Cerezyme® (imiglucerase) at Home

Manual for Patients with Gaucher Disease who Receive Home Infusion of Imiglucerase

Version 5

Read all of this information carefully before you start home infusion.

- Keep this information in an easily accessible place; you may need to read it again.
- If you have further questions, ask your treating physician.
- This medicine has been prescribed for you. Do not pass it on to others even if their symptoms are the same as yours as it may harm them.
- If you experience any side effects, you and/or your caregiver should notify your treating physician or homecare nurse. Please refer to the patient information leaflet (PIL) Cerezyme 400 U Powder for concentrate for solution for infusion for more 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.

Suspected side effects can also be reported to Sanofi: Tel: 0800 090 2314.
email: UK-drugsafety@sanofi.com

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ABBREVIATIONS

ADR: Adverse Drug Reaction

CVAD: Central Venous Access Device

IAR: Infusion Associated Reaction

IV: Intravenous

PIL: Patient Information Leaflet

1. YOUR DISEASE, TREATMENT AND HOME INFUSION

Together with your treating physician, you have decided to start home infusion therapy with imiglucerase. The objective of this document is to provide you with guidance on how to receive imiglucerase at home.

Gaucher disease and treatment

People with Gaucher disease have low levels of an enzyme called acid β-glucosidase. This enzyme helps the body to control the levels of glucosylceramide. Glucosylceramide is a natural substance in the body, made of sugar and fat. In Gaucher disease glucosylceramide levels can get too high inside specific cells called macrophages. When this happens, the cells are called "Gaucher cells". These large cells are mainly present in the bone marrow and organs like the spleen and the liver and can lead to disrupted function causing low number of blood cells, enlarged liver and spleen, and weaker bones. The presenting symptoms of Gaucher disease include pain in the bones and easy bruising or bleeding. Often the spleen and the liver are enlarged.

Imiglucerase is a synthetic enzyme which replaces the natural enzyme acid β-glucosidase which is lacking or not active enough in patients with Gaucher disease. Imiglucerase is used to treat patients who have a confirmed diagnosis of Type 1 or Type 3 Gaucher disease, who show signs of the disease.

Refer to the PIL of imiglucerase for additional information.

Home infusion

Currently, in some countries, people suffering from Gaucher disease, and treated with imiglucerase, receive their infusions at home. The decision to receive home treatment should be made by you and your treating physician after several months of hospital treatment to ensure satisfactory tolerance of the infusions.

Home infusion of imiglucerase will make it possible for you to do the following:

- Receive treatment within your own living environment.
- Be more flexible on the treatment timing.
- Avoid spending time travelling to and from the hospital and being hospitalised.
- Follow a normal schooling programme.
- Organise social and professional activities more easily.
- Facilitate arranging treatment around family and friends.

A homecare nurse, with the appropriate training, will train and assist you and/or your caregiver in the beginning to ensure optimal treatment. However, should you prefer full support for your infusion at home, the homecare nurse will carry out the entire procedure.

If you experience side effects with the treatment you must immediately seek the attention of your treating physician or your homecare nurse. Side effects can occur during treatment administration or shortly after.

If you experience side effects during the infusion, **the infusion should be stopped immediately**, and advice should be sought from your treating physician or your homecare nurse. Subsequent infusions may need to occur in a clinical setting. Please refer to Section 5 of this patient manual for a list of hypersensitivity related symptoms and the PIL for a full list of side effects.

Note: The dose and rate of the infusion while at home should follow the guidelines provided by your treating physician as noted in the Logbook, and not be changed without the agreement of your treating physician and supervision of the homecare nurse.

2. TRAINING IN ADMINISTRATION OF IMIGLUCERASE

In principle, the initial instructions will be given in the hospital. The level of support required from the homecare nurse will be discussed and agreed by you and/or your care giver and your treating physician.

Should you prefer full support to receive your infusion at home, the homecare nurse will carry out the entire procedure for you.

