

Medical Policy:

Fabrazyme® (agalsidase) intravenous infusion

| POLICY NUMBER | LAST REVIEW | ORIGIN DATE |
|---------------|---------------|---------------|
| MG.MM.PH.311 | April 8, 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

Fabrazyme, a human α -galactosidase A (α -Gal), is indicated for use in patients with Fabry disease. It is the same amino acid sequence as the native enzyme and is produced in Chinese hamster ovary cells via recombinant DNA technology. Fabrazyme catalyzes the breakdown of globotriaosylceramide (GL-3) and other α -galactyl-terminated neutral glycosphingolipids to ceramide and galactose and reduces the deposition of GL-3 in the capillary endothelium of the kidney and certain other cell types.

Length of Authorization

Coverage will be provided for 12 months and may be renewed.

Dosing Limits [Medical Benefit]

Each dose must not exceed 1 mg/kg administered intravenously no more frequently than once every 2 weeks.

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

- 115 billable units every 14 days

Guideline

I. INITIAL CRITERIA

1. **Fabry Disease.** Approve for 1 year if the patient meets the following criteria (A, B, and C):
 - A. The diagnosis is established by ONE of the following (i **or** ii):
 - i. Patient has a laboratory test demonstrating deficient α -galactosidase A activity in leukocytes or fibroblasts; **OR**
 - ii. Patient has a molecular genetic test demonstrating a pathogenic variant in the galactosidase alpha (*GLA*) gene; **AND**
 - B. Patient has a baseline of ONE or more of the following:
 - i. Tissue globotriaosylceramide (Gb3/GL-3) inclusions
 - ii. Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lyso-Gb3)
 - iii. Clinical signs and/or symptoms of disease (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations) ; **AND**
 - C. Fabrazyme is prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

II. RENEWAL CRITERIA

1. Patient continues to meet Initial Criteria; **AND**
2. Absence of unacceptable toxicity from the drug. (*Examples of unacceptable toxicity include: anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, compromised cardiac function, etc.*); **AND**
3. Disease response with treatment as defined by a reduction or stabilization in one or more of the following, as compared to pre-treatment baseline:
 - A. Tissue globotriaosylceramide (Gb3/GL-3) inclusions
 - B. Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lysoGb3); **OR**
4. Disease response with treatment as defined by an improvement or stabilization of clinical signs and/or symptoms (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations)

Applicable Procedure Codes

| Code | Description |
|-------|----------------------------------|
| J0180 | Injection, agalsidase beta, 1 mg |

Applicable NDCs

| Code | Description |
|---------------|---------------------------------------|
| 58468-0041-01 | Fabrazyme 5MG Solution Reconstituted |
| 58468-0040-01 | Fabrazyme 35MG Solution Reconstituted |

ICD-10 Diagnoses

| Code | Description |
|--------|---------------|
| E75.21 | Fabry disease |

Revision History

| Company(ies) | DATE | REVISION |
|--------------|------|----------|
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|-----------------------------|----------|---|
| EmblemHealth & ConnectiCare | 4/8/2025 | Annual Review: Initial Criteria: Removed: “mutations in the galactosidase alpha gene; AND” reworded to: “ a pathogenic variant in the galactosidase alpha (GLA) gene; AND “ Added: “Patient has a baseline of one or more of the following: Tissue globotriaosylceramide (Gb3/GL-3) inclusions Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lyso-Gb3) Clinical signs and/or symptoms of disease (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations) ; AND” Renewal criteria: removed and reworded: “Disease response with treatment per physician attestation” reworded to: “Disease response with treatment as defined by a reduction or stabilization in one or more of the following, as compared to pre-treatment baseline: Tissue globotriaosylceramide (Gb3/GL-3) inclusions Plasma or urinary globotriaosylceramide (Gb3/GL-3) or globotriaosylsphingosine (lysoGb3); OR Disease response with treatment as defined by an improvement or stabilization of clinical signs and/or symptoms (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations)” |
| EmblemHealth & ConnectiCare | 3/4/2024 | Annual Review: updated dosing limits, added renewal criteria |
| EmblemHealth & ConnectiCare | 4/7/2023 | Transfer from CCUM Template to Co-Branded Medical template Retired MG.MM.PH.82 |
| EmblemHealth & ConnectiCare | 4/6/2022 | Annual revision: No criteria changes |
| EmblemHealth & ConnectiCare | 4/7/2021 | Annual revision: No criteria changes |

References

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