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) , areis subject to the prior authorization process.

PROCEDURE
Initial Authorization Criteria:
Must meet all of the general criteria listed belowand 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

        - 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)

Initial Authorization	Up to 4 months
Reauthorization	Up to 1 year
Quantity Level Limit	
Juxtapid 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.
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
Criteria Update	*01/16*
Annual Review	*02/16, 02/17, 02/18, 02/19, 02/20, 03/21*
Addition of Evkeeza	*07/21*
Evkeeza assigned HCPCS code update	*10/21*