

UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2021 P 2251-2
Program	Prior Authorization/Medical Necessity
Medication	Bylvay™ (odevixibat)
P&T Approval Date	10/2021, 12/2021
Effective Date	4/1/2022; Oxford only: 4/1/2022

1. Background:

Bylvay (odevixibat) is an ileal bile acid transporter inhibitor indicated for the treatment of pruritis in patients aged 3 months or older with progressive familial intrahepatic cholestasis (PFIC).

PFIC is a heterogeneous group of liver disorders of autosomal recessive inheritance, characterized by an early onset of cholestasis (usually during infancy) with pruritus and malabsorption, which rapidly progresses and ends up as liver failure. Pruritus is the most obvious and the most unbearable symptom in cholestasis. It has been proposed that it is induced by the stimulation of nonmyelinated subepidermal free nerve ends because of increased serum bile acids. Conventional treatments for pruritis associated with PFIC include: ursodeoxycholic acid, cholestyramine, rifampin, naltrexone and phenobarbital.

Limitation of Use:

Bylvay 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).

2. Coverage Criteria^a:

A. Initial Authorization

1. **Bylvay** will be approved based upon **all** of the following criteria:

a. Confirmed molecular diagnosis of progressive familial intrahepatic cholestasis (PFIC) type 1 or 2.

-AND-

b. Patient does not have a ABCB11 variant resulting in non-functional or complete absence of bile salt export pump protein (BSEP-3)

-AND-

c. Patient is experiencing moderate to severe pruritus associated with PFIC.

-AND-

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

-AND-

- e. Patient has had an inadequate response to at least **two** other conventional treatments for the symptomatic relief of pruritus (e.g., bile acid-binding agents, naltrexone, phenobarbital, rifampin, ursodeoxycholic acid)

-AND-

- f. Prescribed by a hepatologist.

Authorization will be issued for 6 months.

B. Reauthorization

1. **Bylvay** will be approved based on **all** the following criterion:

- a. Documentation of positive clinical response to Bylvay therapy (e.g., reduced serum bile acids, improved pruritis and less sleep disturbance)

-AND-

- b. Prescribed by a hepatologist.

Authorization will be issued for 12 months.

^a State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

4. Reference:

1. Bylvay [package insert]. Boston, MA: Albireo Pharma, Inc.; July 2021.
2. Gunaydin M, Bozkurter Cil AT. Progressive familial intrahepatic cholestasis: diagnosis, management, and treatment. *Hepat Med.* 2018;10:95-104. Published 2018 Sep 10. doi:10.2147/HMER.S137209
3. Baumann U, Sturm E, Lacaille F, et al. Effects of odeixibat on pruritus and bile acids in children with cholestatic liver disease: Phase 2 study [published online ahead of print, 2021 Jun 26]. *Clin Res Hepatol Gastroenterol.* 2021;45(5):101751. doi:10.1016/j.clinre.2021.101751
4. Roy-Chowdhury J, Roy-Chowdhury N. Inherited disorders associated with conjugated hyperbilirubinemia. In: Post TW, ed. *UpToDate.* UpToDate, 2021. Accessed August 26, 2021. <https://www.uptodate.com/contents/inherited-disorders-associated-with-conjugated-hyperbilirubinemia>

5. Poupon R, Chopra S. Pruritus associated with cholestasis. In: Post TW, ed. *UpToDate*. UpToDate, 2021. Accessed August 26, 2021. <https://www.uptodate.com/contents/pruritus-associated-with-cholestasis>

Program	Prior Authorization/Medical Necessity - Bylvay (odevixibat)
Change Control	
10/2021	New program
12/2021	Updated criteria with addition of elevated sBA level requirement.