

Patient Information



DOJOLVI is available for children and adults living with



What is DOJOLVI used for?

DOJOLVI is used as a source of calories and fatty acids to treat adults and children with long-chain fatty acid oxidation disorders (LC-FAOD).

Read the Patient Medication Information leaflet for information and instructions on how to prepare and administer DOJOLVI. Use this brochure as a discussion tool with your healthcare professional. It does not take the place of talking with your healthcare professional about your condition or treatment.



Not actual patients.

DOJOLVI is a source of calories and fatty acids for adults and children with LC-FAOD.

What are LC-FAOD?

LC-FAOD are a group of rare, life-threatening metabolic genetic disorders that prevent the body from breaking down long-chain fatty acids into energy during metabolism due to specific enzyme deficiencies.



CPT I Carnitine palmitoyltransferase I deficiency

CACT

Carnitine-acylcarnitine translocase deficiency

CPT II Carnitine palmitoyltransferase II deficiency

VLCAD

Very long-chain acyl-CoA dehydrogenase deficiency

TFP Trifunctional protein deficiency

LCHAD

Long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency

People living with LC-FAOD have difficulty producing enough energy. This is because their bodies cannot use long-chain fatty acids as an energy source during times of:

Fasting
 Illness

Prolonged exercise

Stress





Not actual patients.

DOJOLVI is a source of medium-chain fatty acids that can provide energy to the body.



How does DOJOLVI work?

The body usually uses glucose (sugar) for energy; however, when all the glucose is used up, the body also gets energy from fat. People with LC-FAOD cannot use long-chain fat for energy. DOJOLVI is a source of medium-chain fatty acids. It can provide energy to the body in two ways:

- 1. By working around the process to break down long-chain fatty acids into energy
- 2. By providing another source of energy

Working with your healthcare professional

Your team of healthcare professionals may include pediatricians, metabolic geneticists, dietitians and nurse practitioners.

You will have regular visits with these individuals so that they can determine how DOJOLVI is working for you, make adjustments to your dose and talk to you about your diet.

Storing DOJOLVI

Store DOJOLVI upright at 15° to 30°C. **Do not** freeze.

Once the bottle of DOJOLVI has been opened, use the contents within 9 months or by the expiration date on the bottle, whichever is earlier. There is a place on the DOJOLVI labels to write the date the bottle is opened.

Do not use or store DOJOLVI in containers made of polystyrene or polyvinyl chloride (PVC).

Always take DOJOLVI exactly as directed by your healthcare professional.

Preparing and taking DOJOLVI

DOJOLVI can be taken by mouth or given through a feeding tube.

Follow instructions in the Patient Medication Information regarding preparing and taking DOJOLVI and the supplies you will need.

Take DOJOLVI 4 or more times throughout the day with a meal or snack. You will take DOJOLVI every 3 to 4 hours.

DOJOLVI must be mixed with liquid or soft food. Taking DOJOLVI on its own may cause side effects.

Your healthcare professional will tell you how much DOJOLVI to take and how much liquid or soft food to use.

Only mix or give DOJOLVI using supplies (including feeding tubes) made of stainless steel, glass, high- or low-density polyethylene, polypropylene, polyurethane and silicone.

Do not mix or give DOJOLVI using items made of polystyrene or PVC.

If taking DOJOLVI by mouth, mix it with soft food or liquid. The mixture may be stored for up to 24 hours in the refrigerator.

If giving DOJOLVI through a feeding tube, mix it with prepared tube feeding formula, as directed by your healthcare professional.

If you miss a dose (one of the portions taken throughout the day), take the next dose as soon as possible. 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 get used to and maintain your treatment plan, and can help your healthcare professional decide if changes to your dose are needed.

