# alglucosidase alfa (LUMIZYME®) J0221 (10mg)
# alglucosidase alfa (alglucosidase alfa) J0220 (10mg)
# avalglucosidase alfa-ngpt (NEXVIAZYME™) J0219 (4mg)

## Covered with prior authorization

**alglucosidase alfa (LUMIZYME® or unbranded if available) may be authorized when the following criteria are met:**

- Individual has a diagnosis of infantile-onset Pompe disease, acid alpha-glucosidase deficiency (GAA), confirmed by documented genetic testing (such as activity in skin fibroblasts of less than 1% of the normal mean or by GAA gene sequencing);
  **OR**
- Individual has a diagnosis of non-infantile onset (late-onset) Pompe disease, acid alpha-glucosidase deficiency (GAA), confirmed by documented genetic testing (such as activity in skin fibroblasts of less than 1% of the normal mean or by GAA gene sequencing); **AND**
- Attestation from the individual and/or guardian indicating adherence to the treatment plan is provided; **AND**
- Provider is a clinical specialist knowledgeable in treatment of Pompe disease, metabolic and neuromuscular disease, and immune tolerance induction. Individuals with Pompe disease require a multidisciplinary healthcare team that is expected to include a neurologist, cardiologist, pulmonologist, nutritionist, orthopedist, gastroenterologist, psychologist, and/or genetic counselor; **AND**
- Indication that adequate precautions are in place for addressing boxed warnings related is provided:
  - Risk Of Anaphylaxis, Hypersensitivity And Immunemediated Reactions; **AND**
  - Risk Of Cardiorespiratory Failure; **AND**
- Actual body weight is provided with the request; **AND**
- Dose is 20 mg per kg body weight every two weeks

**avalglucosidase alfa-ngpt (NEXVIAZYME™) may be authorized when the following criteria are met:**

- Individual has a diagnosis of non-infantile onset (late-onset) Pompe disease, acid alpha-glucosidase deficiency (GAA), confirmed by documented genetic testing (such as activity in skin fibroblasts of less than 1% of the normal mean or by GAA gene sequencing); **AND**

- Individual age is ≥ 1 year; **AND**
- Attestation from the individual and/or guardian indicating adherence to the treatment plan is provided; **AND**
- Provider is a clinical specialist knowledgeable in treatment of Pompe disease, metabolic and neuromuscular disease, and immune tolerance induction. Individuals with Pompe disease require a multidisciplinary healthcare team that is expected to include a neurologist, cardiologist, pulmonologist, nutritionist, orthopedist, gastroenterologist, psychologist, and/or genetic counselor; **AND**
- Indication that adequate precautions are in place for addressing boxed warnings related is provided:
    - Risk Of Anaphylaxis, Hypersensitivity And Immunemediated Reactions; **AND**
    - Risk Of Cardiorespiratory Failure; **AND**
- Actual body weight is provided with the request; **AND**
- The dose is appropriate for actual body weight:
    - 40 mg/kg every two weeks for individuals <30 kg
    - 20 mg/kg every two weeks for individuals ≥30 kg

**Exclusion criteria:**

Requests may not be approved for the following:

- Co-therapy utilizing both agents [Lumizyme (alglucosidase alfa) in combination with Nexviazyme (avalglucosidase alfa)]
- Product use for non-FDA approved indications or indications not supported by industry-accepted guidelines;
- Doses, durations, or dosing intervals that exceed FDA maximum limits for any FDA-approved indication or are not supported by industry-accepted practice guidelines or peer-reviewed literature for the relevant off-label use;
- Individuals with significant known risk factors unless the record provides an assessment of clinical benefit that outweighs the risk;

**Step/Alternative Therapies:**

Enzyme replacement therapy (ERT) of these two agents is an approved treatment for individuals with Pompe disease. Additional supportive therapies are also typically required.

**Initial authorization is up to 12 months.**

**Annual reauthorizations will require medical chart documentation that the patient has been seen within the past 12 months and that markers of disease are improved by therapy.**

**Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.**

**U.S. Food and Drug Administration:**

This section is to be used for informational purposes. FDA approval alone is not a basis for coverage.

- alglucosidase alfa (LUMIZYME®) is a hydrolytic lysosomal glycogen-specific enzyme indicated for individuals with Pompe disease (GAA deficiency). Note: product is marketed as Myozyme® outside the United States
- alglucosidase alfa (alglucosidase alfa) (no products currently marketed but HCPC code exists)
- avalglucosidase alfa-ngpt (NEXVIAZYME™ ) is a hydrolytic lysosomal glycogen-specific enzyme indicated for the treatment of individuals 1 year of age and older with late-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency)

**References**:

*Alglucosidase Alfa (Lexi-Drugs)*. (2022, April 19). Lexicomp. Retrieved June 19, 2022, from https://online.lexi.com/lco/action/doc/retrieve/docid/patch_f/511903?cesid=4KjKDJb6fZP&searchUrl=%2Flco%2Faction%2Fsearch%3Fq%3Dlumizyme%26t%3Dname%26acs%3Dtrue%26acq%3Dlumi
*Avalglucosidase Alfa (Lexi-Drugs)*. (2022, April 19). Lexicomp logo. Retrieved June 19, 2022, from https://online.lexi.com/lco/action/doc/retrieve/docid/patch_f/7130191?cesid=09UPTov0xPH&searchUrl=%2Flco%2Faction%2Fsearch%3Fq%3Dnexviazyme%26t%3Dname%26acs%3Dtrue%26acq%3DNexv#dop
*LUMIZYME® (alglucosidase alfa)*. (2020, February). Accessdata.fda.gov. Retrieved June 19, 2022, from https://www.accessdata.fda.gov/drugsatfda_docs/label/2020/125291s151lbl.pdf
*LUMIZYME® (alglucosidase alfa)*. (2022, May). Sanofi US. Retrieved June 19, 2022, from https://products.sanofi.us/lumizyme/lumizyme.pdf
*NEXVIAZYME™ (avalglucosidase alfa-ngpt)*. (2021, August). Sanofi US. Retrieved June 19, 2022, from https://products.sanofi.us/nexviazyme/nexviazyme.pdf
*Pompe Disease - NORD*. (2020). National Organization for Rare Disorders. Retrieved June 19, 2022, from https://rarediseases.org/rare-diseases/pompe-disease/

**Criteria History/ Revision Information**:

| Date | Summary of Changes |
|---|---|
| June 2022 | Criteria for use summary developed by Ascension Medical Specialty Prior Authorization Team |
| July 2022 | Criteria for use summary approved by Ascension Therapeutic Affinity Group |

If you have questions, call 833-980-2352 to speak to a member of the Ascension Rx prior authorization team or email your questions to smarthealthspecialty@ascension.org.