Home Administration and Dosing Guide

Eligibility for home administration

Home administration under the supervision of a healthcare professional may be considered only for those patients who have received at least three infusions and were tolerating their infusions well. Appropriate medical support, including adequately trained personnel in emergency measures, should be readily available when VPRIV® (velaglucerase alfa for infusion) is administered. The eligibility for home infusion guide will help you determine if a patient is eligible for home infusion.

Training in administering VPRIV® (velaglucerase alfa for infusion)

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

Should the patient prefer full support when having their infusion at home, the homecare nurse will carry out the entire procedure for the patient.

Should the patient prefer to carry out the procedure him/herself or with the assistance of a caregiver, the patient and/or caregiver will receive training from the homecare nurse while the infusion is being prepared and administered. The homecare nurse will explain and demonstrate the complete infusion procedure to the patient and/or caregiver.

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

While reconstituting and administering VPRIV®, the procedure described in the Summary of Product Characteristics must be closely observed.

A homecare agency, care provider or hospital will provide the equipment required to administer the home infusion.

Shire will provide the patient care team with home infusion training and educational material.

Organisation of home infusion

The following information is intended to provide information and guidance to all persons involved in the procedures for organising home infusion of VPRIV®.

Patient

General

- The patient and/or caregiver, and/or homecare nurse 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.
- The patient and/or caregiver have an understanding of the illness and are able to recognise side effects and how they differ from the disease and understand the procedures to follow and who should be contacted should they occur.
- The patient and/or caregiver must agree to the treatment at home.
- The patient and/or caregiver have been adequately trained in the procedures of VPRIV® reconstitution and infusion.
- The home environment should be conductive to the provision of the home infusion including a clean environment with electricity, water, telephone, refrigerator and physical space to store VPRIV® and additional infusion supplies.
- In case the patient carries out the procedure him/herself:
  - The patient/caregiver will strictly follow the prescribed method of administration of VPRIV® as stated in the Home Infusion guide.
  - The patient/caregiver records each administration of VPRIV® in the Infusion Diary.
- In the event the patient experiences an adverse event during the infusion, the homecare nurse, the patient or caregiver should discontinue the infusion immediately and telephone the treating physician plus the country-specific national emergency number provided in the Infusion Diary and Emergency Plan.

Medical considerations

- The patient must be physically and mentally able to undergo the infusions at home.
- The treating physician is responsible for the recommendation to receive VPRIV® infusions at home.
- The patient has venous access or a central venous access device that allows for adequate infusion.

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Treating physician

- The treating physician is ultimately responsible for the initiation of all necessary administrative actions allowing other stakeholders (pharmacy, homecare nurse, patient and caregiver) to proceed.
- The treating physician is ultimately responsible for determining the dose and infusion rate to be added to the Infusion Diary. Any changes must be communicated to the patient and updated in the Infusion Diary.
- The treating physician is ultimately responsible for regular monitoring of the patient receiving home infusion of VPRIV®.

Hospital/pharmacy

- Treatment and all necessary equipment will be provided dependant on local arrangements and regulation.

Homecare nurse

- The homecare nurse is qualified to give intravenous (IV) infusions.
- The homecare nurse has been trained in administering VPRIV® and is aware of the possible side effects and the necessary actions to be taken should they occur.
- The homecare nurse will establish with the patient and/or caregiver the level of support required.
- The homecare nurse will coordinate together with the treating physician and the patient and/or caregiver in organizing treatment in your home.
- The homecare nurse will strictly follow the prescribed dose and rate of administration of VPRIV® as determined by the treating physician and given in the Infusion Diary.
- The homecare nurse will record each administration of VPRIV® in the Infusion Diary.
- In the event of an infusion-related reaction, the homecare nurse, the patient or caregiver should discontinue the infusion immediately and telephone the treating physician plus the country specific national emergency number provided in the Infusion Diary.

