POWERED FOR CHANGE FROM THE INSIDE OUT

Nexviazyme®
(avalglucosidase alfa-ngpt)

MAKE THE NEX MOVE FOR YOUR PATIENTS WITH LOPD.
NEXVIAZYME is a monotherapy* for your patients with LOPD who are newly diagnosed or on another ERT.[1]

INDICATION

NEXVIAZYME (avalglucosidase alfa-ngpt) is indicated for the treatment of patients 1 year of age and older with late-onset Pompe disease [lysosomal acid alpha-glucosidase (GAA) deficiency].

IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

See full prescribing information for complete boxed warning.

Hypersensitivity Reactions Including Anaphylaxis

- **Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available. If a severe hypersensitivity reaction occurs, NEXVIAZYME should be discontinued immediately and appropriate medical treatment should be initiated.**

Infusion-Associated Reactions (IARs)

- **If severe IARs occur, consider immediate discontinuation and initiation of appropriate medical treatment.**

Risk of Acute Cardiorespiratory Failure in Susceptible Patients

- **Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function, may be at risk of serious exacerbation of their cardiac or respiratory status during NEXVIAZYME infusion.**

*Not including premedication or pretreatment. **ERT**=enzyme replacement therapy; **LOPD**=late-onset Pompe disease.

Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

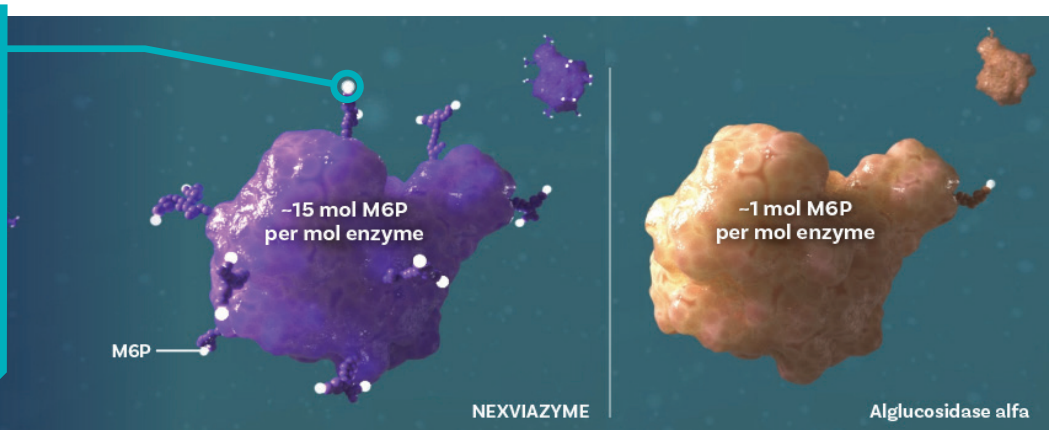
THE NEX STEP IN ERT TECHNOLOGY

NEXVIAZYME: AN M6P-ENRICHED ERT[1]

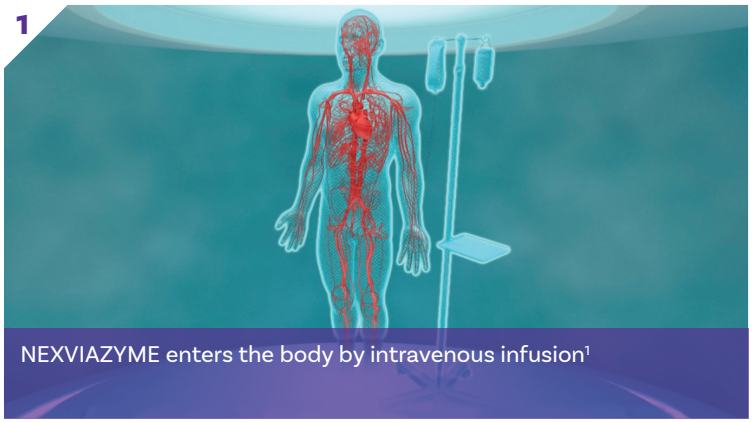
NEXVIAZYME was engineered with 15X more M6P than alglucosidase alfa (Lumizyme).[1,2]

WHAT IS M6P?

