sanofi

DESIGNED FOR CHANGE FROM THE INSIDE OUT

For your patients with Pompe disease

NEXVIAZYME is indicated for long-term enzyme replacement therapy for the treatment of patients one year of age and older with Pompe disease (acid α-glucosidase deficiency).[1]

▼ This medicinal product is subject to additional monitoring in Australia. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse events at www.tga.gov.au/reporting-problems.

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme®
(avalglucosidase alfa)

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Introducing NEXVIAZYME

Nexviazyme®
(avalglucosidase alfa)

What is NEXVIAZYME (avalglucosidase alfa)?

The development of NEXVIAZYME marks the next step in Sanofi's commitment to the Pompe community

- NEXVIAZYME is indicated for long-term enzyme replacement therapy for the treatment of patients 1 year of age and older with Pompe disease (acid α-glucosidase deficiency)[1]
- Monotherapy administered every other week via intravenous infusion, supervised by an experienced physician in the management of Pompe disease[1]
- Replaces the GAA enzyme, which is lacking or dysfunctional in people living with Pompe disease[1]

NEXVIAZYME is an ERT that has the same mechanism of action as MYOZYME® – **enhanced with more M6P for increased cellular uptake[1–3]**

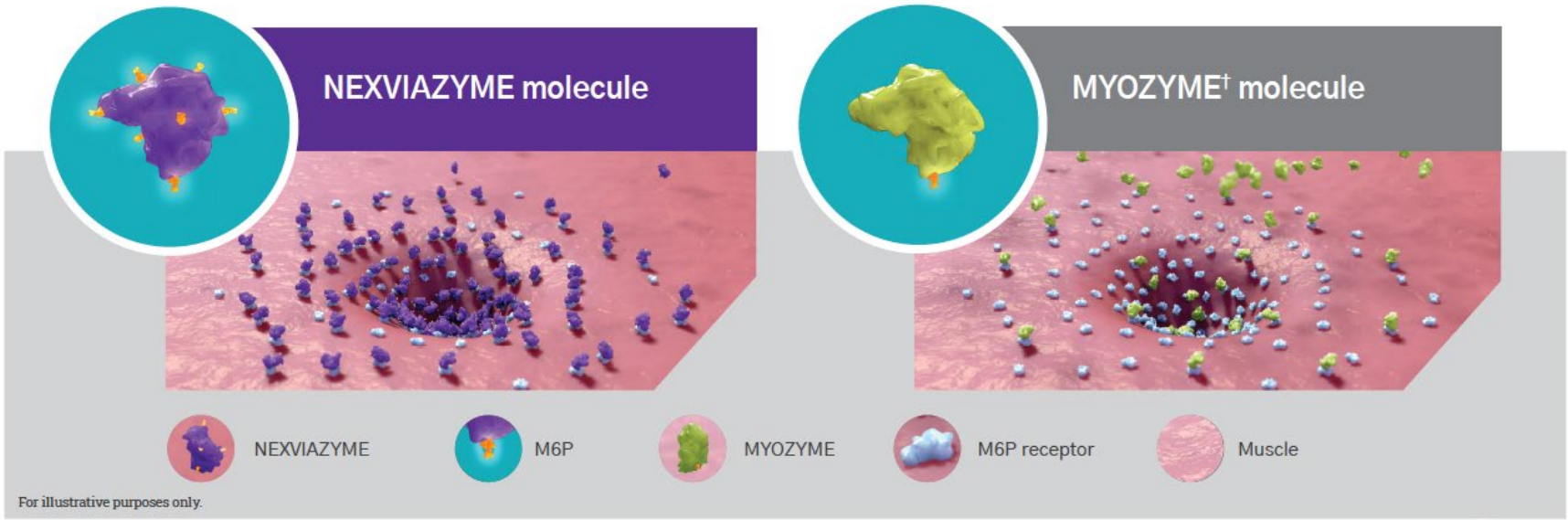
Abbreviations: ERT, enzyme replacement therapy; GAA, acid alpha-glucosidase; M6P, mannose 6-phosphate.

1. NEXVIAZYME Australian Approved Product Information. 2. Zhu Y *et al. Mol Ther* 2009; 17(6): 954–63. 3. Zhu Y *et al. Biochem J* 2005; 389(3): 619–28.

Nexviazyme (avalglucosidase alfa)

NEXVIAZYME has 15x more M6P moieties than MYOZYME®[1–3]

NEXVIAZYME is an enzyme replacement therapy that has been designed to have enhanced M6P content vs. MYOZYME, for increased uptake into muscle cells.[1–3*]



Upon uptake, the GAA enzyme can degrade and clear lysosomal glycogen to help prevent irreversible muscle damage[1,2]

*NEXVIAZYME displayed increased uptake into murine cells vs MYOZYME (P-vale not assessed). Animal data does not necessarily predict human clinical effects.

†MYOZYME was the first ERT approved for the treatment of Pompe disease.[4]

Abbreviations: ERT, enzyme replacement therapy; M6P, mannose 6-phosphate.

