



PHARMACY POLICY STATEMENT North Carolina Marketplace

DRUG NAME	Dojolvi (triheptanoin)
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
COVERAGE REQUIREMENTS	Prior Authorization Required (Non-Preferred Product) QUANTITY LIMIT— see “dosage allowed”
LIST OF DIAGNOSES CONSIDERED NOT MEDICALLY NECESSARY	Click Here

Dojolvi (triheptanoin) is a **non-preferred** product and will only be considered for coverage under the **pharmacy** benefit when the following criteria are met:

Members must be clinically diagnosed with one of the following disease states and meet their individual criteria as stated.

LONG-CHAIN FATTY ACID OXIDATION DISORDERS (LC-FAOD)

For **initial** authorization:

1. Medication must be prescribed by or in consultation with a physician specializing in genetic metabolic disorders; AND
2. Chart notes must show the member has a molecularly confirmed diagnosis of an LC-FAOD (examples include: Very long-chain acylCoA dehydrogenase (VLCAD) Deficiency, Carnitine Palmitoyltransferase 2 (CPT2) Deficiency, Mitochondrial Trifunctional Protein (TFP) Deficiency, Long-chain 3 hydroxyacylCoA dehydrogenase (LCHAD) deficiency); AND
3. Member is symptomatic despite dietary management (e.g. a low-fat diet) and medium-chain triglyceride (MCT) oil for at least 90 days, unless contraindicated; AND
4. Member does not have pancreatic insufficiency; AND
5. Member will discontinue any other medium-chain triglyceride products before starting Dojolvi.
6. **Dosage allowed:** See package insert for titration details and equation for dose calculations based on individual’s daily caloric intake (DCI). Increase up to a total daily dose of 35% DCI.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must show improvement per 1 or more of the following parameters:
 - a) Reduced frequency or severity of major clinical events related to hypoglycemia, cardiomyopathy, and/or rhabdomyolysis.
 - b) Increased endurance and/or exercise tolerance (e.g. 6-minute walk test).

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

CareSource considers Dojolvi (triheptanoin) not medically necessary for the treatment of the diseases that are not listed in this document.

Qualified Health Plans offered in North Carolina by CareSource North Carolina Co., d/b/a CareSource.



DATE	ACTION/DESCRIPTION
09/25/2020	New policy for Dojolvi created.
11/17/2021	Annual review, no changes

References:

1. Dojolvi (triheptanoin) [package insert]. Novato, CA: Ultragenyx Pharmaceutical Inc.; 2020.
2. Vockley J, Burton B, Berry G, et al. UX007 for the treatment of long chain-fatty acid oxidation disorders: Safety and efficacy in children and adults following 24 weeks of treatment. *Molecular Genetics and Metabolism*. 2017;120(4):370-377. doi:10.1016/j.ymgme.2017.02.005
3. Vockley J, Burton B, Berry GT, et al. Results from a 78-week, single-arm, open-label phase 2 study to evaluate UX007 in pediatric and adult patients with severe long-chain fatty acid oxidation disorders (LC-FAOD). *J Inherit Metab Dis*. 2019;42(1):169-177. doi:10.1002/jimd.12038
4. Gillingham MB, Heitner SB, Martin J, et al. Triheptanoin versus trioctanoin for long-chain fatty acid oxidation disorders: a double blinded, randomized controlled trial. *J Inherit Metab Dis*. 2017;40(6):831-843. doi:10.1007/s10545-017-0085-8
5. Knottnerus SJG, Bleeker JC, Wüst RCI, et al. Disorders of mitochondrial long-chain fatty acid oxidation and the carnitine shuttle. *Rev Endocr Metab Disord*. 2018;19(1):93-106. doi:10.1007/s11154-018-9448-1
6. Merritt JL 2nd, Norris M, Kanungo S. Fatty acid oxidation disorders. *Ann Transl Med*. 2018;6(24):473. doi:10.21037/atm.2018.10.57
7. Merritt JL, Macleod E, Jurecka A, Hainline B. Clinical manifestations and management of fatty acid oxidation disorders. *Reviews in Endocrine and Metabolic Disorders*. July 2020. doi:10.1007/s11154-020-09568-3
8. Vockley J, Burton B, Berry G, et al. Effects of triheptanoin (UX007) in patients with long-chain fatty acid oxidation disorders: Results from an open-label, long-term extension study. *J Inherit Metab Dis*. September 2020. doi:10.1002/jimd.12313

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Revised date: 11/17/2021