



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

 PHYO  
 CMC0028-001NS Rev. 8/2022

**AVALglucosidase Alpha (NEXVIAZYME)  
 Infusion Therapy Plan**
**Baseline Patient Demographic**

To be completed by the ordering provider.

 Diagnosis: \_\_\_\_\_ Height: \_\_\_\_\_ cm Weight: \_\_\_\_\_ kg Body Surface Area: \_\_\_\_\_ (m<sup>2</sup>)
 NKDA - No Known Drug Allergies Allergies: \_\_\_\_\_**Therapy Plan orders extend over time (several visits) including recurring treatment.**

Please specify the following regarding the entire course of therapy:

Duration of treatment: \_\_\_\_\_ weeks \_\_\_\_\_ months \_\_\_\_\_ unknown

Treatment should begin:  as soon as possible (within a week)  within the month**\*\*Plans must be reviewed / re-ordered at least annually. \*\*****ORDERS TO BE COMPLETED FOR EACH THERAPY****ADMIT ORDERS** Height and weight Vital signs**HYPOTENSION DEFINED ADMIT** Nursing communication

Notify Provider for Hypotension:

Prior to starting infusion, please determine the patient's threshold for hypotension as defined by the following parameters. This information will be needed in the event of an infusion reaction.

Hypotension is defined as follows:

1 month to 1 year - systolic blood pressure (SBP) less than 70

1 year to 11 years - systolic blood pressure (SBP) less than 70 + (2 x age in years)

11 years to 17 years - systolic blood pressure (SBP) less than 90

OR any age - systolic blood pressure (SBP) drop of more than 30% from baseline.

Baseline systolic blood pressure (SBP) x 0.7 = value below defined as hypotension.

**NURSING ORDERS**

Please select all appropriate therapy

**IV START NURSING ORDERS** Insert Peripheral IV

Place PIV if needed or access IVAD if available.

 lidocaine 1% BUFFERED (J-TIP LIDOCAINE)

0.2 mL, INTRADERMAL, PRN

 when immediate procedure needed  when procedure will take about 1 minute  patient / family preference for procedureAdministration Instructions: NOTE: Do not use this medication in patients with bleeding disorders, platelets  $\leq$  20,000, or in patients taking anticoagulants, when accessing implanted ports or using a vein that will be utilized for chemotherapy administration, nor for pre-term infants or neonates. lidocaine - prilocaine (EMLA) cream

TOPICAL, PRN

 when more than 60 minutes are available before procedure  when procedure will take more than 1 hour patient / family preference for procedure

Administration Instructions: NOTE: In children &lt; 3 months of age, or &lt; 5 kg in weight, maximum application time is 1 hour.

 lidocaine - tetracaine (SYNERA) patch

TOPICAL, PRN

 when 20 - 30 minutes are available before procedure  when procedure will take more than 1 hour when anticipated pain is less than 5 mm from skin surface  patient / family preference for procedure



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

 PHYO  
 CMC0028-001NS Rev. 8/2022

**Avalglucosidase Alpha (NEXVIAZYME)  
 Infusion Therapy Plan**
**ORDERS TO BE COMPLETED FOR EACH THERAPY**
**NURSING ORDERS, CONTINUED**
 **lidocaine with transparent dressing 4% kit**

TOPICAL, PRN

 when 20 - 30 minutes are available before procedure     when procedure will take more than 1 hour

 patient / family preference for procedure

 **Heparin flush**
**heparin flush**

10 - 50 units, INTRAVENOUS, PRN, IV line flush. Per protocol, heparin should not be used to flush peripheral IVs. This heparin flush should be used with all central lines including IVADs, with the exception of de-accessing the IVAD.

**heparin flush**

100 - 300 units, INTRAVENOUS, PRN, IV line flush. Per protocol, heparin should not be used to flush peripheral IVs. For use only when de-accessing IVADs.

 **Sodium chloride flush**
**Sodium chloride flush 0.9% injection**

1 - 20 mL, INTRAVENOUS, PRN, IV line flush

**Sodium chloride - preservative free 0.9% injection**

1 - 30 mL, INTRAVENOUS, PRN, IV line flush

**PRE-PROCEDURE LABS**
 **Complete Blood Count With Differential**

Unit collect

INTERVAL: Every 12 weeks

DURATION: Until Discontinued

 **Blood Urea Nitrogen**

Unit collect

INTERVAL: Every 12 weeks

DURATION: Until Discontinued

 **Creatinine**

Unit collect

INTERVAL: Every 12 weeks

DURATION: Until Discontinued

 **Miscellaneous Send Out Test**

Urine Hex4 (Urine Glucose Tetrasaccharide Biomarker Assay)

Unit collect

INTERVAL: Every 12 weeks

DURATION: Until Discontinued

 **Miscellaneous Send Out Test**

IgG antibody level (Genzyme Lab)

Unit collect

INTERVAL: Every 12 weeks

DURATION: Until Discontinued

**PRE-MEDICATIONS**
 **Acetaminophen pre-medication 30 minutes prior (15 mg / kg, maximum 650 mg)**
**Nursing communication**

Administer only one of the acetaminophen orders, suspension or tablets, do not give both.

