Package leaflet: Information for the user

Elaprase 2 mg/ml concentrate for solution for infusion
idursulfase

▼This medicine is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may get. See the end of section 4 for how to report side effects.

Read all of this leaflet carefully before you start using this medicine because it contains important information for you.

- Keep this leaflet. You may need to read it again.
- If you have any further questions, ask your doctor, pharmacist or nurse.
- This medicine has been prescribed for you only. Do not pass it on to others. It may harm them, even if their signs of illness are the same as yours.
- If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in this leaflet. See section 4.

What is in this leaflet:

1. What Elaprase is and what it is used for

Elaprase is used as enzyme replacement therapy to treat children and adults with Hunter syndrome (Mucopolysaccharidosis II) when the level of the enzyme iduronate-2-sulfatase in the body is lower than normal, helping improve the symptoms of the disease. If you suffer from Hunter syndrome, a carbohydrate called glycosaminoglycan which is normally broken down by your body, is not broken down and slowly accumulates in various organs in your body. This causes cells to function abnormally, thereby causing problems for various organs in your body which can lead to tissue destruction and organ malfunction and failure. Typical organs where glycosaminoglycan accumulates are spleen, liver, lungs, heart, and connective tissue. In some patients glycosaminoglycan accumulates also in the brain. Elaprase contains an active substance called idursulfase which works by acting as a replacement for the enzyme that is at a low level, thereby breaking down this carbohydrate in affected cells.

Enzyme replacement therapy is usually administered as a long-term treatment.

2. What you need to know before you use Elaprase

Do not use Elaprase

If you have experienced severe or potentially life-threatening allergic-type reactions to idursulfase or any of the other ingredients of this medicine (listed in section 6) and these cannot be controlled with appropriate medical treatment.

Warnings and precautions

Talk to your doctor or nurse before using this medicine.

If you are treated with Elaprase you may experience side effects during or following an infusion (see section 4 Possible side effects). The most common symptoms are itching, rash, hives, fever, headache, increased blood pressure, and flushing (redness). Most of the time you can still be given this medicine even if these symptoms occur. If you experience an allergic side effect following administration of this medicine, you should contact your doctor immediately. You may be given additional medicines such as antihistamines and corticosteroids to treat or help prevent allergic-type reactions.

If severe allergic reactions occur, your doctor will stop the infusion immediately, and start giving you suitable treatment. You may need to stay in hospital.

The nature of your genotype (a genetic make up of all active genes in human cells, which determines one's specific, individual characteristics) may influence your therapeutic response to this medicine, as well as your risk of developing antibodies and infusion-related side effects. In individual cases, so-called 'neutralising antibodies' may develop, which may diminish activity of Elaprase and your response to treatment. The longer term effects of antibody development on response to treatment have not been established. Please consult your doctor for additional information.

Keeping a record

In order to improve the traceability of biological medicinal products, the name and batch number of the administered product should be clearly recorded by your healthcare professional. Speak with your healthcare professional if you are not sure.

Other medicines and Elaprase

There is no known interaction of this medicine with other medicines.

Tell your doctor, pharmacist or nurse if you are taking, have recently taken or might take any other medicines.

Pregnancy and breast-feeding

If you are pregnant or breast-feeding, think you may be pregnant or are planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.

Driving and using machines

This medicine has no or negligible influence on the ability to drive and use machines.

Elaprase contains sodium

This medicine contains 11.1 mg sodium (main component of cooking/table salt) in each vial. This is equivalent to 0.6% of the recommended maximum daily dietary intake of sodium for an adult.

3. How to use Elaprase

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

This medicine will be given to you under the supervision of a doctor or nurse who is knowledgeable in the treatment of Hunter syndrome or other inherited metabolic disorders.

The recommended dose is an infusion of 0.5 mg (half a milligram) for every kg you weigh

Elaprase has to be diluted in sodium chloride 9 mg/ml (0.9%) solution for infusion before use. After dilution this medicine is given through a vein (drip feed). The infusion will normally last for 1 to 3 hours and will be given every week.

Use in children and adolescents

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

If you use more Elaprase than you should

Consult your doctor if you have an overdose of this medication.

If you forget to use Elaprase

If you have missed an Elaprase infusion, please contact your doctor.

If you have any further questions on the use of this medicine, ask your doctor or nurse.

4. Possible side effects

Like all medicines, this medicine can cause side effects, although not everybody gets them.