M6P is a residue that binds to muscle cell receptors (called CI-MPRs), mediating the uptake of ERT into muscle cells.[3-5]

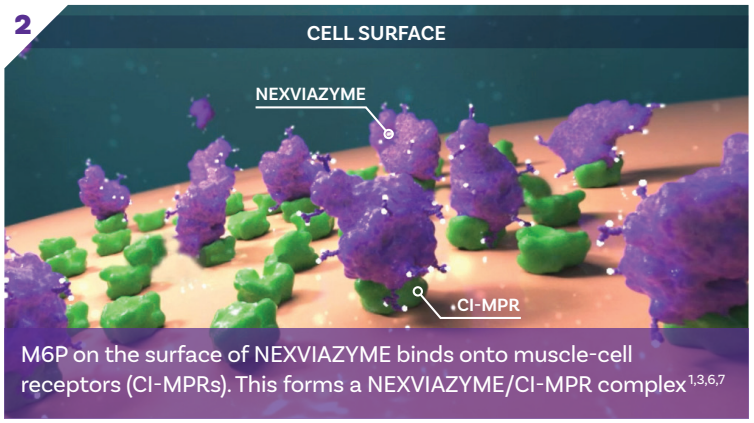


1



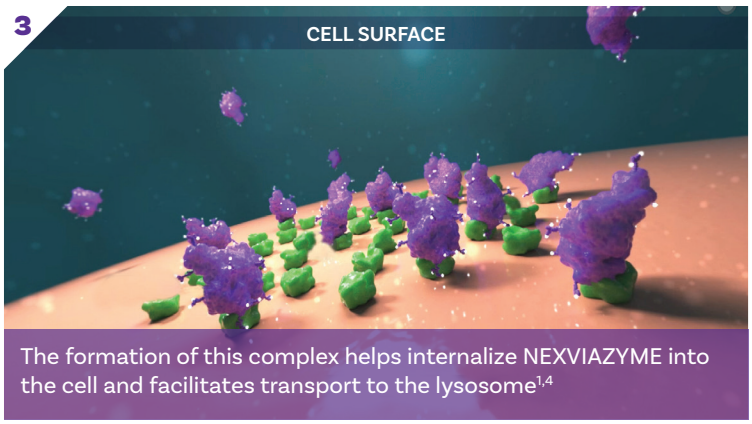
NEXVIAZYME enters the body by intravenous infusion[1]

2



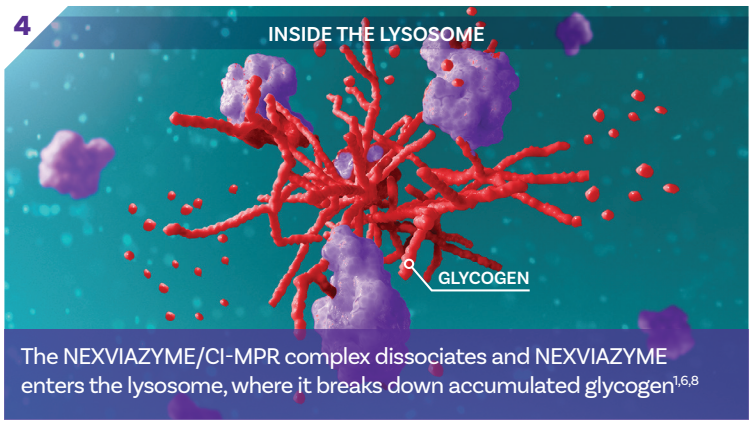
M6P on the surface of NEXVIAZYME binds onto muscle-cell receptors (CI-MPRs). This forms a NEXVIAZYME/CI-MPR complex[1,3,6,7]

3



The formation of this complex helps internalize NEXVIAZYME into the cell and facilitates transport to the lysosome[1,4]

4



The NEXVIAZYME/CI-MPR complex dissociates and NEXVIAZYME enters the lysosome, where it breaks down accumulated glycogen[1,6,8]

CI-MPR=cation-independent mannose-6-phosphate receptor; **M6P**=mannose-6-phosphate.

IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Boxed WARNING: Hypersensitivity Reactions Including Anaphylaxis

Patients treated with NEXVIAZYME have experienced life-threatening hypersensitivity reactions, 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.

