Chiesi TOTAL care

Chiesi Total Care℠ is here to help.
Visit chiesitotalcare.com or
call 1-833-656-1056

ELFABRIO®
(pegunigalsidase alfa-iwxj)

Access Resource Guide

Using This Access Resource Guide

This guide is intended for informational purposes only, not to take the place of the provider's diagnosis and treatment decisions. Providers are responsible for the accuracy, legitimacy, and completeness of any claims, invoices, and other documentation supplied to payers. Providers should contact the payer for answers to specific questions about payment or coverage. Specific direction from the payer supersedes the guidance provided in this guide. Using the guidance provided in this guide does not guarantee coverage or reimbursement.

Indication and Important Safety Information

Indication

Elfabrio® (pegunigalsidase alfa-iwxj) is indicated for the treatment of adults with confirmed Fabry disease.

Important Safety Information

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with Elfabrio have experienced hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Elfabrio administration. If a severe hypersensitivity reaction (eg, anaphylaxis) occurs, discontinue Elfabrio immediately and initiate appropriate medical treatment. In patients with severe hypersensitivity reaction, a desensitization procedure to Elfabrio may be considered.

Please see Important Safety Information on page 2 and Full Prescribing Information, including Boxed Warning on hypersensitivity reactions including anaphylaxis.

Important Safety Information

Indication and Important Safety Information

Indication

Elfabrio® (pegunigalsidase alfa-iwxj) is indicated for the treatment of adults with confirmed Fabry disease.

Important Safety Information

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with Elfabrio have experienced hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Elfabrio administration. If a severe hypersensitivity reaction (eg, anaphylaxis) occurs, discontinue Elfabrio immediately and initiate appropriate medical treatment. In patients with severe hypersensitivity reaction, a desensitization procedure to Elfabrio may be considered.

Prior to Elfabrio administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. Inform patients and caregivers of the signs and symptoms of hypersensitivity reactions and infusion-associated reactions (IARs), and instruct them to seek medical care immediately if such symptoms occur.

- If a severe hypersensitivity reaction (including anaphylaxis) or severe IAR occurs, immediately discontinue Elfabrio administration and initiate appropriate medical treatment.
- If a mild to moderate hypersensitivity reaction or IAR occurs, consider slowing the infusion rate or temporarily withholding the dose.

In clinical trials, 20 (14%) Elfabrio-treated patients experienced hypersensitivity reactions. Four Elfabrio-treated patients (3%) experienced anaphylaxis reactions that occurred within 5 to 40 minutes of the start of the initial infusion. The signs and symptoms of hypersensitivity reactions and anaphylaxis included headache, nausea, vomiting, throat tightness, facial and oral edema, truncal rash, tachycardia, hypotension, rigors, urticaria, intense pruritus, moderate upper airway obstructions, macroglossia, and mild lip edema.

In clinical trials, 41 (29%) Elfabrio-treated patients experienced one or more infusion-associated reactions, including hypersensitivity, nausea, chills, pruritus, rash, chest pain, dizziness, vomiting, asthenia, pain, sneezing, dyspnea, nasal congestion, throat irritation, abdominal pain, erythema, diarrhea, burning sensation, neuralgia, headache, paresthesia, tremor, agitation, increased body temperature, flushing, bradycardia, myalgia, hypertension, and hypotension.

A case of membranoproliferative glomerulonephritis with immune depositions in the kidney was reported during clinical trials. Monitor serum creatinine and urinary protein-to-creatinine ratio. If glomerulonephritis is suspected, discontinue treatment until a diagnostic evaluation can be conducted.

Table of Contents

ELFABRIO® (pegunigalsidase alfa-iwxj)

Important Safety Information > 2

Patient Support Services > 4

Getting Your Patient Started on ELFABRIO > 6

Coverage Considerations > 7

Prior Authorization > 8

Formulary Exceptions > 9

Medical Necessity > 10

- Sample Letter of Medical Necessity >

Navigating Appeals and Denials > 12

- Sample Letter of Appeal >

Payer Landscape > 14

How to Acquire ELFABRIO > 15

Coding and Billing > 16

- Sample CMS-1500 Claim Form >
- Sample CMS-1450 Claim Form >

Important Safety Information (continued)

When switching to Elfabrio from a prior enzyme replacement therapy, the risk of hypersensitivity reactions and infusion-associated reactions may be increased in certain patients with pre-existing anti-drug antibodies (ADAs). Consider monitoring IgG and IgE ADAs and clinical or pharmacodynamic response (eg, plasma lyso-Gb3 levels).

The most common adverse reactions (≥15%) were infusion-associated reactions, nasopharyngitis, headache, diarrhea, fatigue, nausea, back pain, pain in extremity, and sinusitis.

Patient Support Services

ELFABRIO® (pegunigalsidase alfa-iwxj)

Chiesi Total Care℠

A single call to your dedicated Chiesi Total Care Team is all it takes—we will guide you through the process of getting a patient started on Elfabrio® (pegunigalsidase alfa-iwxj) therapy.

Insurance eligibility—Chiesi Total Care assists you and your patients with:

Commercial insurance
If your patients have private insurance through their job or their own business

Government insurance
If your patients have Medicare, Medicaid, Veterans Affairs healthcare, or other government insurance

No insurance
If your patients have no insurance, they may be eligible for financial assistance

Copay Assistance

Chiesi Total Care offers 2 copay programs for eligible patients*:

- Prescription copay: This covers the medication itself. Patients may pay as little as $0 for their Elfabrio prescription
- Infusion services copay: This covers infusion supplies and administration (including home infusion). Patients may pay as little as $0 for their Elfabrio infusion supplies and administration

*Government-funded plans are not eligible for patient support services that provide financial support through the Programs. Patients receiving treatment or residing in MA, MI, MN, or RI are not eligible for infusion services. To receive home infusion support, patients must be referred to home infusion by their prescribing physician.

Program eligibility:

- Patient must be enrolled in Chiesi Total Care. (Enrollment and Authorization Form will be mailed to your patient's home)
- Patient has commercial insurance and a valid prescription for a US Food and Drug Administration (FDA)-approved indication for Elfabrio
- Patient must be a resident of the United States or one of its territories

Please refer to the accompanying full Terms and Conditions for additional eligibility requirements.