Example

Not actual patient data; for illustrative purposes only.	Ma	al daily amount ximum fasting p	period: <u>6hr</u>		
Divide the total daily amount of DOJOLVI by 4 or more. This		commended da commended da	ily fat total from	n food:	_
will give the amount		Amount:	Time taken:	Notes:	
for each dose or portion to take	Dose 1 Dose 2 Dose 3	● 14 m l	7:45 am		
throughout the day. Take DOJOLVI every 3 to 4 hours. Example: 56 mL divided by 4 doses equals 14 mL per dose		14 ml	11:15 am		
		14 ml	3:00 pm		
	Dose 4	14 ml	6:20 pm	Mixed with fat-free milk	દ
Record the exact time you take your doses. This will help you track your DOJOLVI for the day.					ł

You may use the Dosing Tracker in this guide to keep track of your daily doses. Ask your healthcare professional about obtaining additional copies of the Dosing Tracker, or contact the UltraCare Patient Support Program at 1-833-388-5872 (U-LTRA).

For your healthcare professional to complete:

Healthcare professional:	Phone:			
Total daily amount of DOJOLVI:				
Maximum fasting period:				
Recommended daily medical food/formula:				
Recommended daily fat total from food:				

Dosing Tracker

Use this Dosing Tracker as directed by your healthcare professional. Use it along with the Dosing Guide and Patient Medication Information.

For additional copies, ask your healthcare professional or contact the UltraCare Patient Support Program at 1-833-388-5872 (U-LTRA).

Total daily amount of DOJOLVI:	Date:	
Maximum fasting period:		
Recommended daily medical food/formula:		
Recommended daily fat total from food:		

Take DOJOLVI 4 or more times throughout the day with a meal or snack.

	Amount:	Time taken:	Notes:
Dose 1			
Dose 2			
Dose 3			
Dose 4			
Dose 5			
Dose 6			
Dose 7			
Dose 8			

Daily fat intake from food

(list foods and fat grams below to help you reach the recommended daily total listed above)

Foods:		Total fat (grams):
	Daily fat total:	
Healthcare		Phone



Not actual patients.

Use the Dosing Tracker as directed by your healthcare professional. Contact the UltraCare Patient Support Program at 1-833-388-5872 (U-LTRA) or ask your healthcare professional for additional copies.

Important Safety Information

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take DOJOLVI. Talk about any health conditions or problems you may have, including if:

- You have **feeding tube problems**. Feeding tubes can break down over time. It is possible that using DOJOLVI with a feeding tube might also contribute to its breakdown. Check the feeding tube regularly to ensure it is not damaged and is working properly
- You have **pancreatic insufficiency**. This is a condition where the pancreas does not make enough enzymes to help digest food properly
- You or your partner are pregnant or planning to get pregnant
- You are breastfeeding or plan to breastfeed

Visits with your healthcare professional: You should have regular visits with this healthcare professional so they can:

- Determine how DOJOLVI is working for you
- Make adjustments to your dose
- Talk to you about your diet

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

The following may interact with DOJOLVI:

- Medicines called pancreatic lipase inhibitors, which act on enzymes in the gastrointestinal tract. An example is the medicine orlistat
- A medicine used to treat seizures, bipolar disorder or migraines called valproic acid

Important Safety Information (continued)

What are possible side effects from using DOJOLVI?

- Abdominal pain
- Vomiting
- Diarrhea
- Nausea

These are not all the possible side effects you may feel when taking DOJOLVI. If you experience any side effects not listed here, contact your healthcare professional.

If you want more information about DOJOLVI:

- Talk to your healthcare professional
- Find the full Product Monograph that is prepared for healthcare professionals and includes the Patient Medication Information by visiting the Health Canada website (<u>https://www.canada.ca/en/health-canada.html</u>), the manufacturer's website (<u>https://www.ultragenyx.com/canada/medicines/dojolvi-product-monograph-CANADA/</u>) or by calling 1-833-388-5872.

Patient support

Our UltraCare Case Managers are experienced professionals who are passionate about supporting you at every step. They can help you:





Determine your eligibility for **financial assistance**



Understand your **insurance coverage**



To learn more about our patient services: Call **1-833-388-5872 (U-LTRA)** Fax **1-833-592-2273 (CARE)**

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LC-FAOD CPTI CACT CPTI VLCAD TFP LCHAD

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