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:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

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

Diagnosis: _____ **ICD Code:** _____

Weight: _____ **Date:** _____

Recommended Dosage: 380 mcg/kg once daily, taken 30 minutes before the first meal of the day. Maximum dosage is 760 mcg/kg per day

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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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**
20 to 24 kg	0.45 mL	0.5 mL	0.9 mL	1 mL
25 to 29 kg	0.5 mL	0.5 mL	1 mL	1 mL
30 to 34 kg	0.6 mL	1 mL	1.25 mL	3 mL
35 to 39 kg	0.7 mL	1 mL	1.5 mL	3 mL
40 to 49 kg	0.9 mL	1 mL	1.75 mL	3 mL
50 to 59 kg	1 mL	1 mL	2.25 mL	3 mL
60 to 69 kg	1.25 mL	3 mL	2.5 mL	3 mL
≥70 kg	1.5 mL	3 mL	3 mL	3 mL

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

Initial Authorization: 6 months

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

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- ❑ 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 **TWO** of the following **(verified by pharmacy paid claims; documentation of failure as evidenced by labs/ItchRO™ MUST be submitted)**:
  - ❑ cholestyramine
  - ❑ colesevelam
  - ❑ 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

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 REVISED/UPDATED:** ~~1/11/2022; 2/24/2022; 3/23/2022; 6/15/2023;~~ **10/17/2023**