CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Livmarli (maralixibat)
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
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, occurring 1 in 30,000 to 1 in 45,000 according to NORD, and can lead to progressive liver disease. Livmarli (maralixibat) was granted orphan drug designation for ALGS by the FDA and breakthrough designation for the treatment of pruritis.

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 1 year of age; AND
2. Medication must be prescribed by or in consultation with gastroenterologist OR hepatologist; AND
3. Member has a diagnosis of Alagille syndrome (ALGS) confirmed by the involvement of at least 3 of the following alterations in features for diagnosis:
   a) Hepatic Features (e.g., hyperbilirubinemia or scleral icterus)
   b) Cardiac Features (e.g., lesions confirmed on imaging or murmur)
   c) Facial Features (e.g., inverted triangular face, straight nose with bulbous tip)
   d) Ocular Features (e.g., embryotoxon, optic disk drusen)
   e) Skeletal Features (e.g., vertebral anomalies, osteopenia
   f) Renal Features (e.g., renal dysplasia, renal tubular acidosis)
   g) Vascular Features (e.g., narrowing of internal carotid artery, moyamoya disease)
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. The member must have at least a 14-day trial and failure, or contraindication, to two out of three of the following:
   a) Cholestyramine
   b) Ursodiol
   c) Rifampin
8. **Dosage allowed:** Starting dose is 190mcg/kg, titrating up to 380 mcg/kg once daily;
   a) Maximum dose allowed is 28.5 mg per day.

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 year.

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.

References:

1. Livmarli. Package insert. Mirum Pharmaceuticals; 2021. Accessed October 13, 2021. https://www.accessdata.fda.gov/drugsatfda_docs/label/2021/214662s000lbl.pdf
2. Alagille Syndrome A Guide. Children's Liver Disease Foundation; 2019. Childliverdisease.org
3. ClinicalTrials.gov. Identifier: NCT02160782. Safety and Efficacy Study of LUM001 (Marlixibat) With a Drug Withdrawal Period in Participants With Alagille Syndrome (ALGS) (ICONIC). Available at: https://clinicaltrials.gov/ct2/show/results/NCT02160782
4. ClinicalTrials.gov. Identifier: NCT02057692. Evaluation of LUM001 in the Reduction of Pruritus in Alagille Syndrome (ITCH). Available at: https://clinicaltrials.gov/ct2/show/NCT02057692
5. Shneider BL, Spino C, Kamatha BM, et al. Placebo-Controlled Randomized Trial of an Intestinal Bile Salt Transport Inhibitor for Pruritus in Alagille Syndrome. *Hepatol Commun.* 2018 Oct; 2(10): 1184-1198. Doi 10.1002/hep4.1244
6. Ayoub MD and Kamath BM. Alagille Syndrome: Diagnostic Challenges and Advances in Management. *Diagnostics*. 2020; 10(11):907. https://doi.org/10.3390/diagnostics10110907
7. Lin, Henry. Alagille Syndrome. National Organization for Rare Disorders; updated 2020. Accessed October 12, 2021. https://rarediseases.org/rare-diseases/alagille-syndrome/
8. Alagille Syndrome. Johns Hopkins Medicine. Hopkins Medicine. Accessed October 10, 2021. https://www.hopkinsmedicine.org/health/conditions-and-diseases/alagille-syndrome

Effective date: 04/01/2022
Revised date: 10/15/2021