



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Juxtapid

Page: 1 of 3

Effective Date: 8/17/2023

Last Review Date: 6/8/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input checked="" type="checkbox"/> Florida Kids
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Michigan
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input type="checkbox"/> Arizona

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Juxtapid under the patient's prescription drug benefit.

Description:

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 Indication

Juxtapid 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 (APOB), and non-high-density lipoprotein cholesterol (non-HDL-C) in patients with homozygous familial hypercholesterolemia (HoFH).

Limitations of Use:

- The safety and effectiveness 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.

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

Applicable Drug List:

Juxtapid

Policy/Guideline:

Documentation:

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

- A. Current LDL-C level for both initial requests and continuation requests. The level must be dated within the six months preceding the authorization request.
- B. Genetic testing or medical records confirming the diagnosis of HoFH.



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Juxtapid

Page: 2 of 3

Effective Date: 8/17/2023

Last Review Date: 6/8/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input checked="" type="checkbox"/> Florida Kids
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Michigan
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input type="checkbox"/> Arizona

C. Medical records confirming the member is currently on lipid lowering therapy for both initial requests and continuation requests.

Criteria for Initial Approval:

Homozygous familial hypercholesterolemia (HoFH)

Authorization of 6 months may be granted for treatment of homozygous familial hypercholesterolemia when all of the following criteria are met:

- A. Member has a documented diagnosis of homozygous familial hypercholesterolemia confirmed by any of the following criteria:
 - 1. Variant in two low-density lipoprotein receptor (LDLR) alleles
 - 2. Presence of homozygous or compound heterozygous variants in apolipoprotein B (APOB) or proprotein convertase subtilisin-kexin type 9 (PCSK9)
 - 3. Member has compound heterozygosity or homozygosity for variants in the gene encoding low-density lipoprotein receptor adaptor protein 1 (LDLRAP1)
 - 4. An untreated LDL-C of greater than 500 mg/dL or treated LDL-C greater than or equal to 300 mg/dL and either of the following:
 - a. Presence of cutaneous or tendinous xanthomas before the age of 10 years
 - b. An untreated LDL-C level of greater than or equal to 190 mg/dL in both parents
- B. Prior to initiation of treatment with the requested medication, both of the following criteria are/were met:
 - 1. Member is/was receiving a combination lipid-lowering regimen consisting of a high-intensity statin, ezetimibe, and PCSK9 directed therapy unless the member has known LDL-receptor negative mutations in both alleles.
 - 2. Member is/was experiencing an inadequate response to such a combination regimen, as demonstrated by a treated LDL-C of greater than or equal to 100 mg/dL (or greater than or equal to 70 mg/dL with clinical atherosclerotic cardiovascular disease [ASCVD]), unless the member has known LDL-receptor negative mutations in both alleles.
- C. Member will continue to receive concomitant lipid-lowering therapy.

Continuation of Therapy:

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

- A. Member meets all initial authorization criteria
- B. Member has had at least 20% reduction of LDL-C from baseline
- C. Member is currently receiving concomitant lipid-lowering therapy

Approval Duration and Quantity Restrictions:

Approval:



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Juxtapid Page: 3 of 3

Effective Date: 8/17/2023 Last Review Date: 6/8/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input checked="" type="checkbox"/> Florida Kids
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Michigan
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input type="checkbox"/> Arizona

- Initial: 6 months; renewal: 12 months

Quantity Level Limits:

- Juxtapid 5 mg Capsule: 28 per 28 days
- Juxtapid 10 mg Capsule: 28 per 28 days
- Juxtapid 20 mg Capsule: 56 per 28 days
- Juxtapid 30 mg Capsule: 56 per 28 days

References:

1. Juxtapid [package insert]. Cambridge, MA: Aegerion Pharmaceuticals, Inc.; September 2020
2. Nordestgaard BG, Chapman MJ, Humphries SE, et al. Familial hypercholesterolaemia is underdiagnosed and undertreated in the general population: guidance for clinicians to prevent coronary heart disease Consensus Statement of the European Atherosclerosis Society. [published correction appears in *Eur Heart J*. 2020 Dec 14;41(47):4517] *Eur Heart J* 2013;34(45):3478–90a. doi:10.1093/eurheartj/eh273
3. 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(32):2146-2157. doi:10.1093/eurheartj/ehu274
4. National Institute for Health and Clinical Excellence (2008). Identification and management of familial hypercholesterolaemia. NICE clinical guideline 71
5. Cuchel M, Meagher EA, du Toit Theron H, et al. Efficacy and safety of a microsomal triglyceride transfer protein inhibitor in patients with homozygous familial hypercholesterolaemia: a single-arm, open-label, phase 3 study. *Lancet*. 2013;381(9860):40-46. doi:10.1016/S0140-6736(12)61731-0
6. Goldberg AC, Hopkins PN, Toth PP, et al. Familial hypercholesterolemia: screening, diagnosis and management of pediatric and adult patients. Clinical guidance from the National Lipid Association Expert Panel on Familial Hypercholesterolemia. *J Clin Lipidol*. 2011;5(3 Suppl):S1–S8. doi:10.1016/j.jacl.2011.04.003
7. Raal JF, Santos RD. Homozygous familial hypercholesterolemia: current perspectives on diagnosis and treatment. *Atherosclerosis*. 2012;223(2):262-268. doi:10.1016/j.atherosclerosis.2012.02.019
8. Bays HE, Jones PH, Orringer CE, Brown WV, Jacobson TA. National Lipid Annual Summary of Clinical Lipidology 2016. *J Clin Lipidol* 2016;10(1 Suppl):S1-S43. doi:10.1016/j.jacl.2015.08.002
9. Lloyd-Jones DM, Morris PB, Ballantyne CM, et al. 2017 focused update of the 2016 ACC expert consensus decision pathway on the role of non-statin therapies for LDL-cholesterol lowering in the management of atherosclerotic cardiovascular disease risk: a report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents. *J Am Coll Cardiol* 2017;70(14):1785–1822. doi:10.1016/j.jacc.2017.07.745