For the use only of a Registered Medical Practitioner or Hospital or a Laboratory

▼Abbreviated Prescribing Information
Avalglucosidase Alfa Powder for concentrate for solution for infusion 100mg/vial
NEXVIAZYME®
Pre-filled syringe with needle shield
Name and Presentation: Nexviazyme®, 100 mg/vial. Each vial contains 10 mg/mL avalglucosidase alfa after reconstitution with sterile water for injection (WFI) (100 mg extractable dose.Nexviazyme is available as Sterile lyophilized powder administered by intravenous (IV) infusion and Powder for concentrate for solution for infusion. **Therapeutic indication:** Nexviazyme® is indicated for the treatment of long-term enzyme replacement therapy for the treatment of patients with Pompe disease (acid α-glucosidase deficiency). **Dosage and administration:** Patients with late-onset Pompe disease (LOPD): The recommended dose of Nexviazyme is 20 mg/kg of body weight administered once every 2 weeks. Patients with infantile-onset Pompe disease (IOPD): For IOPD patients who experience lack of improvement or insufficient response in cardiac, respiratory, and/or motor function while receiving 20 mg/kg, a dose increases to 40 mg/kg every other week should be considered in the absence of safety concerns (e.g., severe hypersensitivity, anaphylactic reactions, or risk of fluid overload). In patients who do not tolerate avalglucosidase alfa at 40 mg/kg every other week (e.g., severe hypersensitivity, anaphylactic reactions, or risk of fluid overload), consider decreasing the dose to 20 mg/kg every other week. **Special Population:** No dose adjustment is required in patients >65 years. The safety and efficacy of avalglucosidase alfa in patients with hepatic impairment have not been evaluated and no specific dose regimen can be recommended for these patients. No dose adjustment is required in patients with mild renal impairment. The safety and efficacy of avalglucosidase alfa in patients with moderate or severe renal impairment have not been evaluated and no specific dose regimen can be recommended for these patients. The safety and efficacy of avalglucosidase alfa in children 6 months of age and younger have not yet been established. There are no data available in patients 6 months of age and younger **Contraindications:** Life-threatening hypersensitivity to the active substance or to any of the excipients when re-challenge was unsuccessful. **Warnings and precautions:** HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS: Hypersensitivity reactions, including anaphylaxis, have been reported in Nexviazyme-treated patients. Appropriate medical support measures, including cardiopulmonary resuscitation equipment especially for patients with cardiac hypertrophy and patients with significantly compromised respiratory function, should be readily available when Nexviazyme is administered. If severe hypersensitivity or anaphylaxis occur, Nexviazyme should be discontinued immediately, and appropriate medical treatment should be initiated. The risks and benefits of re-administering Nexviazyme following anaphylaxis or severe hypersensitivity reaction should be considered. Some patients have been re-challenged using slower infusion rates at a dose lower than the recommended dose. In patients with severe hypersensitivity, desensitization procedure to Nexviazyme may be considered. If the decision is made to re-administer the product, extreme caution should be exercised, with appropriate resuscitation measures available. Once a patient tolerates the infusion, the dose may be increased to reach the approved dose. If mild or moderate hypersensitivity reactions occur, the infusion rate may be slowed or temporarily stopped. INFUSION-ASSOCIATED REACTIONS: In clinical studies, IARs were reported to occur at any time during and/or within a few hours after the infusion of Nexviazyme and were more likely with higher infusion rates. The majority of IARs were assessed as mild to moderate and included symptoms such as chills, cough, diarrhea, erythema, fatigue, headache, influenza like illness, nausea, ocular hyperemia, pain in extremity, pruritus, rash, rash erythematous, tachycardia, urticaria, vomiting, chest discomfort, dizziness, hyperhidrosis, lip swelling, oxygen saturation decreased, pain, palmar erythema, swollen tongue and tremor.Patients with an acute underlying illness at the time of Nexviazyme infusion appear to 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. Antihistamines, antipyretics, and/or corticosteroids can be given to prevent or reduce IARs. However, IARs may still occur in patients after receiving pretreatment. If severe IARs occur, immediate discontinuation of the administration of Nexviazyme should be considered and appropriate medical treatment should be initiated. The benefits and risks of re- administering Nexviazyme following severe IARs should be considered. Some patients have been re- challenged using slower infusion rates at a dose lower than the recommended dose. Once a patient tolerates the infusion, the dose may be increased to reach the approved dose. If a mild or moderate IARs occur regardless of pre-treatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms. IMMUNOGENICITY: Treatment emergent anti-drug antibodies (ADA) were reported in both treatment naïve (95%) and treatment experienced patients (49%)). IARs and hypersensitivity reactions may occur independent of the development of ADA. The majority of IARs and hypersensitivity reactions were mild or moderate and were managed with standard clinical practices. In clinical studies, the development of ADA did not impact clinical efficacy. ADA testing may be considered if patients do not respond to therapy. Adverse-event-driven immunologic

