OPTIMA HEALTH PLAN

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-800-750-9692**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process may be delayed.**

Drug Requested: DOJOLVI™ (triheptanoin)

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength/Month: _____

Dosing Schedule: _____ **Length of Therapy:** _____

Diagnosis: _____ **ICD Code:** _____

RECOMMENDED DOSING:

- Caloic value of DOJOLVI = 8.3 kcal/mL
- Round the total daily dosage to the nearest whole number.
- Divide the total daily dosage into at least four approximately equal individual doses.

$$Total\ Daily\ Dose\ (\_\_\_mL) = \frac{Patients\ DCI\ (\_\_\_kcal)\ x\ Target\ \_\_\_\%\ dose\ of\ DCI}{8.3\frac{kcal}{mL}\ of\ DOJOLVI}$$

- Initiate DOJOLVI at a total daily dosage of approximately 10% DCI divided into at least four times per day and increase to the recommended total daily dosage of up to 35% DCI over a period of 2 to 3 weeks
- AGE: _____
- Total DCI (KCAL): _____

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization Length: 6 months

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- ❑ Patient must have a diagnosis of a long-chain fatty acid oxidation disorder (LC-FAOD) of either: CPTII, VLCAD, LCHAD, OR TFP/MTP AND confirmed by **two of the following** assessments:
  - ❑ FAOD Deficiency **(please document VLCAD, LCHAD, CPTII, or TFP/MTP)**:

Diagnosis	**Age & Date of assessment**	**FAOD Deficiency: (Please document: VLCAD, LCHAD, CPTII, MTP/TFP)**	**RESULTS FROM DIAGNOSIS (fill in or send the assessment)**	**Confirmed Diagnosis**
❑ Tandem mass spectrometry (MS/MS)				Acylcarnitine analysis: elevations of acylcarnitines on a newborn blood spot or in plasma https://www.acmg.net/ACMG/Medical-Genetics-Practice-Resources/ACT_Sheets_and_Algorithms.aspx
❑ Genetic Analysis			ACADVL, HADHA, HADHP, CPT2: _____	Splice variants or nonsense mutations were identified
❑ Enzyme assay (lymphocytes)				Low enzyme activity in cultured fibroblasts
❑ IVP assay				Elevations of long chain acyl CoA

AND

- ❑ Patient must have severe LC-FAOD confirmed by a history of ≥ 1 of the following despite therapy: (≥ 2X upper limit of age/gender-matched normal, or ≥ 500 units/L if age-matched reference not established)
  - ❑ Chronic elevated creatine kinase ([CK] ≥ 2 times the upper limit of normal) with ≥ 2 major clinical events (e.g., hospitalizations, hypoglycemia, cardiomyopathy, and rhabdomyolysis); **OR**
  - ❑ Episodic elevated CK with reported muscle dysfunction (e.g., frequent muscle fatigue, exercise intolerance, limitation of exercise); **OR**
  - ❑ Highly elevated CK (≥ 4 times the upper limit of normal); **OR**
  - ❑ Frequent (≥ 3 within a year or ≥ 5 within 2 years) severe major clinical events (e.g., hospitalizations, hypoglycemia, cardiomyopathy, and rhabdomyolysis); **OR**
  - ❑ Severe susceptibility to hypoglycemia after short periods of fasting (≥ 2 events within a year that require ongoing prophylactic management or recurrent symptomatic hypoglycemia requiring intervention ≥ 2 times per week); **OR**
  - ❑ Evidence of functional cardiomyopathy (echocardiogram documenting poor ejection fraction);

AND

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- ❑ Patient is being followed by a clinical specialist knowledgeable in appropriate disease-related dietary management (e.g., medical geneticist, genetic metabolic disorders, or a physician with a board certification in nutrition);

AND

- ❑ Patient is practicing appropriate dietary measures for their age and specific disorder (high carbohydrate, low long-chain fatty acids, avoidance of fasting);

AND

- ❑ Patient has tried and failed medium chain triglyceride and continue to have ONE of the following:
  - ❑ elevated CK] ≥ 2 times the upper limit of normal
  - ❑ hospitalizations
  - ❑ hypoglycemia
  - ❑ cardiomyopathy, **OR**
  - ❑ rhabdomyolysis

AND

- ❑ Patient is **NOT** taking a pancreatic lipase inhibitor (e.g., orlistat);

AND

- ❑ Patient will NOT receive an additional medium chain triglyceride while taking triheptanoin.

Reauthorization Approval Length: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ❑ Patient must continue to meet the above criteria;

AND

- ❑ Patient must demonstrate disease improvement and/or stabilization (e.g., cardiac function, exercise tolerance, reduction in major clinical events, including hospitalization) as evidenced by all of the following:
  - ❑ Creatinine kinase is within normal limits
  - ❑ Normal glycemic control
  - ❑ No documentation of recent hospitalization
  - ❑ No evidence of muscle fatigue

AND

- ❑ Patient does **NOT** experience serious treatment-related adverse effects (e.g., gastrointestinal effects).

Medication being provided by Specialty Pharmacy - PropriumRx

(Continued on next page; signature page is required to process request.)

(Please ensure signature page is attached to form.)

Not all drugs may be covered under every Plan

If a drug is non-formulary on a Plan, documentation of medical necessity will be required.

****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****

Member Name: _____

Member Optima #: _____ Date of Birth: _____

Prescriber Name: _____

Prescriber Signature: _____ Date: _____

Office Contact Name: _____

Phone Number: _____ Fax Number: _____

DEA OR NPI #: _____

REVISED/UPDATED: 12/7/2020