

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 acid α -glucosidase gene mutation
- 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

Approve up to 40 mg/kg administered intravenously not more frequently than once every 2 weeks.

References

1. Nexviazyme® intravenous infusion [prescribing information]. Cambridge, MA: Genzyme; August 2021.
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.