Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	December 8, 2021 March 16, 2023, December 20, 2023, July 10, 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 pruritis treatment (e.g., ursodiol, cholestyramine, antihistamine).
- Patient has baseline liver function and fat-soluble vitamin tests and is monitored during treatment.

Bylvay®

Cholestatic pruritus in patients with progressive familial intrahepatic cholestasis (PFIC)

- Patient must be ≥ 3 months of age.
- Patient must have a diagnosis of pruritus with progressive familial intrahepatic cholestasis (PFIC), type 1 or 2, confirmed by a genetic test.
    - *(Note: Bylvay® 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).*

Cholestatic pruritus in patients with Alagille syndrome (ALGS)

- Patient must be ≥ 12 months of age.
- Patient must have a diagnosis of pruritus with Alagille Syndrome (ALGS) confirmed by a genetic test.
- **Patient has had an inadequate response, intolerance, or contraindication to Livmarli®** (clinical documentation demonstrating response to previous therapy must be submitted).

Livmarli®

- **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).*
    - **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/