Pharmacy Drug Policy & Procedure

Policy Name:	Livmarli (maralixibat)	Policy#:	3122P

Purpose of the Policy

The purpose of this policy is to define coverage criteria for Livmarli (maralixibat).

Statement of the Policy

Health Alliance Medical Plans will approve the use of Livmarli (maralixibat) under the specialty pharmacy benefit if the following criteria are met.

Criteria

1. Coverage Criteria for Pruritus due to Familial Intrahepatic Cholestasis

1.1 Diagnosis of moderate to severe pruritus due to progressive familial intrahepatic cholestasis (PFIC)
- Diagnosis confirmed by genetic testing showing biallelic pathogenic mutations in the PFIC1, PFIC3, PFIC4 or PFIC6 genes

1.2 Member has cholestasis, as indicated by one of the following:
- Total serum bile acid >3 × upper limit of normal (ULN) for age
- Conjugated bilirubin >2 mg/dL
- Fat soluble vitamin deficiency that is otherwise unexplainable
- Gamma Glutamyl Transferase (GGT) >3 × ULN for age
- Intractable pruritus explainable only by liver disease

1.3 Age 12 months or older

1.4 Prescribed by or in consultation with a hepatologist (liver doctor)

1.5 Documented concurrent use or previous trial and failure, intolerance or contraindication ursodiol and cholestyramine

1.6 Review of chart notes documenting diagnosis and confirming that the patient has met all of the above requirements for treatment with Livmarli by both a pharmacist and medical director

2. Coverage Criteria for Pruritus due to Alagille Syndrome

2.1 Diagnosis of Alagille syndrome (ALGS) as confirmed by presence of the JAG1 or NOTCH2 mutation and documentation of moderate to severe pruritus (severe itching)

2.2 Age 3 months or older

2.3 Prescribed by or in consultation with a hepatologist (liver doctor)

2.4 Documented trial and failure of or contraindication to at least TWO of the following therapies for pruritus:
- Ursodiol
- Cholestyramine
- Rifampin
- Naltrexone (not for kids)
- Sertraline

2.5 Member has cholestasis, as indicated by one of the following:
- Total serum bile acid >3 × upper limit of normal (ULN) for age
- Conjugated bilirubin >2 mg/dL

- Fat soluble vitamin deficiency that is otherwise unexplainable
- Gamma Glutamyl Transferase (GGT) >3 × ULN for age
- Intractable pruritus explainable only by liver disease

2.6 Review of chart notes documenting diagnosis and confirming that the patient has met all of the above requirements for treatment with Livmarli by both a pharmacist and medical director

3. Exclusion Criteria

3.1 Member has chronic diarrhea requiring ongoing fluids or nutritional intervention
3.2 History of surgical interruption of enterohepatic circulation (partial external biliary diversion [PEBD] surgery)
3.3 History of liver transplant
3.4 Member has decompensated cirrhosis
3.5 Concomittant therapy with Bylvay
3.6 Livmarli is not recommended in PFIC type 2 patients with certain ABCB11 variants resulting in non-functional or complete absence of bile salt export pump (BSEP) protein

4. Approval Period

4.1 Initial: 12 months
4.2 Subsequent Approvals: 12 months with documentation of positive response to therapy

CPT Codes	
HCPCS Codes	

References

1. Livmarli (maralixibat) [prescribing information]. Foster City, CA: Mirum Pharmaceuticals Inc; July 2024.
2. Gonzales E, Hardikar W, Stormon M, et al. Efficacy and safety of maralixibat treatment in patients with Alagille syndrome and cholestatic pruritus (ICONIC): a randomised phase 2 study. Lancet. 2021;398(10311):1581-1592.
3. Kamath BM, Ye W, Goodrich NP, et al; Childhood Liver Disease Research Network (ChiLDReN). Outcomes of Childhood Cholestasis in Alagille Syndrome: Results of a Multicenter Observational Study. Hepatol Commun. 2020 Jan 22;4(3):387-398.
4. Randomized Double-blind Placebo-controlled Phase 3 Study to Evaluate the Efficacy and Safety of Maralixibat in the Treatment of Subjects With Progressive Familial Intrahepatic Cholestasis (PFIC) - MARCH-PFIC.
5. Cies JJ, Giamalis JN. Treatment of cholestatic pruritis in children. *Am J Health Syst Phar* 2007; 64:1157.
6. Jacquemin E. Progressive familial intrahepatic cholestasis. Clin Res Hepatol Gastroenterol. 2012;36 Suppl 1:S26-S35.

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DISCLAIMER

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