INSTRUCTIONS FOR USE

UCEDANE 200 MG DISPERSIBLE TABLET

For oral use.

Active ingredient: Carglumic acid

Excipient(s): Contains microcrystalline cellulose, mannitol, colloidal anhydrous silica, sodium stearyl fumarate, crospovidone type B and copovidone K 28.

▼This medicinal product 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 this ENTIRE INSTRUCTIONS FOR USE carefully before you start using this medicine, as it contains important information for you.

- ***Keep these instructions for use. You may need to read them again later.***
- ***If you have additional questions, please consult your doctor or pharmacist.***
- ***This medicine has been prescribed for you personally, do not give it to others.***
- ***When you visit your doctor or hospital during the use of this medicine, tell your doctor that you are using this medicine.***
- ***Follow exactly what is written in these instructions. Do not use a high or low dose other than the dose recommended to you for this medicine.***

In this instructions for use:

1. What is UCEDANE and what is it used for?

2. Things to consider before using UCEDANE

3. How to use UCEDANE?

4. What are the possible side effects?

5. How to store UCEDANE?

are included.

1. What is UCEDANE and what is it used for?

UCEDANE is in the form of white, biconvex and breakable (with 3 notches on both sides) dispersible tablets with "L/L/L/L" imprinted on one side. The approximate tablet dimensions are 11 mm in length and 6 mm in width. The tablet can be divided into four equal doses. It is supplied in packages containing 12 or 60 tablets in aluminium blisters on both sides, placed in a carton box.

UCEDANE can help to reduce high ammonia levels in the blood. Ammonia is particularly

toxic to the brain and, in severe cases, can lead to reduced consciousness and coma.

High ammonia levels in the blood may be caused by the following:

- N-acetylglutamate synthase deficiency, a special enzyme found in the liver. Patients with this rare disease cannot excrete accumulated nitrogen waste products from the body after protein intake. This condition persists lifelong in affected patients and therefore requires lifelong treatment.
- Isovaleric acidemia, methylmalonic acidemia, or propionic acidemia. Patients with any of these disorders require treatment during a hyperammonemic crisis.

2. Things to consider before using UCEDANE

DO NOT USE UCEDANE in the following situations

If,

- You are allergic to carglumic acid or any of the excipients in UCEDANE.
- You are pregnant.
- You are breastfeeding.

USE UCEDANE CAREFULLY in the following situations

Consult your doctor and pharmacist before taking UCEDANE.

UCEDANE treatment should be initiated under the supervision of a physician experienced in the treatment of metabolic disorders.

Before starting any long-term treatment, your doctor will test your individual response to carglumic acid.

The dose should be adjusted individually to maintain normal ammonia plasma levels.

Your doctor may give you arginine supplements or limit your protein intake.

To monitor your condition and treatment, your doctor may regularly check your liver, kidneys, heart and blood.

Tell your doctor if you have kidney failure. Your daily dose needs to be reduced according to the degree of your kidney failure.

If these warnings apply to you at any time in the past, please consult your doctor.

Using UCEDANE with food and drink

UCEDANE should be taken orally before meals or feeding.

The tablets should be dissolved in at least 5 to 10 mL of water and drunk immediately.

Pregnancy

Consult your doctor or pharmacist before using the medicine.

The effects of UCEDANE on pregnancy and the unborn child (fetus) are unknown. If you are pregnant or planning to become pregnant, consult your doctor or pharmacist.

UCEDANE should not be used during pregnancy.

If you realize you are pregnant during your treatment, consult your doctor or pharmacist immediately.

Breastfeeding

Consult your doctor or pharmacist before using the medicine.

If you are breastfeeding, consult your doctor or pharmacist before taking this medicine.

It has not been studied whether carglumic acid passes into breast milk in women. However, since carglumic acid has been shown to be present in the milk of lactating rats and to have potential toxic effects on fed offspring, you should not breastfeed your baby while taking UCEDANE.

Driving and using machines

Its effect on driving and using machines is unknown.

Important information about some excipients in UCEDANE

Ucedane contains sodium. This medicine contains less than 1 mmol (23 mg) of sodium per maximum daily dose, essentially "sodium-free".

Use with other medicines

If you are currently using or have recently used any prescription or over-the-counter medicine, please inform your doctor or pharmacist about them.

3. How to use UCEDANE?

Instructions for proper use and dose/frequency of administration:

Always follow your doctor's instructions exactly when using UCEDANE. If you are unsure, consult your doctor and pharmacist.

