

INJECTION GUIDE

for parents/caregivers



40 mg/ml solution for injection

18 mg/0.45 ml

28 mg/0.7 ml

40 mg/1 ml

100 mg/ml solution for injection

80 mg/0.8 ml

asfotase alfa



This medicinal product is subject to additional monitoring

Introduction

This Injection Guide is for the parents and caregivers of patients with paediatric-onset hypophosphatasia who would benefit from long-term enzyme replacement therapy and have been prescribed Strensiq®.

The guide gives detailed instructions, with clear diagrams, on how to choose the injection site and how to carry out and record the injection of your infant or child.

This guide is designed as a support document to the training given by your healthcare professional.

Please contact your healthcare professional if you have any questions.

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Important Information

Do not inject yourself with Strensiq® until you have had proper training from a healthcare professional.

- Before starting, check the expiry date of Strensiq®. DO NOT use it if it has expired.
- Always use a new vial and carefully examine the liquid to make sure it is clear, colourless to slightly yellow and contains no particles.
- If you suspect that the liquid in your vial has been contaminated, DO NOT use it, select another vial and contact your healthcare professional for advice.
- Always use the specific dose (in number of milligrams) that has been prescribed by your doctor.
- Vials are for single use and should only be punctured once. Used vials must be discarded at the end of the injection.
- Syringes and needles are for single use only and should be handled with care. Keep them out of reach from children and dispose of them safely in a special sharps container. Needles and syringes must not be mixed with normal household rubbish.
- Store your Strensiq® vials in the refrigerator at a temperature between 2°C and 8°C. DO NOT put Strensiq® in or near the freezer compartment and never inject Strensiq® that you know, or suspect, has been frozen.
- Store your Strensiq® in the original package in order to protect the medication from light.

For detailed information on Strensiq® please refer to the Patient Information Leaflet which you can find in the Strensiq® package.

Warnings and Precautions

- Some patients receiving asfotase alfa have had life threatening allergic reactions similar to anaphylaxis requiring medical treatment. Patients who experienced anaphylaxis-like symptoms had difficulty breathing, choking sensation, nausea, swelling around the eyes, and/or dizziness. The reactions occurred within minutes after injecting asfotase alfa in patients, and can occur in patients who have been taking asfotase alfa for any period of time (e.g. more than one year). If you experience any of these symptoms, discontinue Strensiq® and seek medical help immediately.
- If you experience an anaphylactic reaction, or an event with similar symptoms, your doctor will discuss with you the next steps and the possibility to restart Strensiq® under medical supervision. Always follow the instructions provided by your doctor.
- Fatty lumps or decreased fatty tissue on the surface of the skin (localised lipodystrophy) has been reported at injection sites after several months in patients using Strensiq®. Review the ‘How is Strensiq® Administered?’ section carefully to know the injection recommendations. It is important to rotate the injection from among the following sites to reduce the risk of lipodystrophy: buttocks, abdomen, thigh, or arms (deltoids).
- If you are treated with Strensiq®, you may experience a reaction at the injection site (e.g. pain, nodule, rash, discoloration) during the injection of the medicine or during the hours following the injection. If you experience any severe reaction at the injection site, tell your doctor immediately.

What is Strensiq®?

Strensiq® is a medication designed as a long-term enzyme replacement therapy in patients with paediatric-onset hypophosphatasia. It is administered subcutaneously, under the skin.

How does my infant or child get started on Strensiq® therapy?

Strensiq® must be prescribed by your infant or child's doctor.

Your healthcare professional will advise you on the range of educational materials available to help with the therapy. This injection guide can help support you when learning how to inject your infant or child.

What dose of Strensiq® is used?

Once Strensiq® has been prescribed for your infant or child you will need to administer the drug according to the specific dose (in number of milligrams) that has been prescribed by your doctor.

The dose you administer is based on your infant or child's body weight. The correct dose will be calculated by your doctor and consists of a total of 6 mg per kg of body weight per week of Strensiq®, administered by subcutaneous injection. Your child may receive either an injection of 1 mg/kg of Strensiq® six times per week or an injection of 2 mg/kg of Strensiq® three times per week depending on the recommendation of your doctor.

