Package leaflet: Information for the user

VPRIV 400 Units powder for solution for infusion
velaglucerase alfa

Read all of this leaflet carefully before you start using this medicine because it contains important information for you.

- Keep this leaflet. You may need to read it again.
- If you have any further questions, ask your doctor.
- If you get any side effects, talk to your doctor. This includes any possible side effects not listed in this leaflet (see section 4).

What is in this leaflet

1. What VPRIV is and what it is used for

VPRIV is a long-term enzyme replacement therapy (ERT) for patients with type 1 Gaucher disease.

Gaucher disease is a genetic disorder caused by a missing or defective enzyme named glucocerebrosidase. When this enzyme is missing or does not work properly, a substance called glucocerebroside builds up inside cells in the body. The build-up of this material causes the signs and symptoms found in Gaucher disease.

VPRIV contains a substance called velaglucerase alfa which is designed to replace the missing or defective enzyme, glucocerebrosidase, in patients with Gaucher disease.

2. What you need to know before VPRIV is used

Do not use VPRIV
- if you are severely allergic to velaglucerase alfa or any of the other ingredients of this medicine (listed in section 6).

Warnings and precautions
Talk to your doctor before VPRIV is used
- If you are treated with VPRIV, you may experience side effects during or following the infusion (see section 4, possible side effects). These are called infusion related reactions and might appear as a hypersensitivity reaction with symptoms like nausea, rash, difficulty in breathing, back pain, chest discomfort (chest tightness), hives, joint pain or headache.
- Apart from symptoms of hypersensitivity reactions infusion-related reactions might show as dizziness, high blood pressure, tiredness, fever, itching, blurry vision, or vomiting.
  If you experience any of the symptoms, **you must tell your doctor immediately**.
- You may be given additional medicines to treat or help prevent future reactions. These medicines may include antihistamines, antipyretics, and corticosteroids.
- If the reaction is severe, your doctor will stop the intravenous infusion immediately and start giving you appropriate medical treatment.
- If the reactions are severe and/or there is a loss of effect from this medicine, your doctor will perform a blood test to check for antibodies which may affect the outcome of your treatment

- Your doctor or nurse may decide to continue to administer VPRIV even if you experience any infusion related-reaction. Your condition will be closely monitored.

Tell your doctor if you have previously experienced an infusion-related reaction with other ERT for Gaucher disease.

Children
Do not use in children under the age of 4 years because there is no experience of using the medicine in this age group.

Other medicines and VPRIV
Tell your doctor if you are taking, have recently taken or might take any other medicines.

Pregnancy
Gaucher disease may become more active in a woman during pregnancy and for a few weeks after birth. Women with Gaucher disease who are pregnant or considering pregnancy should talk with their doctor before this medicine is used.

Breast-feeding
It is not known whether VPRIV can pass into breast milk. If you are breast-feeding or considering breast-feeding, you should talk to your doctor before this medicine is used.

Your doctor will then help you decide whether to stop breast-feeding, or whether to stop using VPRIV, considering the benefit of breast-feeding to the baby and the benefit of VPRIV to the mother.

Driving and using machines
VPRIV has no or negligible influence on your ability to drive or use machines.

VPRIV contains sodium
This medicine contains 12.15 mg sodium (main component of cooking/table salt) in each vial. This is equivalent to 0.6% of the recommended maximum daily dietary intake of sodium for an adult.

3. How VPRIV is used

This medicine is only to be used under appropriate medical supervision of a doctor who is knowledgeable in the treatment of Gaucher disease. It is given by a doctor or nurse by intravenous infusion.

Dose
The recommended dose is 60 Units/kg given every other week.

If you are currently being treated for Gaucher disease with another ERT and your doctor wants to change you to VPRIV, you can initially receive VPRIV at the same dose and frequency you had been receiving the other ERT.

Use in children and adolescents
VPRIV may be given to children and adolescents (4 to 17 years of age) at the same dose and frequency as in adults.

Use in elderly
VPRIV may be given to the elderly (aged over 65 years) at the same dose and frequency as in adults.

