PATIENT INFORMATION LEAFLET

SCHEDULING STATUS: S4

MYOZYME® 50 mg powder for concentrate for solution for infusion

Alglucosidase alfa

Contains sugar alcohol: after reconstitution, each vial contains 20 mg/mL mannitol.

Read all of this leaflet carefully before MYOZYME is administered to you.

- Keep this leaflet. You may need to read it again.
- If you have further questions, please ask your doctor, pharmacist, nurse or other health care provider.

What is in this leaflet

1. What MYOZYME is and what it is used for

MYOZYME is used to treat patients who have a confirmed diagnosis of Pompe disease. People with Pompe disease have low levels of an enzyme called alfa-glucosidase. This enzyme helps the body control levels of glycogen (a type of carbohydrate). Glycogen provides the body with energy, but in Pompe disease the levels can get too high.

MYOZYME contains an artificial enzyme called alglucosidase alfa – this can replace the natural enzyme which is lacking in Pompe disease.

2. What you need to know before MYOZYME is administered to you

MYOZYME should not be given to you

- If you are allergic (hypersensitive) to alglucosidase alfa or any of the other ingredients of MYOZYME (listed in section 6 of this leaflet).

Warnings and precautions

Take special care with MYOZYME:

- If you are treated with MYOZYME, you may experience a reaction while you are being given MYOZYME or during the next 2 hours following your infusion. This is known as an infusion-associated reaction and can sometimes be very severe. Such a reaction comprises of different symptoms (see section 4). If you experience a reaction like this, you should tell your doctor immediately. You may need to be given additional medicines to prevent an allergic reaction (e.g. antihistamines, corticosteroids or paracetamol).
- If you experience a severe infection of your airways or lungs, speak to your doctor. Avoid any medicines which suppress the immune system, as this may worsen your infection (see section “Other medicines and MYOZYME”).
- If you experience severe ulcerative lesions of your skin, please inform your doctor.
- If you experience swelling of your lower limbs or generalised swelling, please inform your doctor.
- If you suffer from cardiac hypertrophy (abnormal thickening of the heart muscle), especially if you are about to receive anaesthesia during surgery, please inform your doctor.

Children and adolescents

The recommended dosage of MYOZYME in children and adolescents is the same as in adults.

Other medicines and MYOZYME

Always tell your health care provider if you are taking any other medicine. (This includes complementary or traditional medicines.)

MYOZYME with food and drink

There are no known interactions between MYOZYME and food.

Pregnancy and breastfeeding

The safe use of MYOZYME in pregnancy and breastfeeding has not been established.

If you are pregnant or breastfeeding, think you may be pregnant or are planning to have a baby, please consult your doctor, pharmacist or other health care provider for advice before using MYOZYME.

Driving and using machines

Take care when driving or using any tools or machines shortly after infusion of MYOZYME, since you may experience dizziness.

3. How MYOZYME is administered

Do not share any medicines prescribed for you with any other person.

Always use MYOZYME exactly as your doctor has told you. Check with your doctor if you are not sure.

You will not be expected to give yourself MYOZYME. It will be given to you by a person who is qualified to do so.

MYOZYME is given through a drip into a vein (by intravenous infusion). It is supplied as a powder which will be mixed with sterile water before it is administered.

MYOZYME is only used under the supervision of a doctor who is knowledgeable in the treatment of Pompe disease.

The dose you receive is based on your body weight. The recommended dosage of MYOZYME is 20 mg/kg body weight given once every 2 weeks.

Your doctor will tell you how long your treatment with MYOZYME will last.

If you have the impression that the effect of MYOZYME is too strong or too weak, tell your doctor or pharmacist.

If you receive more MYOZYME than you should

Since a health care provider will administer MYOZYME, he/she will control the dosage. However, in the event of overdosage your doctor will manage the overdosage.

If you forget to use MYOZYME

MYOZYME is administered under the supervision of your doctor. If you think that you have missed an infusion, please contact your doctor.

4. Possible side effects

MYOZYME can have side effects.

Not all side effects reported for MYOZYME are included in this leaflet. Should your general health worsen or if you experience any untoward effects while using MYOZYME, please consult your doctor, pharmacist or other health care provider for advice.

