ELFABRIO®▼ (PEGUNIGALSIDASE ALFA)

IMPORTANT INFORMATION:

on minimising the risk of hypersensitivity reactions and medication errors in home settings

Information for Healthcare Professionals

This educational material is provided by Chiesi Limited as part of the Risk Management Plan for Elfabrio

This booklet fulfills the conditions of the marketing authorisation and has been approved by the HPRA.
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Adverse events should be reported to HPRA Pharmacovigilance, Website: www.hpra.ie. Adverse events should also be reported to Chiesi Limited on 1800 817459 (IE) or PV.UK@Chiesi.com. ▼ This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse reactions.

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LIST OF ABBREVIATIONS

ADR	Adverse drug reaction
CVAD	Central venous access device
ERT	Enzyme replacement therapy
HCP	Healthcare professional
HPRA	Health Products Regulatory Authority
IRR	Infusion-related reaction
IV	Intravenous
PF	Pre-Filled (Heparin syringe)
SmPC	Summary of Product Characteristics

1. OBJECTIVE OF THE HCP BROCHURE

The objective of this document is to:

a) Provide guidance to healthcare professionals (HCPs) for the selection and management of patients suitable for receiving pegunigalsidase alfa at home.

b) Provide relevant information for the HCP to train the patient and/or caregiver(s) to administer pegunigalsidase alfa at home. (See also the document "Information for Patients, Caregivers and Healthcare Professionals").

1.1. Role and Responsibility of the Treating Physician

It is the responsibility of the treating physician to ensure a safe administration to the patient in the home environment, as well as to choose the dosage regimen.

For this, he/she will:

- Initiate and supervise all necessary administrative actions which will allow the other parties involved to proceed (patient and/or caregiver(s), home infusion nurse, pharmacist or other HCPs, as per local implementation of the home infusion therapy).
- Evaluate patient's eligibility to receive home infusion therapy.
- Ensure that a HCP is available at all times during the home infusion and a specified time after infusion, as per national regulations.
- Review regularly the Logbook (see Appendix 9.3) and make sure that all medical instructions regarding dose and infusion frequency and rate, pre-medication and special considerations, as well as emergency actions and treatment, are clearly documented and up to date.
- Regularly monitor the home-infused patient with regards to both disease and infusions.
- Ensure that a clear, rapid and reliable line of communication is available to expedite emergency response in case immediate medical attention is required during the home infusion (treating physician's and caregiver's contact and number).
- Ensure that the patient and caregiver(s) are trained on the practical aspects of the home infusion: administration of the pre-infusion medication, preparation and administration of the infusion, recognition of signs of potential reactions and management of them. Training will be recorded in the Logbook and proper educational material will be distributed to the patient/caregiver(s).
- Ensure that proper (if prescribed) pre-infusion treatment (e.g. antihistamines, corticosteroids), as well as emergency treatment and equipment, is provided to the patient/caregiver(s).

2. ASSESSING ELIGIBILITY FOR HOME INFUSION

Administration of pegunigalsidase alfa at home may be considered for patients who are tolerating their infusions well and have no history of moderate or severe infusion-related reactions (IRRs) for a few months. The decision to transfer pegunigalsidase alfa treatment to the patient's home setting is made by the treating physician and should consider patient preferences and medical status.

The following primary criteria for home infusion must be fulfilled by the patient:

2.1 Checklist for Eligibility for Home Infusion

- ☐ Infusion of pegunigalsidase alfa at home may be considered if the patient is tolerating their infusions well and has no history of moderate or severe IRRs for a few months.
- ☐ The decision to move to home infusion should be made after evaluation and recommendation by the treating physician.
- ☐ Home infusion infrastructure, resources, and procedures, including training, must be established and available to the home infusion nurse in charge of home infusion.
- ☐ The home infusion nurse should be available at all times during the home infusion and for a specified time after infusion.
- ☐ Appropriate training should be given by the treating physician and/or home infusion nurse to the patient and/or caregiver prior to initiation of home infusion.

- ☐ The dose and infusion rate used in the home setting should remain the same as was used in the hospital setting; they should be changed only under the supervision of the treating physician.
- ☐ For patients switching to pegunigalsidase alfa 2 mg/kg every 4 weeks from pegunigalsidase alfa 1 mg/kg every 2 weeks, the first dose of the changed dosing regimen needs to be administered in a hospital/clinical setting.

