

<b>Clinical Policy Title:</b>	triheptanoin
<b>Policy Number:</b>	RxA.632
<b>Drug(s) Applied:</b>	DOJOLVI®
<b>Original Policy Date:</b>	09/14/2020
<b>Last Review Date:</b>	09/14/2020
<b>Line of Business Policy Applies to:</b>	All line of business

## Background

DOJOLVI is a medium-chain triglyceride indicated as a source of calories and fatty acids for the treatment of pediatric and adult patients with molecularly confirmed long-chain fatty acid oxidation disorders.

## Dosing Information

Drug Name	Indication	Dosing Regimen	Maximum Dose
triheptanoin (DOJOLVI ®)	Pediatric and adults with Long-chain fatty acid oxidation disorders (LC-FAOD)	<p><b>For patients not currently taking a MCT product:</b></p> <p>Initiate DOJOLVI at a total daily dosage of approximately 10% DCI divided into at least four times per day and increase to the recommended total daily dosage of up to 35% DCI over a period of 2 to 3 weeks.</p> <p><b>For patients switching from another MCT product</b></p> <p>Discontinue use of MCT products before starting DOJOLVI. Initiate DOJOLVI at the last tolerated daily dosage of MCT divided into at least four times per day. Increase the total daily dosage by approximately 5% DCI every 2 to 3 days until the target dosage of up to 35% DCI is achieved</p>	There is no defined maximum dosage; however, the recommended target daily dosage is up to 35% of the patient's total prescribed daily caloric intake (DCI)

This clinical policy has been developed to authorize, modify, or determine coverage for individuals with similar conditions. Specific care and treatment may vary depending on individual need and benefits covered by the plan. This policy is not intended to dictate to providers how to practice medicine, nor does it constitute a contract or guarantee regarding payment or results. This document may contain prescription brand name drugs that are trademarks of pharmaceutical manufacturers that are not affiliated with RxAdvance.

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		<p><b>Tolerability</b></p> <p>If a patient has difficulty tolerating 1/4 of the total daily dosage at one time, more frequent smaller doses may be considered</p>	
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**Dosage Forms**

- Oral Liquid, 100% w/w of triheptanoin

**Clinical Policy**

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

**I. Initial Approval Criteria**

**A. Long chain fatty acid oxidation disorders (LC-FAOD)**

1. Documented diagnosis of CPT I, CACT, CPT II, VLCAD, TFP, or LCHAD in long chain fatty acid oxidation disorders;
2. Target daily dosage does not exceed 35% of the patient’s total prescribed daily caloric intake (DCI).

**Approval Duration**

**Commercial:** 12 months

**Medicaid:** 12 months

**II. Continued Therapy Approval**

**A. Long chain fatty acid oxidation disorders (LC-FAOD)**

1. Currently receiving medication that has been authorized by RxAdvance or member has previously met initial approval criteria listed in this policy;
2. Member is responding positively to therapy.

**Approval Duration**

**Commercial:** 12 months

**Medicaid:** 12 months

**III. Appendices**

**APPENDIX A: Abbreviation/Acronym Key**

- LC-FAOD – long chain fatty acid oxidation disorders
- DCI – daily caloric intake
- CPT I – carnitine palmitoyltransferase I
- CACT – carnitine-acylcarnitine translocase
- CPT II – carnitine palmitoyltransferase II
- VLCAD – very-long-chain acyl-CoA dehydrogenase deficiency
- TFP – trifunctional protein
- LCHAD – long-chain L-3 hydroxyacyl-CoA dehydrogenase deficiency

**APPENDIX B: Therapeutic Alternatives**

Not applicable

**APPENDIX C: Contraindications/Boxed Warnings**

- Contraindication(s):
  - None reported
  
- Boxed Warning(s):
  - None reported

**APPENDIX D: General Information**

Not applicable

**References**

1. Product Information: DOJOLVI (triheptanoin) oral liquid. Ultragenyx Pharmaceutical Inc. (per FDA), Novato, CA, June 2020. Accessed July 23, 2020
2. Clinical Pharmacology [database online] powered by ClinicalKey. Tampa, FL: Elsevier, 2020. Accessed with subscription at: <http://www.clinicalkey.com>. Accessed June 29, 2020.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy established.	09/14/2020	09/14/2020