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: Livmarli® (maralixibat)

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:** _____

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:** _____

Provider please note:

- Livmarli® oral solution 9.5 mg/mL should be prescribed for the treatment of ALGS only
- Livmarli® oral solution 19 mg/mL should be prescribed for the treatment of PFIC only

Quantity Limits:

- Livmarli® oral solution 9.5 mg/mL – 3 mL per day
- Livmarli® oral solution 19 mg/mL – 2 mL per day
- Livmarli® oral tablets 10 mg, 15 mg & 20 mg – 2 tablets per day
- Livmarli® oral tablets 30 mg – 1 tablet 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.

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❑ DIAGNOSIS: Cholestatic pruritus due to Alagille syndrome

Recommended Dosage: 380 mcg/kg once daily, taken 30 minutes before a meal in the morning. Start dosing at 190 mcg/kg administered orally once daily; after one week, increase to 380 mcg/kg once daily, as tolerated. The maximum daily dose should not exceed 28.5 mg (3 mL) per day for Livmarli oral solution and 30 mg per day for Livmarli tablets. Select the appropriate product based on the patient's weight and ability to swallow tablets.

9.5 mg/mL LIVMARLI Oral Solution for Patients with ALGS: Volume per Dose (mL) by weight

Patient weight (kg)	Days 1 to 7 (190 mcg/kg once daily)	Beginning day 8 (380 mcg/kg once daily)
	9.5 mg/mL Solution (for ALGS) Volume per Dose (mL)	
5 to 6	0.1	0.2
7 to 9	0.15	0.3
10 to 12	0.2	0.45
13 to 15	0.3	0.6
16 to 19	0.35	0.7
20 to 24	0.45	0.9
25 to 29	0.5	1
30 to 34	0.6	1.25
35 to 39	0.7	1.5
40 to 49	0.9	1.75
50 to 59	1	2.25
60 to 69	1.25	2.5
70 or higher	1.5	3

LIVMARLI Tablets for Patients with ALGS: Dosage by Weight

Patient weight (kg)	Days 1 to 7 (190 mcg/kg once daily)	Beginning day 8 (380 mcg/kg once daily)
Less than 25	Use Oral Solution	Use Oral Solution
25 to 32		10 mg
33 to 43		15 mg
44 to 65	10 mg	20 mg
66 or higher	15 mg	30 mg

Initial Authorization: 6 months

- ❑ Medication is prescribed by or in consultation with a hepatologist, gastroenterologist, cardiologist or a physician who specializes in Alagille syndrome
- ❑ Member is 3 months of age or older
- ❑ Member has been diagnosed with Alagille syndrome

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- ❑ Provider has submitted the results of genetic testing confirming a JAG1 or NOTCH2 deletion or mutation **(submit results)**
- ❑ Provider has submitted clinical confirmation of disease met by **ALL** the following **(submit labs and/or chart notes)**:
  - ❑ Bile duct paucity on liver biopsy
  - ❑ THREE (3) or more of the following major criteria:
    - ❑ Liver/cholestasis
    - ❑ Dysmorphic facies
    - ❑ Heart disease
    - ❑ Axial skeleton/vertebral anomalies
    - ❑ Eye/posterior embryotoxin
- ❑ Member is experiencing evidence of cholestasis confirmed by **TWO** of the following **(submit labs and/or chart notes)**:
  - ❑ Total serum bile acid > 3 x ULN for age
  - ❑ Conjugated bilirubin > 1 mg/dL
  - ❑ Fat soluble vitamin deficiency otherwise unexplainable
  - ❑ GGT > 3 x ULN for age
  - ❑ Intractable pruritus explainable only by liver disease
- ❑ Member has an average daily score >2 on the itch-reported outcome (ItchRO™)
- ❑ Member does **NOT** have any of the following:
  - Surgical interruption of the enterohepatic circulation
  - Liver transplantation
  - Decompensated liver cirrhosis
- ❑ Member has failed an adequate trial, is intolerant to, or has a contraindication to **ONE** of the following **(verified by pharmacy paid claims; documentation of failure as evidenced by labs/ItchRO™ MUST be submitted)**:
  - ❑ ursodeoxycholic acid (ursodiol)
  - ❑ rifampin

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.

- ❑ Provider has submitted documentation of ItchRO™ score decrease from baseline by <1 and serum bile acid decrease
- ❑ Member does **NOT** have any of the following:
  - Surgical interruption of the enterohepatic circulation
  - Liver transplantation
  - Decompensated liver cirrhosis

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❑ DIAGNOSIS: Progressive Familial Intrahepatic Cholestasis

Recommended Dosage: 570 mcg/kg twice daily 30 minutes before a meal. The starting dose is 285 mcg/kg orally once daily in the morning, and should be increased to 285 mcg/kg twice daily, 428 mcg/kg twice daily, and then to 570 mcg/kg twice daily, as tolerated. The maximum daily dose should not exceed 38 mg (2 mL) per day for Livmarli oral solution and 40 mg per day for Livmarli tablets. Select the appropriate product based on the patient's weight and ability to swallow tablets.

