CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Bylvay (odevixibat)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Bylvay, approved by the FDA in July 2021, is indicated for the treatment of pruritus in patients 3 months of age and older with progressive familial intrahepatic cholestasis (PFIC). It is the first drug approved for PFIC and is available as oral pellets or capsules, taken once daily.
PFIC is an ultra-rare group of genetic disorders that disrupt bile formation in the liver. It usually presents during infancy and is characterized by cholestasis, jaundice, and intense itching. Most patients will eventually require biliary diversion surgery or liver transplant. PFIC types 1, 2, and 3 are the most common and types 1 and 2 are the most severe. PFIC1 involves extrahepatic manifestations while PFIC2 does not. However, PFIC2 can be complicated by hepatocellular carcinoma.
As of June 2023, Bylvay is also approved for the treatment of cholestatic pruritus in patients 12 months of age and older with Alagille Syndrome (ALGS), a rare genetic disorder that can affect multiple organ systems, most commonly the liver, with a paucity of interlobular ducts.
In cholestatic liver disease, biliary substances aren't eliminated from the liver, thus they re-enter circulation. Cholestatic itch is thought to be related to the accumulation of bile acids in the skin. Bylvay is a reversible inhibitor of the ileal bile acid transporter (IBAT). Inhibiting IBAT decreases reuptake of bile salts.

Bylvay (odevixibat) will be considered for coverage when the following criteria are met:

Progressive familial intrahepatic cholestasis (PFIC)

For **initial** authorization:

1. Member is at least 3 months of age; AND
2. Medication must be prescribed by or in consultation with a gastroenterologist or hepatologist; AND
3. Member has a diagnosis of PFIC type 1 or 2 confirmed by genetic testing identifying pathogenic mutations of the ATP8B1 or ABCB11 genes (results must be provided for review); AND
4. Member has significant pruritis not attributed to another cause; AND
5. Documentation of serum bile acid level ≥ 100 µmol/L; AND
6. Documentation of baseline liver function tests (e.g., ALT, AST, bilirubin, INR); AND
7. Trial and failure of ursodiol (may also continue concurrently); AND
8. Member does NOT have any of the following:
   a) Decompensated cirrhosis
   b) Variants of the ABCB11 gene (PFIC type 2) that code for non-functional or complete absence of the bile salt export pump (BSEP-3) protein (per submitted genetic test result)
   c) Biliary diversion surgery in the past 6 months
   d) Liver transplant
9. **Dosage allowed/Quantity limit:** 40 mcg/kg orally once daily. If no improvement after 3 months, may increase in 40 mcg/kg increments up to 120 mcg/kg. Max dose per day 6 mg.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes must show improvement of pruritis compared to baseline; AND
2. Chart notes must show reduced serum bile acid from baseline; AND
3. Member has NOT experienced portal hypertension or a hepatic decompensation event (e.g., variceal hemorrhage, ascites, hepatic encephalopathy) while being treated with Bylvay.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Alagille Syndrome (ALGS)

For **initial** authorization:
1. Member is at least 12 months of age; AND
2. Medication must be prescribed by or in consultation with a gastroenterologist or hepatologist; AND
3. Member has a diagnosis of Alagille syndrome (ALGS) confirmed by the involvement of at least 3 of the following major clinical features:
   a) Hepatic (e.g., hyperbilirubinemia, cholestasis, xanthomas)
   b) Cardiac (e.g., heart murmur, peripheral pulmonic stenosis)
   c) Facial (e.g., inverted triangular face)
   d) Ocular (e.g., embryotoxon, optic disk drusen)
   e) Skeletal (e.g., butterfly vertebrae)
   f) Renal (e.g., renal dysplasia, renal tubular acidosis)
   g) Vascular (e.g., neurovascular accident, moyamoya disease)

NOTE: Member also meets criterion if has one or more clinical features and an affected first-degree relative; AND
4. Member must have liver biopsy demonstrating reduced number of the interlobular bile ducts OR confirmed finding of JAG1 or NOTCH2 gene mutation; AND
5. Member has moderate to severe pruritus; AND
6. Member's serum bile acid (sBA) level is elevated; AND
7. Member does NOT have any of the following:
   a) Previous liver transplant
   b) Decompensated cirrhosis; AND
8. Member must have a trial and failure of at least 2 of the following:
   a) Cholestyramine
   b) Ursodiol
   c) Rifampin
   d) Naltrexone; AND
9. Documented rationale why Livmarli cannot be used instead of Bylvay; AND
10. Documentation of baseline liver function tests (e.g., ALT, AST, bilirubin, INR).
11. **Dosage allowed/Quantity limit:** 120 mcg/kg taken orally once daily.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Pruritis has improved in response to therapy with Bylvay; AND
2. Member has NOT experienced portal hypertension or a hepatic decompensation event (e.g., variceal hemorrhage, ascites, hepatic encephalopathy) while being treated with Bylvay

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Bylvay (odevixibat) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
11/17/2021	New policy for created for Bylvay.
06/27/2023	Added indication for Alagille Syndrome.

References:

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Effective date: 01/01/2024
Revised date: 06/27/2023