Preparation and Administration of XENPOZYME for the Treatment of ASMD

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INDICATION

XENPOZYME® (olipudase alfa-rpcp) is indicated for treatment of non–central nervous system manifestations of acid sphingomyelinase deficiency (ASMD) in adult and pediatric patients.

IMPORTANT SAFETY INFORMATION

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with XENPOZYME have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Appropriate medical monitoring and support measures, including cardiopulmonary resuscitation equipment, should be readily available during XENPOZYME administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue XENPOZYME immediately, and initiate appropriate medical treatment. In patients with severe hypersensitivity reactions, a desensitization procedure to XENPOZYME may be considered.

Xenpozyme®
(olipudase alfa-rpcp)
For Injection, 20 mg

ASMD=acid sphingomyelinase deficiency.

XENPOZYME. Prescribing Information.

Please see Important Safety Information throughout this presentation and full Prescribing Information for complete details, including Boxed WARNING.

Contents

Xenpozyme® (olipudase alfa-rpcp) For Injection, 20 mg

Sanofi's Commitment

For nearly 40 years, Sanofi has pledged to help improve the care, the health, and the lives of people affected by rare diseases.

- Until now, there has historically been no FDA-approved therapy indicated for ASMD, which can be difficult to diagnose and manage.
- Patients can endure long diagnostic journeys involving multiple specialists, tests, and missed diagnoses.

Sanofi is committed to offering healthcare options to patients with unmet or underserved medical needs.

Understanding ASMD

What Is ASMD?

Historically known as Niemann-Pick disease types A, A/B, and B, ASMD impacts both children and adults.[1]

- As an autosomal recessive genetic disease, if both parents are carriers, every child has a 1-in-4 chance of being affected by ASMD.[1,2]
- The 3 subtypes of ASMD have variable onset, phenotype, and impacts on life expectancy.[1,3,4]

	Type A	Type A/B	Type B (Most Common*)
Onset	Early infancy	Infancy to childhood	Infancy to adulthood
Phenotype	Rapid progression, severe multiorgan manifestations, and neurodegeneration	Slower progression, variable multiorgan manifestations, and neurodegeneration	Slower progression, multiorgan manifestations, and little to no neurological involvement
Life expectancy	Typically less than 3 years	Childhood to mid-adulthood	Childhood to late adulthood

*Based on a patient population from a multicenter, historical cohort study (N=100).[4]

1. McGovern MM, et al. *Orphanet J Rare Dis*. 2017;12(1):41. 2. Wasserstein MP, et al. In: Adam MP, et al, eds. *GeneReviews®* [Internet]. University of Washington; 2006:1993-2019. 3. McGovern MM, et al. *Genet Med*. 2017;19(9):967-974. 4. Cox GF, et al. *JIMD Rep*. 2018;41:119-129.

What Causes ASMD?

ASMD is a progressive disease with serious, multisystemic consequences.[1]

- ASMD is caused by reduced activity of the enzyme acid sphingomyelinase (ASM).[1]
- Insufficient ASM activity causes an accumulation of sphingomyelin, which can lead to multisystemic damage, morbidity, and early mortality.[1]
- The signs and symptoms of ASMD include interstitial lung disease, splenomegaly, hepatomegaly, thrombocytopenia, and pediatric growth delay.[1-3]

1. McGovern MM, et al. *Orphanet J Rare Dis*. 2017;12(1):42. 2. McGovern MM, et al. *Genet Med*. 2017;19(9):967-974. 3. Cox GF, et al. *JIMD Rep*. 2018;41:119-129.

XENPOZYME® (olipudase alfa-rpcp): The First and Only FDA-Approved Treatment for ASMD (Non-CNS Manifestations)

CNS=central nervous system.

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XENPOZYME: The First and Only FDA-Approved Treatment for ASMD

XENPOZYME is indicated for treating non-CNS manifestations of ASMD

XENPOZYME targets the underlying cause of ASMD. As an enzyme replacement therapy, XENPOZYME provides an exogenous source of ASM.

The safety and efficacy of XENPOZYME were evaluated in 3 clinical trials including adult and pediatric patients with ASMD.

XENPOZYME is administered in 2 phases: (1) dose escalation, followed by (2) maintenance phase,* with an option of home infusion during the maintenance phase.

*3 mg/kg is the target maintenance dose, which can be administered following the dose escalation schedule.

