VPRIV®

[Velaglucerase alfa]

Velaglucerase alfa infusion guide for homecare nurse/patient/caregiver

This guide contains important information on how to administer and manage risk of infusion-related reactions (IRR) including allergic-type hypersensitivity reactions in a home setting.

Please read this guide thoroughly and carefully before using this medicine

This guide is intended for patients who have been prescribed Velaglucerase alfa 400 Units powder for solution for infusion, and their caregivers and healthcare professionals (HCPs) who may assist them with home infusion.

This guide is intended for use by Homecare Nurse / Patient / Caregiver only, in conjunction with the VPRIV® SmPC.
This resource was developed by Takeda as part of a commitment made in the Risk Minimisation Measures for velaglucerase alfa.

Please report suspected adverse drug event(s) (AEs) to the MHRA through the Yellow Card scheme. You can report via the Yellow Card website www.mhra.gov.uk/yellowcard or the free. Yellow Card app available from the Apple App Store or Google Play Store. For any patients who do not have online access to report a suspected side effect to the Yellow Card scheme call 0800 731 6789 for free Monday to Friday between 10am and 2pm. Adverse event(s) should also be reported to Takeda at AE.GBR-RL@takeda.com.

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Takeda

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Purpose of this guide

You have received this guide because your treating physician has prescribed you velaglucerase alfa as a treatment for Gaucher Disease. If your initial infusions have gone well, your medication may be able to be administered at home, to make things easier for you.

Depending on your situation, the homecare nurse may administer your medication, or you may be allowed to infuse the medicine yourself (self-administer) at home after you or your caregiver have been appropriately trained. This Guide is designed to help the homecare nurse/patient/carer infuse velaglucerase alfa at home.

With this guide, please also read the Patient Information Leaflet carefully. Your clinical team might also provide you with additional training materials or support to help you.

- Keep this guide throughout the duration of your home infusions. You may need to reread it, and it contains your Infusion Diary, which you will need to fill in after each infusion. You must always bring your Infusion Diary to each appointment with your treatment team.
- Ask your doctor or another healthcare professional who assists or supports you if you have any further questions.
- Talk to your doctor or healthcare professional if you experience any side effects. This includes any possible side effects, even if not listed in this guide.

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Abbreviations

EXP	Expiry
HCP	Healthcare Professional
IV	Intravenous (into a vein)
mg	Milligram
mL	Mililitre

01. Before home infusion

The decision to receive infusions at home should be made by the patient and treating physician after three well-tolerated velaglucerase alfa infusions under medical supervision in a hospital, clinic or office setting to ensure satisfactory tolerance of the infusions.

How to Qualify for Home infusion of Velaglucerase alfa

Your treating physician may decide home infusions are appropriate for you if you have had at least three consecutive well-tolerated velaglucerase alfa infusions and has considered you to be medically stable with a history of adherence to the infusion schedule. You will be instructed on how to infuse velaglucerase alfa at home, about any associated adverse events or possible complications and what do to do in the event of a complication. The home environment must be, safe (clean, hygienic, storage area for supplies, drug, and emergency medication), and adequately equipped. You must maintain rapid and reliable communication measures with the treating physician, or the caregiver must be adequately trained. In addition, the homecare nurse or the caregiver are adequately trained in administering velaglucerase alfa.

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02. Suspected side effects related to your infusion

If an IRR occurs during administration, including a hypersensitivity reaction, discontinue the infusion immediately. You must contact your treating physician as the management of these reactions would be based on severity of the reaction, such as slowing the infusion rate, treatment with medications such as antihistamines, antipyretics and/or corticosteroids, and/or stopping and resuming treatment with an increased infusion time. Subsequent infusions may need to occur in hospital. If you have had a previous experience with adverse reactions during an infusion, you may be told to take an antihistamine and/or corticosteroid before the infusion to help prevent an allergic reaction from happening.

Infusion-related reaction and hypersensitivity

The nurse, patient and caregiver must communicate any events during and after the infusion to the treating physician, and to update the Infusion Diary

In the event of an IRR or hypersensitivity, the homecare nurse, the patient or caregiver should discontinue the infusion immediately and telephone/contact the treating physician using the information provided in the Infusion Diary. Such events must be documented in the Infusion Diary also. When such a reaction is observed, it is important that antibody blood testing be considered and promptly obtained, as warranted.

