

Consumer Medicine Information (CMI) summary

The [full CMI](#) on the next page has more details. If you are worried about using this medicine, speak to your doctor or pharmacist.

▼ This medicine is new. Please report side effects. See the [full CMI](#) for further details.

1. Why am I being given Xenpozyme?

Xenpozyme contains the active ingredient olipudase alfa. Xenpozyme is used to treat patients who have acid sphingomyelinase deficiency (ASMD) type A/B (Niemann-Pick type A/B) or type B (Niemann-Pick type B).

For more information, see Section [1. Why am I being given Xenpozyme?](#) in the full CMI.

2. What should I know before I am given Xenpozyme?

Do not use if you have ever had an allergic reaction to Xenpozyme or any of the ingredients listed at the end of the CMI.

Talk to your doctor if you have any other medical conditions, take any other medicines, or are pregnant or plan to become pregnant or are breastfeeding.

For more information, see Section [2. What should I know before I am given Xenpozyme?](#) in the full CMI.

3. What if I am taking other medicines?

Some medicines may interfere with Xenpozyme and affect how it works.

A list of these medicines is in Section [3. What if I am taking other medicines?](#) in the full CMI.

4. How will I be given Xenpozyme?

- Your doctor or nurse will give you Xenpozyme into a vein (intravenously) as a drip infusion

More instructions can be found in Section [4. How will I be given Xenpozyme?](#) in the full CMI.

5. What should I know whilst being given Xenpozyme?

Things you should do	<ul style="list-style-type: none">Remind any doctor, dentist or pharmacist you visit that you are receiving XenpozymeIt is important to keep all appointments so that your doctor can see if Xenpozyme works for you
Things you should not do	<ul style="list-style-type: none">Do not stop your treatment with Xenpozyme unless you have discussed this with your doctor.
Driving or using machines	<ul style="list-style-type: none">Xenpozyme may cause low blood pressure. Be careful driving or operating machinery until you know how Xenpozyme affects you.
Looking after your medicine	<ul style="list-style-type: none">Xenpozyme will be stored in a refrigerator.

For more information, see Section [5. What should I know whilst being given Xenpozyme?](#) in the full CMI.

6. Are there any side effects?

Infusion reactions have been observed with Xenpozyme treatment.

Common side effects include headache, fever, hives and/or rash, nausea, vomiting, abdominal pain, muscle pain, itchiness.

Serious side effects include allergic reactions with symptoms such as difficulty breathing, chest discomfort, flushing, dizziness, nausea, redness on the palms and/or feet, swollen lips and/or tongue and rash.

For more information, including what to do if you have any side effects, see Section [6. Are there any side effects?](#) in the full CMI.

▼ This medicine is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may get. You can report side effects to your doctor, or directly at www.tga.gov.au/reporting-problems.

Xenpozyme

Active ingredient(s): *olipudase alfa*

Consumer Medicine Information (CMI)

This leaflet provides important information about using Xenpozyme. **You should also speak to your doctor or pharmacist if you would like further information or if you have any concerns or questions about using Xenpozyme.**

A Patient card is also available to you to provide important information for your use of Xenpozyme.

Where to find information in this leaflet:

1. [Why am I being given Xenpozyme?](#)
2. [What should I know before I use Xenpozyme?](#)
3. [What if I am taking other medicines?](#)
4. [How do I receive Xenpozyme?](#)
5. [What should I know while using Xenpozyme?](#)
6. [Are there any side effects?](#)

1. Why am I being given Xenpozyme?

Xenpozyme contains the active ingredient olipudase alfa. Xenpozyme is an enzyme replacement therapy, which can replace the natural enzyme that has reduced activity in Acid Sphingomyelinase Deficiency (ASMD).

Xenpozyme is used to treat manifestations of ASMD not related to the brain in adults and children.

Patients with ASMD lack a properly working version of the enzyme acid sphingomyelinase. This results in a build-up of a substance called sphingomyelin, which damages organs such as the spleen, liver, heart, lungs and blood. Olipudase alfa acts in the same way as the natural enzyme would, and so acts as a replacement, reducing the build-up of sphingomyelin in the organs and treating the signs and symptoms of ASMD.

2. What should I know before I am given Xenpozyme?

Warnings

Do not use Xenpozyme if:

- you are allergic to olipudase alfa, or any of the ingredients listed at the end of this leaflet.
- always check the ingredients to make sure you can use this medicine.

