



**St. Charles Health System**

Adult Ambulatory Infusion Order  
AvaglucoSIdase-alfa (NEXVIAZYME)

Patient Name:  
Date of Birth:

*Patient Identification*

**ALL ORDERS MUST BE MARKED IN INK WITH A CHECKMARK ( ✓ ) TO BE ACTIVE**

Treatment Start Date: \_\_\_\_\_ Allergies: \_\_\_\_\_  
Weight: \_\_\_\_\_ kg Height: \_\_\_\_\_ cm

**REQUIRED ITEMS for all orders – necessary for insurance approval, scheduling, and patient safety**

- 1. FACE SHEET with complete INSURANCE information and patient CONTACT information**
- 2. Recent VISIT NOTE to support treatment (if not available in Epic)**
- 3. LAB RESULTS for any required prescreening (if not available in Epic)**
- 4. DIAGNOSIS CODE \_\_\_\_\_**
- 5. Patient NAME and DATE OF BIRTH on EVERY page faxed**

**GUIDELINES FOR ORDERING**

1. Send FACE SHEET and H&P or most recent chart note.
2. Life-threatening hypersensitivity reactions, including anaphylaxis, and severe infusion-associated reactions (IARs) have occurred in some patients during and after avaglucoSIdase alfa infusions. Patients with an acute underlying illness at the time of infusion may be at greater risk for IARs. Patients with advanced Pompe disease may have compromised cardiac and respiratory function, which may predispose them to a higher risk of severe complications from IARs. Inform patients of the signs and symptoms of anaphylaxis, hypersensitivity reactions, and IARs and have them seek immediate medical care should signs and symptoms occur.

**LABS:**

- CK, Plasma, ONCE, every 8 weeks
- Liver function panel (AST, ALT, BILI TOTAL, BILI DIRECT, ALK PHOS, ALB, PROT TOTAL), ONCE, every 8 weeks

**NURSING ORDERS:**

1. Follow facility policies and/or protocols for vascular access maintenance with appropriate flush solution, dec clotting (alteplase), and/or dressing changes.
2. VITAL SIGNS: Vital signs at baseline, prior to each infusion rate increase, and following infusion. Observe patient for 15 minutes post-infusion.
3. AvaglucoSIdase alfa 20 mg/kg will be administered in a step-wise manner, beginning at an initial rate of 1 mg/kg/hr and increasing by 2 mg/kg/hr every 30 minutes (if there are no signs of infusion-associated reactions (IARs), until a maximum rate of 7 mg/kg/hr is reached.

**PRE-MEDICATIONS: (Administer 30 minutes prior to infusion)**

**Note to provider: Please select which medications below, if any, you would like the patient to receive prior to treatment by checking the appropriate box(s)**

- acetaminophen (TYLENOL) tablet, 650 mg, oral, ONCE, every visit
- cetirizine (ZYRTEC) tablet, 10 mg, oral, ONCE, every visit



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**MEDICATIONS:**

- AvaglucoSIDase alfa (NEXVIAZYME) 20 mg/kg in 0.9% sodium chloride 500 mL, intravenous, EVERY 2 WEEKS, at least 10 days apart. (Pharmacist to round dose for vial size)
  - Use in-line, low protein binding, 0.2 micrometer filter.
  - Refer to nursing orders for infusion instructions. Start infusion no more than 1 mg/kg/hr. May increase by 2 mg/kg/hr every 30 minutes as tolerated to a maximum of 7 mg/kg/hr.

**INFUSION MONITORING/REACTION:**

Infusion Reaction. Acute Infusion and Hypersensitivity Medication Protocol will be used unless the provider selects the option below. If opting out, alternative orders must be included.

1. diphenhydramine 25 mg IV, AS NEEDED x1 for hypersensitivity reaction
2. famotidine 20 mg IV, AS NEEDED x1 dose for hypersensitivity reaction
3. methylprednisolone 125 mg IV, AS NEEDED x1 dose for hypersensitivity reaction
4. epinephrine 0.3 mg IM, AS NEEDED x1 dose for hypersensitivity reaction
5. sodium chloride 0.9% 1000 mL IV, 200 mL/hr, AS NEEDED x 1 dose for alteration in hemodynamic status
6. albuterol 2.5 mg/3 mL nebulizer, AS NEEDED x1 dose for hypersensitivity reaction

Opting out of standard protocol. Alternative orders are attached, or deviations are documented:

\_\_\_\_\_

Patient will be treated at the following infusion location:

- St. Charles Outpatient Infusion Center  
2500 NE Neff Road, Bend, OR 97701  
Phone: (541) 706-5820 Fax: (541) 706-5825

By signing below, I represent the following:

- I am responsible for the care of the patient identified on this form
- I hold an active, unrestricted license to practice medicine
- I am acting within my scope of practice and authorized by law to order the medication described above for the patient identified on this form

**ALL ITEMS BELOW MUST BE COMPLETED TO BE A VALID PRESCRIPTION**

Signature: \_\_\_\_\_ License #: \_\_\_\_\_ Date: \_\_\_\_\_

Print Name: \_\_\_\_\_ Phone: \_\_\_\_\_ Fax: \_\_\_\_\_



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**Plan will expire 1 year after signature date at which time a new order will need to be placed**