Commercial PA Criteria

Effective: May 2017

Prior Authorization: Chenodial Products

Products Affected: Chenodal (chenodiol) oral tablets, Ctexli oral tablets

Medication Description:

Gallstones

The most widely used treatment for symptomatic gallstones is cholecystectomy.[3] Two naturally occurring bile acids are used in the treatment of gallstones: ursodeoxycholic acid (UrsoForte®, Urso-250®, [ursodiol tablets, generic], Actigall® [ursodiol capsules, generic]) and chenodeoxycholic acid/chenodiol (Chenodal). These agents reduce biliary cholesterol; however, their exact mechanisms differ. Both Chenodal and ursodiol promote the gradual dissolution of radiolucent gallstones over a period of 6 months to 2 years.[3]

Cerebrotendinous xanthomatosis (CTX)

CTX is a lipid storage and bile acid synthesis disorder with various clinical manifestations including juvenile cataracts, tendon xanthomas, premature atherosclerosis, and progressive neurologic disturbance (e.g., ataxia, seizures, psychiatric disorders, and peripheral neuropathy).[5] Other conditions associated with CTX include osteoarthritis, skeletal fractures, pulmonary insufficiency, renal and hepatic calculi, and childhood chronic diarrhea. CTX is caused by pathogenic variants in the cytochrome P450 (*CYP)27A1* gene.[7] This gene encodes for sterol 27-hydroxylase, an enzyme responsible for the conversion of cholesterol to cholic acid and chenodeoxycholic acid (primary bile acids). Mutations in this gene lead to 27-hydroxylase deficiency and a subsequent reduction in primary bile acid synthesis. In CTX, reduced synthesis of cholic and chenodeoxycholic acids results in failed feedback inhibition of cholesterol production, in turn leading to hallmark laboratory findings of the disorder: increased serum cholestanol, a cholesterol metabolite, and elevated urinary bile alcohols, like 23S-pentol.[6] Replacement therapy with chenodiol inhibits abnormal bile acid synthesis and is most effective in reducing elevated plasma cholestanol concentrations and eliminating bile alcohols.[5] As such, a CTX expert treatment panel concluded that treatment with chenodiol is necessary to improve/stabilize prognosis in the majority of patients, regardless of age or the presence of symptoms.[7] Alongside the clinical manifestations, biochemical and molecular genetic tests are typically used to diagnose CTX. Diagnostic biochemical tests include detection of elevated serum cholestanol levels while genetic tests include identification of pathogenic variants in the *CYP27A1* gene.

Covered Uses:

1. Chenodiol is indicated in high surgical-risk adults (due to systemic disease or age) with radiolucent stones in well-opacifying gallbladders.
2. Ctexli is indicated for the treatment of cerebrotendinous xanthomatosis (CTX) in adults.

Exclusion Criteria:

Chenodial

1. Gallstone complications, including need for surgery (e.g., unremitting acute cholecystitis, cholangitis, biliary obstruction, gallstone pancreatitis, or biliary gastrointestinal fistula)
2. Hepatocyte dysfunction or bile ductal abnormalities (e.g., intrahepatic cholestasis, primary biliary cirrhosis, or sclerosing cholangitis)
3. Non-visualizing gallbladder, after 2 consecutive single doses of dye
4. Pregnancy

5. Radiopaque stones

Required Medical Information:
1. Medical history
2. Previous therapies tried and failed
3. Current medication regimen

Age Restrictions: 18 years of age or older

Prescriber Restrictions: N/A

Coverage Duration: 12 months

Other Criteria:
I. Coverage of **Chenodal** is recommended in those who meet the following criteria:
1. Gallstones. Approve for 1 year if the patient meets ONE of the following (A **OR** B):
A) Patient has tried an ursodiol product; **OR**
B) Patient is currently receiving an ursodiol product.

II. Coverage of **Ctexli** is recommended in those who meet the following criteria:
1. Cerebrotendinous Xanthomatosis. Approve for 1 year if the patient meets BOTH of the following (A **AND** B):
A) The diagnosis is established by ONE of the following (i **OR** ii):
i. Patient has a molecular genetic test demonstrating a pathogenic variant in the cytochrome P450 27A1 *(CYP27A1)* gene; **OR**
ii. Patient has a laboratory test demonstrating elevated serum cholestanol levels; **AND**
B) The medication is prescribed by or in consultation with a geneticist, neurologist, ophthalmologist, metabolic specialist who treats patients with cerebrotendinous xanthomatosis or a specialist who focuses in the treatment of cerebrotendinous xanthomatosis.

Renewal Criteria
1. Member has responded positively to therapy according to the prescriber; **AND**
2. Member has not experienced unacceptable toxicity from the medication

References:

1. Chenodal™ tablets [prescribing information]. San Diego, CA: Travere; May 2021.
2. Ctexli™ tablets [prescribing information]. Foster City, CA: Mirum; February 2025.
3. Gaby AR. Nutritional approaches to prevention and treatment of gallstones. *Altern Med Rev.* 2009;14(3):258-267.
4. Abraham S, Rivero HG, Erlikh IV, Griffith LF, and Hondamudi VK. Surgical and nonsurgical management of gallstones. *Am Fam Physician*. 2014;89(10):795-802.
5. Moghadasian MH, Salen G, Frohlich JJ, et al. Cerebrotendinous xanthomatosis. *Arch Neurol*. 2002;59:527-529.
6. Lorincz MT, Rainier S, Thomas D and Fink JK. Cerebrotendinous xanthomatosis: possible higher prevalence than previously recognized. *Arch Neurol.* 2005;62:1459-1463.
7. Stelten B, Dotti M., Verrips A, et al. Expert opinion on diagnosing, treating and managing patients with cerebrotendinous xanthomatosis (CTX): a modified Delphi study. *Orphanet J Rare Dis* 16, 353 (2021). Available at: https://doi.org/10.1186/s13023-021-01980-5. Accessed on: March 12, 2025.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	05/2017
2	Annual Review	Updated to new template Added Coverage Duration: Safety of use beyond 24 months is not established.	All	04/22/2020
3	Update	The policy name was changed from "Chenodal" to "Chenodiol Products," with the addition of Ctexli tablets to the policy. Also, divided criteria based on specific agent intended for approval.	All	04/30/2025