Should you prefer to carry out the procedure yourself, or with the assistance of your caregiver, you and/or your caregiver will receive training from the homecare nurse while the infusion is being prepared. The homecare nurse will explain and demonstrate the complete infusion procedure to you and/or your caregiver.

At subsequent visits, the homecare nurse will be present to assist if required, but you and/or your caregiver will gradually transition to performing more of the administration under the homecare nurse's supervision until you feel confident with the entire infusion procedure.

While reconstituting and administering imiglucerase, the procedure described in the PIL and in Section 4 of this patient manual must be closely followed.

3. ORGANISATION

Patient

- You and/or your caregiver must agree to receive the treatment at home.
- The home environment should be conducive to the provision of the home infusion therapy including a clean environment with electricity, water, telephone access, refrigeration, and physical space to support storage of imiglucerase and other infusion supplies.
- You must be physically and mentally able to undergo the infusions at home. The treating physician is responsible for the indication to receive imiglucerase infusions at home.
- You have accessible blood veins that allow an infusion needle to be inserted. When you have a central venous access device (CVAD) you should be able to insert the infusion needle into the septum.
- You and/or your caregiver have been informed by the treating physician about the treatment to be provided at home, the associated risks, the possible complications, and the provision of medical assistance at home.
- You and/or your caregiver have knowledge of Gaucher disease and are able to recognise side effects and understand the procedures to be followed should they occur.
- If you are carrying out the infusion procedure yourself, please ensure that you and/or your caregiver:
    - have been adequately trained in the procedures of imiglucerase reconstitution and infusion.
    - always refer to the Home Infusion Patient Manual and strictly follow the prescribed method of administration of imiglucerase.
    - always record each administration of imiglucerase, including any side effects, in the Logbook.
    - discontinue the infusion immediately and phone the treating physician/homecare nurse and/or the emergency services number provided in the Logbook to seek advice if you experience a side effect during or shortly after the infusion.

Homecare Nurse

- The homecare nurse is qualified to give intravenous (IV) infusions.
- The homecare nurse has been trained in administering imiglucerase and is aware of the possible side effects and the actions to be taken should they occur.
- The homecare nurse will establish with the patient and/or caregiver the level of support necessary.
- The homecare nurse will have a coordinating task together with the treating physician and you and/or your caregiver in organising the treatment at home.
- If the homecare nurse is carrying out the infusion procedure, they will:
    - strictly follow the prescribed method of administration of imiglucerase.
    - record each administration of imiglucerase, including any side effects, in the Logbook.
- In the event of a side effect occurring during or shortly after the infusion (i.e., infusion associated reaction [IAR]), the homecare nurse/patient/caregiver should discontinue the infusion and phone the treating physician and/or the emergency services number provided in the Logbook.

Treating physician

- The treating physician is responsible for the initiation of all necessary administrative actions, allowing other stakeholders (pharmacy, nurse, patient, caregiver) to proceed.
- The treating physician is responsible for the dose and the infusion rate, to be described in the Logbook. Any changes must be clearly communicated to the patient and described in the Logbook.

Third Person / Caregiver

It is preferable for a caregiver/third party to be present during home infusion.

The Logbook (Appendix 6.1)

- The Logbook serves as a means of communication for everyone involved in administering imiglucerase at home.
- In the Logbook, the treating physician clearly states the dose and the infusion rate, as well as any changes. These instructions on prescribed dose and rate of infusion should be strictly followed by you and/or your caregiver and/or homecare nurse.
- The Logbook should be kept at your home and will be kept up to date by you, your caregiver or the homecare nurse, depending on who is carrying out the infusion procedure.
    - The homecare nurse should record the finding and actions from the initial interview.
    - If the homecare nurse continues to carry out the infusion procedure for you at subsequent visits, they will record each adminstration of imiglucerase in the Logbook.
    - If you or your caregiver carry out the infusion procedure, you or your caregiver are responsible for recording each administration of imiglucerase in the Logbook.
- You and/or your caregiver must take the Logbook along to the hospital at each appointment for a check-up and bring it home afterwards.
- In the Logbook, the patient/caregiver/homecare nurse clearly describes what actions have been taken for the infusion side effect based on the advice of the treating physician or the homecare nurse.

