BlueCross BlueShield Arizona

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PHARMACY COVERAGE GUIDELINE

BYLVAY™ (odevixibat)
LIVMARLI™ (maralixibat)
LYNAVOY® (linerixibat)
Generic Equivalent (if available)

This Pharmacy Coverage Guideline (PCG):
- Provides information about the reasons, basis, and information sources we use for coverage decisions
- Is not an opinion that a drug (collectively "Service") is clinically appropriate or inappropriate for a patient
- Is not a substitute for a provider's judgment (Provider and patient are responsible for all decisions about appropriateness of care)
- Is subject to all provisions e.g. (benefit coverage, limits, and exclusions) in the member's benefit plan; and
- Is subject to change as new information becomes available.

Scope
- This PCG applies to Commercial and/or Marketplace plans
- This PCG does not apply to the Federal Employee Program, Medicare Advantage, Medicaid or members of out-of-state Blue Cross and/or Blue Shield Plans

Instructions & Guidance
- To determine whether a member is eligible for the Service, read the entire PCG.
- This PCG is used for FDA approved indications including, but not limited to, a diagnosis and/or treatment with dosing, frequency, and duration.
- Use of a drug outside the FDA approved guidelines, refer to the appropriate Off-Label Use policy.
- The "Criteria" section outlines the factors and information we use to decide if the Service is medically necessary as defined in the Member's benefit plan.
- The "Description" section describes the Service.
- The "Definition" section defines certain words, terms or items within the policy and may include tables and charts.
- The "Resources" section lists the information and materials we considered in developing this PCG
- **We do not accept patient use of samples as evidence of an initial course of treatment, justification for continuation of therapy, or evidence of adequate trial and failure.**
- Information about medications that require prior authorization is available at www.azblue.com/pharmacy. You must fully complete the request form and provide chart notes, lab workup and any other supporting documentation. The prescribing provider must sign the form. Fax the form to BCBSAZ Pharmacy Management at (602) 864-3126 or email it to Pharmacyprecert@azblue.com.

Medical Necessity Requirements for BYLVAY (odevixibat), LIVMARLI (maralixibat), and LYNAVOY (linerixibat)

Criteria for Initial Therapy:

Prescriber Qualifications
- Prescribed by a physician specializing in the diagnosis or in consultation with a Gastroenterologist or Hepatologist

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Indication

- **Bylvay:**
  - Treatment of **cholestatic pruritus** in individuals with Alagille syndrome (ALGS)
  - Treatment of **cholestatic pruritus** in individuals with Progressive Familial Intrahepatic Cholestasis (PFIC) type 1
  - Treatment of **cholestatic pruritus** in individuals with Progressive Familial Intrahepatic Cholestasis (PFIC) type 2 that does not have *ABCB11* variant
- **Livmarli:**
  - Treatment of **cholestatic pruritus** in individuals with Alagille syndrome (ALGS)
  - Treatment of **cholestatic pruritus** in individuals with Progressive Familial Intrahepatic Cholestasis (PFIC) types 1, 3, 4, and 6
  - Treatment of **cholestatic pruritus** in individuals with Progressive Familial Intrahepatic Cholestasis (PFIC) type 2 that does not have *ABCB11* variant
- **Lynavoy:**
  - Treatment of **cholestatic pruritus** in individuals with Primary Biliary Cholangitis (PBC)

Age Requirement

- **Bylvay:**
  - Alagille Syndrome (ALGS): 12 months or older
  - Progressive Familial Intrahepatic Cholestasis (PFIC) types 1 and 2: 3 months or older
- **Livmarli:**
  - Alagille Syndrome (ALGS): 3 months or older
  - Progressive Familial Intrahepatic Cholestasis (PFIC) types 1, 2, 3, 4, and 6: 12 months or older
- **Lynavoy:**
  - Primary Biliary Cholangitis (PBC): 18 years of age or older

