

Effective Date: 04/01/2022
Last Reviewed: 02/2022, 01/2023
Pharmacy Scope: Medicaid*
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 Medical Benefit 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; AND
- Members 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
- 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: 12 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable.***

Dosage/Administration:

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

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; August 2021.