Nexviazyme®
(avalglucosidase alfa-ngpt)

NEXVIAZYME® (avalglucosidase alfa-ngpt) BILLING & CODING GUIDE FOR REIMBURSEMENT

Please see Important Safety Information, including Boxed WARNING on page 3, and accompanying full Prescribing Information.

Using This Billing & Coding Guide

This document is intended as a general guide for submitting information to payers for reimbursement. Use of this guide does not guarantee that the payer will provide coverage for NEXVIAZYME and is not intended to be a substitute for, or an influence on, the independent medical judgment of the prescriber. Prescribers should follow payer-specific coding requirements and exercise clinical judgment when selecting codes and submitting claims to truthfully and accurately reflect the services and products furnished to a specific patient.

The coding information discussed in this guide:

- is provided for informational purposes only
- is subject to change
- should not be construed as legal advice

The codes listed herein may not apply to all patients or to all health plans. Conversely, additional codes not listed in this guide may apply to some patients.

Sanofi is committed to working with providers, as well as public and private payers, to help ensure access to NEXVIAZYME as indicated. If you still have questions after reviewing this guide, please contact CareConnectPSS® Services at 1-800-745-4447, Option 3. Our CareConnectPSS Case Managers have expertise in reimbursement, insurance, case management, and the health care delivery system, and can provide information to physicians and their patients about the reimbursement process.

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Indication and Important Safety Information

INDICATION

NEXVIAZYME (avalglucosidase alfa-ngpt) is indicated for the treatment of patients 1 year of age and older with late-onset Pompe disease [lysosomal acid alpha-glucosidase (GAA) deficiency].

IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Hypersensitivity Reactions Including Anaphylaxis
Patients treated with NEXVIAZYME have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during NEXVIAZYME administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, NEXVIAZYME should be discontinued immediately and appropriate medical treatment should be initiated. In patients with severe hypersensitivity reaction, a desensitization procedure to NEXVIAZYME may be considered.

Infusion-Associated Reactions (IARs)
Patients treated with NEXVIAZYME have experienced severe IARs. If severe IARs occur, consider immediate discontinuation of NEXVIAZYME, initiation of appropriate medical treatment, and the benefits and risks of readministering NEXVIAZYME following severe IARs. Patients with an acute underlying illness at the time of NEXVIAZYME infusion may be at greater risk for IARs. Patients with advanced Pompe disease may have compromised cardiac and respiratory function, which may predispose them to a higher risk of severe complications from IARs.

Risk of Acute Cardiorespiratory Failure in Susceptible Patients
Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during NEXVIAZYME infusion. More frequent monitoring of vitals should be performed during NEXVIAZYME infusion.

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: See Boxed WARNING. Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. The risks and benefits of readministering NEXVIAZYME following severe hypersensitivity reaction (including anaphylaxis) should be considered. If a mild or moderate hypersensitivity reaction occurs, the infusion rate may be slowed or temporarily stopped.

Infusion-Associated Reactions: See Boxed WARNING. IARs may still occur in patients after receiving pretreatment. If mild or moderate IARs occur regardless of pretreatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms.

Risk of Acute Cardiorespiratory Failure in Susceptible Patients: See Boxed WARNING.

ADVERSE REACTIONS

The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia and urticaria.

Please see full Prescribing Information for complete details, including Boxed WARNING.

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Coding Summary

Diagnosis

The ICD-10-CM diagnosis code for Pompe disease which should be used in conjunction with the administration of NEXVIAZYME is E74.02.[1]

ICD-10-CM Codes[1]	
E00-E89	Endocrine, nutritional and metabolic diseases
➤ E74	Other disorders of carbohydrate metabolism
➤ **E74.02**	**Pompe disease**

National Drug Code (NDC)

NEXVIAZYME has a 10-digit NDC code displayed on its packaging. In most cases, this should be converted to an 11-digit NDC code for billing purposes.[2] Payer requirements for NDC use and format may vary. Please contact each payer for specific coding policies. Below are both NDC codes for NEXVIAZYME.

