

Nexviazyme® (avalglucosidase alfa-ngpt)

When requesting Nexviazyme® (avalglucosidase alfa-ngpt), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indications.

FDA-Approved Indication

Nexviazyme (avalglucosidase alfa-ngpt) is indicated for the treatment of individuals with late-onset Pompe disease (lysosomal acid α -glucosidase deficiency).

Coverage Guidelines

Late-Onset Pompe Disease

The individual must meet all of the following criteria for approval:

- Patient is 1 year of age or older;
- Diagnosis is confirmed by one of the following:
 - Laboratory test demonstrating deficient acid α -glucosidase activity in blood, fibroblasts, or muscle tissue; OR
 - Molecular genetic test demonstrating biallelic pathogenic or likely pathogenic acid alpha-glucosidase gene variants;
- Nexviazyme is prescribed by or in consultation with a geneticist, a neurologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Approval duration: 12 months

Dosing Recommendation

Nexviazyme is administered as an intravenous infusion. For patients weighing:

- ≥ 30 kg, the recommended dosage is 20 mg/kg every two weeks
- < 30 kg, the recommended dosage is 40 mg/kg every two weeks

References

1. Nexviazyme® intravenous infusion [prescribing information]. Cambridge, MA: Genzyme; September 2023.
2. Chien YH, Hwu WL, Lee NC. Pompe disease: Early diagnosis and early treatment make a difference. *Pediatr Neonatol*. 2013;54:219-227.

3. Llerena Junior JC, Nascimento OJM, Oliveira ASB, et al. Guidelines for the diagnosis, treatment and clinical monitoring of patients with juvenile and adult Pompe disease. *Arq Neuropsiquiatr*. 2016;74:166-176.
4. Cupler EJ, Berger KI, Leshner RT, et al. Consensus treatment recommendations for late-onset Pompe disease. *Muscle Nerve*. 2012;45:319-333.