BYLVAY® (odevixibat) Dosage and Administration

INDICATIONS

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

- cholestatic pruritus in patients ≥12 months of age with Alagille syndrome (ALGS)
- pruritus in patients ≥3 months of age with progressive familial intrahepatic cholestasis (PFIC)

Limitation of Use:

BYLVAY is not recommended 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

Contraindications

IBAT inhibitors, including BYLVAY, are contraindicated in patients with prior or active hepatic decompensation events (e.g., variceal hemorrhage, ascites, hepatic encephalopathy).

Please see Important Safety Information throughout and on page 7, full Prescribing Information, and Instructions for Use.

Bylvay® (odevixibat)
200 | 400 | 600 | 1200 mcg capsules

Preparation and Administration Instructions

BYLVAY is taken once daily with a morning meal[1]

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

Oral Pellets[1]:

Oral Pellets 200 mcg

Oral Pellets 600 mcg

Administration Instructions:


Mix the contents of the shell containing Oral Pellets into soft food or an age-appropriate liquid.


Discard the emptied shells. Do not swallow the shell containing Oral Pellets whole.

To open Oral Pellets and mix with **soft food**[1]:

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 the soft 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 step 2
3. Gently mix until well dispersed and administer the entire dose immediately.

To open Oral Pellets and mix with **liquid (using an oral dosing syringe)**[1]:

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.
   - If the dose requires more than one shell of Oral Pellets, repeat step 1
2. Add 1 teaspoon (5 mL) of an age-appropriate liquid.
3. Let the pellets sit in the liquid for about 5 minutes to allow complete wetting. 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
7. If any pellet/liquid mixture remains in the mixing cup, repeat steps 5 and 6 until the entire dose has been administered.

IMPORTANT SAFETY INFORMATION (CONT'D)

WARNINGS AND PRECAUTIONS

Hepatotoxicity

BYLVAY treatment is associated with a potential for drug-induced liver injury (DILI). In the PFIC and ALGS trials, treatment-emergent elevations or worsening of liver tests occurred. Of the six patients who experienced DILI, two underwent liver transplant.

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Preparation and Administration Instructions (cont'd)

Capsules[1]:

Capsules 400 mcg

Capsules 1200 mcg

Administration Instructions:

Swallow the Capsule whole with a glass of water.

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 on previous page for Oral Pellets to prepare and administer such a mixture.

Do not crush or chew Capsules.

Whether mixing Oral Pellets or the contents of Capsules with soft food or liquid[1]:
- Follow the dose with an age-appropriate liquid
- Check the child's mouth to make sure all of the mixture has been swallowed
- Do not store mixture for future use

Click to access the BYLVAY Dosing Calculator

IMPORTANT SAFETY INFORMATION (CONT'D)

WARNINGS AND PRECAUTIONS

Hepatotoxicity (cont'd)

Obtain baseline liver tests because some ALGS and PFIC patients have abnormal liver tests at baseline and monitor patients frequently for the first 6 to 8 months, and as clinically needed thereafter, for elevations in liver tests, for the development of liver-related adverse reactions, and for physical signs of hepatic decompensation. If liver test abnormalities or signs of clinical hepatitis occur in the absence of other causes, consider dose reduction or treatment interruption. Permanently discontinue BYLVAY if a patient experiences the following: persistent or recurrent liver test abnormalities, or upon rechallenge, signs and symptoms consistent with clinical hepatitis, or a hepatic decompensation event.

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Recommended Dosing

Dosage Forms and Strengths

BYLVAY is currently available in 200 mcg and 600 mcg Oral Pellets and 400 mcg and 1200 mcg Capsules.[1]

BYLVAY Dosage of 40 mcg/kg/day[1]

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

For patients with PFIC, 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 daily dosage of 6 mg/day.[1]

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.[1]

How Supplied/Storage and Handling[1]

BYLVAY Oral Pellets and Capsules are supplied in bottles of 30 with child-resistant closure.
Store at 20°C to 25°C (68°F to 77°F); excursions permitted between 15°C and 30°C (between 59°F and 86°F).