For example, if the body weight of your infant or child is 6 kg, he or she will receive 6 mg six times per week or 12 mg three times per week.

The maximum volume of medication per injection should not exceed 1 ml. If more than 1 ml is required, multiple injections may be administered, one after another.

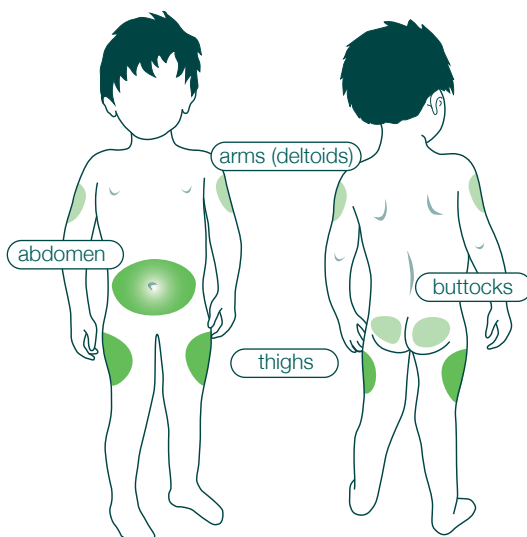
How is Strensiq® administered?

Strensiq® is administered either three or six times per week as a subcutaneous injection into the fatty layer just below the skin. This is called the subcutaneous layer and it is just above the muscle.

There are specific areas of the body suitable for giving subcutaneous injections to infants and children, shown on the diagram below. As a general rule, these areas are those with a substantial amount of fat below the skin. The thighs and abdomen tend to be the most suitable for infants and children.

When injecting regularly, the position on the body where the injections are given should be rotated, as this may help reduce pain and irritation.

As your infant or child grows, other areas may become suitable, such as the arms and buttocks. Please discuss with your healthcare professional the best sites for your infant or child.



Please remember, this guide is designed as a support document to the training given by your **healthcare professional**.

How to inject Strensiq®

Before you begin, take your vial(s) out of the refrigerator 15 to 30 minutes before injecting to allow the liquid to reach room temperature (around 23°C to 27°C). Do not warm Strensiq® in any other way (for example, do not warm it in a microwave or in hot water). Strensiq® should be administered within 3 hours. For more information about the storage of Strensiq®, see page 14.

Please remember your vials are for single use and should only be punctured once. Used vials must be discarded at the end of the injection.

Wash your hands thoroughly with soap and water.

Place all the items you will need on a clean area where they will not be disturbed. These should include:

- Your vial(s) for injection
- Your syringe(s)
- One larger bore syringe needle (e.g. 25G)
- One smaller bore syringe needle (e.g. 27 or 29G, with length appropriate for subcutaneous injection)
- Alcohol swabs (if required)
- Gauze or cotton wool
- Your sharps container*
- Plaster or adhesive bandage (if required)
- Injection diary (or other recording means)

Choose the injection site (see diagram on page 7) and prepare it according to the advice given by your healthcare professional.

** Your healthcare professional will advise on how you can obtain a sharps container and will also advise you on the disposal procedure.*

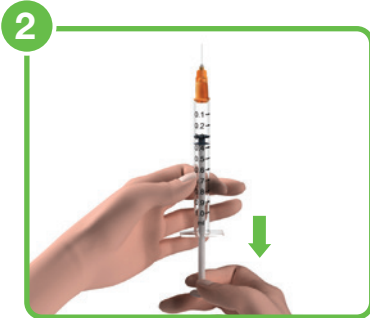
How to inject Strensiq®

Step 1: Preparing the Strensiq® dose



1. Always use a new vial and carefully examine the liquid to make sure it is clear, colourless to slightly yellow, and contains no particles.

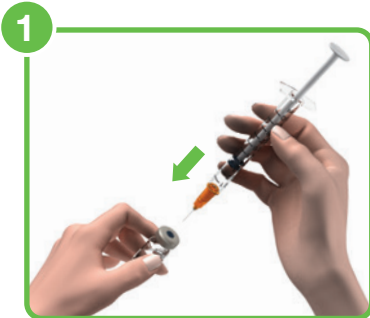
Remove the protective cap (shown as red on the image) from the vial to reveal the sterile rubber seal.