The dedicated *Chiesi Total Care Team* is made up of:

Pharmacists | Patient Service Coordinators | Reimbursement support | Nursing support

Additional *Offerings*

Infusion services*

- Chiesi Total Care clinicians can help patients understand their medication and the infusion process and coordinate a suitable infusion site if needed
- If your patient moves to home infusion, Chiesi Total Care may be able to assist eligible patients with delivery of medication and infusion supplies

*Patients receiving treatment or residing in MA, MI, MN, or RI are not eligible for infusion services. To receive home infusion support, patients must be referred to home infusion by their prescribing physician. Please refer to the full Terms and Conditions for additional eligibility requirements.

Routine testing†

- We provide support for globotriaosylsphingosine (lyso-Gb3) and anti-drug antibodies (ADAs) screenings, with no out-of-pocket costs for patients with commercial insurance. Contact your Chiesi representative for more information

†Tests must be ordered for eligible patients by the prescribing physician. Patients receiving treatment or residing in MA, MI, MN, or RI are not eligible. Please see full Terms and Conditions for additional eligibility requirements.

Chiesi TOTAL care

BY PHONE
1-833-656-1056

BY FAX
1-636-355-3610

HOURS OF OPERATION
7:00 AM - 6:00 PM (Central Time)

For more information, visit chiesitotalcare.com

Indication and Important Safety Information

Indication

Elfabrio® (pegunigalsidase alfa-iwxj) is indicated for the treatment of adults with confirmed Fabry disease.

Important Safety Information

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with Elfabrio have experienced hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Elfabrio administration. If a severe hypersensitivity reaction (eg, anaphylaxis) occurs, discontinue Elfabrio immediately and initiate appropriate medical treatment. In patients with severe hypersensitivity reaction, a desensitization procedure to Elfabrio may be considered.

Getting Your Patient Started on ELFABRIO

A step-by-step guide to the coverage process

Step 1

Complete the **Physician Order/Prescription Form for ELFABRIO,** then enroll the patient in the Chiesi Total Care Program by completing the **Patient Enrollment Form**

✓ Call Chiesi Total Care Program: 1-833-656-1056

or

✓ **Download the Prescription Order Form** ↓ **Download the Patient Enrollment Form** ↓

Visit the Chiesi Total Care website >

Step 2

Chiesi Total Care will determine patient insurance eligibility for ELFABRIO coverage

Step 3

You may need to prepare and submit a **prior authorization (PA)** or **formulary exception** (or tiering exception) and include a **letter of medical necessity**

- **If denied,** prepare and submit a **letter of appeal** using the resources provided in this guide
- **If approved,** work with the patient to set up delivery of ELFABRIO through the preferred specialty pharmacy

Chiesi TOTAL care

Chiesi Total Care can help healthcare providers with benefit investigations, PAs, and the appeal process.

Coverage Considerations

ELFABRIO® (pegunigalsidase alfa-iwxj)

ELFABRIO may be covered under the pharmacy benefit or medical benefit, depending on the patient's health plan[1]

For newly FDA-approved specialty drugs, in the first several months following approval, health plans have yet to make formulary decisions or establish coverage policies.[3]

- Conducting a thorough benefit investigation will uncover if there are any specific health plan restrictions or utilization requirements, such as step therapy or PAs[1]
- If you have determined a patient receiving a different treatment for Fabry disease should be treated with ELFABRIO, there may be additional requirements or approval processes for the premedication required before treatment

Important Safety Information (continued)

Prior to Elfabrio administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. Inform patients and caregivers of the signs and symptoms of hypersensitivity reactions and infusion-associated reactions (IARs), and instruct them to seek medical care immediately if such symptoms occur.

- If a severe hypersensitivity reaction (including anaphylaxis) or severe IAR occurs, immediately discontinue Elfabrio administration and initiate appropriate medical treatment.
- If a mild to moderate hypersensitivity reaction or IAR occurs, consider slowing the infusion rate or temporarily withholding the dose.

Please see Important Safety Information on page 2 and Full Prescribing Information, including Boxed Warning on hypersensitivity reactions including anaphylaxis.

Prior Authorization

For most patients, their health plan will require a PA for treatment with ELFABRIO

In some cases, it may be appropriate to submit a letter of medical necessity with a PA[5]

Considerations for submitting PAs:

- Requirements for PAs will vary by health plan[5]
- Check to see if the health plan has a plan-specific PA form and process for submission[5]
  –This information can generally be found on a plan's website or by contacting their customer service department

Information generally required on a PA:

Patient information
- Name
- Address
- Date of birth
- Name of policyholder
- Policy ID and group numbers

Provider information
- Name
- NPI number
- Plan provider number
- Address
- Phone/fax numbers

Patient clinical information
- Diagnosis/ICD-10-CM code
- Description of treatment or procedure being requested
- Previous treatment history
- Treatment start date

If approved, it is important to understand the length of time the authorization is valid. This can vary by health plan, and in some cases, reauthorization may be required annually.

Important Safety Information (continued)

In clinical trials, 20 (14%) Elfabrio-treated patients experienced hypersensitivity reactions. Four Elfabrio-treated patients (3%) experienced anaphylaxis reactions that occurred within 5 to 40 minutes of the start of the initial infusion. The signs and symptoms of hypersensitivity reactions and anaphylaxis included headache, nausea, vomiting, throat tightness, facial and oral edema, truncal rash, tachycardia, hypotension, rigors, urticaria, intense pruritus, moderate upper airway obstructions, macroglossia, and mild lip edema.

Formulary Exceptions

ELFABRIO® (pegunigalsidase alfa-iwxj)

A formulary exception under the pharmacy benefit is a request for coverage of a drug that is not included on a plan's formulary, or it may be a request to have a utilization requirement waived[6]

Formulary exceptions may include requests for[6]:

- A drug that is not on the plan's list of covered drugs
- A drug that was previously included on the plan's list of covered drugs but was or is being removed from this list during the plan year
- PA for the prescribed drug
- Forgoing the requirement to use another drug before receiving the prescribed drug (step therapy)
- An exception to the plan's requirement of a quantity limit

Tiering exceptions may include situations when[6]:

- A health plan charges a higher copay for the prescribed drug than it charges for another drug that treats the condition, and the lower copayment is preferred
- A patient has been using a drug that was previously included on a lower copayment tier, but was or is being moved to a higher copayment tier
- A health plan charged a higher copay for a drug than it should have
- A patient wants to be reimbursed for a covered prescription drug that was paid for out-of-pocket

Medicare Part D Prescription Drug Plans (PDPs), including Medicare Advantage plans that offer prescription drug coverage (MA-PD), and Medicare/Medicaid plans (for dually eligible individuals) that vary in their requirements for making exception requests. Centers for Medicare and Medicaid Services (CMS) provides some guidance for coverage determination requests.[2,4,6]

A model coverage determination request form and instructions can be downloaded from the CMS website.

