

NEXVIAZYME™ INFUSION ORDER

(avalglucosidase alfa)

1 PATIENT INFORMATION Patient Name: DOB:		PRESCRIBER INFORMATION Prescriber's Name:				
,	Gend			Phone:		
Allergy: Lbs Kg Date Weighed:						
Venous Access Peripheral IV Port G Inch needle			Fax:			
Device: PIV for port access complications			Diagnosis:	271.0 Glucogenosis / E74.02 Pompe disease		
3 CATHETE	R ACCESS ANI	O FLUSH PROT	TOCOL			
ACCESS TYPE		CATHETER FLUSH ORDERS				
Peripheral	☐ 0.9% Saline Flush: Dispense : 30 Days		efills: x 1yr OR Refills	CathFlo: 2 mg/2 mL as directed.		
PORT (Also include	•	mL for patency/SASH protoco		Pharmacy authorized to dispense upon home health nurse validated port occlusion.		
Peripheral IV for Port Malfunction)	I IV for Port Heparin Flush: Dispense: 30 Day		efills: x 1yr OR Refills	☐ Dispense: 1 Kit Refills: x 1yr OR Refills		
			p p			
4 PRE-MED	ICATION Premed	ication for administration	30-60 min. prior to drug infus	sion:		
			-	oort QTY: 1 Refill: x 1yr ORRefills		
5 TREATMENT REGIMEN						
IKEAIIVIE	NT REGIMEN					
DOSE	NT REGIMEN		PRESCRIPTION			
	2 30 kg) Nexviazyme	mg IV every_ ext whole vial (100 mg/10 mL)	PRESCRIPTIONweeks	vials/infusion Dispense 28 days Refill: 12 Months		
DOSE 20 mg/kg (Patient Wt. ≥	: 30 kg) Nexviazyme < 30 kg) Round up to the ne	ext whole vial (100 mg/10 mL)				
DOSE 20 mg/kg (Patient Wt. ≥ 40 mg/kg (Patient Wt. < 40 mg/kg (Patient Wt. < • Add final re • Infuse via i	: 30 kg) Nexviazyme < 30 kg) Round up to the ne INFUSION RA constituted drug volume to a nfusion pump according to ra	ext whole vial (100 mg/10 mL) TE FOR 1 ST AND SUBSEQUE ppropriate volume of Dextro tes in the table below throu	weeksweeks NT INFUSIONS (20 MG/KG OR Use 5% to reach Total Infusion Vo gh 0.2µ or 0.22µ low protein b	t 40 MG/KG) olume in table below		
DOSE 20 mg/kg (Patient Wt. ≥ 40 mg/kg (Patient Wt. < 40 mg/kg (Patient Wt. < • Add final re • Infuse via i	: 30 kg) Nexviazyme < 30 kg) Round up to the ne INFUSION RA constituted drug volume to a nfusion pump according to ra	ext whole vial (100 mg/10 mL) TE FOR 1 ST AND SUBSEQUE ppropriate volume of Dextro tes in the table below throu). If there are no signs of Info	weeksweeks NT INFUSIONS (20 MG/KG OR Use 5% to reach Total Infusion Vo gh 0.2µ or 0.22µ low protein b	d 40 MG/KG) olume in table below binding in-line filter), gradually increase the infusion rate every 30 minutes		
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DOSE 20 mg/kg (Patient Wt. ≥ 40 mg/kg (Patient Wt. < • Add final re • Infuse via i • Start infusio Total Infusion Volume (mL)	Round up to the ne INFUSION RA constituted drug volume to a nfusion pump according to ra on at rate of 1mg/kg/hr (ML/hr) STEP 1: 1 mg/kg/hr (mL/hr)	rext whole vial (100 mg/10 mL) TE FOR 1 ⁵¹ AND SUBSEQUE ppropriate volume of Dextro tes in the table below throu). If there are no signs of Info STEP 2: 3 mg/kg/hr (mL/hr INFUSION RATE FOR SUBS	weeks NT INFUSIONS (20 MG/KG OR USE 5% to reach Total Infusion Vigh 0.2µ or 0.22µ low protein busion Associated Reactions (IARS) STEP 3: 5 mg/kg/hr (mL/hr)	classical design of the state o		
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DOSE 20 mg/kg (Patient Wt. ≥ 40 mg/kg (Patient Wt. < • Add final re • Infuse via i • Start infusio Total Infusion Volume (mL)	Round up to the net of	propriate volume of Dextro tes in the table below throu). If there are no signs of Info STEP 2: 3 mg/kg/hr (mL/hr INFUSION RATE FOR SUBS of 40mg/kg may use either STEP 2: 3 mg/kg/hr (mL/hr	weeks NT INFUSIONS (20 MG/KG OR use 5% to reach Total Infusion V gh 0.2µ or 0.22µ low protein b usion Associated Reactions (IARs) STEP 3: 5 mg/kg/hr (mL/hr) EQUENT INFUSIONS (40MG/K the above 4-step process or the	colume in table below binding in-line filter), gradually increase the infusion rate every 30 minutes STEP 4: 7 mg/kg/hr (mL/hr) GG ONLY) 5-step process below		
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DOSE 20 mg/kg (Patient Wt. ≥ 40 mg/kg (Patient Wt. < • Add final re • Infuse via i • Start infusio Total Infusion Volume (mL) Total Infusion Flush IV wivital Signs: At baseline, e	Nexviazyme	ppropriate volume of Dextro tes in the table below throu). If there are no signs of Info STEP 2: 3 mg/kg/hr (mL/hr INFUSION RATE FOR SUBS of 40mg/kg may use either STEP 2: 3 mg/kg/hr (mL/hr SKILLED NU inical monitoring ate of drug infusion minutes during infusion, a	weeks NT INFUSIONS (20 MG/KG OR USE 5% to reach Total Infusion Vigh 0.2µ or 0.22µ low protein busion Associated Reactions (IARS STEP 3: 5 mg/kg/hr (mL/hr) EQUENT INFUSIONS (40MG/K) the above 4-step process or the STEP 3: 6 mg/kg/hr (mL/hr) JRSING VISIT Administration procedures to business and the statement of the stateme	olume in table below binding in-line filter), gradually increase the infusion rate every 30 minutes STEP 4: 7 mg/kg/hr (mL/hr) 5-step process below STEP 4: 8 mg/kg/hr (mL/hr) STEP 5: 10 mg/kg/hr (mL/hr) De followed per pharmacy protocol th and 30 minutes after completion of post infusion flush supplies.		

