| CHILDREN'S HEALTH  | Page 1 of 4 Patient Name: Date of Birth:                                |
|--|---|
| PHYO         Onasemnogene Abeparvovec (ZOLGENSMA)           CMC0032-001NS         Rev. 8/2022         Infusion Therapy Plan  | )   |
|  | Weight: kg Body Surface Area: (m <sup>2</sup> )                         |
|  | ment.<br>unknown<br>hin the month                                       |
| ORDERS TO BE COMPLETED FOR EACH THERAPY  |   |
| ADMIT ORDERS   |   |
| Height and weight  |   |
| ✓ Vital signs  |   |
| Notify Provider for Hypotension:<br>Prior to starting infusion, please determine the patient's threshold for hypotens<br>needed in the event of an infusion reaction.<br>Hypotension is defined as follows:<br>1 month to 1 year - systolic blood pressure (SBP) less than 70<br>1 year to 11 years - systolic blood pressure (SBP) less than 70 + (2 x age in yea<br>11 years to 17 years - systolic blood pressure (SBP) less than 90<br>OR any age - systolic blood pressure (SBP) drop of more than 30% from baselin<br>Baseline systolic blood pressure (SBP) x 0.7 = value below defined as hypotens | ars)<br>ine.  |
| NURSING ORDERS   |   |
| Please select all appropriate therapy  |   |
| IV START NURSING ORDERS  |   |
| Insert Peripheral IV Place PIV x 2 per onasemnogene aberparvovec protocol  |   |
| Insert Peripheral IV<br>Place PIV if needed or access IVAD if available.   |   |
| Iidocaine 1% BUFFERED (J-TIP LIDOCAINE)<br>0.2 mL, INTRADERMAL, PRN  |   |
| when immediate procedure needed when procedure will take about 1<br>Administration Instructions: NOTE: Do not use this medication in patients<br>anticoagulants, when accessing implanted ports or using a vein that will be<br>neonates.  | with bleeding disorders, platelets $\leq$ 20,000, or in patients taking |
| ☐ Iidocaine - prilocaine (EMLA) cream<br>TOPICAL, PRN  |   |
| <ul> <li>when more than 60 minutes are available before procedure</li> <li>patient / family preference for procedure</li> <li>Administration Instructions: NOTE: In children &lt; 3 months of age, or &lt; 5 kg in w</li> </ul>  |   |
|  |   |

# CHILDREN'S HEALTH



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#### ORDERS TO BE COMPLETED FOR EACH THERAPY

#### NURSING ORDERS, CONTINUED

#### □ lidocaine - tetracaine (SYNERA) patch

TOPICAL, PRN

u when 20 - 30 minutes are available before procedure u 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

#### INTRA-PROCEDURE

#### ☑ Onasemnogene abeparvovec - XIOI (ZOLGENSMA) 2 x 10EXP13 VG / mL

1 Kit, INTRAVENOUS, ONCE, for 1 dose. Dose:

# Vital Signs

Monitor vital signs every 15 minutes x 1 hour during 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 Neurology

Clinically required scheduled time: Next available, Morning

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Onasemnogene Abeparvovec (ZOLGENSMA) Infusion Therapy Plan

#### ORDERS TO BE COMPLETED FOR EACH THERAPY

#### **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.

#### 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: Ves 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:

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#### ORDERS TO BE COMPLETED FOR EACH THERAPY

#### 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:

#### Vital Signs

Monitor vital signs every 15 minutes x 1 hour for the first hour following the infusion. Then, monitor vital signs every 30 minutes x 1 hour for the second hour following the infusion. Observe for signs and symptoms and / or complaints of infusion related reactions. In the event of any hypersensitvity or other infusion related symptoms, the infusion should be stopped and notify the ordering provider.

|                       | (circle one):<br>MD DO |      |      |
|-----------------------|------------------------|------|------|
| Signature of Provider | Credentials            | Date | Time |

Printed Name of Provider