UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2025 P 2072-13
Program	Prior Authorization/Medical Necessity
Medication	Juxtapid® (lomitapide)
P&T Approval Date	10/2015, 9/2016, 9/2017, 9/2018, 9/2019, 9/2020, 7/2021, 7/2022, 7/2023, 2/2024, 2/2025
Effective Date	5/1/2025

1. Background:

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). The safety and efficacy 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.

2. Coverage Criteria[a]:

A. Initial Authorization

1. **Juxtapid** will be approved based on **all** of the following criteria:

   a. Diagnosis of homozygous familial hypercholesterolemia (HoFH) as confirmed by **one** of the following:

      (1) Submission of medical records (e.g., chart notes, laboratory values) confirming genetic confirmation of bi-allelic pathogenic/likely pathogenic variants on different chromosomes at the low-density lipoprotein receptor *(LDLR)*, apolipoprotein B *(APOB)*, proprotein convertase subtilisin kexin type 9 (*PCSK9)*, or low-density lipoprotein receptor adaptor protein 1 (*LDLRAP1)* genes or ≥ 2 such variants at different loci

      -OR-

      (2) **Both** of the following:

         (a) Untreated low-density lipoprotein cholesterol (LDL-C) greater than 400 mg/dL

         -AND-

         (b) **One** of the following:

            i. Xanthoma before 10 years of age
            ii. Evidence of familial hypercholesterolemia in at least one parent

-AND-

b. Patient is on a low-fat diet

-AND-

c. Patient is receiving other lipid-lowering therapy (e.g., statin, ezetimibe, LDL apheresis)

-AND-

d. Prescribed by **one** of the following:

(1) Cardiologist
(2) Endocrinologist
(3) Lipid specialist

-AND-

e. **One** of the following:

(1) **Both** of the following:

(a) History of intolerance, failure or contraindication to Repatha (evolocumab)

-AND-

(b) History of intolerance, failure or contraindication to Evkeeza (evinacumab)

-OR-

(2) Patient is currently on Juxtapid therapy

-AND-

f. Not used in combination with a proprotein convertase subtilisin/kexin type 9 (PCSK9) inhibitor [e.g., Praluent (alirocumab), Repatha (evolocumab)]

-AND-

g. Not used in combination with Evkeeza (evinacumab-dgnb)

Authorization will be issued for 12 months.

B. Reauthorization

1. **Juxtapid** will be approved based on **all** of the following criteria:

a. Patient is on a low-fat diet

-AND-

b. Patient continues to receive other lipid-lowering therapy (e.g., statin, LDL apheresis)

-AND-

c. Documentation of a positive clinical response to therapy from pre-treatment baseline

-AND-

d. Prescribed by **one** of the following:

(1) Cardiologist
(2) Endocrinologist
(3) Lipid specialist

-AND-

e. Not used in combination with a proprotein convertase subtilisin/kexin type 9 (PCSK9) inhibitor [e.g., Praluent (alirocumab), Repatha (evolocumab)]

-AND-

f. Not used in combination with Evkeeza (evinacumab-dgnb)

Authorization will be issued for 12 months.

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

4. Reference:

1. Juxtapid [package insert]. Cambridge, MA: Amryt Pharmaceuticals; September 2020.
2. Cuchel M, Bruckert E, Ginsberg HN, et al. Homozygous familial hypercholesterolaemia: new insights and guidance for clinicians to improve detection and clinical management. A position paper from the Consensus Panel on Familial Hypercholesterolaemia of the European Atherosclerosis Society. Eur Heart J. 2014; 35:2146-57.
3. Cuchel M, Raal FJ, Hegele RA, et al. 2023 Update on European Atherosclerosis Society Consensus Statement on Homozygous Familial Hypercholesterolaemia: new treatments and clinical guidance. Eur Heart J. 2023;44(25):2277-2291. doi:10.1093/eurheartj/ehad197

Program	Prior Authorization/Medical Necessity - Juxtapid® (lomitapide)
Change Control	
10/2015	New program.
7/2016	Added Indiana and West Virginia coverage information.

9/2016	Annual Review. Updated references.
11/2016	Administrative change. Added California coverage information.
9/2017	Annual review. Removed requirement of medical record submission for diagnosis documentation. Updated state mandate verbiage.
9/2018	Annual review with no changes to coverage criteria. Updated reference.
9/2019	Annual review. Removed criteria regarding combination therapy with Kynamro as Kynamro no longer on market.
9/2020	Annual review with no changes to coverage criteria. Updated reference.
7/2021	Added continuation of coverage to background and criteria. Added Evkeeza as step through agent. Updated reference.
7/2022	Annual review. No updates to criteria.
7/2023	Annual review. Updated background. Updated diet requirement, not used in combination with PCSK9, and removed submission of medical records. Removed genetic testing coverage footnote.
2/2024	Updated diagnostic criteria per European Atherosclerosis Society guidance. Changed initial authorization period to 12 months. Updated references.
2/2025	Updated diet requirement per label. Added requirement to not be used in combination with Evkeeza. Revised HoFH criteria to include more precise genetic terminology to account for genetic test result interpretation complexity as well as digenic mutations.