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XENPOZYME Dosing: Overview

XENPOZYME is an intravenous infusion administered by a healthcare professional every 2 weeks.

XENPOZYME is administered in 2 phases:

- Initial dose escalation
- Maintenance phase (target dose of 3 mg/kg)

XENPOZYME administration should take place in a clinical setting throughout dose escalation, to enable management of potential severe reactions. Home infusion under the supervision of a healthcare provider may be an option during the maintenance phase, if recommended by the treating physician.

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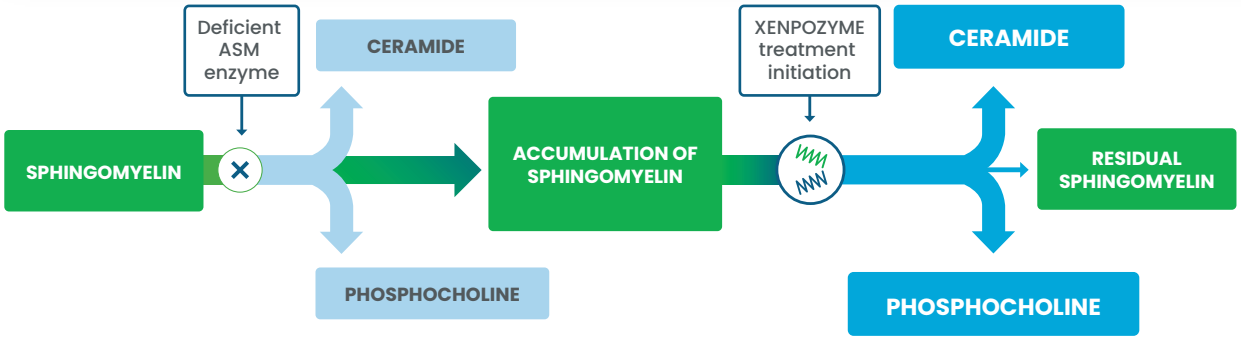
Why Dose Escalation Is Necessary

- XENPOZYME provides an exogenous source of ASM. When infused, it metabolizes accumulated sphingomyelin into ceramide and phosphocholine components.[1]
- At the start of treatment with XENPOZYME, the rapid metabolism of accumulated sphingomyelin generates pro-inflammatory breakdown products, which may induce infusion-associated reactions (IARs) and/or transient transaminase elevations.[2]



- The dose escalation regimen provides a gradual debulking of sphingomyelin and gradual release of ceramide, decreasing the inflammatory response.[1,2]

A dose escalation regimen may reduce the risk of IARs or transient transaminase elevations at treatment initiation.[1]

1. XENPOZYME. Prescribing Information. 2. Wasserstein MP, et al. *Mol Genet Metab*. 2015;116(1-2):88-97.

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Steps to Take Prior to Treatment Initiation

- **Verify pregnancy status in females of reproductive potential.**
  - XENPOZYME dosage initiation or escalation, at any time during pregnancy, is not recommended.
  - Advise female patients of reproductive potential to use effective contraception during treatment with XENPOZYME and for 14 days after the last dose if XENPOZYME is discontinued.
- **Consider administering pretreatment medication.**
  - Antihistamines, antipyretics, and/or corticosteroids may be given prior to XENPOZYME administration to reduce the risk of IARs. However, IARs may still occur in patients after receiving pretreatment.
- **Assess baseline transaminase (ALT and AST) levels** in all patients within 1 month prior to treatment initiation.

ALT=alanine aminotransferase; AST=aspartate aminotransferase.

XENPOZYME. Prescribing Information.

Important Safety Information:

Risk of Fetal Malformations

Risk of Fetal Malformations During Dosage Initiation or Escalation in Pregnancy

- XENPOZYME dosage initiation or escalation, at any time during pregnancy, is not recommended as it may lead to elevated sphingomyelin metabolite levels that may increase the risk of fetal malformations.
- The decision to continue or discontinue XENPOZYME maintenance dosing in pregnancy should consider the female's need for XENPOZYME, the potential drug-related risks to the fetus, and the potential adverse outcomes from untreated maternal ASMD disease.

XENPOZYME. Prescribing Information.

