Juxtapid (lomitapide) capsules

For Healthcare Providers

All prescribers of Juxtapid must be certified in the Juxtapid REMS Program prior to prescribing Juxtapid.
Prescribers who were certified *prior to* February 20, 2017 must recertify under the new program requirements.
Prescribers who certified *after* February 20, 2017 may skip the certification steps and go directly to the prescribe section.

To Certify follow these steps:

1. **Review** the Juxtapid Prescribing Information, Fact Sheet, and complete the Prescriber Training Module at www.juxtapidremsprogram.com.
2. **Print** the Certificate of Completion for the Prescriber Training Module.
3. **Complete, sign, and fax or email** the Juxtapid REMS Prescriber Enrollment Form and a printed copy of the Certificate of Completion to 1-855-898-2498 or REMS@aegerion.com.

To Prescribe follow these steps:

1. **Schedule appointment** to counsel patient. **Complete** the Juxtapid Patient-Prescriber Acknowledgement Form with each patient.
   - **Counsel your patient** on the Juxtapid REMS Program requirements using the Patient Guide available on www.juxtapidremsprogram.com. Provide the patient with a copy of the guide.
   - **Complete and sign** the Patient-Prescriber Acknowledgement Form (attached to the Patient Guide).
2. **Complete and sign** the Juxtapid REMS Prescription Authorization Form found at www.juxtapidremsprogram.com.
3. **Complete and sign** the Compass Enrollment Form (on page 5) to provide Aegerion support services to your patient such as insurance coverage information, nutritional counseling, and financial assistance (if eligible).
4. **Have your patient complete and sign** the Compass Patient Authorization Form (on page 6) to allow a Compass Coordinator to work with your patients to navigate their access and affordability options and to coordinate shipments.
5. **Fax the following completed and signed forms and patient information to Compass at 1-855-898-2498**

- ☐ Juxtapid REMS Prescription Authorization Form
- ☐ Patient-Prescriber Acknowledgement Form
- ☐ Compass Enrollment Form
- ☐ Compass Patient Authorization Form
- ☐ Copy of patient's medical and pharmacy insurance cards (front and back)
- ☐ Clinical notes including documentation of recent medications

Please see Indication and Important Safety Information on pages 3-4 and accompanying full Prescribing Information including Medication Guide and Boxed Warning.

Questions? Call Compass at **1-85-JUXTAPID** (1-855-898-2743), Monday through Friday, 8:00am-8:00pm ET

Juxtapid (lomitapide) capsules

For Patients

1. **Read and sign** the enclosed Patient Authorization Form (on page 6) if you would like to receive Aegerion support services such as insurance coverage information, nutritional counseling, and financial assistance (if eligible).

What's Next?

2. Your Healthcare Provider will fax the necessary paperwork to us.

3. You will be contacted by a Compass Nurse Case Manager within 2 business days to confirm your insurance coverage and delivery details, and to coordinate additional Compass support services. *Please note this call might come from an unfamiliar phone number.*

COMPASS SUPPORT

To help support you with your current treatment plan, a Compass support team is built around you to help provide the support services and resources you need, when you need them. When you sign up for Compass, you can access these support services and your dedicated team of experts at no cost to you.

What you can expect from Compass:

- A phone consultation with your Compass Registered Dietitian to customize your low-fat eating plan
- An in-person session with your Compass Registered Dietitian to learn about your treatment
- A phone session with your Compass Nurse Case Manager to navigate the insurance process and help you schedule the delivery of your Juxtapid

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Juxtapid® (lomitapide) capsules

INDICATION

JUXTAPID® (lomitapide) capsules is a microsomal triglyceride transfer protein inhibitor indicated as an adjunct to a low-fat diet and other lipid-lowering treatments, including LDL apheresis where available, to reduce low-density lipoprotein cholesterol, total cholesterol, apolipoprotein B, and non-high-density lipoprotein cholesterol in patients with homozygous familial hypercholesterolemia (HoFH).

LIMITATIONS OF USE

The safety and effectiveness of JUXTAPID have not been established in patients with hypercholesterolemia who do not have HoFH, including those with heterozygous familial hypercholesterolemia (HeFH). The effect of JUXTAPID on cardiovascular morbidity and mortality has not been determined.

IMPORTANT SAFETY INFORMATION

WARNING: RISK OF HEPATOTOXICITY

JUXTAPID can cause elevations in transaminases. In the JUXTAPID clinical trial, 10 (34%) of the 29 patients treatedwith JUXTAPID had at least one elevation in alanine aminotransferase (ALT) or aspartate aminotransferase (AST) ≥3x upper limit of normal (ULN). There were no concomitant clinically meaningful elevations of total bilirubin, international normalized ratio (INR), or alkaline phosphatase.