The Infusion Diary should include the infusion plan determined by the treating physician as well as a record of the actual infusions administered including health status of the patient before, during and after infusion. The document should accompany the patient and be shared with the treating physician on each visit.

Antibody testing

In case of suspected development of infusion-related reactions/hypersensitivity, types of allergic reactions or side effects/ adverse reactions or at the discretion of the treating physician based on his/her assessment of the response to the medication, a blood sample may be collected promptly in order to test if the patient has developed antibodies against the medication. It is very important that the homecare nurse, and/or patient caregiver report infusion-related reactions, hypersensitivity reactions, adverse reactions, or any suspicion about the infusion not working normally to the treating physician.

Possible side effects

Like all medicines, this medicine can cause side effects, although not everybody gets them. Commonly (may affect up to 1 in 10 people), patients experienced a severe allergic reaction, with difficulty breathing, chest discomfort (chest tightness), feeling sick (nausea), swelling of the face, lips, tongue or throat (anaphylactic/anaphylactoid reactions), common is also an allergic skin reaction such as hives, severe rash or itching. If any of these happen, tell your doctor immediately.

Most side effects, including the allergic reactions, occurred during the infusion or shortly after. These are called IRRs Other infusion related reactions that occurred very commonly (may affect more than 1 in 10 people) include headache, dizziness, fever/body temperature increased, back pain, joint pain and tiredness, as well as high blood pressure (commonly reported), blurry vision, and vomiting (uncommonly reported). If any of these happen tell your doctor immediately.

Other side effects include:

Very common side effects (may affect more than 1 in 10 people) are:

- bone pain
- weakness/loss of strength
- stomachache

Common side effects (may affect up to 1 in 10 people) are:

- lengthening of the time it takes for a cut to stop bleeding may lead to easy/spontaneous bleeding/ easy bruising
- skin flushing
- rapid heart beat
- developing antibodies to velaglucerase alfa **(see section 3)**
- decreased blood pressure

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03. Reporting an adverse event

Report any suspected adverse reactions or complications of self-administration of treatment to your treating physician. Healthcare professionals are asked to 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. Some clinical IT systems (EMIS/ SystmOne/Vision/ MiDatabank) for healthcare professionals. 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. Alternatively adverse events should be reported to Takeda on +44(0) 3333 0001.

If you are feeling unwell, please check with your healthcare provider to decide if your infusion should be postponed until you are feeling better. The homecare nurse may need to take a blood sample to see if you are producing antibodies against velaglucerase alfa, especially if you have an adverse reaction during the infusion process. Your doctor will have discussed these requirements with you beforehand.

Emergency plan

An emergency plan (section 8 page 10) has been provided to you by your physician. This plan should be followed in events of a serious infusion reaction, hypersensitivity reaction and/or adverse reaction.

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04. Facilitation of home infusion

Medication and materials will be supplied by the hospital/ pharmacy or via a third party with the appropriate prescription:

Medication and materials

Appropriate number of vials of velaglucerase alfa (400 U per vial) for the prescribed dose; vials should be stored in a refrigerator at a temperature between 2°C and 8°C.

- Sterile water for injections to reconstitute velaglucerase alfa
- NaCl 0.9% intravenous solution, one (1) to two (2) 100ml bag(s) for IV administration
- NaCl 0.9% intravenous solution, two (2) 50ml bags or vials to flush infusion line pre- and post-infusion
- Chlorhexidine 0.5% in alcohol 70% (antiseptic solution)
- Appropriate number of 5ml and 50ml syringes depending upon dose of velaglucerase alfa
- Sterile hypodermic needles and one (1) butterfly needle
- Tourniquet
- One (1) in-line low protein-binding 0.2 micron filter
- One (1) infusion set or one (1) combined infusion set with filter
- Hypodermic needle tray; Micropore tape; Mediswabs; Sharps bin; Hand wash
- Additional material may be needed if there is a central venous access device for the delivery of the medication. The caregiver will be shown how to care for the device.
- If required, pre-medication (antihistamines and/ or corticosteroids) to be given as per the healthcare provider's instructions. These will be prescribed and used on an individual patient basis as outlined in the Emergency Plan.