Important Safety Information:

Monitoring Transient Transaminase (ALT and AST) Elevations

Transaminase levels should be assessed to manage the risk of transient transaminase elevations prior to and during dose escalation, or upon resuming treatment following a missed dose

- XENPOZYME may be associated with transaminase elevations within 24 to 48 hours after infusion.
- Transaminase elevations generally returned to pre-infusion levels at the time of the next scheduled infusion.

Transaminase testing is recommended to be continued as part of routine clinical management.

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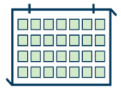
Important Safety Information:

Monitoring Transient Transaminase (ALT and AST) Elevations (Cont.)

The following monitoring guidance must be followed to manage the risk of transient transaminase elevations:

WITHIN 1 MONTH PRIOR TO TREATMENT INITIATION:



- Assess baseline ALT and AST levels.

DURING DOSE ESCALATION OR UPON RESUMING TREATMENT FOLLOWING A MISSED DOSE:

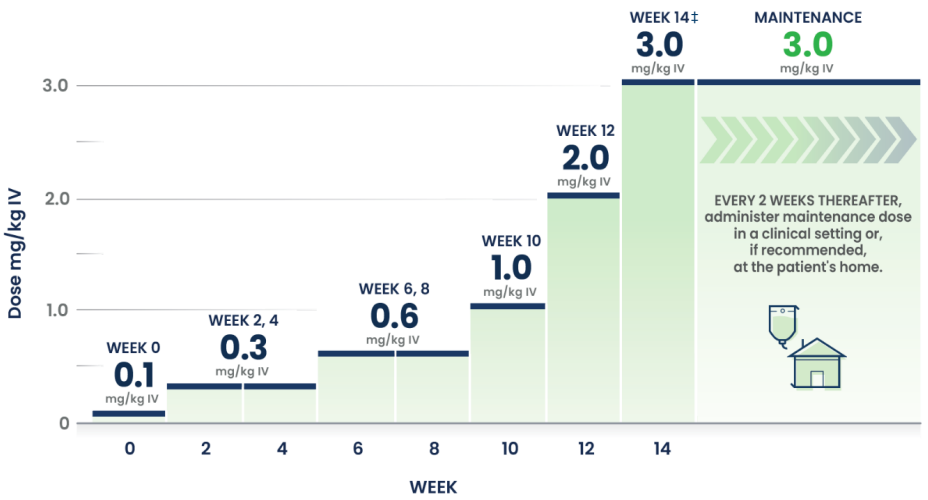
- Assess transaminase levels within 72 hours prior to any infusion during dose escalation, or prior to the next scheduled XENPOZYME infusion upon resuming treatment following a missed dose.
  - If transaminase levels are elevated above baseline and >2 times the ULN, the XENPOZYME dose can be adjusted (prior dose repeated or reduced) or treatment can be temporarily withheld until the liver transaminases return to the patient's baseline value.
  - If either the baseline or pre-infusion transaminase level (during the dose escalation phase) is >2 times the ULN, repeat assessment of transaminase levels within 72 hours after the end of the infusion to monitor trends in transaminase elevations.

ULN=upper limit of normal.

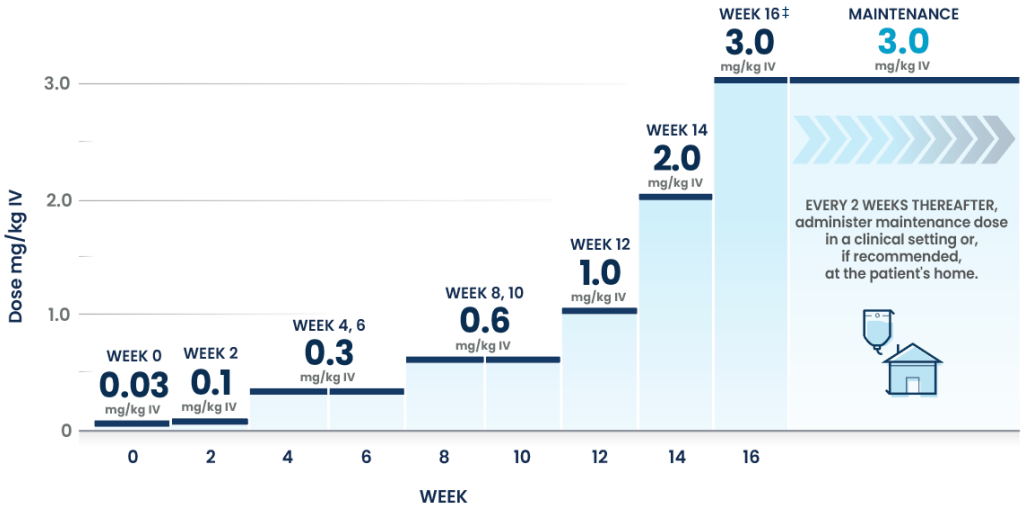
XENPOZYME. Prescribing Information.

