

**DOJOLVI
(triheptanoin)****RATIONALE FOR INCLUSION IN PA PROGRAM****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



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BlueShield.**

Federal Employee Program.

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older (1).

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.