Commercial/Healthcare Exchange PA Criteria
Effective: September 14, 2021

Prior Authorization: Bylvay

Products Affected: Bylvay (odevixibat) oral capsule, coated pellets

Medication Description: Odevixibat is a reversible inhibitor of the ileal bile acid transporter (IBAT). It decreases the reabsorption of bile acids (primarily the salt forms) from the terminal ileum.
Pruritus is a common symptom in patients with PFIC and the pathophysiology of pruritus in patients with PFIC is not completely understood. Although the complete mechanism by which odevixibat improves pruritus in 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.

Covered Uses: Progressive intrahepatic cholestasis, Familial – Pruritus

Limitations of use: 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).

Exclusion Criteria: None

Required Medical Information:
1. Diagnosis
2. Medical history

Age Restrictions: 3 months of age and older

Prescriber Restrictions: Prescribed by, or in consultation with, hepatologist, gastroenterologist, or a physician who specializes in progressive familial intrahepatic cholestasis.

Coverage Duration:
Initial approval: 6 months. Continuation: 1 year

Other Criteria:

I. Initial Approval Criteria

1. Pruritus in progressive familial intrahepatic cholestasis (PFIC)
Patient must meet all the below criteria:

A. Patient has moderate-to-severe pruritus, according to prescriber; AND
B. Diagnosis of progressive familial intrahepatic cholestasis was confirmed by genetic testing demonstrating a gene mutation affiliated with progressive familial intrahepatic cholestasis; AND

Note: Gene mutations affiliated with progressive familial intrahepatic cholestasis include the *ATP8B1* gene, *ABCB11* gene, *ABCB4* gene, *TJP2* gene, *NR1H4* gene, and *MYO5B* gene.

C. Patient has a serum bile acid concentration above the upper limit of the normal reference range for the reporting laboratory; AND

D. Patient has tried at least at least one systemic medication for progressive familial intrahepatic cholestasis, unless contraindicated; AND
Note: Systemic medications for progressive familial intrahepatic cholestasis include cholestyramine, rifampicin, and ursodeoxycholic acid (ursodiol).

E. Patient does not have any of the following (a, b, or c):
   i. Cirrhosis; OR
   ii. Portal hypertension; OR
   iii. History of a hepatic decompensation event
   Note: Examples of a hepatic decompensation event include variceal hemorrhage, ascites, and hepatic encephalopathy.

II. Continued Therapy

1. Pruritus in progressive familial intrahepatic cholestasis (PFIC)

A. Patient does not have any of the following (a, b, or c):
   i. Cirrhosis; OR
   ii. Portal hypertension; OR
   iii. History of a hepatic decompensation event; AND
   Note: Examples of a hepatic decompensation event include variceal hemorrhage, ascites, and hepatic encephalopathy.

B. Patient had response to therapy, as determined by the prescriber; AND
Note: Examples of response to therapy include decrease in serum bile acids and decrease in pruritus.

C. Member has not experienced unacceptable toxicity from the drug.

References:

1. Bylvay (odevixibat) [prescribing information]. Boston, MA: Albireo Pharma Inc; Updated July 2021. Accessed August 17, 2021.
2. Bylvay ®. IBM Micromedex® [database online]. Greenwood Village, CO. Truven Health Analytics. Available at: https://www.micromedexsolutions.com. Updated August 18, 2021. Accessed August 25, 2021.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	9/14/2021