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# 5.30.034

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| **Section:** | Prescription Drugs | **Effective Date:** | April 1, 2024 |
| **Subsection:** | Endocrine and Metabolic Agents | **Original Policy Date:** | July 1, 2010 |
| **Subject:** | Lumizyme | **Page:** | 1 of 4 |

***Last Review Date:*** March 8, 2024

# Lumizyme

## Description

### Lumizyme (alglucosidase alfa)

**Background**
Lumizyme is indicated for Pompe disease, a rare genetic disorder. In Pompe disease, a gene mutation prevents the body from making an enzyme or making enough of the enzyme called acid alpha-glucosidase (GAA), necessary for proper muscle functioning. GAA is used by the heart and muscle cells to convert a form of sugar called glycogen into energy. Without the enzyme action, glycogen builds up in the cells and, ultimately, weakens the heart and muscles. Lumizyme replaces the deficient GAA, thereby reducing the accumulated glycogen in heart and skeletal muscle cells (1).

**Regulatory Status**
FDA-approved indication: Lumizyme (alglucosidase alfa) is a hydrolytic lysosomal glycogen-specific enzyme indicated for patients with Pompe disease (acid α-glucosidase (GAA) deficiency) (1).

Acute cardiorespiratory failure has been observed in a few infantile-onset Pompe disease patients with underlying cardiac hypertrophy, possibly associated with fluid overload with intravenous administration of alglucosidase alfa (1).

Lumizyme has a boxed warning that anaphylactic, severe allergic and immune mediated reactions have been observed during administration and up to 3 hours after. Patients with acute underlying respiratory illness or compromised cardiac and/or respiratory function may be at risk

of serious exacerbation of their cardiac or respiratory compromise during infusions. Appropriate medical support should be available during infusion (1).

Patients should be monitored for IgG antibody formation every 3 months for 2 years and then annually thereafter. Testing for IgG titers may also be considered if patients develop allergic or other immune mediated reactions. Patients who experience anaphylactic or allergic reactions may also be tested for IgE antibodies to alglucosidase alfa and other mediators of anaphylaxis. Patients who develop IgE antibodies to alglucosidase alfa appear to be at a higher risk for the occurrence of anaphylaxis and severe allergic reactions. Therefore, these patients should be monitored more closely during administration of Lumizyme (1).

The safety and effectiveness of alglucosidase alfa have been established in pediatric patients with Pompe disease (1).

**Related policies**

Nexviazyme

Policy

*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.*

Lumizyme may be considered **medically necessary** if the conditions indicated below are met.

Lumizyme may be considered **investigational** for all other indications.

## Prior-Approval Requirements

**Diagnosis**

Patient must have the following:

Pompe disease (acid alpha-glucosidase (GAA) deficiency)

**AND** the following:

1. Monitored for IgG antibody formation every 3 months for 2 years and then annually thereafter

## Prior – Approval *Renewal* Requirements

**Diagnosis**

Patient must have the following:

Pompe disease (acid alpha-glucosidase (GAA) deficiency)

**AND** the following:

1. Monitored for IgG antibody formation every year

## Policy Guidelines

### Pre - PA Allowance

None

### Prior - Approval Limits

**Duration** 2 years

### Prior – Approval *Renewal* Limits

Same as above

## Rationale

**Summary**

Lumizyme (alglucosidase alfa) is a lysosomal glycogen-specific enzyme indicated for patients with Pompe disease (acid α-glucosidase (GAA) deficiency). Lumizyme has a boxed warning that anaphylactic, severe allergic and immune mediated reactions have been observed. Patients with acute underlying respiratory illness or compromised cardiac and/or respiratory function may be at risk of serious exacerbation of their cardiac or respiratory compromise during infusions. Appropriate medical support should be available during infusion. Patients should be monitored for IgG antibody formation every 3 months for 2 years and then annually thereafter (1).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Lumizyme while maintaining optimal therapeutic outcomes.

**References**

1. Lumizyme [package Insert]. Cambridge, MA: Genzyme Corporation; March 2023.

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## Policy History

| Date | Action |
|---|---|
| September 2011 | New Policy |
| September 2012 | Annual editorial review and reference update. |
| June 2013 | Annual editorial review and reference update |
| August 2014 | Removal of age limit and the Lumizyme ACE Program and the revision of the diagnosis to Pompe disease to align with the new package insert. |
| September 2014 | Annual editorial review and reference update |
| September 2015 | Annual editorial review |
| September 2016 | Annual editorial review and reference update<br>Policy number change from 5.08.15 to 5.30.34 |
| December 2017 | Annual review |
| September 2018 | Addition of no history of cardiomyopathy to requirements |
| November 2018 | Annual review |
| December 2019 | Annual editorial review and reference update. Changed approval duration from lifetime to 2 years |
| December 2020 | Annual review and reference update |
| June 2021 | Annual review |
| December 2021 | Annual review |
| June 2022 | Annual review |
| March 2023 | Annual review and reference update. Changed policy number to 5.30.034. Per SME, removed initiation requirement of no cardiac hypertrophy or cardiomyopathy |
| March 2024 | Annual review and reference update |

## Keywords

**This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 8, 2024 and is effective on April 1, 2024.**