New Hampshire Medicaid Fee-for-Service Program Cholestatic Pruritus Criteria

Approval Date: November 21, 2024

Medications

Brand Names	Generic Names	Indication
Bylvay®	odevixibat	• Treatment of pruritus in patients ≥ 3 months of age with progressive familial intrahepatic cholestasis (PFIC) • Treatment of cholestatic pruritus in patients ≥ 12 months of age with Alagille syndrome (ALGS)
Livmarli®	maralixibat	• Treatment of cholestatic pruritus in patients ≥ 5 years of age with PFIC • Treatment of cholestatic pruritus in patients ≥ 3 months of age with ALGS

Criteria for Approval

Progressive Familial Intrahepatic Cholestasis

1. Patient is 3 months of age or older (Bylvay®) or 5 years of age or older (Livmarli®); **AND**
2. Prescribed by or in consultation with a gastroenterologist, hepatologist, or dermatologist; **AND**
3. Patient has progressive familial intrahepatic cholestasis (PFIC) type 1 or type 2, confirmed by a genetic test; **AND**
4. Patient has an elevated serum bile acid concentration; **AND**
5. The patient experiences persistent moderate to severe pruritus; **AND**
6. The benefits outweigh the risks for patients with:
   - Chronic diarrhea requiring ongoing intravenous fluid or nutritional intervention
   - Prior hepatic decompensation event
   - Decompensated cirrhosis
   - An international normalized ration (INR) > 1.4
   - Another concomitant liver disease
7. The patient has failed an adequate trial, or is intolerant to, or has a contraindication to at least one pruritus treatment (e.g., ursodeoxycholic acid [ursodiol], cholestyramine, rifampin, naloxone, naltrexone, antihistamine). **Note**: use of these agents are off-label.

Alagille syndrome (ALGS)

1. Patient is 12 months of age or older (Bylvay®) or 3 months of age or older (Livmarli®); **AND**
2. Prescribed by or in consultation with a gastroenterologist, hepatologist, or dermatologist; **AND**

3. The patient has been diagnosed with Alagille syndrome; **AND**
4. The patient has evidence of cholestasis as evidenced by at least one of the following:
   - Serum bile acid > 3 times upper limit of normal (ULN) for age
   - Conjugated bilirubin > 1 mg/dL
   - Gamma glutamyl transferase (GGT) > 3 times ULN for age
   - Fat soluble vitamin deficiency not otherwise explained
   - Intractable pruritus only explained by liver disease
5. The patient experiences persistent moderate to severe pruritus; **AND**
6. The benefits outweigh the risks for patients with:
   - Chronic diarrhea requiring ongoing intravenous fluid or nutritional intervention
   - Prior hepatic decompensation event
   - Decompensated cirrhosis
   - An international normalized ration (INR) > 1.4
   - Another concomitant liver disease
7. The patient has failed an adequate trial, or is intolerant to, or has a contraindication to at least one pruritus treatment (e.g., ursodeoxycholic acid [ursodiol], cholestyramine, rifampin, naloxone, naltrexone, antihistamine). **Note**: use of these agents are off-label.

Initial approval period: 6 months

Criteria for Renewal

1. Patient must continue to meet the above criteria; **AND**
2. Patient has experienced a reduction in serum bile acids from baseline; **AND**
3. Patient must experience improvement in pruritus; **AND**
4. Patient has not experienced any treatment-restricting adverse effects (e.g., persistent diarrhea, persistent fat-soluble vitamin deficiency despite supplementation, persistent or recurrent worsened liver function tests [alanine aminotransferase (ALT), aspartate aminotransferase (AST), total bilirubin (TB), direct bilirubin (DB)], decompensated cirrhosis, and significant portal hypertension).

Renewal period: 12 months

Criteria for Denial

1. Failure to meet approval criteria.

References

Available upon request.

Revision History

Reviewed by	Reason for Review	Date Approved
DUR Board	New	10/15/2024
Commissioner designee	Approval	11/21/2024