Tell your doctor if you:

- have any other medical conditions
- take any medicines for any other condition

During treatment, you may be at risk of developing certain side effects. It is important you understand these risks and

how to monitor for them. See additional information under Section [6. Are there any side effects?](#)

Pregnancy and breastfeeding

Check with your doctor if you are pregnant or intend to become pregnant. If you are a woman who could become pregnant, your doctor may ask you to take a pregnancy test before you start taking this medicine. There is no 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 breast milk. Xenpozyme was detected in animal milk. Talk to your doctor if you are breastfeeding or intend to breastfeed. Your doctor will then help you decide whether to stop breastfeeding, or whether to stop taking Xenpozyme, considering the benefit of breastfeeding the baby and the benefit of Xenpozyme to the mother.

Xenpozyme contains sodium

This medicine contains 3.02 mg sodium in each vial. This is equivalent to 0.15% of the recommended maximum daily dietary intake of sodium for adults.

3. What if I am taking other medicines?

Tell your doctor or pharmacist if you are taking any other medicines, including any medicines, vitamins or supplements that you buy without a prescription from your pharmacy, supermarket or health food shop.

Some medicines may interfere with Xenpozyme and affect how it works.

Check with your doctor or pharmacist if you are not sure about what medicines, vitamins or supplements you are taking and if these affect Xenpozyme.

4. How will I be given Xenpozyme?

How much will I be given

- You will be given Xenpozyme under the supervision of your doctor or nurse.
- The dose you will receive is based on your body weight.
- For adults, the recommended starting dose of Xenpozyme is 0.1 mg for each kg of bodyweight. This is increased every 2 weeks in a planned way with each subsequent dose until the recommended dose of 3 mg for each kg of bodyweight is reached. It typically takes

up to 14 weeks to reach the recommended dose but may take longer based on your doctor's judgement.

- For children, the recommended starting dose of Xenpozyme is 0.03 mg per kg of bodyweight. This is increased every 2 weeks in a planned way with each subsequent dose until the recommended dose of 3 mg for each kg of bodyweight is reached. It typically takes up to 16 weeks to reach the recommended dose but may take longer based on your doctor's judgement.

How often will you be given Xenpozyme

- Xenpozyme will be given to you every 2 weeks.

How will Xenpozyme be given

- Xenpozyme is given to you as a drip (infusion) under the supervision of a healthcare professional who is experienced in the treatment of ASMD or other metabolic diseases.
- Your doctor may consider home infusion of Xenpozyme if you are on a stable dose and are tolerating your infusions well. This decision should be made after evaluation and recommendation by your doctor.
- If you experience a side effect during infusion, the person giving your infusion may stop the infusion and start other appropriate medical treatment.

If you miss a Xenpozyme dose

It is very important to have your infusion every 2 weeks. An infusion is considered missed if not given within 3 days from the scheduled infusion.

If you have missed an infusion or are unable to attend a scheduled appointment, call your doctor or nurse as soon as possible. Depending on the number of missed doses, your doctor may have to restart the infusion from a lower dose.

If you are given too much Xenpozyme

Your doctor or nurse will determine the correct dose for you to receive meaning an overdose is unlikely. If you receive too much (an overdose) your doctor or nurse will treat and monitor your side effects.

You can also contact the Poisons Information Centre (phone: 13 11 26) if you have any concerns.

5. What should I know whilst being given Xenpozyme?

Things you should do

Keep your medical appointments

It is important to have your infusion of Xenpozyme at the appropriate time to ensure that you do not miss an infusion.

Remind any doctor, nurse, pharmacist or dentist that you visit that you are being given Xenpozyme.

Infusion associated reactions

You may have side effects called infusion-associated reactions (IARs) that may be caused by the infusion (drip)

of the medicine. They may occur while you are being given Xenpozyme or within 24 hours after the infusion. They may include allergic reactions (see Section [6. Are there any side effects?](#)) and symptoms such as headache, a raised, itchy rash (hives), fever, nausea and vomiting.

If you think you are having an IAR, tell your doctor or nurse straight away.

If you have a severe allergic reaction during your infusion, your doctor or nurse will stop your infusion and provide appropriate medical treatment. Your doctor will make a judgement about the risks and benefits of giving you further doses of Xenpozyme.

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 allergic reactions.

