SENTARA HEALTH PLANS

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 1-800-750-9692.** No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization may be delayed.**

Drug Requested: Livmarli® (maralixibat)

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

Member Name: _____

Member Sentara #: _____ **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:** _____

Weight: _____ **Date:** _____

Dose Volume Recommendations Based on Patient Body Weight

Patient weight	Days 1 to 7 (190 mcg/kg once daily)		Beginning day 8 (380 mcg/kg once daily)	
	Volume (once daily)	Dosing dispenser size	Volume (once daily)	Dosing dispenser size
5 to 6 kg	0.1 mL	0.5 mL	0.2 mL	0.5 mL
7 to 9 kg	0.15 mL	0.5 mL	0.3 mL	0.5 mL
10 to 12 kg	0.2 mL	0.5 mL	0.45 mL	0.5 mL
13 to 15 kg	0.3 mL	0.5 mL	0.6 mL	1 mL
16 to 19 kg	0.35 mL	0.5 mL	0.7 mL	1 mL

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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.

❑ 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)
	Volume Per Dose (mL)	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 (4 mL) per day

Patient Weight (kg)	285 mcg/kg	428 mcg/kg	570 mcg/kg
	Volume Per Dose (mL)	Volume Per Dose (mL)	Volume Per Dose (mL)
10 to 12	0.35	0.5	0.6
13 to 15	0.4	0.6	0.8
16 to 19	0.5	0.8	1
20 to 24	0.6	1	1.25
25 to 29	0.8	1.25	1.5
30 to 34	0.9	1.5	2
35 to 39	1.25	1.5	2
40 to 49	1.25	2	2
50 to 59	1.5	2	2
60 or higher	2	2	2

Initial Authorization: 6 months

- ❑ Member is 5 years 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)**

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- ❑ 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
- ❑ 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

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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**: ~~1/20/2022; 5/25/2023;~~ **5/23/2024**
REVISED/UPDATED/REFORMATTED: ~~1/11/2022; 2/24/2022; 3/23/2022; 6/15/2023; 5/17/2024;~~ **6/6/2024**