3. REQUIREMENTS AND ORGANISATION OF HOME INFUSION

Once the patient has been eligible for home infusion based on the primary criteria, a set of requirements must be considered to ensure that pegunigalsidase alfa infusions can be safely, efficiently, and reliably delivered at the patient's home.

3.1 Checklist for Home Infusion Organisation

- ☐ The patient and/or caregiver(s) have been informed by the treating physician about the treatment to be provided at home, the associated risks, and the provision of medical assistance at home, and agrees to the treatment at home.
- ☐ The patient and/or caregiver(s) have an understanding of the illness and have been trained to recognise possible adverse events, including IRRs and understand the procedure to be followed in case they occur (i.e., notify symptoms suggestive of adverse drug reactions [ADRs] to the HCP for proper assessment and management).
- ☐ The home environment must be conducive to home infusion therapy including a clean environment with electricity, water, telephone access, refrigeration, and physical space to support storage of pegunigalsidase alfa and other infusion supplies.
- ☐ Ensure that a HCP is available at all times during the home infusion and a specified time after infusion, as per national regulations. The patient has been informed that the infusion should always be administered in the presence of the caregiver adequately trained on how to manage in case of ADRs, IRRs and medication errors in agreement with the local requirements for the implementation of the home infusion therapy. The patient should not be alone at home, but with a caregiver capable of stopping the infusion and giving the alert in the event of an IRR.

3.2 Drug and Infusion Equipment

Treatment medicine, pre-medication and emergency treatment and supplies, as well as all necessary equipment will be provided to the patient's home according to local arrangements and regulations (hospital/pharmacy to the patient or to a third party with the appropriate prescription).

Transport from the pharmacy/warehouse must comply with the following details of the transport chain as well as compliance with the following activities:

- Temperature control of drug during transport from pharmacy/warehouse to patient's home.
- The temperature monitoring device must be checked to confirm the drug experienced no temperature deviation during the shipping process (it is considered a deviation if temperature <2 or >8 °C).

PRODUCT - VIALS OF PEGUNIGALSIDASE ALFA (20 MG PER 10 ML VIAL OR 5 MG PER 2.5 ML VIAL)

Vials will be provided as a liquid in clear glass 10 ml or 2.5 ml vials closed with coated rubber stoppers and sealed with aluminium flip off cap. They must be stored in a clean refrigerator at a temperature of between +2 °C and +8 °C. Do not freeze or shake.

INFUSION EQUIPMENT

- IV Pole
- Infusion pump
- Bio-waste container
- Alcohol wipes
- Non-sterile gloves
- 30 ml syringe
- 2 x Needle free valves
- 2 x 0.9% Sodium chloride 10 ml syringes
- IV catheter/Huber/extension set (as needed)
- IV Start Kit/Central Line Kit per access type
- Cadd In-line 0.2-micron IV tubing
- Vented vial access spike
- 18-gauge needle
- Tape
- 10 ml syringe
- 3 ml syringe
- Heparin 100 IU/ml PF 5 ml/12 ml syringe (for central lines only)
- Hibiclens
- Sodium chloride 0.9% IV bag(s) according to the dilution needs
- Emergency Kit
- Tourniquet
- Pretreatment medication (if applicable)

NOTE: This is the typical requirement for the infusion equipment but this may vary depending on local arrangements.

Where the home infusion nurse is not required for the entire course of the infusion, he/she will prepare and remove the infusion and will remain reachable by phone in a perimeter close to the patient's home. In these situations, the patient must be accompanied all the time with a caregiver capable of stopping the infusion and giving the alert in the event of an IRR.

3.3 Pre-infusion Treatment and Emergency Treatment

PRE-INFUSION TREATMENT

- Pre-treatment with antihistamines and/or corticosteroids, if administered in the hospital or other medical setting, must be provided based on the patient-specific prescription and should be described in the Logbook.
- This treatment must not be altered in the home setting, unless medically warranted at the discretion of the treating physician.

