Oregon Health & Science University Hospital and Clinics Provider's Orders PO7071 ADULT AMBULATORY INFUSION ORDER **Cipaglucosidase Alfa (POMBILITI) Infusion** Page 1 of 4	ACCOUNT NO. MED. REC. NO. NAME BIRTHDATE *Patient Identification*
ALL ORDERS MUST BE MARKED IN INK WITH A CHECKMARK (✓) TO BE ACTIVE.	

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. For use in combination with migLUstat (OPFOLDA). An outpatient prescription must be ordered by the referring provider and approved for patient prior to scheduling referral to infusion center. MigLUstat is not supplied by ambulatory infusion clinics. Patients must be instructed to bring their prescription with them to the infusion clinic and take the dose of at the start of their infusion visit.
3. Provider verifies patient is not pregnant prior to treatment initiation. Use is contraindicated in patients who are pregnant. Patients who may become pregnant should use effective contraception during therapy with cipaglucosidase alfa (POMBILITI) in combination with migLUstat and for at least 60 days after the last dose.
4. Life-threatening hypersensitivity reactions, including anaphylaxis, and severe infusion-associated reactions (IARs) have occurred in some patients during and after cipaglucosidase 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.
5. Initiate 2 weeks after last enzyme replacement therapy dose. Cipaglucosidase alfa is approved for patients weight greater than or equal to 40 kg who are not improving on their current enzyme replacement therapy.

LABS:

- ☐ CK, PLASMA, every 8 weeks
- ☐ Liver Set (AST, ALT, BILI TOTAL, BILI DIRECT, ALK PHOS, ALB, PROT TOTAL), every 8 weeks
- ☐ Hex4, URINE, every 8 weeks
- ☐ Anti-cipaglucosidase alfa (POMBILITI) antibody, every 8 weeks

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NURSING ORDERS:

1. VITAL SIGNS – At baseline, prior to each infusion rate increase, and following infusion.
2. Confirm patient has been fasting for 2 hours prior to appointment. Confirm patient has migLUstat with them and instruct them take dose following 2 hour fast and at beginning of infusion appointment. Patient should continue to fast for an additional 2 hours following migLUstat administration
3. Cipaglucosidase alfa infusion is started approximately 1 hours after migLUstat dose. Patient will need to be rescheduled if they are unable to take migLUstat at the start of the infusion visit OR if the cipaglucosidase alfa infusion cannot be started within 3 hours of the migLUstat dose.
4. Infuse at a rate of 0.25 mL/kg/hour. Gradually increase infusion rate by 0.5 mL/kg/hour every 30 minutes if no signs of hypersensitivity or infusion-associated reactions, up to a maximum rate of 1.75 mL/kg/hour. Then, maintain infusion rate at 1.75 mL/kg/hour until infusion is complete (total infusion duration: ~4 hours).
   STEP 1 – 0.25 mL/kg/hr for 30 minutes
   STEP 2 – 0.75 mL/kg/hr for 30 minutes
   STEP 3 – 1.25 mL/kg/hr for 30 minutes
   STEP 4 – 1.75 mL/kg/hr (MAX RATE)
5. Follow facility policies and/or protocols for vascular access maintenance with appropriate flush solution, declotting (alteplase), and/or dressing changes.

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

- ☐ acetaminophen (TYLENOL) tablet, 650 mg, oral, ONCE, every visit
- ☐ diphenhydrAMINE (BENADRYL) capsule, 50 mg, oral, ONCE, every visit. ***Give either loratadine or diphenhydrAMINE, not both.***
- ☐ loratadine (CLARITIN) tablet, 10 mg, oral, ONCE AS NEEDED if diphenhydrAMINE is not given, every visit. ***Give either loratadine or diphenhydrAMINE, not both.***
- ☐ methylPREDNISolone sodium succinate (SOLU-MEDROL), 40 mg, intravenous, ONCE, every visit

MEDICATIONS:

- ☐ cipaglucosidase alfa (POMBILITI) 20 mg/kg in sodium chloride 0.9%, intravenous, EVERY 2 WEEKS
  Begin infusion approximately 1 hour after patient takes migLUstat dose from home supply. Administer through a separate line via an in-line, low protein-binding, 0.2 micron filter. Do not infuse in the same IV line as other products. Refrigerate.

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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, 0.5 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**: _____

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OLC Central Intake Nurse:

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