PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Nexviazyme (avalglucosidase alfa-ngp)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Nexviazyme is an enzyme replacement therapy for the treatment of late-onset Pompe disease, also known as acid alpha-glucosidase (GAA) deficiency or glycogen storage disease type II. Pompe disease is a rare, genetic lysosomal storage disorder that results in the buildup of glycogen in cell lysosomes causing serious and life-threatening muscle damage and weakness. Nexviazyme provides an exogenous source of the deficient GAA enzyme to cleave glycogen and reduce its accumulation. In the COMET trial, Nexviazyme was found to be non-inferior to Lumizyme.
Pompe disease can be broadly classified as infantile-onset within the first few months of life (IOPD) or late-onset beyond infancy (LOPD). Classic IOPD is rapidly progressive with severe cardiomyopathy. Non-classic IOPD progresses slower with less severe cardiomyopathy. LOPD does not typically present with cardiomyopathy and has more variable symptoms, especially skeletal muscle weakness. Nexviazyme is only indicated to treat late-onset Pompe disease.

Nexviazyme (avalglucosidase alfa-ngp) will be considered for coverage when the following criteria are met:

Pompe disease (acid α-glucosidase [GAA] deficiency)

For **initial** authorization:
1. Member is at least 1 year of age; AND
2. Medication must be prescribed by or in consultation with a geneticist, neurologist, pulmonologist, or metabolic specialist; AND
3. Member has a diagnosis of late onset Pompe disease confirmed by an enzyme activity assay showing GAA deficiency (2% to 40% of normal); AND
4. Molecular genetic testing shows pathogenic mutation of the GAA gene; AND
5. Member must show signs or symptoms (i.e., motor weakness, reduced respiratory parameters).
6. **Dosage allowed/Quantity limit:**
   Actual body weight 30 kg or greater: 20 mg/kg IV infusion every 2 weeks
   Actual body weight less than 30 kg: 40mg/kg IV infusion every 2 weeks

If all the above requirements are met, the medication will be approved for 12 months.

For **reauthorization**:
1. Chart notes must document positive clinical response such as improved or stabilized respiratory muscle strength (i.e., forced vital capacity (FVC)) or functional endurance (e.g., 6-minute walk test).

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Nexviazyme (avalglucosidase alfa-ngp) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
08/20/2021	New policy for Nexviazyme created.
05/24/2022	Updated J code.
11/22/2023	Annual review; updated reference.

References:

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3. Diaz-Manera J, Kishnani PS, Kushlaf H, et al. Safety and efficacy of avalglucosidase alfa versus alglucosidase alfa in patients with late-onset Pompe disease (COMET): a phase 3, randomised, multicentre trial [published correction appears in Lancet Neurol. 2022 Apr;21(4):e4]. *Lancet Neurol*. 2021;20(12):1012-1026. doi:10.1016/S1474-4422(21)00241-6
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6. Kronn DF, Day-Salvatore D, Hwu WL, et al. Management of Confirmed Newborn-Screened Patients With Pompe Disease Across the Disease Spectrum. *Pediatrics*. 2017;140(Suppl 1):S24-S45. doi:10.1542/peds.2016-0280E
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8. Wang RY, Bodamer OA, Watson MS, Wilcox WR; ACMG Work Group on Diagnostic Confirmation of Lysosomal Storage Diseases. Lysosomal storage diseases: diagnostic confirmation and management of presymptomatic individuals. *Genet Med*. 2011;13(5):457-484. doi:10.1097/GIM.0b013e318211a7e1
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