



**Naglazyme**<sup>®</sup>  
(GALSULFASE)

# PREPARING FOR HOME INFUSIONS

A medication guide  
for patients



**B.OMARIN**

Please see Important Safety Information and  
Prescribing Information on back cover.

# MAKING THE TRANSITION TO HOME INFUSION

Your doctor has determined that you can safely receive your NAGLAZYME® (galsulfase) treatments at home. With home infusion, you'll save time on travel by not going to a clinic each week. You'll also receive your infusion in the comfort of familiar surroundings.

This guide will help you make the transition to home infusion. The following pages will explain the steps to take to prepare your home, how to store NAGLAZYME vials safely, and what you'll need to do on your infusion day.

Home infusion is not an option for every person with Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Doctors make the decision on the specifics of each case, and insurance can be an issue—some private insurance plans, as well as some state Medicaid programs, do not cover home infusion. Depending on your individual circumstances and how you tolerate home treatment, your doctor may decide to move your infusions back to the medical center.



# WHAT WILL CHANGE IF I RECEIVE NAGLAZYME® (galsulfase) INFUSIONS AT HOME?



The most important difference with home infusion is that you will receive the NAGLAZYME® (galsulfase) vials directly from a specialty pharmacy, and you will need to store them in your refrigerator.



Then a nurse will come to your home on the scheduled day, prepare the medication, perform the infusion, and monitor your vital signs throughout—just like when you receive your infusions in a medical center. You will also still receive all the same support you've had from BioMarin and BioMarin RareConnections™ to help make sure everything runs smoothly.



Please see Important Safety Information and Prescribing Information on back cover.

# PREPARING FOR YOUR FIRST HOME INFUSION

Both the infusion supplies and the NAGLAZYME® (galsulfase) vials will be delivered directly to your home from a specialty pharmacy via FedEx® or another delivery service. You will need to be home to receive your shipment of NAGLAZYME vials.

**Infusion supplies:** Prepare ahead for your first shipment of supplies, which will be several boxes of tubing, equipment, saline, etc. Select a cool, dry place for storage. If you store supplies in your kitchen cabinets, be sure the shelf is not close to a heat source, such as a water heater, stove, or refrigerator fan.

**NAGLAZYME vials:** You should also prepare an area in your refrigerator for the medication. As soon as you receive your shipment of NAGLAZYME, store it in your refrigerator at a temperature of 2°C to 8°C (36°F to 46°F). Do not freeze it or heat it in any way.

## Storage and care of NAGLAZYME

- Store the medicine in your refrigerator as soon as you receive it at a temperature of 2°C to 8°C (36°F to 46°F). DO NOT FREEZE.
- Never freeze the medicine. Do not heat the vials with hot water or in a microwave.
- Take the medicine out of the refrigerator 30 to 60 minutes before your scheduled infusion. Do not shake the vials.
- If you do not receive your infusion, your nurse should be capping the vials (or tubing, if already mixed) and putting them back in the refrigerator. Do not use medicine that has been left at room temperature for more than 24 hours.

**NAGLAZYME that has been heated, frozen, left unrefrigerated for more than 24 hours, or damaged in any way, should be separated and placed in the refrigerator. Tell your nurse, and then contact BioMarin RareConnections™ at 1-866-906-6100 for help and instructions.**

Please see Important Safety Information and Prescribing Information on back cover.

## Steps for infusion day

- 1 Before the infusion, the NAGLAZYME® (galsulfase) vials will need to warm up to room temperature.** Remove them from the refrigerator 30 to 60 minutes before your home infusion nurse is scheduled to arrive. Be careful not to shake the vials.

Your doctor may recommend you take an antihistamine (like Benadryl®) and perhaps a fever reducer (like Tylenol®) 30 to 60 minutes before your infusion begins. The purpose of these medicines is to help prevent allergic reactions during or after infusion.

If skin numbing medication (like Emla®) is prescribed, this would be a good time to apply the cream to the IV site area. It may need up to 60 minutes to work.

It's important to stay hydrated—it can make finding a vein for IV access easier for the infusion nurse when he or she arrives.

You may also eat before the infusion.

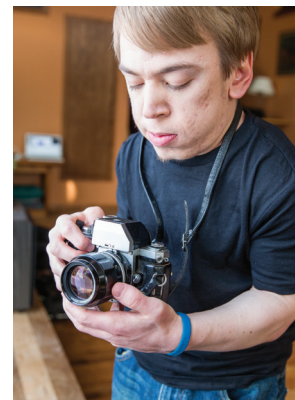
- 2 Upon arrival, your nurse will check for several things before she or he begins the infusion:**

- Your vital signs to make sure you don't have a fever or infection. Be sure to tell your nurse of any illnesses or changes in your health status. It may affect whether you should delay your weekly infusion.
- The infusion supplies to make sure everything needed for the infusion is available, including emergency medications in the event of an infusion reaction.

- 3 Your nurse will then start an IV, prepare the NAGLAZYME solution, and begin the infusion.** During the 4-hour infusion period, your nurse will monitor your vital signs and may adjust the rate of infusion as necessary. You will have some mobility during this time, but be careful not to disrupt your IV line.

The antihistamines you receive may make you drowsy. Until you know how the medication affects you and how quickly it wears off, you should avoid driving, using machines, or doing anything else that could be dangerous if you are not alert.

