



Commercial/Healthcare Exchange PA Criteria Effective: December 9th 2021

Prior Authorization: Nexviazyme®

Products Affected: Nexviazyme® (avalglucosidase alfa-ngpt) intravenous infusion

Medication Description: Nexviazyme, a hydrolytic lysosomal glycogen-specific recombinant human α -glucosidase enzyme, is indicated for patients \geq 1 year of age with late-onset Pompe disease (lysosomal acid α -glucosidase deficiency).

Covered Uses: Treatment of Pompe disease, late onset (acid α -glucosidase deficiency).

Exclusion Criteria: N/A

Required Medical Information: Diagnosis

Prescriber Restriction: The medication is prescribed by or in consultation with a geneticist, neurologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Age Restriction: 1 year and older

Coverage Duration: 12 Months

Other Criteria:

I. INITIAL: Acid Alpha-Glucosidase Deficiency (Pompe Disease):

Approve if the patient meets all the following criteria (A and B):

- A.** Patient has late-onset acid alpha-glucosidase deficiency (late-onset Pompe disease); **AND**
- B.** The diagnosis is established by one of the following (i or ii):
 - i.** Patient has a laboratory test demonstrating deficient acid alpha-glucosidase activity in blood, fibroblasts, or muscle tissue, **OR**
 - ii.** Patient has a molecular genetic test demonstrating acid alpha-glucosidase gene mutation

II. CONTINUATION:

- A.** Patient has experienced a clinical response as determined by the prescribing physician; **AND**
- B.** Patient has not experienced unacceptable toxicity from the drug; **AND**
- C.** The medication is prescribed by or in consultation with a geneticist, neurologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

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Confidential Information

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References:

1. Nexviazyme (avalglucosidase alfa) [prescribing information]. Cambridge, MA: Genzyme Corporation; December 2021.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	12/9/2021

December 2021



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