

# Specialty Guideline Management

## Juxtapid

### Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Juxtapid	lomitapide

### Indications

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 Indications<sup>1</sup>

Juxtapid is indicated as an adjunct to a low-fat diet and exercise and other low density lipoprotein cholesterol (LDL-C) therapies to reduce low-density lipoprotein cholesterol (LDL-C) in adults and pediatric patients aged 2 years and older with homozygous familial hypercholesterolemia (HoFH).

All other indications are considered experimental/investigational and not medically necessary.

### Documentation

Submission of the following information is necessary to initiate the prior authorization review:

Reference number(s)
2071-A

## Initial Requests:

- Genetic testing, chart notes, or medical record documentation confirming the diagnosis of HoFH.
- Current LDL-C level dated within the six months preceding the authorization request.
- With clinical atherosclerotic cardiovascular disease (ASCVD): Chart notes confirming clinical ASCVD (if applicable) (see Appendix).
- For members 10 years of age and older: chart notes, medical record documentation, or claims history confirming the member is currently on maximally tolerated lipid-lowering therapy.
- For members 7 years of age to less than 10 years of age: chart notes, medical record documentation, or claims history confirming the member is currently on maximally tolerated lipid-lowering therapy (if applicable). If therapy is not advisable, documentation of clinical reason to avoid therapy.

## Continuation Requests:

- Current LDL-C level dated within the six months preceding the authorization request.
- For members 10 years of age and older: chart notes, medical record documentation, or claims history confirming the member is currently on maximally tolerated lipid-lowering therapy.
- For members 7 years of age to less than 10 years of age: chart notes, medical record documentation, or claims history confirming the member is currently on maximally tolerated lipid-lowering therapy (if applicable). If therapy is not advisable, documentation of clinical reason to avoid therapy.

## Prescriber Specialties

This medication must be prescribed by or in consultation with a cardiologist, endocrinologist, lipid specialist, geneticist, or a prescriber specialized in the treatment of HoFH.

## Coverage Criteria

### Homozygous Familial Hypercholesterolemia (HoFH)<sup>1-5</sup>

Authorization of 6 months may be granted for treatment of homozygous familial hypercholesterolemia in members 2 years of age and older when all of the following criteria are met:

- Member has a documented diagnosis of homozygous familial hypercholesterolemia confirmed by any of the following criteria:

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- Variant in two low-density lipoprotein receptor (LDLR) alleles.
- Presence of homozygous or compound heterozygous variants in apolipoprotein B (APOB) or proprotein convertase subtilisin-kexin type 9 (PCSK9) gene.
- Member has compound heterozygosity or homozygosity for variants in the gene encoding low-density lipoprotein receptor adaptor protein 1 (LDLRAP1).
- Member has an untreated LDL-C of greater than 400 milligrams per deciliter (mg/dL) and has either of the following:
  - Presence of cutaneous or tendinous xanthomas before the age of 10 years.
  - An untreated LDL-C level of greater than or equal to 190 mg/dL in both parents.
- Prior to initiation of treatment with the requested medication, member meets/has met either of the following criteria:
  - Member has a treated LDL-C level greater than or equal to 70 mg/dL.
  - Member has a treated LDL-C level greater than or equal to 55 mg/dL and meets either of the following criteria:
    - Member has a history of a clinical ASCVD event (see Appendix).
    - Member has major ASCVD risk factors (e.g., 65 years of age or older, familial hypercholesterolemia, diabetes, chronic kidney disease, history of congestive heart failure).
- Prior to initiation of treatment with the requested medication, member meets/has met one of the following criteria:
  - Member is 10 years of age or older and meets both of the following criteria:
    - Member is receiving stable treatment with at least 3 lipid-lowering therapies (e.g., statins, ezetimibe, PCSK9 directed therapy) at the maximally tolerated dose.
    - Member will continue to receive concomitant lipid-lowering therapy at the maximally tolerated dose.
  - Member is 7 years of age to less than 10 years of age and meets either of the following criteria:
    - Member is receiving stable treatment with at least one lipid-lowering therapy (e.g., statins, LDL apheresis) at the maximally tolerated dose and will continue to receive concomitant lipid-lowering therapy at the maximally tolerated dose.
    - Member has an intolerance or contraindication to other lipid-lowering therapies.
  - Member is 2 years of age to less than 7 years of age.

## Continuation of Therapy

Authorization of 12 months may be granted for continued treatment in members 2 years of age and older (including new members) who meet all of the following criteria:

- Member has had at least 20% reduction of LDL-C from baseline.
- Member meets one of the following criteria:
  - Member is 10 years of age or older and is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
  - Member is 7 years of age to less than 10 years of age and meets either of the following criteria:
    - Member is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
    - Member has an intolerance or contraindication to other lipid-lowering therapies.
  - Member is 2 years of age to less than 7 years of age.

## Appendix

### Clinical ASCVD<sup>5-8</sup>

- Acute coronary syndromes
- Myocardial infarction
- Stable or unstable angina
- Coronary or other arterial revascularization procedure (e.g., percutaneous coronary intervention [PCI], coronary artery bypass graft [CABG] surgery)
- Stroke of presumed atherosclerotic origin
- Transient ischemic attack (TIA)
- Non-cardiac peripheral arterial disease (PAD) of presumed atherosclerotic origin (e.g., carotid artery stenosis, lower extremity PAD)
- Obstructive coronary artery disease (defined as greater than or equal to 50% stenosis on cardiac computed tomography angiogram or catheterization)
- Coronary artery calcium (CAC) Score greater than or equal to 300

## References

1. Juxtapid [package insert]. Parma, Italy: Chiesi Farmaceutici S.p.A.; February 2026.
2. Mach F, Koskinas KC, Roeters van Lennep JE, et al. 2025 Focused update of the 2019 ECS/EAS guidelines for the management of dyslipidaemias. *Eur Heart J.* 2025; 46: 4359-4378.
3. Cuchel M, Raal FJ, Hegele RA, et al. Update on European atherosclerosis society consensus statement on homozygous familial hypercholesterolaemia: new treatments and clinical guidance. *Eur Heart J.* 2023;44(25):2277-2291.
4. Lloyd-Jones DM, Morris PB, Ballantyne CM, et al. 2022 ACC Expert consensus decision pathway on the role of nonstatin therapies for LDL-cholesterol lowering in the management of atherosclerotic

Reference number(s)
2071-A

cardiovascular disease risk: A report of the American college of cardiologic solution set oversight committee. J Am Coll Cardiol. 2022;80(14):1366–1418.

5. Grundy SM, Stone NJ, Bailey AL, et al. 2018 AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA guideline on the management of blood cholesterol: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation. 2019;139:e1082– e1143.
6. Jacobson TA, Ito MK, Maki KC, et al. National Lipid Association recommendations for patient-centered management of dyslipidemia: part 1 — full report. J Clin Lipidol. 2015;9:129–169.
7. Min JK, Labounty TM, Gomez MJ, et al. Incremental prognostic value of coronary computed tomographic angiography over coronary artery calcium score for risk prediction of major adverse cardiac events in asymptomatic diabetic individuals. Atherosclerosis. 2014;232(2):298-304.
8. Budoff MJ, Kinninger A, Gransar H, et al. When does a calcium score equate to secondary prevention?: Insights from the multinational CONFIRM registry. JACC Cardiovasc Imaging. 2023;16(9):1181-1189.