SENTARA HEALTH PLANS

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 can be delayed.**

Drug Requested: Ctexli™ (chenodiol)

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Sentara #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

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

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

Quantity Limit: 3 tablets per day

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: 6 months

- ❑ Member is 18 years of age or older
- ❑ Prescribed by or in consultation with a physician experienced in treatment of cerebrotendinous xanthomatosis (CTX) (e.g., neurologist, endocrinologist, hepatologist, gastroenterologist or other metabolic specialist)
- ❑ Member has a diagnosis of cerebrotendinous xanthomatosis (CTX) confirmed by genetic testing **(must submit test results documenting a pathogenic variant in the CYP27A1 gene)**

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- ❑ Member has symptoms of CTX which include at least **ONE** of the following **(check all that apply)**:
  - ❑ Neonatal cholestasis
  - ❑ Infantile-onset diarrhea
  - ❑ Juvenile cataracts
  - ❑ Adolescent- to young adult-onset tendon xanthomas
  - ❑ Adult-onset progressive neurologic dysfunction (e.g., dementia, psychiatric disturbances, pyramidal and/or cerebellar signs, or seizures)
- ❑ Members' lab test results taken within the last 30 days must be submitted to document **ALL** the following:
  - ❑ alanine aminotransferase (ALT)
  - ❑ aspartate aminotransferase (AST)
  - ❑ total bilirubin
  - ❑ urine 23S-pentol (ng/mL – must be above normal limits per reference range)
  - ❑ plasma cholestanol (µg/mL – must be above normal limits per reference range)
- ❑ Provider attests member will receive liver function monitoring at least annually due to the potential for hepatotoxicity during treatment with Ctexli
- ❑ Member will **NOT** use Ctexli™ (chenodiol) with medications that impact bile acid absorption or synthesis (e.g., cholestyramine, colestipol, colesevelam, aluminum-based antacids, Cholbam (cholic acid), Iqirvo (elafibranor), Livdelzi (seladelpar), Ocaliva (obeticholic acid), Bylvay (odevixibat) or Livmarli (maralixibat))

Reauthorization: 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.

- ❑ Member continues to meet **ALL** initial authorization criteria
- ❑ Member must have reduction in urine 23S-pentol and plasma cholestanol from baseline after initial approval, and reduction or stabilization in 23S-pentol and plasma cholestanol after subsequent approvals **(current lab test results must be submitted for documentation)**
- ❑ Member has experienced an absence of unacceptable toxicity from requested medication (e.g., hepatotoxicity)

Medication being provided by Specialty Pharmacy – Proprium Rx

*****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.****

***Approved by Pharmacy and Therapeutics Committee: 5/22/2025**
REVISED/UPDATED/REFORMATTED: 06/09/2025