

Medical Policy:

Lumizyme (alglucosidase) Intravenous

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

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The treating physician or primary care provider must submit to EmblemHealth, or ConnectiCare, as applicable (hereinafter jointly referred to as “EmblemHealth”), the clinical evidence that the member meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request preauthorization or post-payment review. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Health care providers are expected to exercise their medical judgment in rendering appropriate care.

EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary.

If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication.

EmblemHealth may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice. EmblemHealth Services Company, LLC, has adopted this policy in providing management, administrative and other services to EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC, and Health Insurance Plan of Greater New York (HIP) related to health benefit plans offered by these entities. ConnectiCare, an EmblemHealth company, has also adopted this policy. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

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. <i>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.</i> ; 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.