XENPOZYME Dose Escalation Regimens

Adult Dose Escalation and Maintenance*



Pediatric Dose Escalation and Maintenance*†



Initial dose escalation should take place in a clinical setting, and takes at least 14 weeks for adults and at least 16 weeks for pediatric patients. If doses are missed, re-escalation may be necessary.

*Use actual body weight for patients with a body mass index (BMI) ≤30. For patients with a BMI >30, calculate adjusted body weight (kg) = (actual height in m)^2 x 30.

†In the clinical trial in pediatric patients, all but 1 patient completed the dose escalation up to the maintenance dose of 3 mg/kg within 22 weeks.

‡The dose escalation phase includes the first 3 mg/kg dose.

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Missed Doses: Re-escalation May Be Necessary if a Patient Misses 1 or More Doses*

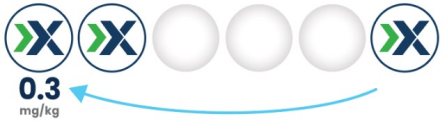
A dose is considered missed when not administered within 3 days of the scheduled date. When a dose of XENPOZYME is missed, administer the next dose as described below as soon as possible. Thereafter, administration should be scheduled every 2 weeks from the date of the last administration.

During the dose escalation phase:

If 1 infusion is missed, administer the last tolerated dose before resuming dose escalation, according to the dose escalation regimens for adult and pediatric patients (see slide 15).

If 2 consecutive infusions are missed, administer 1 dose below the last tolerated dose before resuming dose escalation, according to the dose escalation regimens for adult and pediatric patients (see slide 15).

If 3 or more consecutive infusions are missed, resume dose escalation at 0.3 mg/kg, according to the dose escalation regimens for adult and pediatric patients (see slide 15).

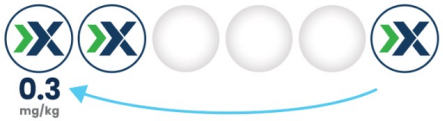


During the maintenance phase:

If 1 maintenance infusion is missed, administer the maintenance dose and adjust the treatment schedule accordingly.

If 2 consecutive maintenance infusions are missed, administer 1 dose below the maintenance dose before resuming the maintenance dose.

If 3 or more consecutive maintenance infusions are missed, restart dosing at 0.3 mg/kg, according to the dose escalation regimens for adult and pediatric patients (see slide 15).



*At the next scheduled infusion after a missed dose, if the dose administered is 0.3 mg/kg or 0.6 mg/kg, that dose should be administered twice per Tables 1 and 2 in the XENPOZYME full Prescribing Information. For complete dose escalation regimens for adult and pediatric patients, please see slide 15 of this guide and Tables 1 and 2 in the XENPOZYME full Prescribing Information.

XENPOZYME. Prescribing Information.

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Storage and Supplies

Storage

- XENPOZYME is supplied as a sterile, white to off-white lyophilized powder for reconstitution in a single-dose vial.
- XENPOZYME is available in 4 mg or 20 mg vials.
- Store XENPOZYME vials refrigerated at 2 °C to 8 °C (36 °F to 46 °F).
- XENPOZYME contains no preservatives.
- **If the reconstituted XENPOZYME vial is not used immediately,** store refrigerated for up to 24 hours at 2 °C to 8 °C (36 °F to 46 °F) or at controlled room temperature for up to 6 hours at 20 °C to 25 °C (68 °F to 77 °F).
- **If the diluted solution is not administered immediately,** refrigerate at 2 °C to 8 °C (36 °F to 46 °F) for up to 24 hours or store at controlled room temperature at 20 °C to 25 °C (68 °F to 77 °F) for up to 12 hours (including infusion time), or discard.
- **Do not freeze.**

Supplies

- Refrigerated XENPOZYME (lyophilized powder for reconstitution)
- Sterile water for injection, USP: 1.1 mL (4 mg vial) or 5.1 mL (20 mg vial)
- Empty syringe or infusion bag (depending on infusion volume)
  - Appropriate infusion pump (for syringe or infusion bag)
- 0.9% sodium chloride injection, USP
- In-line low protein-binding 0.2-µm filter

20 mg vial pictured. XENPOZYME is available in 4 mg or 20 mg vials.

