# SENTARA COMMUNITY PLAN
# (MEDICAID)

## 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 the information provided is not complete, correct, or legible, the authorization process can be delayed.**

**Drug Requested: Bylvay™** (odevixibat)

**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, if applicable:** _____

**Weight:** _____ **Date:** _____

**Quantity Limits:**

- ❑ NDC 74528-020-01: 200 mcg oral pellets: 8 per day; 240 per 30 days
- ❑ NDC 74528-040-01: 400 mcg capsule: 10 per day; 300 per 30 days
- ❑ NDC 74528-060-01: 600 mcg oral pellets: 4 per day; 120 per 30 days
- ❑ NDC 74528-120-01: 1,200 mcg capsule: 5 per day; 150 per 30 days

**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: Progressive Familial Intrahepatic Cholestasis

**Recommended Dosage:**

- ❑ 40 mcg/kg/day. If no improvement in pruritus after 3 months, the dosage may be increased in 40 mcg/kg increments up to 120 mcg/kg/day not to exceed a total daily dose of 6 mg
- ❑ Bylvay oral pellets are for patients weighing <19.5 kg, while the capsules are intended for use by patients weighing ≥ 19.5 kg

| Body weight (kg) | Total daily dose (mcg) for 40 mcg/kg/day | Total daily dose (mcg) for 80 mcg/kg/day | Total daily dose (mcg) for 120 mcg/kg/day |
|---|---|---|---|
| ≤ 7.4 | 200 (1 oral pellet) | 400 (2 oral pellets) | 600 (1 oral pellet) |
| 7.5-12.4 | 400 (2 oral pellets) | 800 (4 oral pellets) | 1200 (2 oral pellets) |
| 12.5-17.4 | 600 (1 oral pellet) | 1200 (2 oral pellets) | 1,800 (3 oral pellets) |
| 17.5-19.4 | 800 (4 oral pellets) | 1600 (8 oral pellets) | 2,400 (4 oral pellets) |
| 19.5-25.4 | 800 (2 capsules) | 1600 (4 capsules) | 2,400 (2 capsules) |
| 25.5-35.4 | 1,200 (1 capsule) | 2,400 (2 capsules) | 3,600 (3 capsules) |
| 35.5-45.4 | 1,600 (4 capsules) | 3,200 (8 capsules) | 4,800 (4 capsules) |
| 45.5-55.4 | 2,000 (5 capsules) | 4,000 (10 capsules) | 6,000 (5 capsules) |
| ≥ 55.5 | 2,400 (2 capsules) | 4,800 (4 capsules) | 6,000 (5 capsules) |

**Initial Authorization: 6 months**

- ❑ Member is 3 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

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- ❑ 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 < 70mL/min/1.73
  - 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 meet but **NOT** exceed **ONE** of the following:
  - ❑ 40 mcg/kg per day, not to exceed the recommended dose and quantity by body weight
  - ❑ 80 mcg/kg per day (up to a maximum of 6 mg per day), and documentation supports no improvement in pruritus after 3 months at a dose of 40 mcg/kg per day
  - ❑ 120 mcg/kg per day (up to a maximum of 6 mg per day), and documentation supports no improvement in pruritus after 3 months at a dose of 80 mcg/kg per day

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❑ **DIAGNOSIS: Cholestatic pruritus due to Alagille syndrome**

**Recommended Dosage:**

- ❑ 120 mcg/kg/day
- ❑ Bylvay oral pellets are for patients weighing <19.5 kg, while the capsules are intended for use by patients weighing ≥ 19.5 kg

| Body weight (kg) | Total daily dose (mcg) for 120 mcg/kg/day |
|---|---|
| ≤ 7.4 | 600 (1 oral pellet) |
| 7.5-12.4 | 1200 (2 oral pellets) |
| 12.5-17.4 | 1,800 (3 oral pellets) |
| 17.5-19.4 | 2,400 (4 oral pellets) |
| 19.5-25.4 | 2,400 (2 capsules) |
| 25.5-35.4 | 3,600 (3 capsules) |
| 35.5-45.4 | 4,800 (4 capsules) |
| 45.5-55.4 | 6,000 (5 capsules) |
| ≥ 55.5 | 7,200 (6 capsules) |

**Initial Authorization: 6 months**

- ❑ Member is 12 months of age or older
- ❑ 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
- ❑ Member has failed an adequate trial, is intolerant to, or has a contraindication to Livmarli® (maralixibat) ***requires prior authorization* (verified by pharmacy paid claims; documentation of failure as evidenced by labs/ItchRO™ MUST be submitted)**

**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.****