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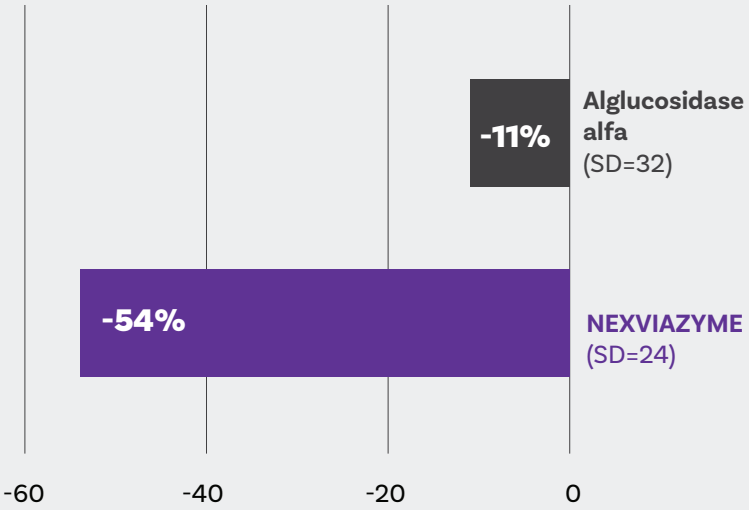
Nexviazyme® (avalglucosidase alfa-ngpt)

NEXVIAZYME PHARMACODYNAMICS: ROLE IN GLYCOGEN DEGRADATION

NEXVIAZYME demonstrated a reduction in urinary glucose tetrasaccharide (Glc4) levels during a phase 3 clinical trial.[1]

CHANGE IN URINARY Glc4 OVER 49 WEEKS



Mean percentage change in urinary Glc4 concentrations from baseline to week 49

The baseline mean urinary/Glc4 concentration was 12.7 mmol/mol Cr and 8.7 mmol/mol Cr in NEXVIAZYME and alglucosidase alfa treatment groups, respectively.[1]

54%

reduction in urinary Glc4 concentration over 49 weeks in a phase 3 clinical trial in treatment-naive patients with LOPD[1]

Urinary Glc4 concentrations were normalized by urine creatinine and reported as mmol Glc4/mol creatinine.[1]

Cr=creatinine; **SD**=standard deviation.

WHAT IS Glc4?

In patients with late-onset Pompe disease, excess of glycogen is degraded to hexose tetrasaccharide (Hex4), which is then excreted in urine. The urinary Hex4 assay measures its major component, Glc4.[1]

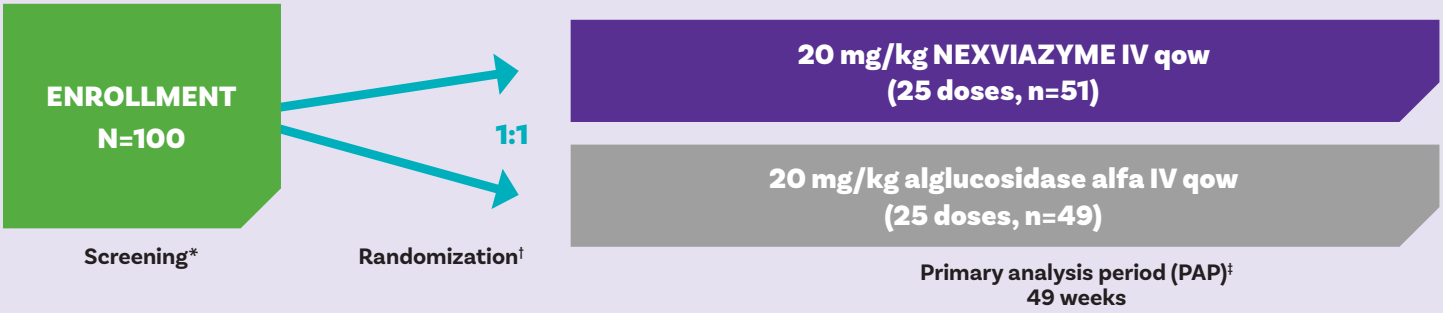
IMPORTANT SAFETY INFORMATION (continued)

Boxed WARNING: Infusion-Associated Reactions (IARs)

Patients treated with NEXVIAZYME have experienced severe IARs. If severe IARs occur, consider immediate discontinuation of NEXVIAZYME, initiation of appropriate medical treatment, and the benefits and risks of readministering NEXVIAZYME following severe IARs. Patients with an acute underlying illness at the time of NEXVIAZYME infusion may 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.

