AvMed

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 1-305-671-0200**. 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: Cholbam® (cholic acid)

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

Member Name: _____

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

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

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

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

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

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

Weight: _____ **Date:** _____

Recommended Dosage: Oral: 10 to 15 mg/kg (once daily or in 2 divided doses); administer 11 to 17 mg/kg (once daily or in 2 divided doses) in patients with concomitant familial hypertriglyceridemia

Quantity Limits:

- 50 mg – 4 capsules per day
- 250 mg – 7 capsules 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.

☐ **Bile Acid Synthesis Disorders due to Single Enzyme Defects (SEDs)**

Initial Authorization: 6 months

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- ❑ Member is 3 weeks of age or older
- ❑ Diagnosis has been confirmed using mass spectrometry (FAB-SM) of serum or urinary bile acid levels
- ❑ Member has a diagnosis of **ONE** of the following single enzyme defects:
  - ❑ 3-beta-hydoxysteroid dehydrogenase (3-β-HSD) deficiency
  - ❑ Aldo-keto reductase 1D1 (AKR1D1)
  - ❑ Cerebrotendinous xanthomatosis (CTX)
  - ❑ Alpha-methylacyl-CoA racemase (AMACR) deficiency
- ❑ Member is **NOT** receiving treatment for extrahepatic manifestations of bile acid synthesis disorders (i.e. neurologic symptoms)
- ❑ Assessment of liver function (AST, ALT & bilirubin) has been performed initially and will be performed with each renewal **(submit lab results)**
- ❑ Member will **NOT** be on concomitant therapy with Bile Salt Efflux Pump (BSEP) Inhibitors (e.g., cyclosporine), or if therapy is unavoidable, member will be monitored closely for adverse reactions

❑ Peroxisomal Disorders (PDs) Including Zellweger Spectrum Disorders

Initial Authorization: 6 months

- ❑ Member is 3 weeks of age or older
- ❑ Diagnosis has been confirmed by **ONE** of the following molecular and biochemical findings:
  - ❑ Detection of abnormalities using mass spectrometry (FAB-MS) of serum or urinary bile acid levels
  - ❑ Detection of pathogenic variants of the PEX gene by molecular genetic testing
- ❑ Member has a diagnosis of **ONE** of the following:
  - ❑ Neonatal Adrenoleukodystrophy
  - ❑ Generalized Peroxisomal Disorder
  - ❑ Refsum Disease
  - ❑ Zellweger Syndrome
  - ❑ Peroxisomal Disorder, Type Unknown
- ❑ Member exhibits at least **ONE** or more of the following:
  - ❑ Manifestations of liver disease
  - ❑ Steatorrhea
  - ❑ Complications from decreased fat-soluble vitamin absorption
- ❑ Member is **NOT** receiving treatment for extrahepatic manifestations of bile acid synthesis disorders (i.e., neurologic symptoms)
- ❑ Medication will be used as adjunctive treatment of peroxisomal disorders (PDs)
- ❑ Assessment of liver function (AST, ALT, & bilirubin) has been performed initially and will be performed with each renewal **(submit lab results)**
- ❑ Member will **NOT** be on concomitant therapy with Bile Salt Efflux Pump (BSEP) Inhibitors (e.g., cyclosporine), or if therapy is unavoidable, member will be monitored closely for adverse reactions

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

- ❑ All initial authorization criteria continues to be met
- ❑ Member has experienced disease response as indicated by **ALL** of the following:
  - ❑ Reduction in ALT or AST to less than 50 U/L, or an 80% reduction from baseline
  - ❑ Reduction in total bilirubin to 1 mg/dL or less
  - ❑ Reduction in steatorrhea and/or jaundice
  - ❑ Body weight increased by 10% or remains stable at greater than the 50th percentile
  - ❑ Member has **NOT** developed cholestasis
- ❑ Member has **NOT** experienced unacceptable toxicity from the drug (e.g., exacerbation of liver impairment)

Medication being provided by Specialty Pharmacy - PropriumRx

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**: 7/21/2022
REVISED/UPDATED: ~~8/10/2022;~~ **10/26/2023**