PATIENT MEDICATION INFORMATION

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

[Pr]NEXVIAZYME™

avalglucosidase alfa for injection, Lyophilized Powder

Read this carefully before you start taking **Nexviazyme** and each time you get a refill. This leaflet is a summary and will not tell you everything about this drug. Talk to your healthcare professional about your medical condition and treatment and ask if there is any new information about **Nexviazyme**.

Serious Warnings and Precautions

- **Hypersensitivity Reactions Including Anaphylaxis**
  If you are treated with Nexviazyme you may experience a life-threatening hypersensitivity reaction, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Nexviazyme administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, Nexviazyme should be discontinued immediately and appropriate medical treatment should be initiated. In patients with severe hypersensitivity reaction, a desensitization procedure to Nexviazyme may be considered.
- **Infusion-Associated Reactions (IARs)**
  If you are treated with Nexviazyme you may experience an infusion-associated reaction. An infusion-associated reaction is defined as any related side effect occurring during the infusion or during the 2 hours following infusion. Life-threatening allergic reactions, including anaphylactic shock, have been observed in patients during Nexviazyme infusion. Because of the potential for severe infusion reactions, immediate discontinuation of Nexviazyme should be considered, initiation of appropriate medical treatment, should be readily available when Nexviazyme is administered, the benefit risks and risks of readministering Nexviazyme following severe infusion reactions should be considered.

  Individuals with an acute underlying illness [e.g. fever, pneumonia or sepsis (severe infection), wheezing/difficulty in breathing, heart failure] at the time of Nexviazyme infusion appear to be at greater risk for infusion reactions. Careful consideration should be given to your clinical status prior to administration of Nexviazyme.

What is Nexviazyme used for?

- Nexviazyme is a medicine that is used to treat adults, children and adolescents who have a confirmed diagnosis of late-onset Pompe disease.

How does Nexviazyme work?

- People with Pompe disease have low levels of an enzyme called acid alpha-glucosidase (GAA)
- This enzyme helps the body control levels of glycogen
  - Glycogen is a type of sugar that provides the body with energy
- In Pompe disease the levels of glycogen can get too high.
  - Too much sugar builds up and damages your muscles and organs

  - Pompe disease causes muscle weakness and trouble breathing:
    - It mostly affects the liver, heart, and muscles
- People with Pompe disease are not able to make enough of this enzyme
- Nexviazyme contains an artificial enzyme called avalglucosidase alfa;
  - it can replace the natural enzyme which is lacking in Pompe disease

What are the ingredients in Nexviazyme?

Medicinal ingredient: avalglucosidase alfa
Non-medicinal ingredients: Glycine, L-Histidine, L-Histidine HCl monohydrate, mannitol, polysorbate 80

Nexviazyme comes in the following dosage forms:

lyophilized powder, 100 mg/vial (10 mg/mL)

Do not use Nexviazyme if:

- If you have experienced life-threatening allergic (hypersensitive) reactions to avalglucosidase alfa or its ingredients or components of the container and re-administration of the medicine was not successful.

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take Nexviazyme. Talk about any health conditions or problems you may have, including if you:

- Have had a severe hypersensitivity or anaphylactic reaction to administration of Nexviazyme.
- Have experienced allergic reactions (see "What are the possible side effects from using Nexviazyme" section) or infusion-associated reactions (IARs) while you were given the medicine or during the hours following the infusion
- Are at increased risk of lung infections due to the progressive effects of the disease on the lung muscles
- Are pregnant, think you may be pregnant or plan to become pregnant or are breast feeding
- Are driving or using any tools or machines shortly after infusion of Nexviazyme, since you may experience dizziness.

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

How to take Nexviazyme:

Nexviazyme will be given to you under the supervision of a healthcare professional who is experienced in the treatment of Pompe disease.

The dose you receive is based on your body weight and will be given to you once every other week.

Nexviazyme is given through a drip into a vein (by intravenous infusion). It is supplied as a powder which will be mixed with sterile water before it is given.

Usual dose:

<u>Late-onset Pompe disease (LOPD)</u>
The recommended dosage of Nexviazyme is 20 mg per kg of body weight once every other week as an intravenous infusion.

Overdose:

There is no experience with overdose of Nexviazyme.

If you think you have taken too much Nexviazyme, contact your healthcare professional, hospital emergency department or regional poison control centre immediately, even if there are no symptoms.

