Nexviazyme™ (avalglucosidase alfa-ngpt) Injection

Humana

Pharmacy Coverage Policy

Effective Date: October 20, 2021
Revision Date: September 27, 2023
Review Date: September 20, 2023
Line of Business: Medicare, Commercial, Medicaid - South Carolina, Medicaid - Ohio
Policy Type: Prior Authorization

Products Affected

Nexviazyme intravenous solution

Listed Indications

Late-Onset Pompe Disease

Late-Onset Pompe Disease

Does the member meet all of the following criteria?	
Criteria #1	The member has a diagnosis of late-onset Pompe Disease
Criteria #2	The diagnosis is supported by 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)
Approval Duration	
Initial	plan year duration

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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

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

Provider Claim 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

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