

Medical Policy:

Nexviazyme™ (avalglucosidase alfa-ngpt) intravenous infusion

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

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The treating physician or primary care provider must submit to EmblemHealth, or ConnectiCare, as applicable (hereinafter jointly referred to as “EmblemHealth”), the clinical evidence that the member meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request preauthorization or post-payment review. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Health care providers are expected to exercise their medical judgment in rendering appropriate care.

EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary.

If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication.

EmblemHealth may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice. EmblemHealth Services Company, LLC, has adopted this policy in providing management, administrative and other services to EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC, and Health Insurance Plan of Greater New York (HIP) related to health benefit plans offered by these entities. ConnectiCare, an EmblemHealth company, has also adopted this policy. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

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. **Acid Alpha-Glucosidase Deficiency (Pompe Disease):**

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 subspecialist, 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.