Mass General Brigham
Health Plan

Livmarli (maralixibat)
Effective 03/01/2025

Plan	☐ MassHealth UPPL ☒ Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit		
Specialty Limitations	N/A		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Exceptions	N/A		

Overview
Livmarli (maralixibat) is an ileal bile acid transporter (IBAT) inhibitor indicated for the treatment of:

- Cholestatic pruritus in patients with Alagille syndrome (ALGS) 3 months of age and older
- Cholestatic pruritus in patients 12 months of age and older with progressive intrahepatic cholestasis (PFIC)

Livmarli is not recommended in a subgroup of PFIC type 2 patients with specific ABCB11 variants resulting in non-functional or complete absence of bile salt export pump (BSEP) protein.

Coverage Guidelines
Authorization may be reviewed for members new to the plan within the past 90 days who are currently receiving treatment with the requested medication, excluding when the product is obtained as samples or via manufacturer's patient assistance programs
OR
Authorization may be granted if the member meets all following criteria:

Progressive Familial Intrahepatic Cholestasis (PFIC)

1. Member has a diagnosis of progressive familial intrahepatic cholestasis (PFIC)
2. Genetic testing does not indicate PFIC type 2 with ABCB11 variants encoding for nonfunction or absence of bile salt export pump protein (BSEP-3)
3. Member is 12 months of age or older
4. Requested medication is being prescribed by or in consultation with a hepatologist, gastroenterologist or a provider who specializes in PFIC
5. Member has pruritis
6. Member has had an inadequate response, adverse reaction, or contraindication to at least ONE of the following:
    a. Ursodiol (UDCA)
    b. Antihistamine

c. Rifampin
d. Cholestyramine
e. Sertraline
f. Naltrexone

Cholestatic Pruritus in Alagille Syndrome (ALGS)

1. The member has a diagnosis of Alagille syndrome (ALGS) confirmed by ONE of the following:
    a. Documentation of genetic testing confirming mutations of the JAG1 or NOTCH2 genes
    b. Bile duct paucity
    c. THREE of the five major clinical features of ALGS:
        i. Cholestasis
        ii. Cardiac defect (e.g., stenosis of the peripheral pulmonary artery and its branches)
        iii. Skeletal abnormality (e.g., butterfly vertebrae)
        iv. Ophthalmologic abnormality (e.g., posterior embryotoxon)
        v. Characteristic facial features (e.g., triangular-shaped face with a broad forehead and a pointed chin, bulbous tip of the nose, deeply set eyes, and hypertelorism)
2. Member is 3 months of age or older
3. Requested medication is prescribed by or in consultation with a hepatologist, gastroenterologist, or provider who specializes in treatment of Alagille Syndrome
4. Member has pruritus
5. Member has had an inadequate response, adverse reaction, or contraindication to at least ONE of the following:
    a. Ursodiol (UDCA)
    b. Antihistamine
    c. Rifampin
    d. Cholestyramine
    e. Sertraline
    f. Naltrexone

Continuation of Therapy

Requests for reauthorization will be approved when the following criteria are met:

1. Prescriber submits documentation of positive clinical response as evidenced by improvement in severity of pruritis.

Limitations

1. Initial approvals and reauthorizations will be granted for 12 months
2. The following quantity limits apply:

Drug Name	Quantity Limit
Livmarli 9.5mg/mL	90mL per 30 days

References

1. Genetic and Rare Diseases Information Center. Alagille syndrome. Rare Disease Database. https://rarediseases.info.nih.gov. Updated October 20, 2017. Accessed October 18, 2021.
2. Livmarli (maralixibat) [prescribing information]. Foster City, CA: Mirum Pharmaceuticals, Inc.; November 2024.
3. National Organization for Rare Disorders (NORD). Alagille syndrome. Rare Disease Database. https://rarediseases.org. Published 2020. Accessed October 18, 2021.

4. Spinner NB, Gilbert MA, Loomes KM, Krantz ID. Alagille syndrome. GeneReviews® [Internet]. December 12, 2019. Last updated December 12, 2019. Accessed October 19, 2021.

Review History

03/16/2022 – Created for March P&T Effective 05/01/2022.
12/11/2024 – Reviewed and updated for December P&T. Added approval criteria for supplemental indication of PFIC. Updated criteria for Alagille syndrome to allow for approval if the member has had genetic testing. Updated approvable age to 3 months or older. Added requirement for specialist prescriber and step through with one alternative agent. Removed requirements that the member cannot have a history of liver transplant or concomitant liver disease. Effective 3/1/2025.
02/12/2025 – Reviewed and updated for February P&T. Updated initial approval to 12 months. Effective 3/1/2025.