It may be appropriate to submit a letter of medical necessity with a formulary exception request.

Medical Necessity

A letter of medical necessity is used to help explain the clinical rationale behind prescribing a specific treatment[7]

May be required if a medication:

- is subject to **step therapy** or **prior authorization**
- is newly FDA-approved
- is not preferred or **not available on the plan's formulary** and a **formulary exception request** is necessary
- has unfavorable coverage or **no established coverage policy** with the plan

Considerations for preparing a letter of medical necessity:

- Understand and adhere to the health plan–specific process and deadlines for PAs and exception request

Recommended information to include in a letter of medical necessity:

Patient information

- Name
- Address
- Date of birth
- Name of policyholder
- Policy ID and group numbers

Provider information

- Name
- NPI number
- Plan provider number
- Address
- Phone/fax numbers

Patient clinical information

- Diagnosis/ICD-10-CM code
- Description of treatment or procedure being requested
- Previous treatment history, including duration and reason for discontinuation
- Severity of the patient's condition

Clinical rationale for treatment with ELFABRIO

- Include clinical trial outcomes that supported FDA approval
- In the case of a formulary exception request, explain the rationale for not treating with the health plan's preferred alternative

Summary of recommendations based on provider clinical expertise

Enclosures that support the rationale for treatment with ELFABRIO

- Peer-reviewed literature
- Patient medical records/notes
- Relevant lab or diagnostic test results

Sample Letter of Medical Necessity

ELFABRIO® (pegunigalsidase alfa-iwxj)

[Insurance Company]
[Address]
[City, State, Zip]

Re: **[Patient Name]**
[Policy #]
[DOB]
[Address]
[City, State, Zip]

To Whom It May Concern:

I am writing this letter of medical necessity on behalf of **[Patient Name, ID#, Group #]** to request coverage for **[Product name (generic name)]**. Included in this letter of medical necessity is information on the treatment rationale, medical records, medical necessity data and medical studies confirming currently prescribed product as an effective treatment for the diagnosis associated with **[ICD10 Code]**.

Treatment Rationale:
[Provide information on patient response and history to past treatments and anticipated prognosis and rationale for the currently prescribed product].

Outline of Medical Studies:
[Outline a brief overview of the studies evaluating the use of the currently prescribed product in this condition and/or patient population. Remember to include the FDA approved indications and usage].

Medical Record Information:
[Highlight key dates and entries of the medical record how the currently prescribed product is used].

Per the included medical information, it is my professional opinion that the currently prescribed product is medically necessary in treating the patient and the denials for the patient's use of the drug should be reversed. Please call my office at **[Office Phone Number]** if I can provide further information.

Sincerely,
[Physician Name and Signature]
[Phone Number]
Enclosure: **[As required]**

This is a sample letter. Use of this template does not guarantee coverage of ELFABRIO.

Important Safety Information (continued)

In clinical trials, 41 (29%) Elfabrio-treated patients experienced one or more infusion-associated reactions, including hypersensitivity, nausea, chills, pruritus, rash, chest pain, dizziness, vomiting, asthenia, pain, sneezing, dyspnea, nasal congestion, throat irritation, abdominal pain, erythema, diarrhea, burning sensation, neuralgia, headache, paresthesia, tremor, agitation, increased body temperature, flushing, bradycardia, myalgia, hypertension, and hypotension.

Please see Important Safety Information on page 2 and Full Prescribing Information, including Boxed Warning on hypersensitivity reactions including anaphylaxis.

Navigating Denials and Appeals

In the case of a denied coverage request, patients and providers have the right to submit an appeal to the health plan[8]

Common reasons for denials[9]:

- New drug that has not been reviewed by the health plan yet for formulary inclusion or medical policy development
- The drug is non-preferred or not on formulary
- Step therapy is required and a different drug must be used first
- PA is required and was not submitted (or PA has expired)
- Missing information on the PA form or coverage determination request

Considerations for navigating denials and appeals:

- Identify the reason for the denial by reviewing the explanation of benefits (EOB) or the denial letter
- Understand and adhere to the health plan–specific appeal's process and deadlines
- Prepare a detailed letter of appeal to submit to the plan

Recommended information to include with a letter of appeal:

Patient information

- Name
- Address
- Date of birth
- Name of policyholder
- Policy ID and group numbers

Provider information

- Name
- NPI number
- Plan provider number
- Address
- Phone/fax numbers

- Date and reason for denial
- Patient diagnosis (ICD-10-CM code), description of treatment being requested, previous treatment history, including duration and reason for discontinuation, if applicable
- Explanation for why an alternative formulary treatment is not appropriate for the patient
- Clinical rationale for treatment with ELFABRIO
  – Include clinical trial outcomes that supported FDA approval
- Summary of recommendations based on provider clinical expertise

Enclosures that support the rationale for treatment with ELFABRIO

- Peer-reviewed literature
- Patient medical records/notes
- Relevant lab or diagnostic test results

Chiesi Total Care can help with denials and appeals.

We have an established success rate in appeals. If your PA submission was denied, call Chiesi Total Care at 1-833-656-1056.

Sample Letter of Appeal

ELFABRIO® (pegunigalsidase alfa-iwxj)

[Insurance Company]
[Address]
[City, State, Zip]

Re: **[Patient Name]**
[Policy #]
[DOB]
[Address]
[City, State, Zip]

To Whom It May Concern:

I am writing to appeal the denial of benefits for the use of **[Product name (generic name)]** for services requested for **[Patient Name, ID#, Group #]**. Included in this letter of appeal are information on the treatment rationale, medical records, medical necessity data and medical studies confirming currently prescribed product as an effective treatment for the diagnosis associated with **[ICD10 Code]**.

Treatment Rationale:
[Provide information on patient response and history to past treatments and anticipated prognosis and rationale for the currently prescribed product].

Outline of Medical Studies:
[Outline a brief overview of the studies evaluating the use of the currently prescribed product in this condition and/or patient population. Remember to include the FDA approved indications and usage].

Medical Record Information:
[Highlight key dates and entries of the medical record how the currently prescribed product is used].

