Juxtapid

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 policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

This policy document describes the status of medical technology or treatment at the time the document was developed. Since that time, new technology or treatment may have emerged or new medical literature may have been published. This policy will be reviewed regularly and be updated as scientific and medical literature becomes available.

DESCRIPTION

The intent of the Juxtapid 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 a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

Juxtapid is 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.

POLICY

Required Documentation

The following information is necessary to initiate the prior authorization review:

- Untreated baseline LDL level, LDL levels while receiving statin therapy (prior to initiating therapy with Juxtapid) and current LDL levels on Juxtapid (if applicable)
- Chart notes demonstrating statin intolerance or contraindication to statin therapy (if applicable)
- Lab results (i.e. LDL-receptor, apo B-100, PCSK9 gain-of-function, or LDL receptor adaptor protein 1/ARH adaptor protein 1 gene locus mutation) or rating scale (i.e. Simon-Broome Diagnostic Criteria or Dutch Lipid Network Criteria) demonstrating homozygous familial hypercholesterolemia diagnosis

Criteria for Initial Approval

A. Juxtapid® (lomitapide) may be considered **medically necessary** when the following criteria are met:

1.) Prescriber must be a lipid specialist or a cardiometabolic specialist, unless the patient resides in an area where access to these specialists are limited, in which case, the prescriber must be a board-certified cardiologist or endocrinologist.

2.) Patient has a diagnosis of homozygous familial hypercholesterolemia confirmed by ONE of the following:

a. Genetic diagnosis with documented mutations in both alleles at LDL receptor, ApoB, PCSK9, or LDL receptor adaptor protein 1/ARH adaptor protein 1 gene locus

OR

b. Clinical diagnosis defined as untreated LDL-C greater than 500 mg/dL OR unknown untreated LDL-C with treated LDL-C >300 mg/dL plus one of the following:

i.) Tendon or cutaneous xanthomas before age 10

ii.) Diagnosis of definite FH by genetic analysis, Simon-Broome Diagnostic Criteria or Dutch Lipid Clinic Network Criteria in both parents (Appendix A)

3.) Patient has been unable to achieve an LDL-C of ≤ 100 mg/dL (or ≤ 70 mg/dL with clinical atherosclerotic cardiovascular disease [ASCVD]) despite adherence† to at least three months of the following lipid lowering therapy:

a. Trial of BOTH high-intensity statins (atorvastatin 40-80 mg and rosuvastatin 20-40 mg) at a maximum tolerated dose in combination with ezetimibe; OR TWO moderate intensity statins (e.g. pravastatin 40-80 mg, lovastatin 40 mg, fluvastatin 80 mg, pitavastatin 2-4 mg, simvastatin 20-40 mg) in combination with ezetimibe, only in the event the patient is unable to complete either of the high-intensity statin trials at the maximum approved dosing

AND

b. Trial of a PCSK9-targeted therapy

4.) Confirmation of provider enrollment in REMS program

5.) If female of childbearing age, documented conversation of no current pregnancy or plans to become pregnant while on treatment

6.) Not to be used in combination with PCSK9-targeted therapy

7.) Member will continue to receive concomitant lipid-lowering therapy.

OR

1.) Prescriber must be a lipid specialist or a cardiometabolic specialist, unless the patient resides in an area where access to these specialists are limited, in which case, the prescriber must be a board-certified cardiologist or endocrinologist.

2.) Patient has a diagnosis of homozygous familial hypercholesterolemia confirmed by ONE of the following:

a. Genetic diagnosis with documented mutations in both alleles at LDL receptor, ApoB, PCSK9, or LDL receptor adaptor protein 1/ARH adaptor protein 1 gene locus

OR

b. Clinical diagnosis defined as untreated LDL-C greater than 500 mg/dL OR unknown untreated LDL-C with treated LDL-C >300 mg/dL plus one of the following:
    i.) Tendon or cutaneous xanthomas before age 10
    ii.) Diagnosis of definite FH by genetic analysis, Simon-Broome Diagnostic Criteria or Dutch Lipid Clinic Network Criteria in both parents (Appendix A)

3.) Patient has been unable to achieve an LDL-C of ≤ 100 mg/dL (or ≤ 70 mg/dL with clinical atherosclerotic cardiovascular disease [ASCVD]) despite adherence† to at least three months of a PCSK9-targeted therapy

