PATIENT MEDICATION INFORMATION

READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE

PrMYOZYME®

Alglucosidase alfa for injection

Read this carefully before you start taking **Myozyme** and each time you get a refill. This leaflet is a summary and will not tell you everything about this drug. Talk to your healthcare professional about your medical condition and treatment and ask if there is any new information about **Myozyme.**

Serious Warnings and Precautions

Do not use Myozyme if you are severely allergic to alglucosidase alfa or any other ingredient of Myozyme .

If you are treated with Myozyme you may experience an infusion-associated reaction. An infusion-associated reaction is defined as any related side effect occurring during the infusion or during the 2 hours following infusion. Life-threatening allergic reactions, including anaphylactic shock, have been observed in patients during Myozyme infusion. Because of the potential for severe infusion reactions, appropriate medical support should be readily available when Myozyme is administered.

Individuals with an acute underlying illness [e.g fever, pneumonia or sepsis (severe infection), wheezing/difficulty in breathing, heart failure] at the time of Myozyme infusion appear to be at greater risk for infusion reactions. Careful consideration should be given to your clinical status prior to administration of Myozyme.

Precaution must be observed when administering general anesthesia to individuals with infantile-onset Pompe's Disease. Reports of intraoperative cardiac arrest following anesthesia induction for invasive procedures have been reported, some of which were fatal. The presence of severe hypertrophic cardiomyopathy in infantile-onset Pompe's Disease may increase the risk of general anesthesia complications.

Infantile onset Pompe patients with heart or breathing problems may be at risk for increasing the seriousness of these problems as a result of Myozyme administration, and may require additional monitoring.

What is Myozyme used for?

- Myozyme is a medicine used for patients with Pompe's Disease (GAA deficiency). Myozyme is used to treat adults, children and adolescents of all ages who have a confirmed diagnosis of Pompe's disease.

How does Myozyme work?

People with Pompe's Disease have low levels of an enzyme called alpha-glucosidase (GAA). This enzyme helps the body control levels of glycogen (a type of carbohydrate). Glycogen provides the body with energy, but in Pompe's Disease the levels can get too high. Glycogen accumulation in Pompe's Disease occurs in various tissues, particularly cardiac, respiratory and skeletal muscle, leading to the development of cardiomyopathy and progressive muscle weakness, including impairment of respiratory function.

Myozyme is an artificial enzyme called alglucosidase alfa – this can replace the natural enzyme which is lacking in Pompe's disease.

What are the ingredients in Myozyme?

Medicinal ingredient: alglucosidase alfa

Non-medicinal ingredients: Mannitol, Polysorbate 80, Sodium phosphate dibasic heptahydrate, Sodium phosphate monobasic monohydrate

Myozyme comes in the following dosage forms:

Sterile lyophilized powder for reconstitution to be used as intravenous infusion, 50 mg

Do not use Myozyme if:

- You have any allergies to this drug or its ingredients or components of the container

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take Myozyme. Talk about any health conditions or problems you may have, including if you:

- Have an acute underlying illness
- Need general anaesthesia for central venous catheter placement
- Have had a severe hypersensitivity or anaphylactic reaction to administration of Myozyme
- Have experienced infusion-associated reactions
- Are at increased risk of lung infections due to the progressive effects of the disease on the lung muscles
- Have underlying heart enlargement
- Are pregnant or plan to become pregnant or are breast-feeding
- Are above the age of 65

Other warnings you should know about:

You may feel sleepy during or after your Myozyme infusion. Use caution in driving or using other machinery following your infusion.

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

The following may interact with Myozyme:

No formal interaction studies have been conducted. Please inform your doctor if you are using any other medicinal products including herbal or dietary supplements, due to the potential risk of interference with the uptake of alglucosidase alfa.

How to take Myozyme:

Myozyme will be given to you under the supervision of a doctor who is knowledgeable in the treatment of Pompe's Disease.

The dose you receive is based on your body weight. Myozyme should be administered as an intravenous infusion.

Infusions will be administered in a step-wise manner beginning at an initial rate of 1 mg/kg/h and gradually increasing until a maximum rate of 7 mg/kg/h if there are no signs of infusion associated reactions (IARs).

You may be at an increased risk of an IAR if you are given Myozyme at a higher dose or infusion rate than recommended. If you experience reactions such as those listed in the Serious Side Effects table below, you should tell your doctor immediately.

Home infusion

Your doctor may consider that you can have home infusion of Myozyme if it is safe and convenient to do so. If you get any side effects during an infusion of Myozyme, your home infusion staff member should stop the infusion immediately and if necessary, start appropriate medical treatment.

