

Effective Date: 04/01/2022
Last Reviewed: 02/2022, 01/2023, 01/2024, 05/2024, 02/2025, 2/2026
Pharmacy Scope: Medicaid*
Medical Scope: Commercial, Medicare

## Nexviazyme (avalglucosidase alfa-ngpt) (Intravenous)

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**\*Effective 04/01/2022- Medication only available on the pharmacy benefit for MEDICAID members**

### **Policy Statement:**

Nexviazyme (avalglucosidase alfa-ngpt) is covered under the Pharmacy Benefit for Medicaid members and covered under the Medical Benefit for Commercial and MMP members when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

### **Procedure:**

Coverage of will be reviewed prospectively via the prior authorization process based on criteria below.

### ***Initial Criteria:***

- Patient is 1 year of age or older; AND
- Patient has documented diagnosis of late-onset Pompe disease (LOPD);
  - a. Diagnosis is evidenced by the following:
    - i. Enzyme assay showing a deficiency of acid alpha-glucosidase (GAA) activity in the blood, skin, or muscle
    - ii. Genetic testing showing a mutation in the GAA gene; AND
- Medication is not being used concurrently with Lumizyme or Pombiliti; AND
- Members weighing <30kg that require a dose of 40 mg/kg must have a documented failure, contraindication or intolerance to Lumizyme; AND
- Patient has measurable signs of Pompe disease (motor weakness, impaired pulmonary function); AND
- Patient has documented baseline percent-predicted forced vital capacity (FVC) and 6-minute walk test; AND
- Patient does not require invasive ventilation, is able to ambulate 40 meters without stopping and without assistive device, has a FVC of >30% but ≤85%, has not previously tried and failed Lumizyme; AND
- Nexviazyme is dosed according to the US Food and Drug Administration labeled dosing for LOPD

### ***Continuation of Therapy Criteria:***

- Patient continues to meet all initial criteria and is tolerating therapy with Nexviazyme; AND

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- Documentation of a positive clinical response to therapy as evidenced by an improvement or stabilization in percent-predicted FVC and/or 6MWT

**Coverage durations:**

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

**Dosage/Administration:**

Indication	Dose	Maximum dose(1 billable unit = 4mg)
LOPD	20mg/kg every 2 weeks <i>*for members weighing &lt;30kg dose of 40mg/kg may be required</i>	575 billable units (2300mg) every 14 days

**Investigational use:** All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

**Applicable Codes:**

Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section. The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J0219	Injection, avalglucosidase alfa-ngpt, 4 mg

**References:**

1. Nexviazyme (avalglucosidase alfa-ngpt) [prescribing information]. Genzyme Corporation. Cambridge, MA; September 2023. Accessed January 2025.