

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 Form/Strength: _____

Dosing Schedule: _____ Length of Therapy: _____

Diagnosis: _____ ICD Code: _____

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

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.

| 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 |

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
- ❑ Provider has submitted the results of genetic testing confirming a JAG1 or NOTCH2 deletion or mutation (submit results)

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- 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

| 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 |

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**)

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- 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 $\mu\text{mol/L}$ (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 $\text{GFR} < 70$ 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
- 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

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- 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.****