**acetaminophen suspension**

15 mg / kg, ORAL, for 1 dose pre-medication, give 30 minutes prior to infusion

**Dose:** \_\_\_\_\_

**acetaminophen tablet**

15 mg / kg ORAL, for 1 dose pre-medication, give 30 minutes prior to infusion

**Dose:** \_\_\_\_\_



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

 PHYO  
 CMC0028-001NS Rev. 8/2022

**Avalglucosidase Alpha (NEXVIAZYME)  
 Infusion Therapy Plan**
**ORDERS TO BE COMPLETED FOR EACH THERAPY**
**PRE-MEDICATIONS**

- 
- Diphenhydramine pre-medication 30 minutes prior (1 mg / kg, maximum 50 mg)**

**Nursing communication**

Administer only one of the diphenhydrAMINE pre-medication orders, liquid, capsule or injection, do not give more than one of the orders as a pre-medication.

**diphenhydrAMINE liquid**

1 mg / kg, ORAL, for 1 dose pre-medication, give 30 minutes prior to infusion

**Dose:** \_\_\_\_\_

**diphenhydrAMINE capsule**

1 mg / kg ORAL, for 1 dose pre-medication, give 30 minutes prior to infusion

**Dose:** \_\_\_\_\_

**diphenhydrAMINE injection**

1 mg / kg, INTRAVENOUS, 1 dose pre-medication, give 30 minutes prior to infusion

**Dose:** \_\_\_\_\_

**INTRA-PROCEDURE**

- 
- Vital Signs**

Check blood pressure, heart rate, pulse oximetry, respirations, temperature and pain prior to the start to the infusion. If any vital signs are not within ranges for age, wait for 5 minutes and repeat. If vital signs continue to be outside the range, contact the neurology provider. After initiation of the infusion, check heart rate and blood pressure with each increase of the infusion rate. Once the patient is at his / her maximum rate, check vitals every hour until completion of the infusion and observe for signs and symptoms and / or complaints of infusion related reactions. In the event of any hypersensitivity or other infusion related symptoms, the infusion should be stopped and notify the ordering provider.

- 
- Nursing Communication**

Flush IV line with 5 mL D5W PRIOR to the AVALglucosidase infusion.

- 
- dextrose 5 % flush**

5 mL, INTRAVENOUS, ONCE, for 1 dose.

Flush IV line prior to administration of AVALglucosidase.

- 
- Nursing Communication**

\*\*Infusion titration varies based on patient weight\*\*

**\*\*AVALglucosidase infusion rates for patients < 30 kg\*\***

Initial infusion: Start infusion at 1 mg / kg / hr and every 30 minutes may increase by 2 mg / kg / hr until a maximum of 7 mg / kg / hr is reached and continue at 7 mg / kg / hr until infusion is completed (~7 hours). Use the following table for the total volume based off weight and a step by step infusion in mL / hr titration.

Patient weight (kg) = Total volume (mL): rate (mL / hr) = step 1, then step 2, then step 3, then step 4. May gradually increase the infusion rate every 30 minutes if the infusion is tolerated and no signs of infusion-related reactions are observed.

Patient weight (kg) = Total volume (mL)	Rate step by step infusion in mL / hr titration
5 - 9.9 kg = 100 mL	rate = 3, then 8, then 13, then 18
10 - 19.9 mg = 200 mL	rate = 5, then 15, then 25, then 35
20 - 29.9 kg = 300 mL	rate = 8, then 23, then 38, then 53

Subsequent infusions: May administer as IV infusion over ~7 hours following the same titration as the initial infusion OR initiate subsequent infusion as follows. Start subsequent infusion rate at 1 mg / kg / hr, may increase to 3 mg / kg / hr, 6 mg / kg / hr, 8 mg / kg / hr, maximum of 10 mg / kg / hr. Maintain the infusion rate at 10 mg / kg / hr until the infusion is complete (~5 hours). Use the following table for the total volume based off weight and a step by step infusion in mL / hr titration.

Patient weight (kg) = total volume (mL): rate (mL / hr) = step 1, then step 2, then step 3, then step 4, then step 5. May gradually increase the infusion rate every 30 minutes if the infusion is tolerated and no signs of infusion-related reactions are observed.

