

PACKAGE LEAFLET: INFORMATION FOR THE USER

CYSTAGON 50 mg Hard Capsules **CYSTAGON 150 mg Hard Capsules** (cysteamine bitartrate) (mercaptamine bitartrate)

Read all of this leaflet carefully before you start using this medicine.

- Keep this leaflet. You may need to read it again.
- If you have further questions, please ask your doctor or your pharmacist.
- This medicine has been prescribed for you. Do not pass it on to others. It may harm them, even if their symptoms are the same as yours.
- If any of the side effects gets serious, or if you notice any side effects not listed in this leaflet, please tell your doctor or pharmacist.

In this leaflet:

1. What CYSTAGON is and what it is used for
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1. WHAT CYSTAGON IS AND WHAT IT IS USED FOR

Cystinosis is a metabolic disease called ‘nephropathic cystinosis’ which is characterized by an abnormal accumulation of the amino acid cystine in various organs of the body such as the kidney, eye, muscle, pancreas, and brain. Cystine build up causes kidney damage and excretion of excess amounts of glucose, proteins and electrolytes. Different organs are affected at different ages.

CYSTAGON is prescribed to manage this rare inherited disorder. CYSTAGON is a medicine that reacts with cystine to decrease its level within the cells.

2. BEFORE YOU USE CYSTAGON

Do not use CYSTAGON

- if you -or your child- are allergic (hypersensitive) to cysteamine bitartrate or penicillamine or any of the other ingredients of Cystagon.
- If you are pregnant, this is particularly relevant during the first trimester
- if you are breast-feeding.

Take special care with CYSTAGON

- When your or your child’s disorder has been confirmed by leucocyte cystine measurements, the therapy with CYSTAGON must be started as soon as possible.
- A few cases of skin lesions on elbows like little hard lumps have been reported in children treated with high doses of different cysteamine preparations. These lesions were associated with skin striae and bone lesions such as fracture and bone deformities, and with laxity of joints. Your doctor could require a regular physical and X-ray examination for the skin and the bones to control the effects of the medicinal product. Self examination of your or your child’s skin is recommended. If any skin or bone abnormalities appear, please inform your doctor immediately.
- Your doctor could require controlling the blood cell count on a regular basis.
- CYSTAGON has not been shown to prevent cystine crystals accumulating in the eye. Where cysteamine ophthalmic solution has been used for that purpose, its usage should continue.

- In contrast to phosphocysteamine, another active substance close to cysteamine bitartrate, CYSTAGON does not contain phosphate. You may already be receiving phosphate supplements and the dose of these may need to be changed when CYSTAGON is substituted for phosphocysteamine.
- To avoid any risk of aspiration in the lungs, the capsules should not be given to children under the age of approximately 6 years.

Using other medicines

Please inform your doctor or pharmacist if you are taking or have recently taken any other medicines, even those not prescribed.

Using CYSTAGON with food and drink

For children under approximately six years of age, the hard capsule may be opened and the contents sprinkled on food (e.g. milk, potatoes or starch based foods) or mixed in formula. Do not add to acidic drinks e.g. orange juice. Consult the doctor for complete directions.

Pregnancy

You should not use CYSTAGON if you are pregnant. Please consult your doctor if you plan to become pregnant.

Breast-feeding

CYSTAGON must not be used during breast-feeding.

Driving and using machines

CYSTAGON may cause some drowsiness. When starting therapy, you or your child should not engage in potentially hazardous activities until the effects of the drug are well known.

3. HOW TO USE CYSTAGON

Always use CYSTAGON exactly as your doctor or your child's doctor has told you. You should check with your doctor if you are not sure.

The dose of CYSTAGON prescribed for you or your child will depend on your or your child's age and weight.

For children up to age 12 years, the dose will be based on the body size (surface area), the usual dose being 1.30 g/m² of body surface area per day.

For patients over age 12 and over 50 kg weight, the usual dose is 2g/day.

In any case the usual dose should not exceed 1.95 g/m²/day.

