



Lamzede® (Velmanase Alfa) Home Infusion Therapy:

Information for Healthcare Professionals

A Guide for Healthcare Professionals Treating Patients with Alpha-Mannosidosis

**Essential Non-Promotional Information
Do not discard**

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

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LIST OF ABBREVIATIONS

ADR	Adverse drug reaction
AE	Adverse event
BLS	Basic life support
BP	Blood pressure
CVAD	Central venous access device
EMA	European Medicines Agency
ERT	Enzyme replacement therapy
GDPR	General Data Protection Regulation
HR	Heart rate
IM	Intramuscular
IRR	Infusion-related reactions
IV	Intravenous
PBLS	Paediatric basic life support
PICC	Peripheral inserted central catheter
PVC	Polyvinyl chloride
RR	Respiratory rate
sADR	Serious adverse drug reaction
SmPC	Summary of Product Characteristics
SpO ₂	Oxygen saturation

RELATED DOCUMENTATION

For more details, see the Summary of Product Characteristics (SmPC, [Appendix 9.1](#)).

1. OBJECTIVES AND GOALS

The objective of this document is to provide guidance to assist healthcare professionals on how to transfer Lamzede® (velmanase alfa) infusion therapy in patients with alpha-mannosidosis from the hospital setting to the patient's home.

This educational material will therefore be distributed in advance to all physicians who are currently administering Lamzede® as ERT for alpha mannosidosis to ensure:

- The therapy treatment is designed in compliance with hospital standards and shared with the Treating Physician, and the procedures operate under safe conditions;
- Full compliance with the infusion schedule allows for therapeutic adherence to treatment;
- All the components and actors that operate within this process are oriented towards satisfying the needs of the patients and his/her relatives to respect his/her quality of life, in compliance with safety standards.

The process encompassing the selection of eligible patients and a careful evaluation of all specifics regarding the organisation of home infusion will be described in detail below, starting with patient evaluation and selection and discussion of home infusion requirements, followed by home infusion organisation and training.

Once specific requirements are met, patients with alpha-mannosidosis may be offered home infusion therapy within the living environment.

Home infusion of Lamzede® may be considered for patients who tolerate their infusions well. The decision to transfer Lamzede® treatment to the patient's home setting is made by the Treating Physician and should consider patient preferences and medical status.

The home infusion will take place under the responsibility of the Treating Physician. Transfer to home care setting should be documented by the Treating Physician in the patient's medical record.

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

2. ASSESSING ELIGIBILITY FOR HOME INFUSION

Before any arrangements are made, the 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 for at least 12 infusions of Lamzede® in a hospital setting; a comprehensive evaluation must be completed before deciding on transfer of therapy.
- The patient has had no previous adverse drug reactions (ADRs) or infusion-related reactions (IRRs) during hospital treatment for at least the last three (3) infusions with or without pre-medication.
- The patient has a proven history of adherence to the previous infusion schedule in hospital.

Any particular clinical situation will be evaluated under the full responsibility of the Treating Physician.

3. REQUIREMENTS AND ORGANISATION OF HOME INFUSION

Once the patient has been considered to be eligible for home infusion based on the primary criteria, a set of requirements must be considered to ensure that Lamzede® 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 initiation of home infusion.

3.1 Patient

3.1.1 General

- The patient and/or caregiver(s) and/or legal tutors 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 must agree to receive the treatment at home;
- The patient and/or caregiver(s) understand the illness and are able to recognise possible ADRs and understand the procedure to be followed in case they occur (i.e., notify symptoms suggestive of ADRs to the healthcare professional for proper assessment and management);
- The home environment must be conducive to home infusion therapy, including a clean environment; access to electricity, water, telephone, refrigeration and physical space to support storage of Lamzede® and/or other infusion supplies;
- Patient/caregiver are trained to recognize IRRs and take the appropriate measures including to immediately inform the infusion nurse, in case the nurse is not at the patient's home for the entire course of the infusion;
- The patient has been informed that the infusion should always be administered in the presence of the Home Nurse/Infusion Nurse or an adult adequately trained in 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.

