



Drug Coverage Policy

Effective Date.....08/15/2024

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 and have discretion in making individual coverage determinations. 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.

Cigna Healthcare Coverage Policy

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 \geq 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.^{2,3} The onset, progression, and severity of Pompe

disease is variable. Infantile-onset Pompe disease usually manifests in the first few months of life and death often occurs in the first year of life, if left untreated.² Clinical manifestations of infantile-onset Pompe disease includes hypotonia, difficulty feeding, and cardiopulmonary failure.⁴ Late-onset Pompe disease has a more variable clinical course and can manifest any time after 12 months of age.^{3,4} Patients typically present with progressive muscle weakness which can progress to respiratory insufficiency. The diagnosis of Pompe disease is established by demonstrating decreased acid α -glucosidase activity in blood, fibroblasts, or muscle tissue; or by genetic testing.

Medical Necessity Criteria

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 \geq 1 year of age; AND

B) Patient has late-onset acid alpha-glucosidase deficiency (late-onset Pompe disease); 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; OR

ii. Patient has a molecular genetic test demonstrating biallelic pathogenic or likely pathogenic acid alpha-glucosidase (GAA) gene variants; 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 \geq 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.

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Receipt of sample product does not satisfy any criteria requirements for coverage.

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven (criteria will be updated as new published data are available).

Coding Information

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; April 2023.
2. Chien YH, Hwu WL, Lee NC. Pompe disease: Early diagnosis and early treatment make a difference. *Pediatr Neonatol.* 2013;54:219-227.
3. Llerena Junior JC, Nascimento OJM, Oliveira ASB, et al. Guidelines for the diagnosis, treatment and clinical monitoring of patients with juvenile and adult Pompe disease. *Arq Neuropsiquiatr.* 2016;74:166-176.
4. Cupler EJ, Berger KI, Leshner RT, et al. Consensus treatment recommendations for late-onset Pompe disease. *Muscle Nerve.* 2012;45:319-333.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	Policy Name Change: Updated Policy Name from "Avalglucosidase" to "Pompe Disease – Enzyme Replacement Therapy – Nexviazyme." No criteria changes.	08/15/2024

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

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