CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Georgia Medicaid

DRUG NAME	Livmarli (maralixibat)
BENEFIT TYPE	Pharmacy
Coverage Requirements	Prior Authorization Required

Livmarli (maralixibat) is an ileal bile acid transport (IBAT) inhibitor initially approved by the FDA in 2021 for the treatment of cholestatic pruritus in patients with Alagille Syndrome (ALGS). ALGS is a rare genetic disorder that can affect multiple organ systems, most commonly the liver, with a paucity of interlobular ducts. In cholestatic liver diseases, biliary substances aren't eliminated from the liver, thus they re-enter circulation. Cholestatic itch is thought to be related to the accumulation of bile acids in the skin. Inhibiting IBAT decreases reuptake of bile salts.

Livmarli (maralixibat) will be considered for coverage when the following criteria are met:

Cholestatic pruritus in patients with Alagille Syndrome (ALGS)

For **initial** authorization:

1. Member is at least 3 months of age; AND
2. Medication must be prescribed by or in consultation with a gastroenterologist OR hepatologist; AND
3. Member has a diagnosis of Alagille syndrome (ALGS) confirmed by the involvement of at least 3 of the following major clinical features:
   a) Hepatic (e.g., hyperbilirubinemia, cholestasis, xanthomas)
   b) Cardiac (e.g., heart murmur, peripheral pulmonic stenosis)
   c) Facial (e.g., inverted triangular face)
   d) Ocular (e.g., embryotoxon, optic disk drusen)
   e) Skeletal (e.g., butterfly vertebrae)
   f) Renal (e.g., renal dysplasia, renal tubular acidosis)
   g) Vascular (e.g., neurovascular accident, moyamoya disease
   NOTE: Member also meets criterion if has one or more clinical features and an affected first-degree relative; AND
4. Member must have liver biopsy demonstrating reduced number of the interlobular bile ducts OR confirmed finding of JAG1 or NOTCH2 gene mutation; AND
5. Member has symptoms of moderate to severe pruritus; AND
6. Member does NOT have any of the following:
   a) Previous liver transplant
   b) Previous surgical disruption of enterohepatic circulation (partial external bile diversion or ileal exclusion)
   c) Decompensated cirrhosis
   d) History or presence of other concomitant liver disease
7. Member must have a trial and failure of at least 2 of the following:
   a) Cholestyramine
   b) Ursodiol
   c) Rifampin

d) Naltrexone

8. **Dosage allowed/Quantity limit:** Starting dose is 190mcg/kg orally once daily, titrating up to 380 mcg/kg once daily. Max dose 28.5 mg (3 mL) per day. QL: 3 bottles (90 mL) per 30 days.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Pruritis has improved in response to therapy with Livmarli.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Livmarli (maralixibat) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
10/15/2021	New policy for Livmarli created.
03/28/2023	Updated/added references. Changed lower age limit from 1 year to 3 months per updated drug label. Added QL. Added naltrexone to list of trial options and removed specific trial duration. Added "note" to diagnostic criteria.

References:

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11. Muntaha HST, Munir M, Sajid SH, et al. Ileal Bile Acid Transporter Blockers for Cholestatic Liver Disease in Pediatric Patients with Alagille Syndrome: A Systematic Review and Meta-Analysis. J Clin Med. 2022;11(24):7526. Published 2022 Dec 19. doi:10.3390/jcm11247526

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