Pharmacy and infusion equipment

Treatment and all necessary equipment will be provided according to the local requirements.

4. HOW DO I PREPARE AND ADMINISTER IMIGLUCERASE?

Requisites

Supplied by the hospital/pharmacy to you or to a third party with the appropriate prescription.

Preparation

1. The vials should be stored in a refrigerator at a temperature between +2°C and +8°C.
2. Prepare the equipment:

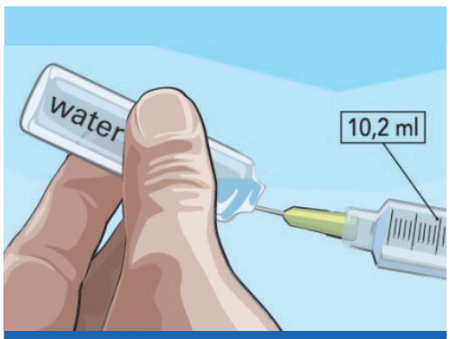
Step 2:
Preparation of the equipment

- The number of vials of imiglucerase required is determined based on the patient's weight and dose. Follow the guidance from your treating physician on the dose and rate of the infusion. Each vial contains 400 units of imiglucerase. Approximately 30 minutes before preparation, the vials should be removed from the refrigerator to reach room temperature. Check the expiry date printed on the the vial pack (do not use imiglucerase after the expiry date).
- Sterile water for injections to reconstitute imiglucerase.
- NaCl 0.9% solution, 2 x 100 ml for IV administration.
- NaCl 0.9% solution, 2 x 50 ml to flush infusion line pre- and post-infusion.
- Chlorhexidine 0.5% in alcohol 70% (antiseptic solution).
- Appropriate number of 10 ml and 50 ml syringes depending upon dose of imiglucerase.
- 3 x sterile hypodermic needles (1.1 x 40 mm); 1 x butterfly needle.
- In-line low protein-binding 0.2 µm filter is recommended.
- Hypodermic needle tray; Micropore tape; Mediswabs; Sharps bin; Handwash.
- Other equipment to be supplied for IV infusion according to local guidelines. Materials required to comply with hygienic and aseptic conditions as well as waste disposal rules.

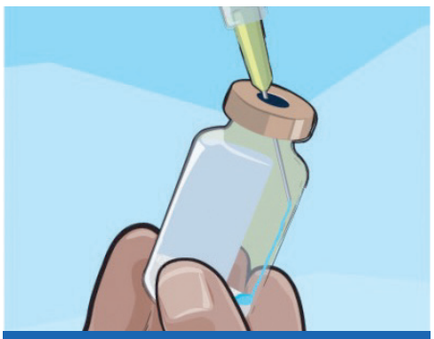
Reconstitution using sterile water

3. Remove the flip-off cap from the imiglucerase vial.
4. Disinfect the rubber stopper of the imiglucerase vial with chlorhexidine and allow to air dry.
5. Open the sterile water for injections.

