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Patient Name:	
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Date of Birth:	

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### Elosulfase (Vimizim) Infusion Therapy Plan

Baseline Patient Demographic
To be completed by the ordering provider.
Diagnosis:
□ 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
□ Iidocaine 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 ≤ 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   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 < 3 months of age, or < 5 kg in weight, maximum application time is 1 hour.



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#### Elosulfase (Vimizim) Infusion Therapy Plan

#### ODDEDS TO BE COMDI ETED FOR EACH THERADY

ORDERS TO BE COMPLETED FOR EACH THERAPY
NURSING ORDERS, CONTINUED
☐ Iidocaine - 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
☐ 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 precedure
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:



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#### Elosulfase (Vimizim) Infusion Therapy Plan

#### ORDERS TO BE COMPLETED FOR EACH THERAPY

PRE-MEDICATIONS, CONTINUED								
<b>Nu</b> Ad	□ 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.							
1 n	diphenhydrAMINE liquid 1 mg / kg, ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion  Dose:							
1 r	diphenhydrAMINE capsule 1 mg / kg ORAL, for 1 dose pre-medication, give 60 minutes prior to infusion  Dose:							
dir	ohenhydrAMINE injectio	on						
1 n	1 mg / kg, INTRAVENOUS, 1 dose pre-medication, give 60 minutes prior to infusion  Dose:							
INTR	A- PROCEDURE							
	ursing Communication  O NOT SHAKE. Administe	er the diluted elosulfa	se with PVC infusion	n set equipped with lo	ow protein binding 0.3	2 micron filter.		
Ev	✓ Vital signs  Every 15 minutes, observe for hypoxia, changes in blood pressure (BP), respiratory distress, angiodema and seizures during infusion. If severe reaction occurs, immediately stop the infusion and contact the ordering provider.							
	Nursing Communication  Administer through port at following schedule:							
D/	060.		< 25 kg			> 25 kg		
D	ose:		Rate	Time at that rate		Rate	Time at that rate	
		Initial rate	3 mL / hour	for 15 minutes	Initial rate	6 mL / hour	for 15 minutes	
		Increase rate to	6 mL / hour	for 15 minutes	Increase rate to	12 mL / hour	for 15 minutes	
		Increase rate to	12 mL / hour	for 15 minutes	Increase rate to	24 mL / hour	for 15 minutes	
		Increase rate to	18 mL / hour	for 15 minutes	Increase rate to	36 mL / hour	for 15 minutes	
		Increase rate to	24 mL / hour	for 15 minutes	Increase rate to	48 mL / hour	for 15 minutes	
		Increase rate to	30 mL / hour	for 15 minutes	Increase rate to	60 mL / hour	for 15 minutes	
		Increase rate to	Maximum rate 36 mL / hour	for 15 minutes	Increase rate to	Maximum rate72 mL / hour	for 15 minutes	
		Then stop infusion		Infusion complete	Then stop infusion		Infusion complete	
Do or	Physician Communication Order  Dose of elosulfase = 2 mg / kg. Vials come as 5 mg / 5 mL. Total volumes and infusion rates are different based on weights of < 25 kg. (100 mL) or ≥ 25 kg (250 mL). Select the appropriate order below that is needed (100 mL for patients < 25 kg and 250 mL for patients ≥ 25 kg. Please enter the dose of elosulfase in 'mg' to facilitate prior authorization requirements.							
☐ eld	elosulfase alfa sodium chloride 0.9% 100 mL infusion INTERVAL: 1 time a week DEFER UNTIL: DURATION:							
Do	ose:		< 25 kg			<u>≥</u> 25 kg		
			Rate	Time at that rate		Rate	Time at that rate	
		Initial rate	3 mL / hour	for 15 minutes	Initial rate	6 mL / hour	for 15 minutes	
		Increase rate to	6 mL / hour	for 15 minutes	Increase rate to	12 mL / hour	for 15 minutes	
	Increase rate to 12 mL / hour for 15 minutes Increase rate to 24 mL / hour for 15 minutes							
		Increase rate to	18 mL / hour	for 15 minutes	Increase rate to	36 mL / hour	for 15 minutes	

for 15 minutes

for 15 minutes

for 15 minutes

Infusion complete

Increase rate to

Increase rate to

Increase rate to

Then stop infusion

48 mL / hour

60 mL / hour

Maximum rate72

mL / hour

for 15 minutes

for 15 minutes

for 15 minutes

Infusion complete

24 mL / hour

30 mL / hour

Maximum rate 36

mL / hour

Increase rate to

Increase rate to

Increase rate to

Then stop infusion



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## Elosulfase (Vimizim) Infusion Therapy Plan

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ORDERS TO BE COMPLETED FOR EACH THERAPY

□ elosulfase alfa in sodium o		L infusion INTER\	/AL: 1 time a week	DEFER UNTIL:	DURATIO	N:	
Dose:					> 25 kg		
		< 25 kg Rate	Time at that rate		Rate	Time at that rate	
	Initial rate	3 mL / hour	for 15 minutes	Initial rate	6 mL / hour	for 15 minutes	
	Increase rate to	6 mL / hour	for 15 minutes	Increase rate to	12 mL / hour	for 15 minutes	
	Increase rate to	12 mL / hour	for 15 minutes	Increase rate to	24 mL / hour	for 15 minutes	
	Increase rate to	18 mL / hour	for 15 minutes	Increase rate to	36 mL / hour	for 15 minutes	
	Increase rate to	24 mL / hour	for 15 minutes	Increase rate to	48 mL / hour	for 15 minutes	
	Increase rate to	30 mL / hour	for 15 minutes	Increase rate to	60 mL / hour	for 15 minutes	
	Increase rate to	Maximum rate 36 mL / hour	for 15 minutes	Increase rate to	Maximum rate72 mL / hour	for 15 minutes	
	Then stop infusion		Infusion complete	Then stop infusion		Infusion complete	
✓ 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 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.
    - 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.

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Elosulfase (Vimizim) Infusion Therapy Plan

#### ORDERS TO BE COMPLETED FOR EACH THERAPY

			1
EMERGENCY MEDICATIONS, CONTINUED			
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 anaphylax 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,  Dose:			sion and respiratory
□ 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 satural Telemetry Required: □ Yes □ No	tion □ Respirator	y rate	
diphenhydrAMINE injection 1 mg / kg, INTRAVENOUS, ONCE PRN, for hives or cutaneous reaction, for Dose:	1 dose maximum do	ose = 50 mg per dose, 300 i	mg per day.
Albuterol for aerosol  0.25 mg / kg., INHALATION ONCE PRN, for wheezing, but oxygen saturate saturation for 1 dose  Dose:	tions stable while wa	aiting for code team, contin	ue to monitor oxygen
POST - PROCEDURE			
■ Nursing communication Flush PIV or IVAD with 20 mL 0.9% sodium chloride (250 mL bag) at the conflush IVAD with saline and heparin flush per protocol prior to de-accessing IVD Discontinue PIV prior to discharge.		on.	
Sodium chloride 0.9% infusion INTRAVENOUS at 0 - 25 mL / hr, ONCE, for 1 dose.  Dose:			
	(circle one): MD DO		
Signature of Provider	Credentials	Date	Time
Printed Name of Provider			