| Effective Date: 04/01/2022 |
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| Last Reviewed: 02/2022, 01/2023, 01/2024, 05/2024<br>Pharmacy Scope: Medicaid*<br>Medical Scope: Commercial, Medicare-Medicaid Plan (MMP) |

# Nexviazyme (avalglucosidase alfa-ngpt) (Intravenous)

***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.

**Summary of Evidence:**

Clinical trials evaluating the efficacy and safety of Nexviazyme in the treatment of Pompe disease have demonstrated significant improvements in respiratory function, motor function, and overall survival compared to placebo or standard of care. Nexviazyme is a recombinant human acid alpha-glucosidase enzyme replacement therapy that targets the underlying cause of Pompe disease by replacing the deficient enzyme, leading to reduced glycogen accumulation in affected tissues.

**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
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

**Continuation of Therapy Criteria:**

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

Per §§ 42 CFR 422.101, this clinical medical policy only applies to INTEGRITY in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD).

**Policy Rationale:**
Nexviazyme was reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use Nexviazyme according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For INTEGRITY (Medicare-Medicaid Plan) members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.

**Dosage/Administration:**

| **Indication** | **Dose** | **Maximum dose(1 billable unit = 4mg)** |
|---|---|---|
| LOPD | 20mg/kg every 2 weeks<br>**for members weighing <30kg dose of 40mg/kg may be required* | 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.