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DRUG POLICY

Livmarli (maralixibat)

NOTICE

This policy contains information which is clinical in nature. The policy is not medical advice. The information in this policy is used by Wellmark to make determinations whether medical treatment is covered under the terms of a Wellmark member's health benefit plan. Physicians and other health care providers are responsible for medical advice and treatment. If you have specific health care needs, you should consult an appropriate health care professional. If you would like to request an accessible version of this document, please contact customer service at 800-524-9242.

BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

DESCRIPTION

The intent of the Livmarli (maralixibat) drug policy is to ensure appropriate selection of members for therapy based on product labeling, clinical guidelines, and clinical studies. The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

- Livmarli (maralixibat) is indicated for the treatment of cholestatic pruritus in members 3 months of age and older with Alagille syndrome (ALGS)
- Livmarli (maralixibat) is indicated for the treatment of cholestatic pruritus in members 12 months of age and older with progressive familial intrahepatic cholestasis (PFIC)

Limitations of Use

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

POLICY

Required Documentation

Submission of the following information is necessary to initiate the prior authorization review:

1. Initial requests: Genetic testing results confirming a diagnosis of Alagille syndrome (ALGS) or progressive familial intrahepatic cholestasis (PFIC), if applicable.
2. Continuation requests: Chart notes or medical records documenting a benefit from therapy (e.g., improvement in pruritus).

Exclusions

1. Coverage will not be provided for members who have PFIC type 2 with variants in the ABCB11 gene resulting in non-functional or complete absence of the bile salt export pump (BSEP) protein.
2. Coverage will not be provided for members with prior or active hepatic decompensation events (e.g., variceal hemorrhage, ascites, hepatic encephalopathy).

Prescriber Specialties

The medication must be prescribed by or in consultation with a hepatologist or gastroenterologist.

Criteria for Initial Approval

Cholestatic pruritus in Alagille syndrome (ALGS)

Authorization of 6 months may be granted for treatment of cholestatic pruritus in Alagille syndrome (ALGS) when all of the following criteria are met:

1. Member has a diagnosis of ALGS established by one of the following (see Appendix for major clinical features of ALGS):
 - A. Genetic testing (e.g., JAG1 or NOTCH2 gene variants)
 - B. Family history of ALGS and one or more major clinical features of ALGS
 - C. Bile duct paucity and three or more major clinical features of ALGS
 - D. Four or more major clinical features of ALGS (see appendix)
2. Member has evidence of cholestasis (e.g., elevated serum bile acid level)
3. Member does not have a history or presence of other concomitant liver disease (e.g., biliary atresia, progressive familial intrahepatic cholestasis [PFIC], liver cancer)
4. Member has not received a liver transplant
5. Member is 3 months of age or older

Pruritus in Progressive Familial Intrahepatic Cholestasis (PFIC)

Authorization of 6 months may be granted for treatment of pruritus in progressive familial intrahepatic cholestasis (PFIC) when all of the following criteria are met:

1. Member has a confirmed molecular diagnosis of PFIC (e.g., ATP8B1, ABCB11, ABCB4, TJP2, or MYO5B gene variants)
2. Member has evidence of cholestasis (e.g., elevated serum bile acid level)
3. Member does not have any other concomitant liver disease (e.g., biliary atresia, liver cancer, alternate non-PFIC related etiology of cholestasis)
4. Member has not received a liver transplant
5. Member is 12 months of age or older

Continuation of Therapy

Authorization of 12 months may be granted for all members (including new members) requesting continuation of therapy when the member is experiencing benefit from therapy (e.g., improvement in pruritus).

Other

The requested medication will not be used concomitantly with any other ileal bile acid transporter (IBAT) inhibitor (e.g., Bylvay).

Livmarli (maralixibat) is considered **not medically necessary** for members who do not meet the criteria set forth above.

Dosage and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

Appendix:

Major clinical features of ALGS

1. Hepatic abnormality (e.g., cholestasis)
2. Cardiac abnormality (e.g., stenosis of the peripheral pulmonary artery and its branches)
3. Skeletal abnormality (e.g., butterfly vertebrae)
4. Ophthalmologic abnormality (e.g., posterior embryotoxon)
5. 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)
6. Vascular abnormalities (e.g., intracranial bleeds, systemic vascular anomalies)
7. Renal structural or functional abnormality (e.g., abnormally small size, cysts)

Quantity Limits:

Medication	Standard Limit	FDA-recommended dosing
Livmarli 9.5 mg/mL oral solution	120 mL (4 bottles) per 30 days	Treatment of ALGS: • Use Livmarli Oral Solution 9.5 mg/mL or tablets • Starting dose is 190 mcg/kg orally once daily • Should be increased to 380 mcg/kg once daily after one week, as tolerated • Not to exceed a maximum dose of 28.5 mg/day and 30 mg/day for tablets Treatment of PFIC: • Use Livmarli Oral Solution 19 mg/mL or tablets • Starting dose is 285 mcg/kg orally once daily • Should be increased to 285 mcg/kg twice daily, 428 mcg/kg twice daily, and then to 570 mcg/kg twice daily, as tolerated • Not to exceed a maximum dose of 38 mg/day and 40 mg/day for tablets <i>Note: Tablets may be used for treatment of both ALGS and PFIC in members weighing 25 kg and above who can swallow tablets</i>
Livmarli 19 mg/mL oral solution	60 mL (2 bottles) per 30 days	
Livmarli 10 mg tablets	60 per 30 days	
Livmarli 15 mg tablets	60 per 30 days	
Livmarli 20 mg tablets	60 per 30 days	
Livmarli 30 mg tablets	30 per 30 days	

PROCEDURES AND BILLING CODES

To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPSC level 2) codes, Revenue codes, and/or ICD diagnostic codes.

- Not applicable (N/A)

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- Menon J, Shanmugam N, Vij M, Rammohan A, Rela M. Multidisciplinary management of Alagille syndrome. J Multidiscip Healthc. 2022;15:353-364. doi: 10.2147/JMDH.S295441.
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- McKiernan P, Bernabeu JQ, Girard M, et al. Opinion paper on the diagnosis and treatment of progressive familial intrahepatic cholestasis. JHEP Rep. 2023;6(1):1000949. doi: 10.1016/j.jhepr.2023.100949.

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POLICY HISTORY

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