

Patient Medication Information

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

Pr **XENPOZYME**[®]

olipudase alfa for injection, lyophilized powder

Read this carefully before you start taking **XENPOZYME** 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 **XENPOZYME**.

Serious warnings and precautions box

Infusion-Associated Reactions (IAR)

- You may have side effects called infusion associated reactions (IARs) that may be caused by infusion of the medicine. They may occur while you are being given **XENPOZYME** or within 24 hours following the infusion. If you think you are having an IAR, tell your doctor straight away. See below section "To help avoid side effects and ensure proper use, talk to your healthcare professional before you take **XENPOZYME**[®]" for more information.

What XENPOZYME is used for:

- **XENPOZYME** contains an enzyme called olipudase alfa. Olipudase alfa is an enzyme replacement therapy, which can replace the natural enzyme that has reduced activity in Acid Sphingomyelinase Deficiency (ASMD). **XENPOZYME** is used in adults and children to treat the signs and symptoms of ASMD not related to the brain.

How XENPOZYME works:

- The reduced activity of acid sphingomyelinase results in a build-up of a fatty substance called sphingomyelin (SM) which can cause damage to various organs. Olipudase alfa replaces the natural enzyme.

The ingredients in XENPOZYME are:

Medicinal ingredients: olipudase alfa

Non-medicinal ingredients: L-methionine, sodium phosphate dibasic heptahydrate, sodium phosphate monobasic monohydrate, sucrose

XENPOZYME comes in the following dosage forms:

Lyophilized powder, one vial contains 4 mg or 20 mg of olipudase alfa.

Do not use XENPOZYME if:

- You have experienced life-threatening allergic (hypersensitive) reactions to olipudase alfa or any of the other ingredients of this medicine.

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

- **Infusion Associated Reactions**
 - IAR may include allergic reactions and symptoms such as headache, hives, fever, nausea, vomiting, itchy skin, reddening of the skin and rash.
 - If you have a severe allergic reaction during your infusion your doctor should stop your infusion and provide appropriate medical treatment.
 - If you have a mild or moderate IAR, your doctor or nurse may temporarily stop the infusion, lower the infusion rate, and/or reduce the dose.
 - Your doctor may also give (or have given) you other medicines to prevent or manage IAR or allergic reactions.
- **Blood tests**
 - Your doctor may order tests to check for anti-olipudase alfa antibodies, particularly if you have experienced an allergic reaction. Anti-olipudase antibodies can be made by the body when a drug such as olipudase alfa is given.
 - Your doctor will order blood tests to check how well your liver is working (liver enzymes) before starting the treatment, and then at regular intervals during the dose escalation.
- **Other medicines and products**

Tell your doctor or nurse if you are using, have recently used, or might use any other medicines.
- **Pregnancy and/or breastfeeding**
 - Talk to your doctor before using XENPOZYME if you are pregnant or breastfeeding, think you may be pregnant, or are planning to have a baby.
 - If you are a woman who could become pregnant, your doctor will verify your pregnancy status before starting treatment with XENPOZYME®.
 - There is very limited experience with the use of XENPOZYME in pregnant women. XENPOZYME may be harmful to unborn children when taken by a woman during pregnancy. XENPOZYME should only be used during pregnancy if clearly necessary. Women who are able to become pregnant should use effective contraception during treatment and for 14 days after the last dose if XENPOZYME is discontinued.
 - It is not known whether XENPOZYME passes into human breast milk. XENPOZYME was detected in animal milk. Tell your doctor if you are breast-feeding or plan to do so. Your doctor will then help you decide whether to stop breast-feeding, or whether to stop taking XENPOZYME®.
- **Caution on driving and using machinery**

XENPOZYME may have a minor influence on the ability to drive and use machines because you may experience low blood pressure.

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

How to take XENPOZYME®:

XENPOZYME will be given to you under the supervision of a healthcare professional who is experienced in giving this type of medication.

The dose you receive is based on your body weight and will be given to you every two weeks.

Infusion usually lasts around 3 hours and 40 minutes; but may be shorter or longer based on your doctor's judgement and may be shorter during the period while your dose is being increased.

Instructions for proper use

XENPOZYME is given through a vein by intravenous (IV) infusion. It is supplied as a powder that will be mixed with sterile water before it is given.

Usual dose:

Adult patients:

The recommended starting dose of XENPOZYME is 0.1 mg for each kg of body weight. The next doses should be increased in a planned way up to the recommended dose of 3 mg for each kg of body weight every 2 two weeks. It typically takes up to 14 weeks to reach the recommended dose but may be longer based on your doctor's judgement.