testing, including IgG and IgE ADA, may be considered for patients who have risk for allergic reaction or previous anaphylactic reaction to Alglucosidase alfa. RISK OF ACUTE CARDIORESPIRATORY FAILURE: Caution should be exercised when administering Nexviazyme to patients susceptible to fluid volume overload or patients with acute underlying respiratory illness or compromised cardiac and/or respiratory function for whom fluid restriction is indicated. These patients may be at risk of serious exacerbation of their cardiac or respiratory status during infusion. Appropriate medical support and monitoring measures should be readily available during Nexviazyme infusion, and some patients may require prolonged observation times that should be based on the individual needs of the patient.: CARDIAC ARRHYTHMIA AND SUDDEN DEATH DURING GENERAL ANESTHESIA FOR CENTRAL VENOUS CATHETER PLACEMENT: Caution should be used when administering general anesthesia for the placement of a central venous catheter or for other surgical procedures in patients with IOPD with cardiac hypertrophy. Cardiac arrhythmia, including ventricular fibrillation, ventricular tachycardia, and bradycardia, resulting in cardiac arrest or death, or requiring cardiac resuscitation or defibrillation, have been associated with the use of general anesthesia in IOPD patients with cardiac hypertrophy. **Drug interactions:** *No drug interaction studies have been conducted with Nexviazyme. Because it is a recombinant human protein, avalglucosidase alfa is an unlikely candidate for cytochrome P450 mediated drug-drug interactions.* **Fertility, pregnancy and lactation:** There are no available data on the use of Nexviazyme in pregnant women. No conclusions can be drawn regarding whether or not Nexviazyme is safe for use during pregnancy. Nexviazyme should be used during pregnancy only if the potential benefits to the mother outweigh the potential risks, including those to the fetus. There are no available data on the presence of Nexviazyme in human milk or the effects of Nexviazyme on milk production or the breastfed infant. No conclusions can be drawn regarding whether Nexviazyme is safe for use during breastfeeding. Nexviazyme should be used during breastfeeding only if the potential benefits to the mother outweigh the potential risks, including those to the breastfed child. **Effects on ability to drive:** No studies on the effects on the ability to drive and use machines have been performed. Because dizziness, hypotension, and fatigue have been reported as IARs, this may affect the ability to drive and use machines on the day of the infusion. **Undesirable effects:** The most common adverse reactions (>5%) were headache, fatigue, diarrhea, nausea, arthralgia, dizziness, myalgia, pruritus, vomiting, dyspnea, erythema, paresthesia and urticaria. **Overdose:** IARs are more likely to occur with higher infusion rates. In a clinical study, pediatric patients received doses up to 40 mg/kg of body weight every other week. **Special precautions for storage:** Store in a refrigerator between 2°C to 8°C (36° to 46°F). Do not use Nexviazyme after the expiration date on the vial.

For full and latest prescribing information, refer to company website www.sanofi.in.

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Updated: Dec 2025
Source: CCDS-v3.1-LRC-30-Jun-2022-revised-19-Jul-2022 & EU SmPC