

ELAPRASE INFUSIONS: WHAT TO EXPECT

What is ELAPRASE[®] (Idursulfase)?

ELAPRASE is a prescription medicine for patients with Hunter syndrome.

ELAPRASE has been shown to improve walking ability in patients 5 yrs and older.

In patients 16 months to 5 yrs old, ELAPRASE did not show improvement in disease-related symptoms or long term clinical result; however, treatment with ELAPRASE has reduced spleen size similarly to patients 5 yrs and older.

It is not known if ELAPRASE is safe and effective in children under 16 months old.

IMPORTANT SAFETY INFORMATION

RISK OF SERIOUS ALLERGIC REACTIONS:

Some patients have experienced serious allergic reactions (including life-threatening anaphylactic reactions) during and up to 24 hours after treatment, regardless of how long they were taking ELAPRASE. Anaphylactic reactions are immediate and include breathing problems, low oxygen levels, low blood pressure, hives and/or swelling of the throat or tongue. If a patient (you or your child) has experienced an anaphylactic reaction, the patient may require an extended period of observation by the patient's healthcare team. If you or your child has breathing problems, a fever, or a respiratory illness, you or your child may be at risk of life-threatening worsening of those conditions due to allergic reactions from ELAPRASE. Your healthcare team should be advised of those conditions before treatment with ELAPRASE because the information may affect the timing of ELAPRASE treatment.

**For Patients
and Caregivers**

Please see additional Important Safety Information on [pages 14-15](#) and [CLICK HERE](#) to see accompanying Full Prescribing Information, including Boxed WARNING for Risk of Anaphylaxis.

**For more information,
please visit www.ELAPRASE.com**

CONTENTS



Introduction	04
Preparing for the first infusion	05
Infusion activity sheet	10
Preparing for weekly infusions	11
Home infusions	12
Storing and handling ELAPRASE vials	13
Important Safety Information	14



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INTRODUCTION

This guide is designed to help patients and their caregivers to prepare for their ELAPRASE infusions. ELAPRASE (Idursulfase) is a prescription medicine for patients with Hunter syndrome. ELAPRASE has been shown to improve walking ability in patients 5 yrs and older. In patients 16 months to 5 yrs old, ELAPRASE did not show improvement in disease-related symptoms or long-term clinical result; however, treatment with ELAPRASE has reduced spleen size similarly to patients 5 yrs and older. It is not known if ELAPRASE is safe and effective in children under 16 months old. ELAPRASE is administered once a week via an intravenous (IV) infusion.

You or someone you are caring for will receive ELAPRASE infusions in a treatment center. This guide has some practical advice about preparing for the first infusion and a section for all following infusions.

This guide also includes information on the storage and handling of ELAPRASE vials. Where permitted by local healthcare and reimbursement policy, physicians may allow patients who tolerate ELAPRASE infusions well over several months to receive infusions at home, administered by a healthcare professional. Speak to your doctor if you would like more information about home infusions.



IMPORTANT SAFETY INFORMATION (CONTINUED)

You or your child should be closely watched during and after ELAPRASE treatment and you should confirm with your healthcare team in advance of treatment that it is prepared to manage serious allergic reactions, including anaphylactic reactions. **Tell your healthcare team immediately if any signs of an allergic reaction happen.** Those signs may include breathing problems, low blood pressure, rash, hives, itching, flushing, fever and/or headache.

When serious allergic reactions happened during clinical trials, later ELAPRASE treatments were managed with allergy-controlling drugs before or during treatment, a slower rate of ELAPRASE treatment, and/or early discontinuation of treatment.

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PREPARING FOR THE FIRST INFUSION

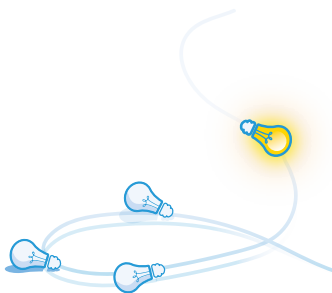
As with any new experience, the first infusion of ELAPRASE may be unfamiliar. This section has been designed to let you know what to expect from the first infusion of ELAPRASE and to help you prepare for this new experience.

