CHILDREN'S HEALTH	Page 1 of 5 Patient Name: Date of Birth:					
PHYO Idursulfase (Elaprase) Infusion CMC85188-001NS Rev. 04/2021 Therapy Plan						
Baseline Patient Demographic						
To be completed by the ordering provider. Diagnosis: Height: cm Weight:kg Body Surface Area: (m ²) 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:						
ORDERS TO BE COMPLETED FOR EACH THERAPY						
✓ 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: month to 1 year - systolic blood pressure (SBP) less than 70 year to 11 years - systolic blood pressure (SBP) less than 70 + (2 x age in years) years to 17 years - systolic blood pressure (SBP) less than 90 OR any age - systolic blood pressure (SBP) and 30% from baseline. 						
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.

Iidocaine - prilocaine (EMLA) cream

TOPICAL, PRN

u when more than 60 minutes are available before procedure u when procedure will take more than 1 hour

patient / family preference for procedure

Administration Instructions: NOTE: In children < 3 months of age, or < 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

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

u when anticipated pain is less than 5 mm from skin surface patient / family preference for procedure

Iidocaine 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 - preserative free 0.9% injection

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

PRE-MEDICATIONS

Acetaminophen pre-medication 60 minutes prior (15 mg / kg, maximum 650 mg)

Nursing communication

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

acetaminophen suspension

15 mg / kg, ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion

Dose: _____

acetaminophen tablet

15 mg / kg ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion **Dose:**

□ Ibuprofen pre-medication 60 minutes prior (10 mg / kg, maximum 600 mg)

Nursing Communication

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

Ibuprofen suspension

10 mg / kg, ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion

Dose:

Ibuprofen tablet

10 mg / kg ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion **Dose:**

Key: BP = blood pressure; cm = centimeter; IV = intravenous; IVAD = implantable venous access device; kg = kilogram; m² = square meters; mg = milligram; mL = milliliter; mL / hr = milliliters per hour; mOsm / I = millisomole per liter; NKDA = No Known Drug Allergies; NS = normal saline; pH = hydrogen ion concentration; PIV = peripheral intravenous; PRN = as needed; PVC = polyvinyl chloride; SBP = systolic blood pressure



Date of Birth:

Patient Name:

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Idursulfase (Elaprase) Infusion Therapy Plan

ORDERS TO BE COMPLETED FOR EACH THERAPY

PRE-MEDICATIONS, CONTINUED

Diphenhydramine pre-medication 60 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 60 minutes prior to infusion **Dose:**

diphenhydrAMINE capsule

1 mg / kg ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion **Dose:**

diphenhydrAMINE injection

1 mg / kg, INTRAVENOUS, 1 dose pre-medication, give 60 minutes prior to infusion **Dose:**

INTRA- PROCEDURE

✓ Nursing Communication

DO NOT SHAKE. Administer with PVC infusion set equipped with low protein binding 0.2 micron filter.

Vital signs

EVERY 15 minute, observe for hypoxia, changes in blood pressure (BP), respiratory distress, angioedema and seizures during infusion. If severe reaction occurs, immedately suspend the infusion and contact ordering provider.

Physician Communication Order

Dose of idursulfase (ELAPRASE) = 0.5 mg / kg. Vials come as 6 mg / 3 mL. Please enter the dose of idursulfase in 'mg' to facilitate prior authorization requirements.

idursulfase in sodium chloride 0.9% 100 r	L infusion INTERVAL: 1 time a week	DEFER UNTIL:	DURATION:
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Dose:

	Rate	Time at that rate
Initial Infusion	8 mL / hour	for 15 minutes
Increase rate to	16 mL / hour	for 15 minutes
Increase rate to	24 mL / hour	for 15 minutes
Increase rate to	32 mL / hour	for 15 minutes
Increase rate to	40 mL / hour	for 15 minutes
Increase rate to	48 mL / hour	for 15 minutes
Increase rate to	56 mL / hour	for 15 minutes
Increase rate to	64 mL / hour	for 15 minutes
Increase rate to	72 mL / hour	for 15 minutes
Increase rate to	80 mL / hour	for 15 minutes
Increase rate to	88 mL / hour	for 15 minutes
Increase rate to	96 mL / hour	for 15 minutes
Then stop infusion	Maximum rate = 100 mL / hour	for 15 minutes until com



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ORDERS TO BE COMPLETED FOR EACH THERAPY

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 Transplant □ Dallas Neurology

Patient Name: _ Date of Birth: _

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 up 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 up 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 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: Ves No



PHYO

Idursulfase (Elaprase) Infusion CMC85188-001NS Rev. 04/2021 Therapy Plan

ORDERS TO BE COMPLETED FOR EACH THERAPY

EMERGENCY MEDICATIONS, CONTINUED

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

Patient Name: Date of Birth:

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

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