AETNA BETTER HEALTH® Coverage Policy/Guideline					
Name:	Juxtapid			Page:	1 of 3
Effective Date:	4/30/2024			Last Review Date:	4/2024
Applies to:	☒Illinois ☐New Jersey ☒Pennsylvania Kids	☐Florida ☒Maryland ☐Virginia		☒Florida Kids ☒Michigan ☐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:

Both initial and continuation requests:
A. Genetic testing or medical records confirming the diagnosis of HoFH.
B. LDL-C level dated within the six months preceding the authorization request.

C. Chart notes, medical record documentation, or claims history confirming the member is currently on lipid-lowering therapy.

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 400 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:

- 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]. Dublin, Ireland: Amryt Pharmaceuticals, Inc.; September 2020
2. 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.
3. 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.
4. McGowan MP, Hosseini Dehkordi SH, Moriarty PM, et al. Diagnosis and treatment of heterozygous familial hypercholesterolemia. *J Am Heart Assoc*. 2019; 8(24):e013225.
5. Bays HE, Jones PH, Orringer CE, et al. National Lipid Annual Summary of Clinical Lipidology 2016. *J Clin Lipidol 2016*;10(1 Suppl):S1-S43.
6. 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 cardiovascular disease risk: A report of the American college of cardiologic solution set oversight committee. *J Am Coll Cardiol.* 2022;80(14):1366–1418.