Caregiver/third party

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

Infusion Diary

- The Infusion Diary serves as a means of communication for everyone involved in administering VPRIV® at home.
- The Infusion Diary should be kept at the patient home and will be maintained and kept up to date by the patient or caregiver or the homecare nurse.
- The patient or caregiver must take the Infusion Diary along to the hospital at each appointment for a check-up and bring it home afterwards.
- In the Infusion Diary, the treating physician clearly states the dose and the infusion rate, as well as any subsequent changes.
- The patient or caregiver or the homecare nurse will strictly follow the prescribed dose and rate of infusion of VPRIV® as stated in the Infusion Diary.
- The homecare nurse records the details and actions from the initial consultation and the patient or caregiver or the homecare nurse notes all relevant information from subsequent visits in the Infusion Diary.
- The patient or caregiver or the homecare nurse will record each administration of VPRIV® in the Infusion Diary.
- In the Infusion Diary, the patient or caregiver or homecare nurse clearly describes what actions have been taken for any infusion reactions or infusion-related side effects based on the advice of the treating physician or the homecare nurse.

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Dosing of VPRIV®

How do I calculate the dose?

VPRIV® is dosed according to body weight. The recommended dose of VPRIV® is 60 Units/kg, administered every 14 days as a 60-minute IV infusion.[1,2]

Dose adjustments can be made on an individual basis based on achievement and maintenance of therapeutic goals. Clinical studies have evaluated doses ranging from 15 to 60 Units/kg every 14 days.

Patients previously treated with imiglucerase enzyme replacement therapy may be switched to VPRIV®, using the same dose and dosing frequency.[1]

Calculating the dosage[1]

- Check the patient's prescribed dose and number of vials needed
- The number of vials needed for the patient's prescribed dose can be calculated as shown below:

Prescribed dose (Total Units)	÷	Vial size 400 Units	=	Number of vials needed

Example: 1600 Units ÷ 400 Units = 4 Vials

How do I prepare VPRIV®?

Prior to infusion[1]

Patient assessment

- Obtain the patient's weight, if required
- Obtain pre-infusion vital signs and blood tests, if required
- Review previous infusion notes, if any, to assess for history of previous infusion reactions
- Notify the prescribing physician or specialist centre if the patient is unable to receive the infusion

What you will need

The following supplies are required for reconstitution, dilution and administration of VPRIV®:

- Sterile Water for Injection
- 0.9% sodium chloride solution for infusion (normal saline IV infusion bags)
- Syringes for reconstitution and dilution
- Needles (≤20 gauge and non-coring)
- IV administration set with in-line low protein-binding 0.22 µm filter
- Infusion control device/pump
- Venous access device/cannula
- Sterile IV infusion pack (e.g. skin preparation solution or wipes, gauze pads)
- Disposable gloves, tourniquet
- Medication label
- Post-infusion dressing
- Access to sharps infectious waste disposal system

If required, pre-medication (antihistamines and/or corticosteroids) to be given as per local/facility infusion protocol. To be prescribed and used on an individual patient basis.

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How do I administer VPRIV®?

1. Before you begin, ensure that the area where you will prepare VPRIV® is thoroughly cleaned. Wash your hands and keep the area clean and germ-free while preparing the solution.
2. Remove the appropriate number of vials from the refrigerator.
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.

Reconstitution[1,2]

1. Using the aseptic technique, add Sterile Water for Injection to each vial as shown in the table below:[2]

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 your hands. **DO NOT SHAKE.**[2]
3. Prior to dilution, visually inspect the solution in the vials. The solution should be clear to slightly opalescent and colourless. Do not use the solution if it is discoloured or if foreign particles are present.[2]

PLEASE NOTE: The patient should be present prior to reconstitution, to avoid waste.[2]

Dilution[1,2]

1. Withdraw the calculated volume of VPRIV® 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 VPRIV® 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

Administration

VPRIV® is intended for IV infusion only. VPRIV® should be administered over 60 minutes.1

VPRIV® 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.[2]

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

Document the following in the infusion diary: date, dose, route of administration, injection site, time infusion started and stopped and patient response to infusion.

References

1. VPRIV® Summary of Product Characteristics. May 2016
2. VPRIV® Patient Information Leaflet. May 2016

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