Bylvay®
(odevixibat)

200 | 400 | 600 | 1200 mcg capsules

Dosage and Administration

Indications and Usage

Bylvay is an ileal bile acid transporter (IBAT) inhibitor indicated for the treatment of cholestatic pruritus in:

- Patients 12 months of age and older with Alagille syndrome (ALGS)
- Patients 3 months of age and older with progressive familial intrahepatic cholestasis (PFIC)
  - **Limitation 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

IMPORTANT SAFETY INFORMATION

Warnings and Precautions:

Liver Test Abnormalities

Patients enrolled in PFIC and ALGS clinical trials had abnormal liver tests at baseline. In clinical trials, treatment-emergent elevations of liver tests or worsening of liver tests relative to baseline values were observed during the clinical trials.

Please see Important Safety Information on page 6. Please see accompanying full Prescribing Information, or at bylvay.com/pdf/Bylvay_PI.pdf

Preparation and Administration Instructions

Take Bylvay in the morning with a meal.

- **For patients taking bile acid binding resins, take Bylvay at least 4 hours before or 4 hours after taking a bile acid binding resin.**

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Capsules:

- Do not crush or chew Capsules.

Administration Instructions:

1. Swallow the Capsule whole with a glass of water.
2. Alternatively, for patients unable to swallow the Capsules whole, Bylvay Capsules may be opened and then sprinkled and mixed with a small amount of soft food or age-appropriate liquid. Follow directions below for Oral Pellets to prepare and administer such a mixture.

Oral Pellets:

- Mix the contents of the shell containing Oral Pellets into soft food or an age-appropriate liquid, such as water, breast milk, or infant formula.
- Discard the emptied shells. Do not swallow the shell containing Oral Pellets whole.

Administration Instructions:

To open and mix with soft food:

1. Place a small amount of soft food (up to 30 mL [2 tablespoons] of apple sauce, oatmeal, banana or carrot puree, chocolate or rice pudding) in a bowl. Keep food at or below room temperature.
2. Open the shell containing Oral Pellets and empty the contents into the bowl of soft food. Gently tap the Oral Pellet shell to ensure that all contents have been dispersed.
   - If the dose requires more than one shell of Oral Pellets, repeat steps 1 and 2.
3. Gently mix until well dispersed and administer the entire dose immediately.

Instructions continue on page 3.

IMPORTANT SAFETY INFORMATION (CONT'D)

Warnings and Precautions (cont'd):

Liver Test Abnormalities (cont'd)

In a clinical trial with PFIC patients, treatment-emergent elevations of liver tests or worsening of liver tests relative to baseline values were observed during the clinical trial. Most abnormalities included elevations in AST, ALT, or total and direct bilirubin. Treatment interruption days ranged from 3 days to 124 days; no PFIC patients permanently discontinued treatment due to liver test abnormalities.

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Preparation and Administration Instructions (cont'd)

To open and mix with liquid (using an oral dosing syringe):

1. Open the shell containing Oral Pellets and empty the contents into a small mixing cup. Gently tap the shell containing Oral Pellets to ensure that all contents have been emptied into the mixing cup.
2. Add 1 teaspoon (5 mL) of an age-appropriate liquid (for example, breast milk, infant formula, or water).
   - If the dose requires more than one shell of Oral Pellets, repeat steps 1 and 2.
3. Let the pellets sit in the liquid for about 5 minutes before administering. Note that Oral Pellets will not dissolve.
4. After 5 minutes, place the tip of the oral syringe completely into the mixing cup. Pull the plunger of the syringe up slowly to withdraw the liquid/pellet mixture into the syringe. Gently push the plunger down again to expel the liquid/pellet mixture back into the mixing cup.
   - Do this 2 to 3 times to ensure complete mixing of the pellets into the liquid.
5. Withdraw the entire contents into the oral syringe by pulling the plunger on the end of the syringe.
6. Place the tip of the syringe into the front of the patient's mouth between the tongue and the side of the mouth, and then gently push the plunger down to squirt the liquid/pellet mixture between your child's tongue and the side of the mouth. Do not squirt liquid/pellet mixture in the back of the child's throat because this could cause gagging or choking.
   - Do not administer via a bottle or "sippy cup" because the Oral Pellets will not pass through the opening. The Oral Pellets will not dissolve in liquid.