Home Infusion

Your treating physician may consider home infusion of Nexviazyme if you are tolerating your clinic infusions well. This decision to move to home infusion should be made after evaluation and recommendation by your treating physician. If you experience an adverse event during an infusion of Nexviazyme, your home infusion staff member may stop the infusion and initiate appropriate medical treatment.

Missed Dose:

If you have missed an infusion, please contact your doctor. If you have any further questions on the use of this medicine, ask your doctor, pharmacist, or nurse.

What are possible side effects from Nexviazyme?

These are not all the possible side effects you may feel when taking Nexviazyme. If you experience any side effects not listed here, contact your healthcare professional.

Side effects were mainly seen while patients were being given the medicine or shortly after ("infusion associated reactions (IARs)"). Allergic reactions may include symptoms such as difficulty breathing, chest pressure, low blood pressure, generalized flushing, cough, dizziness, nausea, redness on palms, swollen lower lip and tongue, decreased breath sounds, difficulty speaking, difficulty swallowing, redness on feet, itchy palms and feet, low level of oxygen in the blood, redness of skin, severe itching, swelling and rash. IARs may include symptoms such as chest discomfort, , fever, headache, dizziness, chills, feeling hot or cold, cough, diarrhea, redness of skin, fatigue, excessive sweating, headache, influenza-like illness, nausea, redness of eye, pain in extremity, itchy skin, rash, red rash, skin lesions, increase in heart rate, increased or decreased blood pressure, hives or vomiting. The majority of the IARs were mild to moderate. If you experience any reaction similar to this, tell your doctor immediately. You may need to be given pre- treatment medicines to prevent an allergic reaction (e.g., antihistamines and/or corticosteroids) or to reduce fever (antipyretics).

If you experience swelling of your lower limbs or generalized swelling, please inform your doctor.

Common: may affect up to 1 in 10 people

- Headache
- Dizziness, sleepiness or fatigue
- Tremor (shaking)
- Burning sensation
- Red and/or itchy eyes
- Swelling of eyelid, face, lip or tongue

- Increased heart rate, increased or lowered blood pressure
- Flushing, excessive sweating, feeling hot/cold
- Pale skin, lips turning blue, low blood oxygen
- Cough, difficulty breathing
- Throat irritation or mouth/throat pain
- Nausea, diarrhea, vomiting, indigestion
- Pain, aches, or discomfort in the abdomen, flank, muscles and/or chest
- Hives, Rash, Itchy and/or red skin or skin plaques
- Muscle spasms
- Flu-like illness, chills, fever
- Infusion site pain

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, talk to your healthcare professional.

Reporting Side Effects

You can report any suspected side effects associated with the use of health products to Health Canada by:

- Visiting the Web page on Adverse Reaction Reporting (https://www.canada.ca/en/health-canada/services/drugs-health-products/medeffect-canada.htmlfor information on how to report online, by mail or by fax; or
- Calling toll-free at 1-866-234-2345.

NOTE: Contact your health professional if you need information about how to manage your side effects. The Canada Vigilance Program does not provide medical advice.

Storage:

Unopened vials – before reconstitution

Store in refrigerator between 2°C - 8°C. Do not use Nexviazyme after the expiration date on the vial. The expiry date refers to the last day of the month.

After reconstitution and dilution

After dilution, an immediate use is recommended. The reconstituted product can be stored up to 24 hours when refrigerated at 2°C -8°C and diluted product can be stored up to 24 hours when refrigerated at 2°C -8°C or up to 9 hours (including infusion time) when stored at room temperature (up to 25°C).

Keep out of reach and sight of children.

If you want more information about Nexviazyme:

- Talk to your healthcare professional.
- Find the full product monograph that is prepared for healthcare professionals and includes this Patient Medication Information by visiting the Health Canada website (https://www.canada.ca/en/health-canada.html); the manufacturer's website www.sanofi.ca, or by calling 1-800-265-7927.

Pompe Registry:

Sanofi Genzyme informs all patients with Pompe Disease that a registry has been established in order to better understand the variability and progression of Pompe Disease and to continue to monitor and evaluate the safety and efficacy of **Nexviazyme**. All patients are encouraged to participate and advised that their participation may involve long-term follow-up. Information regarding the registry program may be found at www.registrynxt.com or by calling 1-800-745-4447, extension 15500.

This leaflet was prepared by sanofi-aventis Canada Inc.

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