

Agalsidase beta (FABRAZYME) Infusion Therapy Plan Orders

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: _____	Diagnosis ICD-10 code (REQUIRED): _____
Weight: _____ kg	ICD-10 description: _____

General Plan Communication

- Annual antibody and GL-3 enzyme testing is supported by Genzyme, ordering can be completed through the LabCorp portal. Contact Genzyme with questions at 1-800-745-4447.
- Next blood/enzyme collection date: _____
- **Special instructions/notes:**

Infusion Therapy

Agalsidase beta (FABRAZYME) in 0.9 % Sodium Chloride

Dose: 1 mg/kg (will be rounded to the nearest 5 mg)

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

Initial infusion rate: 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.

Infusion Rate:

Patients ≥30 kg: 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.

Note any changes to above regimen:

Pre-Medications

<input checked="" type="checkbox"/> acetaminophen (TYLENOL) tablet	Dose: 650 mg	Route: Oral	Frequency: Once, 30 minutes prior to infusion. May also be given once as needed during infusion for fever, headache, or myalgia
<input checked="" type="checkbox"/> cetirizine (ZYRTEC) tablet	Dose: 10 mg	Route: Oral	Frequency: Once, 30 minutes prior to infusion

Provider Signature: _____ **Date:** _____

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

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<input checked="" type="checkbox"/>	Other: _____	Dose: _____	Route: Oral	Frequency: Once, 30 minutes prior to infusion
IV Line Care				
<input checked="" type="checkbox"/>	0.9% Sodium Chloride (NS) infusion 250 mL	Rate: 30 mL/hr	Route: Intravenous	Frequency: Run continuously to keep vein open. Start peripheral IV if no central line
PRN & Hypersensitivity Reaction Medications				
<input checked="" type="checkbox"/>	acetaminophen (TYLENOL) tablet	Dose: 650 mg	Route: Oral	Frequency: Take 650 mg PO every 4 hours PRN for fever (greater than 100.4 F), myalgias, arthralgias or headache.
<input checked="" type="checkbox"/>	alteplase (CATHFLO ACTIVASE) injection	Dose: 2 mg	Route: Intracatheter	Frequency: 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"/>	diphenhydrAMINE (BENADRYL) injectable	Dose: 50 mg	Route: Intravenous	Frequency: Once IV push over 2 minutes PRN for urticaria, pruritus, shortness of breath. Notify provider if patient experiences a hypersensitivity reaction.
<input checked="" type="checkbox"/>	famotidine (PEPCID) (PF) injection	Dose: 20 mg	Route: Intravenous	Frequency: Give once IV push over 2 minutes PRN 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"/>	methylPREDNISolone Sod Succ (PF) Inj 125 mg (SOLU-Medrol PF)	Dose: 125 mg	Route: Intravenous	Frequency: 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"/>	sodium Chloride 0.9% IV bolus	Dose: 1000 mL	Route: Intravenous	Frequency: 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"/>	EPINEPHrine (Epi-Pen) 0.3 mg/0.3 mL IM Auto-Injector	Dose: 0.3 mg	Route: Intramuscular	Frequency: Give IM once PRN for anaphylaxis/allergic reaction. May repeat one time. KPWA Asthma/Allergy and Pediatrics endorse: For patients < 25 kg (55 lbs): 0.15 mg. For patients ≥ 25 kg (55 lbs): 0.3 mg. Inject into anterolateral aspect of the thigh.
Nursing Orders				
<ul style="list-style-type: none"> • Begin 0.9% NS as primary line to keep vein open. • 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: <ol style="list-style-type: none"> 1. STOP infusion immediately 2. Begin primary solution at wide open rate 				

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

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3. Notify MD
 4. Begin monitoring vital signs, and administer prn medication for infusion reaction
 5. Once symptoms have resolved, consult with MD on rate to resume infusion
- Following infusion, flush line with Normal Saline until all drug is infused.
 - Monitor patient for at least one hour after medication has infused for evidence of adverse reaction.
 - Review discharge medications, instructions, and future appointments.
 - Discontinue IV line when therapy complete and patient stabilized.

References

Agalsidase beta® (FABRAZYME) injection for subcutaneous or intravenous use Prescribing Information. Revised July 2024.

Kaiser Permanente Infusion Locations

Please refer to the link below for the current list and contact information:
<https://wa-provider.kaiserpermanente.org/patient-services/ambulatory-infusion>

Provider Signature: _____ **Date:** _____

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