EMERGENCY TREATMENT

- Any patients experiencing adverse events during the home infusion need to immediately stop the infusion process and seek the attention of a HCP.
- The Irish national emergency number and/or the treating physician must also be called if an IRR occurs after completion of the infusion. Any IRR must be reported according to local rules and regulations.
- Emergency treatment must be provided based on the patient-specific prescription. (Please see Sections 6.4 and 6.5 for instructions on how to proceed in case of severe allergic or anaphylactic-type reactions during the infusion) and should be described in the Logbook. *Proper education on the use of emergency medications must be provided to the patient and/or caregiver(s).*
- An available, rapid and reliable line of communication must be ensured to expedite emergency response in case immediate medical attention is required as per indications included in the "Serious Allergic Reactions to pegunigalsidase alfa" section (Section 6.5) of this Brochure and the Logbook (Section 3.4).
- Should patients experience or should the home infusion nurse or caregiver(s) identify any ADR or any problem with the dilution and administration of pegunigalsidase alfa, they need to contact the treating physician and caregiver immediately. Subsequent infusions may need to occur in a hospital or other medical setting at the discretion of the treating physician and caregiver.
- Equipment and medications must be available to respond to an emergency, if necessary. They will be provided by the Home Infusion Organisation and/or treating physician (to be decided based on local requirements) and they will replace items prior to expiration. *Proper education on the use of emergency medications and material must be provided by the treating physician to the patient and/or caregiver(s).*

EMERGENCY KIT

Emergency Kit will consist of:

- Airway
- Ambu Mask
- Pulse Oximeter
- 1000cc Hartman or Lactated Ringer's
- Antihistamine, as prescribed by the treating physician
- Any additional items per the treating physician's order (i.e., adrenaline, methylprednisolone)
- 2 IV 0.2 µm filters

NOTE: This is the typical requirement for the Emergency Kit but this may vary depending on local arrangements.

This Emergency Kit will be provided in a locked Emergency Box.

In the event the patient experiences an adverse event during or shortly after the infusion, the procedures indicated in Sections 6.4 and 6.5 are to be followed.

Any patients experiencing adverse events during the home infusion need to immediately stop the infusion process and seek the attention of a HCP. Subsequent infusions may need to occur in a clinical setting.

The management of IRRs must be based on the severity of the reaction, and include slowing the infusion rate and treatment with medicinal products such as antihistamines, antipyretics and/or corticosteroids, for mild to moderate reactions.

If severe allergic or anaphylactic-type reactions occur, immediate discontinuation of pegunigalsidase alfa is recommended and current medical standards for emergency treatment are to be followed. Subsequent infusions may need to occur in a hospital or other medical setting.

Adverse events should be reported to Health Products Regulatory Authority (HPRA) Pharmacovigilance, *Website: www.hpra.ie.* Adverse events should also be reported to Chiesi Limited on 1800 817459 (IE) or *PV.UK@Chiesi.com* by the treating healthcare professional (reporting instructions are provided in this Brochure in Section 6.2 [Safety Reporting]).

3.4 The Logbook

The Logbook serves as a means of communication for all involved in administering pegunigalsidase alfa in the home-setting.

- The patient/caregiver(s) will record the findings and actions from the initial interview and all relevant information from subsequent visits in the Logbook.
- A resource contact list must be completed and available at home in the Logbook for the patient and/or caregiver(s).
- The Logbook must be kept at the patient's home and will be updated by the home infusion nurse/ patient/caregiver(s) each time pegunigalsidase alfa is administered.
- The patient must take the Logbook along to the hospital at each appointment and bring it home afterwards.
- In the Logbook, the treating physician clearly states the dose, the required infusion volume, infusion rate, as well as any changes. The treating physician clearly states what has to be done and which procedures are to be followed and what medications are to be administered in the event of a serious IRR in line with current medical standards for emergency treatment. The contact details of the treating physician and the Irish national emergency number are documented in the Logbook.
- The pegunigalsidase alfa dose required, frequency, volume, infusion rate, pre-medication, emergency medication, as well as any changes will be determined by the treating physician. The prescription must be described in the Logbook (Appendix 9.3). Any changes to this prescription (dose or infusion rate) must again be reported in the Logbook. It is important to keep this guide handy and review the method of administration regularly. This will ensure optimal practice as well as an effective way of communicating with the treating physician.

4. TRAINING ON PREPARING AND ADMINISTERING PEGUNIGALSIDASE ALFA

The initial training of infusion administration and periodic update training is considered a fundamental activity to ensure treatment compliance and patient safety.

In principle, the initial instructions will be given in the hospital and the level of support required from the home infusion nurse in the home setting will be discussed and agreed by the treating physician and the patient and/ or caregiver(s).