Usual dose:

The recommended dosage regimen of Myozyme is 20 mg/kg body weight administered every 2 weeks as an intravenous infusion.

Overdose:

There is no experience with overdoses of Myozyme for doses up to 40 mg/kg of body weight. Infusion associated reactions are more likely to occur at higher doses.

If you think you, or a person you are caring for, have taken too much Myozyme, contact a healthcare professional, hospital emergency department, or regional poison control centre immediately, even if there are no symptoms.

Missed Dose:

If you have missed a Myozyme infusion, please contact your doctor. It is important to have your infusion on a regular basis. The total dose administered each month should remain substantially unchanged.

What are possible side effects from using Myozyme?

These are not all the possible side effects you may have when taking Myozyme. If you experience any side effects not listed here, tell your healthcare professional.

Side effects were mainly seen while patients were being given the medicine or shortly after ("infusion related effects"). Some of these infusion related side effects became serious. Should you experience any reaction like this, please **tell your doctor immediately**. Regardless of pre-treatment, your infusion may need to be slowed or stopped and you may need to be given additional medicines to treat an allergic reaction.

The most significant infusion reactions included allergic reactions and allergic shock to Myozyme. Other serious infusion reactions included hives, abnormal breathing sounds, elevated heart rate, difficulty in breathing, elevated respiration, swelling around the eyes, high blood pressure, decreased oxygen concentration in blood and fever, heart attack, chest pain, abdominal pain, low blood pressure, shortness of breath.

Some patients have experienced infusion related side effects in the form of flu-like symptoms or a combination of events such as fever, chills, muscle pain, joint pain, pain or fatigue, which lasted for a few days after completion of the infusion.

In addition, patients also experienced the following non-serious events:

cough, infusion site reaction including pain and bruising, feeling unwell, itching, diarrhea, nausea, vomiting, dry heaves, constipation, stomach bloating, indigestion, inability to sleep, agitation, irritability, restlessness, tremor, headache, dizziness, tingling sensation, lack of energy, ringing in the ears, blood in the urine and sleepiness.

Serious side effects (Infusion Reactions), How Often they happen and what to do about them	
Symptom / effect	**Talk to your healthcare professional**
COMMON (occurred in ≥ 5% of patients)	
fever, decreased oxygen concentration in blood, hives, flushing, elevated heart rate, rash, shivering, low blood pressure, high blood pressure, cough, elevated respiration, agitation, irritability, vomiting, chest discomfort, burning sensation	✓
UNCOMMON (occurred in <5% of patients)	
increased sweating, mottling, itching, rash, fever, pallor, cyanosis, restlessness, retching, tremor, chest pain, throat tightness, tongue swelling	✓
UNKNOWN FREQUENCY	

Serious side effects (Infusion Reactions), How Often they happen and what to do about them	
Symptom / effect	**Talk to your healthcare professional**
fainting	✓

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, tell your healthcare professional.

If you opt for the infusion of Myozyme through central catheter, discuss with your doctor potential complications related to use of such a delivery system.

Reporting Side Effects

You can report any suspected side effects associated with the use of health products to Health Canada by:

- Visiting the Web page on Adverse Reaction Reporting (https://www.canada.ca/en/health-canada/services/drugs-health-products/medeffect-canada.html) for information on how to report online, by mail or by fax; or
- Calling toll-free at 1-866-234-2345.

NOTE: Contact your health professional if you need information about how to manage your side effects. The Canada Vigilance Program does not provide medical advice.

Storage:

Store Myozyme in a refrigerator between 2°-8°C. DO NOT FREEZE OR SHAKE. DO NOT USE Myozyme after the expiration date on the vial.

It is recommended that Myozyme is used immediately after it has been mixed with sterile water. However it can be kept for up to 24 hours if it is kept cool (2°C – 8°C) and in the dark.

Keep out of reach and sight of children.

Pompe Registry:

Sanofi informs all patients with Pompe's Disease that a registry has been established in order to better understand the variability and progression of Pompe's Disease and to continue to monitor and evaluate the safety and efficacy of Myozyme treatments. All patients are encouraged to participate and advised that their participation may involve long-term follow-up. Information regarding the registry program may be found at www.pomperegistry.com or by calling 1-800-745-4447.

If you want more information about Myozyme:

- Talk to your healthcare professional
- Find the full product monograph that is prepared for healthcare professionals and includes this

Patient Medication Information by visiting the Health Canada website: (https://www.canada.ca/en/health-canada/services/drugs-health-products/drug-products/drug-product-database.html); the manufacturer's website www.sanofi.com/en/canada, or by calling 1-800-265-7927.

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