ELFABRIO®▼ (PEGUNIGALSIDASE ALFA)

IMPORTANT INFORMATION
on minimising the risk of hypersensitivity reactions and medication errors in home settings
Information for Patients, Caregivers and Healthcare Professionals

Please read this guide thoroughly and carefully before using this medicine as it contains important information for you.

The processes presented in this document serve as overall guidance but are subject to local medical practise and national rules and regulations

Version No: 2.0 August 2024

▼ This medicinal product is subject to additional monitoring. This will allow quick identification of new safety information. Please report suspected adverse drug reactions (ADRs) to the MHRA through the Yellow Card scheme. You can report via:
• the Yellow Card website www.mhra.gov.uk/yellowcard
• the free Yellow Card app available from the Apple App Store or Google Play Store
Alternatively you can report a suspected side effect to the Yellow Card scheme by calling 0800 731 6789 for free, Monday to Friday between 9am and 5pm. You can leave a message outside of these hours.

When reporting please provide as much information as possible. By reporting side effects, you can help provide more information on the safety of this medicine.

Alternatively, suspected adverse events can be reported to Chiesi Limited on 0800 0092329 (UK) or PV.UK@Chiesi.com

For patients and caregivers:

With this Guide, please also read the Patient Information Leaflet carefully. Your medical team will also provide you with additional training and support to help you.

- Keep this guide throughout the duration of home infusions. You may need to reread it.
- Ask your doctor (Treating Physician), nurse or other healthcare professional who assists or supports you if you have any further questions.
- Talk to your doctor (Treating Physician), nurse or other healthcare professional if you experience any side effects. This includes any possible side effects, even if not listed in this guide.

CONTENT

TABLE OF CONTENTS

1. OBJECTIVES AND GOALS

The objective of this document is to provide guidance to assist patients, caregivers and healthcare professionals in the administration of pegunigalsidase alfa. Administration of pegunigalsidase alfa at home may be considered for patients who are tolerating their infusions well.

The decision to transfer pegunigalsidase alfa treatment to the patient's home setting is made by the Treating Physician according to patient preferences and medical status.

An additional objective of this document is to inform patients and caregivers of the associated risks of adverse reactions from the use of pegunigalsidase alfa including how to recognise signs and symptoms of infusion-related reactions (IRRs) and hypersensitivity/anaphylactic reactions and the actions to take should these occur (see Section 6.2.1 of this document for further information on IRRs and hypersensitivity reactions including anaphylactic reactions).

2. ASSESSING ELIGIBILITY FOR HOME INFUSION

Home infusion of pegunigalsidase alfa may be considered for patients who tolerate their infusions well. The home infusion will take place under the responsibility of the Treating Physician. The Treating Physician overseeing the patient's clinical care must determine if the patient meets the following primary criteria for transfer of hospital-based infusion therapy to the patient's home setting:

- The patient is clinically stable and in good general clinical condition in a hospital setting
- The patient is tolerating their infusions well and has no history of moderate or severe IRRs for a few months
- The patient has a proven history of adherence to the previous infusion schedule in hospital

3. REQUIREMENTS AND ORGANISATION OF HOME INFUSION

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

Appropriate training should be given by the Treating Physician and/or nurse to the patient and/or caregiver prior to starting home infusion therapy.

3.1 Treating Physician

3.1.1 General

Before home infusions can start, the Treating Physician and medical team will ensure the following has been undertaken:

- The patient and/or caregiver(s) have been informed by the Treating Physician about the treatment to be provided at home and the associated risks and must agree to receive the treatment at home
- The patient and/or caregiver(s) understand the illness and are able to recognise possible adverse drug reactions (ADR) and understand the procedure to be followed in case they occur
- The Treating Physician should confirm with the patient that the home environment is appropriate for home infusion therapy, such as including a clean environment, access to electricity, water, telephone, refrigeration and physical space to store pegunigalsidase alfa and/or other infusion supplies
- Patient/caregivers are trained to recognise IRRs and hypersensitivity reactions and take the appropriate measures including to immediately inform the Home Infusion Nurse if suspected
- The patient has been informed that the infusion should always be administered in the presence of the Home Infusion Nurse or an adult adequately trained in how to manage in case of ADRs, IRRs and medication errors.