19 mg/mL LIVMARLI Oral Solution for Patients with PFIC: Volume per Dose (mL) by Weight

Patient Weight (kg)	285 mcg/kg (once daily titrated to twice daily)	428 mcg/kg (twice daily)	570 mcg/kg (twice daily as tolerated)
	19 mg/mL Solution (for PFIC) Volume per Dose (mL)		
5	0.1	0.1	0.15
6 to 7	0.1	0.15	0.2
8	0.1	0.2	0.25
9	0.15	0.2	0.25
10 to 12	0.15	0.25	0.3
13 to 15	0.2	0.3	0.4
16 to 19	0.25	0.4	0.5
20 to 24	0.3	0.5	0.6
25 to 29	0.4	0.6	0.8
30 to 34	0.45	0.7	0.9
35 to 39	0.6	0.8	1
40 to 49	0.6	0.9	1
50 to 59	0.8	1	1
60 or higher	0.9	1	1

LIVMARLI Tablets for Patients with PFIC: Dosage by Weight

Patient Weight (kg)	285 mcg/kg (twice daily)	428 mcg/kg (twice daily)	570 mcg/kg (twice daily)
Less than 25	Use Oral Solution	Use Oral Solution	Use Oral Solution
25 to 32			15 mg
33 to 43	10 mg	15 mg	20 mg
44 or higher	15 mg	20 mg	20 mg

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Initial Authorization: 6 months

- ❑ Member is 12 months of age or older
- ❑ Prescribed by or in consultation with a hepatologist, gastroenterologist or a physician who specializes in progressive familial intrahepatic cholestasis
- ❑ Member is experiencing pruritus requiring at least medium scratching (≥ 2 on 0-4 scale) according to prescriber **(please submit pruritus assessment)**
- ❑ Diagnosis has been confirmed by genetic testing demonstrating a gene mutation affiliated with progressive familial intrahepatic cholestasis ***Note: Gene mutations affiliated with progressive familial intrahepatic cholestasis include the ATP8B1 gene, ABCB11 gene (BSEP 1 AND BSEP 2)**
- ❑ Member's total serum bile acids ≥ 100 µmol/L **(please submit labs)**
- ❑ Member has failed, is intolerant to, or has a contraindication to at least **ONE** of the following therapies used for the treatment of progressive familial intrahepatic cholestasis **(verified by pharmacy paid claims)**:
  - ❑ cholestyramine
  - ❑ rifampicin
  - ❑ ursodiol
- ❑ Member has failed an adequate trial, is intolerant to, or has a contraindication to Bylvay® (odevixibat) *requires prior authorization* **(verified by pharmacy paid claims; documentation of failure as evidenced by labs/ItchRO™ MUST be submitted)**
- ❑ Member does **NOT** have any of the following medical conditions:
  - Cirrhosis
  - Portal hypertension
  - History of a hepatic decompensation event (e.g., variceal hemorrhage, ascites, and hepatic encephalopathy)
  - Pathologic variations of the ABCB11 gene that predict complete absence of the BSEP protein (BSEP 3 gene)
  - Past medical history or current liver disease (i.e., biliary atresia, benign recurrent intrahepatic cholestasis, liver cancer or metastases, non-PFIC, liver transplant)
  - Chronic Kidney Disease with GFR < 70 mL/min/1.73 m^2
  - Medical history of persistent diarrhea

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 has experienced a reduction in serum bile acids from baseline
- ❑ Member has experienced a decrease of at least 1 in the pruritus scratching score

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- ❑ Member has **NOT** experienced any treatment-restricting adverse effects (e.g., persistent diarrhea; persistent fat-soluble vitamin deficiency despite vitamin A, D, E, K supplementation; elevated liver function tests [alanine aminotransferase (ALT), aspartate aminotransferase (AST), total bilirubin (TB), direct bilirubin (DB)])
- ❑ Member has **NOT** developed decompensated cirrhosis
- ❑ Member has **NOT** developed significant portal hypertension
- ❑ Member has experienced a positive response to therapy, as determined by the prescriber (e.g., decrease in serum bile acids and decrease in pruritus)
- ❑ Prescribed dose must **NOT** exceed FDA approved labeling

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**: ~~1/20/2022; 5/25/2023; 5/23/2024; 9/26/2024;~~ **7/24/2025**
REVISED/UPDATED/REFORMATTED: ~~1/11/2022; 2/24/2022; 3/23/2022; 6/15/2023; 5/17/2024; 6/6/2024; 10/15/2024;~~ **8/22/2025**