Baseline Clinical Evaluation

- Diagnosis confirmed by:
  - **For Alagille Syndrome ALGS: ONE** of the following:
    1. *JAG1*, *JAGGED1*, or *NOTCH2* gene mutation
    2. Three or more major clinical features of ALGS: cholestasis, cardiac disease, ocular abnormalities, skeletal abnormalities, characteristic facial features
  - **For Progressive familial intrahepatic cholestasis PFIC: ONE** of the following:
    1. Biallelic pathogenic gene mutations (*ATP8B1, ABCB11, ABCB4, TJP2, MYO5B)*
    2. Clinical signs and symptoms of PFIC (ex., growth failure, progressive liver disease)
  - **For Primary Biliary Cholangitis: TWO** of the following:
    1. Positive antimitochondrial antibodies (AMA) at a titer of 1:40 or more or other primary biliary cholangitis specific autoantibody titer
    2. Presence of cholestasis with alkaline phosphatase at least 1.5 times ULN for more than 6 months
    3. Liver biopsy consistent with primary biliary cholangitis showing chronic nonsuppurative cholangitis of small and medium bile ducts
- Baseline tests completed and will have continued monitoring as clinically appropriate:
  - Liver tests (ALT, AST, total bilirubin, direct bilirubin, INR)
  - Fat soluble vitamins (A, D, E, K) with or without use of supplementation

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- Evidence of cholestasis with **ONE** of the following:
  - Elevated alkaline phosphatase
  - Gamma glutamyl transpeptidase (GGT) greater than three times upper limit of normal for age
  - Direct bilirubin greater than 1 mg/dL
  - Fasting total serum bile acid greater three times upper limit of normal for age
  - Unexplained fat soluble vitamin deficiency
  - Intractable pruritus explainable only by liver disease
- Evaluation of pruritus is **ONE** of the following:
  - **For Alagille Syndrome or Progressive familial intrahepatic cholestasis EITHER** of the following:
    1. Scratch Severity Score greater than or equal to 2 (moderate/medium scratching)
    2. Itch Reported Outcome instrument (ItchRO) greater than or equal to 2 (moderate pruritus)
  - **For Primary Biliary Cholangitis:** Numerical Rating Scale (NRS) Pruritus greater than 3 (moderate pruritus)

Alternative Therapies

- Failure, contraindication per FDA label, intolerance, or is not a candidate for **TWO** of the following:
  - Cholestyramine
  - Naltrexone
  - Phenobarbital
  - Rifampin
  - Sertraline
  - Ursodeoxycholic acid (ursodiol)

Brand Specific Criteria

- Have failure, contraindication or intolerance with **THREE** generic equivalents (if available) for at least three months each. **Note**: Any failure, contraindication, or intolerance to the generic drugs should be reported to the FDA (see Definitions section)

Safety

- **NONE** of the following:
  - Prior or active hepatic decompensation event (variceal hemorrhage, ascites, hepatic encephalopathy)
  - Clinically significant portal hypertension
  - Past medical history or ongoing chronic cirrhosis
  - Decompensated cirrhosis
  - Other liver disease
  - Surgical history of disruption of enterohepatic circulation (biliary diversion surgery) within previous 6 months
  - Liver transplantation or planned within next 6 months
  - Uncontrolled pruritic condition other than PFIC or ALGS
  - PFIC type 2 with *ABCB11* variant resulting in nonfunctioning bile salt export pump protein (BSEP 3)
- Bylvay, Livmarli, and Lynavoy will not be used together

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Documentation Requirements

- A completed request form must be submitted including:
  - Chart notes
  - Lab
  - Supporting clinical documentation

Initial Therapy Criteria Approval Duration

- 6 months OR end of plan year
- **Note**: Livmarli is available as two oral solutions and as a tablet:
  - Oral Solution 9.5 mg/mL for treatment of ALGS
  - Oral Solution 19 mg/mL for treatment of PFIC
  - The two strengths should not be substituted for one another when treating PFIC
  - Tablets can be used for treatment of both ALGS and PFIC in patients weighing 25 kg and above who can swallow tablets