Per the included medical information, it is my professional opinion that the currently prescribed product is medically necessary in treating the patient and the denials for the patient's use of the drug should be reversed. Please call my office at **[Office Phone Number]** if I can provide further information or speak with a review board to appeal the denial of coverage decision. I look forward to reaching resolution of overturning the denied status of the currently prescribed product for this patient.

Sincerely,
[Physician Name and Signature]
[Phone Number]
Enclosure: **[Original denial notification copy]**

This is a sample letter. Use of this template does not guarantee coverage of ELFABRIO.

Important Safety Information (continued)

A case of membranoproliferative glomerulonephritis with immune depositions in the kidney was reported during clinical trials. Monitor serum creatinine and urinary protein-to-creatinine ratio. If glomerulonephritis is suspected, discontinue treatment until a diagnostic evaluation can be conducted.

Payer Landscape

Commercial, Medicare Advantage, and Medicare Part D PDP coverage for ELFABRIO will vary by plan and by patient. A thorough benefit investigation will determine coverage, including requirements for PAs or step therapy.[1]

Original Medicare does not offer prescription coverage. When covered by Medicare, ELFABRIO will most likely be covered under the pharmacy benefit, or Part D, and coverage will be determined based on the Part D formulary.[10]

In the case where ELFABRIO is covered under the Medicare Part B benefit, a local coverage determination will dictate coverage parameters and, in these cases, infusions would have to be delivered in a setting that buys and bills ELFABRIO directly versus the provider acquiring it through a specialty pharmacy.[1,10,11]

Eligibility and benefit plans through state Medicaid programs (administered by the state) and **Managed Medicaid** (administered by commercial payers) vary from state to state.[2,12] Usually, treatment with ELFABRIO will need to be considered medically necessary to be covered under the Medicaid program. Depending on the state, initial treatment with ELFABRIO may require prior authorization by the state Medicaid program.

Tips on engaging with health plans

- **Follow the individual health plan's guidelines** and processes for submitting any type of coverage determination, including PAs, exception requests, and letters of medical necessity and appeal
- **Review and thoroughly understand** the health plan's formulary criteria or coverage policy when preparing communications
- **Thoroughly consider the reason(s) for denial** and address those reason(s) specifically when preparing an appeal letter
- **Provide a clear and concise description** of the patient's medical history and the clinical rationale for the treatment request
- **Clearly state the provider's credentials** and relevant affiliations in communications, including experience in treating the disease state
- **Submit all requested forms,** medical records, and any additional supporting clinical documentation through the health plan's preferred or recommended method
- **Document and keep record** of all communication with the health plan, including phone calls and written communication
- **Adhere to any specific deadlines** for coverage determinations and appeals, including requests from the health plan for additional information

How to Acquire ELFABRIO

ELFABRIO (pegunigalsidase alfa-iwxj)

Specialty pharmacy

ELFABRIO prescriptions can be fulfilled through the preferred specialty pharmacy or by enrolling the patient in the Chiesi Total Care Program and completing the ELFABRIO Physician Order/Prescription Form

Chiesi TOTAL care

- Call Chiesi Total Care Program: 1-833-656-1056

or

- Download the Prescription Order Form
- Download the Patient Enrollment Form

Visit the Chiesi Total Care website

Buy and bill

For healthcare providers who prefer to buy and bill, ELFABRIO can be acquired through EVERSANA® Life Science Services.

For more information or to place an order, email: CTCPurchasing@eversana.com

Chiesi TOTAL care | EVERSANA®

Chiesi Total Care is offered through EVERSANA® Life Science Services Specialty Pharmacy.

Important Safety Information (continued)

When switching to Elfabrio from a prior enzyme replacement therapy, the risk of hypersensitivity reactions and infusion-associated reactions may be increased in certain patients with pre-existing anti-drug antibodies (ADAs). Consider monitoring IgG and IgE ADAs and clinical or pharmacodynamic response (eg, plasma lyso-Gb3 levels).

Coding and Billing

ELFABRIO® (pegunigalsidase alfa-iwxj)

The codes provided here are commonly associated with the administration of ELFABRIO; however, providers should contact the patient's health plan for specific guidance on coding and site of care requirements before administration.

Healthcare Common Procedure Coding System (HCPCS) Codes

Provider-administered drugs are typically reported with **HCPCS** Level II J-codes and assigned by CMS.[13] Miscellaneous or "unclassified" J-codes allow healthcare providers to begin billing immediately for a service or item as soon as the FDA allows it to be marketed and until a permanent code is assigned.[13,14] A unit of "1" should always be billed with a miscellaneous J-code.[15]

Code[16]	Description
J3590	Unclassified biologics
C9399	Unclassified drugs or biologicals (hospital outpatient use)

When billing with a miscellaneous HCPCS code, the plan may require additional information and documents, such as:

- Drug name
- Drug strength
- Unit of measure
- Number of units administered
- Number of units wasted
- Total dosage
- Route of administration
- 11-digit NDC
- A copy of the ELFABRIO invoice[15,17]

National Drug Codes (NDC)

NDCs are unique numbers that identify a drug's labeler, product, and trade package size. The FDA uses a 10-digit format when registering NDCs; however, payer requirements regarding use of a 10-digit or 11-digit NDC on claim forms varies. CMS requires an 11-digit NDC format. It is important to check with individual health plans before billing.[18,19]

10-Digit Code[19] (5-3-2 format)	11-Digit Code (5-4-2 format)	Description[19]
10122-160-01	10122-0160-01	Single-dose glass vial
10122-160-02	10122-0160-02	Carton containing one vial
10122-160-05	10122-0160-05	Carton containing five vials
10122-160-10	10122-0160-10	Carton containing ten vials

International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM) Codes

ICD-10-CM is the diagnosis code set used for all healthcare settings for medical claims reporting.[20]

Code[16]	Description
E75.21	Fabry (Anderson) Disease

Current Procedural Terminology (CPT®) Codes

CPT is the code set used to describe procedures and services performed by healthcare providers. Evaluation and management codes, or E/M codes, are CPT codes that allow providers to bill for the total time and level of service spent treating the patient.[21]

Code[16]	Description
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
96366	Each additional hour (list separately in addition to primary procedure code, 96365)

Modifiers

Modifiers are 2-digit codes that are added to a CPT or HCPCS code and used to provide additional information about an item or service provided.[14]