General dose:

The initial daily dose applied is usually 100 mg per kilogram of body weight, and doses up to a maximum of 250 mg per kilogram of body weight can be used (for example, if you are 10 kg, you should take 1 g or 5 tablets per day). In patients with N-acetylglutamate synthase deficiency, in the long term, the daily dose usually ranges from 10 mg to 100 mg per kilogram of body weight.

Your doctor will determine the appropriate dose to keep your blood ammonia levels normal.

Route and method of administration:

Tablets should be dispersed in at least 5-10 mL of water and drunk immediately. UCEDANE should ONLY be

administered orally or via a feeding tube into the stomach.

If the patient is in hyperammonemic coma (excessive elevation of ammonia levels in the blood), UCEDANE is rapidly administered with a syringe into the tube inserted for feeding.

Tell your doctor if you have kidney failure. Your daily dose needs to be reduced according to the degree of your kidney failure.

Different age groups:

Use in children:

'Your doctor will determine and administer your medicine's dose based on your illness.'

Use in the elderly:

'Your doctor will determine and administer your medicine's dose based on your illness.'

Special use cases:

Kidney/liver failure:

Your doctor will adjust your daily dose according to the degree of your kidney failure as follows:

Dose adjustment should be made according to GFR (Glomerular filtration rate).

- Patients with moderate renal impairment (GFR 30-59 mL/minute)
  - The recommended initial dose for patients with hyperammonemia due to N-acetylglutamate synthase (NAGS) deficiency or organic acidemia is 50 mg/kg/day to 125 mg/kg/day,
  - For long-term use, the daily dose should be in the range of 5 mg/kg/day to 50 mg/kg/day and should be adjusted individually to maintain normal ammonia plasma levels.
- Patients with severe renal impairment (GFR ≤ 29 mL/minute)
  - The recommended initial dose for patients with hyperammonemia due to N-acetylglutamate synthase (NAGS) deficiency or organic acidemia is 15 mg/kg/day to 40 mg/kg/day,
  - For long-term use, the daily dose should be in the range of 2 mg/kg/day to 20 mg/kg/day and should be adjusted individually to maintain normal ammonia plasma levels.

If you have used more UCEDANE than you should:

If you have used more UCEDANE than you should, talk to a doctor or pharmacist.

Undesirable effects such as tachycardia (increased heart rate), excessive sweating, increased bronchial secretion (respiratory tract secretion), increased body temperature and restlessness may be observed.

Do not take a double dose to make up for forgotten doses.

Effects that may occur when treatment with UCEDANE is discontinued

No effects are expected when treatment is discontinued.

If you have the impression that the effect of UCEDANE is too strong or too weak, talk to your doctor or pharmacist.

4. What are the possible side effects?

Like all medicines, UCEDANE may cause side effects in people sensitive to the substances contained in it.

Side effects are listed in the following categories: Very common: May be seen in at least one in 10 patients. Common: May be seen in less than one in 10 patients, but more than one in 100 patients. Uncommon: May be seen in less than one in 100 patients, but more than one in 1,000 patients. Rare: May be seen in less than one in 1,000 patients. Very rare: May be seen in less than one in 10,000 patients. Unknown: Cannot be estimated from the available data.

Common:

- Increased sweating

Uncommon:

- Bradycardia (decreased heart rate),
- Diarrhea
- Fever
- Increased transaminases (liver enzymes),
- Vomiting

Unknown:

- Rash

If you encounter any side effects not mentioned in these instructions for use, inform your doctor or pharmacist.

Reporting of side effects

If you experience any side effects, whether or not they are mentioned in the Instructions for Use, talk to your doctor, pharmacist or nurse. You can also report side effects to the Turkish Pharmacovigilance Centre (TÜFAM) by clicking on the "Drug Side Effect Notification" icon on the website www.titck.gov.tr or by calling the side effect notification line at 0 800 314 00 08.

By reporting side effects, you will contribute to obtaining more information about the safety of the medicine you are using.

5. How to store UCEDANE?

Keep UCEDANE out of the sight and reach of children and in its packaging.

Use in accordance with the expiry date.

Do not use UCEDANE after the expiry date stated on the packaging.

The expiry date is the last day of the specified month (written after “exp” on the box or label).

Store at room temperature below 30°C.

Do not dispose of expired or unused medicines in the trash! Hand them over to the collection system determined by the Ministry of Environment, Urbanization and Climate Change.

Marketing authorisation holder:

LUCANE PHARMA Sağlık Hizmetleri Ltd.Şti

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