

Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	December 8, 2021 March 16, 2023, December 20, 2023, May 8, 2024

Ileal Bile Acid Transporter Inhibitor Agents Bylvay[®] (odevixibat) and Livmarli[®] (maralixibat)

LENGTH OF AUTHORIZATION: Up to 6 months

INITIAL REVIEW CRITERIA:

- Patient has an elevated serum bile acid concentration.
- Patient experiences persistent pruritus.
- Patient has trial and failure or contraindication to at least 1 pruritus treatment (e.g., ursodiol, cholestyramine, antihistamine).
- Patient has baseline liver function and fat-soluble vitamin tests and is monitored during treatment.

Bylvay[®]

- Patient must be ≥ 3 months of age.
- Patient must have a diagnosis of pruritus with progressive familial intrahepatic cholestasis (PFIC) confirmed by a genetic test.

Livmarli[®]

- **Patient must have one of the following diagnoses and meets all associated requirements:**
 - Cholestatic pruritus in patients with Alagille syndrome (ALGS)**
 - Patient must be ≥ 3 months of age.
 - Diagnosis must be confirmed by a genetic test.
 - Cholestatic pruritus in patients with progressive familial intrahepatic cholestasis (PFIC)**
 - Patient must be ≥ 5 years of age.
 - Diagnosis must be confirmed by a genetic test.
(Note: Livmarli[®] is not recommended in a subgroup of PFIC type 2 patients with specific ABCB11 variants resulting in non-functional or complete absence of bile salt export pump (BSEP) protein).
 - Patient has had an inadequate response, intolerance, or contraindication to Bylvay[®] (clinical documentation demonstrating response to previous therapy must be submitted).

CONTINUATION OF THERAPY:

- Patient met initial review criteria.
- Documentation of improved clinical response.
- Dosing is appropriate as per labeling or is supported by compendia.

DOSING AND ADMINISTRATION:

- Refer to product labeling at <https://www.accessdata.fda.gov/scripts/cder/daf/>