3.1.2 Medical

- The patient must be judged physically and mentally able to undergo infusions at home by the Treating Physician. The patient/caregiver/legal tutor must be able to understand and accept the implications of home infusion therapy;
- The patient has an easy access to blood veins or a CVAD or a PICC that allows adequate infusion;

3.2 Treating Physician

The Treating Physician is responsible for:

- The initiation of all the necessary administrative actions which will allow the other parties involved to proceed (patient and/or caregiver(s) and/or legal tutors, Home

Nurse/Infusion Nurse, pharmacist or other healthcare professionals as per local implementation of the home infusion therapy);

- 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 Lamzede® infusion that was tolerated by the patient in a more controlled setting (e.g., hospital or other medical setting) must not be changed in the home setting, unless necessary due to safety considerations. Any changes in Lamzede® administration must be clearly documented in the patient's medical record and in the Infusion Diary (Appendix 9.3);
- Regular monitoring (at least once every 6 months) of the home-infused patient with regards to both disease and infusions;
- Ensuring a rapid and reliable line of communication is available to expedite emergency response in case immediate medical attention is required.

3.3 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.

3.4 Home Nurse/Infusion Nurse

The Home Nurse/Infusion Nurse is qualified to give IV infusions and has been appropriately trained on the administration of Lamzede®, and is trained on the possible adverse events (including serious adverse events such as anaphylactoid reactions) and the actions to be taken should they occur.

The Home Nurse/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 will establish with the Treating Physician, patient and/or caregiver(s) the level of support necessary in the patient's home.

The Home Nurse/Infusion Nurse will be also responsible for:

- Strictly following the prescribed method of preparation and administration of Lamzede® as stated in [Section 5](#) of this manual and in the approved SmPC ([Appendix 9.1](#));
- Strictly following the prescribed dose and infusion rate of Lamzede® reported in the Treating Physician's prescription and as stated in the Infusion Diary (Appendix 9.3);
- Recording each administration of Lamzede® according to the Treating Physician's prescription in the Infusion Diary (Appendix 9.3) and sending a copy of each Infusion session form to the Treating Physician.
- In the event of an IRR, immediately call the Treating Physician and follow the "Emergency Treatment Plan" (Appendix 9.2), take life saving actions should clinical signs of anaphylactic reaction occur during the visit and record any action taken in the Infusion Diary (Appendix 9.3).

The country-specific 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.

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

- Check patients' vital signs and, if deemed appropriate, communicate the information to the Treating Physician to get authorisation for the administration of the drug;
- Prepare the drug as described in [Section 5.3](#) to [Section 5.5](#) of this manual and proceed with the infusion as described in [Section 5.6](#) of this manual following the instructions according to the via used for the infusion.

For those countries where the Home Nurse/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 in a perimeter 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.

3.5 Pre-Treatment and Emergency Treatment

- Pre-infusion treatment, if administered in the hospital or other medical setting (e.g., antihistamines, paracetamol, ibuprofen, corticosteroids), must be provided based on the patient-specific prescription and should be described in the Infusion Diary (Appendix 9.3). This treatment must not be altered in the home setting, unless medically warranted at the discretion of the Treating Physician;
- Emergency treatment must be provided based on the patient-specific prescription (see also [Appendix 9.2](#) “Emergency Treatment Plan” for instructions on how to proceed in case of emergency during the infusion) and should be described in the Infusion Diary (appendix 9.3). Proper education on the use of emergency medications must be provided to the patient and/or caregiver(s) and/or legal tutors;
- 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 “Emergency Treatment Plan” ([Appendix 9.2](#));
- Should patients experience or should the caregiver(s) identify any ADR or any problem with the reconstitution and administration of Lamzede[®], they need to contact the Treating Physician or his/her medical designee immediately. Subsequent infusions may need to occur in a hospital or other medical setting at the discretion of the Treating Physician or his/her medical designee.

3.6 The Infusion Diary

- The Infusion Diary serves as a means of communication for all involved in administering Lamzede[®] in the home-setting.
- The 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.
- A resource contact list must be completed and available at home in the Infusion Diary for the patient and/or caregiver(s) and the infusion nurse.