1. NEXVIAZYME Australian Approved Product Information. 2. Zhu Y *et al. Mol Ther* 2009; 17(6): 954–63. 3. Zhu Y *et al. Biochem J* 2005; 389(3): 619–28. 4. van der Ploeg A *et al. N Engl J Med* 2010; 362(15): 1396–406.

Nexviazyme® (avalglucosidase alfa)

When to use NEXVIAZYME

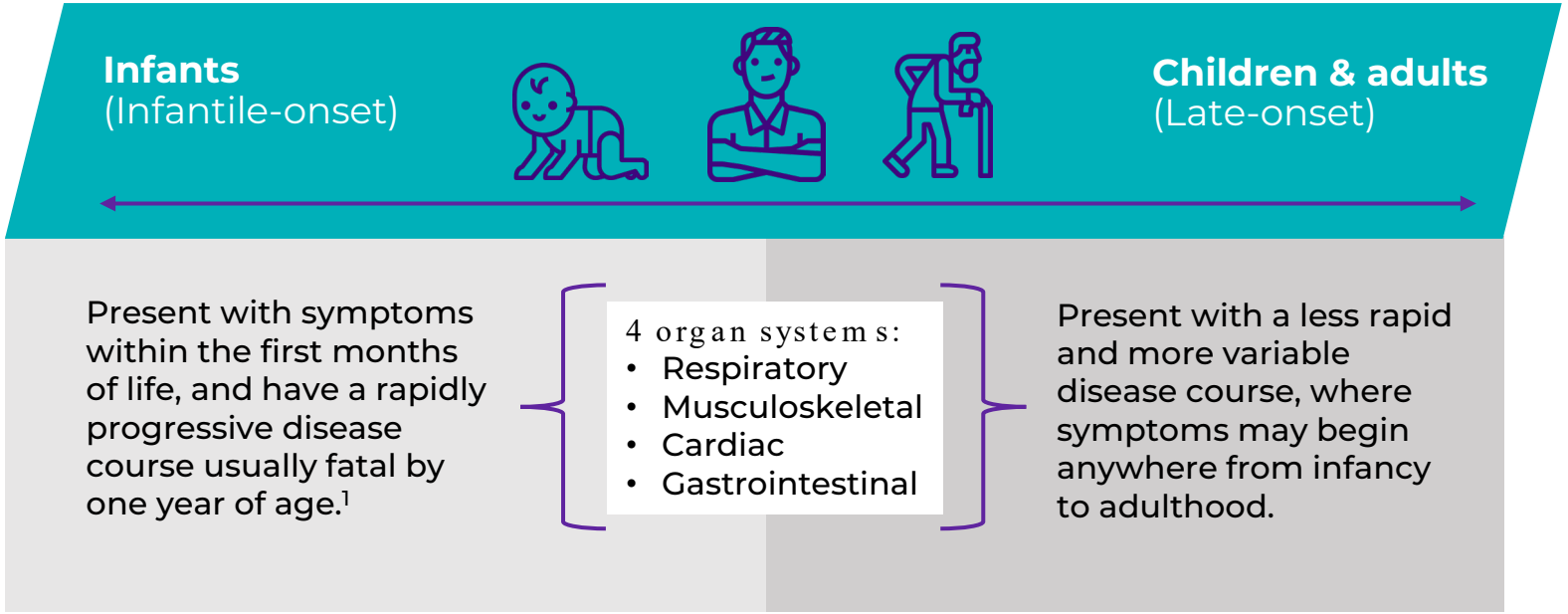
Nexviazyme®
(avalglucosidase alfa)

Disease spectrum: one disease with variable presentations

Differences in disease presentation are observed among infants, children and adults. Patients typically present with symptoms that manifest in four organ systems.[1]

Myozyme® (alglucosidase alfa)

For patients <1 year of age



Nexviazyme® (avalglucosidase alfa)

A new ERT for patients ≥1 year of age

The level of residual acid alfa-glucosidase (GAA) activity is considered to be associated with severity and rate of disease progression.[2]

Abbreviations: ERT, enzyme replacement therapy..

1. Kishnani PS *et al. Gen Med* 2006; 8: 267–88. 2. Hirschhorn and Reuser. The Metabolic and Molecular Basis of Inherited Disease. 2001, 8th edition.

Common signs and symptoms in children and adults with late-onset Pompe Disease (LOPD)

Children and adults experience steady degeneration of respiratory and skeletal muscles.[1]

Respiratory

- Respiratory failure / insufficiency
- Diaphragm weakness
- Sleep-disordered breathing
- Orthopnoea
- Dyspnoea

