Orsini
Rare Disease Pharmacy Solutions

Fax To: 847-427-7976 Phone: 800-240-9572

AVLAYAH™ INFUSION ORDER

(tividenofusp alfa-eknm)

1 PATIENT INFORMATION

Patient Name: _____ DOB: _____
Primary Phone: _____ Gender: ○ Male ○ Female
Alternate Contact Name: _____
Address: _____ Phone: _____
City, State, Zip: _____
Allergy: _____
Patient Weight: _____Lbs _____Kg Date Weighed: _____

2 PRESCRIBER INFORMATION

Prescriber's Name: _____
Contact Person: _____ Phone: _____
NPI #: _____
Address: _____
City, State, Zip: _____
Phone: _____ Fax: _____
ICD-10 Diagnosis: *E76.1 Hunter Syndrome*

3 CATHETER ACCESS AND FLUSH PROTOCOL

ACCESS TYPE	CATHETER FLUSH ORDERS	
○ Peripheral PORT *(Also include Peripheral IV for Port Malfunction)*	☒ **0.9% Saline Flush:** **Dispense:** 28 Days **Refill:** x 1 Yr Flush line/port with 10mL for patency/SASH protocol. **Heparin Flush:** **Dispense:** 28 Days **Refill:** x 1 Yr Patients < 20kg: Flush port with 3-5mL of Heparin 10units/mL per SASH protocol Patients > 20kg: Flush port with 3-5mL of Heparin 100units/mL per SASH protocol Alternative Heparin Order: _____	**CathFlo:** 2 mg/2 mL as directed. *Pharmacy authorized to dispense upon home health nurse validated port occlusion.* **Dispense:** 1 Kit **Refill:** x 1 Yr

4 PRE-MEDICATION

Premedication for administration 30-60 min. prior to drug infusion:

☐ Antipyretic: _____ q.s. 28 days **Refill:** x 1 Yr
☐ Anthistamine: _____ q.s. 28 days **Refill:** x 1 Yr
☐ Corticosteroid: _____ q.s. 28 days **Refill:** x 1 Yr
☐ **No Premedication Needed**
☐ LMX 4 or (EMLA) Lidocaine 2.5%/Prilocaine 2.5% cream - Apply topically 1 hour prior to starting IV or accessing port **QTY:** 1 **Refill**: x 1Yr
☐ Other Premedications: _____

5 TREATMENT REGIMEN

Site of Care: ○ Clinic (Titration) ○ Clinic (Maintenance Dose) ○ Home

AVLAYAH DOSE	PRESCRIPTION
☐ **3** mg/kg once weekly ☐ **7.5** mg/kg once weekly ☐ **15** mg/kg once weekly ☐ Other: _____	Infuse ____mg of AVLAYAH IV once weekly Disp: 28 days Refills: 12 months *If dose is not specified, pharmacy to calculate dose rounded to the nearest whole vial and dispense appropriate number of vials for infusion* *(Each vial of AVLAYAH yields 150mg/5mL upon reconstitution with 5.2mL of Sterile Water for Injection.)*

PREPARATION AND ADMINISTRATION

Reconstitute each required AVLAYAH vial with 5.2mL of Sterile Water for Injection to yield 150mg/5mL of drug solution per vial
Dilute reconstituted AVLAYAH dose in appropriate volume of 0.9% Sodium Chloride to reach the Total Infusion Volume below. (*Final concentration between 0.6mg/mL and 15mg/mL*)
Total Infusion Volume: ☐25mL ☐50mL ☐100mL ☐250mL ☐Other: _____ mL
Infusion Rate: Infuse at rate schedule indicated in the manufacturer package insert based upon total infusion volume.
Infusion Rate Schedule
Step 1: _____mL/hr for _____min if tolerated increase to
Step 2: _____mL/hr for _____min if tolerated increase to
Step 3: _____mL/hr final rate for remainder of infusion duration
Other Rate Directions: _____
Infuse diluted AVLAYAH solution through in-line low protein-binding 0.2µ filter at rate schedule specified above
Post Infusion: Flush IV with 20mL 0.9% Sodium Chloride Injection, USP at final rate of drug infusion
Vital Signs: At baseline and at every _____ minutes during infusion, at completion of post-infusion flush and 30 minutes after completion of post-infusion flush

SKILLED NURSING VISIT

☒ As needed for IV access, administration, and proper clinical monitoring | Administration procedures to be followed per pharmacy protocol
☒ **Supplies:** Provide infusion pump if needed, IV pole, back-up peripheral IV kit and all necessary infusion supplies including Sterile Water for Injection

6 LAB ORDERS

☐ Protein and Creatinine Random Urine to measure UPCR every 3 months or _____
☐ Cystatin C every 3 months or _____
☐ Hemoglobin every 3 months or _____ ☐ Other Labs: _____

ALSO SEE PAGE 2 (AVLAYAH INFUSION REACTION MANAGEMENT ORDERS)

7 PROVIDER SIGNATURE ("AVLAYAH Infusion Order")

X _____ Product Substitution Permitted Signature _____ Date of Signature

X _____ Dispense as Written Signature _____ Date of Signature

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AVLAYAH™ INFUSION REACTION MANAGEMENT ORDERS

Patient Name: _____ DOB: _____

8 EMERGENCY MEDICATIONS - #Q.S. REFILL X 1 YR

☒ 0.9% Saline 250mL – administer at 50mL/hr once infusion stops OR
☐ 0.9% Saline _____mL – administer at _____ml/hr once infusion stops

☐ **For mild infusion reactions**, give:
Acetaminophen 500mg po or alternate dose: _____
Diphenhydramine 25-50mg po or alternate dose: _____
☐ **For moderate infusion reactions**, give diphenhydramine 25-50mg IV or alternate dose: _____
☐ **For severe infusion reactions**, give Methylprednisolone 1mg/kg IV prn or alternate dose: _____
☐ **Other infusion reaction medication protocol:** _____

☒ **For severe bronchospasm/anaphylaxis, administer appropriate Epinephrine Autoinjector IM and call 911.**
Epinephrine 2-Pak 0.3mg Autoinjector IM for patients weighing ≥ than 30kg
Epinephrine 2-Pak 0.15mg Autoinjector IM for patients weighing < than 30kg

EXAMPLES OF AVLAYAH INFUSION REACTIONS

- Bronchospasm
- Itching
- Hypotension
- Abdominal Pain
- Headache
- Fever and/or Chills
- Cough
- Vomiting
- Angioedema
- Tachycardia
- Rash or Hives
- Flushing

INSTRUCTIONS DURING REACTION

1) STOP AVLAYAH and start 0.9% normal saline at 50mL/hr or at rate specified below:
☐ Other Rate: _____mL/hr

2) ADMINISTER EMERGENCY MEDS FROM BOX 8 ACCORDING TO PHYSICIAN ORDERS
☒ For severe anaphylaxis, administer appropriate Epinephrine Autoinjector IM – may repeat in 20 minutes if needed. Call 911.

3) CALL PHYSICIAN

9 PROVIDER SIGNATURE ("AVLAYAH™ Infusion Reaction Management Orders")

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X _____ Dispense as Written Signature _____ Date of Signature