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Criteria for Continuation of Therapy (renewal therapy):

Note: Manufacturer assistance (e.g., coupons, samples, etc.) are not considered for continuation of therapy

Prescriber Qualifications

- Continues to be seen by a physician specializing in diagnosis or in consultation with a Gastroenterologist or Hepatologist

Clinical Response

- Positive response defined as **ALL** the following:
  - Improvement in liver tests (alanine aminotransferase, aspartate aminotransferase, total bilirubin)
  - Reduction in alkaline phosphatase and gamma glutamyl transpeptidase
  - At least 1 point reduction in either daily Itch, Scratch score, or Numerical Rating Scale over baseline
  - Decrease in serum bile acids

Adherence

Adherence to the prescribed therapy regimen has been documented

Age Requirement

- Age is consistent per product label for diagnosis and medication

Brand Specific Criteria

- Have failure, contraindication or intolerance with **THREE** generic equivalents (if available) for at least three months each. **Note**: Any failure, contraindication, or intolerance to the generic drugs should be reported to the FDA (see Definitions section)

Safety

- **NONE** of the following:
  - Prior or active hepatic decompensation event (variceal hemorrhage, ascites, hepatic encephalopathy)

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  - Clinically significant portal hypertension
  - Past medical history or ongoing chronic cirrhosis
  - Decompensated cirrhosis
  - Other liver disease
  - Surgical history of disruption of enterohepatic circulation (biliary diversion surgery) within previous 6 months
  - Liver transplantation or planned within next 6 months
  - Uncontrolled pruritic condition other than PFIC or ALGS
  - PFIC type 2 with *ABCB11* variant resulting in nonfunctioning bile salt export pump protein (BSEP 3)
  - Persistent or recurrent liver test abnormalities (ALT, AST, total bilirubin, direct bilirubin, INR)
  - Persistent diarrhea, abdominal pain, vomiting without alternative cause
  - Persistent or worsening fat soluble vitamin deficiency despite supplementation
  - Concomitant drugs that cause adverse reaction or drug interaction
  - **Additionally for Livmarli only:** suspected propylene glycol toxicity in children under 5 years (hemolysis, hyperosmolarity with anion gap metabolic acidosis, acute kidney injury, and CNS toxicity)
- Bylvay, Livmarli, and Lynavoy will not be used together

Documentation Requirements

- Chart notes
- Lab values confirming safe use
- Supporting clinical documentation with evidence of improvement

Continuation Therapy Criteria Approval Duration

- 12 months OR end of plan year
- **Note**: Livmarli is available as two oral solutions and as a tablet:
  - Oral Solution 9.5 mg/mL for treatment of ALGS
  - Oral Solution 19 mg/mL for treatment of PFIC
  - The two strengths should not be substituted for one another when treating PFIC
  - Tablets can be used for treatment of both ALGS and PFIC in patients weighing 25 kg and above who can swallow tablets

Criteria for Off-Label Use Requests:

Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

1. Off-Label Use of Non-Cancer Medications

2. Off-Label Use of Cancer Medications

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Description:

Livmarli (maralixibat) is a reversible inhibitor of the ileal bile acid transporter (IBAT) indicated for the treatment of cholestatic pruritus in patients with Alagille syndrome (ALGS) who are 3-months of age and older. It is also indicated for the treatment of cholestatic pruritus in patients 12 months of age and older with progressive familial intrahepatic cholestasis (PFIC). Livmarli (maralixibat) is not recommended in a subgroup of PFIC type 2 patients with specific *ABCB11* variants resulting in non-functional or complete absence of bile salt export pump (BSEP) protein. Livmarli (maralixibat) decreases the reabsorption of bile acids (primarily the salt forms) from the terminal ileum. Although the complete mechanism by which maralixibat improves pruritus in ALGS and PFIC patients is unknown, it may involve inhibition of the IBAT, which results in decreased reuptake of bile salts, as observed by a decrease in serum bile acids.