If at any point you have questions or concerns about the treatment, please speak to a healthcare professional and they will be able to help you.

Caregivers: it's important to be honest with your child when describing the process so that they can feel prepared. Try to think about the words that you use when talking to your child about infusions. We often already associate certain words with feelings; you could try using "discomfort" instead of "pain" or "hurt" and try to use positive language. When checking how your child feels, try not to use leading questions, which may prompt a specific response, and instead try asking more open questions: rather than "Does this hurt?" you could try "How does that feel?" or even try a number scale (e.g., from 1 to 10) of discomfort to gauge how your child is feeling.

What will happen at the first infusion?

The first infusion will take place in a hospital or infusion center. When you arrive at the infusion center for the first infusion of ELAPRASE, there may be others also receiving treatment. Different centers have different guidelines on interactions between patients and activities that are appropriate during infusions. Check with your center before you arrive to see which activities are suitable.



IMPORTANT SAFETY INFORMATION (CONTINUED)

Children with serious genetic mutations may be at risk for allergic reactions, serious side effects and antibody development. In a clinical study of children 7 years and younger, patients with certain types of genetic mutations experienced a higher number of allergic reactions, serious side effects, and development of an immune response to treatment. This immune response may interfere with the effectiveness of ELAPRASE. Talk to your healthcare team about whether you or your child may be at risk.

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PREPARING FOR THE FIRST INFUSION (CONTINUED)

The dose of ELAPRASE given depends on weight, so the person receiving treatment will need to be weighed to determine the best dose of ELAPRASE.

ELAPRASE is administered via an IV infusion, which means that ELAPRASE is delivered to your blood stream through your arm using a needle and tube. This may feel slightly uncomfortable and if you are concerned about this step, please inform your nurse and they will be able to support you.

Initial infusions may be administered over a period of approximately 3 hours; this time may be gradually reduced to 1 hour if no hypersensitivity reactions occur. There may also be additional time needed for preparation and observation.



Speak to your healthcare provider for more details about what to expect and to help you plan ahead.

Caregivers: you know your child best, so you and the nurse can work together as a team to help make your child feel most comfortable.



IMPORTANT SAFETY INFORMATION (CONTINUED)

If you or your child has breathing problems, other respiratory illness, heart problems, or susceptibility to fluid overload, you or your child may be at higher risk of fluid overload during ELAPRASE treatment. Your healthcare team should be advised of those problems before treatment and you should confirm with your healthcare team in advance of treatment that it is appropriately trained to watch for signs of fluid overload and provide the necessary medical support. Patients at risk for fluid overload may require longer observation time.

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PREPARING FOR THE FIRST INFUSION (CONTINUED)

How can we prepare for the first infusion?

Here are some practical things that can be done to prepare for the first infusion:



Plan how to travel to the infusion center in advance to ensure you arrive in plenty of time.



Drink lots of water to stay hydrated.



Each infusion center has its own guidelines about what can be done during therapy. You might be able to engage in a quiet activity such as reading a book, watching TV, or doing homework. Ask your treatment center before you arrive to find out which activities are suitable.



Ensure you and the person you are caring for has a good night's sleep before the first infusion, as this will help them feel less anxious the next day.



Eat a good balanced meal before the infusion.



Make sure the person receiving treatment wears loose and comfortable clothing so they feel relaxed and the infusion nurse has easy access to their arm.



Plan a treat for after the first infusion. Any new experience can make you and the person you are caring for feel uneasy or nervous, so it might be nice to have something to look forward to after completing the first infusion.



Ask your doctor any questions you have before the first infusion —they are there to help!

Caregivers: it's also important for you to rest too as children will likely pick up on any feelings of nervousness. Children are often very sensitive to how you are feeling, so try to remain calm and relaxed to help them feel the same.