Response to treatment
Your doctor will monitor your response to treatment and may change your dose (up or down) over time.

If you are tolerating your infusions well in the clinic, your doctor or nurse may administer your infusions at home.

Administration
VPRIV is supplied in a vial as a packed powder which is mixed with sterile water and further diluted in sodium chloride 9 mg/ml (0.9%) solution for infusion prior to intravenous infusion.

After preparation, your doctor or nurse will give the medicine to you through a drip into a vein (by intravenous infusion) over a period of 60 minutes.

If you have any further questions on the use of this medicine, ask your doctor.

4. 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 infusion-related reactions. 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
- stomach ache

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 VPRIV (see section 2)
- decreased blood pressure

Reporting of side effects
If you get any side effects, talk to your doctor. This includes any possible side effects not listed in this leaflet. You can also report side effects directly via Yellow Card Scheme, Website: www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store. By reporting side effects you can help provide more information on the safety of this medicine.

5. How to store VPRIV

Keep this medicine out of the sight and reach of children.

Do not use this medicine after the expiry date which is stated on the carton and label after ‘EXP’. The expiry date refers to the last day of that month.

Store in the refrigerator (2 ºC – 8 ºC).
Do not freeze.
Keep the vial in the outer carton in order to protect from light.

Reconstituted and diluted solution for infusion:
Use immediately. Do not exceed 24 hours at 2 °C to 8 °C.

Do not use if the solution is discoloured or if foreign particles are present.

Do not throw away any medicines via wastewater or household waste. Ask your pharmacist how to throwaway medicines you no longer use. These measures will help to protect the environment.

6. Contents of the pack and other information

What VPRIV contains

- The active substance is velaglucerase alfa.
  Each vial contains 400 Units of velaglucerase alfa.
  After reconstitution, one ml of solution contains 100 Units of velaglucerase alfa

- The other ingredients are sucrose, sodium citrate dihydrate, citric acid monohydrate and polysorbate 20 (see section 2 “VPRIV contains sodium”).

What VPRIV looks like and contents of the pack
20 ml glass vial containing a white to off-white powder for solution for infusion.

Packs of 1, 5 or 25 vials.
Not all pack sizes may be marketed.

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This leaflet was last revised in 10/2022.

The following information is intended for healthcare professionals only.

VPRIV is a powder for solution for infusion. It requires reconstitution and dilution and is intended for intravenous infusion only. VPRIV is for single–use only and is administered through a 0.2 or 0.22 µm filter. Discard any unused solution. VPRIV should not be infused with other medicines in the same infusion as the compatibility in solution with other medicines has not been evaluated. The total volume of infusion should be delivered over a period of 60 minutes.

Use aseptic technique.
Prepare VPRIV as follows:

1. Determine the number of vials to be reconstituted based on the individual patient's weight and the prescribed dose.
2. Remove the required vials from the refrigerator. Reconstitute each vial using sterile water for injections:

Vial size	Water for injections
400 Units	4.3 ml

3. Upon reconstitution, mix vials gently. Do not shake.
4. Prior to dilution, visually inspect the solution in the vials; the solution should be clear to slightly opalescent and colourless; do not use if the solution is discoloured, or if foreign particulate matter is present.
5. Withdraw the calculated volume of medicine from the appropriate number of vials. Some solution will remain in the vial:

Vial size	Extractable volume
400 Units	4.0 ml

6. Dilute the total volume required in 100 ml of sodium chloride 9 mg/ml (0.9%) solution for infusion. Mix gently. Do not shake. Initiate the infusion within 24 hours from the time of reconstitution.

From a microbiological point of view, use the medicine immediately. If you do not use immediately, in-use storage times and conditions prior to use are the responsibility of the user. Do not exceed 24 hours at 2 ºC to 8 ºC.

Do not dispose of the medicine via waste water or household waste. Dispose of any unused medicine or waste material in accordance with local requirements.

Keeping a record
In order to improve the traceability of biological medicine, record the name and batch number of the administered medicine clearly.