Effective Date **1/15/2024**
Next Review Date… **1/15/2025**
Coverage Policy Number **IP0279**

Avalglucosidase

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Related Coverage Resources

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. 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

This policy supports medical necessity review for avalglucosidase alfa-ngpt intravenous infusion (**Nexviazyme®**).

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

Medical Necessity Criteria

Avalglucosidase alfa-ngpt (Nexviazyme) is considered medically necessary when ONE of the following is met:

1. **Acid Alpha-Glucosidase Deficiency (Pompe Disease).** Individual meets **ALL** of the following criteria:
   A. Age 1 year or older
   B. Diagnosis of late-onset acid alpha-glucosidase deficiency (late-onset Pompe disease) is confirmed by documentation of **ONE** of the following:

i. A laboratory test demonstrating deficient acid alpha-glucosidase activity in dry blood spot, fibroblasts, lymphocytes, or muscle tissue
ii. A molecular genetic test demonstrating biallelic pathogenic or likely pathogenic variants in the *GAA* gene

C. 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. **ONE** of the following dosing regimens:

1. Individual greater than or equal to 30 kg: Dose is 20 mg/kg administered by intravenous infusion once every 2 weeks
2. Individual less than 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.

Reauthorization Criteria

Continuation of avalglucosidase alfa-ngpt (Nexviazyme) is considered medically necessary for the treatment of late-onset acid alpha-glucosidase deficiency (late-onset Pompe disease) when the above medical necessity criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration: up to 12 months.
Reauthorization approval duration: up to 12 months.

Conditions Not Covered

Any other use is considered experimental, investigational or unproven.

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

Background

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.[2,3] 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.

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.