Clinical Policy Title:	triheptanoin
Policy Number:	RxA.632
Drug(s) Applied:	Dojolvi™
Original Policy Date:	09/14/2020
Last Review Date:	09/14/2021
Line of Business Policy Applies to:	All line of business

Background

Triheptanoin (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)	**For patients not currently taking a MCT product:** 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. **For patients switching from another MCT product:** 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. **Tolerability:** If a patient has difficulty tolerating 1/4 of the total daily dosage at one time, more frequent smaller doses may be considered.	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).

Dosage Forms

- Oral Liquid, 100% w/w of triheptanoin

Clinical Policy

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.

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria. The provision of provider samples does not guarantee coverage under the terms of the pharmacy benefit administered by RxAdvance. All criteria for initial approval must be met in order to obtain coverage.

I. Initial Approval Criteria

A. Long chain fatty acid oxidation disorders (LC-FAOD) (must meet all):

1. Documented diagnosis of CPT I, CACT, CPT II, VLCAD, TFP, or LCHAD in long chain fatty acid oxidation disorders;
2. Prescribed by or in consultation with a metabolic disease specialist or a physician who specializes in the management of long-chain fatty acid oxidation disorders;
3. 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) (must meet all):

1. Member is 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.
3. 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

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

- Feeding Tube Dysfunction: Regularly monitor the tube to ensure proper functioning and integrity.
- Intestinal Malabsorption in Patients with Pancreatic Insufficiency: Low or absent pancreatic enzymes may reduce absorption of Dojolvi™. Avoid administration of Dojolvi™ in patients with pancreatic insufficiency.

References

1. Dojolvi™ Prescribing Information. Novato, CA: Ultragenyx Pharmaceutical Inc. September 2020. Available at: https://www.ultragenyx.com/medicines/dojolvi-full-prescribing-information/. Accessed June 28, 2021.
2. Clinical Pharmacology [database online] powered by ClinicalKey. Tampa, FL: Elsevier, 2020. Accessed with subscription at: http://www.clinicalkey.com. Accessed June 28, 2021.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy established.	09/14/2020	09/14/2020
Policy was reviewed: 1. Statement about provider sample ‘‘The provision of provider samples does not guarantee coverage…” was added to Clinical Policy. 2. Initial Approval Criteria I.A.2 was updated to include prescriber criteria, “Prescribed by or in consultation with a metabolic disease specialist…”. 3. Continued Therapy Approval Criteria II.A.1 was rephrased to “Member is currently receiving medication that has been authorized by RxAdvance…”. 4. Continued Therapy Approval Criteria II.A.3 was updated to include “Target daily dosage does not exceed 35%...”. 5. Appendix D was updated to include warnings and precautions, “Feeding Tube Dysfunction…” and “Intestinal Malabsorption in Patients with Pancreatic Insufficiency…”. 6. References were reviewed and updated.	06/28/2021	09/14/2021