The home infusion nurse will carry out the entire procedure for the first infusions at the patient's home.

Subsequently, should the patient then prefer to carry out the procedure with the assistance of a caregiver, the following conditions must be followed:

- The patient and/or caregiver(s) will receive adequate training from the home infusion nurse on how the infusion is being prepared and administered. The home infusion nurse will explain and demonstrate the complete infusion procedure to the patient and/ or caregiver(s), including training in hand hygiene, proper disinfection and aseptic handling when preparing the infusion.
- At subsequent visits, the home infusion nurse will be present to assist, if required, until the patient and/or caregiver(s) feels confident with the entire infusion procedure.
- While diluting and administering pegunigalsidase alfa, the procedures described in the ELFABRIO® Summary of Product Characteristics (Appendix 9. 1) and in Section 5 "Administration of pegunigalsidase alfa" of this document must be adhered to, and each administration of pegunigalsidase alfa should be recorded in the Logbook (Appendix 9.3). In case of any problems with the dilution and administration of pegunigalsidase alfa, the patient or caregiver(s) should contact the home infusion nurse or treating physician to determine appropriate action before starting or continuing with the infusion as detailed in the Logbook.

The infusion should always be administered in the presence of a caregiver knowledgeable about the infusion procedures and adequately trained on how to handle the situation in case of an IRR and medication errors, as assessed by the treating physician or home infusion nurse.

5. ADMINISTRATION OF PEGUNIGALSIDASE ALFA

Instructions for use relating to the dilution and administration can be found in the Summary of Product Characteristics (SmPC, Appendix 9.1).

5.1 Preparations

NOTE: The instructions for use (dilution and administration) can be found in the SmPC (Appendix 9.1). A detailed description is provided in this document.

Maintain strict asepsis while performing all preparation activities

1. Prepare a clean, flat, work area and lay out the requisites.
2. Keep the provided Emergency Kit nearby during the infusion.

Verify if the number of vials received is correct and the temperature monitoring device shows the correct information.

NOTE: IF THERE IS AN ALARM ON THE TEMPERATURE LOG, DO NOT START THE INFUSION. CALL THE HOME INFUSION ORGANISATION CONTACT IMMEDIATELY FOR FURTHER INSTRUCTION.

3. Check lot numbers, expiration dates (do not use pegunigalsidase alfa after the labelled expiry date), and current prescription, then remove the correct number of boxes to prepare the prescribed dose. Vials are for single use only.
4. Allow the required number of vials to reach room temperature prior to dilution (approximately 30 minutes). Ensure vials of pegunigalsidase alfa have been allowed to warm to room temperature. Do not heat vials with hot water or in the microwave.
5. Wash hands with soap and water.
6. Prepare the infusion bag provided to initiate the process.
7. Remove the vials of pegunigalsidase alfa from their boxes, inspect vials. Do not use if cap is missing or broken. Do not use if medication is discoloured or contains particulate matter.

5.2 Dilution of pegunigalsidase alfa

The recommended dose should be diluted in 0.9% sodium chloride, to a total volume based on patient body weight. The recommended dose and infusion volume are detailed in the Logbook (Appendix 9. 3).

NOTE: In some specific cases, pegunigalsidase alfa may be prepared at the pharmacy and shipped (in a cooler box) under temperature-controlled conditions (2 – 8 °C) with a temperature monitoring device to the patient's location for administration.

1. Remove the protective aluminium flip off cap from the pegunigalsidase alfa vials, and aseptically wipe each rubber seal with an alcohol pad, using one pad for each vial, and allow to dry.
2. Wipe the injection port of the IV bag of 0.9% sodium chloride with an alcohol pad and allow to dry.
3. Attach an 18-gauge needle to the needle free valve.
4. Remove needle cap and insert the needle into the IV bag injection port.
5. Secure the connection of the needle-free valve to injection port of the IV bag with tape.
6. Cleanse the valve with a new alcohol pad and allow to dry completely.
7. Prior to adding pegunigalsidase alfa to the 0.9% sodium chloride IV bag, an equal volume of sodium chloride must be removed from the IV bag.

