Orsini Specialty Pharmacy

NEXVIAZYME™ INFUSION ORDER

(avalglucosidase alfa)

1 PATIENT INFORMATION

Patient Name: _____ DOB: _____
Primary Phone: _____ Gender: ◯ Male ◯ Female
Address: _____
City, State, Zip: _____
Allergy: _____
Patient Weight: _____ Lbs _____ Kg Date Weighed: _____
Venous Access Device: ◯ Peripheral IV ◯ Port ____ G ____ Inch needle
☐ PIV for port access complications

2 PRESCRIBER INFORMATION

Prescriber's Name: _____
Contact Person: _____ Phone: _____
NPI #: _____
Address: _____
Address: _____
City, State, Zip: _____
Phone: _____ Fax: _____
Diagnosis: *271.0 Glucogenosis / E74.02 Pompe disease*

3 CATHETER ACCESS AND FLUSH PROTOCOL

ACCESS TYPE	CATHETER FLUSH ORDERS	
◯ Peripheral ◯ PORT *(Also include Peripheral IV for Port Malfunction)*	☐ **0.9% Saline Flush:** **Dispense:** 30 Days **Refills:** x 1yr OR ____ Refills Flush line/port with 10mL for patency/SASH protocol. ☐ **Heparin Flush:** **Dispense:** 30 Days **Refills:** x 1yr OR ____ Refills Flush port with _____ mL of Heparin _____units/mL per SASH protocol.	**CathFlo:** 2 mg/2 mL as directed. *Pharmacy authorized to dispense upon home health nurse validated port occlusion.* ☐ **Dispense:** 1 Kit **Refills:** x 1yr OR ____ Refills

4 PRE-MEDICATION

Premedication for administration 30-60 min. prior to drug infusion:

☐ Antipyrectic: _____ q.s. 1 month **Refill:** x 1yr OR ____ Refills ☐ Not Needed
☐ Anthistamine: _____ q.s. 1 month **Refill:** x 1yr OR ____ Refills ☐ Not Needed
☐ LMX 4 or (EMLA) Lidocaine 2.5%/Prilocaine 2.5% cream - Apply topically 1 hour prior to starting IV or accessing port **QTY:** 1 **Refill**: x 1yr OR ____ Refills
☐ _____

5 TREATMENT REGIMEN

DOSE	PRESCRIPTION
◯ 20 mg/kg (Patient Wt. ≥ 30 kg) ◯ 40 mg/kg (Patient Wt. < 30 kg)	Nexviazyme _____ mg IV every _____ weeks _____ vials/infusion Dispense 28 days **Refill:** 12 Months *Round up to the next whole vial (100 mg/10 mL)*

INFUSION RATE FOR 1ST AND SUBSEQUENT INFUSIONS (20 MG/KG OR 40 MG/KG)

- Add final reconstituted drug volume to appropriate volume of Dextrose 5% to reach Total Infusion Volume in table below
- Infuse via infusion pump according to rates in the table below through 0.2µ or 0.22µ low protein binding in-line filter
- Start infusion at rate of 1mg/kg/hr (Step 1). If there are no signs of Infusion Associated Reactions (IARs), gradually increase the infusion rate every 30 minutes

Total Infusion Volume (mL)	**STEP 1:** 1 mg/kg/hr (mL/hr)	**STEP 2:** 3 mg/kg/hr (mL/hr)	**STEP 3:** 5 mg/kg/hr (mL/hr)	**STEP 4:** 7 mg/kg/hr (mL/hr)

OPTIONAL INFUSION RATE FOR SUBSEQUENT INFUSIONS (40MG/KG ONLY)

- The infusion process for doses of 40mg/kg may use either the above 4-step process or the 5-step process below

Total Infusion Volume (mL)	**STEP 1:** 1 mg/kg/hr (mL/hr)	**STEP 2:** 3 mg/kg/hr (mL/hr)	**STEP 3:** 6 mg/kg/hr (mL/hr)	**STEP 4:** 8 mg/kg/hr (mL/hr)	**STEP 5:** 10 mg/kg/hr (mL/hr)

SKILLED NURSING VISIT

☐ As needed for IV access, administration, and proper clinical monitoring | Administration procedures to be followed per pharmacy protocol

Post Infusion: Flush IV with 25 mL Dextrose 5% at final rate of drug infusion
Vital Signs: At baseline, each rate change, then every _____ minutes during infusion, at completion of post infusion flush and 30 minutes after completion of post infusion flush

☒ **Supplies:** Provide infusion pump if needed, IV Pole, back-up peripheral IV kit and all necessary infusion supplies.

ALSO SEE PAGE 2 ("NEXVIAZYME INFUSION REACTION MANAGEMENT ORDERS)

6 PROVIDER SIGNATURE ("Nexviazyme™ Infusion Reaction Management Orders")

X _____ Product Substitution Permitted Signature _____ Date of Signature

X _____ Dispense as Written Signature _____ Date of Signature

Orsini Specialty Pharmacy

INFUSION REACTION MANAGEMENT ORDERS

Patient Name: _____ DOB: _____

7 EMERGENCY MEDICATIONS - #q.s. Refill: x 1yr OR___ Refills

- ☐ Diphenhydramine 50 mg/mL _____ mg IV if Infusion Reaction
- ◯ Epinephrine 2-Pak 0.3 mg injector IM once for severe brochospasm/anaphylaxis and call 911
- ◯ Epinephrine 2-Pak 0.15 mg injector - IM once for severe brochospasm/anaphylaxis and call 911
- ☐ Corticosteroid: _____
- ☐ 0.9% Saline 250 mL - administer at 50 mL/hr once Nexviazyme infusion stops:
- ☐ **Supplies:** Provide infusion pump if needed, IV Pole, back-up peripheral IV kit and all necessary infusion supplies.

EXAMPLES OF COMPLEX THERAPY INFUSION REACTIONS

- Fever
- Chills/Rigors
- Headache
- Rash/Itching
- Swelling/Edema
- Nausea/Vomiting
- Abdominal Pain
- Irritability
- Hypotension
- Dyspnea
- Respiratory Distress

INSTRUCTIONS DURING REACTION

1) STOP NEXVIAZYME™, flush with 10 mL D5W, and start 0.9% normal saline at 50 mL/hr.

2) ADMINISTER EMERGENCY MEDS FROM BOX 7 ACCORDING TO PHYSICAN ORDERS

☒ For severe anaphylaxis, administer appropriate Epinephrine Autoinjector IM - may repeat in 20 minutes if needed. Call 911.

3) CALL PHYSICIAN

8 PROVIDER SIGNATURE

X _____
Product Substitution Permitted Signature Date of Signature

X _____
Dispense as Written Signature Date of Signature