JUXTAPID also increases hepatic fat, with or without concomitant increases in transaminases. The median absolute increase in hepatic fat was 6% after both 26 and 78 weeks of treatment, from 1% at baseline, measured by magnetic resonance spectroscopy. Hepatic steatosis associated with JUXTAPID treatment may be a risk factor for progressive liver disease, including steatohepatitis and cirrhosis.

Measure ALT, AST, alkaline phosphatase, and total bilirubin before initiating treatment and then ALT and AST regularly as recommended. During treatment, adjust the dose of JUXTAPID if the ALT or AST are ≥3x ULN. Discontinue JUXTAPID for clinically significant liver toxicity.

Because of the risk of hepatotoxicity, JUXTAPID is available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS) called the JUXTAPID REMS Program.

Prescribe JUXTAPID only to patients with a clinical or laboratory diagnosis consistent with HoFH. The safety and effectiveness of JUXTAPID have not been established in patients with hypercholesterolemia who do not have HoFH.

CONTRAINDICATIONS

- Pregnancy
- Concomitant administration of moderate or strong CYP3A4 inhibitors
- Moderate or severe hepatic impairment or active liver disease including unexplained persistent elevations of serum transaminases

WARNINGS AND PRECAUTIONS

JUXTAPID can cause elevations in transaminases and hepatic steatosis. Although cases of hepatic failure have not been reported, there is concern that JUXTAPID could induce steatohepatitis, which can progress to cirrhosis over several years. Modify the dose of JUXTAPID if elevations of transaminases are observed and discontinue JUXTAPID for persistent or clinically significant elevations. If transaminase elevations are accompanied by clinical symptoms of liver injury, such as nausea, vomiting, abdominal pain, fever, jaundice, lethargy, flu-like-symptoms, increases in bilirubin ≥2x ULN, or active liver disease, discontinue treatment with JUXTAPID and identify the probable cause. Use JUXTAPID with caution when co-administered with agents known to be hepatotoxic. Alcohol may increase levels of hepatic fat and induce or exacerbate liver injury.

Measure ALT, AST, alkaline phosphatase, and total bilirubin before initiating treatment. During the first year, measure liver-related tests (ALT and AST, at a minimum) prior to each increase in dose or monthly, whichever occurs first. After the first year, do these tests at least every 3 months and before any increase in dose.

JUXTAPID may cause fetal harm when administered to a pregnant woman. Females of reproductive potential should have a negative pregnancy test before starting JUXTAPID and should use effective contraception during therapy with JUXTAPID. The recommended maximum dosage of JUXTAPID is 40 mg daily when used concomitantly with oral contraceptives.

Given its mechanism of action in the small intestine, JUXTAPID may reduce the absorption of fat-soluble nutrients. Patients treated with JUXTAPID should take daily supplements that contain 400 international units vitamin E and at least 200 mg linoleic acid, 210 mg alpha-linolenic acid (ALA), 110 mg eicosapentaenoic acid (EPA), and 80 mg docosahexaenoic acid (DHA).

Gastrointestinal adverse reactions are common and may lead to treatment discontinuation. Instruct patients to stop JUXTAPID and contact their healthcare provider if severe diarrhea occurs, or if they experience symptoms of volume depletion such as lightheadedness, decreased urine output, or tiredness. In such cases, consider reducing the dose

Please see Indication and Important Safety Information on pages 3-4 and accompanying full Prescribing Information, including Medication Guide and Boxed Warning.

or suspending use of JUXTAPID. To reduce the risk of gastrointestinal adverse reactions, patients should adhere to a low-fat diet supplying less than 20% of energy from fat and the dosage of JUXTAPID should be increased gradually.

Weak CYP3A4 inhibitors can increase the exposure of lomitapide approximately 2-fold; therefore, when JUXTAPID is administered with weak CYP3A4 inhibitors, the dose of JUXTAPID should be decreased by half and the recommended maximum dosage of JUXTAPID is 30 mg daily. The recommended maximum dosage is 40 mg daily when used concomitantly with oral contraceptives. Strong and moderate CYP3A4 inhibitors should not be used with JUXTAPID. Patients taking JUXTAPID 5 mg daily may continue with the same dosage.

Due to risk of myopathy associated with simvastatin or lovastatin, doses of these agents should be limited when co-administered with JUXTAPID.

JUXTAPID increases the plasma concentrations of warfarin. Increases or decreases in the dose of JUXTAPID may lead to supra- or subtherapeutic anticoagulation, respectively. Patients taking warfarin should undergo regular monitoring of the INR, especially after any changes in JUXTAPID dosage.

Avoid use of JUXTAPID in patients with rare hereditary diseases of galactose intolerance.