Dosing and reconstitution

Velaglucerase alfa is dosed according to body weight. Your doctor would have provided your homecare nurse or caregiver with the necessary information for calculating the correct dose in the Infusion Diary. The medication is supplied as a powder and must be reconstituted immediately before use. The homecare nurse or caregiver will mix the required amount of powder with the appropriate amount of sterile water. To make sure that no medication is wasted, you should be present when it is being prepared. If you are not able to start the infusion right away, reconstituted velaglucerase alfa can be stored in a fridge at 2–8 ^{O}C for up to 24 hours. The homecare nurse or caregiver will mix the reconstituted velaglucerase alfa with a bag of saline solution and attach the bag to the IV administration set. The infusion will take about an hour, the homecare nurse will monitor you regularly and will make further notes in the Infusion Diary.

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05. Preparation for the infusion

How to get ready for an infusion

1. Approximately 30 minutes before the infusion, remove the appropriate number of vials from the refrigerator to reach room temperature.
2. The healthcare provider will explain how many vials to use to provide the correct dose. **DO NOT** alter this dose.
3. Confirm that each vial is within the expiry date, which is printed on the vial and outer carton (the expiry date refers to the last day of the month indicated). **DO NOT** use after the expiry date.
4. Before beginning, ensure that the area used for preparing velaglucerase alfa is thoroughly cleaned.
5. Lay out the material.
6. Wash hands and keep the area clean and germ-free while preparing the solution.

How will the homecare nurse or caregiver insert the needle in the vein (if a central venous access device is not available)?

1. Ensure that the infusion system (infusion line connected to IV bag containing velaglucerase alfa) is within reach and that swabs, plasters, chlorhexidine and medical tape are close by.
2. Remove the butterfly needle from the packaging.
3. The patient will sit down and rest one arm on a table (preferably on a clean cloth).
4. The homecare nurse or caregiver will apply a tourniquet above the site of the infusion.
5. Prepare the infusion site by carefully wiping the skin with a disinfection swab. Allow the skin to dry before inserting the butterfly. Always use a new sterile needle for the infusion. Never re-use needles or syringes.
6. Remove the cap from the butterfly needle and insert the needle into a vein at as shallow an angle as possible.
7. Loosen the tourniquet and make sure that the needle is in a vein by pulling back the plunger gently (you should see backflow of blood into the butterfly tube).
8. To avoid needle movement during the infusion, tape the winged adapter to the skin using medical tape.

Wash hands

Clean area

Wear gloves

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06. Description of reconstitution and dilution technique

The homecare nurse and patient caregiver should ensure that they have received training and understand the administrative logistics of home-infusion of velaglucerase alfa:

Instructions for reconstitution

1. Using the aseptic technique, add Sterile Water for Injection to each vial as shown in the table below:

Solution	400 Units/vial
Volume of Sterile Water for Injection	4.3ml
Concentration after reconstruction	100 Units/ml
Withdrawal volume	4.0ml

2. Upon reconstitution, mix vials by gently rolling between hands. **DO NOT SHAKE**.
3. Prior to dilution, visually inspect the solution in the vials. The solution should be clear to slightly opalescent and colourless. The solution must not be use if it is discoloured or if foreign particles are present.

PLEASE NOTE: The patient should be present prior to reconstitution, to avoid waste.

Instructions to dilute for intravenous administration

1. Withdraw the calculated volume of velaglucerase alfa from the appropriate number of reconstituted vials. Some solution will remain in the vial (withdrawal volume = 4.0ml for 400 Unit vial).
2. Dilute the total volume of velaglucerase alfa required in 100ml of 0.9% sodium chloride solution for infusion.
3. Mix gently. **DO NOT SHAKE**.

From a microbiological point of view, the medicine should be used immediately. If not used immediately, in-use storage times and conditions prior to use are the responsibility of the user and must not exceed 24 hours at 2–8°C. Any unused solution should be disposed of in accordance with local requirements.

