UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2026 P 2221-7
Program	Prior Authorization/Medical Necessity
Medication	Dojolvi® (triheptanoin)
P&T Approval Date	10/2020, 12/2020, 5/2022, 5/2023, 5/2024, 5/2025, 5/2026
Effective Date	8/1/2026

1. Background:

Dojolvi (triheptanoin) 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 (LC-FAOD).

2. Coverage Criteria[a]:

A. Initial Authorization

1. **Dojolvi** will be approved based on **all** of the following criteria:

   a. Submission of medical records confirming the diagnosis of long-chain fatty acid oxidation disorders (LC-FAOD) with at least **two** of the following diagnostic criteria:

      (1) Disease specific elevation of acylcarnitines on a newborn blood spot or in plasma
      (2) Low enzyme activity in cultured fibroblasts
      (3) Genetic testing demonstrating one or more pathogenic mutations in a gene associated with long-chain fatty acid oxidation disorders (e.g., CPT2, ACADVL, HADHA, or HADHB)

   -AND-

   b. Patient is not receiving Dojolvi in combination with any other medium-chain triglyceride (MCT) products

   -AND-

   c. Prescribed by a board-certified medical geneticist experienced in the treatment of long-chain fatty acid oxidation disorders (LC-FAOD)

   -AND-

   d. Target recommended daily dosage does not exceed 35% of the patient's total prescribed daily caloric intake (DCI)

   -AND-

   e. Patient is receiving disease related dietary management

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-AND-

f. If not diagnosed by newborn screening, patient has a history of clinical manifestations of long-chain fatty acid oxidation disorders LC-FAOD (e.g., rhabdomyolysis)

Authorization will be issued for 12 months

B. Reauthorization

1. **Dojolvi** will be approved based on all of the following criteria:

a. Documentation of positive clinical response to Dojolvi therapy (e.g., increased cardiac efficiency, decreased left ventricular wall mass, decreased incidence of rhabdomyolysis, etc.)

-AND-

b. Patient is not receiving Dojolvi in combination with any other medium-chain triglyceride (MCT) product

-AND-

c. Prescribed by a board-certified medical geneticist experienced in the treatment of long-chain fatty acid oxidation disorders (LC-FAOD)

-AND-

d. Target recommended daily dosage does not exceed 35% of the patient's total prescribed daily caloric intake (DCI)

-AND-

e. Patient is receiving disease related dietary management

Authorization will be issued for 12 months

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

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4. References:

1. Dojolvi [package insert]. Novato, CA: Ultragenyx Pharmaceutical, Inc.; October 2023.
2. 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
3. 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

Program	Prior Authorization/Medical Necessity – Dojolvi® (triheptanoin)
Change Control	
Date	Change
10/2020	New program
12/2020	Change to prescriber requirement criteria.
5/2022	Annual review with no change to clinical criteria. Updated reference.
5/2023	Annual review with no change to clinical criteria.
5/2024	Annual review. Revised listing of genes associated with long-chain fatty acid disorders. Revised initial authorization to 12 months. Updated references.
5/2025	Annual review with no change to clinical criteria.
5/2026	Annual review with no change to clinical criteria.

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