COMET TRIAL: STUDYING THE NEX MOVE IN PATIENTS WITH LOPD

THE FIRST PIVOTAL, HEAD-TO-HEAD, ERT CLINICAL TRIAL IN LOPD[1]



NEXVIAZYME TRIAL STUDY DESIGN[1,9]	
Study design	• Pivotal, phase 3, multicenter, multinational, randomized, double-blinded trial • 100 patients, naive to alglucosidase alfa or any investigational Pompe therapy
Select baseline characteristics	• Upright FVC levels ≥32% and ≤85% predicted • 6MWT distance between ≥118 m and ≤630 m • Median age was 49 years (range from 16 to 78)
Primary endpoint	• Change in FVC (% predicted) in the upright position from baseline to week 49
Key secondary endpoint	• Change in total distance walked in 6 minutes (6MWT) from baseline to week 49

6MWT=6-Minute Walk Test; **FVC**=forced vital capacity; **qow**=every other week.
*Screening phase (up to 14 days), may be extended to up to 8 weeks in prespecified situations.
†Randomization at a 1:1 ratio with stratification factors based on baseline FVC, sex, age, and country (Japan or ex-Japan).
‡NEXVIAZYME infusion, safety assessments, and efficacy evaluations.

IMPORTANT SAFETY INFORMATION (continued)

Boxed WARNING: Risk of Acute Cardiorespiratory Failure in Susceptible Patients

Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during NEXVIAZYME infusion. More frequent monitoring of vitals should be performed during NEXVIAZYME infusion.

Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

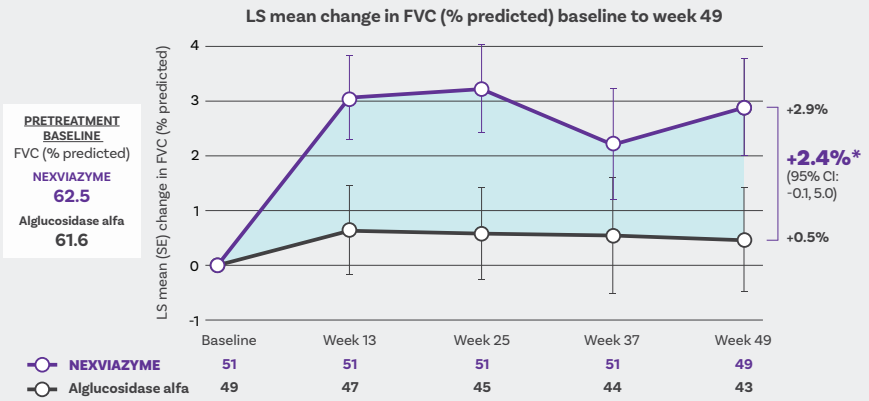
Nexviazyme® (avalglucosidase alfa-ngpt)

COMET TRIAL: STUDYING THE NEX MOVE IN PATIENTS WITH LOPD

PATIENTS RECEIVING NEXVIAZYME DEMONSTRATED A MEANINGFUL IMPROVEMENT IN BREATHING AND WALKING COMPARED WITH ALGLUCOSIDASE ALFA[1]

NEXVIAZYME helped patients improve their ability to walk and breathe compared with baseline.[1]

PRIMARY ENDPOINT



At 49 weeks, NEXVIAZYME improved FVC (% predicted)[1]

