avalglucosidase alfa-ngpt (Nexviazyme)

Medical Benefit Drug Policy

Place of Service
Home Infusion Administration
Infusion Center Administration
Office Administration
Outpatient Facility Administration

Drug Details

USP Category: GENETIC OR ENZYME OR PROTEIN DISORDER: REPLACEMENT, MODIFIERS, TREATMENT
Mechanism of Action: Hydrolytic lysosomal glycogen-specific enzyme
HCPCS:
J0219:Injection, avalglucosidase alfa-ngpt, 4 mg
How Supplied:
100 mg single-dose vial

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

- Late-Onset Pompe Disease

Any condition not listed in this policy requires a review to confirm it is medically necessary. For conditions that have not been approved for intended use by the Food and Drug Administration (i.e., off-label use), the criteria outlined in the California Code of Regulations (CCR), Title 22, Section 51303 and 51313 must be met.

Special Instructions and Pertinent Information

Provider must submit documentation (such as office chart notes, lab results or other clinical information) to ensure the member has met all medical necessity requirements.

The member's specific benefit may impact drug coverage. Other utilization management processes, and/or legal restrictions may take precedence over the application of this clinical criteria.

Coverage Criteria
The following condition(s) require Prior Authorization/Preservice.

Late-Onset Pompe Disease
Meets medical necessity if all the following are met:
1. Diagnosis is late-onset Pompe disease (also known as Glycogen Storage Disease Type II or acid maltase deficiency)
2. Meets ONE of the following:
   a. Genetic testing showing acid alpha-glucosidase (GAA) mutation
   b. An enzyme assay showing absent or decreased acid alpha-glucosidase (GAA) activity from blood, skin, or muscle tissues

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Covered Doses:

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

Coverage Period:
yearly, based on continued response to therapy

ICD-10:
E74.02

References

1. AHFS. Available by subscription at http://www.lexi.com
2. DrugDex. Available by subscription at http://www.micromedexsolutions.com/home/dispatch
3. National Organization for Rare Disorders. Pompe Disease. https://rarediseases.org/rare-diseases/pompe-disease/. Last updated January 18, 2024. Accessed May 19, 2025
4. Nexviazyme (avalglucosidase alfa-ngpt) Prescribing Information. Genzyme Corporation, Cambridge, MA: 9/2023.
5. The American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) Consensus Treatment Recommendations for Late-Onset Pompe Disease Muscle Nerve 2012 Mar 45(3): 319-333. Available at https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3534745/.
6. Stevens D, Milani-Nejad S, Mozaffar T. Pompe Disease: a Clinical, Diagnostic, and Therapeutic Overview. Curr Treat Options Neurol. 2022 Nov;24(11):573-588. Epub 2022 Aug 4. Available at https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10035871/pdf/nihms-1847668.pdf

Review History

Date of Last Annual Review: 3Q2026
Changes from previous policy version:

- No change to policy following routine annual review

Blue Shield of California Medication Policy to Determine Medical Necessity
Reviewed by P&T Committee