

POLICY NAME	Bylvay (odevixibat)	POLICY #	3176P
--------------------	---------------------	-----------------	-------

Criteria

Coverage Criteria for Pruritus due to Familial Intrahepatic Cholestasis

- ☐ Diagnosis of pruritus (itching) due to progressive familial intrahepatic cholestasis (PFIC)
 - Diagnosis confirmed by genetic testing showing biallelic pathogenic mutations in the ATP8B1 (ie, PFIC1) or ABCB11 (ie, PFIC2) genes
- ☐ Member has cholestasis, as indicated by one of the following:
 - Total serum bile acid $>3 \times$ upper limit of normal (ULN) for age
 - Conjugated bilirubin >2 mg/dL
 - Fat soluble vitamin deficiency that is otherwise unexplainable
 - Gamma Glutamyl Transferase (GGT) $>3 \times$ ULN for age
 - Intractable pruritus explainable only by liver disease
- ☐ Age 3 months or older
- ☐ Prescribed by or in consultation with a hepatologist (liver doctor)
- ☐ Documented concurrent use or previous trial and failure, intolerance or contraindication ursodiol and cholestyramine
- ☐ Review of chart notes documenting diagnosis and confirming that the patient has met all of the above requirements for treatment with Bylvay by both a pharmacist and medical director

Coverage Criteria for Pruritus due to Alagille Syndrome

- ☐ Diagnosis of moderate to severe pruritus due to Alagille syndrome (ALGS)
 - Diagnosis of ALGS confirmed by genetic testing showing pathogenic variants in the JAG1 or NOTCH2 genes
- ☐ Member has cholestasis, as indicated by one of the following:
 - Total serum bile acid $>3 \times$ upper limit of normal (ULN) for age
 - Conjugated bilirubin >2 mg/dL
 - Fat soluble vitamin deficiency that is otherwise unexplainable
 - Gamma Glutamyl Transferase (GGT) $>3 \times$ ULN for age
 - Intractable pruritus explainable only by liver disease
- ☐ Age 12 months or older
- ☐ Prescribed by or in consultation with a hepatologist (liver doctor)

- ☐ Documented trial and failure, contraindication or intolerance to TWO of the following:
 - Ursodiol
 - Rifampin
 - Cholestyramine
 - Sertraline
 - Naltrexone (not for pediatric patients)
- ☐ Review of chart notes documenting diagnosis and confirming that the patient has met all of the above requirements for treatment with Bylvay by both a pharmacist and medical director

Exclusion Criteria – Any of the following prevents coverage

- ☐ Genetic testing indicates PFIC with ABCB11 variants encoding for non-function or absence of BSEP-3
- ☐ Pregnancy
- ☐ Chronic diarrhea requiring consistent fluid or nutritional intervention
- ☐ History of liver transplant or biliary diversion surgery within the past 6 months
- ☐ Evidence of decompensated cirrhosis
- ☐ Concurrent use with Livmarli