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Section:	Prescription Drugs	**Effective Date:**	April 1, 2024
Subsection:	Endocrine and Metabolic Agents	**Original Policy Date:**	August 7, 2020
Subject:	Dojolvi	**Page:**	1 of 4

Last Review Date: March 8, 2024

Dojolvi

Description

Dojolvi (triheptanoin)

Background
Dojolvi (triheptanoin) is a medium-chain triglyceride consisting of three odd-chain 7-carbon length fatty acids (heptanoate) that provide a source of calorie and fatty acids to bypass the long-chain fatty acid oxidation disorder (FAOD) enzyme deficiencies for energy production and replacement (1).

Regulatory Status
FDA-approved indication: 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 (LC-FAOD) (1).

Patient metabolic requirements should be determined by their daily caloric intake (DCI) prior to calculating the dose of Dojolvi. For patients receiving another medium-chain triglyceride (MCT) product, the product should be discontinued prior to the first dose of Dojolvi (1).

Dojolvi should be administered mixed with semi-solid food or liquids orally or enterally via a silicone or polyurethane feeding tube. Dojolvi should not be administered alone to avoid gastrointestinal upset (1).

Pancreatic enzymes hydrolyze triheptanoin and release heptanoate as medium-chain fatty acids in the small intestine. Low or absent pancreatic enzymes may result in reduced absorption of

heptanoate subsequently leading to insufficient supplementation of medium-chain fatty acids. Administration of Dojolvi in patients with pancreatic insufficiency should be avoided (1).

The most common adverse reactions to Dojolvi reported in the pooled safety population of Study 1 and Study 2 were gastrointestinal (GI)-related, and included abdominal pain (abdominal discomfort, abdominal pain, abdominal distention, abdominal pain upper, GI pain; 60%), diarrhea (44%), vomiting (44%), and nausea (14%). If a patient experiences a gastrointestinal (GI) adverse reaction, a dose reduction should be considered until the symptoms resolve (1).

The safety and effectiveness of Dojolvi have been established in pediatric patients aged birth and older (1).

Related policies

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Dojolvi may be considered **medically** if the conditions indicated below are met.

Dojolvi may be considered **investigational** for all other indications.

Prior-Approval Requirements

Diagnosis

Patient must have the following:

Long-chain fatty acid oxidation disorder (LC-FAOD)

AND ALL of the following:

1. Diagnosis of LC-FAOD has been molecularly confirmed
2. Patient will not be using Dojolvi with another medium-chain triglyceride (MCT) product
3. Prescriber agrees to monitor gastrointestinal (GI) adverse reactions and adjust the dose as needed

4. Patients using a feeding tube **only**: patient or caregiver will be advised to regularly inspect the feeding tube for proper functioning and integrity
5. **NO** pancreatic insufficiency

Prior – Approval *Renewal* Requirements

Diagnosis

Patient must have the following:

Long-chain fatty acid oxidation disorder (LC-FAOD)

AND ALL of the following:

1. Patient has had an improvement in symptoms (e.g., less episodes of rhabdomyolysis)
2. Patient will not be using Dojolvi with another medium-chain triglyceride (MCT) product
3. Patients using a feeding tube **only**: patient or caregiver has been advised to regularly inspect the feeding tube for proper functioning and integrity
4. **NO** pancreatic insufficiency
5. **NO** unacceptable gastrointestinal (GI) toxicity

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 12 months

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Dojolvi (triheptanoin) is a medium-chain triglyceride consisting of three odd-chain 7-carbon length fatty acids (heptanoate) that provide a source of calorie and fatty acids to bypass the long-chain fatty acid oxidation disorder enzyme deficiencies for energy production and replacement. The safety and effectiveness of Dojolvi have been established in pediatric patients aged birth and older (1).

Prior approval is required to ensure the safe, clinically appropriate, and cost-effective use of Dojolvi while maintaining optimal therapeutic outcomes.

References

1. Dojolvi [package insert]. Novato, CA; Ultragenix Pharmaceutical Inc.; October 2023.

Policy History

Date	Action
August 2020	Addition to PA
September 2020	Annual review
December 2020	Annual review and reference update. Addition of requirements per FEP: “prescriber agrees to monitor GI adverse reactions and adjust dose as needed” added to initiation and “no unacceptable GI toxicity” added to renewal
March 2021	Annual editorial review
March 2022	Annual review and reference update
March 2023	Annual review. Changed policy number to 5.30.070
March 2024	Annual review and reference update

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 8, 2024 and is effective on April 1, 2024.