Example (for once every 2 weeks):

- *Patient weight is 80 kg*
- *Patient prescribed dose is 1 mg/kg = 80 mg*
- *Pegunigalsidase alfa vial concentration is 20 mg/ 10 ml or 5 mg/2.5 ml (2 mg/ml)*
- *An 80 kg patient would receive 40 ml of pegunigalsidase alfa and need 40 ml of sodium chloride removed from the IV bag prior to adding pegunigalsidase alfa*

Example (for once every 4 weeks):

- *Patient weight is 80 kg*
- *Patient prescribed dose is 2 mg/kg = 160 mg*
- *Pegunigalsidase alfa vial concentration is 20 mg/ 10 ml or 5 mg/2.5 ml (2 mg/ml)*
- *An 80 kg patient would receive 80 ml of pegunigalsidase alfa and need 80 ml of sodium chloride removed from the IV bag prior to adding pegunigalsidase alfa*

8. Attach 30 ml syringe to needle free valve/clave and remove appropriate amount of 0.9% sodium chloride from IV bag, discard in the trash.
9. Attach a vented vial access spike to a sterile 10 ml syringe (and 3 ml syringe as needed).
10. Remove the protective cap of the vented vial access spike. While holding the vial of pegunigalsidase alfa firmly on the table, insert the spike into the centre of the rubber seal.
11. Invert the vial and withdraw the contents into the syringe.
12. Unscrew the syringe from the spike and attach the syringe directly from the needle free valve at the injection port of the IV bag. Slowly inject the medication into the IV bag. Do NOT inject in the airspace within the infusion bag.
13. Reattach the syringe to the spike and remove the spike from the empty vial. Now insert it in to the next vial of pegunigalsidase alfa, while maintaining aseptic technique.
14. Repeat these steps until the total calculated dose of pegunigalsidase alfa has been transferred into the IV bag.

NOTE: calculated volume may require removal of less than maximum volume (10 ml or 2.5 ml - depending on which vial used) from the last vial used for the infusion (partial vial use).

15. Remove the needle free valve and 18-gauge needle from the injection port and dispose of in the bio-waste receptacle.
16. Discard all pegunigalsidase alfa vials in the bio-waste container and document any amount of medication discarded in the Logbook.
17. Gently invert IV bag to mix the solution, avoiding vigorous shaking or agitation.

5.3 Administration

Diluted solutions of pegunigalsidase alfa should be used immediately. If not used immediately, in-use storage times and conditions would normally not be longer than 24 hours in the refrigerator (2°C – 8°C) or 8 hours if stored below 25 °C.

If medication cannot be used during these time frames it must be discarded. In such case, IMMEDIATELY CONTACT the treating physician's emergency line.

Time of preparation should be the time when the infusion preparation is finished and ready to be administered to the patient.

The pegunigalsidase alfa dose, frequency, infusion rate, as well as any changes, will be determined by the treating physician. The treatment must not be altered in the home setting, unless medically warranted at the discretion of the treating physician.

Infusion will be administered intravenously (IV) using a pre-programmed pump over a specified time period. The pump may be pre-set by the treating physician's team before the first home infusion.

NOTE: Settings on the pump will remain the same as programmed infusion settings. Monitor the pump screen display that indicates the amount infused. Note it in the Logbook (Appendix 9.3).

- Remove the protective cap from the 0.2-micron Cadd administration tubing spike and insert into the infusion port of the IV bag containing pegunigalsidase alfa.
- Hang IV bag on IV pole and attach Cadd Cassette to pump.

- Obtain IV access.
- Prime the tubing and connect to the patient to start infusion. DO NOT prime fluid with the tubing connected to the patient.
- Ensure medication is administered at infusion rate per orders.
- The patient should be sat down and relaxed while the infusion takes place.
- Should any alarm occur, resolve the problem as per pump specific instructions:
  - In case of "air in line", stop the infusion, disconnect the line from the patient and gently tap the line to move all bubbles close to the end of the line (to limit any drug wasting) and prime the line to ensure all air is removed.
  - In case of "down occlusion alarm" check patency of the infusion line and cannula. If the needle or cannula is occluded, do not flush; instead place a new needle or cannula in a different insertion point and remove the occluded cannula.
- In the case of a hypersensitivity reaction to the medication or emergency, refer to Section 3.3 "Pre-infusion Treatment and Emergency Treatment" and Sections 6.4 and 6.5.
- The pump will alarm at the end of the infusion. An empty infusion bag indicates the end time of infusion and the start time of the clinical observation period. (see Section 5.4)

NOTE: Do not remove the IV access at this time.