Code[22,23]	Description
59	Distinct Procedural Service: Under certain circumstances, it may be necessary to indicate that a procedure or service was distinct or independent from other non-E/M services performed on the same day
JW	Discarded drug not administered
JZ	Zero drug wasted (effective July 1, 2023, Medicare plans will require the JZ Modifier to attest that there was no discarded amount from a single vial. Requirements vary by plan)

Place of Service (POS) Codes

POS codes are 2-digit numeric codes used to indicate the setting in which a healthcare service was provided and are only used on professional claims.[24]

Code[24]	Description
11	Office
12	Home
19	Off-Campus Outpatient Hospital
22	On-Campus Outpatient Hospital

Revenue Codes

Revenue codes are 4-digit numeric codes used only by hospital-based facilities to indicate what department and where a procedure was performed within the facility or to identify supplies used in the procedure. Revenue codes are only used on institutional claims.[25]

Code[26]	Description
0258	Pharmacy, IV solutions
0260	IV Therapy, general IV
0261	IV Therapy, infusion pump
0636	Pharmacy, drugs requiring detailed coding

Please see Important Safety Information on page 2 and Full Prescribing Information, including Boxed Warning on hypersensitivity reactions including anaphylaxis.

Sample CMS-1500 Claim Form for a Provider Office

ELFABRIO® (pegunigalsidase alfa-iwxj)

Billing for reimbursement or administration of ELFABRIO in the provider office setting should be submitted on the CMS-1500 manual claim form, or its electronic equivalent 837P (Professional).[27]

Use HCPCS code J3590 for ELFABRIO and bill one (1) unit regardless of dosage.

Additional Claim Information: Some payers may require additional detail about the drug administered. **Item 19**

TIP: When completing a claim form for a drug that does not have a permanent HCPCS code, check with the payer on their requirements. It may be required to include the 11-digit NDC, the drug name (brand and generic), drug strength, total dosage, total wastage (if applicable), and route of administration.

Diagnosis/Nature of Illness or Injury: Enter the clinically appropriate ICD-10-CM code. **Item 21**

Prior Authorization Number: Enter the prior authorization number if one was issued. **Item 23**

Date of Service: Enter the appropriate date for each procedure, service, or supply. Some payers require that the shaded area of 24A includes the NDC, and the appropriate qualifier (N4) and NDC units (UN, ML). **Item 24A**

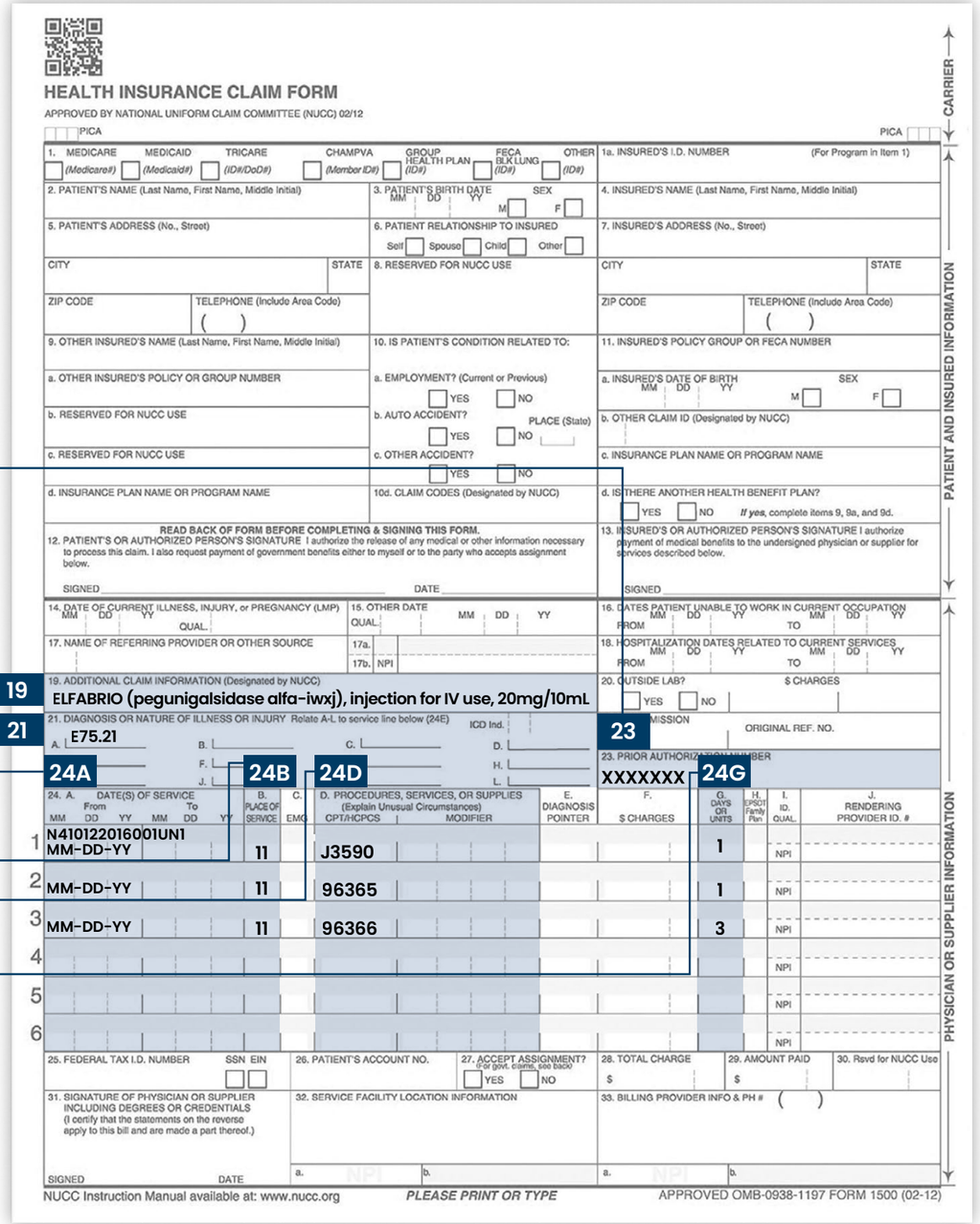
Place of Service: Enter the appropriate site of care code indicating where the medical service was rendered. **Item 24B**

Procedures, Services, or Supplies: Enter the appropriate CPT and HCPCS codes and modifiers, if required. **Item 24D**

Days or Units: Enter the number of units that were administered. **Item 24G**

TIP: When billing with a miscellaneous J-code, enter 1 unit, regardless of dosage.