2. Place a larger bore needle (e.g. 25G) on the empty syringe and with the protective cap on, push down and turn clockwise the needle onto the syringe until it is tight. Remove the plastic cap covering the syringe needle and dispose of it in your sharps container. Pay attention to not hurt yourself with the needle.

Pull the plunger back to draw air into the syringe equal to your dose.

Step 2: Withdrawing Strensiq® solution from the vial



1. Holding the syringe and vial, at a 45° angle, insert the needle through the sterile rubber seal and into the vial.

Push the plunger in completely to inject the air into the vial.

How to inject Strensiq®

2



2. Invert the vial and syringe.

With the needle in the solution, pull the plunger to withdraw the correct dose into the syringe.

3



3. Before removing the needle from the vial, check the syringe for air bubbles.

In the event that bubbles appear in the syringe, hold the syringe with the needle pointing upwards and gently tap the side of the syringe until the bubbles rise to the top.

4



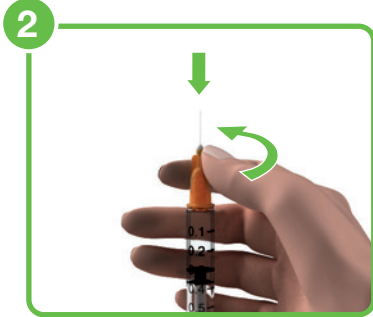
4. Once all the bubbles are at the top of the syringe, gently push on the plunger to force the bubbles out of the syringe and back into the vial.

After removing the bubbles, check the dose of medication in the syringe to be sure you have drawn up the correct amount. You may need to use several vials to withdraw the complete amount needed to reach the correct dose.

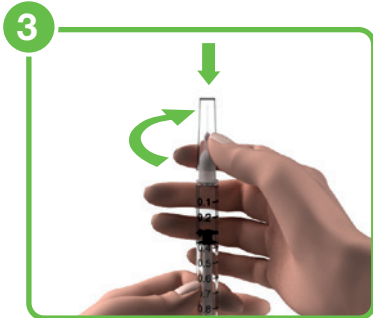
How to inject Strensiq®

Step 3: Placing the needle for injection on the syringe

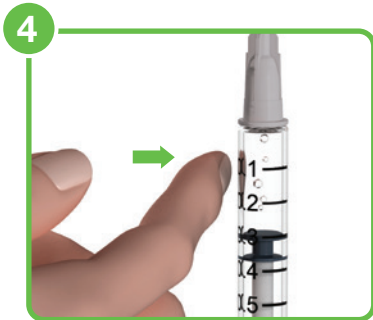
1. Remove the needle from the vial and place back the protective cap over the needle, as instructed by your healthcare professional.



2. Remove the recapped larger bore needle pushing down and turning counterclockwise and dispose of it in your sharps container.



3. Place a smaller bore needle (e.g. 27 or 29G) on the filled syringe and with the protective cap on, push down and turn clockwise the needle onto the syringe until it is tight. Pull the cap straight off the needle and dispose of it in your sharps container.



4. Hold the syringe with the needle pointing up and tap the barrel of the syringe with your finger to remove any air bubbles. After removing the bubbles, ensure the volume of medication in the syringe you have drawn up is the correct amount. You are now ready to inject the correct dose.

How to inject Strensiq®

Step 4: Injecting Strensiq®

Please refer to Section “How Strensiq® is administered” on page 7 for determining the most suitable injection site for your infant or child (thighs, abdomen, arms (deltoids), buttocks).

1



1. Hold your infant or child firmly in a comfortable position.

Your healthcare professional will advise you on the best position to hold your infant or child for the chosen injection site.

2



2. Gently pinch the skin of the chosen injection area (in this case the infant or child's thigh) between your thumb and index finger.

3



3. Holding the syringe like a pencil or a dart, insert the needle into the raised skin so it is at an angle of between 45° and 90° to the skin surface. For small children who have little subcutaneous fat or thin skin, a 45° angle may be preferable.