- Flush the infusion line with 20 ml of saline.
- Once the pump indicates 20 ml has been infused, manually stop the pump.
- Remove the infusion tubing from the patient's IV canula or central venous access device (CVAD).

NOTE: The IV access should remain in place throughout the end of infusion monitoring period.

- Note: At the end of the infusion, *all IV bags and administration tubing* can be disposed of into the household trash unless contaminated with visible blood. *Contaminated tubing and IV needles* should be disposed of into the bio-waste container.

5.4 Observation Period

- The patient should be observed for IRRs for two hours after the infusion.

6. PEGUNIGALSIDASE ALFA SAFETY INFORMATION

The clinical status of patients receiving home infusions should be monitored according to standard of care.

For patients switched to pegunigalsidase alfa 2 mg/kg body weight once every 4 weeks, regular monitoring (e.g.: after 3, 6, 12, 18 and 24 months) should be performed. Monitoring should include at least the evaluation of lyso-Gb3, renal (eGFR, proteinuria), cardiac (LVMi, NT-proBNP, troponin or ECG), and biochemical parameters. A change in any individual parameter should be interpreted in the context of the patient's overall clinical status, and clinically relevant deterioration should prompt re-evaluation of the treatment regimen.

Please refer to Section 4 of the current Summary of Product Characteristics (link provided in Appendix 9.1) for complete information on the safety of pegunigalsidase alfa.

6.1 Safety Procedures

Pegunigalsidase alfa has been shown to have good tolerability. However, IRRs, including hypersensitivity reactions, cannot be ruled out. For this reason, emergency management procedures are described in Sections 6.4 and 6.5. However, the home infusion nurse is a HCP with the ability to manage enzyme replacement therapy (ERT) and medical emergencies and is trained by the treating physician or the company responsible for the home infusion therapy, as per local clinical practice, at the beginning of his/her participation. pegunigalsidase alfa will also be closely monitored for evidence of any ADRs involving treated patients, following required safety procedures. The emergency treatment and reporting procedures to follow, in accordance with clinical standards and current legislation, are indicated in the following subsections.

6.2 Safety Reporting

The patient/caregiver or the home infusion nurse should inform the treating physician if an ADR/IRR occurs in a patient treated with pegunigalsidase alfa in the home infusion setting. Should an anaphylactoid reaction occur during or after the infusion, the home infusion nurse/caregiver(s) must immediately call the treating physician. Anaphylactoid reactions which require immediate contact of the treating physician are reported in the "Serious Allergic Reactions to pegunigalsidase alfa" section (Section 6.5).

In addition, if the patient/caregiver or the home infusion nurse becomes aware that a mistake was made in the preparation and/or administration of pegunigalsidase alfa, the patient or home infusion nurse should inform the treating physician to determine appropriate action. Any healthcare professional is then responsible for reporting any suspected adverse reaction, including medication errors, via the national reporting system according to the current local regulation. The record and reporting of medication errors ensure that systematic and recurring problems can be recognised and consecutive actions performed within the vigilance system.

6.3 Possible Type of Reactions to pegunigalsidase alfa

Pegunigalsidase alfa has been shown to have good tolerability, however, being an IV protein product, hypersensitivity reactions including severe ones cannot be ruled out and these are commonly known as IRRs.

IRRs defined as any related adverse events with onset after start of infusion and up to 2 hours after end of infusion have been reported (see also Section 4.8 of SmPC).

The most commonly observed symptoms of IRRs were hypersensitivity, itching, nausea, dizziness, chills and muscular pain. As with any IV protein product, allergic-type hypersensitivity reactions may manifest and can include localised angioedema (including swelling of the face, mouth, and throat), bronchospasm, hypotension, generalised urticaria, dysphagia, rash, dyspnea, flushing, chest discomfort, pruritus, and nasal congestion.

6.4 Management of Adverse Drug Reactions to pegunigalsidase alfa

Any patients experiencing adverse events during the home infusion need to immediately stop the infusion process and seek the attention of a HCP. Subsequent infusions may need to occur in a clinical setting.

The management of IRRs must be based on the severity of the reaction, and include slowing the infusion rate and treatment with medicinal products such as antihistamines, antipyretics and/or corticosteroids, for mild to moderate reactions. Pre-treatment with antihistamines and/or corticosteroids may prevent subsequent reactions in those cases where symptomatic treatment was required, although IRRs occurred in some patients after receiving pre-treatment (see section 4.2 of the SmPC).

