

Agalsidase Beta (FABRAZYME) Infusion Therapy Plan Orders

Name: _____

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 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 of the following:

Order Date: _____ Weight: _____ kg	Diagnosis: ICD-10 code (REQUIRED): _____ ICD-10 description _____ _____
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General Plan Communication

- Every year collect the following blood sample for antibody and GL-3 enzyme testing. Genzyme provides kit with instructions and blood collection tubes. Kits can be ordered from Genzyme by any staff member at 1-800-745-4447.
- Next blood/enzyme collection date _____

Infusion Therapy

- Agalsidase Beta (FABRAZYME) IV infusion in 0.9% Sodium Chloride**
Dose: _____ mg (will be rounded the nearest 5 mg) = 1 mg/kg x weight (kg)

Route: Intravenous

 Frequency: every 2 weeks _____

Infusion Volume (based upon patient weight):

Patient weight (kg)	Minimum Total Volume (mL)
≤ 35	50
35.1 – 70	100
70.1 - 100	250
> 100	500

Infusion Rate:

Initial infusion rate should not exceed 0.25 mg/minute.

Interrupt or decrease rate in the event of an infusion reaction; may be restarted after resolution of symptoms and/or after administration of antipyretics, antihistamines, and/or steroids. After patient tolerance to the infusion is established, rate may be increased in increments of 0.05-0.08 mg/minute with each subsequent infusion.

Maximum infusion rate: Patients <30 kg: 0.25 mg/minute; patients ≥30 kg: Infuse over at least 1.5 hours.

If infusion-related reaction:

- 1) STOP infusion immediately;
- 2) Increase primary infusion to wide open rate;
- 3) Administer PRN medications per hypersensitivity protocol;
- 4) Notify MD

Note any changes to above regimen: _____

Pre-Meds

- acetaminophen (TYLENOL) tablet
 Dose: 975 mg Route: Oral Frequency: Once, 30 minutes prior to agalsidase beta (Fabrazyme) infusion.
 May also be given once as needed during infusion for fever, headache, or myalgia.
- cetirizine (ZYRTEC) tablet
 Dose: 10 mg Route: Oral
 Frequency: Once, at least 60 minutes prior to agalsidase beta (Fabrazyme) infusion.

Provider Signature: _____ **Date:** _____

Printed Name: _____ **Phone:** _____ **Fax:** _____

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- No routine pre-medications necessary. Above pre-meds may be given if patient has reaction and requires pre-medications for future doses.

IV Line Care

- 0.9% sodium chloride infusion 250 mL
Rate: 30 mL/hr Route: Intravenous Frequency: Run continuously to keep vein open
 Start peripheral IV if no central line

Infusion Reaction Meds

- Acetaminophen Tab 650 mg (TYLENOL)
Dose: 650 mg Route: Oral Frequency: EVERY 4 HOURS AS NEEDED for fevers greater than 100.4 F, myalgias, arthralgias or headache.
- albuterol (PROVENTIL) nebulizer solution 0.083%
Dose: 2.5 mg Route: Nebulization Frequency: PRN for shortness of breath/wheezing
- diphenhydramine (BENADRYL) injectable
Dose: 25 mg Route: Intravenous Frequency: Once PRN, May repeat x1 for urticaria, pruritus, shortness of breath. May repeat in 15 minutes if symptoms not resolved.
- EPINEPHrine (EpiPen) 0.3 mg/0.3 mL IM Auto-Injector
Dose: 0.3 mg Route: Intramuscular Frequency: Once PRN for anaphylaxis. Inject into lateral thigh and hold for 10 seconds. Massage the injected area. Use for patients weighing greater than 27.3 kg (60 lbs). Use amp and 1.5 inch needle for patients with BMI greater than 30. Notify physician if administered.
- MethylPREDNISolone Sod Succ (PF) Inj 125 mg (SOLU-Medrol PF).
Dose : 125 mg Route : IV push Frequency: Once PRN for hypersensitivity reaction. Notify MD upon giving medication.

Lab Review for Nursing

- Ensure baseline lab (e.g., SCr) is drawn within 3 months of initial treatment if provider has ordered.

Nursing Orders

- ~ Anaphylaxis (e.g., hypotension, angioedema, urticaria or other rash, pruritus, and dyspnea) has been reported in patients. Discontinue infusion if any of these symptoms are observed and report to MD for additional instructions.
- ~ Infusion related reaction: STOP infusion immediately, begin primary solution at wide open rate, notify MD, begin monitoring vital signs, and administer prn medication for infusion reaction; once symptoms have resolved, consult with MD on rate to resume infusion.
- ~ Monitor patient for at least one hour after medication has infused for evidence of adverse reaction.
- ~ Review discharge medications, instructions, and future appointments.
- ~ Following infusion, flush line with Normal Saline until all drug is infused.
- ~ Discontinue IV line when therapy complete and patient stabilized.

References

- [Fabrazyme Prescribing Information Revised May 2019](#)

Kaiser Permanente Infusion Locations

Bellevue Medical Center
 11511 NE 10th St, Bellevue, WA 98004
Fax: 425-502-3512 Phone: 425-502-3510

Capitol Hill Medical Center
 201 16th Ave E, Seattle WA 98112
Fax: 206-326-2104 Phone: 206-326-3109

Riverfront Medical Center – Spokane
 W 322 North River Drive, Spokane, WA 99201
Fax: 509-324-7168 Phone: 509-241-2073

Silverdale Medical Center
 10452 Silverdale Way NW, Silverdale, WA 98383
Fax: 360-307-7421 Phone: 360-307-7316

Provider Signature: _____ Date: _____

Printed Name: _____ Phone: _____ Fax: _____

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Everett Medical Center
2930 Maple St, Everett, WA 98201
Fax: 425-261-1578 Phone: 425-261-1566

Tacoma Medical Center
209 Martin Luther King Jr Way, Tacoma, WA 98405
Fax: 253-383-6262 Phone: 253-596-3666

Olympia Medical Center
700 Lilly Road N.E., Olympia, WA 98506
Fax: 360-923-7106 Phone: 360-923-7164

Provider Signature: _____ Date: _____

Printed Name: _____ Phone: _____ Fax: _____