Providers should contact the health plan for questions about coverage, coding, and payment. Specific direction from the plan supersedes the codes included here.



HEALTH INSURANCE CLAIM FORM

APPROVED BY NATIONAL UNIFORM CLAIM COMMITTEE (NUCC) 02/12

PICA

1. MEDICARE (Medicare#) MEDICAID (Medicaid#) TRICARE (ID#/DoD#) CHAMPVA (Member ID#) GROUP HEALTH PLAN (ID#) FECA BLK LUNG (ID#) OTHER (ID#)

1a. INSURED'S I.D. NUMBER (For Program in Item 1)

2. PATIENT'S NAME (Last Name, First Name, Middle Initial)

3. PATIENT'S BIRTH DATE MM DD YY SEX M F

4. INSURED'S NAME (Last Name, First Name, Middle Initial)

5. PATIENT'S ADDRESS (No., Street)

6. PATIENT RELATIONSHIP TO INSURED Self Spouse Child Other

7. INSURED'S ADDRESS (No., Street)

CITY STATE

8. RESERVED FOR NUCC USE

CITY STATE

ZIP CODE TELEPHONE (Include Area Code) ( )

ZIP CODE TELEPHONE (Include Area Code) ( )

9. OTHER INSURED'S NAME (Last Name, First Name, Middle Initial)

10. IS PATIENT'S CONDITION RELATED TO:

11. INSURED'S POLICY GROUP OR FECA NUMBER

a. OTHER INSURED'S POLICY OR GROUP NUMBER

a. EMPLOYMENT? (Current or Previous) YES NO

a. INSURED'S DATE OF BIRTH MM DD YY SEX M F

b. RESERVED FOR NUCC USE

b. AUTO ACCIDENT? YES NO PLACE (State)

b. OTHER CLAIM ID (Designated by NUCC)

c. RESERVED FOR NUCC USE

c. OTHER ACCIDENT? YES NO

c. INSURANCE PLAN NAME OR PROGRAM NAME

d. INSURANCE PLAN NAME OR PROGRAM NAME

10d. CLAIM CODES (Designated by NUCC)

d. IS THERE ANOTHER HEALTH BENEFIT PLAN? YES NO If yes, complete items 9, 9a, and 9d.

READ BACK OF FORM BEFORE COMPLETING & SIGNING THIS FORM.

12. PATIENT'S OR AUTHORIZED PERSON'S SIGNATURE I authorize the release of any medical or other information necessary to process this claim. I also request payment of government benefits either to myself or to the party who accepts assignment below.

SIGNED DATE

13. INSURED'S OR AUTHORIZED PERSON'S SIGNATURE I authorize payment of medical benefits to the undersigned physician or supplier for services described below.

SIGNED

14. DATE OF CURRENT ILLNESS, INJURY, or PREGNANCY (LMP) MM DD YY QUAL.

15. OTHER DATE QUAL. MM DD YY

16. DATES PATIENT UNABLE TO WORK IN CURRENT OCCUPATION MM DD YY FROM TO MM DD YY

17. NAME OF REFERRING PROVIDER OR OTHER SOURCE

17a.

17b. NPI

18. HOSPITALIZATION DATES RELATED TO CURRENT SERVICES MM DD YY FROM TO MM DD YY

19. ADDITIONAL CLAIM INFORMATION (Designated by NUCC)
ELFABRIO (pegunigalsidase alfa-iwxj), injection for IV use, 20mg/10mL

20. OUTSIDE LAB? YES NO $ CHARGES

21. DIAGNOSIS OR NATURE OF ILLNESS OR INJURY Relate A-L to service line below (24E) ICD Ind.
A. E75.21 B. C. D. F. H. J. L.

22. RESUBMISSION ORIGINAL REF. NO.

23. PRIOR AUTHORIZATION NUMBER
XXXXXXX

24. A. DATE(S) OF SERVICE From MM DD YY	To MM DD YY	B. PLACE OF SERVICE	C. EMG	D. PROCEDURES, SERVICES, OR SUPPLIES (Explain Unusual Circumstances) CPT/HCPCS	MODIFIER	E. DIAGNOSIS POINTER	F. $ CHARGES	G. DAYS OR UNITS	H. EPSDT Family Plan	I. ID. QUAL.	J. RENDERING PROVIDER ID. #
1 N410122016001UN1 MM-DD-YY		11		J3590				1		NPI	
2 MM-DD-YY		11		96365				1		NPI	
3 MM-DD-YY		11		96366				3		NPI	
4										NPI	
5										NPI	
6										NPI	

25. FEDERAL TAX I.D. NUMBER SSN EIN

26. PATIENT'S ACCOUNT NO.

27. ACCEPT ASSIGNMENT? (For govt. claims, see back) YES NO

28. TOTAL CHARGE $

29. AMOUNT PAID $

30. Rsvd for NUCC Use

31. SIGNATURE OF PHYSICIAN OR SUPPLIER INCLUDING DEGREES OR CREDENTIALS (I certify that the statements on the reverse apply to this bill and are made a part thereof.)

SIGNED DATE

32. SERVICE FACILITY LOCATION INFORMATION

a. NPI b.

33. BILLING PROVIDER INFO & PH # ( )

a. NPI b.

NUCC Instruction Manual available at: www.nucc.org PLEASE PRINT OR TYPE APPROVED OMB-0938-1197 FORM 1500 (02-12)

CARRIER

PATIENT AND INSURED INFORMATION

PHYSICIAN OR SUPPLIER INFORMATION

Important Safety Information (continued)

The most common adverse reactions (≥15%) were infusion-associated reactions, nasopharyngitis, headache, diarrhea, fatigue, nausea, back pain, pain in extremity, and sinusitis.

Sample CMS-1450 (UB-04) Claim Form for the Institutional or Hospital Outpatient Setting

ELFABRIO® (pegunigalsidase alfa-iwxj)

Billing for reimbursement or administration of ELFABRIO in the institutional or hospital outpatient setting should be submitted on the CMS-1450 (UB-04) manual claim form, or its electronic equivalent 837I (Institutional).[28]

Use HCPCS code J3590 or C9399 (in the hospital outpatient setting) for ELFABRIO and bill one (1) unit regardless of dosage.