IMPORTANT SAFETY INFORMATION (CONTINUED)

What are possible side effects of ELAPRASE? The most common side effects of ELAPRASE include:

- In patients aged 5 and older:
 - Headache
 - Itching
 - Muscle and bone pain
 - Hives
 - Diarrhea
 - Cough
- In patients aged 7 years or younger:
 - Fever
 - Rash
 - Vomiting
 - Hives

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PREPARING FOR THE FIRST INFUSION (CONTINUED)

Here are some questions that you may like to ask your infusion nurse or doctor before the first infusion:

- What can we do during an infusion?
- How do we go to the bathroom during an infusion?
- Can we eat or drink during an infusion?

Here is some space for you to note down any other questions you may like to ask your infusion nurse or doctor:

IMPORTANT SAFETY INFORMATION (CONTINUED)

The most common side effects needing medical attention were allergic reactions, and included rash, hives, itching, flushing, fever, and headache. **Tell your healthcare team immediately if any signs of an allergic reaction happen.** These are not all the possible side effects of ELAPRASE.

Please see additional Important Safety Information on [pages 14-15](#) and [CLICK HERE](#) to see accompanying Full Prescribing Information, including Boxed WARNING for Risk of Anaphylaxis.

PREPARING FOR THE FIRST INFUSION (CONTINUED)

What should we bring to the first infusion?

Here are some practical things that can be done to prepare for the first infusion:



Wear or bring layers of clothing or even a blanket in case you feel too hot or cold, so you can make yourself or the person you're caring for comfortable.

Caregivers: you could pick a new special and cozy blanket for your child's infusions



You may be able to bring light snacks to eat during an infusion. Ask the treatment center before arrival.



A water bottle to help stay hydrated.



Your selected activity, whether that's a book, electronic device, or homework. You may also want to bring a charger for your electronic device.



Children may want to bring a cuddly toy with them as an infusion companion.

After the first infusion

You and the person you are caring for may feel tired after your first infusion, so it's important to rest.

After completing the first infusion of ELAPRASE, speak with your doctor about next steps to ensure you or the person you are caring for receive treatment with ELAPRASE every week or as prescribed by your healthcare provider.

