HAP CareSource™

PHARMACY POLICY STATEMENT
HAP CareSource™ Marketplace

DRUG NAME	Juxtapid (lomitapide)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Juxtapid, approved by the FDA in 2012, 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).

Juxtapid (lomitapide) will be considered for coverage when the following criteria are met:

Homozygous Familial Hypercholesterolemia (HoFH)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a cardiologist or lipid specialist; AND
3. Member has a diagnosis of homozygous familial hypercholesterolemia (HoFH) confirmed by one of the following:
   a) Genetic testing confirmation of two mutant alleles in the *LDLR, Apo-B, PCSK9*, or *LDLRAP1* gene locus; OR
   b) LDL-C > 400 mg/dL before any lipid-lowering drug treatment AND at least one of the following:
      i) Cutaneous or tendon xanthoma before 10 years of age; and/or
      ii) Evidence of heterozygous familial hypercholesterolemia (HeFH) in both parents; AND
4. Chart notes must include documentation of baseline LDL-C above goal within the past 90 days; AND
5. Member is unable to achieve LDL-C goal (see Note) after 8-week trials with ALL of the following:
   a) High-intensity /max-tolerated statin in combination with ezetimibe (unless there is documentation of clearly established statin intolerance or statin contraindication—see note*); and
   b) PCSK9 inhibitor (e.g., Repatha or Praluent; prior authorization required) unless there is evidence of no LDL receptor function (receptor-negative type HoFH); AND
6. Juxtapid will be used as an adjunct to other lipid-lowering treatments (e.g., statins, ezetimibe, PCSK9 inhibitor, LDL apheresis, etc.), unless contraindicated or intolerant; AND
7. Prescriber attests that the member will be on a low-fat diet during treatment; AND
8. Member does NOT have moderate to severe hepatic impairment; AND
9. Juxtapid is not being concomitantly initiated with Evkeeza.
10. **Dosage allowed/Quantity limit:** Start 5 mg once daily and titrate up to 60 mg once daily per prescribing information.

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(QL: 60 capsules per 30 days for the 20 mg or 30 mg capsules; 30 capsules per 30 days for the 5 mg or 10 mg capsules)

NOTE: The LDL-C goals for adults are <70 mg/dL or <55 mg/dL with additional ASCVD risk factors.

**NOTE: If not on statin therapy, member must have documented contraindication to all statin drugs or documentation of intolerance to at least 2 different statins, including low/moderate intensity or alternate dosing such as every other day*.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes/labs show at least 15% LDL-C reduction since starting Juxtapid.

If all the above requirements are met, the medication will be approved for an additional 12 months.

HAP CareSource considers Juxtapid (lomitapide) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
7/22/2020	New policy for Juxtapid created.
04/05/2021	Added Praluent to list of alternative products. Changed trials to include Praluent in addition to Repatha and increased trial length to 90 days. Added a trial requirement for concomitant request of Juxtapid and Evkeeza. Updated genetic testing requirement to ask for specific alleles (previously not specified). Updated atorvastatin high-intensity requirement to reflect pediatric vs. adult dosing.
02/22/2022	Policy for Juxtapid transferred to new template. Corrected the quantity limit from 30/30 to 60/30; highest capsule strength is 30mg.
05/25/2023	Added references. Changed “baseline cholesterol” to “baseline LDL-C above goal.” Added receptor-negative as exception to PCSK9 requirement. Simplified statement regarding Evkeeza (does not prohibit using them together, but they may not be started at the same time).
05/07/2024	Updated references. Removed LDL cutoff for HeFH in parents. Changed LDL cutoffs of 500 or 300 to 400; specified at least 15% LDL reduction for reauth; changed LDL goals of 100 or 70 to 70 or 55; changed statin/ezetimibe and PCSK9 trials from 90 days to 8 weeks (EAS/Cuchel 2023). Added note about statin intolerance. Added that the member does not have moderate to severe hepatic impairment (contraindication).

References:

1. Juxtapid [Package insert]. Cambridge, MA: Aegerion Pharmaceuticals, Inc; 2020.
2. Alonso R, Cuevas A, Mata P. Lomitapide: a review of its clinical use, efficacy, and tolerability. Core Evid. 2019;14:19-30. Published 2019 Jul 1.
3. Robinson JG. Management of familial hypercholesterolemia: a review of the recommendations from the National Lipid Association Expert Panel on Familial Hypercholesterolemia. *J Manag Care Pharm*. 2013;19(2):139-149. doi:10.18553/jmcp.2013.19.2.139

4. Gidding SS, Champagne MA, de Ferranti SD, et al. The Agenda for Familial Hypercholesterolemia: A Scientific Statement From the American Heart Association [published correction appears in Circulation. 2015 Dec 22;132(25):e397]. *Circulation*. 2015;132(22):2167-2192. doi:10.1161/CIR.0000000000000297
5. France M, Rees A, Datta D, et al. HEART UK statement on the management of homozygous familial hypercholesterolaemia in the United Kingdom. *Atherosclerosis*. 2016;255:128-139. doi:10.1016/j.atherosclerosis.2016.10.017
6. Mach F, Baigent C, Catapano AL, et al. 2019 ESC/EAS Guidelines for the management of dyslipidaemias: lipid modification to reduce cardiovascular risk [published correction appears in Eur Heart J. 2020 Nov 21;41(44):4255]. *Eur Heart J*. 2020;41(1):111-188. doi:10.1093/eurheartj/ehz455
7. Watts GF, Gidding SS, Hegele RA, et al. International Atherosclerosis Society guidance for implementing best practice in the care of familial hypercholesterolaemia. *Nat Rev Cardiol*. 2023;20(12):845-869. doi:10.1038/s41569-023-00892-0
8. 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

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