Drug Coverage Policy

Effective Date……08/15/2026
Coverage Policy Number……IP0279
Policy Title…… Nexviazyme

Pompe Disease – Enzyme Replacement Therapy – Nexviazyme

- Nexviazyme® (avalglucosidase alfa-ngpt intravenous infusion – Genzyme)

INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

OVERVIEW

Nexviazyme, a hydrolytic lysosomal glycogen-specific recombinant human α-glucosidase enzyme, is indicated for **late-onset Pompe disease** (lysosomal acid α-glucosidase deficiency) in patients ≥ 1 year of age.[1]

Disease Overview
Pompe disease (glycogen storage disease type II, or acid maltase deficiency), is a rare lysosomal storage disorder characterized by a deficiency in acid α-glucosidase activity leading to the accumulation of glycogen, particularly in muscle.[2] In general, the condition is defined by the age of onset, organ involvement, severity and rate of progression. Late-onset Pompe disease has a more variable clinical course and can manifest any time after 12 months of age. Patients typically present with progressive muscle weakness which can progress to respiratory insufficiency. The diagnosis of Pompe disease is established a deficiency of acid α-glucosidase activity or by identification of biallelic pathogeneic variants by genetic testing. Enzyme replacement therapy has an important role in the management of Pompe disease.

Coverage Policy

POLICY STATEMENT
Prior Authorization is required for benefit coverage of Nexviazyme. Approval is recommended for those who meet the **Criteria** and **Dosing** for the listed indication. Extended approvals are allowed if the patient continues to meet the Criteria and Dosing. Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Nexviazyme as well as the monitoring required for adverse events and long-term efficacy, approval requires Nexviazyme to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Documentation: Documentation is required where noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes, laboratory tests, claims records, prescription receipts and/or other information. All documentation must include patient-specific identifying information.

Nexviazyme is considered medically necessary when the following criteria are met:

FDA-Approved Indication

1. **Acid Alpha-Glucosidase Deficiency (Pompe Disease).** Approve for 1 year if the patient meets ALL of the following (A, B, C, and D):
   - **A)** Patient is ≥ 1 year of age; AND
   - **B)** Patient has late-onset acid alpha-glucosidase deficiency (late-onset Pompe disease) **[documentation required]**; AND
   - **C)** The diagnosis is established by ONE of the following (i or ii):
     - **i.** Patient has a laboratory test demonstrating deficient acid alpha-glucosidase activity in blood, fibroblasts, or muscle tissue **[documentation required]**; OR
     - **ii.** Patient has a molecular genetic test demonstrating biallelic pathogenic or likely pathogenic acid alpha-glucosidase (GAA) gene variants **[documentation required]**; AND
   - **D)** The medication is prescribed by or in consultation with a geneticist, neurologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Dosing. Approve ONE of the following dosing regimens (A or B):

A) Patient ≥ 30 kg: Dose is 20 mg/kg administered by intravenous infusion once every 2 weeks; OR

B) Patient < 30 kg: Dose is 40 mg/kg administered by intravenous infusion once every 2 weeks.

Conditions Not Covered

Nexviazyme for any other use is considered not medically necessary. Criteria will be updated as new published data are available.

Coding Information

Note: 1) This list of codes may not be all-inclusive.
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

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

References

1. Nexviazyme® intravenous infusion [prescribing information]. Cambridge, MA: Genzyme; September 2023.
2. Sperry E, Leslie N, Berry L, et al. Pompe Disease. 2007 Aug 31 [Updated 2025 Aug 21]. In: Adam MP, Bick S, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2026. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1261/. Accessed on May 4, 2026.

Revision Details

Summary of Changes	Review Date	Effective Date
Policy Name Change: Updated Policy Name from "Avalglucosidase" to "Pompe Disease – Enzyme Replacement Therapy – Nexviazyme." No criteria changes.	06/20/2024	08/15/2024
No criteria changes.	05/29/2026	07/15/2025
No criteria changes.	05/21/2026	07/15/2026
Added documentation requirements.	07/09/2026	08/15/2026

The policy effective date is in force until updated or retired.

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