10-Digit NDC	11-Digit NDC	How Supplied
58468-0426-1	58468-0426-01	Each single-dose vial contains 100 mg of a sterile, white to pale-yellow lyophilized powder for intravenous use after reconstitution and dilution. After reconstitution, the resultant concentration is 10 mg/mL.[3]

CPT® Code

CPT codes are used to describe the procedures performed on a patient and/or how a drug or supply being billed was administered.[4] The CPT codes most commonly associated with the administration of IV-infused biologic therapies like NEXVIAZYME are listed below. Confirm preferred coding policy with payer prior to administration whenever possible.

Primary Codes[5]	
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
96366	Each additional hour (list separately in addition to code for primary procedure; report 96366 for infusion intervals of greater than 30 minutes beyond 1-hour increments)*

*Per CMS guidelines, if the incremental amount of infusion time is 30 minutes or less, the time is not to be billed separately. Note that some payers may require reporting the actual number of minutes on the claim.

HCPCS Procedure Code

As part of the standardized Level II HCPCS coding system,[6] NEXVIAZYME has been issued a permanent J code as of April 1, 2022, which is listed below. Temporary codes, such as C9085, will no longer be valid after April 1, 2022. Note that the coding system is not a methodology for making coverage or payment determinations. The fact that a HCPCS code exists does not imply coverage, only that the product may be reimbursed if covered. Each payer makes determinations on coverage and payment outside of this coding process.[6]

Permanent J Code[7]		
Code #	**Short Description**	**Long Description**
J0219	Inj aval alpha-ngpt 4 mg	Injection, avalglucosidase alpha-ngpt, 4 mg

JW modifier: Medicare and some commercial payers require providers and suppliers to report the JW modifier on Part B drug claims for discarded drugs and biologicals.[8] Refer to each payer's policy for coding and documentation requirements.

Place of Service Codes

Because NEXVIAZYME can be administered in various settings (infusion center, physician office, patient's home if deemed clinically appropriate by the prescribing physician), it is important to populate a claim with the appropriate 2-digit place-of-service (POS) code.[9] Always verify the preferred POS codes for your patient's health plan before submitting a claim.

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CMS, Centers for Medicare & Medicaid Services; CPT®, Current Procedural Terminology; HCPCS, Healthcare Common Procedure Code System; ICD-10-CM, International Classification of Diseases, Tenth Revision, Clinical Modification; IV, intravenous.

Sample Reimbursement Forms

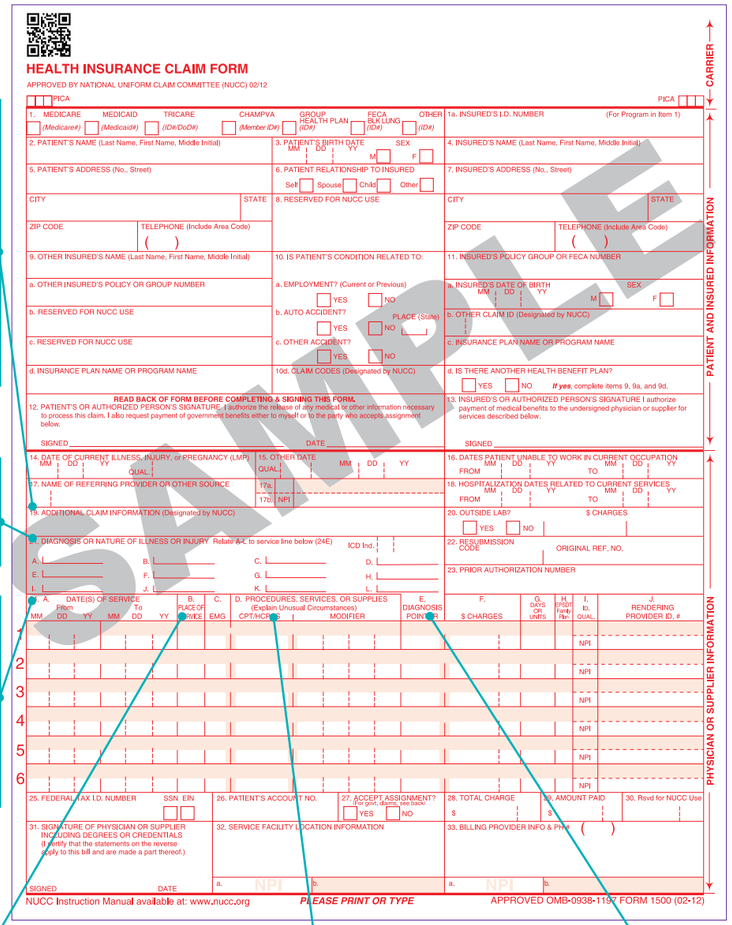
These sample claim forms are intended for use only as a reference. Reimbursement codes are subject to continual change. Please confirm the accuracy of the codes used with each payer.