If severe allergic or anaphylactic-type reactions occur, immediate discontinuation of pegunigalsidase alfa is recommended and current medical standards for emergency treatment are to be followed.

In patients who have experienced severe hypersensitivity reactions during pegunigalsidase alfa infusion, caution should be exercised upon re-challenge and appropriate medical support should be readily available. Moreover, for patients who experienced severe hypersensitivity reactions with ERT infusion including pegunigalsidase alfa, appropriate medical support should be readily available.

6.5 Serious Allergic Reactions to pegunigalsidase alfa

Allergic-type hypersensitivity IRRs can be severe, therefore, appropriate medical support should be readily available when pegunigalsidase alfa is administered.

In clinical studies, 5 patients over a total of 141, experienced a severe hypersensitivity reaction and recovered within the day after of occurrence with appropriate treatment (also see section 4.8 of SmPC)

If a severe allergic or anaphylactic-type reaction occurs, immediate discontinuation of pegunigalsidase alfa is recommended and current medical standards for emergency treatment are to be followed.

7. CALL FOR REPORTING

Reporting suspected adverse reactions is important. It allows continued monitoring of the benefit/risk balance of the medicinal product. Adverse events should be reported to HPRA Pharmacovigilance, *Website: www.hpra.ie. Adverse events should also be reported to Chiesi Limited on 1800 817459 (IE) or PV.UK@Chiesi.com.*

If the patient, caregiver or home infusion nurse becomes aware that a mistake was made in the preparation and/or administration of the drug, they should inform the treating physician to determine appropriate action. Any medication errors should be reported as a spontaneous report to the treating physician.

8. FURTHER INFORMATION

Please refer to the Summary of Product Characteristic (Appendix 9.1) for complete indication statements and further information about the approved use of pegunigalsidase alfa. Other detailed information on pegunigalsidase alfa is available at the following website: The European Medicines Agency (EMA) *(see http://www.ema.europa.eu).*

9. APPENDICES

Appendix 9.1 – ELFABRIO® Summary of Product Characteristics

The SmPC for pegunigalsidase alfa can be accessed at the HPRA website (https://www.hpra.ie/) and at medicines information online (https://www.medicines.ie/).

Appendix 9.2 - Adverse Event Form

Chiesi	Adverse Event Form	ELFABRIO® (pegunigalsidase alfa)	
Patient initials			
Country			
Date of birth (DD/MM/YY)			
Age			
Sex	MALE ☐	FEMALE ☐	
Weight (Kg)			
Reaction onset (DD/MM/YY)			
Event seriousness: Check all that applies		YES	NO
	Death	☐	☐
	Involved or prolonged inpatient hospitalisation	☐	☐
	Involved persistent or significant disability or incapacity	☐	☐
	Life threatening	☐	☐
	Other		
The outcome of the event may be captured as:	Ongoing	☐	☐
	Recovered/resolved	☐	☐
	Not recovered/not resolved	☐	☐
	Unknown	☐	☐
Description of reaction (as reported by HCP)			
Information(s) on pegunigalsidase alfa			
Dose and posology			
Route(s) of administration			
Treatment start (DD/MM/YY)			
Treatment stop (DD/MM/YY)			
Reaction abated after treatment stop	YES ☐	NO ☐	
Concomitant drugs			
Other relevant information			

Appendix 9.3 - Logbook

This Logbook is designed to assist with the administration of pegunigalsidase alfa. The Logbook should be kept as a record of when pegunigalsidase alfa is administered. Included within the Logbook is relevant information such as contact details for relevant healthcare professionals (HCPs) associated with the administration of pegunigalsidase alfa and how and where to report adverse drug reactions (details of which are included on the front and back covers of this Logbook).

If you require additional/replacement copies of the Logbook or if you, or one of your healthcare team have any questions, please contact Chiesi at **medinfo.uk@chiesi.com** or **+44 161 488 5555**.

