RX.PA.062.MPC Nexviazyme ® (avalglucosidase alfa)

The purpose of this policy is to define the prior authorization process for **Nexviazyme ® (avalglucosidase alfa).**

Nexviazyme ® (avalglucosidase alfa) is indicated for the treatment of Pompe disease, late onset.

PROCEDURE

1. Pompe Disease

A. <u>Initial Authorization Criteria:</u>

Must meet all of the criteria listed below:

- Must be age 1 year or older
- Must be prescribed by or in consultation with a neurologist or a physician who specializes in the treatment of inherited metabolic disorders
- Must have a diagnosis of alpha glucosidase deficiency (Pompe disease) confirmed through GAA enzyme assay (from blood, skin fibroblasts, lymphocytes, or muscle) and/or identification of GAA gene mutation
- Member must have clinical signs and symptoms of Pompe disease such as cardiac hypertrophy, respiratory distress, skeletal muscle weakness, etc.
- Nexviazyme must not be used in combination with other enzyme replacement therapies (i.e. alglucosidase-alfa)
- Member must not be susceptible to fluid volume overload, have acute underlying respiratory illness, or have compromised cardiac or respiratory function

B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.

C. Nexviazyme will be considered investigational or experimental for any other use and will not be covered.

D. Reauthorization Criteria:

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.

MPC Renewal:

- Must be prescribed by or in consultation with a neurologist or a physician who specializes in the treatment of inherited metabolic disorders
- Must have chart documentation from the provider that the member's condition has stabilized or improved based upon the prescriber's assessment while on therapy (i.e stabilization or improvement of motor function, walking capacity, cardiac/respiratory function, muscle strength)
- Member is monitored for antibody formation/neutralizing antibodies

Renewal from Previous Insurer:

- Members who have received prior approval (from insurer other than MPC) and have been taking Nexviazyme have been receiving medication samples, should be considered under criterion A (Initial Authorization Criteria).; AND
- Provider has a documented clinical response of the member's condition which has stabilized or improved based upon the prescriber's assessment.

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	3 months
Reauthorization	1 year

HCPCS Code(s):

Code	Description
J0219	Injection, avalglucosidase alfa-ngpt, 4 mg

REFERENCES

1. Nexviazyme (avalglucosidase alfa) [prescribing information]. Cambridge, MA: Genzyme Corporation; August 2021
2. van der Ploeg AT, Kruijshaar ME, Toscano A, et al; European Pompe Consortium. European consensus for starting and stopping enzyme replacement therapy in adult patients with Pompe disease: a 10-year experience. Eur J Neurol. 2017;24(6):768-e31. doi:10.1111/ene.13285

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review	*02/2026*
Annual Review	*02/2025*
Annual policy review. Update to reauthorization criteria for non-MPC renewals	*02/2024*
Annual Review	*02/2023*
New Policy	*10/2022*