Annotated claim form CMS-1500[10]

Field 19: Provide any required detailed information such as drug name, total dosage and strength, method of administration, 11-digit NDC, and basis of measurement (attach separately if needed)

Field 21: Enter the appropriate ICD-10-CM diagnosis codes

Field 24A: Enter the date of service for each procedure. Include NDC information, if required, in the shaded area above each date



Field 24B: Enter appropriate place of service code (office, infusion center, etc)

Field 24D: Include payer-required details such as HCPCS (J code), CPT codes and modifiers.

Field 24E: Enter the diagnosis code reference letter or number from Field 21 that relates to the date of service and the services or procedures performed that are entered on the same line under 24D

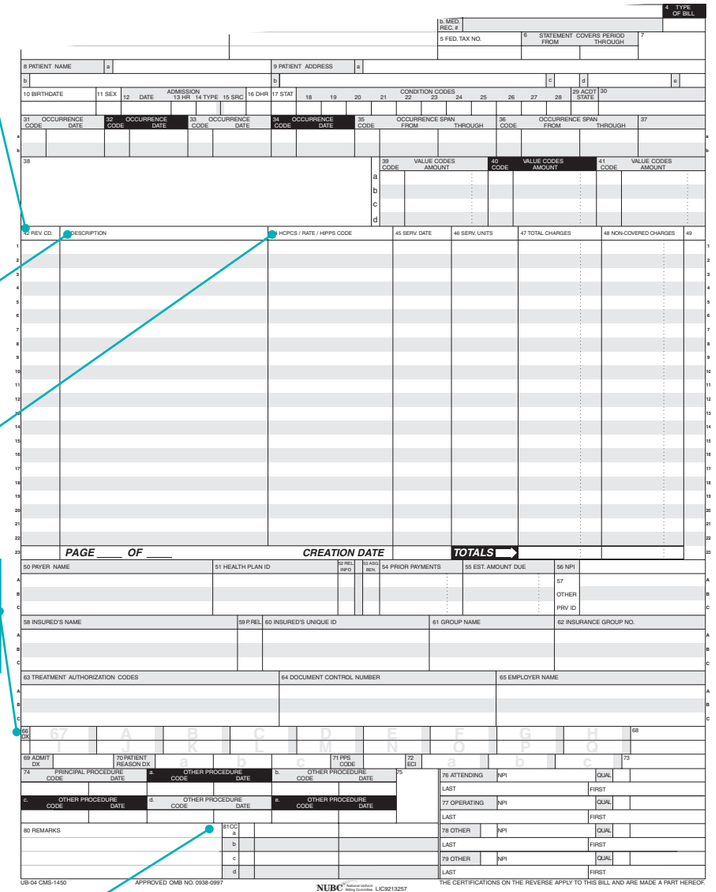
Annotated claim form CMS-1450[11]

This form is used for claims submitted by hospitals, nursing facilities, and other inpatient institutions. Although fields are organized differently than in the CMS 1500, the information captured is essentially the same.

Field 42: Enter the 4-digit revenue code that best describes the service provided, in accordance with the hospital billing policy

Field 43: Enter the corresponding description of service (eg, IV therapy)

Field 44: Include payer-required details such as relevant HCPCS and CPT codes



Field 66: Enter the appropriate ICD-10-CM diagnosis codes

Field 80: Provide any required detailed information such as drug name, total dosage and strength, method of administration, and 11-digit NDC (attach separately if needed)

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Nexviazyme® (avalglucosidase alfa-ngpt)

Dosing Information[3]

NEXVIAZYME is for intravenous infusion only. Recommended dosage for patients with LOPD is 20 mg/kg (of actual body weight) for patients weighing ≥30 kg, and 40 mg/kg (of actual body weight) for patients weighing <30 kg. The recommended starting infusion rate is 1 mg/kg/hour. The recommended infusion duration is between 4 and 7 hours. Physicians may pretreat with antihistamines, antipyretics, and/or corticosteroids prior to NEXVIAZYME administration.

