Medical Policy:

Nexviazyme™ (avalglucosidase alfa-ngpt) intravenous infusion

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.343	March 17, 2025	December 9, 2021

Definitions

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

Length of Authorization

Approvals will be granted for 12 months and may be renewed.

Dosing Limits [Medical Benefit]

Approve up to 40 mg/kg (of actual body weight) every two weeks.

Max Units (per dose and over time) [HCPCS Unit]:

575 billable units (2300 mg) every 14 days

Guideline

I. INITIAL APPROVAL CRITERIA

1. **<u>Acid Alpha-Glucosidase Deficiency (Pompe Disease):</u>**
   Approve if the patient meets all the following criteria:
   A. Patient is ≥ 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. Patient has documented baseline values for percent predicted forced vital capacity (FVC) and/or 6-minute walk test (6MWT); **AND**
E. Will not be used in combination with other enzyme replacement therapies (i.e., alglucosidase-alfa, cipaglucosidase alfa, etc.); **AND**
F. 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.

II. RENEWAL CRITERIA

Coverage can be renewed based on the following criteria:

1. Patient continues to meet identified Initial criteria; **AND**
2. Absence of unacceptable toxicity from the drug. *Examples of unacceptable toxicity include: anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, acute cardiorespiratory failure, etc.*; **AND**
3. Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following: stabilization or improvement in FVC and/or 6-MWT; **AND**
4. Patient is being monitored for antibody formation (including neutralizing antibodies)

Limitations/Exclusions

Coverage is not recommended for circumstances not listed in the Guideline. Criteria will be updated as new published data are available

Applicable Procedure Codes

Code	Description
J0219	Injection, avalglucosidase alfa-ngpt, 4 mg

Applicable NDCs

Code	Description
58468-0426-01	Solution reconstituted , 100 mg per single dose vial

ICD-10 Diagnoses

Code	Description
E74.02	Pompe disease

Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	3/17/2025	Annual Review: Initial Criteria: Removed" acid alpha-glucosidase gene mutation; AND" from the following statement: "Patient has a molecular genetic test demonstrating acid alpha-glucosidase gene mutation; AND" replaced with:

		"biallelic pathogenic or likely pathogenic acid alpha-glucosidase (GAA) gene variants;" Added: "Patient has documented baseline values for percent predicted forced vital capacity (FVC) and/or 6-minute walk test (6MWT); AND Will not be used in combination with other enzyme replacement therapies (i.e., alglucosidase-alfa, cipaglucosidase alfa, etc.); AND" Added Renewal Criteria: "Coverage can be renewed based on the following criteria: Patient continues to meet identified Initial criteria; AND Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, acute cardiorespiratory failure, etc.; AND Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following: stabilization or improvement in FVC and/or 6-MWT; AND Patient is being monitored for antibody formation (including neutralizing antibodies)"
EmblemHealth & ConnectiCare	2/1/2024	Annual Review: no criteria changes
EmblemHealth & ConnectiCare	5/30/2023	Annual Review: no criteria changes
EmblemHealth & ConnectiCare	09/13/2022	Transferred policy to new template. Updated billing codes from C9085 and J3590 to J0219.
EmblemHealth & ConnectiCare	12/9/2021	New Policy

References

1. NEXVIAZYME™ intravenous infusion [package insert]. Cambridge, MA. Genzyme Corporation. Updated Aug 6, 2021. Accessed Sep 15, 2021.
2. Kishnani P, Attarian S, Lindolfo Borges J, et al. Efficacy and safety results of the avalglucosidase alfa phase 3 COMET trial in late-onset Pompe disease patients [poster]. Presented at: The World Muscle Society Virtual Congress; September 25-October 2, 2020.