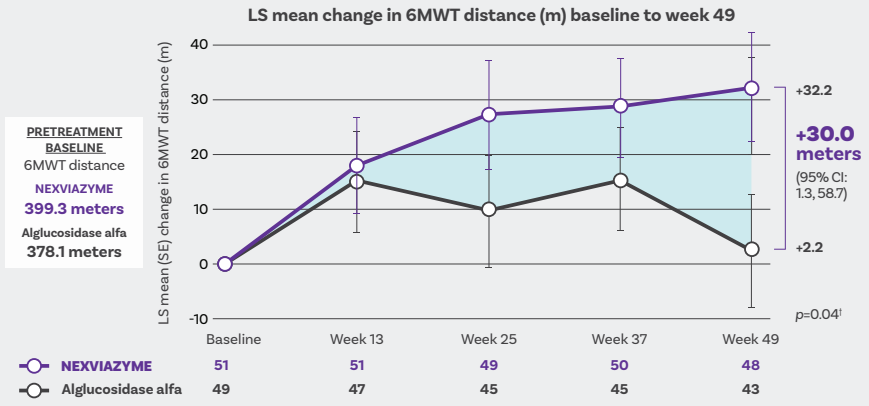
+2.9 pp compared with baseline

+2.4 pp compared with alglucosidase alfa

*Noninferiority margin of 1.1% (p=0.0074). Statistical superiority of NEXVIAZYME over alglucosidase alfa was not achieved (p=0.06). Statistical threshold for significance was p=0.05.

LS=least squares; **pp**=percentage points.

KEY SECONDARY ENDPOINT



At 49 weeks, NEXVIAZYME improved 6MWT[1]

+32.2 meters compared with baseline

+30.0 meters compared with alglucosidase alfa

†P value at nominal level, without multiplicity adjustment.

IMPORTANT SAFETY INFORMATION (continued)

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: See Boxed WARNING. Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. The risks and benefits of readministering NEXVIAZYME following severe hypersensitivity reaction (including anaphylaxis) should be considered. If a mild or moderate hypersensitivity reaction occurs, the infusion rate may be slowed or temporarily stopped.

COMET TRIAL: STUDYING THE NEX MOVE IN PATIENTS WITH LOPD

THE SAFETY PROFILE OF NEXVIAZYME WAS WELL ESTABLISHED

No patients reported a severe IAR and 1 (2%) patient reported a serious AR over 49 weeks in the COMET study.[1]

Adverse reactions reported in ≥6% of patients treated with NEXVIAZYME[1]

Adverse Reaction	NEXVIAZYME (N=51) n (%)	ALGLUCOSIDASE ALFA (N=49) n (%)
Headache	11 (22%)	16 (33%)
Fatigue	9 (18%)	7 (14%)
Diarrhea	6 (12%)	8 (16%)
Nausea	6 (12%)	7 (14%)
Arthralgia	5 (10%)	8 (16%)
Dizziness	5 (10%)	4 (8%)
Myalgia	5 (10%)	7 (14%)
Pruritus	4 (8%)	4 (8%)
Vomiting	4 (8%)	3 (6%)
Dyspnea	3 (6%)	4 (8%)
Erythema	3 (6%)	3 (6%)
Paresthesia	3 (6%)	2 (4%)
Urticaria	3 (6%)	1 (2%)

ARs=adverse reactions; **IARs**=infusion-associated reactions.

NEXVIAZYME IARs AND ARs IN THE COMET STUDY

IARs[1]

25% (13/51) of patients receiving NEXVIAZYME

33% (16/49) of patients receiving alglucosidase alfa

Mild-to-moderate IARs reported in >1 patient receiving NEXVIAZYME were headache, diarrhea, pruritus, urticaria, and rash.

SEVERE IARs[1]

0 patients receiving NEXVIAZYME

4% (2/51) of patients receiving alglucosidase alfa

SERIOUS ARs[1]

2% (1/51) of patients receiving NEXVIAZYME

6% (3/49) of patients receiving alglucosidase alfa

The COMET trial was not designed to demonstrate a statistically significant difference in the incidence of ARs between NEXVIAZYME and alglucosidase alfa.[1]

IMPORTANT SAFETY INFORMATION (continued)

WARNINGS AND PRECAUTIONS

Infusion-Associated Reactions: See Boxed WARNING. IARs may still occur in patients after receiving pretreatment. If mild or moderate IARs occur regardless of pretreatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms.

Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

COMPREHENSIVE SAFETY ANALYSIS

A LONG-TERM LOOK AT THE NEX MOVE

Treatment discontinuations

4% (5/141) of NEXVIAZYME-treated patients permanently discontinued treatment due to adverse reactions.[1]

2 of the 5 patients discontinued because of serious adverse reactions.[1]

Serious adverse reactions

In 2 or more NEXVIAZYME-treated patients, respiratory distress, chills, and pyrexia were reported.[1]

Serious adverse events were similar across both adult and pediatric populations.[1]

Pooled safety analysis was gathered for 141 patients with Pompe disease treated with NEXVIAZYME (118 adult and 23 pediatric), including patients who switched from alglucosidase alfa.[1,9]

Infusion-associated reactions

34% (48/141) of NEXVIAZYME-treated patients reported IARs.[1]

IARs reported in >1 patient included chills, cough, diarrhea, erythema, fatigue, headache, influenza-like illness, nausea, ocular hyperemia, pain in extremity, pruritus, rash, rash erythematous, tachycardia, urticaria, vomiting, chest discomfort, dizziness, hyperhidrosis, lip swelling, oxygen saturation decreased, pain, palmar erythema, swollen tongue, abdominal pain upper, burning sensation, eyelid edema, feeling cold, flushing, respiratory distress, throat irritation, and tremor.[1]

COMPREHENSIVE SAFETY ANALYSIS

IMMUNOGENICITY AND ANTIDRUG ANTIBODIES (ADA)

ADA activity was monitored in NEXVIAZYME-treated patients for up to 7 years.[1]

Incidence of anti-NEXVIAZYME antibodies in patients with Pompe disease

Most adults and some children developed ADA following treatment with NEXVIAZYME. Most ADA were not neutralizing antibodies (NAbs).[1]

	NEXVIAZYME			
	Treatment-Naive Patients: ADA* (N=61)†	Treatment-Experienced Patients: ADA (N=74)‡		
	Adults/ Pediatrics 20 mg/kg every 2 weeks (N=61)†	Adults 20 mg/kg every 2 weeks (N=58)	Pediatrics 20 mg/kg every 2 weeks (N=6)	Pediatrics 40 mg/kg every 2 weeks (N=10)
	n (%)	n (%)	n (%)	n (%)
ADA at baseline	2 (3%)	43 (74%)	1 (17%)	1 (10%)
ADA after treatment	58 (95%)	32 (55%)	1 (17%)	5 (50%)
Neutralizing Antibody (NAb)				
Both NAb types	13 (21%)	3 (5%)	0	0
Inhibition of enzyme activity	17 (28%)	10 (18%)	0	0
Inhibition of enzyme cellular uptake	24 (39%)	12 (21%)	0	1 (10%)

*Includes one pediatric patient.

†Treatment naive: treated only with NEXVIAZYME.

‡Treatment experienced: previously treated with alglucosidase alfa. Treatment-experienced patients received alglucosidase alfa treatment within a range of 0.9-9.9 years for adult patients and 0.5-11.7 years for pediatric patients before receiving NEXVIAZYME.

IN NEXVIAZYME-TREATED PATIENTS (MEAN 26 MONTHS, UP TO 7 YEARS OF TREATMENT)

Incidence of IARs[1]:
62% (8/13) in those with ADA peak titer ≥12,800
19% (8/43) in those with ADA titer <12,800
33% (1/3) in those who were ADA negative

Incidence of hypersensitivity reactions[1]:
31% (4/13) in those with higher ADA titers
14% (2/14) in those with lower ADA titers

In ERT-experienced adult patients who switched to NEXVIAZYME[1]:
Increased incidence of IARs and hypersensitivity reactions in those patients who developed ADA versus those ADA negative.

One treatment-naive patient (ADA peak titer 3200) and 2 treatment-experienced patients (ADA peak titers; 800 and 12,800, respectively) developed anaphylaxis.

Seroconversion[1]:
The median time to seroconversion was 8 weeks.

The development of ADA did not have an apparent impact on clinical efficacy or pharmacokinetics.

A trend toward decreased pharmacodynamic response was observed in patients with ADA peak titer ≥12,800.

Antibody detection is highly dependent on assay performance and can be influenced by several factors. For these reasons, comparing antibody incidence rates between studies may be misleading.

ADA=antidrug antibodies.

