Guidance for the Sample Order Form

The sample order form on page 2 is for the sole purpose of providing a template that can be used to order NEXVIAZYME (avalglucosidase alfa-ngpt). If you choose to use this sample order form, please copy and paste the information on page 2.

IMPORTANT SAFETY INFORMATION AND INDICATION

INDICATION

NEXVIAZYME (avalglucosidase alfa-ngpt) is indicated for the treatment of patients 1 year of age and older with late-onset Pompe disease [lysosomal acid alpha-glucosidase (GAA) deficiency].

IMPORTANT SAFETY INFORMATION

BOXED WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, AND RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

BOXED WARNING: Hypersensitivity Reactions Including Anaphylaxis: Patients treated with NEXVIAZYME have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during NEXVIAZYME administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, NEXVIAZYME should be discontinued immediately and appropriate medical treatment should be initiated. In patients with severe hypersensitivity reaction, a desensitization procedure to NEXVIAZYME may be considered

BOXED WARNING: Infusion-Associated Reactions (IARs): Patients treated with NEXVIAZYME have experienced severe IARs. If severe IARs occur, consider immediate discontinuation of NEXVIAZYME, initiation of appropriate medical treatment, and the benefits and risks of readministering NEXVIAZYME following severe IARs. Patients with an acute underlying illness at the time of NEXVIAZYME 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.

BOXED WARNING: Risk of Acute Cardiorespiratory Failure in Susceptible Patients: Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during NEXVIAZYME infusion. More frequent monitoring of vitals should be performed during NEXVIAZYME infusion.

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: Life-threatening hypersensitivity reactions, including anaphylaxis, have been reported in NEXVIAZYME-treated patients. Some of the hypersensitivity reactions were IgE mediated. Anaphylaxis signs and symptoms included respiratory distress, chest discomfort, flushing, cough, erythema, lip swelling, pruritus, swollen tongue, dysphagia, and rash. Symptoms of severe hypersensitivity reactions included respiratory distress, erythema, urticaria, tongue edema, and rash. Increased incidence of hypersensitivity reactions was observed in patients with higher antidrug antibody (ADA) titers. Prior to NEXVIAZYME administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids.

Infusion-Associated Reactions: In clinical studies, IARs were reported to occur at any time during and/or within a few hours after the NEXVIAZYME infusion and were more likely to occur with higher infusion rates. IARs that led to treatment discontinuation were chest discomfort, cough, dizziness, erythema, flushing, nausea, ocular hyperemia, and respiratory distress. Increased incidence of IARs was observed in patients with higher ADA titers. Antihistamines, antipyretics, and/or corticosteroids can be given prior to NEXVIAZYME administration to reduce the risk of infusion-associated reactions (IARs). However, IARs may still occur in patients after receiving pretreatment.

Risk of Acute Cardiorespiratory Failure in Susceptible Patients: Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during the NEXVIAZYME infusion. More frequent monitoring of vitals should be performed during NEXVIAZYME infusion. Some patients may require prolonged observation times.

ADVERSE REACTIONS

The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia, and urticaria.

Please see full Prescribing Information, including **Boxed WARNING.**

Sample Order Form

It is recommended that the following information, at a minimum, be included in the physician order to the pharmacy and infusion staff.

Patient Name: _____ Diagnosis: _____
Date of Birth: _____ Patient Weight (kg): _____
Today's Date: _____
Notify Dr. _____at phone/pager _____ when patient arrives.

1) Obtain and record patient weight (kg) above.
2) Obtain vital signs prior to infusion.
3) Prepare:
   - ❒ 20 mg/kg (for patients weighing ≥30 kg) avalglucosidase alfa-ngpt in 5% dextrose injection, to a total volume of ___ mL.
   - ❒ 40 mg/kg (for patients weighing <30 kg) avalglucosidase alfa-ngpt in 5% dextrose injection, to a total volume of ___ mL.

   Monitor vital signs during NEXVIAZYME infusion as prescribed by the healthcare provider. It is recommended that patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated, have more frequent monitoring of vitals.

4) Pre-treatment medication (if prescribed by the treating physician): _____

5) Using an infusion pump, administer intravenously in a stepwise manner. Use a 0.2 µm in-line, low protein binding filter.
   a) Begin the infusion at rate of 1 mg/kg/hr (____mL/hr) and administer for at least 30 minutes. If no signs of IARs…
   b) Increase the infusion rate to ____mg/kg/hr (____mL/hr) and administer for at least 30 minutes. If no signs of IARs…
   c) Increase the infusion rate to ____mg/kg/hr (____mL/hr) and administer for at least 30 minutes. If no signs of IARs…
   d) Increase the infusion rate to ____mg/kg/hr (____mL/hr) and administer at this rate for the remainder of the infusion if 4-step process. If 5-step process, this rate should be administered for at least 30 minutes. If no signs of IARs…
   e) If 5-step process, increase the infusion rate to ____mg/kg/hr (____mL/hr) and administer at this rate for the remainder of the infusion.

6) Flush infusion line with 5% dextrose at the last tolerated infusion rate to ensure the entire dose is administered. Do not IV push the flush.

7) Obtain vital signs _____ hours after completion of the infusion. Some patients may require prolonged observation times.

Contact Dr. _____immediately in the event of a hypersensitivity reaction or infusion-associated reaction.

To report **suspected adverse reactions**, contact Sanofi at 1-800-745-4447, option 2 or the FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Physician's Signature: _____ Date: _____
Phone/Pager Number: _____