Bylvay (odevixibat) is a reversible inhibitor of IBAT, is indicated for the treatment of pruritus in patients 3-months of age and older with progressive familial intrahepatic cholestasis (PFIC) and for the treatment of cholestatic pruritus in patients with ALGS in patients 12-months of age and older. Bylvay (odevixibat) may not be effective in PFIC type 2 patients with ABCB11 variants resulting in non-functional or complete absence of bile salt export pump protein (BSEP-3). Odevixibat decreases the reabsorption of bile acids (primarily the salt forms) from the terminal ileum. Although the complete mechanism by which odevixibat improves pruritus in patients is unknown, it may involve inhibition of the IBAT, which results in decreased reuptake of bile salts, observed by a decrease in serum bile acids.

Lynavoy (linerixibat) is a reversible ileal bile acid transporter (IBAT) inhibitor indicated for the treatment of cholestatic pruritus associated with primary biliary cholangitis (PBC) in adult patients. is an ileal bile acid transporter (IBAT) inhibitor indicated for the treatment of cholestatic pruritus associated with primary biliary cholangitis (PBC) in adult patients.

ALGS is characterized by chronic cholestasis with lack of the interlobular bile ducts on liver biopsy. Associated features seen in most patients include cardiac anomalies, butterfly vertebrae, posterior embryotoxon of the eye, and characteristic facial features.

ALGS is an autosomal dominant inherited condition. The syndrome is also known as syndromic lack of the interlobular bile ducts or arteriohepatic dysplasia. The clinical diagnosis of ALGS in an infant with cholestasis includes the characteristic clinical features and a liver biopsy demonstrating reduced number of the interlobular bile ducts. In patients with clinical characteristics suggestive of ALGS, the diagnosis can also be made or confirmed by the finding of a *JAG1(*or *JAGGED1)* or *NOTCH2* gene mutation.

Medical management of patients with ALGS depends on establishing the diagnosis and treating each affected organ system. Pruritus is a common symptom in patients with ALGS. It occurs in over 60% of patients. The pathophysiology of pruritus in patients with ALGS is not completely understood. Cholestasis is seen with many hepatobiliary disorders. One difficult symptom associated with cholestasis is pruritus, which can range in severity from mild to extreme.

Pruritus may be treated with ursodeoxycholic acid (ursodiol), rifampin, bile acid sequestrants (such as cholestyramine, colesevelam), naltrexone, sertraline, and intestinal bile acid transport (IBAT) inhibitors.

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Initial studies of the IBAT inhibitor maralixibat have shown some benefit. Pruritus was assessed using Itch Reported Outcome (ItchRO[Obs]) measure, using an electronic diary (eDiary) completed by the patient or caregiver twice daily (morning and evening). ItchRO(Obs) score ranges from 0 to 4, with the higher score indicating increasing itch severity. The highest score between the morning and evening ItchRO(Obs) reports represented the daily score: a measure of the worst itching over the previous 24-hour period. Approximately 40% of the patients with ALGS and pruritus, the pruritus is refractory to medical treatment. In these cases, biliary diversion or liver transplantation may be indicated.

Progressive familial intrahepatic cholestasis (PFIC) is a heterogeneous group of disorders caused by homozygous or compound heterozygous variants and is characterized by defective secretion of bile acids or other components of bile. It presents in infancy or childhood and is associated with growth failure and progressive liver disease.

