# POLICY Document for ELFABRIO (pegunigalsidase alfa-iwxj)

The overall objective of this policy is to support the appropriate and cost-effective use of the medication, specific to use of preferred medication options, lower cost site of care and overall, clinically appropriate use. This document provides specific information to each of the three sections of the overall policy.

**Section 1: Site of Care**

- Policy information specific to site of care (outpatient, hospital outpatient, home infusion)

**Section 2: Clinical Criteria**

- Policy information specific to the clinical appropriateness for the medication

## Section 1: Site of Care

### Site of Care Criteria
### Administration of Intravenous Elfabrio

**POLICY**

**I. CRITERIA FOR APPROVAL FOR ADMINISTRATION IN OUTPATIENT HOSPITAL SETTING**

This policy provides coverage for administration of Elfabrio in an outpatient hospital setting for up to 106 days when a member is new to therapy or is reinitiating therapy after not being on therapy for at least 6 months.

This policy provides coverage for administration of Elfabrio in an outpatient hospital setting for a longer course of treatment when ANY of the following criteria are met:

A. The member has experienced an adverse reaction to the drug that did not respond to conventional interventions (eg, acetaminophen, steroids, diphenhydramine, fluids, pre-medications or slowing of infusion rate) or a severe adverse event (anaphylaxis, anaphylactoid reactions, myocardial infarction, thromboembolism, or seizures) during or immediately after an infusion.
B. The member has developed IgG or IgE anti-drug antibodies which increases the risk for infusion related reactions.
C. The member is medically unstable (eg respiratory, cardiovascular, or renal conditions).
D. The member has severe venous access issues that require the use of special interventions only available in the outpatient hospital setting.
E. The member has significant behavioral issues and/or physical or cognitive impairment that would impact the safety of the infusion therapy AND the patient does not have access to a caregiver.
F. Alternative infusion sites (pharmacy, physician office, ambulatory care, etc.) are greater than 30 miles from the member's home.
G. The member is less than 14 years of age.

For situations where administration of Elfabrio does not meet the criteria for outpatient hospital infusion, coverage for Elfabrio is provided when administered in alternative sites such as; physician office, home infusion or ambulatory care.

**II. REQUIRED DOCUMENTATION**

The following information is necessary to initiate the site of care prior authorization review (where applicable):

A. Medical records supporting the member has experienced an adverse reaction that did not respond to conventional interventions or a severe adverse event during or immediately after an infusion
B. Medical records supporting the member has developed IgG or IgE anti-drug antibodies
C. Medical records supporting the member is medically unstable
D. Medical records supporting the member has severe venous access issues that requires specialized interventions only available in the outpatient hospital setting
E. Medical records supporting the member has behavioral issues and/or physical or cognitive impairment and no access to a caregiver
F. Records supporting alternative infusion sites are greater than 30 miles from the member's home.
G. Medical records supporting the member is new to therapy

# Section 2: Clinical Criteria

## SPECIALTY GUIDELINE MANAGEMENT

### ELFABRIO (pegunigalsidase alfa-iwxj)

**POLICY**

**III. INDICATIONS**

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Elfabrio is indicated for the treatment of adults with confirmed Fabry disease.

All other indications are considered experimental/investigational and not medically necessary.

**IV. DOCUMENTATION**

Submission of the following information is necessary to initiate the prior authorization review:
A. Initial requests: alpha-galactosidase enzyme assay or genetic testing results supporting diagnosis. In the case of obligate carriers, the documentation must be submitted for the parent.
B. Continuation requests: lab results or chart notes documenting a positive response to therapy.

**V. CRITERIA FOR INITIAL APPROVAL**

**Fabry disease**
Authorization of 12 months may be granted for treatment of Fabry disease when both of the following criteria are met:
A. The diagnosis of Fabry disease was confirmed by enzyme assay demonstrating a deficiency of alpha-galactosidase enzyme activity or by genetic testing, or the member is a symptomatic obligate carrier; and
B. The requested medication will not be used in combination with Galafold.

**VI. CONTINUATION OF THERAPY**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III who are responding to therapy (e.g., reduction in plasma globotriaosylceramide [GL-3, Gb3] or GL-3/Gb3 inclusions, improvement and/or stabilization in renal function, pain reduction).

## REFERENCES


**SECTION 1**

1. Elfabrio [package insert]. Cary, NC: Chiesi USA, Inc.; May 2023.

**SECTION 2**

1. Elfabrio [package insert]. Cary, NC: Chiesi USA, Inc.; May 2023.
2. Biegstraaten M, Arngrimsson R, Barbey F, et al. Recommendations for initiation and cessation of enzyme replacement therapy in patients with Fabry disease: the European Fabry Working Group consensus document. *Orphanet J Rare Dis.* 2015; 1036.