Patient weight (kg) = Total volume (mL)	Rate step by step infusion in mL / hr titration
5 - 9.9 kg = 100 mL	rate = 3, then 8, then 15, then 20, then 25
10 - 19.9 mg = 200 mL	rate = 5, then 15, then 30, then 40, then 50
20 - 29.9 kg = 300 mL	rate = 8, then 23, then 45, then 60, then 75



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

 PHYO  
 CMC0028-001NS Rev. 8/2022

**Avalglucosidase Alpha (NEXVIAZYME)  
 Infusion Therapy Plan**
**ORDERS TO BE COMPLETED FOR EACH THERAPY**
**INTRA-PROCEDURE, CONTINUED**
**\*\*AVALglucosidase infusion rates for patients  $\geq 30$  kg\*\***

Infusion rate: Start infusion at 1 mg / kg / hr and every 30 minutes may increase by 2 mg / kg / hr until a maximum of 7 mg / kg / hr is reached and continue at 7 mg / kg / hr until infusion is completed (~4 to 5 hours). Use the following table for the total volume based off weight and a step by step infusion in mL / hr titration.

Patient weight (kg) = Total volume (mL): rate (mL / hr) = step 1, then step 2, then step 3, then step 4. May gradually increase the infusion rate every 30 minutes if the infusion is tolerated and no signs of infusion-related reactions are observed.

Patient weight (kg) = Total volume (mL)	Rate step by step infusion in mL / hr titration
30 - 34.9 kg = 200 mL	rate = 10, then 30, then 50, then 70
35 - 49.9 kg = 250 mL	rate = 13, then 38, then 63, then 88
50 - 59.9 kg = 300 mL	rate = 15, then 45, then 75, then 105
60 - 99.9 kg = 500 mL	rate = 25, then 75, then 125, then 175
100 - 119.9 kg = 600 mL	rate = 30, then 90, then 150, then 210
120 - 140 kg = 700 mL	rate = 35, then 105, then 175, then 245

 **Physician Communication Order**

\*\*\*Dose of AVALglucosidase based on weight. Select the appropriate section below. Vital sizes are 100 mg, if clinically acceptable, round to nearest 100 mg. Please enter the dose of AVALglucosidase in 'mg' facilitate prior authorization requirements:

AVALglucosidase < 30 kg: 40 mg / kg, round to nearest 100 mg, if clinically acceptable.

AVALglucosidase  $\geq$  30 kg: 20 mg / kg, round to nearest 100 mg, if clinically acceptable.

 **AVALglucosidase alpha for patients < 30 kg**
**INTERVAL: Every 2 weeks**
**DURATION: Until Discontinue**
 **AVALglucosidase alfa-ngpt in dextrose 5 % infusion**

INTRAVENOUS, ONCE, for 1 dose

Flush the line with 5 mL of D5W flush prior to starting infusion. Administer AVALglucosidase via an in-line, low protein binding, 0.2 micrometer filter. Do not mix or infuse with other medications.

**Dose:** \_\_\_\_\_

 **AVALglucosidase alpha for patients  $\geq 30$  kg**
**INTERVAL: Every 2 weeks**
**DURATION: Until Discontinue**
 **AVALglucosidase alfa-ngpt in dextrose 5 % infusion**

INTRAVENOUS, ONCE, for 1 dose

Flush the line with 5 mL of D5W flush prior to starting infusion. Administer AVALglucosidase via an in-line, low protein binding, 0.2 micrometer filter. Do not mix or infuse with other medications. Initiate infusion at 1 mg / kg / hr. If tolerated and no signs of infusion-related reactions are observed, gradually increase the infusion rate every 30 minutes to 3 mg / kg / hr, 5 mg / kg / hr and 7 mg / kg / hr respectively. Maintain infusion rate at 7 mg / kg / hr until the infusion is complete (~ 4 to 5 hours). After infusion is complete, flush line with D5W.

**Dose:** \_\_\_\_\_

 **Nursing Communication**

Flush PIV or IVAD with 20 mL D5W (50 mL bag) at the completion of the infusion. Flush IVAD with D5W and heparin flush per protocol prior to de-accessing IVAD. Discontinue PIV prior to discharge on the last day of infusion.

 **dextrose 5 % flush 0 - 50 mL**

0 - 50 mL, INTRAVENOUS, ONCE, starting 5 hours after treatment start time, for 1 dose.

Flush IV line after completion of AVALglucosidase.

 **Therapy Appointment Request**

Please select department for the therapy appointment request:

Expires in 365 days

 Dallas Special Procedures  
 Plano Infusion Center  
 Dallas Allergy  
 Dallas Transplant  
 Dallas Neurology

Schedule appointment at most 1 days before or at most 1 days after, internal referral.

Please check Avalglucosidase intervals for appropriate scheduling.



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

PHYO  
CMC0028-001NS Rev. 8/2022

**Avalglucosidase Alpha (NEXVIAZYME)  
Infusion Therapy Plan**

**ORDERS TO BE COMPLETED FOR EACH THERAPY**

**INTRA-PROCEDURE, CONTINUED**

**Vital Signs**

Obtain vital signs at the end of the infusion and 1 hour after the infusion. In the event of any hypersensitivity or other infusion related symptoms, notify the ordering provider. EVERY HOUR, starting when released, until specified, obtain vital signs at the end of the infusion and 1 hour after the infusion. In the event of any hypersensitivity or other infusion related symptoms, notify the ordering provider.

**EMERGENCY MEDICATIONS**

**Nursing communication**

1. Hives or cutaneous reaction only – no other system involvement

**PATIENT IS HAVING A DRUG REACTION:**

- Stop the infusion
- Give diphenhydramine as ordered
- Check heart rate, respiratory rate and blood pressure every 5 minutes until further orders from provider.
- Connect patient to monitor (cardiac / apnea, blood pressure and oxygen saturation) if not already on one
- Notify provider for further orders

2. Hives or cutaneous reaction plus one other system, i.e. abdominal cramping, vomiting, hypotension, altered mental status, respiratory distress, mouth / tongue swelling

**PATIENT IS HAVING ANAPHYLAXIS:**

- Stop the infusion
- Call code – do not wait to give epinephrine
- Give epinephrine as ordered
- Notify provider
- Check heart rate, respiratory rate and blood pressure every 5 minutes until the code team arrives.
- Connect patient to monitor (cardiac / apnea, blood pressure and oxygen saturation), if not already on one.
- Give diphenhydramine once as needed for hives
- May repeat epinephrine every 5 minutes x 2 doses for persistent hypotension and respiratory distress with desaturation until code team arrives.
- May give albuterol as ordered for wheezing with oxygen saturation stable while waiting for code team, continue to monitor oxygen saturation.

**Hypotension is Defined as Follows:**

1 month to 1 year – systolic blood pressure (SBP) less than 70  
 1 year to 11 years – systolic blood pressure (SBP) less than 70 + (2 x age in years)  
 11 years to 17 years – systolic blood pressure (SBP) less than 90  
 OR any age – systolic blood pressure (SBP) drop more than 30% from baseline.  
 Baseline systolic blood pressure (SBP) x 0.7 = value below defined as hypotension.

**EPINEPHrine Injection Orderable For Therapy Plan  
(AMPULE / EPI - PEN JR. / EPI - PEN) 0.01 mg / kg**

0.01 mg / kg, INTRAMUSCULAR, EVERY 5 MINUTES PRN, for anaphylaxis and may be repeated for persistent hypotension and respiratory distress with desaturation until the code team arrives, for 3 doses.

Use caution with PIV administration. This solution has a pH < 5, or a pH > 9, or an osmolality > 600 mOsm / L.

**Dose:** \_\_\_\_\_

**Cardio / Respiratory Monitoring**

**Rationale for Monitoring: High risk patient (please specify risk)**

- Clinically significant cardiac anomalies or dysrhythmias
- Recent acute life-threatening event
- Unexplained or acutely abnormal vital signs
- Artificial airway (stent, tracheostomy, oral airway)
- Acute, fluctuating or consistent oxygen requirements

Monitor Parameters (select all that apply):  Heart rate  Oxygen saturation  Respiratory rate

Telemetry Required:  Yes  No

**diphenhydrAMINE injection**

1 mg / kg, INTRAVENOUS, ONCE PRN, for hives or cutaneous reaction, for 1 dose. Maximum dose = 50 mg per dose, 300 mg per day.

**Dose:** \_\_\_\_\_



Patient Name: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

PHYO  
CMC0028-001NS Rev. 8/2022

**Avalglucosidase Alpha (NEXVIAZYME)  
Infusion Therapy Plan**

**ORDERS TO BE COMPLETED FOR EACH THERAPY**

**EMERGENCY MEDICATIONS, CONTINUED**

**Albuterol for aerosol**

0.25 mg / kg., INHALATION ONCE PRN, for wheezing, but oxygen saturations stable while waiting for code team, continue to monitor oxygen saturation for 1 dose.

**Dose:** \_\_\_\_\_

(circle one):  
MD DO

\_\_\_\_\_  
Signature of Provider

\_\_\_\_\_  
Credentials

\_\_\_\_\_  
Date

\_\_\_\_\_  
Time

\_\_\_\_\_  
Printed Name of Provider