Weight: ___________ kg  Height: ___________ cm

Allergies: ____________________________________________________________

Diagnosis Code: ______________________________________________________

Treatment Start Date: ___________  Patient to follow up with provider on date: ___________

**This plan will expire after 365 days at which time a new order will need to be placed**

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 set (AST, ALT, BILI TOTAL, BILI DIRECT, ALK PHOS, ALB, PROT TOTAL), ONCE, every 8 weeks
- Hex4, Urine, ONCE, every 8 weeks
- Anti-Avalglucosidase Alfa (NEXVIAZYME) antibody to Labcorp-Sanofi Genzyme, ONCE, every 8 weeks

NURSING ORDERS:
1. Vital signs at baseline, prior to each infusion rate increase, and following infusion. Observe patient for 15 minutes post-infusion.
2. Actual Body Weight ≥30 kg: Avaglucosidase alfa (NEXVIAZYME) 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.
   a. Initial and Subsequent Infusions-DO NOT PRE-PROGRAM PUMP FOR AUTOMATIC TITRATIONS!
      Step 1: 1 mg/kg/hr (0.25 mL/kg/hr) administered over 30 mins - If no signs of IARs, go to next step
      Step 2: 3 mg/kg/hr (0.75 mL/kg/hr) administered over 30 mins - If no signs of IARs, go to next step
      Step 3: 5 mg/kg/hr (1.25 mL/kg/hr) administered over 30 mins - If no signs of IARs, go to next step
      Step 4: 7 mg/kg/hr (1.75 mL/kg/hr) administered over 30 mins - If no signs of IARs, complete infusion at this rate
3. Follow facility policies and/or protocols for vascular access maintenance with appropriate flush solution, declotting (alteplase), and/or dressing changes.
ADULT AMBULATORY INFUSION ORDER

Avalglucosidase Alfa (NEXVIAZYME) Infusion

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
☐ loratadine (CLARITIN) tablet, 10 mg, oral, ONCE, every visit

MEDICATIONS (must check one):

☐ Actual body weight ≥ 30 kg: avalglucosidase alfa (NEXVIAZYME) 20 mg/kg in dextrose 5%, intravenous, ONCE, every 2 weeks
☐ Actual body weight < 30 kg: avalglucosidase alfa (NEXVIAZYME) 40 mg/kg in dextrose 5%, intravenous, ONCE, every 2 weeks

HYPERSENSITIVITY MEDICATIONS:

1. NURSING COMMUNICATION – If hypersensitivity or infusion reactions develop, temporarily hold the infusion and notify provider immediately. Administer emergency medications per the Treatment Algorithm for Acute Infusion Reaction (OHSU HC-PAT-133-GUD, HMC C-132). Refer to algorithm for symptom monitoring and continuously assess as grade of severity may progress.
2. diphenhydRAMINE (BENADRYL) injection, 25-50 mg, intravenous, AS NEEDED x 1 dose for hypersensitivity or infusion reaction
3. EPINEPHrine HCl (ADRENALIN) injection, 0.3 mg, intramuscular, AS NEEDED x 1 dose for hypersensitivity or infusion reaction
4. hydrocortisone sodium succinate (SOLU-CORTEF) injection, 100 mg, intravenous, AS NEEDED x 1 dose for hypersensitivity or infusion reaction
5. famotidine (PEPCID) injection, 20 mg, intravenous, AS NEEDED x 1 dose for hypersensitivity or infusion reaction

By signing below, I represent the following:
I am responsible for the care of the patient (who is identified at the top of this form);
I hold an active, unrestricted license to practice medicine in: ☐ Oregon ☐ ______________________ (check box that corresponds with state where you provide care to patient and where you are currently licensed. Specify state if not Oregon);

My physician license Number is # ______________________ (MUST BE COMPLETED TO BE A VALID PRESCRIPTION); and I am acting within my scope of practice and authorized by law to order Infusion of the medication described above for the patient identified on this form.

Provider signature: ____________________________ Date/Time: ____________________________
Printed Name: ____________________________ Phone: __________________ Fax: __________________
Central Intake:
Phone: 971-262-9645 (providers only) Fax: 503-346-8058

Please check the appropriate box for the patient's preferred clinic location:

- **Beaverton**
  OHSU Knight Cancer Institute
  15700 SW Greystone Court
  Beaverton, OR 97006
  Phone number: 971-262-9000
  Fax number: 503-346-8058

- **NW Portland**
  Legacy Good Samaritan campus
  Medical Office Building 3, Suite 150
  1130 NW 22nd Ave.
  Portland, OR 97210
  Phone number: 971-262-9600
  Fax number: 503-346-8058

- **Gresham**
  Legacy Mount Hood campus
  Medical Office Building 3, Suite 140
  24988 SE Stark
  Gresham, OR 97030
  Phone number: 971-262-9500
  Fax number: 503-346-8058

- **Tualatin**
  Legacy Meridian Park campus
  Medical Office Building 2, Suite 140
  19260 SW 65th Ave.
  Tualatin, OR 97062
  Phone number: 971-262-9700
  Fax number: 503-346-8058

Infusion orders located at: [www.ohsuknight.com/infusionorders](http://www.ohsuknight.com/infusionorders)