- The Infusion Diary must be kept at the patient's home and will be updated by the infusion nurse/patient/caregiver(s) each time Lamzede® 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 vials to be reconstituted with water, the total reconstituted volume (ml), the total volume to be infused (ml), 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 infusion related reaction IRR in line with current medical standards for emergency treatment.
- The contact details of the treating physician and the country-specific national emergency number are documented in the Infusion Diary

4. TRAINING ON PREPARING AND ADMINISTERING LAMZEDE® (VELMANASE ALFA)

The initial training of operators and their periodic updating is considered a fundamental activity to ensure treatment compliance and patient safety.

While reconstituting Lamzede®, the procedure described in the Lamzede® SmPC ([Appendix 9.1](#)) and in [Section 5](#) of this manual must be followed. Each administration of Lamzede® must be recorded according to local rules and regulations. In case of any problems with the reconstitution and administration of Lamzede®, the Treating Physician must be contacted immediately to determine the appropriate action before starting or continuing with the infusion.

If the patient/caregiver(s)/legal tutor feels that the treatment is not effective, he/she should consult the Treating Physician.

5. ADMINISTRATION OF LAMZEDE® INFUSION

Instructions relating to the reconstitution and administration can be found in the SmPC ([Appendix 9.1](#)). A detailed description is provided in [Section 5.3](#) to [Section 5.6](#).

5.1 Prescription

The dose, the required reconstituted 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 ([Appendix 9.3](#)) as well as any change in the dose or infusion rate.

It is important to keep this guide handy and review the method of administration regularly to ensure optimal practice.

5.2 Supplies

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

- Vital signs equipment (sphygmomanometer, stethoscope, pulse oximeter, thermometer);
- Syringe pump;
- Syringe pump tubing;
- Line for drug administration (if applicable);
- 0.22 µm filter;
- Water for injections;
- Vials of sterile sodium chloride 9 mg/mL (0.9%) solution;
- Disposable gloves;
- Sterile drape, cotton wool and gauze;
- Adhesive for cannula;
- 22G and 24G IV cannula depending on age and weight, if needed;
- 5 mL, 10 mL and 20 mL syringes;
- Skin disinfectant: chlorhexidine 2% or povidone-iodine 10% disinfectant;
- Hand disinfectant;
- Waste disposal container;
- Heparin of the required dilution (10, 100, 250 or 500 U/mL) (if applicable);
- Huber needle (if applicable);
- Ambu bag for adults and paediatric patients, with attached mask (if applicable);
- Guedel cannula for adult and paediatric use (if applicable).

Provision of supplies may vary according to country-specific regulations and local requirements.

In addition to the above equipment, the Home Nurse/Infusion Nurse has medications for the pre-treatment and/or management of IRRs and ADRs, as specified in [Section 0](#).

5.3 Preparation of Lamzede®

Note: the instructions for use (reconstitution, dilution and administration) can be found in the SmPC ([Appendix 9.1](#)). A detailed description is also provided in this section.

1. Prepare a clean work area and place the material required;
2. Wash your hands and put on disposable gloves;
3. Check with the Treating Physician the dose and number of Lamzede® vials needed for the infusion and make sure they are at room temperature at the time of the reconstitution.

5.4 Reconstitution and Dilution of Lamzede®

1. Carefully check each vial for any signs of damage and inspect the powder for any foreign particles or change in colour before reconstitution. If these are found, do not use the drug and inform the Treating Physician immediately;
2. Reconstitute the powder in the vial by slowly injecting 5 mL water for injections, gently directing the jet towards the inside wall of the vial and not directly onto the freeze-dried powder, to keep foaming to a minimum;
3. Place the reconstituted vials on the table then gently tilt and rotate each vial for 15 to 20 seconds to encourage the powder to dissolve. Reconstituted vials should never be

turned upside down, spun rapidly or shaken. The concentration in the vial should now be 2 mg/mL;

