

## **RX.PA.040.MPC Juxtapid® (Lomitapide) and Evkeeza (Evinacumab-dgnb)**

The purpose of this policy is to define the prior authorization process for Juxtapid® (lomitapide) and Evkeeza (evinacumab-dgnb).

Juxtapid® (lomitapide) 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).

Evkeeza (evinacumab-dgnb) is indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 12 years and older, with homozygous familial hypercholesterolemia (HoFH).

### **DEFINITIONS**

N/A

### **POLICY**

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Pharmacy & Therapeutics Committee of the criteria for prior authorization, as described in RX.002 Pharmacy and Therapeutics Committee and RX.003-Prior Authorization Process. The drugs, Juxtapid® (lomitapide) and Evkeeza (evinacumab-dgnb), are subject to the prior authorization process.

### **PROCEDURE**

#### **Initial Authorization Criteria:**

*Must meet all of the general criteria listed below and product-specific criteria:*

#### **General Criteria (applies to all drugs):**

- Must be prescribed by or in consultation with a clinical lipidologist
- Must have a diagnosis of homozygous familial hypercholesterolemia. Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other possible diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as ONE of the four following scenarios:
  - Must have documented functional mutation(s) in both LDL receptor alleles or alleles known to affect LDL receptor functionality
  - Must have untreated total cholesterol (TC) >500mg/dL and triglycerides (TG) <300mg/dL and have both parents with untreated TC >250mg/dL and LDL-C level >500mg/dL
  - Must have untreated TC >500mg/dL and TG <300mg/dL and have both parents with untreated TC >250mg/dL and ONE of the following:
    - Skin fibroblast LDL receptor activity <20% normal
    - Presence of cutaneous and tendon xanthomas and corneal arcus in the first decade of life
  - Must have untreated LDL-C level >500mg/dL and ONE of the following:
    - Skin fibroblast LDL receptor activity <20% normal

**Juxtapid® (Lomitapide) and Evkeeza (evinacumab-dgnb)**

**POLICY NUMBER: RX.PA.040.MPC**

**REVISION DATE: 08/2021**

**PAGE NUMBER: 2 of 3**

- Presence of cutaneous and tendon xanthomas and corneal arcus in the first decade of life
- Must be on at least TWO LDL-lowering medications from different classes (e.g., statins, nicotinic acid, ezetimibe)
  - One of these medications must be an HMG-CoA Reductase Inhibitor unless contraindicated or intolerant
- Must have an adequate trial (of at least 3 months) of Praluent (18 years and older) or Repatha (13 years and older) with an inadequate response or significant side effects/toxicity or have a contraindication to therapy

**For Juxtapid:**

- Must be age 18 years and older
- Must have the following baseline tests (within one month of initiation and dates of tests must be provided):
  - For females of reproductive potential:
    - Must have a negative pregnancy test prior to starting Juxtapid
    - Must be using effective contraception
  - Transaminase (ALT and AST), alkaline phosphatase, and bilirubin levels
  - LDL-C level
- Must not have moderate or severe hepatic impairment (Child-Pugh category B or C) or active liver disease
- Must not be on concomitant treatment with moderate or strong CYP3A4 inhibitors (e.g., amprenavir, aprepitant, atazanavir, ciprofloxacin, crizotinib, darunavir/ritonavir, diltiazem, erythromycin, fluconazole, fosamprenavir, imatinib, verapamil, boceprevir, clarithromycin, conivaptan, indinavir, itraconazole, ketoconazole, lopinavir/ritonavir, mibefradil, nefazodone, nelfinavir, posaconazole, ritonavir, saquinavir, telaprevir, telithromycin, voriconazole)

**For Evkeeza:**

- Must be age 12 years and older
- Must have the following baseline tests (within one month of initiation and dates of tests must be provided):
  - For females of reproductive potential:
    - Must have a negative pregnancy test prior to starting Juxtapid
    - Must be using effective contraception
  - LDL-C level

**Reauthorization Criteria:**

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon chart documentation from the prescriber that the member's condition has improved based upon the following:

- Documentation of improvement in the condition based upon the prescriber's assessment while on treatment
- Documentation of reduction in LDL levels since starting treatment
- **For Juxtapid Only:** Documentation of laboratory monitoring of transaminase, alkaline phosphatase, and bilirubin levels during treatment

**Limitations:**

Length of Authorization (if above criteria met)
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**Juxtapid® (Lomitapide) and Evkeeza (evinacumab-dgnb)****POLICY NUMBER: RX.PA.040.MPC****REVISION DATE: 08/2021****PAGE NUMBER: 3 of 3**

Initial Authorization	Up to 4 months
Reauthorization	Up to 1 year
<b>Quantity Level Limit</b>	
<u>Juxtapid</u> Capsule	30 capsules per 30 days

If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.

**HCPCS Codes:**

Code	Description
J1305 (previously C9079)	Injection, Evinacumab-dgnb, 5 mg

**REFERENCES**

1. Cuchel M, Meagher E, du Toit Theron H, et al. Efficacy and Safety of a Microsomal Triglyceride Transfer Protein Inhibitor in Patients with Homozygous Familial Hypercholesterolemia: a Single Arm, Open-Label, Phase 3 Study. *The Lancet*. 2012; published online at [http://dx.doi.org/10.1016/S0140-6736\(12\)61731-0](http://dx.doi.org/10.1016/S0140-6736(12)61731-0).
2. Juxtapid [package insert]. Cambridge, MA: Aegerion Pharmaceuticals; May 2015 .
3. Repatha [prescribing information]. Thousand Oaks, CA: Amgen Inc; August 2015.
4. Raal FJ, Honarpour N, Blom DJ et al. Inhibition of PCSK9 with evolocumab in homozygous familial hypercholesterolaemia (TESLA Part B): a randomised, double-blind, placebo-controlled trial. *Lancet*. 2015 Jan 24;385(9965):341-50.
5. Raal FJ, Santos RD. Homozygous familial hypercholesterolemia: current perspectives on diagnosis and treatment. *Atherosclerosis* 2012; 223: 262–68.

**RECORD RETENTION**

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

**REVIEW HISTORY**

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
<i>Criteria Update</i>	01/16
<i>Annual Review</i>	02/16, 02/17, 02/18, 02/19, 02/20, 03/21
<i>Addition of Evkeeza</i>	07/21