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(olipudase alfa-rpcp)
For Injection, 20 mg

XENPOZYME. Prescribing Information.

Infusion Preparation

Determine the number of vials to be reconstituted
based on patient's weight and the prescribed dose.*

Patient dose (mg)
Patient weight (kg) **x dose** (mg/kg)

Vials required
Patient dose (mg) **X** mg/vial

If the number of vials includes a fraction, round up to the next whole number.
X = the vial size (either 4 mg or 20 mg, depending on which vial size is being used).

XENPOZYME is supplied as 4 mg or 20 mg of lyophilized powder in a single-dose vial for reconstitution.

*In patients with a BMI >30, calculate an adjusted body weight (kg) based on height in meters as described:
Adjusted body weight (kg) = (actual height in m)^2 x 30.

XENPOZYME. Prescribing Information.

Infusion Preparation (Continued)

1 **Complete the reconstitution and dilution steps under aseptic conditions.**

2 **Remove the required number of vials** from refrigeration and set aside for approximately 20 to 30 minutes to allow them to reach room temperature.

3 **Reconstitute each vial** with 1.1 mL (4 mg vial) or 5.1 mL (20 mg vial) of sterile water for injection, USP, by directing the diluent flow to the inside wall of the vial to avoid foaming.

4 **Tilt and roll each vial gently** between the palms and avoid foaming. Each reconstituted vial will yield a 4 mg/mL clear, colorless solution.

XENPOZYME. Prescribing Information.

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Infusion Preparation (Continued)

5 **Visually inspect the reconstituted solution** in the vials for particulate matter and discoloration.

The solution should be clear and colorless.

Discard if the solution is discolored or if visible particulate matter is present.

Do not infuse XENPOZYME in the same intravenous line with other products.

XENPOZYME. Prescribing Information.

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Infusion Administration

6 **Withdraw the required volume of XENPOZYME from the vial(s) and dilute the XENPOZYME solution for infusion with 0.9% sodium chloride injection, USP, in a syringe or infusion bag depending on the volume of infusion.**

- For patients who weigh less than 10 kg receiving 0.03 mg/kg and 0.1 mg/kg and patients who weigh between 10 to 20 kg receiving 0.03 mg/kg dose, the volume of infusion will vary to achieve a fixed final concentration of 0.1 mg/mL. Prepare the required dose diluted to a final concentration of 0.1 mg/mL in a syringe for infusion.
- For all other patient weights and doses, the final concentration will vary to achieve a fixed total volume.

XENPOZYME. Prescribing Information.

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Infusion Administration (Continued)

6 **Withdraw the required volume of XENPOZYME from the vial(s) and dilute the XENPOZYME solution for infusion with 0.9% sodium chloride injection, USP, in a syringe or infusion bag depending on the volume of infusion.**

For a total volume ≤20 mL, prepare a syringe for infusion:

- Inject the required volume of the reconstituted XENPOZYME solution (4 mg/mL) slowly down the inside wall of the syringe.
- Slowly add the quantity sufficient of 0.9% sodium chloride injection, USP, to obtain the required total infusion volume.
- Avoid foaming within the syringe.

For a total volume ≥50 mL, prepare an infusion bag:

- Slowly add the required volume of the reconstituted XENPOZYME solution (4 mg/mL) into the appropriate size 0.9% sodium chloride injection, USP, infusion bag to achieve a fixed total volume based on infusion volumes table on the next slide.
- Avoid foaming within the bag.

If the dose calls for only a portion of the vial volume, do not use the vial in its entirety. Discard any unused solution.

XENPOZYME. Prescribing Information.