Musculoskeletal

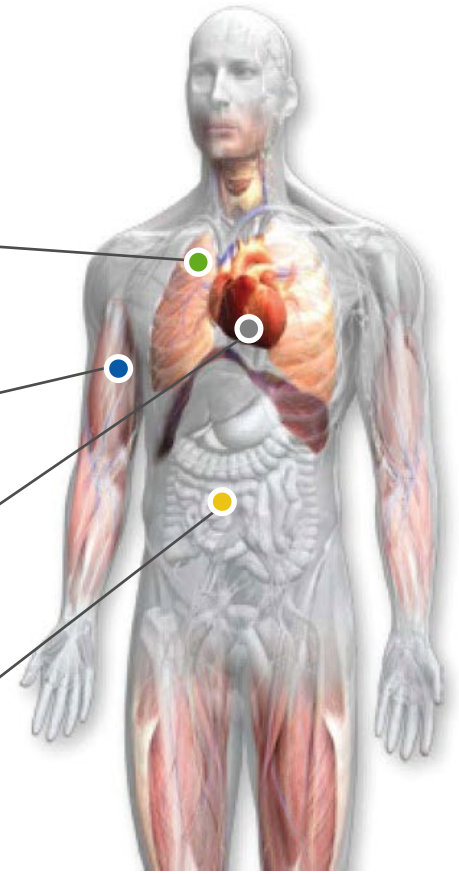
- Limb-girdle muscle weakness
- Muscle pain
- Frequent falls
- Gait abnormalities
- Difficulty walking, climbing stairs
- Scoliosis / scapular winging

Cardiac

- Less common among adults

Gastrointestinal

- Difficulty chewing / jaw muscle fatigue
- Poor weight gain / maintenance
- Swallowing difficulties / weak tongue/macroglossia
- Gastroesophageal reflux

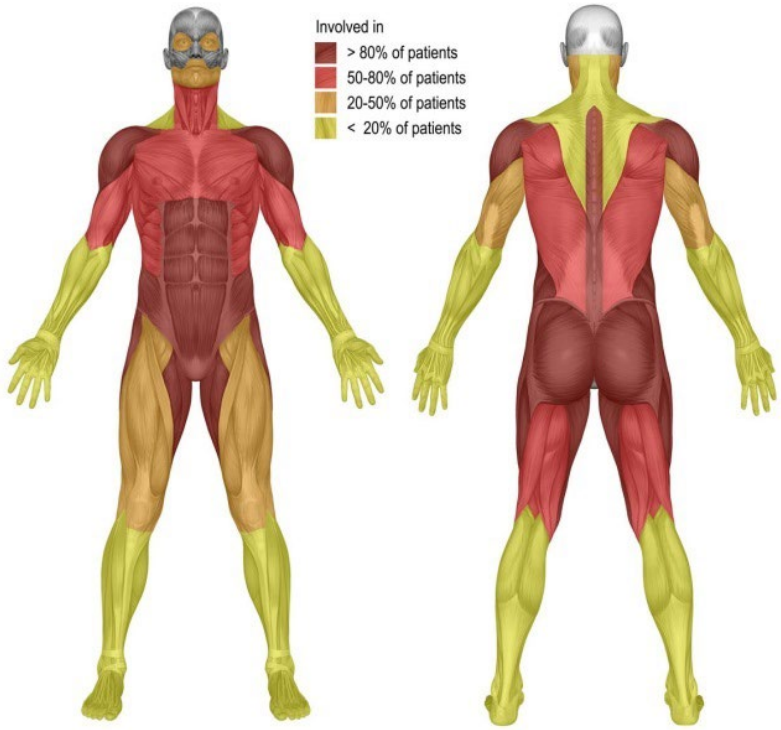


1. Kishnani PS, et al. Gen Med 2006; 8: 267-88

Nexviazyme® (avalglucosidase alfa)

Proximal muscle weakness affects most LOPD patients

- In a natural history study of 94 adults with LOPD, proximal trunk and limb muscles were weakened in 50–80% of patients.
- Fewer patients had weakness more distally.



Abbreviations: LOPD, late-onset Pompe disease

1. Van der Beek NA et al. Orphanet J Rare Dis. 2012;7(1):88, Fig. 2A

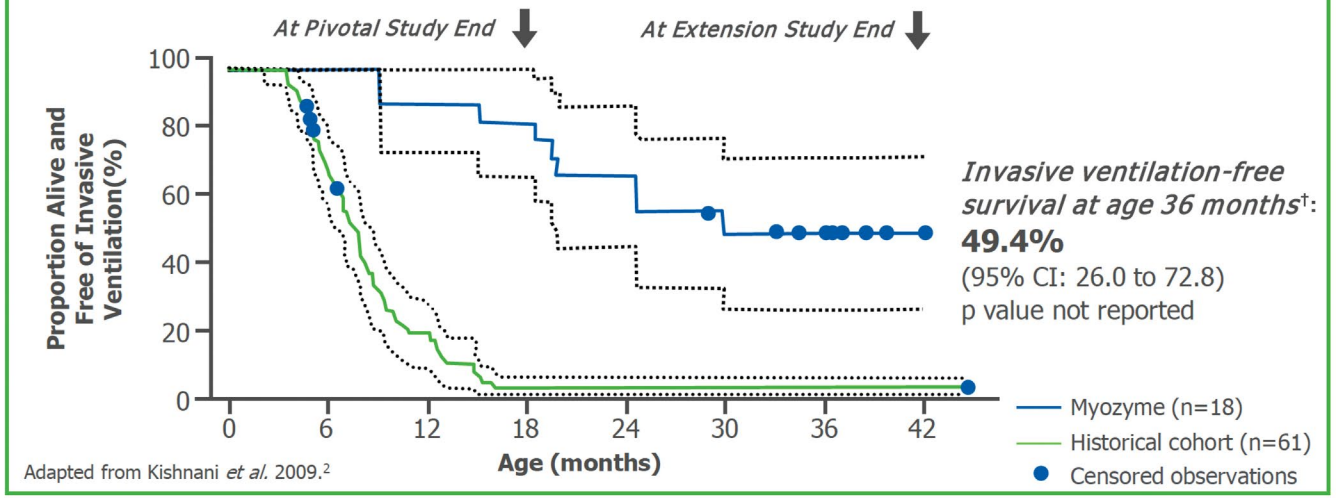
Nexviazyme® (avalglucosidase alfa)