ADVERSE REACTIONS

The most common adverse reactions were gastrointestinal, reported by 27 (93%) of 29 patients. Adverse reactions reported by 8 (28%) or more patients in the HoFH clinical trial included diarrhea, nausea, vomiting, dyspepsia and abdominal pain. Other common adverse reactions, reported by 5 to 7 (17-24%) patients, included weight loss, abdominal discomfort, abdominal distension, constipation, flatulence, increased ALT, chest pain, influenza, nasopharyngitis, and fatigue.

REPORTING OF ADVERSE REACTIONS

All healthcare professionals should report all suspected adverse reactions. Please contact Aegerion Pharmaceuticals, Inc. at 1-855-303-2347 or the FDA at 1-800-FDA-1088 or *http://www.fda.gov/medwatch*.

Please see accompanying full Prescribing Information, including Medication Guide and Boxed Warning.

COMPASS ENROLLMENT FORM

HEALTHCARE PROVIDER: Please complete all sections of this form and fax to 1-855-898-2498.

Juxtapid (lomitapide) capsules

I. PATIENT INFORMATION

Patient is (choose one): ☐ New ☐ Currently Receiving JUXTAPID
Patient Name (First MI Last): _____
Address: _____ City: _____ State: _____ Zip: _____
Date of Birth: ____ /____ / ____ Gender: ☐ Male ☐ Female Email: _____
Primary Contact: ☐ Patient ☐ Legal Representative (if applicable): _____
Preferred Phone: _____ ☐ OK to leave message Alternate Phone: _____ ☐ OK to leave message

II. INSURANCE INFORMATION

Please send a copy of the front and back of the medical and pharmacy insurance cards. If not available, please complete the fields below.

Medical Insurance
Policy Holder Name: _____
Medical Insurance Phone: _____
Medical Policy #: _____ Group # _____

Prescription Insurance
Policy Holder Name: _____
Prescription Insurance Phone: _____
Prescription Policy #: _____ Group # _____

III. MEDICAL ASSESSMENT

REMS Attestation:
☐ Patient has clinical/laboratory diagnosis consistent with HoFH.

ICD-10 Diagnosis Code:
☐ E78.0/Pure Hypercholesterolemia (including HoFH)
☐ Other: _____ /_____

Allergies: ☐ None or ☐ Specify: _____

Height: _____ inches **Weight:** _____ lbs.

Pretreatment Lab Information:
Measured ALT, AST, ALP and total bilirubin? ☐ Yes ☐ No
During the first year, measure liver-related tests (ALT and AST, at a minimum) prior to each increase in dose or monthly, whichever occurs first.
Negative pregnancy test for childbearing women? ☐ Yes ☐ No

Additional Pretreatment Lab Information (most recent):

Test	Result	Date
LDL – C (Treated)	mg/dL	
LDL – C (Untreated)	mg/dL	
Triglycerides	mg/dL	

Patient on Apheresis? ☐ Yes ☐ No If Yes, frequency: _____
Date of First Apheresis: _____

Positive patient history of hypercholesterolemia or premature CVD? ☐ Yes ☐ No

Family History on Both Sides? ☐ Yes ☐ No ☐ Unknown

Current Cholesterol Lowering Drugs and Dosage: *(if applicable)*

✓	Therapy	Dose	Duration
	Alirocumab	☐ 75 ☐ 150	
	Evolocumab	☐ 140 ☐ 420	
	Atorvastatin	☐ 10 ☐ 20 ☐ 40 ☐ 80	
	Rosuvastatin	☐ 5 ☐ 10 ☐ 20 ☐ 40	
	Ezetimibe	☐ 10	
	Niacin		
	BAS (Bile Acid Sequestrants)		
	Fibrates		
	Other: _____		

Tried/Failed Cholesterol Lowering Drugs and Dosage:

✓	Therapy	Dose	Duration
	Alirocumab	☐ 75 ☐ 150	
	Evolocumab	☐ 140 ☐ 420	
	Atorvastatin	☐ 10 ☐ 20 ☐ 40 ☐ 80	
	Rosuvastatin	☐ 5 ☐ 10 ☐ 20 ☐ 40	
	Ezetimibe	☐ 10	
	Niacin		
	BAS (Bile Acid Sequestrants)		
	Fibrates		
	Other: _____		

IV. PRESCRIBER INFORMATION

Prescriber Name: _____
Phone: _____ Fax: _____
Tax ID #: _____
NPI #: _____

Office Contact Name: _____
Office Contact Phone: _____
Office Contact Email: _____
License # _____

REVIEW AND SIGN THE ACKNOWLEDGMENT AND AUTHORIZATION

☐ I acknowledge that I have obtained any required authorization or other permission necessary to release the patient's protected health information and the information on this form and any prescription to Aegerion Pharmaceuticals, its affiliates and their representatives, agents, and contractors ("Aegerion"), for the purposes of providing product support services, including but not limited to conveying personal information to dispensing pharmacies. I further certify that any service provided through Compass on behalf of any patient is not made in exchange for any express or implied agreement or understanding that I would recommend, prescribe, or use JUXTAPID or any other Aegerion product or service for anyone, and any decision to prescribe JUXTAPID was and will in the future be based solely on my determination of medical necessity, and that I will not seek reimbursement for any medication or service provided by or through Compass from any government program or third-party insurer.

