

# avalglucosidase alfa-ngpt (NEXVIAZYME) Infusion Therapy Plan Orders

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Name: _____
Kaiser Permanente Member I.D. #: _____
Date of Birth: _____

### Instructions to Provider

Review orders and note any changes. All orders with  will be placed unless otherwise noted. Please fax completed order form to the infusion center where the patient will be receiving treatment (see fax numbers using the link at the end of this protocol).  
Lab orders are not included on this form – place orders via usual method. Lab monitoring is the responsibility of the ordering physician.

Please complete all the following:

Order Date: _____	<b>Diagnosis</b> ICD-10 code (REQUIRED): _____
Weight: _____ kg	ICD-10 description: _____

### General Plan Communication

- **Recommended dosing** (using actual body weight): ≥ 30 kg: 20 mg/kg every 2 weeks; < 30 kg: 40 mg/kg every 2 weeks
- **Special instructions/notes:**  
\_\_\_\_\_

### Infusion Therapy

**Avalglucosidase alfa-ngpt (NEXVIAZYME) in D5W**

**Dose:**  20 mg/kg (patient ABW ≥ 30 kg)  
 Other dose: \_\_\_\_\_

**Route:** Intravenous

**Frequency:** Every 2 weeks

- Infusion Rate:**
- 1 to 7 mg/kg/hour, titrated
  - Start infusion rate at 1 mg/kg/hour
  - Slowly increase by 2 mg/kg/hour every 30 minutes to max 7 mg/kg/hour if no reaction

Please refer to product insert for suggested infusion rates when dose is 40 mg/kg (i.e. patient < 30 kg)

**If infusion-related reaction:**

- 1) STOP infusion immediately
- 2) Begin primary infusion to wide open rate
- 3) Notify MD
- 4) Monitor vital signs
- 5) Administer PRN medications
- 6) 30 minutes after symptoms have resolved, restart infusion at 50% of rate when reaction occurred

**Note any changes to above regimen:**  
\_\_\_\_\_

### Pre-Medications

acetaminophen (TYLENOL) tablet  
**Dose:** 650 mg    **Route:** Oral    **Frequency:** Once, 30 minutes prior to avalglucosidase alfa-ngpt infusion if patient has history of prior reaction. May also be given once as needed during infusion for achiness, headache, or fever

cetirizine (ZYRTEC) tablet  
**Dose:** 10 mg    **Route:** Oral    **Frequency:** Once, 60 minutes prior to avalglucosidase alfa-ngpt infusion (if not taken at home) if patient has history of prior reaction

Other: \_\_\_\_\_  
**Dose:** \_\_\_\_\_    **Route:** Oral    **Frequency:** Once, 30 minutes prior to avalglucosidase alfa-ngpt infusion

### IV Line Care

**Provider Signature:** \_\_\_\_\_ **Date:** \_\_\_\_\_

**Printed Name:** \_\_\_\_\_ **Phone:** \_\_\_\_\_ **Fax:** \_\_\_\_\_

**avalglucosidase alfa-ngpt (NEXVIAZYME)  
Infusion Therapy Plan Orders**

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<input checked="" type="checkbox"/>	<b>5% Dextrose (D5W) infusion 250 mL</b> <i>Rate:</i> 30 mL/hr <i>Route:</i> Intravenous <i>Frequency:</i> Run continuously to keep vein open. Start peripheral IV if no central line
<b>PRN &amp; Hypersensitivity Reaction Medications</b>	
<input checked="" type="checkbox"/>	<b>acetaminophen (TYLENOL) tablet</b> <i>Dose:</i> 650 mg <i>Route:</i> Oral <i>Frequency:</i> Take 650 mg PO every 4 hours PRN for fever (greater than 100.4 F), myalgias, arthralgias or headache.
<input checked="" type="checkbox"/>	<b>alteplase (CATHFLO ACTIVASE) injection</b> <i>Dose:</i> 2 mg <i>Route:</i> Intracatheter <i>Frequency:</i> Instill 2 mg to affected port(s) of central venous catheter if sluggish or occluded. Allow to dwell for 30 minutes, if unable to aspirate blood allow to dwell for an additional 90 minutes. May repeat one time if unsuccessful.
<input checked="" type="checkbox"/>	<b>diphenhydrAMINE (BENADRYL) injectable</b> <i>Dose:</i> 50 mg <i>Route:</i> Intravenous <i>Frequency:</i> Once PRN for urticaria, pruritus, shortness of breath. May repeat one time in 15 minutes if symptoms not resolved. Notify MD upon giving medication.
<input checked="" type="checkbox"/>	<b>famotidine (PEPCID) (PF) injection</b> <i>Dose:</i> 20 mg <i>Route:</i> Intravenous <i>Frequency:</i> Give IV push over 2 minutes for hives, rash, itching, flushing, and/or swelling in a suspected hypersensitivity reaction. Give immediately after diphenhydrAMINE. Notify provider if patient experiences a hypersensitivity reaction.
<input checked="" type="checkbox"/>	<b>methylPREDNISolone Sod Succ (PF) Inj 125 mg (SOLU-Medrol PF)</b> <i>Dose:</i> 125 mg <i>Route:</i> Intravenous <i>Frequency:</i> Give 125 mg IV push one time PRN for shortness of breath, bronchospasm, or other symptoms of a suspected hypersensitivity reaction not otherwise specified. Notify provider if patient experiences a hypersensitivity reaction.
<input checked="" type="checkbox"/>	<b>sodium Chloride 0.9% IV bolus</b> <i>Dose:</i> 1000 mL <i>Route:</i> Intravenous <i>Frequency:</i> Give IV over 1 hour one time PRN for hypotension due to presumed anaphylaxis. Notify provider if patient experiences a hypersensitivity reaction.
<input checked="" type="checkbox"/>	<b>EPINEPHrine (Epi-Pen) 0.3 mg/0.3 mL IM Auto-Injector</b> <i>Dose:</i> 0.3 mg <i>Route:</i> Intramuscular <i>Frequency:</i> Once PRN for anaphylaxis. Give IM one time for severe cardiovascular or respiratory symptoms (e.g. dyspnea, wheeze/bronchospasm, stridor, hypoxemia) of a suspected hypersensitivity reaction. Provider must be present upon given medication.
<b>Nursing Orders</b>	
<ul style="list-style-type: none"> <li>• Begin D5W as primary line to keep vein open.</li> <li>• Perform assessment for toxicity and tolerance.</li> <li>• Monitor for temperature greater than 100.4F, chills, pruritus, chest pain, blood pressure changes (notify MD if greater than 10% drop in systolic blood pressure or if patient is symptomatic), or dyspnea.</li> <li>• For hypersensitivity: stop avalglucosidase alfa-ngpt, give diphenhydramine and steroid as ordered.</li> <li>• Review discharge medications, instructions, and future appointments.</li> </ul>	
<b>References</b>	
Avalglucosidase alfa-ngpt® (NEXVIAZYME) injection for intravenous use Prescribing Information. Revised September 2023.	
<b>Kaiser Permanente Infusion Locations</b>	
Please refer to the link below for the current list and contact information: <a href="https://wa-provider.kaiserpermanente.org/patient-services/ambulatory-infusion">https://wa-provider.kaiserpermanente.org/patient-services/ambulatory-infusion</a>	

**Provider Signature:** \_\_\_\_\_ **Date:** \_\_\_\_\_

**Printed Name:** \_\_\_\_\_ **Phone:** \_\_\_\_\_ **Fax:** \_\_\_\_\_