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Cufence (*trientine dihydrochloride*)

An overview of Cufence and why it is authorised in the EU

What is Cufence and what is it used for?

Cufence is a medicine used to treat patients aged 5 years and older with Wilson's disease, a genetic condition in which copper absorbed from food builds up in the body, particularly in the liver and the brain, causing damage. Cufence is used in patients who cannot take D-penicillamine, another medicine for this condition.

Cufence contains the active substance trientine dihydrochloride.

How is Cufence used?

Cufence can only be obtained with a prescription and treatment should be started by a specialist with experience in the management of Wilson's disease.

Cufence is available as 200 mg capsules. The recommended daily dose is 4 to 8 capsules in adults, and 2 to 5 capsules in children. The capsules are taken in 2 to 4 divided doses. Doses are adjusted according to patient response and levels of copper in the body. Cufence should be taken on an empty stomach, at least one hour before or two hours after meals. Cufence must be stored in a refrigerator (between 2ºC and 8ºC).

For more information about using Cufence, see the package leaflet or contact your doctor or pharmacist.

How does Cufence work?

The active substance in Cufence, trientine, is a chelating agent. It works by attaching to copper in the body and forming a complex that is then eliminated in the urine and faeces.

What benefits of Cufence have been shown in studies?

The active substance in Cufence, trientine, has been shown to improve symptoms of liver and neurological disease in patients with Wilson's disease who could no longer take D-penicillamine.

In a study of medical records of 77 patients treated with trientine for at least six months, symptoms of liver disease improved in almost half (49%) of the patients treated, and neurological symptoms

improved in 14% of the patients. A small proportion of patients had deteriorating symptoms: 5% with worsening liver symptoms and 3% with worsening neurological symptoms.

What are the risks associated with Cufence?

The most common side effect with Cufence (which may affect up to 1 in 10 people) is nausea (feeling sick), especially when starting treatment. Skin rash may affect up to 1 in 100 people. Duodenitis (inflammation of the duodenum, the part of the gut leading out of the stomach) and severe colitis (inflammation in the large bowel causing pain and diarrhoea) have also been reported. In some patients, neurological deterioration can occur at the start of the treatment, with symptoms such as dystonia (involuntary muscle contractions), stiffness, tremor (shaking) and dysarthria (difficulty speaking).

For the full list of side effects and restrictions of Cufence, see the package leaflet.

Why is Cufence authorised in the EU?

Trientine has been used for over 30 years to treat patients with Wilson's disease. Although D-penicillamine is the main treatment for this condition, trientine is effective at improving the symptoms of liver and neurological disease in patients who cannot take this medicine. The safety of Cufence has been shown to be similar to that of other trientine medicines.

The European Medicines Agency therefore decided that Cufence's benefits are greater than its risks and it can be authorised for use in the EU.

What measures are being taken to ensure the safe and effective use of Cufence?

The company that markets Cufence will conduct a study to further characterise the effectiveness of trientine in the treatment of Wilson's disease, including its effect on the associated liver, neurological or psychiatric symptoms and what the dose should be during early treatment.

Recommendations and precautions to be followed by healthcare professionals and patients for the safe and effective use of Cufence have also been included in the summary of product characteristics and the package leaflet.

As for all medicines, data on the use of Cufence are continuously monitored. Side effects reported with Cufence are carefully evaluated and any necessary action taken to protect patients.

Other information about Cufence

Cufence received a marketing authorisation valid throughout the EU on 25 July 2019.

Further information on Cufence can be found on the Agency's website: www.ema.europa.eu/medicines/human/EPAR/cufence

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