Whether mixing with soft food or liquid, follow the dose with an age-appropriate liquid and do not store mixture for future use.

IMPORTANT SAFETY INFORMATION (CONT'D)

Warnings and Precautions (cont'd):

Liver Test Abnormalities (cont'd)

Obtain baseline liver tests and monitor during treatment. Dose reduction or treatment interruption may be required if abnormalities occur. For persistent or recurrent liver test abnormalities, consider treatment discontinuation.

Bylvay was not evaluated in PFIC or ALGS patients with cirrhosis. Closely monitor for liver test abnormalities; permanently discontinue Bylvay if a patient progresses to portal hypertension or experiences a hepatic decompensation event.

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Recommended Dosing for Bylvay® (odevixibat)

Dosage Forms and Strengths

Bylvay is currently available in 200 mcg and 600 mcg Oral Pellets, and 400 mcg and 1200 mcg Capsules.

Recommended Dosage of Bylvay for 40 mcg/kg/day

Body Weight (kg)	Total Daily Dose (mcg)	Number of Oral Pellets or Capsules
7.4 and below	200	1 (200 mcg Oral Pellet)
7.5 to 12.4	400	2 (200 mcg Oral Pellets)
12.5 to 17.4	600	1 (600 mcg Oral Pellet)
17.5 to 19.4	800	4 (200 mcg Oral Pellets)
19.5 to 25.4	800	2 (400 mcg Capsules)
25.5 to 35.4	1200	1 (1200 mcg Capsule)
35.5 to 45.4	1600	4 (400 mcg Capsules)
45.5 to 55.4	2000	5 (400 mcg Capsules)
55.5 and above	2400	2 (1200 mcg Capsules)

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.

Oral Pellets are intended for use by patients weighing less than 19.5 kilograms. Capsules are intended for use by patients weighing 19.5 kilograms or above.

How Supplied/Storage and Handling

Bylvay Capsules are supplied in bottles of 30 with child-resistant closure. Store at 20°C to 25°C (68°F to 77°F).

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Recommended Dosing for Bylvay® (odevixibat)

Recommended Dosage of Bylvay for 80 mcg/kg/day

Body Weight (kg)	Total Daily Dose (mcg)	Number of Oral Pellets or Capsules
7.4 and below	400	2 (200 mcg Oral Pellets)
7.5 to 12.4	800	4 (200 mcg Oral Pellets)
12.5 to 17.4	1200	1 (1200 mcg Capsule)*
17.5 to 19.4	1600	8 (200 mcg Oral Pellets)
19.5 to 25.4	1600	4 (400 mcg Capsules)
25.5 to 35.4	2400	2 (1200 mcg Capsules)
35.5 to 45.4	3200	8 (400 mcg Capsules)
45.5 to 55.4	4000	10 (400 mcg Capsules)
55.5 and above	4800	4 (1200 mcg Capsules)

*Open 1200 mcg Capsules and sprinkle on food or mix with liquid for child unable to swallow Capsule whole.

Recommended Dosage of Bylvay for 120 mcg/kg/day

Body Weight (kg)	Total Daily Dose (mcg)	Number of Oral Pellets or Capsules
7.4 and below	600	1 (600 mcg Oral Pellet)
7.5 to 12.4	1200	1 (1200 mcg Capsule)*
12.5 to 17.4	1800	3 (600 mcg Oral Pellets)
17.5 to 19.4	2400	2 (1200 mcg Capsules)*
19.5 to 25.4	2400	2 (1200 mcg Capsules)
25.5 to 35.4	3600	3 (1200 mcg Capsules)
35.5 to 45.4	4800	4 (1200 mcg Capsules)
45.5 and above	6000	5 (1200 mcg Capsules)

*Open 1200 mcg Capsules and sprinkle on food or mix with liquid for child unable to swallow Capsule whole.

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  - **Limitation of Use:**
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IMPORTANT SAFETY INFORMATION

Warnings and Precautions:
Liver Test Abnormalities

Patients enrolled in PFIC and ALGS clinical trials had abnormal liver tests at baseline. In clinical trials, treatment-emergent elevations of liver tests or worsening of liver tests relative to baseline values were observed during the clinical trials.

In a clinical trial with PFIC patients, treatment-emergent elevations of liver tests or worsening of liver tests relative to baseline values were observed during the clinical trial. Most abnormalities included elevations in AST, ALT, or total and direct bilirubin. Treatment interruption days ranged from 3 days to 124 days; no PFIC patients permanently discontinued treatment due to liver test abnormalities.