4. Inspect the solution for any particles or change in colour, immediately after reconstitution. The solution must be clear and must not be used if opaque particles or a change in colour is observed. Because of the nature of the medicine, the reconstituted solution may occasionally contain protein particles that look like thin white threads or translucent fibres, which will be removed by the in-line filter during the infusion;
5. Withdraw the reconstituted solution from each vial slowly and carefully to avoid foaming in the syringe. If the volume of solution exceeds the capacity of one (1) syringe, prepare the necessary number of syringes so that they can be quickly replaced during the infusion;
6. The reconstituted solution should be administered using an infusion set with a pump and a low protein binding 0.22 µm in-line filter. The total infusion volume is based on the patient's weight and should be administered over at least 50 minutes. In patients weighing < 18 kg, who will receive < 9 mL reconstituted solution, the infusion rate must be calculated so that the infusion time is ≥ 50 minutes. The maximum infusion rate is 25 mL/h;
7. The infusion volume and time can be calculated using Section 6.6 of the approved SmPC (Appendix 9.1) and should be recorded in the Infusion Diary (Appendix 9.3).

5.5 Filling the Infusion Line

1. Fill the infusion set fitted with a 0.22 µm filter with sodium chloride 0.9% solution and connect the filter to the syringe;
2. Replace the syringe containing sodium chloride 0.9% solution with the syringe containing the diluted drug;
3. Set the total volume to infuse and record it in the Infusion Diary (Appendix 9.3).

As the solution does not contain preservatives, it should be administered as soon as possible after reconstitution. In case of any delay or pause of the infusion, the solution should be stored at +2 to +8°C and used the same day, and it must not be mixed with other drugs.

The solution is single use. Therefore, any unused product or waste from preparation must be disposed of in accordance with local requirements.

5.6 Administration of Lamzede® with Syringe Pump

The Home Nurse/Infusion Nurse will administer Lamzede® with syringe pump, following the instructions according to the way used for infusion ([Section 5.6.1](#) to [Section 5.6.3](#)) and record each infusion in the Infusion Diary (Appendix 9.3).

Before starting to prepare the equipment required ensuring that some strips of tape are hanging ready for use and that the start of the infusion system is within reach. Place the chlorhexidine close by along with some gauzes

5.6.1 Via Peripheral Vein

1. Prepare the patient's arm; select the arm which has better venous access and proceed with venepuncture;

2. Once the vein is cannulated, check the stability of venous access, securing the needle or cannula wings with adhesive (hypoallergenic if necessary);
3. Enter the infusion rate in the pump display in accordance with the instructions received from the Treating Physician;
4. If the needle is displaced, stop the infusion, remove the needle and use another venous access point.

5.6.2 Via Central Venous Catheter

1. Advise the patient or caregiver of the position to take or how to position the patient;
2. Locate the septum of the reservoir by palpation;
3. Prepare the Huber needle, prefilling it with saline solution;
4. Disinfect the skin over the implant site with an appropriate antiseptic (repeat this procedure three times);
5. Insert the needle through the skin and the septum, until it reaches the reservoir floor;
6. Check if the catheter is working by performing a blood withdrawal;
7. Enter the infusion rate in the pump display in accordance with the instructions received from the Treating Physician.

5.6.3 Via Peripheral Inserted Central Catheter

1. Advise the patient or caregiver of the position to take or how to position the patient; Assess the catheter site by palpating it through the dressing or inspecting it visually if a transparent dressing has been used; If the patient shows signs of inflammation (swelling, redness, fever, etc.) on palpation contact the Treating Physician for an assessment;
2. Disinfect the hub before injecting into the infusion line;
3. Close with a needleless connector (neutral pressure) and close unused ports;
4. Disinfect the skin over the implant site with an appropriate antiseptic;
5. Connect the drug line to the needleless connector;
6. Prepare a syringe of 10 mL heparin solution (10-100-250-500 U/mL as prescribed or as indicated on the catheter);
7. Enter the infusion rate in the pump display in accordance with the instructions received from the Treating Physician.

5.7 Completing the Infusion Cycle

1. When the last syringe is empty, replace the dosing syringe with a 20 mL syringe filled with sodium chloride 0.9% solution for injection. Administer a volume of 10 mL sodium chloride solution into the infusion system to flush any velmanase alfa remaining in the infusion line;
2. At the end of the drug infusion and after at least 20 minutes of observation period, check the patient's vital signs before removing the needle in accordance with each method of administration. Report any clinical finding in the Infusion Diary (Appendix 9.3) before leaving patient's home.