MYOZYME in IOPD

MYOZYME prolongs survival and survival free of invasive ventilation in infants

Study 1 and the extension study demonstrated that MYOZYME markedly extended survival (72% survival rate at 36 months, 95% CI: 47.9 - 96.0, p value not reported) and survival free of invasive ventilation (49.4% invasive ventilation-free survival rate at 36 mo, 95% CI: 26.0 - 72.8, *P*-value not reported), compared with a historical control group.[1,2]

Proportion alive and free of invasive ventilation at end of Study 1 Extension



†Four patients were right-censored from this analysis because they had not reached age 36 months by the end of the study, although they remained free of invasive ventilation that time.

Abbreviations: IOPD, infantile-onset Pompe disease..

Study design: Patients aged <7 months with documented symptoms of IOPD (n=18) were treated with MYOZYME 20 or 40* mg/kg administered by iv infusion every other week. A closely matched untreated historical control group (n=61) was used as comparator. The 52-week study was extended (n=16) with a median treatment duration of 2.3 years. Primary endpoint: invasive ventilation-free survival.[1,2]

* The recommended dosage regimen of MYOZYME in Australia is 20 mg/kg of body weight administered once every 2 weeks as an intravenous infusion.[3]

1. Kishnani PS *et al. Neurol* 2007; 68: 99–109. 2. Kishnani PS *et al. Pediatr Res* 2009; 66(3): 329–35. 3. MYOZYME Australian Approved Product Information.

NEXVIAZYME in LOPD

NEXVIAZYME offers patients with LOPD favourable differences in their motor and respiratory function at Week 97 (6MWT estimated difference: 30.01 m, nominal *P*=0.04; FVC [% predicted] estimated difference: 2.43%, *P*=0.06 [not significant])[1]

MORE M6P:
NEXVIAZYME has 15× more M6P moieties than MYOZYME[1]

EFFICACY OUTCOMES:
Favourable improvements as compared to MYOZYME[1]

(6MWT estimated difference: 30.01 m, nominal *P*=0.04; FVC [% predicted] estimated difference: 2.43%, *P*=0.06 [not significant])

SIMPLE TO START OR SWITCH:
NEXVIAZYME contains 100mg of medication per vial, compared to 50mg per vial for MYOZYME[1,2]

Nexviazyme is a **monotherapy** administered by intravenous infusion without fasting or stabilisers.

SAFETY DEMONSTRATED IN A PHASE 3 TRIAL:
With NEXVIAZYME, fewer patients with LOPD experienced IARs and SARs as compared to MYOZYME[1]

(*P*–value not assessed)

Abbreviations: LOPD, late-onset Pompe disease

1. NEXVIAZYME Australian Approved Product Information. 2. MYOZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Safety profile of NEXVIAZYME across 4 clinical trials[1]

Most common adverse drug reactions

- The most frequently reported adverse drug reactions (>5%) were headache, nausea, pruritus, rash, urticaria, fatigue, and chills.

Undesirable effects

- In clinical studies, IARs were reported to occur in patients at any time during and/or within a few hours after the infusion of NEXVIAZYME and were more likely with higher infusion rates.
    - 3 (2.2%) patients reported severe IARs including symptoms of chest discomfort, nausea, and increased blood pressure

Immunogenicity

- Treatment-emergent ADAs were reported in both treatment-naïve (95%) and treatment-experienced patients (49%). In the COMET trial, ADAs did not impact measures of efficacy while limited impacts on pharmacokinetics and pharmacodynamics were observed primarily with high-titre patients.
- In adult patients with LOPD, 1 treatment-naive patient and 1 treatment-experienced patient developed anaphylaxis.
- In paediatric patients with IOPD or LOPD, no patients developed anaphylactic reactions.

Abbreviations: ADAs, antidrug antibodies; IAR, infusion-associated reaction; IOPD, infantile-onset Pompe disease; LOPD, late-onset Pompe disease.

