

**Dojolvi (triheptanoin)**  
**Effective 06/01/2021**

<b>Plan</b>	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange		<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input type="checkbox"/> Medical Benefit			<input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Specialty Limitations</b>	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.			
<b>Contact Information</b>	<b>Medical Benefit</b>	<b>Pharmacy Benefit</b>	Phone: 833-895-2611 Phone: 800-711-4555	Fax: 888-656-6671 Fax: 844-403-1029
<b>Exceptions</b>	N/A			

### Overview

Long chain fatty acid oxidation disorders are rare, autosomal recessive genetic disorders characterized by acute crises of energy production and chronic energy deficiency.

Dojolvi is 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 (LC-FAOD).

### Coverage Guidelines

Authorization may be reviewed for members new to the plan who are currently receiving treatment with Dojolvi, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

#### OR

Authorization may be granted when ALL of the following criteria are met:

1. Member has a diagnosis of long-chain fatty acid oxidation disorders (LC-FAOD)
2. Prescriber has submitted documentation of results from genetic testing or molecular analysis to confirm diagnosis (ex. CPT I or II, LCHAD, TFP, VLCAD deficiency)
3. Prescriber is a specialist in genetic or metabolic disease or medication is being prescribed in consultation with a specialist.
4. Documentation of a trial with a diet consisting of low-fat, high-carbohydrate, and avoidance of fasting
5. Member's current caloric intake

### Continuation of Therapy

Reauthorization may be granted for patient is experiencing benefit from therapy as evidenced by disease stability or disease improvement.

### Limitations

1. Initial approvals will be granted for 6 months.
2. Reauthorizations will be granted for 12 months.

### References

1. Dojolvi [package insert]. Novato, CA; Ultragenyx Pharmaceutical Inc.; November 2021.

2. 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; 42:169.
3. Vockley J, Burton B, Berry GT, 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. *Mol Genet Metab* 2017;120:370-377.
4. Merritt JL 2nd, Norris M, Kanungo S. Fatty acid oxidation disorders. *Ann Transl Med.* 2018;6(24):473.
5. American College of Medical Genetics and Genomics. ACT Sheet and Algorithms. Available at [https://www.acmg.net/ACMG/Medical-Genetics-Practice-Resources/ACT\\_Sheets\\_and\\_Algorithms.aspx](https://www.acmg.net/ACMG/Medical-Genetics-Practice-Resources/ACT_Sheets_and_Algorithms.aspx). Accessed July 20, 2020.

#### **Review History**

03/17/2021 – Created and Reviewed at March P&T

05/19/2021 – Updated; Added Specialty Pharmacy Limitation. Effective 06/01/2021.

09/21/2022 - Reviewed P&T; references updated; separated out Comm/Exch vs. MH.