There are several types of PFIC involving several gene mutations. PFIC1 is caused by variants in the aminophospholipid flippase (*ATP8B1*) gene, which encodes the familial intrahepatic cholestasis 1 (FIC1) protein, while PFIC2 results from variants in the *ABCB11* gene, which encodes the bile salt export pump (BSEP) protein. PFIC2 is further categorized into BSEP subgroups based on specific variants. The BSEP-1 subgroup includes patients with at least one p.D482G (c.1445A>G) or p.E297G (c.890A>G) variant, BSEP-2 includes patients with at least one missense variant other than p.D482G or p.E297G (non BSEP-1), and BSEP-3 includes patients with variants that are predicted to encode a non-functional protein. PFIC3 is caused by variants in the *ABCB4* gene, which encodes multidrug resistance protein 3 (*MDR3*). PFIC4 is caused by variants in the tight junction protein 2 gene (*TJP2*), which encodes TJP2. PFIC5 is caused by variants in *NR1H4* gene which encodes for nuclear receptor subfamily 1 group h member 4. PFIC6 is caused by variants in myosin 5B (*MYO5B*), which encodes MYO5B.

Intractable pruritus is a dominant feature of PFIC types 1 and 2 and both are associated with life-threatening cholestasis. In all forms of PFIC, use of ursodeoxycholic acid, may improve liver function in some patients, and may relieve the pruritus associated with the disorder.

Definitions:

U.S. Food and Drug Administration (FDA) MedWatch Forms for FDA Safety Reporting
MedWatch Forms for FDA Safety Reporting | FDA

Major clinical criteria/features for Alagille syndrome include:

- Liver biopsy demonstrating reduced number of the interlobular bile ducts
- Cholestasis, cardiac, renal, vascular, ocular, skeletal involvement, or characteristic “Alagille” facies

Hepatic	Bile duct paucity, chronic cholestasis, jaundice, conjugated hyperbilirubinemia, cirrhosis, pruritus, xanthomas, hepatic osteodystrophy, fat soluble vitamin deficiencies, portal hypertension, end-stage liver disease
Cardiac	Murmur or cardiac anomalies. The most common finding is peripheral pulmonary stenosis/hypolasia; other anomalies include tetralogy of Fallot, ductus arteriosus, septal defects, or coarctation of the aorta

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Vascular	Intracranial bleeds: epidural, subdural, subarachnoid and intraparenchymal, systemic vascular anomalies: aorta, kidney, celiac, superior mesenteric and subclavian arteries, Moyamoya syndrome
Renal	Renal involvement (19 to 74 percent). The most common finding is renal dysplasia; other disorders include glomerular mesangiolipidosis or renal tubular acidosis, ureteropelvic obstruction
Vertebral/Skeletal	Butterfly vertebrae, occasionally hemivertebrae, fusion of adjacent vertebrae, and spina bifida occulta
Facies	Dysmorphic facies, consisting of broad nasal bridge, triangular facies (broad forehead and pointed chin), and deep-set eyes with moderate hypertelorism
Ocular	Most commonly posterior embryotoxon (prominent Schwalbe line). Other findings may include optic disc abnormalities, hypopigmentation of the peripheral retina, pseudopapilledema, and true papilledema (associated with intracranial hypertension)
Ears	Hearing loss

Alagille facies:

- Facial features are triangular, with a prominent, broad forehead, deep-set eyes, prominent ears, a pointed chin, and a straight nose with a bulbous tip
- The facial features change over time, with a more triangular shape during childhood and prominence of the jaw as patients reach adulthood

Scratch Severity Score:
The Clinician Scratch Scale (CSS) uses a 0-to-4-point rating system to measure physical signs and observable severity of scratching.
The 0 to 4 Scratch Severity Score:

- **0 — None:** No evidence of scratching or scratch marks
- **1 — Mild:** Rubbing or mild scratching only when the patient is undistracted
- **2 — Moderate:** Active scratching present, but without visible skin abrasions
- **3 — Severe:** Abrasions or excoriations (scratched skin) are clearly evident
- **4 — Very Severe:** Cutaneous mutilation with significant hemorrhage (bleeding) and scarring

Itch Reported Outcome (ItchRO):

