Juxtapid®
(lomitapide) capsules

Getting Started Guide

To get a patient started on Juxtapid® follow 3 steps outlined in this guide.

Important Safety Information about Juxtapid

INDICATION:

Juxtapid® (lomitapide) 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 (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high-density lipoprotein cholesterol (non-HDL-C) 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.

WARNING: RISK OF HEPATOTOXICITY

JUXTAPID can cause elevations in transaminases. In the JUXTAPID clinical trial, 10 (34%) of the 29 patients treated with 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.

Please see additional Important Safety Information throughout and accompanying full Prescribing Information, including Boxed Warning.

Visit chiesitotalcare.com or call 1-855-898-2743. We're ready to help!

Chiesi TOTAL care℠

STEP 1
REMS Training

Complete Risk Evaluation and Mitigation Strategy (REMS) training

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**.

A Risk Evaluation and Mitigation Strategy (REMS) is a strategy to manage known or potential serious risks associated with a drug product and is required by the Food and Drug Administration (FDA) to ensure that the benefits of a drug outweigh its risks.

To complete your REMS Prescriber Certification:

- Review the JUXTAPID Prescribing Information and the Fact Sheet
- Visit the Juxtapid REMS program website to complete the online prescriber training module and THEN FAX THE PRESCRIBER ENROLLMENT FORM to 1- 855-898-2498.

Counsel each new patient on the risk of hepatotoxicity and the need for baseline and periodic monitoring using the Patient Guide. Complete a Patient-Prescriber Acknowledgement Form with each patient.

FPO

Scan the QR code to visit the site.

STEP 2
Prescription

Fill out the Prescription Forms

A prescription for Juxtapid can only be written on the Juxtapid REMS Authorization Form. Complete, sign, and fax the completed form to 1-855-898-2498

FPO

Scan the QR code to download the form.

IMPORTANT SAFETY INFORMATION

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.

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PRESCRIBER ATTESTATION

By signing this form, I attest that:

- JUXTAPID® (lomitapide) capsules are only available through the JUXTAPID REMS and that I must comply with the program requirements in order to prescribe JUXTAPID.
- I have reviewed the **Prescribing Information**, **Fact Sheet** and **Prescriber Training Module**.
- Successfully completed the **Prescriber Knowledge Assessment** and submitted it to the JUXTAPID REMS.

Use:

- I understand that JUXTAPID is only 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 (LDL-C), total cholesterol (TC), apolipoprotein B (apo B) and non-high density lipoprotein cholesterol (non-HDL-C) in patients with homozygous familial hypercholesterolemia (HoFH).
- I understand that the safety and effectiveness of JUXTAPID has not been established in patients with hypercholesterolemia who do not have HoFH, including those with heterozygous familial hypercholesterolemia (HeFH).

Hepatotoxicity Risk:

- I understand that there is a risk of hepatotoxicity associated with JUXTAPID.
- I understand the Recommendations for Monitoring of Transaminases with JUXTAPID treatment:

Before treatment initiation (first dose), I must:

- Counsel patients on the approved indication for use in patients with HoFH, the risk of hepatotoxicity with JUXTAPID and the need for regular monitoring using the **Patient Guide**.
- Provide the patient a copy of the **Patient Guide**.
- Enroll the patient by completing and submitting the **Patient-Prescriber Acknowledgement Form** to the JUXTAPID REMS.
- Assess the patient to confirm a clinical or laboratory diagnosis consistent with the approved indication.
- Assess the patient's liver function.
- Order the prescription using the **Prescription Authorization Form**.

Lab Requirements:

- I must assess liver-related tests for this patient as recommended in the JUXTAPID Prescribing Information and in the chart below.

Monitoring of Transaminases

Before Initiating therapy
– Measure ALT, AST, alkaline phosphatase, and total bilirubin.

During the first year
– Measure liver-related tests (ALT and AST, at a minimum) **monthly** or prior to each increase in dose, whichever occurs first.

After the first year
– Measure liver-related tests (ALT and AST, at a minimum) at least **every 3 months** and before any increase in dose.

During Treatment:

- I agree to complete and sign the **Prescription Authorization Form** for each prescription.
- I agree that personnel from the JUXTAPID REMS may contact me to gather further information or resolve discrepancies or to provide other information related to JUXTAPID or the JUXTAPID REMS.
- I agree that Chiesi, its agents and contractors such as the pharmacy providers may contact me via phone, mail, or email to survey me on the effectiveness of the program requirements for the JUXTAPID REMS.

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A

Prescriber Signature:_____ Date:_____

Prescriber Name:_____ Phone:_____

Prescriber Enrolln

IMPORTANT

Prescription Authorization Form

Juxtapid® (lomitapide) capsules

REMS Program

This form must be completed and signed for each JUXTAPID prescription.
Fax this form to 1-855-898-2498.

ALL FIELDS ARE REQUIRED | PLEASE PRINT

PATIENT INFORMATION

First Name:_____ Middle Initial:____ Last Name:_____

Address:_____ Phone:_____

City:_____ Email:_____

State:_____ Zip:_____ Date of Birth:_____

JUXTAPID PRESCRIPTION

B

Dose:_____ *mg po q hs* (recommended starting dosage is 5 mg daily). Quantity to dispense:_____ Refills:_____

Additional Instructions:_____

PRESCRIBER INFORMATION

First Name:_____ Middle Initial:____ Last Name:_____

Practice/Facility Name:_____

A

Complete prescriber attestation

Juxtapid Can Only Be Prescribed in Accordance with the FDA-Approved Indication: Juxtapid is indicated only as an adjunct to a low-fat diet and other lipid-lowering treatments, including LDL apheresis where available, to reduce low-density lipoprotein cholesterol (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high-density lipoprotein cholesterol (non-HDL-C) in patients with homozygous familial hypercholesterolemia (HoFH).

B

Complete the prescription and confirm dosing

Attest that patients have a diagnosis consistent with homozygous familial hypercholesterolemia (HoFH) and affirm.

STEP 3
Support

Once you have completed the form

1. Attach copies of patient insurance and prescription cards – front and back.

2. First prescription for the patient:

Fax completed form to Chiesi Total Care℠ at 1-855-898-2743.
Please complete one form per patient.

3. Subsequent prescriptions:

Each new prescription must be submitted on the REMS PAF form which may include up to 11 refills.

4. Let your patients know they will be receiving a call from Chiesi Total Care.

A member of the Chiesi Total Care team will contact patients to complete and fulfill their prescriptions for Juxtapid.

FPO

The fillable PDF can be downloaded and saved for future use.
Scan the QR code to download a copy.

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LIMITATIONS OF USE:

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WARNING: RISK OF HEPATOTOXICITY

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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.

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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 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 Chiesi Farmaceutici S.p.A. at 1-888-661-9260 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Please see additional Important Safety Information throughout and accompanying full Prescribing Information, including Boxed Warning.

Reference: 1. Juxtapid® (lomitapide) Prescribing Information. Amryt, February 2022.

For more information, visit juxtapid.com.

Chiesi Total Care℠ Program offered through Accredo Specialty Pharmacy.

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