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07. Ongoing Administration

Velaglucerase alfa is intended for IV infusion only and should be administered over a period of 60 minutes. Velaglucerase alfa should not be infused with other products in the same infusion tubing, as the compatibility in solution with other products has not been evaluated. The diluted solution should be filtered through an in-line low protein-binding 0.22 µm filter during administration.

1. Attach the IV tubing to the diluted bag of velaglucerase alfa and prime the IV tubing with normal saline solution, expelling all air.
2. Set the infusion rate. Velaglucerase alfa should be administered over a period of 60 minutes.
3. Obtain IV access and attach the IV giving set. Follow local/facility protocols for IV insertion and infusion of medication.
4. Monitor the infusion regularly for IRRs.
5. When the infusion is complete, flush the tubing with normal saline to ensure residual velaglucerase alfa remaining in the tubing is infused.
6. Remove the venous access device and discard in an infectious waste disposal container.

The homecare nurse/patient caregiver must document the following information in the infusion diary: date, dose, route of administration, injection site, time infusion started and stopped and patient response to infusion.

Procedure for administration

If the patient has a central venous access device (in- dwelling central line), the homecare nurse or the caregiver have been shown how to care for the device.

1. Attach the infusion line to the butterfly needle or to the patients in-dwelling central line as shown by the doctor.
2. Attach the IV bag containing velaglucerase alfa to the drip stand and open the valve. Set the infusion rate determined by the treating physician.
3. Closely observe for any occurrence of IRRs **(see section 2)**.
4. At the end of the infusion, to ensure that the total treatment dose is administered, rinse the tubing using a 50ml bag of 0.9% NaCl, without increasing the infusion rate. In case of failure to gain venous access; or development of excessive bleeding, pain, swelling or severe bruising; or failure to infuse velaglucerase alfa into a vein correctly, please contact the treating physician immediately.
5. Remove the butterfly needle and discard in an infectious waste disposal container. For a central venous access device, follow the technique for proper care as shown by the healthcare provider or homecare nurse.
6. Any unused solution should be disposed of in accordance with local requirements as indicated by the healthcare provider or nurse.
7. Document the following in the infusion diary: date, dose, route of administration, injection site, time infusion started and stopped and patient response to infusion.
8. If aware that a mistake was made while preparing and/or administering the drug, please contact the healthcare provider. If the error occurred during the preparation step, do not administer the drug. If the error occurred during the administration, check with the healthcare provider before giving another infusion.

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08. Emergency plan for home infusion

[Treating Physician to provide individual instructions for homecare nurse/patient caregiver below.]

Remind the homecare nurse, patient/caregiver of necessary actions in the event of a serious infusion reaction, hypersensitivity reaction and/or adverse reaction:

1. Stop the infusion	☐
2. Call the national emergency number	☐
3. Call the treating physician	☐
4. If the homecare nurse is present, they will obtain a blood sample for antibody testing	☐
5. If the homecare nurse is not present, they will arrange for collection of the sample	☐

09. Infusion Diary for home infusion

The Infusion Diary should be completed and maintained by the treating physician, homecare nurse, patient/caregiver in collaboration.

Patient
Name:
Address:
City:
Telephone:
Email:
Caregiver (if applicable)
Name:
Address:
City:
Telephone:
Email:
Treating Physician
Name:
Address:
City:
Telephone:
Email:
Nurse
Name:
Address:
City:
Telephone:
Email:
Pharmacy
Name:
Address:
City:
Telephone:
Email:
National Emergency Number
Telephone:

General Information

Administration Details

Administered since (DD/MM/YYYY):
First infusion at home (DD/MM/YYYY):
Dose and frequency:
Infusion rate:
Indicate support to be provided by nurse:

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10. Complete your infusion diary

Infusion number:
Date of infusion:
Name of person giving the infusion (patient, caregiver or homecare nurse):
Patient's general health:
Patient's weight (kg):
Dose and rate of infusion:
Lot number:
Numbers of vials used:
Expiry date:
Time infusion started:
Time infusion stopped:
General remarks:
Any problems related to infusion? *
Any action taken:

*Remember that a blood sample should be obtained promptly for suspected IRR or hypersensitivity reactions.

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