Bylvay (odevixibat)

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 Bylvay (odevixibat) drug policy is to ensure appropriate selection of patients 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

A. Bylvay (odevixibat) is indicated for the treatment of pruritus in patients 3 months of age and older with progressive familial intrahepatic cholestasis (PFIC).

B. Bylvay (odevixibat) is indicated for the treatment of cholestatic pruritus in patients 12 months of age and older with Alagille syndrome (ALGS).

Limitations of Use

Bylvay may not be effective in a subgroup of PFIC type 2 patients with specific ABCB11 variants resulting in non-functional or complete absence of the bile salt export pump protein.

POLICY

Required Documentation

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

- Initial requests: Genetic testing results confirming a diagnosis of progressive familial intrahepatic cholestasis (PFIC) or Alagille syndrome (ALGS), if applicable.

- Continuation requests: Chart notes or medical record documentation showing a benefit from therapy (e.g., improvement in pruritus).

Exclusions

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.

Prescriber Specialties

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

Criteria for Initial Approval

A. 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 3 months of age or older.

B. 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):
    i. Genetic testing (e.g., JAG1 or NOTCH2 gene variants).
    ii. Family history of ALGS and one or more major clinical features of ALGS
    iii. Bile duct paucity and three or more major clinical features of ALGS
    iv. Four or more major clinical features of ALGS
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, PFIC, liver cancer).
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

Member cannot use the requested medication concomitantly with any other ileal bile acid transporter (IBAT) inhibitor (e.g., Livmarli).

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

Bylvay 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.

Quantity Limits:

Medication	Standard Limit	FDA-recommended dosing
Bylvay (odevixibat) pellets 200 mcg	360 per 30 days	**Progressive familial intrahepatic cholestasis (PFIC)** • 40 mcg/kg once daily • If there is no improvement in pruritus after 3 months, the dosage may be increased in 40 mcg/kg increments up to 120 mcg/kg once daily not to exceed a total daily dose of 6 mg. **Alagille syndrome (ALGS)** • 120 mcg/kg once daily • Dose reduction to 40 mcg/kg/day may be considered if tolerability issues occur in the absence of other causes. Once tolerability issues stabilize, increase to 120 mcg/kg/day.
Bylvay (odevixibat) pellets 600 mcg	120 per 30 days	
Bylvay (odevixibat) capsule 400 mcg	540 per 30 days	
Bylvay (odevixibat) capsule 1200 mcg	180 per 30 days	

PROCEDURES AND BILLING CODES

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

- Not applicable (N/A)

REFERENCES

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- Spinner NB, Gilbert MA, Loomes KM, Krantz ID. Alagille syndrome. GeneReviews® [Internet]. Published May 19, 2020. Last updated January 4, 2024. Accessed March 6, 2024.
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POLICY HISTORY

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