Product Substitution Permitted Signature

| Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Date of Signature | Da

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INFUSION REACTION MANAGEMENT ORDERS

Patient Name:			DOB:		
☐ Diphenhydramine 50 mg/mL _ Epinephrine 2-Pak 0.3 mg inje Epinephrine 2-Pak 0.15 mg inj ☐ Corticosteroid: ☐ 0.9% Saline 250 mL - administ	mg IV if Infusion Reaction mg IV if Infusion Reaction ctor IM once for severe brochospasm/anaphyl ector - IM once for severe brochospasm/anaphyl er at 50 mL/hr once Nexviazyme infusion stop mp if needed, IV Pole, back-up peripheral IV k	axis and call 911 hylaxis and call 911			
EXAMPLES OF COMPLEX THERAPY INFUSION REACTIONS					
FeverChills/RigorsHeadache	Rash/ItchingSwelling/EdemaNausea/Vomiting	Abdominal PainIrritabilityHypotension	DyspneaRespiratory Distress		
	INSTRUCTION	S DURING REACTION			
1) STOP NEXVIAZYME [™] , flush	with 10 mL D5W, and start 0.9% normal sa	line at 50 mL/hr.			
•	MEDS FROM BOX 7 ACCORDING TO PHY dminister appropriate Epinephrine Autoinjec		eded. Call 911.		
8 PROVIDER SI		Dispense as Written Signature			
Product Substitution Permitted Signati IMPORTANT NOTICE: This facsimile transmiss other than the named addressee, the recipie retained by anyone other than the named ad	ure Date of Signature ion is intended to be delivered only to the named addressee and may cont nt should immediately notify the sender at the address and telephone nun dressee, except by the express authority of the sender to the named addre		Date of Signature opt from disclosure under applicable law. If it is received by anyone e transmitted material. In no event should such material be read or		

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