4.) Patient has a documented contraindication (e.g., active liver disease, pregnancy, breastfeeding), or medically justifiable reason that precludes statin use (e.g. patients has experienced rhabdomyolysis, CK elevations ≥ 10x ULN, or statin intolerance).
    a. Statin intolerance shall be defined in accordance with the National Lipid Association definition:
        i.) Inability to tolerate at least two statins (one at any dose, one at lowest daily dose) due to objectionable symptoms or abnormal biomarkers temporally related to statin use, reversible upon statin discontinuation and reproducible by re-challenge while excluding other known determinants. Other known determinants include low body mass index (BMI), acute infection, untreated or undertreated hypothyroidism, severe renal or hepatic dysfunction, organ transplant, recent severe trauma, HIV infection, Vitamin D deficiency, history of creatine kinase elevation, history of preexisting or unexplained muscle or joint pain, high level of physical activity, illicit drug abuse, excess alcohol use. Each statin trial, both initial and re-challenge shall be at least two weeks duration.
        ii.) A trial of one statin at lowest starting daily dose
            - Rosuvastatin 5mg
            - Atorvastatin 10mg
            - Simvastatin 10mg
            - Lovastatin 20mg
            - Pravastatin 40mg
            - Fluvastatin 40mg
            - Pitavastatin 2mg
        iii.) One statin at any daily dose

5.) Confirmation of provider enrollment in REMS program

6.) If female of childbearing age, documented conversation of no current pregnancy or plans to become pregnant while on treatment

7.) Not to be used in combination with PCSK9-targeted therapy

Approval will be for **6 months**

Continuation of Therapy

All members (including new members) requesting authorization for continuation of therapy with Juxtapid must meet all initial authorization criteria and have achieved or maintained an LDL-C reduction greater than 20% from the levels immediately prior to initiation of treatment with Juxtapid.

Approval will be for **12 months**

Juxtapid is considered **not medically necessary** for patients who do not meet the criteria set forth above.

†Please note: Documentation of LDL-C levels are required (untreated baseline and current [within 30 days of prior authorization request]);

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 Apply

- Juxtapid 30 capsules per 30 days

APPENDIX

APPENDIX A: Diagnosis of familial hypercholesterolemia (FH)

A definite diagnosis of FH is made when one of the following diagnostic criteria is met:

1. Genetic diagnosis
   a) An LDL-receptor mutation, familial defective apo B-100, or a PCSK9 gain-of-function mutation
2. Simon-Broome Diagnostic Criteria for definite FH
   a) Total cholesterol > 290 mg/dL or LDL-C > 190 mg/dL in patients over 16 years of age or total cholesterol > 260 mg/dL or LDL-C > 155 mg/dL in patients less than 16 years of age
   AND
   b) Tendon xanthomas in the patient, first (parent, sibling or child) or second degree relative (grandparent, uncle or aunt)
3. Dutch Lipid Clinic Network Criteria for definite FH
   a) Total score > 8 points

CLINICAL RATIONALE

Homozygous familial hypercholesterolemia (HoFH) is quite rare, affecting around 1:1,000,000 persons (approximately 300 people in the United States). HoFH is generally identified via severely elevated low density lipoprotein cholesterol (LDL-C) in the absence of secondary causes of hypercholesterolemia; cardiovascular events can occur in the second decade of life. There are two sets of diagnostic criteria used for diagnosis (the World Health Organization/Dutch Lipid Network and Simon-Broome Register); genetic testing is also available. Juxtapid is approved for the treatment of HoFH patients. Juxtapid is an oral microsomal triglyceride transfer protein inhibitor that can reduce LDL-C by up to 40 percent when combined with maximally tolerated lipid-lowering therapies and LDL apheresis. Both drugs have black box warnings and are available through risk evaluation and mitigation strategy (REMS) because of the risk of hepatotoxicity.

PROCEDURES AND BILLING CODES

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

- Not applicable

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*Some content reprinted from CVSHealth

POLICY HISTORY

Policy #: 05.01.91
Policy Creation: December 2015
Reviewed: April 2022
Revised: April 2022
Current Effective Date: July 1, 2022