JOHNS HOPKINS HEALTH PLANS	Johns Hopkins Health Plans **Pharmacy Public** **Pharmacy Management Drug Policies**	*Policy Number*	MEDS138
		Effective Date	10/20/2021
		Review Date	10/18/2023
	Subject **Bylvay**	*Revision Date*	10/18/2023
		Page	1 of 3

This document applies to the following Participating Organizations:

Priority Partners

Keywords: Bylvay

Table of Contents		Page Number
I.	**POLICY**	**1**
II.	**POLICY CRITERIA**	**1**
	A. Bylvay	**1**
III.	**AUTHORIZATION PERIOD/LIMITATIONS**	**2**
IV.	**EXCLUSIONS**	**2**
V.	**REFERENCES**	**2**
VI.	**APPROVALS**	**3**

I. POLICY

Bylvay (odevixibat) will require prior authorization to ensure appropriate use. The process for initiating a prior authorization request can be found in policy PHARM 20.

1. PPMCO members are subject to the Priority Partners formulary, available at www.ppmco.org.
2. USFHP members are subject to prior authorization criteria, step-edits and days-supply limits outlined in the Tricare Policy Manual. Tricare Policy supersedes JHHC Medical/Pharmacy Policies. Tricare limits may be accessed at: http://pec.ha.osd.mil/formulary_search.php?submenuheader=1

II. POLICY CRITERIA

A. **Bylvay** may be approved for patients meeting the following:

1. **Progressive Familial Intrahepatic Cholestasis (PFIC)**
   a. Patient is 3 months of age or older
   b. Documented diagnosis of PFIC with both of the following:
      I. genetic confirmation of PFIC type 1 or PFIC type 2
      II. elevation of serum bile acid concentration
   c. Documentation showing the patient is experiencing significant pruritis
   d. Documentation of trial and inadequate response to both of the following:
      I. ursodiol (ursodeoxycholic acid)
      II. an agent used for symptomatic relief of pruritus (e.g. cholestyramine, rifampin)
   e. Prescriber is, or has consulted with, a hepatologist or gastroenterologist with experience in treating PFIC
2. **Alagille Syndrome (ALGS)**
   a. Patient is 1 year of age or older
   b. Documentation has been submitted showing the following:
      I. Diagnosis of ALGS, confirmed with genetic testing showing a JAG1 or NOTCH2 mutation
      II. Patient has significant moderate-to-severe pruritus
      III. Evidence of cholestasis, shown by at least one of the following;
         i. Total serum bile acid > 3 times upper limit of normal (ULN) for age
         ii. Conjugated bilirubin > 1 mg/dL

iii. Fat-soluble vitamin deficiency otherwise unexplainable
iv. Gamma-glutamyl transferase > 3 times ULN for age
v. Intractable pruritus explainable only by liver disease

IV. Patient does not have a history or ongoing presence of other types of liver disease (eg. biliary atresia, progressive familial intrahepatic cholestasis, hepatocellular carcinoma)
V. Patient has had trial and inadequate response, or contraindication to both of the following:
i. ursodiol (ursodeoxycholic acid)
ii. an agent used for symptomatic relief of pruritus (e.g. cholestyramine, rifampin)
VI. Prescriber is, or has consulted with, a hepatologist or gastroenterologist with experience in managing ALGS

III. AUTHORIZATION PERIOD/LIMITATIONS

A. Initial therapy may be approved for 12 months
B. Continuation of therapy may be approved in 12-month intervals with documentation showing the patient is tolerating treatment, as well as one of the following:
1. A decrease in pruritis from baseline
2. A decrease in serum bile acid concentration

IV. EXCLUSIONS

A. Bylvay will not be approved for the following:
1. Pediatric patients that are less than 3 months of age
2. Patients with decompensated liver disease
3. Patients with a history of liver transplant
4. Patients with portal hypertension
5. Patients with pathologic variations of the ABCB11 gene that predict complete absence of the BSEP protein
6. Any indications or uses that are not FDA-approved, or guideline-supported
B. The use of physician samples, or manufacturer product discounts, does not guarantee coverage under the provisions of the medical and/or pharmacy benefit. All pertinent criteria must be met in order to be eligible for benefit coverage.

V. REFERENCES

1. Bylvay [prescribing information]. Boston, MA: Albireo Pharma Inc; June 2023.
2. European Association for the Study of the Liver. EASL Clinical Practice Guidelines: management of cholestatic liver diseases. J Hepatol. 2009 Aug;51(2):237-67
3. Davit-Spraul et al. Progressive familial intrahepatic cholestasis. Orphanet J Rare Dis. 2009;4:1.
4. Jacquemin E et al. Ursodeoxycholic acid therapy in pediatric patients with progressive familial intrahepatic cholestasis. Hepatology. 1997;25(3):519.
5. Squires RH. Evaluation of the pediatric patient for liver transplantation: 2014 practice guideline by the American Association for the Study of Liver Diseases, American Society of Transplantation and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Hepatology. 2014;60(1):362.
6. Kremer AE. Advances in pathogenesis and management of pruritus in cholestasis. Dig Dis 2014;32:637-645.
7. Webb GJ, Rahman SR, Levy C, Hirschfield GM. Low risk of hepatotoxicity from rifampicin when used for cholestatic pruritus: a cross-disease cohort study. Aliment Pharmacol Ther. 2018;47(8):1213

8. Roy-Chowdhury J, Roy-Chowdhury, N. Inherited disorders associated with conjugated hyperbilirubinemia. In: UpToDate, Lindor KD, Grover S (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Accessed on September 20, 2021.
9. Turnpenny PD and Ellard S. Alagille syndrome: pathogenesis, diagnosis and management. Eur J Hum Genet. 2012 Mar; 20(3): 251–257.

VI. APPROVALS

Signature on file at JHHC

DATE OF REVISION	SUMMARY OF CHANGE
10/20/2021	Policy Creation
10/18/2023	Updated clinical criteria based on FDA-approved indication expansion

Review Date: 10/20/2021, 10/18/2023

Revision Date: 10/18/2023