- 4 After the infusion, your nurse will remove the IV line and carefully dispose of the needles, tubing, and any unused medicine.** Your nurse may wait with you for a short period of time after the infusion to watch for any signs of a delayed allergic reaction.



# SAFETY INFORMATION FOR HOME INFUSIONS

NAGLAZYME® (galsulfase) is generally well tolerated, but can result in side effects. Talk to your doctor and your home infusion nurse to develop a plan everyone is comfortable with in order to handle any complications that may arise.

## During the infusion, let your nurse know immediately if you experience any of the following:

- Itching or hives
- Swelling on your face or hands
- Swelling or tingling in your mouth or throat
- Tightness in your chest
- Difficulty breathing
- Feeling very drowsy or sleepy
- Fever or chills
- Lightheadedness or fainting
- Any other side effects you think may be caused by the medicine

If you experience any of these side effects after your nurse has left, call your doctor's office immediately for guidance.

## Possible side effects of NAGLAZYME

In people taking NAGLAZYME, some serious, even life-threatening, reactions could occur, including allergic reactions (anaphylaxis). Because NAGLAZYME is given by infusion, there are also reactions that may happen as a result of the infusion.

In clinical studies, the most common side effects of NAGLAZYME were:

- rash
- pain
- hives
- fever
- itching
- chills
- headache
- nausea
- vomiting
- abdominal pain
- difficulty breathing

Please see Important Safety Information and Prescribing Information on back cover.



## Protecting NAGLAZYME® (galsulfase) during power outages

It is important to do everything you can to make sure your medication stays safely refrigerated until use. If there is an electrical outage in your home, follow these steps to protect NAGLAZYME® (galsulfase):

- 1 Avoid opening the door of the refrigerator until power is restored.
- 2 Write down the time when the power outage began.
- 3 As soon as the electricity comes back on, write down the time and the temperature inside the refrigerator. If the power outage exceeds 24 hours, or if the temperature inside the refrigerator rises to 77°F or higher, call BioMarin RareConnections™ at 1-866-906-6100.

For power outages longer than 24 hours, you may want to move NAGLAZYME to a location that has refrigeration. If transportation will take no longer than 1 hour, follow these steps:

- 1 Remove NAGLAZYME from the refrigerator. Write down the time and temperature inside the refrigerator when you remove the medication.
- 2 Place NAGLAZYME in an insulated container, such as a small ice chest with a lid. There is no need to add ice or coolant.
- 3 Transport the insulated container to the new location and place NAGLAZYME in the new refrigerator.
- 4 Write down the time and the internal temperature of the new refrigerator.

### Important:

**NAGLAZYME that has been heated, frozen, left unrefrigerated for more than 24 hours, or damaged in any way, should be separated and placed in the refrigerator. Tell your nurse, and then contact BioMarin RareConnections™ at 1-866-906-6100 for help and instructions.**

## Indication

NAGLAZYME® (galsulfase) is indicated for patients with Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). NAGLAZYME has been shown to improve walking and stair-climbing capacity.

## Important Safety Information

Severe and life-threatening allergic reactions can occur during NAGLAZYME® (galsulfase) infusions and up to 24 hours after infusion. Typical signs of an allergic reaction include shock, difficulty breathing, wheezing, swelling of the throat, and low blood pressure. If a severe allergic reaction occurs during infusion, the infusion should be stopped immediately and you should receive medical attention. Contact your doctor or get medical help right away if you develop any severe symptoms after infusion.

In clinical trials, most patients developed antibodies to NAGLAZYME treatment. There was no clear relationship between antibody formation and the safety or effectiveness of NAGLAZYME.

Serious and severe infusion reactions are associated with NAGLAZYME, including hives, chest pain, rash, abdominal pain, difficulty breathing, swelling, fever, and eye irritation. You should receive medication such as antihistamines before NAGLAZYME infusions to reduce the risk of infusion reactions. If an infusion reaction occurs, the infusion should be slowed or stopped and you may be given additional medication.

The most common side effects of NAGLAZYME seen in clinical trials were rash, pain, hives, fever, itching, chills, headache, nausea, vomiting, abdominal pain and difficulty breathing. The most common side effects requiring medical attention are infusion-related effects.

These are not all of the possible side effects with NAGLAZYME. Talk to your doctor if you have any symptoms that bother you or that do not go away.

NAGLAZYME is a prescription medicine. Before treatment with NAGLAZYME, it is important to discuss your medical history with your doctor. Tell your doctor if you are taking any medication and if you are allergic to any medicines. Your doctor will decide if NAGLAZYME is right for you. If you have questions or would like more information about NAGLAZYME, contact your doctor.

Spinal cord damage may occur due to the natural MPS VI disease process. Signs of spinal cord injury include back pain, loss of bladder and bowel control, numbness, and paralysis. Contact your doctor immediately if you develop any of these symptoms.

**Call your doctor for medical advice about side effects. You may report side effects to BioMarin Pharmaceutical Inc. at 1-866-906-6100, or FDA at 1-800-FDA-1088 or go to [www.fda.gov/medwatch](http://www.fda.gov/medwatch).**

**For more information, call BioMarin RareConnections™ at 1-866-906-6100.**

**Please see accompanying full Prescribing Information or visit [www.naglazyme.com](http://www.naglazyme.com).**