## Agalsidase beta (Fabrazyme)

**Provider Order Form** 



## PATIENT INFORMATION

Date: Patient Name:	DOB:
ICD-10 code (required):	ICD-10 description:
□ NKDA Allergies:	Weight lbs/kg:
Patient Status: ☐ New to Therapy ☐ Continuing Therapy	Next Due Date (if applicable):
PROVIDER INFORMATION	
Ordering Provider:	Provider NPI:
Referring Practice Name:	Phone: Fax:
Practice Address:	City: State: Zip Code:
NURSING	THERAPY ADMINISTRATION
<ul> <li>Provide nursing care per IVX Standard Nursing Procedures, including reaction management</li> </ul>	<ul> <li>□ Agalsidase beta (Fabrazyme) in 0.9% sodium chloride, intravenous infusion, administer with 0.2 micron filter</li> <li>□ 1mg/kg / □ other</li> </ul>
LABORATORY ORDERS	■ Frequency: □ every 2 weeks
□ CBC □ at each dose □ every   □ CMP □ at each dose □ every   □ Other: □ Other:    PRE-MEDICATION ORDERS  □ acetaminophen (Tylenol) □ 500mg / □ 650mg / □ 1000mg PO   □ cetirizine (Zyrtec) 10mg PO   □ loratadine (Claritin) 10mg PO   □ diphenhydramine (Benadryl) □ 25mg / □ 50mg □ PO / □ IV   □ methylprednisolone (Solu-Medrol) □ 40mg / □ 125mg IV   □ Other: □ Dose: □ Route:   Frequency: □ SPECIAL INSTRUCTIONS	<ul> <li>Infuse over one hour</li> <li>Initial intravenous infusion rate is no more than 0.25 mg/min (15mg/hr). After tolerance to the infusion is well established, increase the infusion rate in increments of 0.05 to 0.08 mg/min (increments of 3 to 5 mg/hr). Maximum infusion rate for patients weighing less than 30kg is 0.25mg/min (15mg/hr). For patients &gt;30kg, the minimum infusion duration is 1.5 hours.</li> <li>✓ Flush with 0.9% sodium chloride at the completion of infusion</li> <li>✓ Patient is required to stay for 30-minute observation post infusion</li> <li>✓ Patient is NOT required to stay for observation time</li> <li>✓ Refills: ✓ Zero / ✓ for 12 months / ✓ (if not indicated order will expire one year from date signed)</li> </ul>
Ordering Provider: Initial here	and proceed to the next page.

## **ADULT REACTION MANAGEMENT PROTOCOL**

- Observe for hypersensitivity reaction: Fever, chills, rigors, pruritus, rash, cough, sneezing, throat irritation, nausea, vomiting
- ☑ If reaction occurs:
  - Stop infusion.
  - Maintain/establish vascular access.
  - Notify referring provider as clinically appropriate and follow clinical escalation protocol.
  - IVX Health clinicians have the following PRN medications available for the following reactions.
    - o Headache, pain, fever >100.4F, chills or rigors- Acetaminophen 650mg PO or Ibuprofen 400mg PO.
    - o Mild Hives, itching, redness, or rash- Loratadine 10mg PO or Diphenhydramine 50mg PO.
    - o Severe hives, itching, redness, or rash- Diphenhydramine 25-50mg SIVP.
    - o Nausea, vomiting, heartburn, acid reflux- Ondansetron 4mg ODT (may repeat x 1 in 20 minutes if nausea continues, max dose 8mg) or Famotidine 20mg PO.
    - Severe Nausea, vomiting, heartburn, acid reflux- Ondansetron 4mg SIVP (may repeat x 1 in 20 minutes if nausea continues, max dose 8mg) or Famotidine 20mg SIVP.
    - Hypotension (90/60), vasovagal response- Place patient in reclined position, administer 0.9% Sodium Chloride IV 250ml. May repeat to keep BP >90/60, maximum of 1000ml, monitor vital signs.
    - Chest pain/discomfort, shortness of breath- Oxygen 2-15 liters, titrate to keep Spo2 >92%.
    - o Solumedrol 125mg IV- Refractory to other treatments given.
    - o Other
  - When symptoms resolve resume infusion at 50% previous rate and increase per manufactures guidelines.

## ☑ Severe allergic/anaphylactic reaction:

- If symptoms are rapidly progressing or continuing after administration of PRN medications above and signs symptoms of severe allergic/anaphylactic reaction (angioedema, swelling of the mouth, tongue, lips, or airway, dyspnea, bronchospasm with or without hypotension or hypertension.)
  - o Call 911
  - o Initiate basic life support as needed.
  - o Bring the **AED** to the patient (Attach pads if indicated).
  - Epinephrine- administer 0.3mg of a 1:1,000 (1mg/ml) concentration intramuscularly (preferably outer thigh), may be repeated every 5-15 minutes as needed to a maximum of 3 doses.
  - Place patient in recumbent position, elevate lower extremities.
  - o **Oxygen** administer 2-15 liters/minute or 100 percent oxygen as needed maintain SpO2 >92 percent.
  - IV Fluids- Treat hypotension with normal saline bolus of 500ml, repeat as needed to maintain systolic BP >90.
  - o Administer **diphenhydramine** 50mg IV or Famotidine 20mg IVP, if not previously given.
  - Administer methylprednisolone 125mg IVP, if not previously given.
  - Continuous monitoring of blood pressure, pulse oximetry, and heart rate.
  - o Notify clinical executive, DON or CMO, when appropriate. Must be done same day. Do not delay treatment.

Patient Name	Patient Date of Birth
Provider Name (Print)	
Provider Signature	Date

Email ivxintake@ivxhealth.com or fax this form, insurance card (both sides), demographics, recent H&P, labs, and supporting clinicals to:		
CHICAGO: 312-253-7244GlenviewSchaumburgLombardNaperville	TAMPA: 844-946-0849BrandonCarrollwoodWesley Chapel	
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