IMPORTANT SAFETY INFORMATION (CONT'D)

WARNINGS AND PRECAUTIONS

Hepatotoxicity (cont'd)

The safety and effectiveness of BYLVAY have not been established in patients with decompensated cirrhosis. Monitor patients with compensated cirrhosis or portal hypertension more frequently and discontinue if hepatic decompensation occurs. IBAT inhibitors, including BYLVAY, are contraindicated in patients with prior or active hepatic decompensation events.

Please see Important Safety Information throughout and on page 7, full Prescribing Information, and Instructions for Use.

Recommended Dosing (cont'd)

BYLVAY Dosage of 80 mcg/kg/day[1]

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	2 × 600 mcg Oral Pellets
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

Maximum total daily dose for PFIC: 6 mg

BYLVAY Dosage of 120 mcg/kg/day[1]

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	2 × 600 mcg Oral Pellets
12.5 to 17.4	1800	3 × 600 mcg Oral Pellets
17.5 to 19.4	2400	4 × 600 mcg Oral Pellets
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 to 55.4	6000	5 × 1200 mcg Capsules
55.5 and above	7200	6 × 1200 mcg Capsules

Maximum total daily dose for pruritus in PFIC: 6 mg
Maximum total daily dose for cholestatic pruritus in ALGS: 7.2 mg

ALGS=Alagille syndrome; PFIC=progressive familial intrahepatic cholestasis.

IMPORTANT SAFETY INFORMATION (CONT'D)

WARNINGS AND PRECAUTIONS

Diarrhea
In the PFIC and ALGS clinical trials, diarrhea was reported more frequently in BYLVAY-treated patients compared to placebo. In the PFIC clinical trials, treatment interruption due to diarrhea occurred in 2 patients with 3 events.

Please see Important Safety Information throughout and on page 7, full Prescribing Information, and Instructions for Use.

Dedicated Support for Your Patients and Their Families

IPSENCARES®
Coverage, Access, Reimbursement & Education Support

IPSEN CARES® helps patients and families get access to their BYLVAY prescription, providing the information and resources they need to better understand and manage their condition.

Financial & Insurance Assistance*

Dedicated, Individualized Support

Educational Materials & Programs

Continuity of Care

Enroll your patients by filling out the IPSEN CARES Enrollment Form

Fill out and submit online

Fill out online and fax

Enroll on paper and fax

We'll take it from there

After enrollment, a member of the IPSEN CARES team will reach out to your patient.

We offer support related to healthcare decisions that have been made between patients and their healthcare providers and are committed to keeping them informed along the way.

*Please see patient eligibility and program terms and conditions at IPSENCARES.com.

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Indications and Important Safety Information

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Limitation of Use:

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IMPORTANT SAFETY INFORMATION

Contraindications

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WARNINGS AND PRECAUTIONS

Hepatotoxicity

BYLVAY treatment is associated with a potential for drug-induced liver injury (DILI). In the PFIC and ALGS trials, treatment-emergent elevations or worsening of liver tests occurred. Of the six patients who experienced DILI, two underwent liver transplant.

Obtain baseline liver tests because some ALGS and PFIC patients have abnormal liver tests at baseline and monitor patients frequently for the first 6 to 8 months, and as clinically needed thereafter, for elevations in liver tests, for the development of liver-related adverse reactions, and for physical signs of hepatic decompensation. If liver test abnormalities or signs of clinical hepatitis occur in the absence of other causes, consider dose reduction or treatment interruption. Permanently discontinue BYLVAY if a patient experiences the following: persistent or recurrent liver test abnormalities, or upon rechallenge, signs and symptoms consistent with clinical hepatitis, or a hepatic decompensation event.