Your doctor will order blood tests to check how well your liver is working (by measuring levels of your liver enzymes) before starting the treatment, and then at regular intervals as the doses are adjusted.

Driving or using machines

Be careful before you drive or use any machines or tools until you know how Xenpozyme affects you.

Xenpozyme may have a minor influence on the ability to drive and use machines because you may experience low blood pressure (which may make you feel faint).

Looking after your medicine

- Do not use this medicine after the expiry date.
- Store in a refrigerator between 2°C to 8°C.
- After reconstitution and 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 room temperature (up to 25°C)
- After dilution the solution can be stored for up to 24 hours at 2°C to 8°C followed by 12 hours (including infusion time) at room temperature (25°C)

Keep it where young children cannot reach it.

Getting rid of any unwanted medicine

Do not throw away any medicines via the wastewater or household waste. Ask your doctor or nurse how to throw away any medicines you no longer need or use.

6. Are there any side effects?

All medicines can have side effects. If you do experience any side effects, most of them are minor and temporary. However, some side effects may need medical attention.

Infusion associated reactions (IARs) have been seen while patients were being given Xenpozyme or within 24 hours after the infusion.

The most serious side effects may include sudden severe allergic reactions, raised, itchy rash (hives), rash, increased liver enzymes and irregular heartbeat.

You must tell your doctor or nurse immediately if you get an IAR or allergic reaction.

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 you appropriate medical treatment.

See the information below and, if you need to, ask your doctor or nurse if you have any further questions about side effects.

Less serious side effects

Less serious side effects	What to do
<p>General body:</p> <ul style="list-style-type: none"> • Muscle aches • Increased blood test levels for inflammation • Catheter site-related reactions including pain, itching, or swelling <p>Eye-related:</p> <ul style="list-style-type: none"> • Itchy or red eyes • Eye discomfort 	<p>Speak to your doctor or nurse if you have any of these less serious side effects and they worry you.</p>

Serious side effects

Serious side effects	What to do
<p>Signs and symptoms of infusion-associated reactions:</p> <ul style="list-style-type: none"> • Headache • Nausea • Vomiting • Raised, itchy rash (hives) • Itchy skin • Muscle aches, abdominal (belly) pain, joint pain • Fever <p>Heart-related:</p> <ul style="list-style-type: none"> • Irregular heart beat, weak or rapid pulse • Low blood pressure (you may feel faint) 	<p>Call your doctor or nurse straight away, if you notice any of these side effects, or if they worsen during or after your infusion.</p>

Very serious side effects

Very serious side effects	What to do
<p>Signs and symptoms of anaphylaxis or another serious reaction:</p> <ul style="list-style-type: none"> • Wheezing, shortness of breath • Swelling of the face, lips, tongue 	<p>Call your doctor straight away, or go straight to the Emergency Department at</p>

Very serious side effects	What to do
<ul style="list-style-type: none"> • Difficulty in breathing or swallowing • Hives, itching, skin rash 	<p>your nearest hospital if you notice any of these serious side effects.</p>

Tell your doctor or pharmacist if you notice anything else that may be making you feel unwell.

Other side effects not listed here may occur in some people.

Reporting side effects

After you have received medical advice for any side effects you experience, you can report side effects to the Therapeutic Goods Administration online at www.tga.gov.au/reporting-problems. By reporting side effects, you can help provide more information on the safety of this medicine.

Always make sure you speak to your doctor or nurse before you decide to stop taking any of your medicines.

7. Product details

This medicine is only available with a doctor's prescription.

What Xenpozyme contains

Active ingredient (main ingredient)	olipudase alfa
Other ingredients (inactive ingredients)	dibasic sodium phosphate heptahydrate methionine monobasic sodium phosphate monohydrate sucrose

Do not use this medicine if you are allergic to any of these ingredients.

What Xenpozyme looks like

Xenpozyme is a white to off-white lyophilised powder in a vial (4 mg/vial or 20 mg/vial).

After mixing with sterile water, it is a clear, colourless solution. The solution must be further diluted before infusion.

Xenpozyme is supplied in packs of 1, 5, 10 or 25 vials.

AUST R 423370 (4 mg/vial)

AUST R 394103 (20 mg/vial)

Who distributes Xenpozyme

Xenpozyme is supplied in Australia by:

sanofi-aventis australia Pty Ltd

12-24 Talavera Road

Macquarie Park NSW 2113

Toll Free No: 1800 818 806

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