Oregon Health & Science University
Hospital and Clinics Provider's Orders

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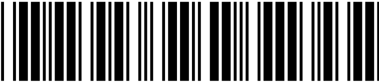

ADULT AMBULATORY INFUSION ORDER
Pegunigalsidase Alfa-iwxj (ELFABRIO) Infusion

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. Patients treated with pegunigalsidase alfa-iwxj have experienced hypersensitivity reactions, including anaphylaxis.
3. Consider premedication in patients who are enzyme replacement therapy (ERT)-naive or who are ERT-experienced but previously required pretreatment medications. After 4 to 6 infusions, a stepwise decrease in pretreatment medication dose(s) and/or discontinuation of pretreatment medication(s) may be considered in patients tolerating treatment.
4. The development of IgG anti-drug antibodies may occur and has been observed within 26 weeks (about 6 months) from the onset of therapy. Monitoring presence of IgG and IgE anti-pegunigalsidase alfa-iwxj antibodies is recommended in patients developing hypersensitivity or infusion reactions
5. Membranoproliferative glomerulonephritis with immune deposits in the kidney leading to decreased renal function has been reported.
6. Patients with advanced Fabry disease may have compromised cardiac function which may predispose them to a higher risk of severe complications from infusion reactions.
7. Infusion rate is dependent on prior enzyme replacement exposure. Provider must indicate if patient is enzyme replacement therapy _________ (naïve) (experienced) – *Circle One*

NURSING ORDERS:

1. VITAL SIGNS – Monitor and record vital signs, tolerance, and presence of infusion-related reactions prior to infusion, with every rate increase, then hourly until infusion is complete.
2. **Initial 4 infusions**: Infuse pegunigalsidase alfa-iwxj using an in-line low protein binding 0.2-micron filter.

   Initial infusion rate (enzyme replacement naive)
   - < 70 kg: 37.5 mL/hour
   - 70-100 kg: 60 mL/hour
   - > 100 kg: 83 mL/hour

   Initial infusion rate (swithing from other enzyme treatment (e.g. fabrazyme))
   - < 70 kg: 50 mL/hour
   - 70-100 kg: 83 mL/hour
   - > 100 kg: 167 mL/hour

3. **Subsequent infusions:** Subsequent infusions if no infusion reactions: rate may be decreased in decrements of 30 minutes every 3rd infusion (5th, 8th, 11th, 14th, 17th, 20th) to allow a total infusion time of no less than 1.5 hours.

Enzyme Replacement Therapy–**Naive** Patients Infusion Rate (mL/hour)

Patient weight (actual body weight)	**Infusion Rate (mL/hr)**									
	Initial 4	**5th**	**8th**	**11th**	**14th**	**17th**	**20th**	**23rd**	**26th**	**29th**
< 70 kg Total volume: 150 mL	37.5	43	50	60	75	100				
70-100 kg Total volume 250 mL	60	68	79	94	115	150				
> 100 kg Total volume 500 mL	83	91	100	111	125	143	167	200	250	333

*May decrease the duration of every third infusion after the initial 4 infusions in decrements of 30 minutes as tolerated.

Enzyme Replacement Therapy–**Experienced** Patients Infusion Rates (mL/hour)

Patient weight (actual body weight)	**Infusion Rate (mL/hr)**			
	Initial 4	**5th**	**8th**	**11th**
< 70 kg Total volume: 150 mL	50	60	75	100
70-100 kg Total volume 250 mL	83	100	125	167
> 100 kg Total volume 500 mL	167	200	250	333

*May decrease the duration of every third infusion after the initial 4 infusions in decrements of 30 minutes as tolerated.

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

- PROVIDER TO PHARMACIST COMMUNICATION - OK to discontinue diphenhydrAMINE (BENADRYL) pre-medication after fourth infusion, if tolerated.

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
☐ 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, administer over 5-60 minutes, every visit.

MEDICATIONS:

- Pegunigalsidase Alfa-iwxj (ELFABRIO) 1 mg/kg = _______ mg in sodium chloride 0.9% IV infusion, ONCE, every 2 weeks

Infuse per nursing order. Administer using an in-line low protein binding 0.2-micron filter. Flush infusion line with NS using the same infusion rate used for the last part of the pegunigalsidase infusion. Refrigerate.

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**:_____________

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