Checklist for Home Infusion Organisation

- ☐ The patient and/or caregiver(s) have been informed by the treating physician about the treatment to be provided at home, the associated risks, and the provision of medical assistance at home, and agrees to the treatment at home.
- ☐ The patient and/or caregiver(s) understand the illness and have been trained to recognise possible adverse events, including infusion-related reactions (IRRs) and understand the procedure to be followed in case they occur (i.e., notify symptoms suggestive of adverse drug reactions [ADRs] to the HCP for proper assessment and management).
- ☐ The home environment must be conducive to home infusion therapy including a clean environment with electricity, water, telephone access, refrigeration, and physical space to support storage of pegunigalsidase alfa and other infusion supplies.
- ☐ Ensure that a HCP is available at all times during the home infusion and a specified time after infusion, as per national regulations. The patient should not be alone at home, but with a caregiver capable of stopping the infusion and giving the alert in the event of an IRR. The patient has been informed that the infusion should always be administered in the presence of the caregiver adequately trained on how to manage in case of ADRs, IRRs and medication errors in agreement with the local requirements for the implementation of the home infusion therapy.

Logbook for pegunigalsidase alfa Home Infusion

General data (*to be completed by treating physician*)

Emergency number:		
CONTACT DETAILS		
Patient	Name:	
	Date of birth:	
	Address:	
	City / Postcode:	
	Telephone:	
Patient's caregiver contact details	Name:	
	Address:	
	City / Postcode:	
	Telephone:	
Home infusion nurse	Name:	
	Organisation:	
	Address:	
	City / Postcode:	
	Telephone:	
Treating physician	Name:	
	Hospital:	
	Address:	
	City / Postcode:	
	Telephone:	
	Emergency number:	
Pharmacy	Name:	
	Address:	
	City / Postcode:	
	Telephone:	
Irish national emergency number		

Administration details *(to be completed by treating physician)*

Pegunigalsidase alfa administered since	Date (dd-mm-yyyy):	
First pegunigalsidase alfa infusion at home	Date (dd-mm-yyyy):	
Pegunigalsidase alfa dosing regimen		
- Weight (kg)		
- Dose (mg)		
- Frequency (once every 2 weeks or once every 4 weeks) and day of the week		
- Number of pegunigalsidase alfa vials to be used	10 ml vials	2.5 ml vials
- Volume of pegunigalsidase alfa to be used (ml)		
- Volume of 0.9% sodium chloride to be removed and discarded (ml)		
- Minimum total volume to be infused (ml) based on body weight <70 kg - 150 ml 70-100 kg - 250 ml >100 kg - 500 ml		
- Rate of infusion (ml/hr)		
Pre-treatment medication (if applicable)		
Reasons for pegunigalsidase alfa infusion at home		
Findings and actions from the initial interview		
Indicate support to be provided by home infusion nurse		

Infusion session form

(To be complete at each infusion session)

- The patient and/or caregiver(s) have been informed about the associated risks of home infusion of pegunigalsidase alfa, and proper education on the use of emergency medications has been provided.
- The management of infusion-related reactions (IRRs) should be based on the severity of the reaction. For mild to moderate reactions, the infusion rate may be reduced and, if needed, medications may be administered to help manage symptoms, such as antihistamines, antipyretics, and/or corticosteroids to control inflammation/ immune response. If severe hypersensitivity reactions (including anaphylaxis) occur, the infusion should be stopped immediately and urgent medical attention sought.
- Any patients experiencing adverse events during the home infusion need to immediately stop the infusion process and seek the attention of a healthcare professional. Subsequent infusions may need to occur in a clinical setting.

Date of Infusion	Date (dd-mm-yyyy)	
Patient's general health status - Describe any new health issues that you are currently experiencing prior to infusion, if any		
Dose (mg) and frequency (once every 2 weeks or once every 4 weeks)		
Required pegunigalsidase alfa volume (ml)		
Number of pegunigalsidase alfa vials used	10 ml vials	2.5 ml vials
Preparation completion time		
Start time of infusion		
Duration of administration		
Infusion rate (ml/hr)		
End time of Observation Period		
Problems/Remarks related to the infusion, if any (including infusion related reaction(s), action taken, and outcome)		
Name of person responsible for infusion, and date - home infusion nurse - caregiver (if different from above)		

Notes

Adverse events should be reported to HPRA Pharmacovigilance, Website: www.hpra.ie. Adverse events should also be reported to Chiesi Limited on 1800 817459 (IE) or PV.UK@Chiesi.com.
▼ This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse reactions.

Chiesi
global rare diseases

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This booklet fulfills the conditions of the marketing authorisation and has been approved by the HPRA. IE Version No: 3.0 September 2026

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