3.1.2 Medical

- The decision to have a patient move to home infusion should be made after evaluation and recommendation by the Treating Physician. The patient must be physically and mentally able to undergo infusions at home
- The patient/caregiver must be able to understand and accept the implications of home infusion therapy

The Treating Physician is responsible for:

- All aspects related to the prescription of the therapy and evaluation of patients' eligibility to receive home infusion therapy, as well as all clinical aspects related to home treatment and patients' safety
- Selection of infusion rate and dose. The rate of pegunigalsidase alfa infusion that was tolerated by the patient in a hospital or other medical setting must not be changed in the home setting; they should be changed only under the supervision of the Treating Physician
- Regular monitoring of the home-infused patient with regards to both disease and infusions
- Ensuring a rapid and reliable line of communication is available in case immediate medical attention is required
- Observation of the patient for IRRs for two hours after the infusion
- Periodic evaluation of the treatment effects of pegunigalsidase alfa with discontinuation of treatment considered in cases where no clear benefits are observed.

3.2 Pharmacy and Infusion Equipment

Treatment and all necessary equipment will be provided and prescribed according to local arrangements and regulations. Any waste materials are disposed of in accordance with local requirements or collected and disposed of by a specialised company.

3.3 Home Infusion Nurse

The Home Infusion Nurse is qualified to give IV infusions and has been appropriately trained on the administration of pegunigalsidase alfa, and the actions to be taken should adverse events occur.

The Home Infusion Nurse will have a coordinating role together with the Treating Physician and the patient and/or caregiver(s) in organising the treatment at home, and establishing the level of support necessary in the patient's home.

The Home Infusion Nurse will be also responsible for:

- Strictly following the method of preparation and administration of pegunigalsidase alfa
- Strictly following the dose and infusion rate of pegunigalsidase alfa reported in the Treating Physician's prescription
- Recording each administration of pegunigalsidase alfa according to the Treating Physician's prescription in the Infusion Diary and sending a copy of each infusion session form to the Treating Physician
- In the event of an IRR, immediately take life saving actions should clinical signs of anaphylactic reaction occur during the visit, call the Treating Physician and record any action taken in the Infusion Diary.

Before starting the infusion, the Home Infusion Nurse has to perform the following activities:

- Check patients' vital signs
- Prepare the drug and proceed with the infusion following the instructions according to the vial used for the infusion.

For those countries where the Home Infusion Nurse stay is not permitted for the entire course of the infusion, he/she will prepare and remove the infusion and will remain reachable by phone close to the patient's home. **The patient should not be alone at home, but with an adult person capable of stopping the infusion and giving the alert in the event of an IRR.**

3.4 Pre-Treatment and Emergency Treatment

- Pre-treatment with antihistamines and/or corticosteroids may prevent subsequent reactions in cases where symptomatic treatment was required
- Pre-infusion treatment must not be altered in the home setting, unless at the discretion of the Treating Physician
- Emergency treatment must be provided based on the patient's prescription and should be described in the Infusion Diary. 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 in case immediate medical attention is required. Should patients experience or should the caregiver(s) identify any ADR or potential medication errors, they need to contact the Treating Physician and/or his/her assigned medical team or emergency services immediately

3.5 The Infusion Diary

- The Infusion Diary serves as a means of communication for all involved in administering pegunigalsidase alfa in the home-setting. The Infusion Diary is available as a standalone document in both print and electronic format
- A resource contact list must be completed in the Infusion Diary for the patient and/or caregiver(s) and the Home Infusion Nurse
- The Home Infusion Nurse/patient/caregiver(s) will record the findings and actions from the initial interview and all relevant information from subsequent visits in the Infusion Diary
- **The Infusion Diary 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 and/or caregivers must take the Infusion Diary along to the hospital at each appointment and bring it home afterwards
- In the Infusion Diary, the Treating Physician clearly states the dose (mg), the number of pegunigalsidase alfa vials to be used, the volume of 0.9% sodium chloride to be removed and discarded (ml), the minimum total volume to be infused (ml) based on body weight , the infusion rate (ml/hr), as well as any relevant additional information
- The Treating Physician should provide clear instructions, including the medications to be administered in the event of an IRR in line with current medical standards for emergency treatment The contact details of the Treating Physician and the national emergency number are included in the Infusion Diary.