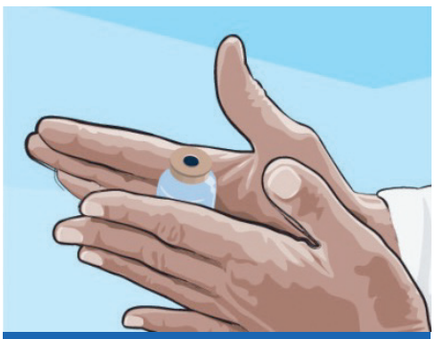
Step 4:
Observe aseptic technique



Step 6:
Drawing of sterile water

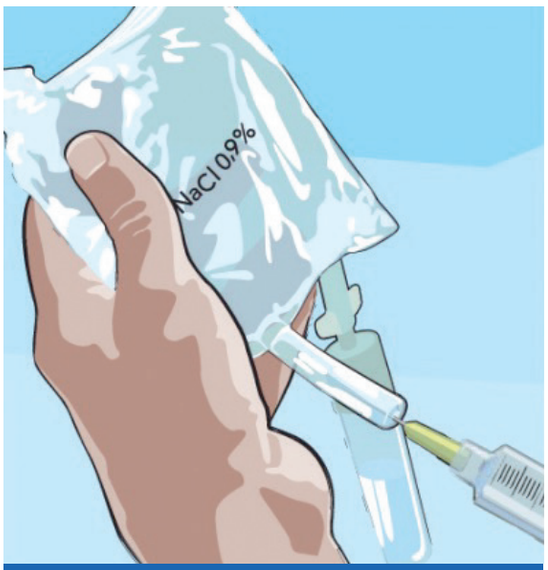


Step 7:
Injection of sterile water



Step 8:
Carefully swirl the vial using a circular movement of the hands

6. Draw 10.2 ml of sterile water for injections into the syringe.
7. Inject the sterile water gently down the glass side of each vial.
8. Carefully swirl the vial(s) to mix the solution (avoid forceful shaking during the reconstitution process to avoid foaming of the solution).
9. Small bubbles may appear after the mixing.
10. Let the solution settle for a few minutes to allow any bubbles present to disappear and to ensure that the powder is properly reconstituted (check that there are no foreign particles or discolouration).

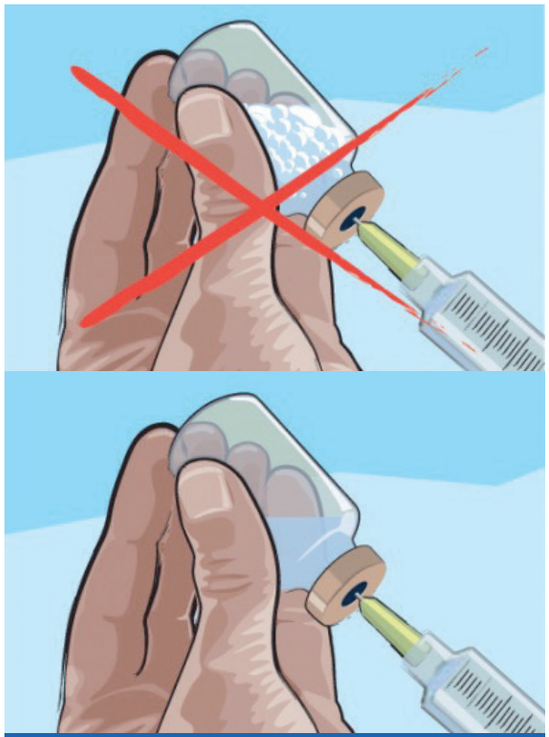


Step 12:
Withdraw and discard 10 ml (400 U vial) from the bag for each vial used

Dilution in 0.9% NaCl

11. Disinfect the cap/opening of 1 or 2 bags of NaCl 0.9% solution using chlorhexidine and allow to air dry.
12. Calculate the quantity of reconstituted imiglucerase solution present in the vials and draw the same quantity from the bag of NaCl solution, thus creating enough space to add the reconstituted imiglucerase solution.

    For instance, if the prescribed quantity is 3 vials of imiglucerase of 400 units each, remove 30 ml (= 3 x 10 ml) of NaCl solution from the bag of NaCl solution. Never remove more than half the content of the bag of NaCl to ensure that at least half the diluted solution consists of NaCl.



Step 13:
At the time of drawing, the reconstituted product should not contain any foam

13. Using one or more 50 ml syringes, draw 10 ml (400 U vial) from the reconstituted vials. When these quantities are drawn, the reconstituted product should not contain any foam. Gently inject the total volume of the reconstituted imiglucerase solution into the bag of NaCl 0.9% solution.
14. Carefully mix this imiglucerase solution.
15. The diluted solution should be filtered through an in-line low protein-binding 0.2 µm filter during administration.