Infusion Volumes Based on Body Weight*

	PEDIATRIC PATIENTS (0 TO 17 YEARS)			ADULT PATIENTS ≥18 YEARS
	BODY WEIGHT (≥2 kg to <10 kg)	BODY WEIGHT (≥10 kg to <20 kg)	BODY WEIGHT (≥20 kg)	BODY WEIGHT (≥20 kg)
XENPOZYME DOSE (mg/kg)	Total infusion volume† (mL)	Total infusion volume† (mL)	Total infusion volume† (mL)	Total infusion volume† (mL)
0.03	Actual volume will vary‡ (0.6 mL to 3 mL)	Actual volume will vary‡ (3 mL to 6 mL)	5	NA
0.1	Actual volume will vary‡ (2 mL to 10 mL)	5	10	20
0.3	5	10	20	100
0.6	10	20	50	100
1.0	20	50	100	100
2.0	50	75	200	100
3.0	50	100	250	100

Prior to administration, inspect the syringe or infusion bag for foaming. If foaming is present, let foam dissipate before administering XENPOZYME.

*Use actual or adjusted body weight per patient BMI. Use actual body weight (kg) for patients with a BMI ≤30. For patients with a BMI >30, calculate an adjusted body weight (kg) based on height in meters = (actual height in m)^2 x 30.
†For a total volume of infusion ≤20 mL, prepare a syringe for infusion. For a total volume of ≥50 mL, prepare an infusion bag.
‡Volume will vary to achieve a final concentration of 0.1 mg/mL.

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Infusion Administration (Continued)

7 **Gently invert the syringe or the infusion bag to mix. Do not shake.**

- Because this is a protein solution, slight flocculation (described as thin translucent fibers) occurs occasionally after dilution.
- The diluted solution must be filtered through an in-line low protein-binding 0.2-µm filter during administration.

8 **Vials are for single dose only.** Discard any unused solution.

9 **After the infusion is complete, the infusion line should be flushed** with 0.9% sodium chloride injection, USP, using the same infusion rate as the one used for the last part of the infusion.

10 **Do not infuse XENPOZYME in the same intravenous line with other products.**

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Infusion Rates for Adult Patients

Total time for infusion is usually between 3.5-4 hours.

DOSE (mg/kg)	INFUSION RATE			
	step 1	step 2	step 3	step 4
0.1	20 mL/hr	60 mL/hr	NA	NA
0.3 to 3	3.33 mL/hr	10 mL/hr	20 mL/hr	33.33 mL/hr

Do not infuse XENPOZYME in the same intravenous line with other products.

- In the absence of IARs, increase infusion rate per the steps of infusion as indicated (+/- 5 min).
- Each step of infusion will last for 20 minutes with the exception of the final step, which should last until completion of the infusion volume.

XENPOZYME. Prescribing Information.

Infusion Rates for Pediatric Patients

DOSE (mg/kg)	INFUSION RATE			
	step 1	step 2	step 3	step 4
0.03	0.1 mg/kg/hr for the full length of the infusion	NA	NA	NA
0.1	0.1 mg/kg/hr	0.3 mg/kg/hr	NA	NA
0.3	0.1 mg/kg/hr	0.3 mg/kg/hr	0.6 mg/kg/hr	NA
0.6	0.1 mg/kg/hr	0.3 mg/kg/hr	0.6 mg/kg/hr	1 mg/kg/hr
1.0				
2.0				
3.0				

Total time for infusion is usually between 3.5-4 hours.

Do not infuse XENPOZYME in the same intravenous line with other products.

- In the absence of IARs, increase infusion rate per the steps of infusion as indicated (+/- 5 min).
- Each step of infusion will last for 20 minutes with the exception of the final step, which should last until completion of the infusion volume.

XENPOZYME. Prescribing Information.

Important Safety Information:

Monitoring Infusion-Associated Reactions and Hypersensitivity

Observe patients closely during and for an appropriate period of time after the infusion, based on clinical judgment.

DURING AND AFTER THE INFUSION:

- If a *severe* hypersensitivity reaction occurs, discontinue XENPOZYME immediately and initiate appropriate medical treatment. Consider the risks and benefits of re-administering XENPOZYME following severe hypersensitivity reactions.
- If a *mild or moderate* hypersensitivity reaction occurs, consider temporarily holding the infusion, slowing the infusion rate, and/or reducing the XENPOZYME dose.
- If *severe* IARs occur, discontinue XENPOZYME immediately and initiate appropriate medical treatment. Consider the risks and benefits of re-administering XENPOZYME following severe IARs.
- If *a mild or moderate* IAR occurs, the infusion rate may be slowed or temporarily withheld, and/or the XENPOZYME dosage may be reduced.
- Consider testing for IgE ADA, serum tryptase, and complement activation in patients who experience anaphylaxis.