Revenue Code: Enter the appropriate revenue code based on the cost center and service provided. **FL 42**

Revenue Description: Enter a narrative description or standard abbreviation for each corresponding revenue code. **FL 43**

HCPCS & Procedure Codes: Enter the appropriate HCPCS and CPT codes. **FL 44**

Service Units: Enter the number of times a single procedure or item was performed or provided for the date of service. **FL 46**

TIP: When billing with a miscellaneous HCPCS code, enter 1 unit, regardless of dosage.

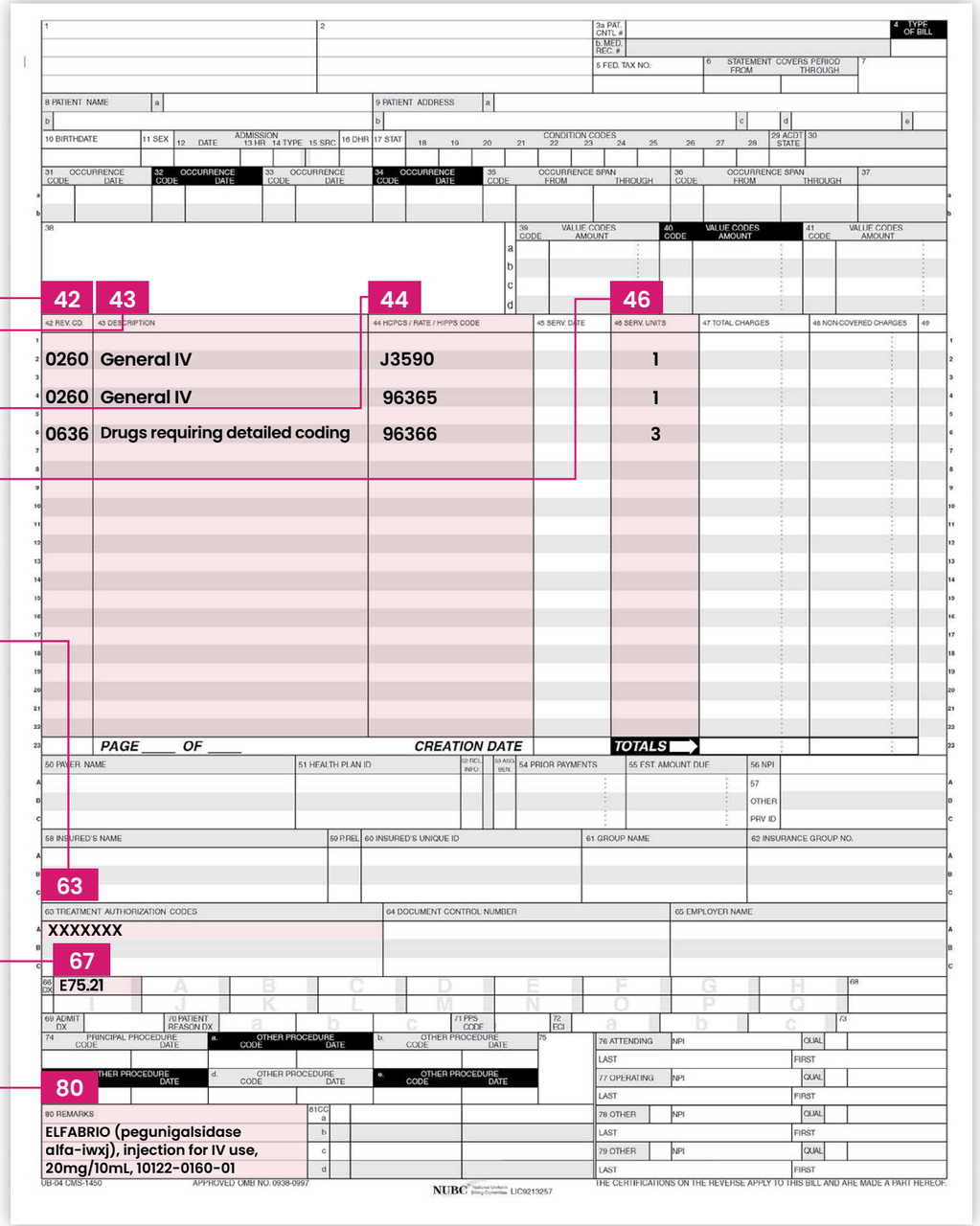
Prior Authorization Number: Enter the prior authorization number if one was issued. **FL 63**

Diagnosis (DX) Code: Enter the clinically appropriate ICD-10-CM code. **FL 67**

Remarks: Some payers may require additional information about the drug administered. **FL 80**

TIP: When completing a claim form for a drug that does not have a permanent HCPCS code, check with the payer on their requirements. It may be required to include the 11-digit NDC, the drug name (brand and generic), drug strength, total dosage, total wastage (if applicable), and route of administration.

Providers should contact the health plan for questions about coverage, coding, and payment. Specific direction from the plan supersedes the codes included here.



Important Safety Information (continued)

A case of membranoproliferative glomerulonephritis with immune depositions in the kidney was reported during clinical trials. Monitor serum creatinine and urinary protein-to-creatinine ratio. If glomerulonephritis is suspected, discontinue treatment until a diagnostic evaluation can be conducted.

Enroll Your Patient in Chiesi Total Care Today

ELFABRIO® (pegunigalsidase alfa-iwxj)

Chiesi TOTAL care

BY PHONE
1-833-656-1056

BY FAX
1-636-355-3610

HOURS OF OPERATION
7:00 AM - 6:00 PM (Central Time)

For more information, visit chiesitotalcare.com

Please see accompanying Full Prescribing Information for Elfabrio and full Terms and Conditions for additional Chiesi Total Care eligibility requirements.