Administration

16. The imiglucerase dose and infusion rate will be determined by the treating physician.
17. Imiglucerase must be administered by intravenous infusion.
18. Prior to infusion start, fill the infusion system with the mixed solution; fill the entire system to remove any air bubbles that may be present.
19. It is recommended that the diluted solution be administered within 3 hours. The product diluted in 0.9% NaCl intravenous solution will retain chemical stability if stored up to 24 hours at +2°C and +8°C under protection from light.

In case of a Central Venous Access Device (CVAD)

When you have a CVAD for the delivery of imiglucerase, you and/or your caregiver will be shown how to care for the device.

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

The following steps are necessary:

- When in use, cover site with transparent occlusive dressing. No dressing required when not in use.
- Flush with 5 mL saline before and after each use.
- Flush with 5 mL heparin (100 U/mL) after each use.

5. SAFETY ASSESSMENT

Should side effects occur during the infusion, or if you feel unwell when taking the treatment or after the treatment, contact the homecare nurse or the treating physician promptly. Side effects can occur during treatment administration or shortly after. These have included: itching, flushing, hives / localised swelling of the skin or lining of the mouth or throat, rash, chest discomfort, increased heart rate, bluish skin, breathlessness, coughing, a sensation of tingling, pricking, burning or numbness of the skin, fall in blood pressure, and backache.

If you experience side effects during the infusion, **the infusion should be stopped immediately**, and advice should be sought from your treating physician or your homecare nurse. Please refer to the PIL for a full list of side effects. You may need to be given additional medicines to prevent an allergic reaction. Please follow the instructions of your doctor or nurse. Any side effects should also be recorded in the Logbook.

In case severe side effects occur during or shortly after the infusion that require immediate attention/intervention, call the emergency services (see Logbook).

Preparation/administration mistake

If you become aware that a mistake was made while preparing and/or administering of the drug, please contact the homecare nurse or the treating physician.

6. APPENDICES

6.1 Logbook

Logbook for Cerezyme (imiglucerase) Home Infusion

Please ensure that old logbooks are not destroyed and are suitably retained so that historical medical data remain available.

General data

Patient	Name:	
	Address:	
	City:	
	Telephone:	
Homecare Nurse	Name:	
	Organisation:	
	Telephone:	
Treating physician	Name:	
	Hospital:	
	Address:	
	City:	
	Telephone:	
Pharmacy	Name:	
	Address:	
	City:	
	Telephone:	

Administration details (to be completed by treating physician)

Imiglucerase administered since	Date (dd-mmm-yyyy)
First infusion at home	Date (dd-mmm-yyyy)
Reasons for imiglucerase infusion at home	
Please indicate support to be provided by nurse	
Imiglucerase dosing regimen (dose, frequency, and rate of infusion)	

Emergency treatment details (to be completed by treating physician)

Necessary actions in the event of a serious infusion associated reaction: 1. Stop the infusion. 2. Call the emergency services number 999 3. Call the treating physician.

Infusion data (to be completed by homecare nurse and/or patient and/or caregiver)

Date of infusion	**Date (dd-mmm-yyyy)**
Patient's general health condition: specific problems/ remarks	
Dose of infusion	
Batch number/Number of vials used	
Duration of administration	
Rate of administration	
Problems/Remarks (related to infusion, e.g. side effects)	

Date of infusion	**Date (dd-mmm-yyyy)**
Patient's general health condition: specific problems/ remarks	
Dose of infusion	
Batch number/Number of vials used	
Duration of administration	
Rate of administration	
Problems/Remarks (related to infusion, e.g. side effects)	

Date of infusion	**Date (dd-mmm-yyyy)**
Patient's general health condition: specific problems/ remarks	
Dose of infusion	
Batch number/Number of vials used	
Duration of administration	
Rate of administration	
Problems/Remarks (related to infusion, e.g. side effects)	

Date of infusion	**Date (dd-mmm-yyyy)**
Patient's general health condition: specific problems/ remarks	
Dose of infusion	
Batch number/Number of vials used	
Duration of administration	
Rate of administration	
Problems/Remarks (related to infusion, e.g. side effects)	