4. TRAINING ON PREPARING AND ADMINISTERING PEGUNIGALSIDASE ALFA

Training on preparing and administering pegunigalsidase alfa is a fundamental activity to ensure treatment compliance and patient safety. In case of any problems with the dilution and administration of pegunigalsidase alfa, the Treating Physician must be contacted immediately to determine the appropriate action before starting or continuing with the infusion.

If the patient/caregiver(s) feel that the treatment is not effective, he/she should discuss this with the Treating Physician.

5. ADMINISTRATION OF PEGUNIGALSIDASE ALFA INFUSION BY HOME INFUSION NURSE

5.1 Prescription

The dose, the required diluted volume, the infusion rate, pre-medications, and emergency medications as well as any changes will be the total responsibility of the Treating Physician. The prescription must be documented in the patient's medical record and in the Infusion Diary as well as any change in the dose or infusion rate.

5.2 Supplies

Where possible and permitted by local regulations, the Home Infusion Nurse is responsible for bringing the following equipment to each home visit, if not kept at the patient's home:

1. IV Pole;
2. Infusion pump;
3. Bio-waste container;
4. Alcohol wipes;
5. Disposable gloves;
6. 30 ml syringe;
7. 2 x Needle free valves;
8. 2 x 0.9% sodium chloride 10 ml syringes;
9. IV catheter/Huber/extension set (as needed);
10. IV Start Kit/Central Line Kit per access type;
11. Cadd In-line 0.2-micron IV tubing;
12. Vented vial access spike;
13. 18-gauge needle;
14. Tape;
15. 10 ml syringe;
16. 3 ml syringe;
17. Heparin 100u/ml PVC 5ml/12ml syringe (for central lines only);
18. Hibiclens;
19. Sodium chloride 0.9% IV bag(s) according to the dilution needs;
20. Tourniquet.

Provision of supplies may vary according to country specific regulations and local requirements. In addition to the above equipment, the Home Infusion Nurse has medications for the pre-treatment and/or management of IRRs and ADRs, as specified in Table 1.

5.3 Preparation of pegunigalsidase alfa

1. Prepare a clean work area and place the material required;
2. Wash your hands with soap and water and put on disposable gloves;
3. Check with the Treating Physician the dose and number of pegunigalsidase alfa vials needed for the infusion and make sure they are at room temperature at the time of dilution. Do not heat vials with hot water or in the microwave.

5.4 Dilution of pegunigalsidase alfa

1. Carefully check each vial for any signs of damage and inspect the solution for any foreign particles or change in colour before dilution. If these are found, do not use the drug and inform the Treating Physician immediately. Please check the expiry date on each vial;
2. Remove the protective lids from the pegunigalsidase alfa vials, and aseptically wipe each rubber seal with an alcohol wipe, using one wipe for each vial, and allow to dry;
3. Wipe the injection port of the IV bag of 0.9% sodium chloride with an alcohol wipe and allow to dry;
4. Attach an 18-gauge needle to the needle free valve;
5. Remove needle cap and insert the needle into the IV bag injection port;
6. Secure the connection of the needle-free valve to injection port of the IV bag with tape;
7. Cleanse the valve with a new alcohol wipe and allow to dry completely;

8. 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;

   The Treating Physician will decide on the dose which should be followed

   Example:

   - 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

9. 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;
10. Attach a vented vial access spike to a sterile 10 ml syringe (and 3 ml syringe as needed);
11. 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 center of the rubber seal;
12. Invert the vial and withdraw the contents into the syringe;
13. Unscrew the syringe from the spike and attach the syringe directly to the needle free valve at the injection port of the IV bag. Slowly inject the medication in to the IV bag;
14. Reattach the syringe to the spike and remove the spike from the empty vial. Now insert it into the next vial of pegunigalsidase alfa, while maintaining aseptic technique;
15. 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);

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

Diluted solutions of pegunigalsidase alfa should be used immediately. If immediate use is not possible, the diluted solution may be stored for up to 24 hours in the refrigerator (2°C-8°C) or 8 hours at room temperature if stored below 25 °C.

If pegunigalsidase alfa cannot be used during these time frames it must be discarded. In such case, **IMMEDIATELY CONTACT** the Treating Physician's emergency line.

