Medical Policy:

Lumizyme (alglucosidase) Intravenous

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.303	March 19, 2025	December 2019

Definitions

Lumizyme, a human hydrolytic lysosomal glycogen-specific enzyme (acid α-glucosidase), is indicated for patients with Pompe disease (acid α-glucosidase deficiency). It is produced in a Chinese hamster ovary cell line via recombinant DNA technology. After administration of Lumizyme, it is internalized into cells and transported to lysosomes where it catalyzes the breakdown of glycogen to glucose.

Length of Authorization

Coverage will be provided for 12 months and may be renewed

Dosing Limits [Medical Benefit]

Each dose must not exceed 20 mg/kg administered intravenously no more frequently than once every 2 weeks

Max Units (per dose and over time) [HCPCS Unit]:

230 billable units every 14 days

Guideline

I. INITIAL CRITERIA

1. **Acid Alpha-Glucosidase Deficiency (Pompe Disease).** Approve if the patient meets the following criteria:
    A. The diagnosis is established by one of the following (i or ii):
        i. Patient has a laboratory test demonstrating deficient acid alpha-glucosidase activity in blood, fibroblasts, or muscle tissue; **OR**
        ii. Patient has a molecular genetic test demonstrating biallelic pathogenic or likely pathogenic acid alpha-glucosidase (GAA) gene variants**; AND**
    B. Will not be used in combination with other enzyme replacement therapies (i.e., avalglucosidase alfa, cipaglucosidase alfa, etc.); **AND**
    C. Documented baseline age-appropriate values for **ONE** or more of the following:
        i. Infantile-onset disease: muscle weakness, motor function, respiratory function, cardiac involvement, percent predicted forced vital capacity (FVC), and/or 6-minute walk test (6-MWT); **OR**
        ii. Late-onset (non-infantile) disease: FVC and/or 6-MWT; **AND**
    D. Lumizyme is prescribed by or in consultation with a geneticist, neurologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

II. RENEWAL CRITERIA

Coverage may be renewed based on the following criteria:

1. Patient continues to meet Initial Criteria; **AND**
2. Absence of unacceptable toxicity from the drug. *Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, immune-mediated cutaneous reactions, systemic immune-mediated reactions, acute cardiorespiratory failure, cardiac arrhythmia during general anesthesia, etc.*; **AND**
3. Patient is being monitored for antibody formation (including neutralizing antibodies); **AND**
4. Patient has demonstrated a beneficial response to therapy compared to pretreatment age-appropriate baseline values in **ONE** or more of the following:
    i. Infantile-onset disease: stabilization or improvement in muscle weakness, motor function, respiratory function, cardiac involvement, FVC and/or 6-MWT; **OR**
    ii. Late-onset (non-infantile) disease: stabilization or improvement in FVC and/or 6-MWT

Applicable Procedure Codes

Code	Description
J0221	Injection, alglucosidase alfa, (Lumizyme), 10 mg

Applicable NDCs

Code	Description
58468-0160-02	Lumizyme 50mg Solution Reconstituted
58468-0160-01	Lumizyme 50mg Solution Reconstituted

ICD-10 Diagnoses

Code	Description
E74.02	Pompe Disease

Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	03/19/2025	Annual Review: Initial Criteria: Removed: "acid alpha-glucosidase gene mutation; AND " from the following statement: "Patient has a molecular genetic test demonstrating acid alpha-glucosidase gene mutation; AND" and replaced with: "biallelic pathogenic or likely pathogenic acid alpha-glucosidase (GAA) gene variants; AND" Added: "Will not be used in combination with other enzyme replacement therapies (i.e., avalglucosidase alfa, cipaglucosidase alfa, etc.); AND Documented baseline age-appropriate values for one or more of the following: Infantile-onset disease: muscle weakness, motor function, respiratory function, cardiac involvement, percent predicted forced vital capacity (FVC), and/or 6-minute walk test (6-MWT); OR Late-onset (non-infantile) disease: FVC and/or 6-MWT; AND" Added RENEWAL CRITERIA "Coverage may be renewed based on the following criteria: Patient continues to meet Initial Criteria; AND Absence of unacceptable toxicity from the drug. *Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, immune-mediated cutaneous reactions, systemic immune-mediated reactions, acute cardiorespiratory failure, cardiac arrhythmia during general anesthesia, etc.*; AND Patient is being monitored for antibody formation (including neutralizing antibodies); AND Patient has demonstrated a beneficial response to therapy compared to pretreatment age-appropriate baseline values in ONE or more of the following: Infantile-onset disease: stabilization or improvement in muscle weakness, motor function, respiratory function, cardiac involvement, FVC and/or 6-MWT; OR Late-onset (non-infantile) disease: stabilization or improvement in FVC and/or 6-MWT"
EmblemHealth & ConnectiCare	2/2/2024	Annual Review: No criteria changes
EmblemHealth & ConnectiCare	4/7/2023	Transfer from CCUM template to Co-Branded medical template Retired MG.MM.PH.303
EmblemHealth & ConnectiCare	4/6/2022	Annual Revision: No criteria changes
EmblemHealth & ConnectiCare	4/7/2021	Annual Revision: No criteria changes

References

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