CYSTAGON should be taken or given only by mouth and exactly as your or your child's doctor directs. In order for CYSTAGON to work correctly, you must do the following:

- Follow your doctor's directions exactly. Do not increase or decrease the amount of medicine without your doctor's approval.
- Hard capsules should not be given to children under approximately six years of age because they may not be able to swallow them and they may choke. For children under approximately six years of age, the hard capsule may be opened and the contents sprinkled on food (e.g. milk, potatoes or starch based foods) or mixed in formula. Do not add to acidic drinks e.g. orange juice. Consult the doctor for complete directions.
- Your or your child's medical treatment may include, in addition to CYSTAGON, one or more supplements to replace important electrolytes lost through the kidneys. It is important to take or give these supplements exactly as instructed. If several doses of the supplements are missed or weakness or drowsiness develops, call the doctor for instructions.
- Regular blood tests to measure the amount of cystine inside white blood cells are necessary to help determine the correct dose of CYSTAGON. Your or your child's doctor will arrange for the blood tests to be done. Regular blood and urine tests to measure the levels of the body's important electrolytes are also necessary to help your or your child's doctor correctly adjust the doses of these supplements.

CYSTAGON should be taken 4 times a day, every 6 hours, preferably just after or with food. It is important to take the dose as close to every 6 hours as possible.

Treatment with CYSTAGON should continue indefinitely, as instructed by your doctor.

If you use more CYSTAGON than you should:

You should contact your or your child's doctor or the hospital emergency department immediately if more medicine has been taken than has been prescribed, drowsiness develops .

If you forget to take CYSTAGON:

If a dose of medicine is missed, it should be taken as soon as possible. However if it is within two hours of the next dose, skip the missed dose and go back to the regular dosing schedule. Do not take a double dose to make up for a forgotten dose.

4. POSSIBLE SIDE EFFECTS

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

CYSTAGON may cause some people to become drowsy or less vigilant than they are normally. Make sure you know how you or your child reacts to this medicine before doing anything that could be dangerous if not alert.

The following side effects were reported as follows: very common (occurring in at least one in 10 patients), common (occurring in at least one in 100 patients), uncommon (occurring in at least one in 1,000 patients), rare (occurring in at least one in 10,000 patients), very rare (occurring in at least one in 100,000 patients).

- Very common: vomiting, nausea, diarrhoea, loss of appetite, fever and sensation of sleep
- Common: abdominal pain or discomfort, unpleasant breath and body odour, skin eruption, gastroenteritis, fatigue, headache, encephalopathy (brain disorder) and liver function test abnormalities.
- Uncommon: skin striae, skin lesion (little-hard lumps on elbows), joint laxity, leg pain, bone fracture, scoliosis (deviation of the vertebral column), bone deformity and fragility, hair discoloration, severe allergic reaction, somnolence, fits, nervousness, hallucination, decrease in white blood cells, gastrointestinal ulcer manifested by bleeding in the digestive tract and effect on the kidney manifested by swelling of the extremities and weight gain.

Since some of these side effects are serious, ask your or your child's doctor to explain their warning signs.

Reporting of side effects

If you get any side effects, talk to your doctor, nurse or pharmacist. This includes any possible side effects not listed in this leaflet. You can also report side effects directly via

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

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

5. HOW TO STORE CYSTAGON

Keep out of the reach and sight of children.

Do not use after the expiry date which is stated on the label. The expiry date refers to the last day of that month.

Do not store above 25°C and keep the container tightly closed in order to protect from light and moisture.

6. FURTHER INFORMATION

What CYSTAGON contains

- The active substance is cysteamine bitartrate (mercaptamine bitartrate). Each hard capsule of CYSTAGON 50 mg contains 50 mg of cysteamine (as mercaptamine bitartrate) Each hard capsule of CYSTAGON 150 mg contains 150 mg of cysteamine (as mercaptamine bitartrate)
- The other ingredients are microcrystalline cellulose, starch, pregelatinized, magnesium stearate/sodium lauryl sulphate, colloidal silicon dioxide, croscarmellose sodium, capsule shells: gelatin, titanium dioxide, black ink on hard capsules (E172).

What CYSTAGON looks like and contents of the pack

Hard Capsules

- Cystagon 50 mg: white, opaque hard capsules with CYSTA 50 on the body and MYLAN on the cap. Bottles of 100 or 500 hard capsules. All pack sizes may be not marketed.

- Cystagon 150 mg: white, opaque hard capsules with CYSTAGON 150 on the body and MYLAN on the cap.

Bottles of 100 or 500 hard capsules. All pack sizes may be not marketed.

Marketing Authorisation Holder

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For any information about this medicine, please contact the local representative of the Marketing Authorisation Holder:

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This leaflet was last approved on April 2019

Detailed information on this medicine is available on the European Medicines Agency web site:

<http://www.ema.europa.eu>