5.5 Administration of pegunigalsidase alfa with syringe pump

The pegunigalsidase alfa dose, 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 determined by the Treating Physician. The pump may be pre-set by the 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 Infusion Diary

- 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 6.3 and 6.4 within this booklet
- 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.

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 cannula or Central Venous Access Device

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.

5.5.1 Venous access device

When the patient has a venous access device for the delivery of pegunigalsidase alfa, the patient and/or caregiver(s) will be shown how to care for the device, under the instruction of the Treating Physician.

Proper home care of a venous access device involves regular irrigation with heparin to prevent clotting and attention to a sterile technique to keep the device free of infectious agents.

The patient and/or caregiver(s) will be informed of the following necessary steps under the guidance of the Treating Physician and according to local standard procedures:

- When in use, cover site with transparent occlusive dressing. No dressing is required when not in use
- Flush with 5 ml sodium chloride 0.9% solution before and after each use
- Flush with 5 ml heparin (100 U/ml) after each use.

5.6 Observation Period

- The patient should be observed for IRRs for two hours after the infusion
- Vital signs should be collected every 60 minutes until the observation duration has concluded, and again at the end time of the observation period
- Once the observation period is complete, disconnect the patient and remove the peripheral IV access (if used) according to local standard procedures and properly dispose of all used supplies in biohazard bag or sharps container as appropriate
- Additionally, the Home Infusion Nurse will call the patient one hour after the observation period to follow up on tolerability post infusion.

5.7 Preparation and Administration – Process Flow Diagram

Preparation of Infusion Bag

- Prior to starting the preparation, confirm that all equipment is available to prepare the infusion bag
- Receive drug vials in a temperature controlled shipment
- Confirm shipping container is sealed and intact and check the temperature monitoring device for any alarm
- Allow vials to sit at room temperature (30 minutes) and check they are colorless and free of any visible particles
- Record batch number and expiration date for each of the vials in the Infusion Diary
- Check the Infusion Diary for dose and diluent specifications
- Proceed with drug preparation as described in the instructions (in Sections 5.3 and 5.4)
- Record dose, and date/time of preparation in the Infusion Diary

Preparation for Administration of Drug Infusion

- Assess pre-medications
- Confirm contents and expiration dates of all items
- Confirm the infusion pump is charged (check battery status and inform the physician when replacement is needed)
- Confirm dose settings on the infusion pump. Collect pre-infusion vital signs (within 10 minutes before the infusion)
- Insert IV catheter or prepare central venous access

Administration of Drug Infusion

- Prime the line with saline. The prime volume may differ based on specific pump/tubing used
- Connect primed tubing to prepared infusion bag and to IV/central venous access
- Administer infusion of drug according to instructions. (as per Section 5.5) Record start time of infusion
- Collect vital signs every 30 minutes until end of infusion
- Monitor for signs of infusion reaction
- Empty infusion bag indicates end time of infusion and start time of clinical observation

Clinical Observation

- Flush the line with 20 ml of saline at the same infusion rate as the infusion
- Disconnect infusion tubing from the IV line
- Leave the cannula/central venous access patent
- Evaluate the infusion site to ensure there is no reaction
- Collect vital signs every 60 minutes and at the end of clinical observation
- Record end time of 2 hour observation period
- Properly dispose of all used supplies, according to local procedures, in biohazard bag or sharps container as appropriate

6. SAFETY INFORMATION

6.1 Safety Procedures

Pegunigalsidase alfa has been shown to have good tolerability. However, IRRs, including hypersensitivity reactions, cannot be ruled out. However, the Home Infusion Nurse is a healthcare professional with the ability to manage enzyme replacement therapy and medical emergencies.

Pegunigalsidase alfa will also be closely monitored for evidence of any ADRs involving treated patients, following required safety procedures. Emergency treatment and reporting procedures are provided 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) will take emergency life saving actions and must immediately call the Treating Physician.

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 the drug, the patient or infusion nurse should inform the Treating Physician to determine appropriate action.

6.2.1 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 infusion-related reactions (IRRs).

IRRs (defined as any related adverse events with an onset after the start of infusion and up to 2 hours after the end of the infusion) have been reported with the use of pegunigalsidase alfa (see also section 4 of the patient information leaflet or section 4.8 of the SmPC).