☐ I understand that my patient may authorize Aegerion to provide HoFH and JUXTAPID education, including compliance and persistency support. I understand that this program does not include individual treatment or medical advice to the patient, and it does not replace the medical treatment and care provided by me the patient's healthcare provider. I acknowledge that the education provided by Aegerion does not replace any obligation I have to inform the patient of the risks associated with JUXTAPID or any other treatment I may prescribe.

☐ I understand and agree that JUXTAPID product support services may include dispensing of vitamin E, linoleic acid, alpha-linolenic acid (ALA), eicosapentaenoic acid (EPA), and docosahexaenoic acid (DHA) supplements as required per package labeling.

Prescriber Signature: _____ Date: _____

Please see accompanying full Prescribing Information including Medication Guide and Boxed Warning.

COMPASS PATIENT AUTHORIZATION FORM

Juxtapid (lomitapide) capsules

The purpose of this Authorization form is to permit Compass participants to receive additional information and support ("Patient Support") from Aegerion Pharmaceuticals, its affiliates, representatives, agents and contractors ("Aegerion"). Please read this form carefully and ask any questions that you may have.

To be read, completed, and signed by patient or patient's personal representative.

PLEASE FAX TO 1-855-898-2498

I. AUTHORIZATION TO SHARE PROTECTED HEALTH INFORMATION

By signing this Authorization, I authorize Accredo Specialty Pharmacy ("Accredo") to disclose my contact information and protected health information (or "PHI") related to JUXTAPID therapy and my disease management, including but not limited to my name, medical and pharmacy records and information relating to payment for my disease treatment, care management and health insurance, as well as all information provided on any JUXTAPID prescription or prescription related to my disease treatment, to Aegerion Pharmaceuticals, Inc., and those working on its behalf (collectively, "Aegerion") to provide the Patient Support.

II. AUTHORIZATION FOR COMPASS SERVICES AND COMMUNICATIONS

The purpose of this Authorization is to enable me to obtain Patient Support from Aegerion, including:

- Investigation of my insurance coverage
- Coordination of benefits and reimbursement support
- Investigation of financial support services and programs, or comparable programs for JUXTAPID that may help me
- Facilitation of claims adjudication and submission of claims to third party payers for payment
- Education and access to patient programs related to JUXTAPID including medication adherence support, nutrition support and access to a registered dietitian, and treatment and medication reminders
- Participation in surveys and quality assessment activities to evaluate the effectiveness of the Patient Support

The Authorization also enables me to receive Marketing communications from Aegerion or those acting on its behalf offering programs, services or products of interest to patients taking JUXTAPID.

Aegerion is authorized to contact me by mail, e-mail, text, telephone, and/or any alternative communication method that I request in connection with the Patient Support.

Once my PHI has been disclosed to Aegerion, I understand that federal privacy laws may no longer protect that PHI. However, Aegerion will take reasonable steps to protect my PHI by using and disclosing it only for the purposes described in this Authorization or as otherwise authorized by law.

I understand that I may refuse to sign this Authorization, and that doing so will not affect my ability to receive treatment or benefits to which I am otherwise entitled. I understand that I am entitled to a copy of this Authorization, and that I may revoke this Authorization at any time, by mailing a letter requesting revocation to: Accredo Health Group, Inc. c/o The JUXTAPID Program, 1640 Century Center Parkway Memphis, TN 38134.

I understand that expiration of or revoking this Authorization will end further use and disclosure of my PHI but that it will not affect use or disclosure of PHI that has already been disclosed by Accredo in reliance upon this Authorization.

This Authorization will expire upon my revocation or one year after I receive my last prescription.

AGREED:

Patient Signature: _____ Date: _____

Patient Name (please print): _____

Personal Representative or Guardian Signature (if applicable): _____

Personal Representative or Guardian Name (please print): _____

Relationship to Patient, including the authority for status as Personal Representative: _____

Address of Patient or Personal Representative: _____

Telephone Number: _____ Email Address: _____

Please see Indication and Important Safety Information on pages 3-4 and accompanying full Prescribing Information including Medication Guide and Boxed Warning.