Stepwise escalation of infusion rate

If there are no signs of infusion-associated reactions (IARs), and at the discretion of the health care provider, careful stepwise escalation of infusion rate every 30 minutes may be considered to safely reduce the overall infusion duration.

- For 20 mg/kg dosage: After starting at 1 mg/kg/hour, HCP may increase rate in 30-minute increments to 3 mg/kg/hour, 5 mg/kg/hour, and finally 7 mg/kg/hour. Once infusion rate of 7 mg/kg/hour is reached, maintain until the infusion is complete. The approximate total infusion duration is 4 hours to 5 hours
- For 40 mg/kg dosage: the same pattern as the 20 mg/kg dosing can be followed, resulting in an infusion duration of approximately 7 hours
  - There is an optional 5-step process for subsequent infusions that may be used to safely reduce the overall infusion duration
- **Please see the full Prescribing Information for complete details regarding infusion rates, as well as additional details regarding preparation and administration.**

Storage

Refrigerate vials of NEXVIAZYME at 36°F to 46°F (2°C to 8°C). Do not use NEXVIAZYME after the expiration date on the vial.[3]

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: See Boxed WARNING. Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. The risks and benefits of readministering NEXVIAZYME following severe hypersensitivity reaction (including anaphylaxis) should be considered. If a mild or moderate hypersensitivity reaction occurs, the infusion rate may be slowed or temporarily stopped.

LOPD, late-onset Pompe disease.

Additional Billing and Coding Considerations

Reimbursement considerations

NEXVIAZYME is designed to be prepared and administered by a health care provider, in both outpatient and patient home settings. The drug costs are expected to be covered under the Medicare Part B benefit.[12] Please refer to the individual patient's plan to determine any applicable coverage requirements. The specifics of coverage may vary by payer.

When filing a claim

It is recommended that NEXVIAZYME coverage be confirmed with all payers prior to patient administration, as patient benefits vary among payers and by plan.

Some payers also have policies that may affect coverage for NEXVIAZYME. These include:

- **Site of care:** Some payers may have coverage rules that restrict where patients can receive certain types of medical care like infusions
- **Network providers:** Some payers have exclusive contracts with in-network or participating providers to administer infusion therapies. These may include contracts for coverage in physician offices and outpatient settings or with specialty pharmacies that provide drugs and biologics to the provider
- **Prior authorization:** Many plans may require providers to obtain prior authorization (eg, medical necessity) to begin a course of treatment. Check with the payer to determine their process, requirements, and method for requesting authorization

Documenting necessity

As a new medication used to treat a rare disease, some insurers may not be familiar with NEXVIAZYME, and may require additional documentation to process either a prior authorization or a claim upon receipt. Examples of documentation that can be required include:

- Statement of medical necessity from the attending physician
- NEXVIAZYME Prescribing Information (available here)
- Details on the patient's case history, previous therapy, and clinical course

Example of a statement of medical necessity

STATEMENT OF MEDICAL NECESSITY
FOR THE TREATMENT OF LATE-ONSET POMPE DISEASE

Patient Information

Patient Name:	Address:
Date of Birth:	City: State: Zip:
Gender: ◉Male ◉Female	Phone No. (Home):
	Phone No. (Work):

Insurance Information

Insurance Co.:	Policy Holder Name:
Subscriber ID No.:	Insurance Phone No.:
Group No.:	

Medical Assessment

Patient Weight: _____(kg/lb) **Patient Height:** _____(cm/in)
Respiratory: **Musculoskeletal:**
Cardiac: **Other:**

Enclosures <include patient medical history, full Prescribing Information, additional supporting clinical documents>
***Attach copy of GAA enzyme assay or GAA Gene Sequencing**

Diagnosis

Pompe Disease E74.02: Date of Confirmed Diagnosis: _____

How was the diagnosis confirmed? Confirmation REQUIRES the presence of #1 OR #2 below.