References:

1. McCain J. The importance of a thorough benefits investigation to help navigate medical vs pharmacy benefit. National Association of Medication Access & Patient Advocacy. Accessed April 23, 2023. https://namapa.org/medical-vs-pharmacy-benefit. **2.** Academy of Managed Care Pharmacy. Guide to pharmaceutical payment methods, 2013 update, version 3.0. Published March 15, 2019. Accessed April 23, 2023. https://www.amcp.org/Resource-Center/guide-pharmaceutical-payment-methods. **3.** Academy of Managed Care Pharmacy. Formulary management. Published November 2009. Updated July 18, 2019. Accessed April 23, 2023. https://www.amcp.org/about/managed-care-pharmacy-101/concepts-managed-care-pharmacy/formulary-management. **4.** Center for Medicare Advocacy. Part D/prescription drug benefits. Accessed April 23, 2023. https://medicareadvocacy.org/medicare-info/medicare-part-d/. **5.** American Medical Association. Tips to help physicians reduce the prior authorization burden in their practice. Published 2015. Accessed April 23, 2023. https://www.ama-assn.org/sites/ama-assn.org/files/corp/media-browser/premium/psa/prior-authorization-tips_0.pdf. **6.** Centers for Medicare & Medicaid Services. Exceptions. Published December 1, 2021. Accessed April 23, 2023. https://www.cms.gov/Medicare/Appeals-and-Grievances/MedPrescriptDrugApplGriev/Exceptions. **7.** National Association of Insurance Commissioners. What is medical necessity. Accessed May 2, 2023. https://content.naic.org/sites/default/files/consumer-health-insurance-what-is-medical-necessity.pdf. **8.** Patient Advocate Foundation. A patient's guide to navigating the insurance appeals process. Accessed April 23, 2023. https://www.patientadvocate.org/wp-content/uploads/sites/8/NavigatingInsuranceAppeals.pdf. **9.** Medicare Interactive. The Medicare prescription drug coverage and your rights notice. Accessed May 2, 2023. https://www.medicareinteractive.org/get-answers/medicare-denials-and-appeals/part-d-appeals/the-medicare-prescription-drug-coverage-and-your-rights-notice. **10.** Medicare Interactive. Prescription drug coverage (Parts A, B, and D). Medicare Rights Center. Accessed April 23, 2023. https://www.medicareinteractive.org/get-answers/medicare-covered-services/medicare-coverage-overview/prescription-drug-coverage-parts-a-b-and-d. **11.** Noridian Healthcare Solutions. Local coverage determination (LCD). Published October 12, 2022. Accessed April 23, 2023. https://med.noridianmedicare.com/web/jddme/policies/lcd. **12.** US Department of Health & Human Services. What's the difference between Medicare and Medicaid? Published December 8, 2022. Accessed April 23, 2023. https://www.hhs.gov/answers/medicare-and-medicaid/what-is-the-difference-between-medicare-medicaid/index.html. **13.** American Academy of Professional Coders. What is HCPCS? Accessed May 2, 2023. https://www.aapc.com/resources/medical-coding/hcpcs.aspx. **14.** Centers for Medicare & Medicaid Services. Healthcare Common Procedural Coding System (HCPCS) level II coding procedures. Published April 21, 2022. Accessed April 23, 2023. https://www.cms.gov/Medicare/Coding/MedHCPCSGenInfo/Downloads/2018-11-30-HCPCS-Level2-Coding-Procedure.pdf. **15.** Noridian Healthcare Solutions. Unlisted and not otherwise classified code billing. Published December 9, 2022. Accessed April 23, 2023. https://med.noridianmedicare.com/web/jeb/topics/claim-submission/submission-errors-solutions/unlisted-procedure-and-noc-codes. **16.** Find-A-Code. CPT code set: 96365, 96366; HCPCS procedure and supply codes: C9399, J3490; ICD-10-CM diagnosis codes E75.21. Accessed April 21, 2023. **17.** WPS Government Health Administrators. Billing and describing not otherwise classified (NOC) codes. Updated July 16, 2020. Accessed May 2, 2023. https://www.wpsgha.com/wps/portal/mac/site/claims/guides-and-resources/not-otherwise-classified-billing **18.** UnitedHealthcare. National Drug Code (NDC) requirement policy, professional and facility. Published 2023. Accessed April 23, 2023. https://www.uhcprovider.com/content/dam/provider/docs/public/policies/comm-reimbursement/COMM-National-Drug-Code-Requirement-Policy.pdf. **19.** ELFBRIO Prescribing Information DRAFT. Accessed May 2, 2023. **20.** American Academy of Professional Coders. What is ICD-10? Published May 20, 2021. Accessed April 23, 2023. https://www.aapc.com/icd-10/. **21.** Medical billing and coding certification. Accessed April 23, 2023. http://www.billing-coding.com/pdf/mbacc_ebook_full_pages.pdf. **22.** Novitas Solutions. Modifier 59: what you need to know. Published July 26, 2021. Accessed April 23, 2023. https://www.novitas-solutions.com/webcenter/portal/MedicareJH/pagebyid?contentId=00144545. **23.** Centers for Medicare & Medicaid Services. Medicare program: discarded drugs and biologicals — JW modifier and JZ policy frequently asked questions. Accessed April 23, 2023. https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/HospitalOutpatientPPS/Downloads/JW-Modifier-FAQs.pdf. **24.** Centers for Medicare & Medicaid Services. Place of service codes. Published December 1, 2021. Accessed April 23, 2023. https://www.cms.gov/Medicare/Coding/place-of-service-codes. **25.** E2E Medical Billing Services. What is revenue code? Published November 12, 2019. Accessed April 23, 2023. https://www.e2emedicalbilling.com/blog/what-is-revenue-code/. **26.** Noridian Healthcare Solutions. Revenue codes. Published June 28, 2022. Accessed April 23, 2023. https://med.noridianmedicare.com/web/jea/topics/claim-submission/revenue-codes. **27.** National Uniform Claim Committee. 1500 health insurance claim form reference instruction manual for form version 02/12. Published July 2022. Accessed April 23, 2023. https://www.nucc.org/images/stories/PDF/1500_claim_form_instruction_manual_2022_07-v10a.pdf. **28.** Centers for Medicare & Medicaid Services. Medicare claims processing manual chapter 25: completing and processing the form CMS-1450 data set. Published August 6, 2021. Accessed April 23, 2023. https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c25.pdf.

Chiesi

Chiesi is a specialty pharmaceutical company with a long history of bringing innovative products to the healthcare marketplace. We are deeply committed to developing products for rare diseases such as Fabry disease.

For more information, visit hcp.elfabrio.com.
Chiesi Total Care℠ is offered through EVERSANA® Life Science Services Specialty Pharmacy.

©Chiesi USA, Inc. 2023. All rights reserved. Elfabrio® is a registered trademark of CHIESI FARMACEUTICI S.p.A. Chiesi Total Care℠ is a service mark of CHIESI FARMACEUTICI S.p.A. EVERSANA® is a registered trademark. All other trademarks are the property of their respective owners.
PP-EF-0326 V2.0