If any of the following happens, stop receiving MYOZYME and tell your doctor immediately or go to the casualty department at your nearest hospital:

- Swelling of your hands, feet, ankles, face, lips, mouth or throat, which may cause difficulty in swallowing or breathing.
- Rash, hives or itching.
- Fainting.
- Infusion-associated reactions that are serious and may be life-threatening (see section “Warnings and precautions”). It includes symptoms such as low blood pressure, chest discomfort, very fast heart rate, difficulty breathing (bronchospasm), throat tightness, coughing, vomiting, facial, lip or tongue swelling (angioedema), hives or rash. You may also

experience infusion-associated reactions in the form of flu-like symptoms, which can last for a few days after completion of your MYOZYME infusion.

These are all serious side effects. If you have them, you may have had a serious allergic or infusion-associated reaction to MYOZYME. You may need urgent medical attention or hospitalisation.

Tell your doctor immediately or go to the casualty department at your nearest hospital if you notice any of the following:

- Changes in the way your heart beats, such as slow heart rate (bradycardia) or fast heart rate (tachycardia).
- Heart stops beating (cardiac arrest).
- Bluish discolouration of the skin.
- Increased or high blood pressure.
- Abnormal breathing sounds, including a whistling sound, shortness of breath, wheezing, stopping breathing.
- Open sores or wounds that develop on the skin, which develop infection.
- Kidney disorder (nephrotic syndrome) that comprises of side effects such as severe swelling around your eyes and of your ankles, foamy urine, weight gain, fatigue and loss of appetite.
- Increased levels of protein in the urine.
- Low levels of oxygen in the blood.

These are all serious side effects. You may need urgent medical attention.

Tell your doctor as soon as possible if you notice any of the following:

Frequent side effects:

- Agitation, tremor.
- Headache.
- Dizziness.
- Tingling or prickling (pins and needles sensation) in the arms, hands, legs or feet.

- Paleness, redness of the skin, (facial) flushing.
- Increased breathing rate, elevated respiration.
- Cough.
- Chest discomfort.
- Nausea, diarrhoea, vomiting.
- Retching.
- Itchy skin.
- Mottled skin.
- Increased sweating.
- Muscle pain, muscle spasms.
- Fever, chills.
- Swelling of the arms and legs.
- Irritability.
- Feeling hot.
- Tiredness.

Side effects with unknown frequency:

- Restlessness.
- Inflammation of the membrane that covers your eyeball and eyelid.
- Narrowing of the blood vessels causing blood flow to be decreased.
- Hypotension.
- Abdominal pain.
- Eyes tearing.
- Joint pain.
- Pain or local reaction at the site of the drip.
- Cold extremities (e.g. hands, feet).

If you notice any side effects not mentioned in this leaflet, please inform your doctor or

pharmacist.

Reporting of side effects

If you get side effects, talk to your doctor or pharmacist. You can also report side effects to:

- The Pharmacovigilance Unit at Sanofi: za.drugsafety@sanofi.com (email) or 011 256 3700 (tel), or
- SAHPRA via the "6.04 Adverse Drug Reaction Reporting Form", found online under SAHPRA's publications: https://www.sahpra.org.za/Publications/Index/8

By reporting side effects, you can help provide more information on the safety of MYOZYME.

5. How to store MYOZYME

- Store under refrigeration between 2 °C and 8 °C.
- Do not use after the expiry date printed on the vial or carton.
- The reconstituted and diluted solution should be administered without delay.
- If immediate use is not possible, the reconstituted and diluted solution is stable for up to 24 hours at 2 °C to 8 °C.
- The reconstituted and diluted infusion solution should be protected from light.
- Storage of the reconstituted solution at room temperature is not recommended.
- DO NOT FREEZE OR SHAKE.
- STORE ALL MEDICINES OUT OF REACH OF CHILDREN.
- Return all unused medicine to your pharmacist.
- Do not dispose of unused medicine in drains and sewerage systems (e.g. toilets).

6. Contents of the pack and other information

What MYOZYME contains

The active ingredient is alglucosidase alfa. Each vial contains 50 mg of alglucosidase alfa.

After reconstitution, the solution contains 5 mg/mL alglucosidase alfa (total extractable dose of 50 mg/10 mL). After dilution, the concentration varies from 0,5 mg/mL to 4 mg/mL.

The other ingredients are mannitol, polysorbate 80, sodium phosphate dibasic heptahydrate and sodium phosphate monobasic monohydrate.

What MYOZYME looks like and contents of the pack

A sterile, non-pyrogenic, white to off-white lyophilised cake or powder.

Single-use, clear type I glass 20 mL (cc) vials. The closure consists of a siliconised butyl stopper and an aluminium seal with a plastic flip-off cap.

Pack size: 1 vial per carton.

Holder of certificate of registration

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