The safety and effectiveness of BYLVAY have not been established in patients with decompensated cirrhosis. Monitor patients with compensated cirrhosis or portal hypertension more frequently and discontinue if hepatic decompensation occurs. IBAT inhibitors, including BYLVAY, are contraindicated in patients with prior or active hepatic decompensation events.

Diarrhea

In the PFIC and ALGS clinical trials, diarrhea was reported more frequently in BYLVAY-treated patients compared to placebo. In the PFIC clinical trials, treatment interruption due to diarrhea occurred in 2 patients with 3 events.

Treatment interruption due to diarrhea ranged between 3 to 7 days. One patient withdrew from the trial. In the ALGS clinical trial, no patients interrupted or discontinued treatment due to diarrhea.

If diarrhea occurs, monitor for dehydration and treat promptly. Interrupt 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 treatment.

Fat-Soluble Vitamin (FSV) Deficiency

Fat-soluble vitamins (FSV) include vitamin A, D, E, and K. PFIC and ALGS patients can have FSV deficiency at baseline. BYLVAY may adversely affect absorption of FSVs. In clinical trials, new onset or worsening of existing FSV deficiency was reported more frequently in BYLVAY-treated patients compared to placebo.

Obtain baseline INR (International Normalized Ratio) and FSV levels and monitor during treatment along with any clinical manifestations. If FSV deficiency is diagnosed, supplement with FSV. Discontinue BYLVAY if FSV deficiency persists, worsens, or complications occur despite adequate FSV supplementation.

If bone fracture occurs, consider interrupting BYLVAY treatment and supplement with FSV if indicated.

If bleeding occurs, interrupt treatment with BYLVAY. Optimize treatment of FSV deficiency and consider restarting BYLVAY once the patient is clinically stable.

Adverse Reactions

The most common adverse reactions for BYLVAY in patients with PFIC are diarrhea, liver test abnormalities, vomiting, abdominal pain, and FSV 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

Limited human data on BYLVAY use in pregnant persons are insufficient 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. As BYLVAY may inhibit the absorption of fat-soluble vitamins, which are essential for normal fetal growth and development, monitor pregnant patients for FSV deficiency and increase supplementation as needed. Consider the woman's need for BYLVAY, the potential drug-related risks to the fetus, and the potential adverse outcomes from untreated maternal PFIC and ALGS.

There is a pregnancy exposure registry that monitors pregnancy outcomes in persons exposed to BYLVAY during pregnancy. Pregnant women exposed to BYLVAY, or their healthcare providers, should report BYLVAY exposure by calling 1-855-463-5127.

To report SUSPECTED ADVERSE REACTIONS, contact Ipsen Biopharmaceuticals, Inc. at 1-855-463-5127 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Please see full Prescribing Information.

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BYLVAY Offers Once-Daily Dosing and Flexible Administration[1]

1 **daily dose** taken with a morning meal[1]

2 **oral formulation options:** oral pellets, capsules[1]

3 **ways to take:** swallow capsules whole; mix contents* with soft food or liquid[1]

*Pellets or capsules.

The **ONLY IBAT inhibitor** that:

- ✔ **CAN BE** mixed with soft food or age-appropiate liquid[2]
- ✔ **DOES NOT** require fasting or delaying of meals[2]
- ✔ **DOES NOT** require dose titration for standard treatment initiation[2]

Recommended starting dose for BYLVAY

- Recommended starting dose for pruritus in PFIC: **40 mcg/kg/day**
  - 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
  - Dose should not exceed 6 mg/day
- Recommended starting dose for cholestatic pruritus in ALGS: **120 mcg/kg/day**
  - 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

For complete instructions on dosage and administration, see the full **Prescribing Information, and Instructions for Use.**

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ALGS=Alagille syndrome; IBAT=ileal bile acid transporter; PFIC=progressive familial intrahepatic cholestasis.

References: 1. BYLVAY. Prescribing Information. Ipsen Biopharmaceuticals, Inc.; 2025. **2.** Data on file. Ipsen US. [NON-US-004496].

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