5.7.1 For Patients Receiving Lamzede® Via Peripheral Vein

1. After the observation period, stop the pump and remove the needle following a sterile procedure;

2. Close the pump, disassemble the set and dispose of the syringe with the remaining sodium chloride 0.9% solution, the set and the needle following the required procedures.

5.7.2 For Patients Receiving Lamzede® Via Central Venous Catheter

1. After the observation period, stop the pump and unscrew the end of the tubing from the Huber needle;
2. Attach a 10 mL syringe containing a sodium chloride 0.9% solution or other solution as per local clinical practice, to the Huber needle tubing and administer the solution using the “positive pressure” manoeuvre;
3. Remove the Huber needle and apply a sterile dressing;
4. Disassemble the set and the vial with the remaining sodium chloride 0.9% solution and dispose of them following the required procedures.

5.7.3 For Patients Receiving Lamzede® Via Peripheral Inserted Central Catheter

1. After the observation period, remove the tubing from the needleless connector. Perform a pulse lavage with 20 mL saline solution, using two 10 mL syringes to inject small boluses of 1–2 mL saline solution and one (1) syringe of 10 mL heparin solution (10-100-250-500 U/mL as prescribed or as indicated on the catheter).
2. Stop the pump, remove the syringe with the remaining sodium chloride 0.9% solution and dispose of the syringe following the required procedures.

5.8 End of the Home Assistance Visit

After at least a 20 minutes observation period, from the end of infusion, the Home Nurse/Infusion Nurse completes patient observations, checking vital signs and, in case of any ADR/IRR or other safety concern, follows the indication included in the “Emergency Treatment Plan” ([Appendix 9.2](#)) and record any clinical finding in the Infusion Diary ([Appendix 9.3](#)) before leaving the patient’s home.

5.9 Home Care Suspension and/or Interruption

Home infusion treatment can be suspended at the discretion of the Treating Physician, who may decide to suspend and/or interrupt the treatment at home in case of a change of the clinical condition and/or changes in the organisational needs of the patient.

In such a case, the Treating Physician will discuss and agree with the patient/caregiver(s)/legal tutors the patient's therapeutic management at the clinical centre.

6. SAFETY INFORMATION

6.1 Safety Procedures

Lamzede® 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 [Section 6.3](#). However, the Home Nurse/Infusion Nurse is a healthcare professional with the ability to manage 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.

Lamzede® 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/care giver or the Home Nurse/Infusion Nurse should inform the Treating Physician if an ADR/IRR occurs in a patient treated with Lamzede® in the home infusion setting.

Should an anaphylactoid reaction occur during or after the infusion, the Home Nurse/Infusion Nurse must immediately call the treating physician. Anaphylactoid reactions which require immediate contact of the Treating Physician are reported in “Emergency Treatment Plan” ([Appendix 9.2, Table 1](#)).

In addition, if the patient/care giver or the Home Nurse/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.

The Treating Physician 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.

6.2.1 Possible Types of Reactions to Lamzede®

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

Infusion-related reactions (IRRs) are defined as those ADRs which occur during or after 2 hours from the end of the infusion of Lamzede® and that are assessed as being infusion-related.

During Lamzede® clinical development programme, the most common adverse reactions observed were weight increase (15%), IRRs (13%), diarrhoea (10%), headache (7%), arthralgia (7%), increased appetite (5%) and pain in extremity (5%).

The majority of these adverse reactions were non-serious.

IRRs, that were reported in 13% of the patients (5 out of 38 patients), included hypersensitivity, cyanosis, nausea, vomiting, pyrexia, chills, feeling hot, malaise, urticaria, anaphylactoid reaction and hyperhidrosis. All were mild or moderate in severity and 2 were reported as a serious adverse event. All patients who experienced IRRs recovered.

A total of 4 serious adverse reactions (loss of consciousness in 1 patient, acute renal failure in 1 patient, chills and hyperthermia in 1 patient) were observed. In all cases the patients recovered without sequelae.

6.3 Management of Adverse Drug Reactions to Lamzede®

In the event of an IRR, the Home Nurse/Infusion Nurse will activate the emergency procedure as required following the instructions provided in the “Emergency Treatment Plan” ([Appendix 9.2](#)) and, if necessary, administer the support medications according to the specific indication of the Treating Physician as reported in the Infusion Diary ([Appendix 9.3](#)).

