



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

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

 PHYO  
 CMC0037-001NS Rev. 9/2022

**CHST Agalsidase Beta (FABRAZYME)  
 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

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 occurring.

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 / Access IVAD

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 procedure

 Administration 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.



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**ORDERS TO BE COMPLETED FOR EACH THERAPY**

**NURSING ORDERS, CONTINUED**

**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

**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**

- |   |                                 |                                      |
|---|---------------------------------|--------------------------------------|
| <input checked="" type="checkbox"/> <b>Complete Blood Count With Differential</b><br>Unit collect | <b>INTERVAL: Every 12 weeks</b> | <b>DEFER UNTIL: 1 time this week</b> |
| <input checked="" type="checkbox"/> <b>Blood Urea Nitrogen</b><br>Unit collect                    | <b>INTERVAL: Every 12 weeks</b> | <b>DEFER UNTIL: 1 time this week</b> |
| <input checked="" type="checkbox"/> <b>Creatinine</b><br>Unit collect                             | <b>INTERVAL: Every 12 weeks</b> | <b>DEFER UNTIL: 1 time this week</b> |
| <input checked="" type="checkbox"/> <b>Plasma LysoGB3</b><br>Unit collect                         | <b>INTERVAL: Every 12 weeks</b> | <b>DEFER UNTIL: 1 time this week</b> |
| <input checked="" type="checkbox"/> <b>IgG Antibody Level (Genzyme lab)</b><br>Unit collect       | <b>INTERVAL: Every 12 weeks</b> | <b>DEFER UNTIL: 1 time this week</b> |



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

- 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 of 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 ordering provider. After initiation of the infusion, 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**

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

**\*\*agalsidase beta infusion rates for patients <30 kg\*\***

Infusion should be initiated at a rate of 15 mg / hour (0.25 mg / minute), no further titration recommended, maximum infusion rate: 15 mg / hour (0.25 mg / minute).

**\*\*agalsidase beta infusion rates for patients ≥ 30 kg\*\***

Initial infusion should be initiated at a rate of 15 mg / hour (0.25 mg / minute). If the initial infusion is tolerated, the subsequent infusions may be increased in increments of 3 to 5 mg / hour (0.05 to 0.08 mg / minute). Administration duration minimum 1.5 hours (based upon individual tolerability).

Infusion rate increases based on no infusion reactions:	
1st infusion:	15 mg / hour
9th infusion:	20 mg / hour
17th infusion:	25 mg / hour
Rates can be increased in these increments until a minimum duration time of 1.5 hours is reached (based upon individual tolerability).	



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**INTRA-PROCEDURE, CONTINUED**

**Physician Communication Order**

Dose of agalsidase beta: 1 mg / kg. Please enter the dose of agalsidase beta in 'mg' to facilitate prior authorization requirements and round to nearest 5 mg, if clinically acceptable. Agalsidase beta vial sizes: 5 mg and 35 mg.

**Agalsidase Beta in sodium chloride 0.9%**

**INTERVAL: Every 2 weeks**

**DEFER UNTIL: \_\_\_\_\_**

For 1 dose.

**DOSE: \_\_\_\_\_ mg**

**Vital Signs**

Obtain vital signs at the end of the infusion and 1 hour after the infusion. 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.

**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

**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 heart rate, respiratory rate and 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 heart rate, respiratory rate and 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 saturation stable while waiting for code team, continue to monitor oxygen saturation.

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**EMERGENCY MEDICATIONS, CONTINUED**

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

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

**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 20 mL 0.9% sodium chloride (250 mL bag) at the completion of the infusion.

Flush IVAD with saline and heparin flush per protocol prior to de-accessing IVAD.

Discontinue PIV prior to discharge.

**Sodium chloride 0.9% infusion**

INTRAVENOUS, at 0 - 25 mL / hour ONCE, for 1 dose

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

(circle one):  
MD DO

Signature of Provider

Credentials

Date

Time

Printed Name of Provider