NEXVIAZYME: A MONOTHERAPY*

THE NEX STEP FOR PATIENTS WITH LOPD 1 YEAR OF AGE AND OLDER[1]

NEXVIAZYME is engineered with 15x more M6P than alglucosidase alfa[1,2]

- NEXVIAZYME
  ~15 mol M6P per mol enzyme
- Alglucosidase alfa
  1 mol M6P per mol enzyme

In a 49-week, head-to-head clinical trial of treatment-naive patients with LOPD, NEXVIAZYME demonstrated:

54% ▼ **reduction in urinary Glc4 concentrations from baseline**

Glc4 is a major component of Hex4.[1] The clinical relevance of urinary Glc4 concentrations has not been determined.

Improved respiratory function[1]

+2.9

percentage point improvement in FVC (% predicted) compared with baseline

Improved walking distance[1]

+32.2

meter improvement in 6MWT compared with baseline

Established safety profile[1]

1 (2%)

patient receiving NEXVIAZYME reported a serious adverse reaction

3 (6%)

patients receiving alglucosidase alfa reported serious adverse reactions

*Not including premedication or pretreatment.

Glc4=glucose tetrasaccharide; **M6P**=mannose-6-phosphate.

IMPORTANT SAFETY INFORMATION (continued)

WARNINGS AND PRECAUTIONS

Risk of Acute Cardiorespiratory Failure in Susceptible Patients: See Boxed WARNING.

ADVERSE REACTIONS

The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia and urticaria.

GETTING STARTED

SELECT NEXVIAZYME for your patients with LOPD. NEXVIAZYME is suitable for ERT-naive or ERT-experienced patients 1 year of age and older.[1]

PRESCRIBE the number of vials based on individual patient weight and dosage.[1]

Calculating dose and vials example:

Total patient weight (kg) **x** Dosage selection (weight based) 20 mg/kg or 40 mg/kg = **Total patient dose** (mg)

$$\frac{\text{Total patient dose (mg)}}{\text{Vial concentration (100 mg/vial)}} = \textbf{Total vial count}$$ (round up to the nearest whole vial)

Example:

42 kg **x** 20 mg/kg = **840 mg**

$$\frac{840\text{ mg}}{100\text{ mg/vial}} = \textbf{8.4 vials}$$

9 vials total of NEXVIAZYME are needed

CHOOSE an infusion location that works best for each patient. Patients already receiving ERT can keep their same infusion center.

CHECK to see if your patient is eligible for the CareConnectPSS® QuickStart Program.

IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Boxed WARNING: Hypersensitivity Reactions Including Anaphylaxis

Patients treated with NEXVIAZYME have experienced life-threatening hypersensitivity reactions, 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.

Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

NEXVIAZYME: GIVEN AS A MONOTHERAPY*

The recommended dosage of NEXVIAZYME for patients weighing ≥30 kg is 20 mg/kg (of actual body weight) every 2 weeks. For patients weighing <30 kg, the recommended dosage is 40 mg/kg (of actual body weight) every 2 weeks.[1] The initial recommended infusion rate is 1 mg/kg/hour. Gradually increase the infusion rate every 30 minutes if there are no signs of infusion-associated reactions (IARs).[1]

- Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids
- NEXVIAZYME must be reconstituted and diluted prior to use

Projected intravenous infusion volume for NEXVIAZYME administration according to patient's weight[1]

4-STEP PROCESS

PATIENT WEIGHT RANGE (kg)	TOTAL INFUSION VOLUME (mL)	RECOMMENDED DOSE (mg/kg)	Step 1 1 mg/kg/hour	Step 2 3 mg/kg/hour	Step 3 5 mg/kg/hour	Step 4 7 mg/kg/hour	APPROXIMATE TOTAL INFUSION DURATION
			INFUSION RATE (mL/hour)				
5 to 9.9	100	40	3	8	13	18	7 hours
10 to 19.9	200		5	15	25	35	
20 to 29.9	300		8	23	38	53	
30 to 34.9	200	20	10	30	50	70	4-5 hours
35 to 49.9	250		13	38	63	88	
50 to 59.9	300		15	45	75	105	
60 to 99.9	500		25	75	125	175	
100 to 119.9	600		30	90	150	210	
120 to 140	700		35	105	175	245	

