication administered at a preferred site of service, including the home, a physician's office, or an independent infusion center not associated with a hospital.

For members that cannot receive infusions in the preferred home or ambulatory setting AND meet one of the following criteria points, drug administration may be performed at a hospital outpatient facility infusion center.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****

AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

ADMINISTRATION OF NEXVIAZYME IN THE HOSPITAL OUTPATIENT FACILITY SITE OF CARE REQUIRES ONE OF THE FOLLOWING: (*Supporting Documentation must be submitted)*

1. **Patient is receiving their first infusion of Nexviazyme or is being re-initiated on Nexviazyme after at least 6 months off therapy.** *Subsequent doses will require medical necessity for continued use in the hospital outpatient facility site of care.*

Additional clinical monitoring is required during administration as evidenced by one of the following:

2. **Patient has experienced a previous severe adverse event to Nexviazyme based on documentation submitted.**

3. **Patient continues to experience moderate to severe adverse events to Nexviazyme based on documentation submitted, despite receiving premedication such as acetaminophen, steroids, diphenhydramine, fluids, etc.**

4. **Patient is clinically unstable based on documentation submitted.**

5. **Patient is physically or cognitively unstable based on documentation submitted.**

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for avalglucosidase alfa-ngpt (Nexviazyme®) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Late-onset Pompe disease

1. Diagnosis is late-onset Pompe disease (also known as Glycogen Storage Disease Type II or acid maltase deficiency), **AND**
2. One of the following:
    a. Genetic testing showing acid alpha-glucosidase (GAA) mutation, or
    b. An enzyme assay showing absent or decreased acid alpha-glucosidase (GAA) activity from blood, skin, or muscle tissues

Covered Doses

Weight range (kg)	Dosage regimen
Less than 30 kg	20 mg/kg (of actual body weight) IV every 2 weeks
Greater than or equal to 30 kg	40 mg/kg (of actual body weight) IV every 2 weeks

Coverage Period
Indefinitely

ICD-10:
E74.02

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for avalglucosidase alfa-ngpt (Nexviazyme®) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s)

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional Information

How supplied:

- 100 mg single-dose vial

(6) References

- AHFS®. Available by subscription at http://www.lexi.com
- DrugDex®. Available by subscription at http://www.micromedexsolutions.com/home/dispatch
- Nexviazyme (avalglucosidase alfa-ngpt) [Prescribing information]. Cambridge, MA: Genzyme Corporation; 4/2023.
- The American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) Consensus Treatment Recommendations for Late-Onset Pompe Disease Muscle Nerve 2012 Mar 45(3): 319-333

(7) Policy Update

Date of last review: 3Q2023
Date of next review: 3Q2024
Changes from previous policy version:

- No clinical change to policy following routine annual review.

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee