



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

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

PHYO  
CMC84727-001NS Rev. 11/2020

### Alglucosidase (LUMIZYME) Infusion Therapy Plan

#### BASELINE PATIENT DEMOGRAPHIC

To be completed by the ordering provider.

 NKDA - No Known Drug Allergies    Height: \_\_\_\_\_ cm    Weight: \_\_\_\_\_ kg    Body Surface Area: \_\_\_\_\_ (m<sup>2</sup>)

 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

##### 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) injection

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



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#### 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

#### Select one:

- 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 0.9% injection**  
1 - 20 mL, INTRAVENOUS, PRN, IV line flush
- Sodium chloride - pres free 0.9% injection**  
1 - 30 mL, INTRAVENOUS, PRN, IV line flush

#### 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:** \_\_\_\_\_

- Ibuprofen pre-medication 30 minutes prior (10 mg / kg, maximum 600 mg)**

##### Nursing communication

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

##### Ibuprofen suspension

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

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

##### Ibuprofen tablet

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

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



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**PRE-MEDICATIONS, CONTINUED**

**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 EVERY 30 MINUTES**, check blood pressure, pulse, pulse oximetry, respirations, temperature and pain prior to the start of the infusion. If any vital signs are not within ranges for age, wait for 5 minutes and repeat. Observe vital signs frequently upon the initiation of the infusion for signs and symptoms and / or complaints of infusion related reactions. Monitor fluid intake and urine output during the infusion and as needed.

**Nursing communication**

Must obtain vital signs prior to each increase and if stable may increase as follows (see physician communication order for volume to be administered and infusion rate)

**Physician communication order**

Dose for alglucosidase (LUMIZYME): 20 mg / kg (50 mg vial size). Infusions should be administered in a step-wise manner using an infusion pump. The initial infusion rate should be no more than 1 mg / kg / hour. The infusion rate may be increased by 2 mg / kg / hour every 30 minutes, after patient tolerance to the infusion rate is established, until a maximum rate of 7 mg / kg / hour is reached. Consult the following chart for infusion rates at each step and total volume to be administered:

\*\*Use the following table for total volume based off weight and step by step infusion in mL / hour by weight\*\*

Patient weight (kg) = total volume (mL); rate = step 1, then step 2, then step 3, then step 4 (in mL / hr) with 30 minutes between each step with no infusion reactions

Patient Weight	Total Volume	Step 1	Step 2	Step 3	Step 4 (maximum rate)
1.25 - 10 kg	50 mL	3 mL / hr	8 mL / hr	13 mL / hr	18 mL / hr
10.1 - 20 kg	100 mL	5 mL / hr	15 mL / hr	25 mL / hr	35 mL / hr
20.1 - 30 kg	150 mL	8 mL / hr	23 mL / hr	38 mL / hr	53 mL / hr
30.1 - 35 kg	200 mL	10 mL / hr	30 mL / hr	50 mL / hr	70 mL / hr
35.1 - 50 kg	250 mL	13 mL / hr	38 mL / hr	63 mL / hr	88 mL / hr
50.1 - 60 kg	300 mL	15 mL / hr	45 mL / hr	75 mL / hr	105 mL / hr
60.1 - 100 kg	500 mL	25 mL / hr	75 mL / hr	125 mL / hr	175 mL / hr
100.1 - 120 kg	600 mL	30 mL / hr	90 mL / hr	150 mL / hr	210 mL / hr
120.1 - 140 kg	700 mL	35 mL / hr	105 mL / hr	175 mL / hr	245 mL / hr
140.1 - 160 kg	800 mL	40 mL / hr	120 mL / hr	200 mL / hr	280 mL / hr
160.1 - 180 kg	900 mL	45 mL / hr	135 mL / hr	225 mL / hr	315 mL / hr
180.1 - 200 kg	1,000 mL	50 mL / hr	150 mL / hr	250 mL / hr	350 mL / hr

**Alglucodiase, sodium chloride 0.9% infusion Every 2 weeks Defer until \_\_\_\_\_ Until discontinued**

INTRAVENOUS, for 1 dose, administer over 4 hours. WARNING: ANAPHYLAXIS and RESTRICTED DISTRIBUTION PROGRAM. Life - threatening anaphylactic reactions, severe allergic reactions and immune mediated reactions have been observed in some patients during LUMIZYME infusions. Therefore, appropriate medical support should be readily available when LUMIZYME is administered.

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



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**INTRA-PROCEDURE, CONTINUED**

**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

**Vital signs** Obtain vital signs every hour for 3 hours after infusion for the first 2 infusions. Then obtain vital signs for 3rd and subsequent infusion only once 1 hour after completion of infusion. In the event of any hypersensitivity or other infusion related symptoms, the infusion should be stopped and the ordering provider will be notified.

**EMERGENCY MEDICATIONS**

**Nursing communication**

1. Hives or cutaneous reaction only – no other system involvement **PATIENT IS HAVING A DRUG REACTION:**

- a. Stop the infusion
- b. Give diphenhydramine as ordered
- c. Check vitals including blood pressure every 5 minutes until further orders from provider.
- d. Connect patient to monitor (cardiac / apnea, blood pressure and oxygen saturation), if not already on one
- e. 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:**

- a. Stop the infusion
- b. Call code – do not wait to give epinephrine
- c. Give epinephrine as ordered
- d. Notify provider
- e. Check vitals including blood pressure every 5 minutes until the code team arrives.
- f. Connect patient to monitor (cardiac / apnea, blood pressure and oxygen saturation), if not already on one.
- g. Give diphenhydramine once as needed for hives
- h. May repeat epinephrine every 5 minutes x 2 doses for persistent hypotension and respiratory distress with desaturation until code team arrives.
- i. May give albuterol as ordered for wheezing with oxygen saturations 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 (SPB) less than 90
- OR any age – systolic blood pressure (SPB) drop more than 30% from baseline.
- Baseline systolic blood pressure x 0.7 = value below defined as hypotension.

**EPINEPHrine Injection  
(AMPULE / EPI - PEN JR. / EPI - PEN)**

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



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**ORDERS TO BE COMPLETED FOR EACH THERAPY**

**EMERGENCY MEDICATIONS, CONTINUED**

**Cardio / respiratory monitoring rationale for monitoring:  
high risk patient (please specify risk)**

(Patient receiving infusion with potential infusion reactions); heart rate, respiratory rate, oxygen saturation  
Rationale for Monitoring: High risk patient (please specify risk)  
Parameters: heart rate, respiratory rate, oxygen saturation  
Alarm limits: preset at age specific limits

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

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

**POST - PROCEDURE**

**Nursing communication**

Flush PIV or IVAD with 10 - 20 mL 0.9% sodium chloride at the completion of the infusion. Flush IVAD with saline and heparin flush per protocol prior to de-accessing IVAD.

**Sodium chloride flush 0.9%**

Intravenous at 0 - 25 mL / hr  
**Dose:** \_\_\_\_\_

(circle one):  
MD DO

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

\_\_\_\_\_  
Credentials

\_\_\_\_\_  
Date

\_\_\_\_\_  
Time

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