1. NEXVIAZYME Australian Approved Product Information.

Fewer patients with LOPD experienced IARs and serious adverse reactions* vs MYOZYME (*P*-value not assessed)[1]

Safety profile of NEXVIAZYME in the phase 3 COMET trial

Patients with LOPD experiencing undesirable effects (%)[1]				
TREATMENT GROUP	IARs	SEVERE IARs	SERIOUS ADVERSE REACTIONS*	DISCONTINUATION
NEXVIAZYME (n=51)	25.5% (n=13)	0% (n=0)	2.0% (n=1)	0% (n=0)
MYOZYME (n=49)	32.7% (n=16)	4.1% (n=2)	6.1% (n=3)	8.2% (n=4)

*Treatment-related.[1]

IARs[1]

Fewer patients (25.5%) experienced IARs with NEXVIAZYME compared with those receiving MYOZYME (32.7%) (*P*-value not assessed).

Discontinuations due to adverse reactions[1]

- No patients withdrew from the NEXVIAZYME arm during the 49-week study period
- 4 patients in the MYOZYME arm withdrew from the COMET trial due to adverse reactions

Abbreviations: IAR, infusion-associated reaction; LOPD, late-onset Pompe disease.

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Infusion-associated reactions (IARs)[1]

- In clinical studies, IARs were reported to occur at any time during and/or within a few hours after the infusion of NEXVIAZYME and were more likely with higher infusion rates.
- IARs were reported in approximately 30.4 % of patients treated with NEXVIAZYME in clinical studies.
- The majority of IARs were assessed as mild to moderate and included symptoms such as chills, cough, diarrhoea, erythema, fatigue, headache, influenza-like illness, nausea, ocular hyperaemia, pain in extremity, pruritus, rash, rash erythematous, tachycardia, urticaria, vomiting, chest discomfort, dizziness, hyperhidrosis, lip swelling, oxygen saturation decreased, pain, palmar erythema, swollen tongue and tremor
- In clinical studies, 3 (2.2%) patients reported severe IARs including symptoms of chest discomfort, nausea and increased blood pressure.
- Patients with an acute underlying illness at the time of NEXVIAZYME infusion appear to be at greater risk for IARs.
- Patients with advanced Pompe disease may have compromised cardiac and respiratory function, which may predispose them to a higher risk of severe complications from IARs.
- Antihistamines, antipyretics, and/or corticosteroids can be given to prevent or reduce IARs. However, IARs may still occur in patients after receiving pre-treatment.

Abbreviations: IAR, infusion-associated reaction.

1. NEXVIAZYME Australian Approved Product Information.

NEXVIAZYME has been evaluated for safety across the disease spectrum in children and adults[1]

Special populations

Paediatric and elderly populations

- The safety and efficacy of NEXVIAZYME were assessed in patients older than one year of age, including patients over the age of 75 years (n=3). NEXVIAZYME was studied in paediatric patients with IOPD (n=19; 1 to 12 years of age) and 1 paediatric patient with LOPD (16 years of age). There is no recommended dose adjustment for patients over the age of 65.[1]

Patients with renal or hepatic impairment

- Safety and efficacy have not been determined in patients with hepatic impairment or patients with moderate or severe renal impairment. No dose adjustment is required in patients with mild renal impairment.[1]

Pharmacokinetic properties

- Population pharmacokinetic analyses in patients with LOPD showed that age and gender did not meaningfully influence the pharmacokinetics of NEXVIAZYME.[1]

Special warnings and precautions for use

Hypersensitivity reactions including anaphylaxis

- Hypersensitivity reactions, including anaphylaxis, have been reported in patients treated with NEXVIAZYME.[1]
  - In clinical studies, 60 (43.5%) patients experienced hypersensitivity reactions, including 6 patients who reported severe hypersensitivity reactions and 2 patients who experienced anaphylaxis.[1]
  - If mild or moderate hypersensitivity reactions occur, infusion rate may be slowed or temporarily ceased. If severe hypersensitivity or anaphylaxis occur, discontinue immediately and appropriate medical treatment should be initiated.[1]

Abbreviations: IOPD, infantile-onset Pompe disease; LOPD, late-onset Pompe disease.

1. NEXVIAZYME Australian Approved Product Information.

NEXVIAZYME dosing and administration

Nexviazyme®
(avalglucosidase alfa)

Simple to start or switch[1]

NEXVIAZYME dosing is weight-based. Ensure recorded weight is up to date for accurate dosage

For patients ≥1 year

	Patient weight (kg)		Dose (mg/kg)		Patient dose (mg)	Patient dose (mg) 100 (mg/vial)	Vials to reconstitute
Example 1	16	x	20	=	320	3.2 vials	4
Example 2	16	x	40	=	640	6.4 vials	7

Abbreviations: IOPD, infantile-onset Pompe disease; LOPD, late-onset Pompe disease.