In a clinical trial with ALGS patients, treatment-emergent elevations or worsening in liver tests relative to baseline values were observed during the trial. Most abnormalities included elevations in ALT or AST. One ALGS patient interrupted treatment for 40 days; no ALGS patients permanently discontinued treatment due to liver test abnormalities.

Obtain baseline liver tests and monitor during treatment. Dose reduction or treatment interruption may be required if abnormalities occur. For persistent or recurrent liver test abnormalities, consider treatment discontinuation.

Bylvay was not evaluated in PFIC or ALGS patients with cirrhosis. Closely monitor for liver test abnormalities; permanently discontinue Bylvay if a patient progresses to portal hypertension or experiences a hepatic decompensation event.

Diarrhea

In a PFIC clinical trial, diarrhea was reported in 2 (10%) placebo-treated patients, 9 (39%) Bylvay-treated 40 mcg/kg/day patients, and 4 (21%) Bylvay-treated 120 mcg/kg/day patients. Treatment interruption due to diarrhea occurred in 2 patients with 3 events during treatment with Bylvay 120 mcg/kg/day. Treatment interruption due to diarrhea ranged between 3 to 7 days. One patient treated with Bylvay 120 mcg/kg/day withdrew from the pivotal clinical trial due to persistent diarrhea.

In an ALGS clinical trial, diarrhea in ALGS patients was reported in 1 (6%) placebo-treated patient and in 10 (29%) Bylvay-treated patients. No patients interrupted or permanently discontinued Bylvay due to diarrhea.

If diarrhea occurs, monitor for dehydration and treat promptly. Interrupt Bylvay dosing if a patient experiences persistent diarrhea. Restart Bylvay at 40 mcg/kg/day when diarrhea resolves and increase the dose as tolerated if appropriate. If diarrhea persists and no alternate etiology is identified, stop Bylvay treatment.

Fat-Soluble Vitamin (FSV) Deficiency

Fat-soluble vitamins (FSV) include vitamin A, D, E, and K (measured using INR levels). PFIC patients can have FSV deficiency at baseline. Bylvay may affect absorption of fat-soluble vitamins. In a clinical trial, new onset or worsening of existing FSV deficiency was reported in 1 (5%) placebo-treated patient and 3 (16%) Bylvay-treated 120 mcg/kg/day patients; none of the Bylvay-treated 40 mcg/kg/day patients had new onset or worsening of existing FSV deficiency. In an ALGS clinical trial, new or worsening of existing FSV deficiency was reported in 3 (17.6%) placebo-treated patients and 3 (8.6%) Bylvay-treated patients.

Obtain serum FSV levels at baseline and monitor during treatment, along with any clinical manifestations. If FSV deficiency is diagnosed, supplement with FSV. Discontinue Bylvay if FSV deficiency persists or worsens despite adequate FSV supplementation.

Adverse Reactions

The most common adverse reactions for Bylvay in patients with PFIC are diarrhea, liver test abnormalities, vomiting, abdominal pain, and fat-soluble vitamin deficiency.

The most common adverse reactions for Bylvay patients with ALGS are diarrhea, abdominal pain, hematoma, and decreased weight.

Drug Interactions

For patients taking bile acid binding resins, take Bylvay at least 4 hours before or 4 hours after taking a bile acid binding resin.

Use in Specific Populations

There are no human data on Bylvay use in pregnant persons to establish a drug-associated risk of major birth defects, miscarriage, or adverse developmental outcomes. Based on findings from animal reproduction studies, Bylvay may cause cardiac malformations when a fetus is exposed during pregnancy.

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Dedicated Support for Your Patients Taking Bylvay

Bylvay is backed by Albireo Assist, an in-house comprehensive support program specifically for patients prescribed Bylvay. Dedicated Albireo Assist Care Coordinators can provide:

- Financial & Insurance Assistance for Eligible Patients
- Care Logistics
- Dedicated, Live Support
- Educational Materials and Programs

For more information, or to speak with an Albireo Assist Care Coordinator, please call 855-ALBIREO (855-252-4736).

Visit Bylvay.com for more information

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Find instructional videos and other tips on taking Bylvay at Bylvay.com or by scanning or clicking this code.

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