The Home Nurse/Infusion Nurse is equipped with the drugs used to manage an ADR according to the specific protocol ([Section 0](#)).

How IRRs are managed should depend on the intensity of the reaction. Examples include slowing the infusion rate, treating the patient with drugs (such as antihistamines, antipyretics and/or corticosteroids) or interrupting administration and resuming treatment with a longer infusion time ([Table 2](#)[Error! Reference source not found.](#)).

6.3.1 Serious Allergic Reactions to Lamzede®

The first signs of an anaphylactic reaction mainly affect the skin and/or mucosa (erythema, redness, pruritus and angioedema), while those that put the patient’s life at risk generally involve the respiratory system (obstruction of upper and lower airways) or the cardiovascular system (hypotensive shock, cardiovascular collapse, cardiac arrhythmia, myocardial ischaemia). Symptoms involving the gastrointestinal tract are also possible (abdominal cramps, vomiting, etc.).

The earlier the onset, the more serious the reaction. Symptoms may appear suddenly a few hours after contact with the causal agent, although **serious** clinical manifestations generally occur **within 30 minutes to 1 hour**.

After an anaphylactic reaction, patients should preferentially be observed in a safe environment.

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:

1. Immediately stop administering the drug;
2. Maintain venous access with saline solution;
3. Place the patient in a comfortable position and, if possible, in the Trendelenburg position (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 2](#) of [Appendix 9.2](#). Drugs Available to the Home Nurse/Infusion Nurse. Any action taken following an IRR will be documented in the Infusion Diary ([Appendix 9.3](#)).

Drug supplies available to the Home Nurse/Infusion Nurse will be managed according to local requirements and regulations.

7. PRIVACY DATA MANAGEMENT

Sensitive patient data will be managed in accordance with the General Data Protection Regulation (GDPR) or any other specific local regulation.

8. FURTHER INFORMATION

Please refer to the SmPC ([Appendix 9.1](#)) for complete indication statements and further information about the approved use of Lamzede[®]. Other detailed information on Lamzede[®] is available at the European Medicines Agency (EMA) website (<https://www.ema.europa.eu/en/medicines/human/EPAR/lamzede>).

9. APPENDICES

9.1 Lamzede® Summary of Product Characteristics

9.2 Emergency Treatment Plan

Lamzede® has been shown to have good tolerability. However, IRRs, including hypersensitivity reactions or anaphylactic reactions, cannot be ruled out. Clinical signs and manifestations of anaphylaxis by system and severity are reported in [Table 1](#) below.

Table 1 - Assessment of the intensity of an anaphylactic reaction

	Skin	Gastrointestinal	Respiratory	Cardiovascular	Neurological
Mild	Sudden itching of eyes and nose, generalised pruritus, skin redness, urticaria	Itch/pruritus in mouth, nausea, mild abdominal pain	Nasal congestion and/or sneezing, rhinorrhoea, chest tightness	-	Change in behaviour, anxiety
Moderate	As above + angioedema	As above + swelling of lips, vomiting, cramping abdominal pain, diarrhoea	As above + hoarseness, barking cough, dysphagia, stridor, dyspnoea, wheezing	Tachycardia (increase by 15 bpm)	Sense of impending doom
Severe	As above	As above + faecal incontinence	As above + cyanosis or O ₂ saturation < 92%, respiratory arrest	Hypotension and/or collapse, arrhythmia, severe bradycardia and/or cardiac arrest	Confusion, loss of consciousness

bpm = Beats per minute.

Source: Muraro A, Roberts G, Clark A, et al. The management of anaphylaxis in childhood: position paper of the European academy of allergology and clinical immunology. *Allergy*. 2007;62(8):857-71.

If any of the above reported clinical signs of anaphylactic reaction occur during the visit, the Infusion Nurse will take live saving actions and then immediately call the Treating Physician who will then provide the guidance to proceed following the instructions indicated in [Table 2](#)**Error! Reference source not found.**

Any procedure must be confirmed by the Treating Physician should an event occur. No specific procedure will be conducted without the Treating Physician confirmation.

