Pharmacy Drug Policy & Procedure

Policy Name:	Juxtapid (lomitapide)	Policy #:	2389P

Purpose of the Policy

The purpose of this policy is to establish the criteria for coverage of Juxtapid.

Statement of the Policy

Health Alliance Medical Plans will approve the use of Juxtapid under the specialty pharmacy benefit when the following criteria have been met.

Criteria

1. Initial Coverage for the Treatment of Homozygous Familial Hypercholesterolemia (HoFH)

1.1 Documented diagnosis of homozygous familial hypercholesterolemia, confirmed by one of the following:
- Documentation of two identified mutations in any one of the following genes: LDL receptor (LDLR), Apo B, or Proprotein convertase subtilisin/kexin type 9 (PCSK9) gene, OR
- A formal clinical diagnosis of familial hypercholesterolemia made using one of the following tools:
  - U.S. Make Early Diagnosis Prevent Early Death (MEDPED)
  - Dutch Lipid Clinical Network
  - Simon-Broome Registry

1.2 Ordered by a cardiologist (heart doctor) or lipid specialist

1.3 Age 18 or older

1.4 Triglyceride level < 400mg/dL

1.5 Documentation of ACC/AHA 10-year risk calculation of 7.5% or greater

1.6 Lab value of treated low-density lipoprotein (LDL) level greater than 100mg/dL within the last 30 days,

1.7 Documentation to support one of the following:
- Documented failure on high-intensity statin therapy (atorvastatin 80mg, Crestor 20mg, Crestor 40mg), in combination with ezetimibe, defined as being unable to decrease LDL level by 50%
  - Failure is defined as an inability to decrease LDL level by 50%, and corresponding claims history supporting that the member has filled at least 150 days of both medications in the last six (6) months
- Documented intolerance to high-intensity statin therapy (defined as severe myalgias/muscle aches and/or creatinine kinase levels greater than 10 times the upper limit of the lab reference range), AND documented failure on medium-intensity statin therapy (atorvastatin 10mg, Crestor 5mg, simvastatin 20 to 40mg, pravastatin 40mg, lovastatin 40mg, fluvastatin XL 80mg, fluvastatin 40mg twice daily, or Livalo 2 to 4mg) used in combination with ezetimibe
  - Failure is defined as an inability to decrease LDL level by 50%, and corresponding claims history supporting that the member has filled at least 150 days of both medications in the last six (6) months

1.8 Documentation to support one of the following:
- Documented failure after 3 doses of Repatha 420mg in combination with high-intensity statin therapy and ezetimibe unless use of these agents is contraindicated, defined as being unable to decrease LDL level by 30%
  - Failure is defined as an inability to decrease LDL level by 30%, and corresponding claims history supporting that the member has filled at least 84 days of Repatha in at least four (4)

months
- Member with a confirmed diagnosis of LDL receptor-negative homozygous familial hyperlipidemia will bypass step through Repatha, OR
  - Repatha was not effective at lowering LDL levels in these patients per TESLA Part B trial

1.9 Documentation that the member will discontinue PCSK9 inhibitor therapy prior to the initiation of Juxtapid
- Safety and efficacy of this combination has not been studied

1.10 Negative urine pregnancy test in female patient of reproductive potential prior to treatment

1.11 Documentation that liver function tests will be collected every 4 weeks and after each dose escalation during the first year

1.12 Documentation that liver function test will be collected at least every 3 months and prior to dosage increases after the first year

1.13 Confirmation that the member will use daily supplements containing vitamin E 400 units, linoleic acid 200mg, alpha-linolenic acid (ALA) 210mg, eicosapentaenoic acid (EPA) 110mg, and docosahexaenoic acid (DHA) 80mg

1.14 Confirmation that the member will initiate and maintain a low-fat diet supplying < 20% of energy from fat

2. Renewal Coverage for the Treatment of Homozygous Familial Hypercholesterolemia (HoFH)

2.1 Submission of subsequent liver function tests taken every 4 weeks for the first year

2.2 Submission of liver function tests taken every 3 months after the first year

2.3 At least 75% compliance with Juxtapid, maximum tolerated statin in combination with ezetimibe

2.4 Documented 20% or more reduction in LDL from pre-treatment level

3. Approval Period

3.1 Initial Approval: 12 months

3.2 Subsequent Approvals: 12 months with documented clinical benefit

4. Managed Dose Limit

4.1 Juxtapid is limited to dispensing of 28 capsules per 28 days

CPT Codes

HCPCS Codes

References

1. Juxtapid (lomitapide) [prescribing information]. Dublin, Ireland US: Amryt Pharmaceuticals DAC; September 2020.
2. 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 hypercholesterolemia: a single-arm, open-label, phase 3 study. Lancet. 2013; 381:40–46.
3. 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, S1–S8.
4. 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. J Am Coll Cardiol. 2019 Jun 25;73(24):e285- e350.

5. American College of Cardiology Solution Set Oversight Committee 2022 ACC Expert Consensus Decision Pathway on the Role of Nonstatin Therapies for LDL-Cholesterol Lowering in the Management of Atherosclerotic Cardiovascular Disease Risk. J Am Coll Cardiol. 2022 Oct 4;80(14):1366-1418.

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