

Drug and Biologic Coverage Policy



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Chenodiol

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Related Coverage Resources

INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This policy supports medical necessity review for chenodiol tablets (**Chenodal™**).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Chenodiol (Chenodal) is considered medically necessary when ONE of the following is met:

- 1. Gallstones.** Individual meets **ALL** of the following criteria:
 - A. Documented presence of radiolucent stones in well-opacifying gallbladder
 - B. Not a candidate for surgery (for example, increased surgical risk due to systemic disease or age)
 - C. Documentation of failure, contraindication, or intolerance to ursodiol capsules **OR** tablets used for a minimum of 6 months
- 2. Cerebrotendinous Xanthomatosis.** Individual meets **ALL** of the following criteria:

- A. Documentation of molecular genetic test demonstrating biallelic pathogenic variants in the *CYP27A1* gene
 - B. Medication is prescribed by, or in consultation with, a neurologist, a metabolic disease specialist or a specialist who focuses in the treatment of cerebrotendinous xanthomatosis
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When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Continuation of chenodiol (Chenodal) is considered medically necessary for **ALL** covered diagnoses when the above medical necessity criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration is up to 12 months.
Reauthorization approval duration is up to 12 months.

Conditions Not Covered

Any other use is considered experimental, investigational or unproven, including the following (this list may not be all inclusive):

1. **Combination Therapy with Cholbam™ (cholic acid capsules).** There are no efficacy data available to support use of combination therapy with Chenodal and Cholbam.
2. **Total Treatment Duration Exceeding 24 months for Gallstones.** There are no safety and efficacy data available to support use beyond 24 months.

Background

OVERVIEW

Chenodal, a naturally occurring bile acid, is indicated for patients with **radiolucent stones** in well-opacifying gallbladders, in whom selective surgery would be undertaken except for the presence of increased surgical risk due to systemic disease or age.¹

The most widely used treatment for symptomatic gallstones is cholecystectomy.² 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).³ 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.²

Other Uses with Supportive Evidence

Cerebrotendinous xanthomatosis (CTX) is a lipid storage 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).⁴ Other conditions associated with CTX include osteoarthritis, skeletal fractures, pulmonary insufficiency, renal and hepatic calculi, and childhood chronic diarrhea. CTX is the result of a mutated enzyme (cytochrome P450 27-sterol hydroxylase) which is normally responsible for the conversion of cholesterol to cholic acid and chenodeoxycholic acid. 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 cholesterol concentrations and elevated urinary bile alcohols.⁵

Replacement therapy with chenodiol inhibits abnormal bile acid synthesis and is most effective in reducing elevated plasma cholesterol concentrations and eliminating bile alcohols.⁴

References

1. Chenodal™ tablets [prescribing information]. San Diego, CA: Travers; May 2021.
2. Gaby AR. Nutritional approaches to prevention and treatment of gallstones. *Altern Med Rev.* 2009;14(3):258-267.
3. 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.
4. Moghadasian MH, Salen G, Frohlich JJ, et al. Cerebrotendinous xanthomatosis. *Arch Neurol.* 2002;59:527-529.
5. Lorincz MT, Rainier S, Thomas D and Fink JK. Cerebrotendinous xanthomatosis: possible higher prevalence than previously recognized. *Arch Neurol.* 2005;62:1459-1463.

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