5-STEP PROCESS*

PATIENT WEIGHT RANGE (kg)	TOTAL INFUSION VOLUME (mL)	RECOMMENDED DOSE (mg/kg)	Step 1 1 mg/kg/hour	Step 2 3 mg/kg/hour	Step 3 6 mg/kg/hour	Step 4 8 mg/kg/hour	Step 5 10 mg/kg/hour	APPROXIMATE TOTAL INFUSION DURATION
			INFUSION RATE (mL/hour)					
5 to 9.9	100	40	3	8	15	20	25	5 hours
10 to 19.9	200		5	15	30	40	50	
20 to 29.9	300		8	23	45	60	75	

40 mg/kg
20 mg/kg

Step 1 indicates the starting infusion rate. If there are no signs of IARs, gradually increase the infusion rate every 30 minutes to the subsequent step.

*The 5-step process should be used only for subsequent infusions.

Dose modifications due to hypersensitivity reactions and/or IARs

Dose modifications with NEXVIAZYME may be necessary due to severe hypersensitivity reactions (including anaphylaxis) or a severe IAR. Please consult the full Prescribing Information for instructions on appropriate dose modifications before beginning administration.[1]

Switching to NEXVIAZYME can be a seamless process

For patients who are switching to NEXVIAZYME, the label does not require a washout period between the final alglucosidase alfa dose and the first NEXVIAZYME dose.[1,9] Patients may also be able to keep their same dosing schedule and their same infusion center.[1]

See full Prescribing Information for administration instructions, including the recommended infusion rate schedule.

*Not including premedication or pretreatment.

DOSING AND ADMINISTRATION FOR NEXVIAZYME

Administer the infusion incrementally, as determined by the patient's response and comfort.

WHEN THE RECOMMENDED DOSE IS 20 MG/KG

Initial and subsequent infusions[1]

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 4 steps:

1 mg/kg/hour → **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.

WHEN THE RECOMMENDED DOSE IS 40 MG/KG

Initial infusion[1]

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 4 steps:

1 mg/kg/hour → **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.

Subsequent infusions[1]

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:

1 mg/kg/hour → **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.

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

IMPORTANT SAFETY INFORMATION (continued)

Boxed WARNING: Infusion-Associated Reactions (IARs)

Patients treated with NEXVIAZYME have experienced severe IARs. If severe IARs occur, consider immediate discontinuation of NEXVIAZYME, initiation of appropriate medical treatment, and the benefits and risks of readministering NEXVIAZYME following severe IARs. Patients with an acute underlying illness at the time of NEXVIAZYME infusion may 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.

Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

INDICATION AND IMPORTANT SAFETY INFORMATION

INDICATION

NEXVIAZYME (avalglucosidase alfa-ngpt) is indicated for the treatment of patients 1 year of age and older with late-onset Pompe disease [lysosomal acid alpha-glucosidase (GAA) deficiency].

IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Hypersensitivity Reactions Including Anaphylaxis

Patients treated with NEXVIAZYME have experienced life-threatening hypersensitivity reactions, 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)

Patients treated with NEXVIAZYME have experienced severe IARs. If severe IARs occur, consider immediate discontinuation of NEXVIAZYME, initiation of appropriate medical treatment, and the benefits and risks of readministering NEXVIAZYME following severe IARs. Patients with an acute underlying illness at the time of NEXVIAZYME infusion may 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.

Risk of Acute Cardiorespiratory Failure in Susceptible Patients

Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during NEXVIAZYME infusion. More frequent monitoring of vitals should be performed during NEXVIAZYME infusion.

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: See Boxed WARNING. Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. The risks and benefits of readministering NEXVIAZYME following severe hypersensitivity reaction (including anaphylaxis) should be considered. If a mild or moderate hypersensitivity reaction occurs, the infusion rate may be slowed or temporarily stopped.

Infusion-Associated Reactions: See Boxed WARNING. IARs may still occur in patients after receiving pretreatment. If mild or moderate IARs occur regardless of pretreatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms.

Risk of Acute Cardiorespiratory Failure in Susceptible Patients: See Boxed WARNING.

ADVERSE REACTIONS

The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia and urticaria.

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Please see **Important Safety Information** on page 15 and full Prescribing Information for complete details, including **Boxed WARNING.**

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