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Infusion Volumes

Projected intravenous infusion volumes for Nexviazyme administration by patient weight at 20 and 40 mg/kg doses

PATIENT WEIGHT RANGE (kg)	TOTAL INFUSION VOLUME (mL) FOR 20 MG/KG DOSE	TOTAL INFUSION VOLUME (mL) FOR 40 MG/KG DOSE
5.1 to 10	50	100
10.1 to 20	100	200
20.1 to 30	150	300
30.1 to 35	200	400
35.1 to 50	250	500
50.1 to 60	300	600
60.1 to 100	500	1000
100.1 to 120	600	1200
120.1 to 140	700	1400
140.1 to 160	800	1600
160.1 to 180	900	1800
180.1 to 200	1000	2000

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Supplies and equipment[1]

Supplies and equipment needed for infusion

- **NEXVIAZYME single-use vials**
- **Intravenous (IV) administration set** with 0.2 µm, low protein binding (in-line) filter
- **Sterile water for injection** for reconstitution – 10 mL for each vial
- **5% dextrose in water (D5W)** for dilution
- **Syringes and needles** for reconstitution and dilution (diameter not larger than 20-gauge-calibre)
- Additional supplies per institution protocol

NOTE: Filtered needles should NOT be used during preparation of NEXVIAZYME Infusion.

1. NEXVIAZYME Australian Approved Product Information.

Reconstitution and dilution in 10 steps[1]

Reconstitution

Note: Use aseptic technique during preparation.

1. Remove the required number of vials needed for the infusion from the refrigerator and set aside for approximately 30 minutes to allow them to reach room temperature.

2. Reconstitute each vial by slowly injecting 10 mL of sterile water for injection (SWFI) into each vial.
   - Inject SWFI by a slow drop-wise addition of the diluent down the inside of the vial and not directly onto the lyophilised powder
   - Avoid forceful impact of the diluent on the lyophilised powder and avoid foaming

3. Tilt and roll each vial gently.
   - Do not invert, swirl, or shake
   - Allow the solution to become dissolved
   - After reconstitution, each vial will yield 100 mg/10 mL (10 mg/mL) of NEXVIAZYME

1. NEXVIAZYME Australian Approved Product Information.

Reconstitution and dilution in 10 steps[1]

Reconstitution

Note: Use aseptic technique during preparation.

4. Perform an immediate visual inspection of the reconstituted solution in vials for particulate matter and discolouration.
   - Reconstituted solution should be clear, colourless to pale yellow
   - Do not use if solution is discoloured or if opaque particles are observed
5. Dilute the reconstituted solution without delay
   - If immediate use is not possible, the reconstituted solution can be stored up to 24 hours at 2°C to 8°C
   - Do not freeze

Acceptable

Clear, colourless to pale yellow

Not acceptable

Discoloured, opaque particles, or foreign matter

1. NEXVIAZYME Australian Approved Product Information.

Reconstitution and dilution in 10 steps[1]

Dilution

Dilute in 5% dextrose in water immediately after reconstitution to a final concentration of 0.5 to 4 mg/mL NEXVIAZYME.

6. Check the volume for dilution.
   - Remove that same volume from the infusion bag
   - Remove air from inside the infusion bag to reduce the risk of foam or protein particle formation

7. Slowly withdraw the volume of reconstituted solution from each vial (calculated according to patient's weight).

8. The reconstituted solution should be diluted slowly and directly into 5% dextrose in water.
   - Make up the recommended total infusion volume based on the patient's weight
   - Avoid foaming or agitation of the infusion bag, and avoid air introduction into the infusion bag
   - Discard any unused reconstituted solution remaining in the vial

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Reconstitution and dilution in 10 steps[1]

Dilution

9. Mix the contents of the infusion bag by gently inverting or massaging the infusion bag.
   - Do not shake
   - After dilution, the solution will have a final concentration of 0.5 to 4 mg/mL of NEXVIAZYME

10. Administer the diluted solution without delay. The recommended infusion duration is between 4 to 7 hours. Discard any unused diluted solution after 9 hours.

1. NEXVIAZYME Australian Approved Product Information.

How to store diluted solution[1]

Can be stored **up to 24 hours** in a refrigerator (2°C to 8°C) and **up to 9 hours** (including infusion time) when stored at room temperature (up to 25 °C)

Once the diluted solution is removed from the refrigerator, **it cannot be re-stored in the refrigerator**

Completely infuse the diluted solution within **9 hours** after removal from the refrigerator

Discard the diluted solution if refrigerated for **more than 24 hours** or if the diluted solution is not able to be completely infused **within 9 hours** after removal from the refrigerator

Do not freeze

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

Administration steps[1]

1. Explain the administration procedure to the patient.
2. Obtain vital signs prior to and during the infusion.
3. Obtain IV access.
4. Draw any required blood work if applicable; flush line with 5% dextrose in water
5. Initiate primary infusion line of 5% dextrose in water to maintain patency of IV access.
6. Use a programmable intravenous infusion pump to control infusion rate.
7. A 0.2µm low protein-binding inline filter is recommended
8. NEXVIAZYME should not be infused in the same IV line with other products.
9. Set up and prime the administration set with the NEXVIAZYME infusion solution.
10. Use care to prevent the appearance of air bubbles in tubing
11. Connect administration set to the 0.2µm in-line low protein binding filter set and prime
12. Connect to lowest additive port on primary administration set
13. Infuse NEXZVIAZYME per the infusion schedule
14. Flush the line with 5% dextrose in water at the last infusion rate
15. After infusion, remove the administration set, any unused product or waste material and dispose in accordance with local requirements

1. NEXVIAZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

How to infuse incrementally[1]

LOPD

Initial and subsequent infusions

The recommended starting infusion rate is 1 mg/kg/hour. If there are no signs of IARs, gradually increase the infusion rate every 30 minutes in each of the following three steps:

3 mg/kg/hour → **5 mg/kg/hour** → **7 mg/kg/hour**

Then maintain the infusion rate at 7 mg/kg/hour until the infusion is complete. The approximate total infusion duration is 4 to 5 hours.

