# Dojolvi (triheptanoin)

| Override(s) | Approval Duration |
|---|---|
| Prior Authorization<br>Quantity Limit | Initial requests: 6 months<br>Continuation requests: 1 year |

| Medications | Quantity Limit |
|---|---|
| Dojolvi (triheptanoin) 100% w/w oral liquid | 1000 mL per 30 days* |

*May approve increased quantity per 30 days as needed based on confirmed daily caloric intake (DCI) and target daily dosage of Dojolvi. The target daily dosage of Dojolvi should not exceed 35% of DCI.

Total daily dose in ML = [DCI in kcal X target % dose of DCI]/8.3

## APPROVAL CRITERIA

Initial requests for Dojolvi (triheptanoin) may be approved if the following criteria are met:

I. Documentation is provided that individual is using for the treatment of long-chain fatty acid oxidation disorders (LC-FAOD) including (Gillingham 2017, Vockley 2021):
   A. Very long-chain acylCoA dehydrogenase (VLCAD) deficiency; **OR**
   B. Carnitine palmitoyltransferase-1 (CPT-1) deficiency; **OR**
   C. Carnitine palmitoyltransferase-2 (CPT-2) deficiency; **OR**
   D. Trifunctional protein (TFP) deficiency; **OR**
   E. Long-chain 3-hydroxyacylCoA dehydrogenase (LCHAD) deficiency; **OR**
   F. Carnitine-acylcarnitine translocase (CACT) deficiency;

**AND**

II. Documentation is provided that diagnosis has been confirmed by genetic testing showing a pathogenic mutation in ACADVL, CPT1A, CPT2, HADHA, HADHB, or SLC25A20; **AND**

III. Use will be in combination with dietary management.

Continuation requests for Dojolvi (triheptanoin) may be approved if the following criteria are met:

I. Documentation is provided that there is clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to improved cardiomyopathy symptoms, decreased hospitalizations or emergency department visits, decreased episodes of severe hypoglycemia or rhabdomyolysis); **AND**

II. Use will be in combination with dietary management.

Dojolvi (triheptanoin) may not be approved for the following:

I. In combination with another medium-chain triglyceride (MCT) agent; **OR**

II. Individual with pancreatic insufficiency.

**Key References**:


1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: December 12, 2021.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. 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 November; 40(6): 831–843. doi:10.1007/s10545-017-0085-8.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2021; Updated periodically.
5. Merritt JL, Norris M, Kanungo S. Fatty acid oxidation disorders. *Ann Transl Med*. 2018 Dec;6(24):473.
6. 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*. 2021 Jan;44(1):253-263.
7. Vockley J, Marsden D, McCracken E, et. al. Long-term major clinical outcomes in patients with long chain fatty acid oxidation disorders before and after transition to triheptanoin treatment – A retrospective chart review. *Mol Genet Metab*. 2015 Sep-Oct; 116(1-2): 53-60.