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: 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:** _____

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.

(Continued on next page)

❑ 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

(Continued on next page)

- ❑ 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

(Continued on next page)

❑ **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

(Continued on next page)

- ❑ 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.****