Patients switching to NEXVIAZYME will likely not need to change their infusion process or center.[1]

IOPD

4–step process

The recommended starting infusion rate is 1 mg/kg/hour. If there are no signs of IARs, gradually increase the infusion rate every 30 minutes in each of the following three steps:

3 mg/kg/hour → **5 mg/kg/hour** → **7 mg/kg/hour**

Then maintain the infusion rate at 7 mg/kg/hour until the infusion is complete (4-step process). The approximate total infusion duration is 7 hours.

5–step process

The recommended starting infusion rate is 1 mg/kg/hour, with gradual increase in infusion rate every 30 minutes if there are no signs of IARs. The process may use either the above 4-step process or the following 5-step process:

3 mg/kg/hour → **6 mg/kg/hour** → **8 mg/kg/hour** → **10 mg/kg/hour**

Then maintain the infusion rate at 10 mg/kg/hour until the infusion is complete. The approximate total 5-step infusion duration is 5 hours.

Abbreviations: IAR, infusion-associated reaction.

1. NEXVIAZYME Australian Approved Product Information.

Monitoring[1]

- Infusion should be administered incrementally as determined by patient response and comfort.
- Vital signs should be obtained at each step, before increasing the infusion rate.
- If anaphylaxis or severe hypersensitivity reaction or severe IARs occur, immediately discontinue administration of NEXVIAZYME and initiate appropriate medical treatment.
  - In mild to moderate hypersensitivity reactions or IARs occur, the infusion rate may be slowed or temporarily stopped and/or appropriate medical treatment initiated
  - Symptoms may persist despite temporarily stopping the infusion; therefore, the treating physician should wait at least 30 minutes for symptoms of the reactions to resolve before deciding to stop the infusion for the remainder of the day
  - If symptoms subside, resume infusion rate for 30 minutes at half the rate, or less, of the rate at which the reactions occurred, followed by an increase in infusion rate by 50% for 15 to 30 minutes
  - If symptoms do not recur, increase the infusion rate to the rate at which the reactions occurred and consider continuing to increase the rate in a stepwise manner until the maximum rate is achieved

▼ This medicinal product is subject to additional monitoring in Australia.
This will allow quick identification of new safety information.
Healthcare professionals are asked to report any suspected adverse events at www.tga.gov.au/reporting-problems.

Abbreviations: IAR, infusion-associated reaction.

1. NEXVIAZYME Australian Approved Product Information.

Simplified preparation with 100 mg of medication per vial with NEXVIAZYME vs 50 mg per vial for MYOZYME®[1,2]

- Preparation is simplified with NEXVIAZYME as the vials are larger vs. MYOZYME® (ie. they contain 100 mg vs 50 mg) and the reconstituted solution is 10 mg/mL (rather than 5 mg/mL)
- Thus, there may be less set up and handling required for preparation of an infusion bag with NEXVIAZYME[1,2]
- This may cut patient wait times and improve clinic efficiency

1. NEXVIAZYME Australian Approved Product Information. 2. MYOZYME Australian Approved Product Information.

Nexviazyme® (avalglucosidase alfa)

How to store[1]

- Each pack contains 1, 5, 10 or 25 single-use vials, each containing 100 mg of avalglucosidase alfa
- Vials should be stored in a refrigerator between 2°C to 8°C
- Do not use NEXVIAZYME after the expiration date
- The reconstituted and diluted solution should be administered without delay
  - The reconstituted product can be stored up to 24 hours when refrigerated at 2°C to 8°C
  - The diluted product can be stored up to 24 hours when refrigerated at 2°C to 8°C and up to 9 hours (including infusion time) when stored at room temperature (up to 25°C)

1. NEXVIAZYME Australian Approved Product Information

Sanofi Support

Nexviazyme®
(avalglucosidase alfa)

Home infusion

Sanofi has carefully selected Care For Rare Nursing Providers based on a number of criteria to ensure patient privacy is maintained, patient safety is upheld, and the quality use of medicines is applied

Arranging the home infusions

- The Care For Rare Nurse will contact patient's doctor and hospital-based nurses to discuss patient specific infusion information.
- Working with the hospital-based nurse, the Care For Rare Nurse will provide their first infusion with each new patient in the hospital setting. Future infusions will be scheduled in the patient's preferred location (eg home, workplace, etc).
- The Care For Rare Nurse will schedule the next infusion with the patient.
- The Care For Rare Nurse will send a report to the patient's doctor.

Healthcare professional reports

- Specific patient-related data will be recorded and sent to the patient's HCP by email from the Care For Rare Nurse's electronic database after each infusion.
- This information will include:
  - date of infusion
  - patient's general health condition
  - dose/rate of infusion
  - number of vials used
  - duration of administration
  - rate of administration
  - problems/remarks (related to infusion, eg side effects).

NEXVIAZYME® Prescribing Information

PBS Information: This product is not listed on the PBS.
This product is funded under the Life Saving Drugs Program.

Please review full Product Information before prescribing. Full Product Information is available from sanofi-aventis australia pty ltd at bit.ly/nexviazyme-pi or by contacting 1800 818 806.

MINIMUM PRODUCT INFORMATION: Nexviazyme (avalglucosidase alfa) **INDICATIONS:** Nexviazyme is indicated for long-term enzyme replacement therapy for the treatment of patients one year of age and older with Pompe disease (acid α-glucosidase deficiency). **DOSAGE AND ADMINISTRATION:** The recommended dose of Nexviazyme is 20 mg/kg of body weight administered every other week as an intravenous infusion. Dose escalation to 40 mg/kg every other week may be considered for patients with infantile onset Pompe disease (IOPD) who experience insufficient control or declining response at the lower dose. Home administration by a trained health care professional may be considered for individual patients after safety and tolerability has been established in the clinical setting. **CONTRAINDICATIONS:** Life-threatening hypersensitivity to the active substance or to any of the excipients when re-challenge was unsuccessful. **PRECAUTIONS:** Hypersensitivity, infusion associated reactions (IARs), anaphylaxis, immunogenicity, monitor for IgG and IgE antibodies, cardiac hypertrophy, compromised cardiac and respiratory function. Refer to full PI. **INTERACTIONS:** No interaction studies have been performed. **ADVERSE EFFECTS:** Serious adverse reactions include headache, dyspnoea, respiratory distress, nausea, skin discoloration, chills, erythema, chest discomfort, pyrexia, blood pressure increase, body temperature increase, heart rate increase, and oxygen saturation decrease. Common IARs include chills, cough, diarrhoea, erythema, fatigue, headache, influenza-like illness, nausea, ocular hyperaemia, pain in extremity, pruritus, rash, rash erythematous, tachycardia, urticaria, vomiting, chest discomfort, dizziness, hyperhidrosis, lip swelling, pain, palmar erythema, swollen tongue and tremor. Refer to full PI. **NAME OF SPONSOR:** sanofi-aventis australia pty ltd, 12-24 Talavera Road, Macquarie Park, NSW 2113. Based on Full Product Information with TGA date of approval of 22 June 2023. Date of Preparation: 22 June 2023.

▼ This medicinal product is subject to additional monitoring in Australia. This will allow quick identification of new safety information. Healthcare professionals are asked to report any suspected adverse events at www.tga.gov.au/reporting-problems.

MYOZYME® Prescribing Information

PBS Information: This product is not listed on the PBS.
This product is funded under the Life Saving Drugs Program.

Please review full Product Information before prescribing. Full Product Information is available from sanofi-aventis australia pty ltd at bit.ly/myozyme-pi or by contacting 1800 818 806.

Minimum Product Information MYOZYME (Alglucosidase alfa 52.5 mg per 50mg vial) **INDICATION:** Long-term treatment of patients with a confirmed diagnosis of Pompe disease (acid alfa-glucosidase deficiency). **DOSAGE AND ADMINISTRATION:** 20 mg/kg administered once every 2 weeks as an intravenous infusion, initial infusion rate 1mg/ kg/hr, infusion rate may be increased by 2 mg/kg/hr every 30 minutes, after patient tolerance to the infusion rate is established, until the maximum rate of 7 mg/kg/hr - see full PI. Dilute prior to use - see full PI. Does not contain preservatives. Use within 24 hours of infusion preparation. **CONTRAINDICATIONS:** Hypersensitivity to the active or any of the excipients. **PRECAUTIONS:** Hypersensitivity, anaphylaxis, infusion reaction history, underlying cardiac hypertrophy, compromised cardiac and respiratory function, monitor for IgG antibodies, monitor following infusion, pregnancy (category B1), lactation, prior history of anaphylactoid reactions. No clinical experience in patients under 1 month or patients older than 65 years. Renal and hepatic impairment. See full PI. **INTERACTIONS:** No drug interaction data. **ADVERSE EFFECTS:** Hypersensitivity; anaphylactoid › reactions, infusion and post-infusion reactions, flushing, fever, headache, rash, bronchospasm, wheezing, acute cardiorespiratory failure, cardiac arrest, decreased oxygen saturation, hypotension, oedema, nephrotic syndrome - see full PI. **NAME OF SPONSOR:** sanofi-aventis australia pty ltd, 12-24 Talavera Road, Macquarie Park, NSW 2113. Date of Preparation: 06 Aug 2020. Based on Full PI with TGA date of approval of 14 Mar 2008, with most recent amendment on 06 Aug 2020.