1. ☐ **GAA Enzyme Activity (must be reduced or absent):** Value:_____(unit/s) Date:_____ Normal Reference Range:_____ for laboratory & sample
   Sample Type: ☐Blood ☐Purified Lymphocytes ☐Mixed Leukocytes ☐Muscle Tissue ☐Cultured Skin Fibroblasts
2. ☐ **GAA Gene Sequencing** Date:_____ List DNA sequence changes 1._____ 2._____
   Additional information (if needed):

Treatment Recommendation

NEXVIAZYME™ (avalglucosidase alfa-ngpt) **NDC #: 58468-0426-1 (carton of 1 single-use vial)**
Dose:_____**mg/kg** **Frequency:**_____
Therapy Start Date:_____

Physician Authorization

I certify that the above-indicated therapy is medically necessary, and the information provided is accurate to the best of my knowledge.

Physician Name (printed):_____Date:_____
Address:_____City:_____State:_____Zip:_____
Phone No._____
Physician Signature:_____
Physician's Medical License No._____**State Issued:**_____

Note that some payers have their own specific form for medical necessity, which should be used in those cases.

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Patient Support Services

Sanofi cares about patients in a variety of ways...

careconnect PSS®
Personalized Support Services

CareConnectPSS®

Personalized support services, designed to support each patient's unique journey. Support includes:

- Dedicated CareConnectPSS Case Managers and Patient Education Liaisons
- Disease-specific content and resources, including information about how rare diseases may run in families
- Information regarding genetic testing options and diagnostics
- Care coordination for treatment
- Help with handling insurance issues

CareConnectPSS Co-pay Program

Helps eligible patients in the US who are prescribed NEXVIAZYME pay for eligible out-of-pocket drug costs and specified infusion-related charges, including co-pays, coinsurance, and deductibles, up to the program maximum.*

CareConnectPSS Patient Assistance Program

Provides NEXVIAZYME at no cost to eligible patients who do not have health insurance or cannot access NEXVIAZYME under the terms of their insurance plan(s), until insurance coverage for NEXVIAZYME is secured.†

To find out more, contact a Case Manager at 1-800-745-4447 (Option 3) or visit www.CareConnectPSS.com

*Patients must be eligible under applicable state law(s). Patients whose medication or infusion-related costs are covered by a state or federal health care program, including but not limited to Medicare, Medicare Part D, Medigap, Medicaid, Veterans Affairs (VA), Department of Defense (DoD), or TRICARE, are not eligible. Patient must live in the US or a US territory. Other terms and conditions of the Program apply.

Co-Pay Program does not cover or provide support for MD office visits/evaluations, nursing services/observation periods, blood work, x-rays or other testing, pre-medications/other medications, epinephrine injection pens, transportation or other related services associated with treatment. In accordance with state law, infusion-related costs are not covered for commercially insured patients residing in MA or RI. Sanofi reserves the right to modify or discontinue the programs at any time. Savings may vary depending on patients' out-of-pocket costs. All program details provided upon registration.

†Patient Assistance Program and eligibility criteria include the following:

- Patient must not have insurance coverage or not have access to NEXVIAZYME under the terms of the patient's insurance plan(s)
- Patient must live in the US or a US territory
- Patient must have a valid prescription from a health care provider licensed in the US or a US territory
- Other terms and conditions of the Program apply

Ordering Information for NEXVIAZYME

To order NEXVIAZYME, contact one of the specialty distributors listed below.

Specialty Distributors	Phone	Web
Cardinal Health	800-926-3161	cardinalhealth.com
Cardinal Health Specialty Distribution	877-453-3972	specialtyonline.cardinalhealth.com
McKesson Corp	855-625-6285	connect.mckesson.com
McKesson Specialty (MSCD)	800-482-6700	oncology.mckessonspecialtyhealth.com
McKesson Plasma & Biologics (MPB)	877-625-2566	connect.mckesson.com
Morris & Dickson	800-388-3833	morrisdickson.com

NEXVIAZYME can be ordered directly from the manufacturer by contacting Sanofi.

Direct Order Contact	Phone	Email
Rare Disease Product Services	800-745-4447, Option 1	CO@Sanofi.com

NEXVIAZYME is also available through most specialty pharmacies.

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

Infusion-Associated Reactions: See Boxed WARNING. IARs may still occur in patients after receiving pretreatment. If mild or moderate IARs occur regardless of pretreatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms.

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The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia and urticaria.

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References

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