Policy Title: **Nexviazyme (avalglucosidase alfa-ngpt)**  
(Intravenous)  

<table>
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<tr>
<th>Department:</th>
<th>PHA</th>
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**Effective Date:** 02/01/2022  
**Review Date:** 01/13/2022

**Purpose:** To support safe, effective and appropriate use of Nexviazyme (avalglucosidase alfa-ngpt).

**Scope:** Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

**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 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  
- 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 Nexviziazyme; 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:

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose</th>
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<tbody>
<tr>
<td>LOPD</td>
<td>20mg/kg every 2 weeks *for members weighing &lt;30kg dose of 40mg/kg may be required</td>
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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:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>C9399</td>
<td>Unclassified drugs or biologicals</td>
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References: