Nexviazyme™ (avalglucosidase alfa-ngpt) Injection

Humana

Pharmacy Coverage Policy

Effective Date: October 20, 2021
Revision Date: September 28, 2022
Review Date: September 21, 2022
Line of Business: Medicare, Commercial, Medicaid - Humana, Medicaid - Ohio
Policy Type: Prior Authorization

Disclaimer
Description
Coverage Determination
Background
Medical Terms
References

Disclaimer

State and federal law, as well as contract language, including definitions and specific inclusions/exclusions, take precedence over clinical policy and must be considered first in determining eligibility for coverage. Coverage may also differ for our Medicare and/or Medicaid members based on any applicable Centers for Medicare & Medicaid Services (CMS) coverage statements including National Coverage Determinations (NCD), Local Medical Review Policies (LMRP) and/or Local Coverage Determinations. See the CMS website at http://www.cms.hhs.gov/. The member's health plan benefits in effect on the date services are rendered must be used. Clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Clinical technology is constantly evolving, and we reserve the right to review and update this policy periodically.

Description

Nexviazyme (avalglucosidase-alfa) is an enzyme replacement therapy.

Avalglucosidase alfa is the lysosomal enzyme acid alpha-glucosidase (GAA). The GAA enzyme is required for lysosomal glycogen degradation. Once it bings to the mannose 6-phosphate receptor, it is transported into the lysosome where it can ultimately cleave the glycogen inside. This results in a decrease accumulation of the lysosomal glycogen in the skeletal and hear muscles, potentially resulting in decrease in muscular wekanes and/or respiratory and heart failure.

Nexviazyme (avalglucosidase-alfa) is indicated for the treatment of patients aged 1 year and older with late-onset Pompe disease (GAA deficiency)

Avalglucosidase alfa is available as brand Nexviazyme in a 100 mg single use vial

Coverage Determination

Please note the following regarding medically accepted indications:

All reasonable efforts have been made to ensure consideration of medically accepted indications in this policy. Medically accepted indications are defined by CMS as those

uses of a covered Part D drug that are approved under the federal Food, Drug and Cosmetic Act, or the use of which is supported by one or more citations included or approved for inclusion in any of the compendia described in section 1927(g)(1)(B)(i) of the Act. These compendia guide review of off-label and off-evidence prescribing and are subject to minimum evidence standards for each compendium. Currently, this review includes the following references when applicable and may be subject to change per CMS:

- American Hospital Formulary Service-Drug Information (AHFS-DI)
- National Comprehensive Cancer Network (NCCN) Drugs and Biologics Compendium
- Truven Health Analytics Micromedex DrugDEX
- Elsevier/Gold Standard Clinical Pharmacology
- Wolters Kluwer Lexi-Drugs

Nexviazyme (avalglucosidase-alfa) will require prior authorization. This agent may be considered medically necessary when the following criteria are met:

Late-Onset Pompe Disease

- The member has a diagnosis of late-onset Pompe Disease **AND**
  - The diagnosis is supported by submission of lab and/or genetic testing consistent with late-onset Pompe disease (e.g. evidence of GAA enzyme deficiency, identification of pathogenic variants in GAA gene) **AND**
- Symptom onset was evident at 1 year of age or older **AND**
- The member is under the care by, or in consultation with, a specialist experienced in the management of late-onset Pompe disease **AND**
- Documentation of baseline functional test results (e.g. FVC, 6 minute-walk test)

Reauthoriziation

- Attestation of improvement from pre-treatment baseline (e.g. improvement in functional status, 6MWT, FVC) or from natural progression of disease

Nexviazyme (avalglucosidase-alfa) will be approved in plan year durations or as determined through clinical review.

Coverage Limitations

Nexviazyme (avalglucosidase-alfa) therapy is not considered medically necessary for members with the following concomitant conditions:

- Experimental/Investigational Use – Indications not supported by CMS recognized compendia or acceptable peer reviewed literature.

Background

This is a prior authorization policy about Nexviazyme (avalglucosidase-alfa).

Pompe is a type of glycogen storage disease, meaning patients have an excessive build-up of a complex sugar molecule known as glycogen within their cells, especially in muscle cells. This is because of a defect in the GAA gene that causes low levels of the acid alpha-glucosidase enzyme to be produced, an enzyme that is crucial in breaking glycogen down into glucose. Different types of mutations in the GAA gene can affect how much of a functional acid alpha-glucosidase enzyme exists in cells. The type and severity of Pompe disease depends on the levels of the working acid alpha-glucosidase enzyme. Symptom onset in late-onset Pompe disease can occur anytime after the age of 1 year. People with late-onset Pompe have higher GAA enzyme levels than are found in the infantile-onset forms of this disease, but generally less than 40 percent of “normal” levels. In comparison, infantile-onset Pompe patients typically have less than 1 percent of GAA enzyme activity, while those with non-classic forms usually have less than 10 percent. The actual frequency of late-onset Pompe disease in the U.S. is unknown but it is estimated to be as high as 1 in 40,000.

Black Box Warnings:

- Hypersensitivity reactions, including anaphylaxis: appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available. If a severe
  hypersensitivity reaction occurs, therapy should be discontinued immediately and appropriate medical treatment should be initiated
- Infusion-associated reactions (IARs): If severe IARs occur, consider immediate discontinuation and initiation of appropriate medical treatment
- 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, may be at risk of serious exacerbation of their cardiac or respiratory status during the infusion

Provider Claims Codes

For medically billed requests, please visit www.humana.com/PAL. Select applicable Preauthorization and Notification List(s) for medical and procedural coding information.

Medical Terms

Nexviazyme; avalglucosidase alfa-ngpt; Pharmacy; Late-onset Pompe disease; Orphan; intravenous

References

1. Barba-romera, MA, Barrot E, Bautista-lorite J, et al. Clinical guidelines for late-onset Pompe disease. *Rev Neurol* 2012; 54 (8): 497-507
2. Clinical Pharmacology [database online]. Tampa, Fl; Gold Standard, Inc. Updated periodically. https://www.clinicalkey.com/pharmacology
3. Culper EJ, Berger KI, Leshner RT, et al. Consensus treatment recommendations for late-onset Pompe disease. *Muscle Nerve 2021:* 45: 319–333
4. DRUGDEX® System [Internet database]. Greenwood Village, Colo: Thompson Reuters (Healthcare) Inc. Updated periodically.
5. Nexviazyme [prescribing information]. Genzyme Corporation. August 2021
6. Pompe Disease: Rare Disease Database; NORD: National Organization for Rare Disorders. 2020. https://rarediseases.org/rare-diseases/pompe-disease