How to inject Strensiq®



4. While continuing to hold the skin, push the syringe plunger to inject the medication slowly and steadily all the way in.

Remove the needle, release the skin fold, place the syringe in the sharps container, and gently place a piece of cotton wool or gauze over the injection site for a few seconds. This will help seal the punctured tissue and prevent any leakage. Do not rub the injection site after injection.



5. Dispose your syringe, used vial and needle into your sharps container. Your doctor, pharmacist or nurse will advise you on how you can obtain a sharps container.

6. If you need a second injection for your prescribed dose, get another Strensiq® vial and repeat all of the previous steps.

Your vials are for single use and should only be punctured once.

Place a small plaster or adhesive bandage over the injection site, if necessary.



7. Record all details of the injection in your injection diary or using other means.

It is recommended that you note:

- where you injected
- the dose injected
- any injection reactions

Storing and Travelling with Strensiq®

Storing your Strensiq® vials

- Store your child's Strensiq® vials in the refrigerator at a temperature between 2°C and 8°C. DO NOT put Strensiq® in or near the freezer compartment and never inject Strensiq® that you know, or suspect, has been frozen.
- Store your child's Strensiq® in the original package in order to protect the medication from light.
- Before starting, check the expiry date of Strensiq® which you will find on the vial label. DO NOT use if it has expired.

Travelling with your Strensiq® vials

- Calculate how many vials you will need for your trip. Take some extra in case you are away for longer than expected.
- Remember to take your sharps container and some extra syringes. It is recommended that you carry your supplies as hand luggage.
- Check to ensure that your travel destination has refrigeration that you can use.
- It is recommended that you carry your vials in a cool bag when you travel. The bag should contain cool blocks to help keep your vials at the correct temperature.
- Keep the cool blocks in your freezer so that they are ready for use when you travel. Ensure that the frozen cool blocks are not in direct contact with your child's medication.

Travelling by air

- It is recommended that you carry your child's medication and supplies as hand luggage.
- It may be advisable to carry a letter from your infant or child's doctor explaining that you need to travel with your medication.

Storing and Travelling with Strensiq®

- Check with your airline, or the airports you are travelling through, for any specific rules for persons carrying injectable medication and associated accessories.
- If you have any concerns about travelling, discuss them with your healthcare professional.

Reporting of side effects

If your infant or child experiences any side effects, talk to your doctor, pharmacist or nurse. This also includes any side effects you experience that are not listed in this guide.

In order to improve the traceability of this medicine, when reporting a side effect, the product name and the batch number of the medicine taken should be clearly recorded.

By reporting side effects you can help provide more information on the safety of this medicine.

Suspected adverse reactions should be reported to the MHRA using the Yellow Card obtained electronically via: MHRA website at <http://www.mhra.gov.uk/yellowcard> or search for MHRA Yellow Card in the Google Play or Apple App Store.

Alternatively, prepaid Yellow Cards for reporting are available by writing to FREEPOST YELLOW CARD (no other address details necessary), by emailing yellowcard@mhra.gov.uk, at the back of the British National Formulary (BNF), by telephoning the Commission on Human Medicines (CHM) free phone line: **0800 731 6789**, or by downloading and printing a form from the Yellow Card section of the MHRA website.

If you are resident in the Republic of Ireland, please visit the HPRA website at <http://www.hpra.ie> with the adverse reaction report form found at <https://www.hpra.ie/homepage/about-us/report-an-issue/human-adverse-reaction-form> (Ireland)

Adverse events and product complaints should also be reported to:

Alexion Pharma UK on 0800 3213902
Ireland Freephone 1 800 936 544 or
by email to uk.adverseevents@alexion.com

United Kingdom:

<https://www.medicines.org.uk/emc/product/7991>

Ireland:

[http://www.hpra.ie/homepage/medicines/medicines-information/
find-a-medicine/results/item?pano=EU/1/15/1015/001-002&t=Strensiq](http://www.hpra.ie/homepage/medicines/medicines-information/find-a-medicine/results/item?pano=EU/1/15/1015/001-002&t=Strensiq)

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