Mass General Brigham
Health Plan

Dojolvi (triheptanoin)
Effective 06/01/2021

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Exceptions	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 with 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. 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.