Ensure appropriate medical support measures, including cardiopulmonary resuscitation equipment, are readily available during XENPOZYME administration.

ADA=antidrug antibody; IgE=immunoglobulin E.

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Important Safety Information:

Monitoring Infusion-Associated Reactions and Adverse Drug Reactions

Acute phase reactions (APRs)

› Acute phase reactions (APRs), acute inflammatory responses accompanied by elevations in inflammatory serum protein concentrations, have been observed. Most APRs occurred at 48 hours post infusion during the dose escalation period. APRs were managed similar to other IARs.

Adverse drug reactions

› Most frequently reported adverse drug reactions in adults (incidence ≥10%) were headache, cough, diarrhea, hypotension, and ocular hyperemia.

› Most frequently reported adverse drug reactions in pediatric patients (incidence ≥20%) were pyrexia, cough, diarrhea, rhinitis, abdominal pain, vomiting, headache, urticaria, nausea, rash, arthralgia, pruritus, fatigue, and pharyngitis.

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Sanofi Resources and Support

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Sanofi Resources and Support

For patients living with rare diseases and their caregivers, having programs and resources developed specifically for them can be an important part of the treatment experience. With this in mind, Sanofi created CareConnect Personalized Support Services.

CareConnect was built to support eligible patients throughout their diagnosis and treatment journey. CareConnect patient support ranges from assistance with insurance coverage to community connection.

STARTING TREATMENT

Whether an eligible patient is newly diagnosed or trying a systemic therapy, CareConnect has people who can help.

INSURANCE CHANGES

Sometimes insurance information changes. CareConnect can help assist eligible patients transition without impacting access to treatment.

TRANSITION OF CARE

CareConnect can assist eligible patients making the transition from the hospital to home infusions.

RESOURCE CONNECTIONS

CareConnect can assist eligible patients and their support system find helpful information and tools.

VISIT CARECONNECTPSS.COM

careconnect™
PERSONALIZED SUPPORT SERVICES

FOR MORE INFORMATION ON XENPOZYME, VISIT XENPOZYME.COM/HCP

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Appendix: Sample Order Form Examples

Xenpozyme®
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XENPOZYME (olipudase alfa-rpcp): Sample Order Form Example Biweekly IV Infusion for Adult Patients

Patient Information

First & Last Name: _____ **Date of Birth:** _____ **Infusion Date:** _____ **Pre-Infusion Transaminase Assessment Date:** _____

Current Weight (kg): _____ **U#/Medical Record:** _____ **ICD-10: E75.249**

Confirm Patient's Stage in XENPOZYME Dosing Regimen

Week	0	2	4	6	8	10	12	14	Maintenance
Dose (mg/kg)	0.1	0.3	0.3	0.6	0.6	1.0	2.0	3.0	3.0
Infusion Vol. (mL)	20	100	100	100	100	100	100	100	100
Check One →									

Allergies: _____

Other Notes: _____

_____

_____

XENPOZYME Preparation (Please reference full Prescribing Information for preparation and administration instructions)

1 Determine the number of vials to be reconstituted based on the individual patient's weight and the prescribed dose.

Required # of Vials: _____

Patient dose (mg)
Patient weight (kg) **x dose** (mg/kg)

Vials required
$\frac{\text{Patient dose (mg)}}{\text{X mg/vial}}$

If the number of vials includes a fraction, round up to the next whole number.
X = the vial size (either 4 mg or 20 mg, depending on which vial size is being used).

In patients with a BMI >30, the body weight used to calculate the dose of XENPOZYME is calculated via the following method:
Body weight (kg) = (actual height in m)^2 x 30.

2 Reconstitute each vial with 1.1 mL (4 mg vial) or 5.1 mL (20 mg vial) of sterile water for injection, USP, by directing the diluent flow to the inside wall of the vial to avoid foaming.

3 Withdraw the required volume of reconstituted solution and dilute to final volume with 0.9% sodium chloride injection, USP, in a syringe for final volumes up to 20 mL, or in an infusion bag for volumes ≥50 mL.

4 Based on patient's dose confirmed in the patient information section above, make note of the infusion rate(s) in the table shown to the right for the infusion team to reference.