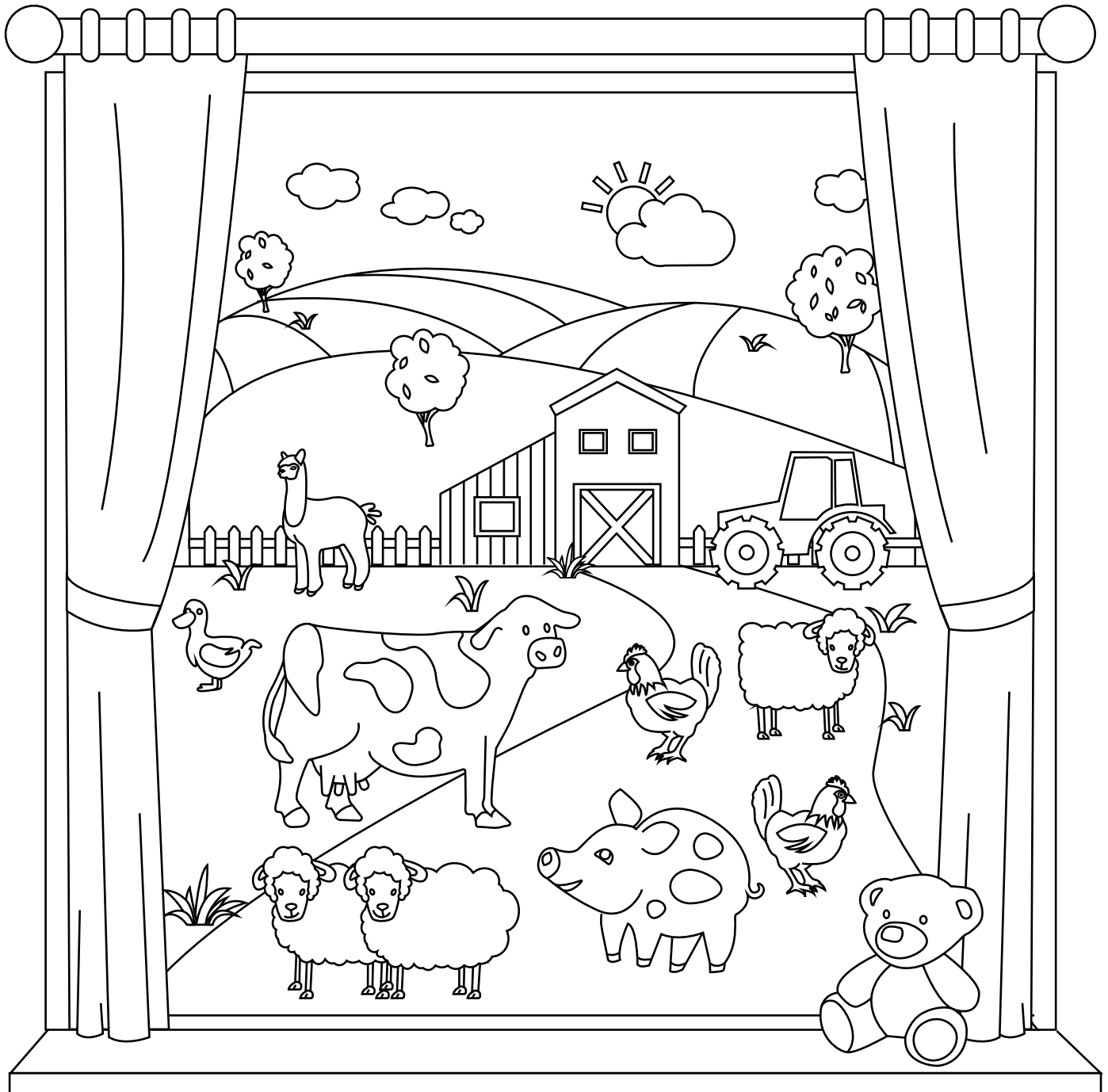
IMPORTANT SAFETY INFORMATION (CONTINUED)

RISK OF SERIOUS ALLERGIC REACTIONS:

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INFUSION ACTIVITY SHEET



Please see Important Safety Information on [pages 14-15](#) and [CLICK HERE](#) to see accompanying Full Prescribing Information, including Boxed WARNING for Risk of Anaphylaxis.

PREPARING FOR WEEKLY INFUSIONS

It is important to continue to take your treatment as prescribed by your doctor.

Use the appointment tracker below and infusion checklist to help as you start your infusion routine.

INFUSION	DATE	TIME	LOCATION	NOTES (e.g., length of infusion)
1				
2				
3				
4				
5				
6				
7				
8				

Caregivers: infusions can make children nervous but there are things you can try to help them feel more comfortable. You could try using a sticker chart for each infusion completed to reward them for their bravery. You could even give a special treat after a certain number of collected stickers or completed infusions so that they have something to look forward to. You know what works best for your child, so trust your instincts.

If your child is still finding infusions difficult, you could try talking through the whole infusion process together to try and determine exactly what is causing your child's anxiety. Once you have identified the main issue that your child is not comfortable with, you can speak to your healthcare team to find ways to make your child feel more at ease.

My infusion checklist:

Use the infusion checklist to help remind you of things to bring to your infusion. There is space for you to write down any other things you might like to bring.