The most commonly observed symptoms of IRRs were hypersensitivity, itching, nausea, dizziness, chills and muscular pain. As with any intravenous protein product, allergic-type hypersensitivity reactions may occur and can include localised swelling of the face, mouth, and throat, coughing and wheezing, low blood pressure, itching, trouble swallowing, feeling short of breath, rash, flushing and nasal congestion.

6.3 Management of Adverse Drug Reactions to pegunigalsidase alfa

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

In the event of an IRR, the Home Infusion Nurse will take life saving actions and then immediately call the Treating Physician who will then provide the guidance to proceed, following the instructions indicated in Table 1.

6.4 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.

The first signs of an anaphylactic reaction (severe allergic reaction) mainly affect the skin and/or mucosa (swelling, itching, redness), while the more significant reactions involve the respiratory system (difficulty breathing, persistently clearing the throat, wheezing) or the cardiovascular system (low blood pressure).

Symptoms involving the gastrointestinal tract are also possible (abdominal cramps, vomiting, etc).

Symptoms may appear suddenly after a few hours although serious clinical manifestations generally occur within 30 minutes to 1 hour.

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. Severe reactions are generally managed with administration of antihistamines, corticosteroids, intravenous fluids, and/or oxygen, when clinically indicated. If the event is clearly anaphylaxis, then intramuscular epinephrine should be used.

After an anaphylactic reaction, patients should preferentially be observed in a safe environment, which will be decided by the attending healthcare professional under the instruction of the Treating Physician.

The following guidelines indicate the first aid procedures that should be used to manage a severe hypersensitivity reaction during home administration of the drug.

At the first signs of a reaction:

- Immediately stop administering the drug
- Maintain venous access with saline solution
- Place the patient in a comfortable position, preferably with the legs raised to prevent hypotension. If the patient has difficulty breathing, a seated position is preferable to lying down
- If the signs and symptoms are severe or deteriorate rapidly, take life saving actions, and then immediately call the Treating Physician who will then provide the guidance to proceed, following instruction reported in Table 1.
- Any action taken following an IRR must be documented in the Infusion Diary
- Drug supplies available to the Home Infusion Nurse will be managed according to local requirements and regulations.

The hypersensitivity symptoms and reaction and their recommended response reactions are shown in Table 1.

Table 1 – Actions to be taken based on hypersensitivity, allergic symptoms and reactions

Symptoms and Reactions	Recommended/Suggested Actions	Recommended/ Suggested Drugs
Mild: Pain in the head, feeling hot, sudden warmth, feeling unsteady, shaking	1. Reduce the infusion rate 25% to 50% or consider stopping the infusion if deemed appropriate; 2. Call the emergency treatment number and then the Treating Physician for instructions, including on any drugs to administer; 3. Reduce the infusion rate by a further 25% or consider stopping the infusion if deemed appropriate if the symptom persists 10 minutes after the first reduction in rate; 4. Call the Treating Physician again for instructions, including on any drugs to administer; 5. Stop the infusion if the symptom still persists after 10 minutes; 6. Complete home infusion **if symptoms resolved** and instructed by the Treating Physician. 7. Complete ADR form.	If instructed by the emergency medical services or Treating Physician, administer: • Paracetamol; or • Ibuprofen; or • Oral/IV antihistamines; or • Other drugs depending on nature of symptoms.
Moderate: Feeling like vomiting, heart beating too fast, pain or discomfort in the chest, widespread skin rash or hives, itchy skin, high blood pressure, very bad headache, vomiting, loose or watery stools, stomach cramps, indigestion or upset stomach, pain in muscles or joints	1. Stop administration, whilst maintaining venous access; 2. Call the emergency treatment number and then the Treating Physician for instructions, including on any drugs to administer; 3. Administer the prescribed therapy; 4. Inform the Treating Physician, requesting an assessment before the next home infusion; 5. Complete home infusion **if symptoms resolved** and instructed by the Treating Physician; 6. Complete ADR form.	If instructed by the Treating Physician and based on the nature of symptoms: • Oral/parenteral antihistamines; or • Oral/parenteral corticosteroids. • And other drugs as indicated by the Treating Physician
Severe: Low blood pressure, cold or clammy skin, a bluish discoloration of the skin, blue or grey lips or fingernails, difficulty in breathing or feeling out of breath, a high-pitched whistling sound when breathing, chest discomfort, irregular heartbeat, cough, confusion, fainting, extreme drowsiness and restlessness. Headache, difficulty swallowing, changes in the voice (such as hoarseness), hives, rash, flushing, itchiness, sneezing, nasal congestion, swelling under the skin - often around the eyes and lips; and severe allergic reaction causing swelling of hands, feet, ankles, face, lips, mouth or throat.	1. Stop administration immediately, whilst maintaining venous access; 2. Call the emergency number; 3. State that you are a nurse/patient/caregiver and describe the seriousness of the situation; 4. Provide the telephone number and address; 5. Ask for an ambulance to be sent immediately; 6. State that you are trained in first aid; 7. State that you have a first aid kit and seek advice on the appropriateness of practising first aid while waiting for the ambulance; 8. State the name of the administered drug(s); 9. If necessary, carry out resuscitation following BLS guidelines; 10. Inform the Treating Physician of the event; 11. Complete ADR form.	Treatment advised by emergency number based on the nature of symptoms: • IM adrenaline; or • Oral/parenteral antihistamines; or • Oral/parenteral corticosteroids; or • Beta-2 agonist spray or nebulised saline solution as per the clinical condition.

