Medical Policy:

Elfabrio (pegunigalsidase alfa-iwxj) intravenous injection

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.389	April 3, 2025	July 28, 2023

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

Fabry disease is caused by deficiency of the lysosomal enzyme alpha-galactosidase A. Pegunigalsidase alfa-iwxj provides an exogenous source of alpha-galactosidase A; it is internalized and transported into lysosomes where it is thought to exert enzymatic activity and reduce accumulated globotriaosylceramide (Gb3)

Length of Authorization

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

Dosing Limits [Medical Benefit]

The recommended dosage of Elfabrio, based on actual body weight, is 1 mg/kg administered by intravenous infusion every 2 weeks.

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

• 120 mg every 14 days

Guideline

I. INITIAL CRITERIA

1. **Fabry Disease.** Approve if the patient meets the following criteria (A, B, and C):
   A. The patient is ≥ 18 years of age; **AND**
   B. 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 pathogenic variants in the galactosidase alpha gene; **AND**
   C. Elfabrio 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, glomerulonephritis, etc.;* **AND**
3. Disease response with treatment as defined by improvement or stabilization in the rate of decline of the estimated glomerular filtration rate (eGFR)

Limitations/Exclusions

1. Concurrent Use with Galafold (migalastat oral capsules)
2. Concurrent Use with Fabrazyme (agalsidase beta intravenous infusion)

Applicable Procedure Codes

Code	Description
J2508	Injection, pegunigalsidase alfa-iwxj, 1 mg

Applicable NDCs

Code	Description
10122-0160-xx	Elfabrio 20mg/10 mL vial
10122-0165-02	Elfabrio 5 mg/2.5 mL

ICD-10 Diagnoses

Code	Description
E75.21	Fabry(-Anderson) disease

Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	4/2/2025	Annual Review: Fabry Disease: For diagnosis confirmed by genetic testing, the term “mutation” was rephrased to “pathogenic variant.”
EmblemHealth & ConnectiCare	3/18/2024	Annual Review: Updated dosing limits, Added Limitations/Exclusions and Renewal criteria, removed J3590 and C9399, added J2508

EmblemHealth & ConnectiCare	07/28/2023	New Policy

References

1. Product Information: Elfabrio intravenous injection, pegunigalsidase alfa-iwxj intravenous injection. Chiesi USA, Inc. (per FDA), Cary, NC, 2023.