- | | |
|---|-----------|
| 1. Relevant medical records | 6. _____ |
| 2. Layers of light, loose-fitting, and comfortable clothing | 7. _____ |
| 3. Blanket | 8. _____ |
| 4. Water bottle | 9. _____ |
| 5. My selected activity (book, electronic device, homework, toy, or coloring pencils) | 10. _____ |

IMPORTANT SAFETY INFORMATION (CONTINUED)

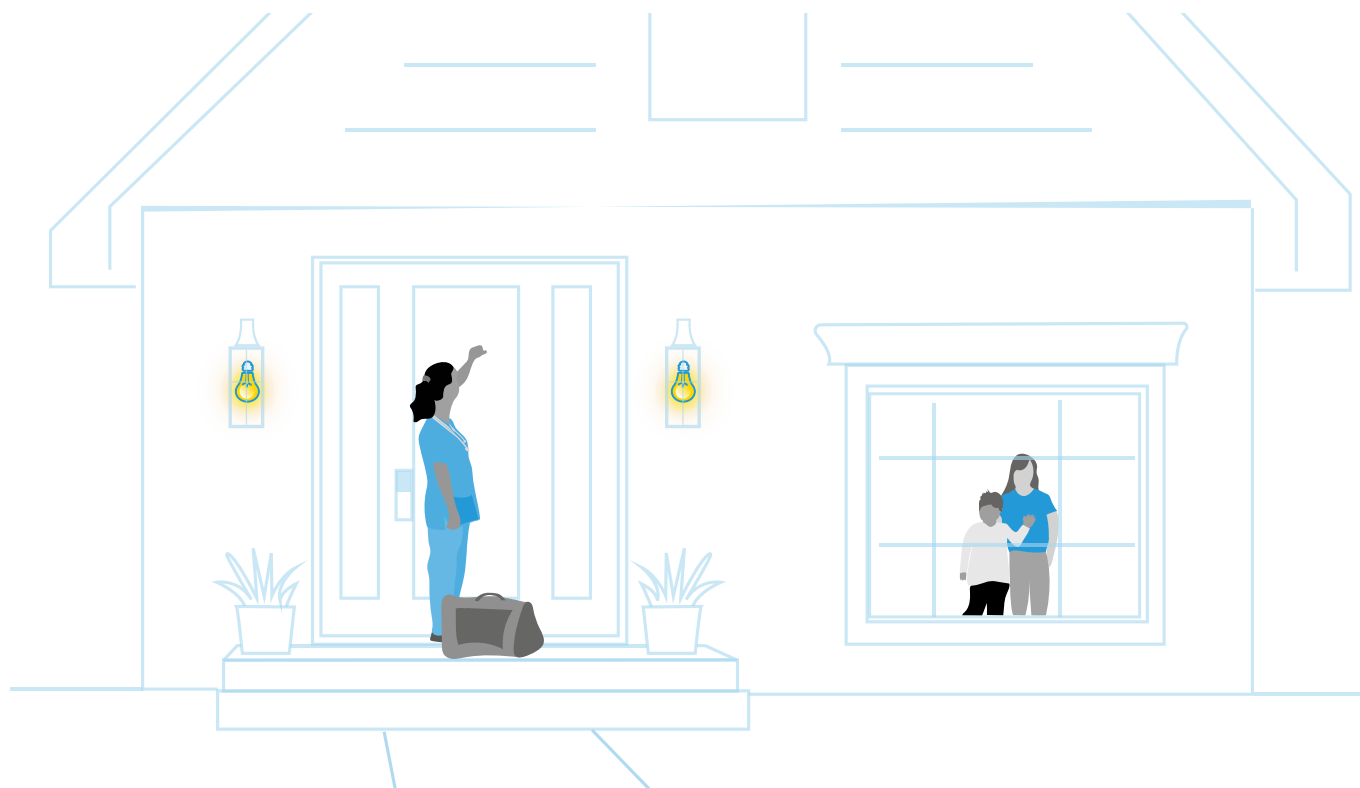
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HOME INFUSIONS

Where permitted by local healthcare and reimbursement policy, physicians may decide whether patients who tolerate ELAPRASE infusions well over several months can receive infusions at home under the supervision of a healthcare professional; appropriate emergency medical services should be readily available if needed. **Please speak to your doctor if you would like to learn more about home infusions and if they are an option for you or the person you are caring for.**

For patients enrolled with Takeda Patient Support, after your doctor has approved home infusions, contact our support specialists as they will be able to help you in setting up the home infusions. They will liaise with the infusion center and your specialty pharmacy to make all the necessary arrangements for your home infusions. If you have any questions about setting up home infusions, please ask our support specialists and they will be able to guide you.



