PrDOJOLVI®

Triheptanoin Oral Liquid

NEW BRUNSWICK

Ultragenyx Canada would like to announce that PrDOJOLVI® (triheptanoin) is eligible for public reimbursement in New Brunswick for patients with LC-FAOD based on the reimbursement criteria detailed below under the NB Drug Plans Formulary.

DOJOLVI® (triheptanoin) is indicated as a source of calories and fatty acids for the treatment of adult and pediatric patients with long-chain fatty acid oxidation disorders (LC-FAOD).

Please consult the Product Monograph at https://dojolvihcp.ca/wp-content/uploads/pdfs/Dojolvi_Final_PM.pdf for important information on contraindications, warnings and precautions, the conditions of clinical use, adverse reactions, drug interactions, and dosing instructions.

The Product Monograph is also available by calling us at 1-833-388-5872.

The DOJOLVI® Reimbursement Criteria for the NB Drug Plans Formulary Are as Follows:

For the treatment of patients with an acute life-threatening long-chain fatty acid oxidation disorder (LC-FAOD) who meet all of the following criteria:

- Alternative therapy to conventional even-chain medium-chain triglyceride (MCT) supplementation is required and
- One of the following circumstances is met:
  - The patient has a confirmed diagnosis of one of the types of LC-FAOD and is experiencing acute life-threatening events; or
  - The patient lacks a confirmed diagnosis of LC-FAOD but is presenting with acute life- threatening events consistent with LC-FAOD

Renewal Criteria

Renewals will be considered for patients meeting all of the following criteria:

- Patient who was initiated on triheptanoin without a confirmed diagnosis of LC-FAOD has subsequently received a confirmed diagnosis established by a specialist in metabolic diseases experienced in the treatment and management of LC-FAOD, with the type of LC- FAOD specified and the genetic and other findings provided to confirm the diagnosis.
- Patient is optimized on, and adherent to, appropriate dietary management.
- Patient continues to benefit from triheptanoin therapy. Requesters must include a description of the patient's current response to triheptanoin therapy and clearly outline how this response meets the clinical treatment goals established at initiation.

Clinical Notes

1. Acute life-threatening events consistent with LC-FAOD may include:
    - A catastrophic presentation with acute or recurrent rhabdomyolysis with severe pain, compartment syndrome, acute renal failure requiring hospitalization and life-saving interventions including dialysis, treatment of hyperkalemia, and surgical treatment of compartment syndrome
    - Severe hypoglycemia, recurrent or acute, with or without seizures
    - Cardiomyopathy with or without arrhythmia

2. Requests should specify the acute life-threatening events that the patient presents with that are consistent with LC-FAOD and include clinical and biochemical findings of impacted organ systems that support warranted triheptanoin initiation.

3. Individualized treatment goals for triheptanoin treatment must be submitted with the initial coverage request.

4. Patient's Daily Caloric Intake (DCI) requirements must be provided with all requests.

Claim Notes

- Must be prescribed by a physician with experience in the management of LC-FAOD.
- Approvals will be for a maximum of 35% of the patient's total DCI.
- Approval period: 1 year.

For more information on applying for DOJOLVI® coverage, we can arrange an appointment at your convenience, or you can visit the UltraCare website. You can also find the UltraCare Enrolment Form on the UltraCare website.