ADR = Adverse Drug Reaction; BLS = Basic Life Support; IM = Intramuscular; IV = Intravenous.

7. FURTHER INFORMATION

Please refer to the Summary of Product Characteristics for complete indication statements and further information about the approved use of pegunigalsidase alfa, which can be found here: https://www.medicines.org.uk/emc/product/14960/smpc. Further information on the use of pegunigalsidase alfa can also be found in the Patient Information Leaflet (PIL) which can be found here: https://www.medicines.org.uk/emc/product/14960/pil.

8. REPORTING ADVERSE EVENTS

Adverse events should be reported. Reporting forms and information can be found at www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store. Adverse events should also be reported to Chiesi Limited on 0800 0092329 (UK) or PV.UK@Chiesi.com.

Alternatively you can report a suspected side effect to the Yellow Card scheme by calling 0800 731 6789 for free, Monday to Friday between 9am and 5pm. You can leave a message outside of these hours. When reporting please provide as much information as possible. By reporting side effects, you can help provide more information on the safety of this medicine.

9. APPENDIX

9.1 The Infusion Diary

Infusion Diary for pegunigalsidase alfa Home Infusion.

A standalone version of the Infusion Diary for patients/caregivers is also available in both print and electronic format. The diary must be retained by the patient at home and infusion details should be shared with the Treating Physician.

General data (*to be completed by Treating Physician*)

Emergency number:

CONTACT DETAILS		
Patient	Name:	
	Date of Birth:	
	Address:	
	Postcode / City:	
	Telephone:	
Patient's caregiver contact details	Name:	
	Address:	
	Postcode / City:	
	Telephone:	
Home Infusion Nurse	Name:	
	Organisation:	
	Address:	
	Postcode / City:	
	Telephone:	
Treating Physician	Name:	
	Hospital:	
	Address:	
	Postcode / City:	
	Telephone:	
	Emergency number:	
Pharmacy	Name:	
	Address:	
	Postcode / City:	
	Telephone:	
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) Day of the week		
- Number of pegunigalsidase alfa vials to be used	10ml vials	2.5ml 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 infusion nurse at home		

Infusion session form

Infusion session form

(To be completed at each infusion session by Home Infusion Nurse)

- 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
- In the event of an IRR, consider slowing down or immediately discontinuing the infusion (depending on the severity of the reaction) and/or administering appropriate treatment following Treating Physician instructions.
- Necessary actions in the event of a serious IRR are described in Sections 6.3 and 6.4 of this document. Keep this information readily available during the infusion procedure.

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)		
Required pegunigalsidase alfa volume (ml)		
Number of pegunigalsidase alfa vials used	10ml vials	2.5ml vials
Start time of preparation of infusion		
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 - Nurse - Caregiver (if different from above)		

Notes

Notes

Notes

CHIESI LIMITED
Manchester Green, 333 Styal Road, Manchester, M22 5LG United Kingdom
Office: +441614885555
XI-ELF-2400038 August 2024