For more information about home infusions and Takeda Patient Support, visit: www.ELAPRASE.com/getting-started-support

IMPORTANT SAFETY INFORMATION (CONTINUED)

When serious allergic reactions happened during clinical trials, later ELAPRASE treatments were managed with allergy-controlling drugs before or during treatment, a slower rate of ELAPRASE treatment, and/or early discontinuation of treatment.

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STORING AND HANDLING ELAPRASE VIALS

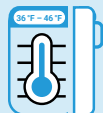
Introduction

This guide is for people who are starting home infusions with ELAPRASE following approval from their doctor. This guide has been designed to inform you how ELAPRASE vials should be stored and handled correctly.

ELAPRASE is supplied as a sterile injection in a 5 mL Type I glass vial and each carton contains a single vial. ELAPRASE vials are single use.



Pick up the vials of ELAPRASE carefully. Do not shake ELAPRASE.



Store the vials of ELAPRASE in a refrigerator at 36°F to 46°F (2°C to 8°C) to keep them cold and away from light.



When it is time to use ELAPRASE, your healthcare professional administering ELAPRASE will check the expiration date on the vial. ELAPRASE should not be used after the expiration date on the vial.



Prior to infusion, the appropriate dose of ELAPRASE will be diluted in a 100 mL bag of 0.9% Sodium Chloride Injection, USP for intravenous infusion by a healthcare professional. The diluted ELAPRASE is then infused at room temperature.



If immediate use is not possible, the diluted solution should be stored refrigerated at 36°F to 46°F (2°C to 8°C) for up to 24 hours. Any unused product or waste material should be discarded and disposed of in accordance with local requirements.

For more information on storing and handling ELAPRASE vials, please [CLICK HERE](#) to refer to the Full Prescribing Information, and speak to your doctor.

IMPORTANT SAFETY INFORMATION (CONTINUED)

Children with serious genetic mutations may be at risk for allergic reactions, serious side effects and antibody development. In a clinical study of children 7 years and younger, patients with certain types of genetic mutations experienced a higher number of allergic reactions, serious side effects, and development of an immune response to treatment. This immune response may interfere with the effectiveness of ELAPRASE. Talk to your healthcare team about whether you or your child may be at risk.

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What is ELAPRASE[®] (Idursulfase)?

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In patients 16 months to 5 yrs old, ELAPRASE did not show improvement in disease-related symptoms or long term clinical result; however, treatment with ELAPRASE has reduced spleen size similarly to patients 5 yrs and older.

It is not known if ELAPRASE is safe and effective in children under 16 months old.

Important Safety Information

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Important Safety Information (continued)

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What are possible side effects of ELAPRASE?

The most common side effects of ELAPRASE include:

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 - Muscle and bone pain
 - Hives
 - Diarrhea
 - Cough
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 - Rash
 - Vomiting
 - Hives

The most common side effects needing medical attention were allergic reactions, and included rash, hives, itching, flushing, fever, and headache. **Tell your healthcare team immediately if any signs of an allergic reaction happen.** These are not all the possible side effects of ELAPRASE.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

For additional safety information, please **[CLICK HERE](#)** for the Full Prescribing Information, including **Boxed WARNING for Risk of Anaphylaxis**, and discuss with your doctor.

For more information, please visit
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1. Wraith JE *et al.* *Eur J Pediatr.* 2008; 167: 267-277. 2. ELAPRASE Prescribing Information.