Check one →		
Dose (mg/kg)	0.1	0.3 - 3.0
Step	Infusion Rate (mL/hr)	
1	20	3.33
2	60	10
3	NA	20
4	NA	33.33

Xenpozyme® (olipudase alfa-rpcp) For Injection, 20 mg

XENPOZYME (olipudase alfa-rpcp): Sample Order Form Example
Bi-weekly IV Infusion for Pediatric Patients

Patient Information

First & Last Name: _____ **Date of Birth:** _____ **Infusion Date:** _____ **Pre-Infusion Transaminase Assessment Date:** _____

Current Weight (kg): _____ **U#/Medical Record:** _____ **ICD-10: E75.249**

Confirm Patient's Body Weight & Stage in XENPOZYME Dosing Regimen

Body Weight (kg)	Week	0	2	4	6	8	10	12	14	16	Maintenance
↓ Check One	Dose (mg/kg)	0.03	0.1	0.3	0.3	0.6	0.6	1.0	2.0	3.0	3.0
	≥2 to <10	Infusion Vol. (mL)	†	††	5	5	10	10	20	50	50	50
	≥10 to <20	Infusion Vol. (mL)	†††	5	10	10	20	20	50	75	100	100
	≥20	Infusion Vol. (mL)	5	10	20	20	50	50	100	200	250	250
		Check One →										

Allergies: _____

Other Notes: _____

Actual volumes will vary for these doses to achieve a final concentration of 0.1 mg/mL:

† (0.6 mL to 3 mL)
†† (2 mL to 10 mL)
††† (3 mL to 6 mL)

XENPOZYME Preparation (Please reference full Prescribing Information for preparation and administration instructions)

1 Determine the number of vials to be reconstituted based on the individual patient's weight and the prescribed dose.

Required # of Vials: _____

Patient dose (mg)
Patient weight (kg) **x dose** (mg/kg)

Vials required
Patient dose (mg) / X mg/vial

If the number of vials includes a fraction, round up to the next whole number.
X = the vial size (either 4 mg or 20 mg, depending on which vial size is being used).

In patients with a BMI >30, the body weight used to calculate the dose of XENPOZYME is calculated via the following method: Body weight (kg) = (actual height in m)^2 x 30.

2 Reconstitute each vial with 1.1 mL (4 mg vial) or 5.1 mL (20 mg vial) of sterile water for injection, USP, by directing the diluent flow to the inside wall of the vial to avoid foaming.

3 Withdraw the required volume of reconstituted solution and dilute to final volume with 0.9% sodium chloride injection, USP, in a syringe for final volumes up to 20 mL, or in an infusion bag for volumes ≥50 mL.

4 Based on patient's dose confirmed in the patient information section above, make note of the infusion rate(s) in the table shown to the right for the infusion team to reference.

Check one →				
Dose (mg/kg)	0.03	0.1	0.3	0.6 – 3.0
Step	Infusion Rate (mg/kg/hr)			
1	0.1	0.1	0.1	0.1
2	NA	0.3	0.3	0.3
3	NA	NA	0.6	0.6
4	NA	NA	NA	1.0

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Estimated Approximate Infusion Durations by XENPOZYME Dose

Note: The tables below indicate estimated approximate total infusion durations by dose for both adult and pediatric patients receiving XENPOZYME. These values should be considered for reference only to help facilitate infusion site planning. All durations shown below are estimates only, and based on infusions that occur without infusion-associated reactions (IARs) or other interruptions.

XENPOZYME Infusion Rates and Estimated Infusion Durations for Adult Patients

Dose (mg/kg)	0.1	0.3 - 3.0
Step	Infusion Rate (mL/hr)	
1	20	3.33
2	60	10
3	NA	20
4	NA	33.33
Estimated Approximate Total Infusion Duration (Minutes)	~35	~220

XENPOZYME Infusion Rates and Estimated Infusion Durations for Pediatric Patients

Dose (mg/kg)	0.03	0.1	0.3	0.6	1.0	2.0	3.0
Step	Infusion Rate (mg/kg/hr)						
1	0.1	0.1	0.1	0.1	0.1	0.1	0.1
2	NA	0.3	0.3	0.3	0.3	0.3	0.3
3	NA	NA	0.6	0.6	0.6	0.6	0.6
4	NA	NA	NA	1.0	1.0	1.0	1.0
Estimated Approximate Total Infusion Duration (Minutes)	~18	~35	~60	~80	~100	~160	~220

Times above do not include the time required to flush the line with 0.9% sodium chloride injection, USP, at the final infusion rate. Times will vary depending on the volume of the tubing.

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