Table 2 - Actions to be taken based on hypersensitivity, allergic symptoms and reactions

Symptoms and Reactions	Recommended/Suggested Actions	Recommended/Suggested Drugs
Mild: Headache, fever, hot flushes, dizziness, tremor.	1. Reduce the infusion rate by 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 instructed by the emergency medical services or Treating Physician, administer: • Paracetamol: 15 mg/kg; dose can be repeated, if necessary, after 4-6 hours; or • Ibuprofen: 10 mg/kg; dose can be repeated, if necessary, after 6-8 hours; or • Oral/IV antihistamines; or • Other drugs depending on nature of symptoms.
Moderate: Nausea, tachycardia, chest pain, generalised rash and/or urticaria, pruritus, hypertension, severe headache, vomiting, diarrhoea, abdominal cramps, dyspepsia, muscle or joint pain.	1. Stop administration; 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 a new home infusion; 5. Complete home infusion..	If instructed by the Treating Physician and based on the nature of symptoms (see Section 6.3 for dosage and indications): • IM/IV antihistamines; or • IV corticosteroids.
Severe: Hypotension and shock, shortness of breath, wheezing, hypoxia, tachypnoea, bronchospasm, cough, laryngeal oedema, respiratory failure, cyanosis, cardiac arrhythmia, angioedema and anaphylactic reactions (swelling of hands, feet, ankles, face, lips, mouth or throat such as to cause difficulties swallowing or breathing)	1. Stop administration immediately; 2. Call the emergency number; 3. State that you are a nurse 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. If necessary, carry out resuscitation following BLS/PBLS guidelines; 9. Inform the Treating Physician of the event;	Treatment advised by emergency number based on the nature of symptoms (see text for dosage and instructions): • IM/IV adrenaline; or • Oral/IM/IV antihistamines; or • IV/oral corticosteroids; or • Beta-2 agonist spray; • Saline solution (in addition, if needed).

ADR = Adverse drug reaction; BLS = Basic life support; IM = Intramuscular; IV = Intravenous; PBLS = Paediatric basic life support.

9.3 The Infusion Diary

Infusion Diary for Lamzede® Home Infusion

General data (to be completed by treating physician)

Emergency number:

CONTACT DETAILS		
Patient	Name:	
	Date of Birth:	
	Address:	
	Zip / City:	
	Telephone:	
Patient's caregiver Contact details	Name:	
	Address:	
	Zip / City:	
	Telephone	
Infusion Nurse/Home Nurse	Name:	
	Organisation:	
	Address:	
	Zip / City:	
Treating Physician	Name:	
	Hospital:	
	Address:	
	Zip / City:	
	Telephone:	
	Emergency number	
Pharmacy	Name:	
	Address:	
	Zip / City:	
	Telephone:	

Administration details (to be completed by treating physician)

Lamzede® administered since	Date (dd-mmm-yyyy):
First Lamzede® infusion at home	Date (dd-mmm-yyyy):
Lamzede® dosing regimen	
- Dose (mg)	
- Frequency	
- Number of Lamzede® vials to be reconstituted with water	
- Total reconstituted volume (ml)	
- Total volume to be infused (ml)	
- Rate of infusion (ml/hr)	
Pre-treatment medication (if applicable)	
Reasons for Lamzede® infusion at home	
Findings and actions from the initial interview	
Indicate support to be provided by infusion nurse at home	

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 Lamzede®, and proper education on the use of emergency medications has been provided.
- In the event of any infusion-associated reaction, the **infusion must be immediately discontinued**.
- Necessary actions in the event of a serious infusion-associated reaction, **including emergency contact details**, are described in section 9.2 Emergency Treatment Plan of the Infusion Manual. Keep this information readily available during the infusion procedure.

Date of Infusion	Date (mm-dd-yyy)
Patient's general health status - Describe any new health issues that you are currently experiencing prior to infusion, if any	
Dose	
Number of Lamzede® vials to be reconstituted with water	
Total reconstituted volume (ml)	
- Total volume to be infused (ml)	
Duration of administration	
- Rate of infusion (ml/hr)	
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)	