- Can be completed by the patient (ItchRO[Pt]) or a caregiver/observer (ItchRO[Obs])
- It is performed in the morning and evening
- The daily score is the highest (worst) score from the morning and evening reports
- Average daily score is the sum of all daily scores divided by the number of days the ItchRO was completed

Completed by the patient (ItchRO[Pt]) in the morning and evening				
I didn't feel itchy	I felt a little bit itchy	I felt pretty itchy	I felt very itchy	I felt very, very itchy
0	1	2	3	4

Completed by caregiver/observer (ItchRO[Obs]) in the morning and evening				
Not itchy at all	A little bit itchy	Somewhat/moderately itchy	Very itchy	Extremely itchy

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0	1	2	3	4

Numerical Rating Scale (NRS) – Pruritus:
The NRS is comprised of one item and represents the numbers 0 ("no itch") to 10 ("worst imaginable itch"). Subjects are asked to rate the intensity of their itch using this scale. It features high reliability and concurrent validity and is a popular choice for all patients due to its simple format.
Time needed for completion: 1 minute

- Validated in several languages
- It can be interpreted as follows:
  NRS 0 - no pruritus
  NRS <3 - mild pruritus
  NRS ≥ 3< 7 - moderate pruritus
  NRS ≥ 7< 9 - severe pruritus
  NRS ≥ 9 - very sever pruritus

Numerical Rating Scale (NRS)	
On a scale of "no itch" (0) to "worst imaginable itch" (10):	Select a number that matches your itch
• How was your itching on average in the last 24 hours?	0, 1, 2, 3, 4, 5, 6, 7, 8, 9, 10
• How was your itching at its most intense in the last 24 hours?	0, 1, 2, 3, 4, 5, 6, 7, 8, 9, 10

Resources:

Bylvay (odevixibat) cap & oral pellets product information, revised by Albireo Pharma, Inc. 12-2025. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed August 17, 2026.

Livmarli (maralixibat) tab & oral solution product information, revised by Mirum Pharmaceuticals, Inc. 04-2025. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed August 17, 2026.

Lynavoy (linerixibat) tab product information, revised by Intercept Pharmaceuticals. 04-2026. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed August 17, 2026.

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Wehrman A, Loomes KM. Causes of cholestasis in neonates and young infants. In: UpToDate, Abrams SA, Rand EB, Hoppin AG (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Literature current through August 2026. Topic last updated September 05, 2025. Accessed September 04, 2026.

Hirschfield GM, Bowlus CL, Jones DEJ, et al. Linerixibat in patients with primary biliary cholangitis and cholestatic pruritus (GLISTEN): a randomized, multicenter, double-blind, placebo-controlled, phase 3 trial. Lancet Gastro Hepatol 2026 Jan;11:22-33; published online Oct 28, 2025. https://doi.org/10.1016/S2468-1253(25)00192-X. Accessed September 06, 2026.

Shneider BL, Spino C, Kamath BM, et al.: Placebo-controlled randomized trial of an Intestinal Bile Salt Transport Inhibitor for pruritus in Alagille Syndrome. Hepatology Communications; 2018; 2(10): 1184-1198. Accessed October 22, 2021. Re-evaluated September 04, 2026.

Kamath BM, Abetz-Webb L, Kennedy C, et al.: Development of a novel tool to assess the impact of itching in pediatric cholestasis. Patient 2017 July 14; DOI 10.1007/s40271-017-0266-4. Accessed October 23, 2021. Re- evaluated September 10, 2025.

Thompson RJ, Arnell H, Artan R, et al.: Odevixibat treatment in progressive familial intrahepatic cholestasis: a randomized, placebo-controlled, phase 3 trial. Lancet Gastroenterol Hepatol 2022; 7: 830–42. https://doi.org/10.1016/ S2468-1253 (22) 00093-0. Access June 18, 2023. Re- evaluated September 04, 2026.

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