Most side effects are mild to moderate and associated with the infusion, however some side effects may be serious. Over time the number of these infusion-associated reactions decreases.

If you have problems breathing, with or without bluish skin, **tell your doctor straight away and seek immediate medical assistance**.

Very common side effects (may affect more than 1 in 10 people) are:
- Headache
- Flushing (redness)
- Shortness of breath, wheezing
- Abdominal pain, nausea, vomiting, frequent and/or loose stools
- Chest pain
- Hives, rash, itching, redness of the skin
- Fever
- Infusion-related reaction (see section entitled “Warnings and precautions”)

Common side effects (may affect up to 1 in 10 people) are:
- Dizziness, tremor
- Rapid heartbeat, irregular heartbeat, bluish skin
- Increased blood pressure, decreased blood pressure
- Difficulty breathing, cough, low oxygen levels in your blood
- Swollen tongue, indigestion
- Pain in the joints
- Infusion-site swelling, swelling of the extremities, facial swelling

Uncommon side effects (may affect up to 1 in 100 people) are:
- Quickened breathing

Side effects for which frequency is not known (cannot be estimated from available data) are:
- Serious allergic reactions

Reporting of side effects

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in this leaflet. You can also report side effects directly via the national reporting system (see below). By reporting side effects you can help provide more information on the safety of this medicine.

United Kingdom:
Yellow Card Scheme
Website: www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store

Ireland:
HPRA Pharmacovigilance
Website: www.hpra.ie

5. How to store Elaprase

Keep this medicine out of the sight and reach of children.

Do not use this medicine after the expiry date which is stated on the label and carton after EXP. The expiry date refers to the last day of that month.
Store in a refrigerator (2°C – 8°C)
Do not freeze

Do not use this medicine if you notice that there is discolouration or presence of foreign particles.

Do not throw away any medicines via wastewater or household waste. Ask your pharmacist how to throw away medicines you no longer use. These measures will help protect the environment.

6. Contents of the pack and other information

What Elaprase contains

The active substance is idursulfase, which is a form of the human enzyme iduronate-2-sulfatase. Idursulfase is produced in a human cell line by genetic engineering technology (it involves the introduction of genetic information into human cells in the lab, which will then produce the desired product).

Each vial of Elaprase contains 6 mg of idursulfase. Each ml contains 2 mg of idursulfase.

The other ingredients are Polysorbate 20, sodium chloride, dibasic sodium phosphate, heptahydrate, monobasic sodium phosphate, monohydrate and water for injections.

What Elaprase looks like and contents of the pack

This medicine is a concentrate for solution for infusion. It is supplied in a glass vial as a clear to slightly opalescent, colourless solution.

Each vial contains 3 ml of concentrate for solution for infusion.

Elaprase is supplied in pack sizes of 1, 4 and 10 vials per carton. Not all pack sizes may be marketed.

Marketing Authorisation Holder

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This leaflet was last revised in 05/2020.

This medicine has been authorised under "exceptional circumstances". This means that because of the rarity of this disease it has been impossible to get complete information on this medicine.

The European Medicines Agency will review any new information on the medicine every year and this leaflet will be updated as necessary.

Other sources of information

Detailed information on this medicine is available on the European Medicines Agency web site: http://www.ema.europa.eu. There are also links to other websites about rare diseases and treatments.

This leaflet is available in all EU/EEA languages on the European Medicines Agency website.

The following information is intended for healthcare professionals only:

Instructions for use, handling and disposal

1. Calculate the total dose to be administered and number of Elaprase vials needed.

2. Dilute the total volume of Elaprase concentrate for solution for infusion required in 100 ml of sodium chloride 9 mg/ml (0.9%) solution for infusion. It is recommended to deliver the total volume of the infusion using a 0.2 µm in line filter. Care must be taken to ensure the sterility of the prepared solutions since Elaprase does not contain any preservative or bacteriostatic agent; aseptic technique must be observed. Once diluted, the solution should be mixed gently, but not shaken.

3. The solution should be inspected visually for particulate matter and discolouration prior to administration. Do not shake.

4. It is recommended that administration is started as soon as possible. The chemical and physical stability of the diluted solution has been demonstrated for 8 hours at 25°C.

5. Do not infuse Elaprase concomitantly in the same intravenous line with other medicinal products.

6. For single use only. Any unused product or waste material should be disposed of in accordance with local requirements.