Health Alliance™ Pharmacy Drug Policy Checklist

POLICY NAME	Livmarli (maralixibat)	POLICY #	3122P

Criteria

Coverage Criteria for Pruritus due to Familial Intrahepatic Cholestasis

- ☐ 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
- ☐ 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 Age 12 months or older Prescribed by or in consultation with a hepatologist (liver doctor) Documented concurrent use or previous trial and failure, intolerance or contraindication ursodiol and cholestyramine 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

Coverage Criteria for Pruritus due to Alagille Syndrome

- ☐ Diagnosis of Alagille syndrome (ALGS) as confirmed by presence of the JAG1 or NOTCH2 mutation and documentation of moderate to severe pruritus (severe itching)
- ☐ Age 3 months or older
- ☐ Prescribed by or in consultation with a hepatologist (liver doctor)
- ☐ 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

- [ ] 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
- [ ] 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

Exclusion Criteria – Any of the following prevents coverage

- [ ] Member has chronic diarrhea requiring ongoing fluids or nutritional intervention
- [ ] History of surgical interruption of enterohepatic circulation (partial external biliary diversion [PEBD] surgery)
- [ ] History of liver transplant
- [ ] Member has decompensated cirrhosis
- [ ] Concomittant therapy with Bylvay
- [ ] 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