

Title: UM-PT131 Nexviazyme (avalglucosidase alfa-ngpt)	Division: Medical Management Department: Pharmacy
Approval Date: 12/16/2025	LOB: Medicaid, SNP, HARP, CHP, QHP, EP, Gold, Goldcare
Effective Date: 12/16/2025	Policy Number: UM-PT131
Review Date: 12/16/2025	Cross Reference Number:
Retired Date:	Page 1 of 5

I. POLICY DESCRIPTION:

Enzyme Replacement Therapy – Hydrolytic Lysosomal Glycogen-Specific Enzyme – Nexviazyme (avalglucosidase alfa-ngpt)

II. RESPONSIBLE PARTIES:

Medical Management Administration, Pharmacy Department, Utilization Management, Integrated Care Management, Claims Department

III. DEFINITIONS:

Nexviazyme is a hydrolytic lysosomal glycogen-specific enzyme indicated for the treatment of late-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency). Nexviazyme is indicated for the treatment of late-onset Pompe disease in patients 1 year of age or older. Pompe disease is an inherited disorder of glycogen metabolism caused by deficiency of lysosomal enzyme acid alpha-glucosidase (GAA). Nexviazyme provides an exogenous source of GAA that exerts enzymatic activity in the cleaving glycogen.

IV. POLICY:

Nexviazyme will be considered medically necessary once the following coverage criteria is met. Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

Chart notes must be submitted to confirm diagnosis and previous treatment(s).

INITIAL REQUEST:

1. Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency)
 - A. Member is 1 year of age or older;
AND
 - B. Medication is prescribed in collaboration with a board certified cardiologist, geneticist, or neurologist
AND
 - C. Member has a diagnosis of late-onset Pompe disease based on GAA enzyme assay which shows reduced enzyme activity (2-40% partial deficiency of the lab specific normal mean value) confirmed with the ONE of the following:
 - a. DNA GAA mutation testing that includes the finding of two pathogenic variants in trans in the GAA gene;
OR
 - b. A second GAA enzyme activity assay in a separate sample;
OR
 - c. Muscle biopsy confirming late-onset Pompe disease
- AND**

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D. Member has documented baseline values for percent predicted forced vital capacity (FVC) and 6-minute walk test (6MWT);

AND

E. Member has not failed a previous course of alglucosidase alfa (Lumizyme) therapy;

AND

F. Medication is NOT prescribed in concomitant use of alglucosidase alfa

AND

G. Authorization is for 12 months

RENEWAL REQUEST:

1. Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency)

A. Initial conditions of coverage have been met;

AND

B. Member has no evidence of unacceptable toxicity or disease progression while on the current regimen (i.e. hypersensitivity reactions (e.g. anaphylaxis), severe infusion associated reactions, acute cardiorespiratory failure);

AND

C. Member demonstrates a positive response to therapy (i.e. improvement in FVC in the upright position from baseline, improvement in 6MWT, improvements in motor function, prevention of progression of disease state);

AND

D. Authorization is 12 months

V. LIMITATIONS/ EXCLUSIONS:

Nexviazyme will be considered experimental and investigational if prescribed for indications that have not been approved by the FDA and will not be covered under this policy.

VI. APPLICABLE PROCEDURE CODES:

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

VII. APPLICABLE DIAGNOSIS CODES:

CODE	Description
E74.02	Pompe disease

VIII. REFERENCES:

1. Nexviazyme (avalglucosidase alfa) [prescribing information]. Cambridge, MA: Genzyme Corporation; September 2023.

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2. US Food & Drug Administration (FDA): Nexviazyme. US Food & Drug Administration (FDA). Silver Spring, MD. 2022. Available from
URL: https://www.accessdata.fda.gov/drugsatfda_docs/label/2021/761194s000lbl.pdf.
(Accessed on November 20, 2025).
3. Kaplan SL. Lysosomal acid alpha-glucosidase deficiency (Pompe disease, glycogen storage disease II, acid maltase deficiency). In: UpToDate, Firth HV (Ed), Wolters Kluwer. (Accessed on November 20, 2025).
4. IPD Analytics. ICD-10-CM CODES.




Policy and Procedure

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REVISION LOG:

REVISIONS	INITIAL	DATE
Creation date	JL	12/16/2025

Approved:	Date:	Approved:	Date:
<i>Suzana Patel</i>	2/18/2026		02.18.2026
Suzana Patel, PharmD Senior Director of Pharmacy		Sanjiv Shah, MD Chief Medical Officer	



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Medical Guideline Disclaimer:

Property of MetroPlus HealthPlan. All rights reserved. The treating physician or primary care provider must submit MetroPlus Health Plan clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, Metroplus Health Plan will not be able to properly review the request for prior authorization. The clinical review criteria expressed in this policy reflects how MetroPlus Health Plan determines whether certain services or supplies are medically necessary. MetroPlus Health Plan 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). MetroPlus Health Plan 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 MetroPlus Health Plan, as some programs exclude coverage for services or supplies that MetroPlus Health Plan considers medically necessary. If there is a discrepancy between this guidelines and a member's benefits program, the benefits program will govern. 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 website links are accurate at time of publication.

MetroPlus HealthPlan has adopted the herein policy in providing management, administrative and other services to our members, related to health benefit plans offered by our organization.