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 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.

To report suspected adverse reactions contact BioMarin Pharmaceutical Inc. at 1-866-906-6100, or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

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Preparing for Home Infusions
A medication guide for patients
Other than the location, home infusion is very similar to what you’ve already experienced in the hospital or outpatient clinic. Your doctor will prescribe the dose of your medication. A nurse will come to your home on the scheduled day, prepare the medication, perform the infusion, and monitor your vital signs throughout. You’ll still receive all of the same support you’ve had from BioMarin and BioMarin Patient and Physician Support (BPPS) to make sure that both the infusion appointments and reimbursements go smoothly.

The most important difference with home infusion is that you will receive the medication directly from a specialty pharmacy, and you will need to store it in your refrigerator until shortly before your infusion.

Increasing your comfort with home infusion

Your doctor has determined that you can safely receive your NAGLAZYME treatments at home. With home infusion, you’ll save time and avoid the hassle of traveling to an outpatient clinic each week. You’ll also receive your medication in the comfort of familiar surroundings.

This guide will help you make the transition to home infusion.

Home infusion is not an option for every person with MPS VI. 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. As always, the goal is to provide you with safe, effective treatment with as little disruption to your life as possible.
How will I get NAGLAZYME and the infusion supplies?

Both the supplies you will need and the NAGLAZYME itself will be delivered directly to your home through a specialty pharmacy via FedEx or another delivery service. Prepare ahead for your first shipment of supplies (tubing, equipment, and saline). Select a cool, dry place with enough shelf space to store several boxes. 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.

You should also prepare an area in your refrigerator for your medication. As soon as you receive your shipment of NAGLAZYME, store it in your refrigerator. Do not freeze it or heat it in any way.

Taking care of NAGLAZYME

- Store the medicine in your refrigerator as soon as you receive it.
- Never freeze the medicine. Do not heat the vials with hot water or 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 use the medicine, put it back in the refrigerator. Do not use medicine that has been left at room temperature for more than 24 hours.

If your medicine has been heated, frozen, left unrefrigerated too long, or damaged in any way, contact your physician for help and instructions.

Protecting NAGLAZYME 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:

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 BPPS 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 of the refrigerator when you take the medication out.
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: If NAGLAZYME remains unrefrigerated for 24 hours or longer, or if the refrigerator’s internal temperature is 77°F or higher, call BPPS at 1-866-906-6100.
What should I watch out for during and after the infusion?

NAGLAZYME is generally well tolerated, but as is the case with any infusion, some patients may experience 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.

Using these few precautions, you can enjoy the improved convenience and comfort of home infusion with NAGLAZYME.

For answers to other questions, ask your home infusion nurse or call your doctor.

What will happen during the infusion?

1. Before the infusion, your medicine will need to warm up to room temperature. You should remove it from the refrigerator 30 to 60 minutes before your home infusion care nurse is scheduled to arrive. Be careful not to shake the vial(s).

2. Upon arrival, your nurse will check your vital signs to make sure you don’t have a fever or infection. 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. Be sure to tell your nurse of any illnesses or changes in your health status. They may affect whether you should delay your weekly infusion.

3. Your nurse will then start an IV, prepare the NAGLAZYME, 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. Quiet activities such as reading, playing cards or computer games, and doing paperwork are ideal. 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 delayed allergic reactions.

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Please see accompanying full Prescribing Information or visit www.NAGLAZYME.com.

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