Pediatric patients:

The recommended starting dose of XENPOZYME is 0.03 mg for each kg of body weight. The next doses should be increased in a planned way up to the recommended dose of 3 mg for each kg of body weight every 2 two weeks. It typically takes up to 16 weeks to reach the recommended dose but may be longer based on your doctor's judgement.

Home Infusion

Your doctor may consider home infusion of XENPOZYME if you are on stable dose and tolerating your infusions well. This decision to move to home infusion should be made after evaluation and recommendation by your doctor. If you experience a side effect during an infusion of XENPOZYME®, your home infusion provider may stop the infusion and start appropriate medical treatment.

Overdose:

Tell your doctor immediately.

If you think you, or a person you are caring for, have been given too much XENPOZYME contact a healthcare professional, hospital emergency department, regional poison control centre or Health Canada's toll-free number, 1-844 POISON-X (1-844-764-7669) immediately, even if there are no symptoms.

Missed Dose:

It is important to have your infusion every 2 weeks. An infusion is considered missed if not given within 3 days from the scheduled infusion. Depending on the number of missed doses, your healthcare provider may have to restart from a lower dose. Dose escalation should take place in a clinic or hospital setting.

If you have missed an infusion or are unable to attend a scheduled appointment, please contact your doctor right away.

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

Possible side effects from using XENPOZYME®:

These are not all the possible side effects you may have when taking XENPOZYME®. If you experience any side effects not listed here, tell your healthcare professional.

Like all medicines, this medicine can cause side effects, although not everybody will experience them. IARs were seen while patients were being given the medicine or within 24 hours after the infusion. The majority of the IARs were mild to moderate.

The most serious side effects may include sudden severe allergic reactions and irregular heartbeat.

If you experience any of those side effects, seek immediate medical attention.

If you have an infusion reaction you may be given additional medicines to treat or help prevent future reactions. If the infusion reaction is severe, your doctor may stop the infusion of XENPOZYME and start giving appropriate medical treatment.

Very Common: may affect more than 1 in 10 people

- Headache
- Fever – body temperature increased
- Raised, itchy rash (hives)
- Nausea
- Vomiting
- Abdominal (belly) pain
- Muscles aches
- Itchy skin
- Increased results of blood test for inflammation
- Pain in upper belly
- Reddening of the skin
- Diarrhea
- Fatigue
- Joint pain
- Back pain
- Rash (different types of rash, sometimes with itch)
- Difficulty breathing
- Feeling very warm
- Weakness

Common: may affect up to 1 in 10 people

- Abnormal blood test for liver function
- Reddening of the skin
- Chills
- Bone pain
- Pain
- Low blood pressure
- Forceful heartbeat that may be rapid or irregular
- Fast heartbeat
- Liver pain
- Severe allergic reactions
- Throat and voice box irritation

- Throat tightness and swelling
- Wheezing
- Rapid swelling under the skin in areas such as the face, throat, arms and legs which can be life threatening if throat swelling blocks the airway.
- Itchy or red eyes
- Eye discomfort
- Abnormal blood test for inflammation
- Abdominal discomfort
- Skin lesions (such as solid elevated or red flat lesions)

Serious side effects and what to do about them

Symptom / effect	Talk to your healthcare professional		Stop taking this drug and get immediate medical help
	Only if severe	In all cases	
COMMON			
Severe allergic reaction			√
Allergic reaction		√	
Hives		√	
Rash		√	

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, tell 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 (canada.ca/drug-device-reporting) for information on how to report online, by mail or by fax; or
- Calling toll-free at 1-866-234-2345.

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

Storage:

Keep out of the reach and sight of children.

Store refrigerated between 2°C and 8°C. Do not use XENPOZYME after the expiration date stated on the label. The expiry date refers to the last day of the month.

After dilution, immediate use is recommended.

If not used immediately, the reconstituted solution may be stored for up to 24 hours at 2°C to 8°C or up to 6 hours at 25°C.

After dilution, the solution can be stored for up to 24 hours at 2-8°C followed by 12 hours (including infusion time) when stored at 25°C.

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

If you want more information about XENPOZYME®:

- 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 Drug Product Database website: ([Drug Product Database: Access the database](#)); the manufacturer's website www.sanofi.com/en/canada, or by calling 1-800-265-7927.

This leaflet was prepared by sanofi-aventis Canada Inc.

Date of Authorization: 2026-02-04