



Dosing guide.



Read the instructions in the enclosed Patient Information Leaflet for information on preparation and administration techniques. This dosing guide does not take the place of talking with your healthcare team about your condition or treatment.

INDICATION

DOJOLVI is a prescription medicine used to treat long-chain fatty acid oxidation disorders (LC-FAOD) in children and adults.

Please see Important Safety Information on the inside spread and continued on the back cover.

LC-FAOD DEFINED

Long-chain fatty acid oxidation disorders (LC-FAOD) are a group of rare but life-threatening metabolic genetic disorders that include specific enzyme deficiencies.



People living with LC-FAOD have difficulty producing enough energy because of their body's inability to use long-chain fatty acids as an energy source during times of:

- Fasting
- Illness
- Prolonged exercise

DOJOLVI is the first and only FDA-approved treatment for patients of all ages diagnosed with long-chain fatty acid oxidation disorders (LC-FAOD).

IMPORTANT SAFETY INFORMATION

What are the possible side effects of DOJOLVI?

- **Feeding tube problems.** Feeding tubes may not work as well or stop working over time when taking DOJOLVI. **Do not use DOJOLVI in feeding tubes made of polyvinyl chloride (PVC),** a solid plastic material. Monitor the feeding tube to make sure it is working properly.
- **Intestinal absorption problems in patients with pancreatic insufficiency.** If you have pancreatic insufficiency, consult with your healthcare provider as it may affect how well DOJOLVI works.

Please see additional Important Safety Information on the back cover.

WORKING WITH YOUR HEALTHCARE TEAM

All patients treated with DOJOLVI should be under the care of clinical specialists knowledgeable in appropriate disease-related dietary management based upon current nutritional recommendations.

These specialists, which may include metabolic geneticists, dietitians, and nurse practitioners, will work together as your healthcare team and:

- May start you on a low dose of DOJOLVI and slowly increase your dose to help avoid side effects
- Ensure you are receiving frequent monitoring and nutritional counseling appropriate to your specific needs when taking DOJOLVI

**Always take DOJOLVI exactly
as your healthcare team advises.**

STORING DOJOLVI



Store DOJOLVI upright at room temperature between 68 and 77 °F (20 and 25 °C)



Do not freeze DOJOLVI



When the bottle of DOJOLVI has been opened, use within 90 days or by the expiration date on the bottle, whichever comes first



Do not store product in containers made of polystyrene or polyvinyl chloride (PVC)

TAKING DOJOLVI



If you are taking another medium-chain triglyceride (MCT) formulation, stop taking the MCT before starting DOJOLVI



Use an oral syringe or a measuring cup to measure the prescribed dose of DOJOLVI



Do not mix or give DOJOLVI using containers, dosing syringes, or measuring cups made of polystyrene (a type of plastic that can be solid or foam) or polyvinyl chloride (PVC), a solid plastic material



Mix or give DOJOLVI using containers, dosing components, or utensils made of materials such as stainless steel, glass, or high-density polyethylene (HDPE), polypropylene, low-density polyethylene, polyurethane, and silicone (types of plastic materials)

**4x
daily**

DOJOLVI should be taken at least 4 times a day with meals or snacks, and always mixed well with soft food or drink

DOJOLVI can be mixed with the following soft food or drink:

- Plain or artificially sweetened fat-free yogurt
- Fat-free milk, formula, or cottage cheese
- Whole grain hot cereal
- Fat-free low-carbohydrate pudding, smoothies, applesauce, or baby food



The mixture may be stored for up to 24 hours in the refrigerator

If you miss a dose, take the next dose as soon as possible. Take the following doses 3 to 4 hours apart. If it is not possible to take all the doses for the day, skip the missed dose.

TRACKING DAILY DOSES

It's important to stick with your treatment plan. Frequent monitoring, especially as you begin taking DOJOLVI, can:

- Help you adapt to and maintain your treatment plan
- Help your healthcare team decide if any dosing modifications are needed

EXAMPLE

Not actual patient data; for illustrative purposes only.

Divide the prescribed daily amount of DOJOLVI by at least 4 to determine each dose amount.

**Ex: 56 mL/4
= 14 mL per dose**

Prescribed daily amount of DOJOLVI: 56 ml

Maximum fasting interval: 6 hr

Daily medical food/formula prescription: _____

Prescribed daily fat intake from food: 16 g

	AMOUNT	TIME TAKEN	NOTES
DOSE 1	14 ml	7:45 am	
DOSE 2	14 ml	11:15 am	
DOSE 3	14 ml	3:00 pm	
DOSE 4	14 ml	6:20 pm	Mixed w/ fat-free milk

Recording the exact time you take your doses will help you track medication for the day.

Use the notes section to communicate important information to your healthcare team.

FOR YOUR HEALTHCARE TEAM

PROVIDER: _____ **PHONE:** _____

Prescribed daily amount of DOJOLVI: _____

Maximum fasting interval: _____

Daily medical food/formula prescription: _____

Prescribed daily fat intake from food: _____

IMPORTANT SAFETY INFORMATION (CONTINUED)

What are the possible side effects of DOJOLVI (continued)?

- The most common side effects of DOJOLVI are:
 - stomach (abdominal) pain
 - diarrhea
 - vomiting
 - nausea

These are not all the possible side effects of DOJOLVI. Call your healthcare provider for medical advice about side effects. You may report side effects to Ultragenyx Pharmaceutical Inc. at 1-888-756-8657 or FDA at 1-800-FDA-1088.

Before taking DOJOLVI, tell your healthcare provider about all of your medical conditions, including if you:

- are pregnant or plan to become pregnant. It is not known if DOJOLVI will harm your unborn baby. **Pregnancy Safety Study:** There is a pregnancy safety study for women who take DOJOLVI during pregnancy. The purpose of this study is to collect information about your health and your baby's health. You can talk to your healthcare provider or contact 1-888-756-8657 to enroll in this study or get more information.
- are breastfeeding or plan to breastfeed. It is not known if DOJOLVI passes into breast milk. Talk to your healthcare provider about the best way to feed your baby if you take DOJOLVI.
- are taking a pancreatic lipase inhibitor, such as orlistat, as it may affect how well DOJOLVI works.

Tell your healthcare provider about all the medicines you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

How should I take DOJOLVI?

- See the detailed **“Instructions for Use”** at the end of the Patient Information Leaflet for instructions about how to mix and take DOJOLVI by mouth in soft foods or drinks or how to mix and give DOJOLVI through feeding tubes.
- Take DOJOLVI exactly as your healthcare provider tells you.
- Your healthcare provider may start you on a low dose of DOJOLVI and slowly increase your dose to help avoid side effects. **If you are taking another medium chain triglyceride (MCT) product, stop taking the MCT before starting DOJOLVI.**

Please see the enclosed full Prescribing Information, including the Patient Information Leaflet, for additional Important Safety Information.