SPECIALTY GUIDELINE MANAGEMENT

JUXTAPID (lomitapide)

POLICY

I. 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 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, apolipoprotein B, and non-high-density lipoprotein cholesterol in patients with homozygous familial hypercholesterolemia (HoFH).

All other indications are considered experimental/investigational and are not a covered benefit.

II. PRESCRIBER SPECIALTIES

The prescriber must be a lipid specialist, cardiometabolic specialist, cardiologist or an endocrinologist.

III. CRITERIA FOR APPROVAL

Homoygous familial hypercholesterolemia (HoFH)
Authorization for 12 months may be granted for members who meet ALL of the criteria listed below:
A. Member has a diagnosis of HoFH confirmed by genetic analysis or clinical criteria (See Appendices):
B. Prior to initiation of treatment with Juxtapid, patient is/was receiving a combination lipid-lowering regimen consisting of a high-intensity statin, ezetimibe, and evolocumab (Repatha).
C. Prior to initiation of treatment with Juxtapid, patient is/was experiencing an inadequate response to such a combination regimen, as demonstrated by one of the following:
   1. Treated LDL-C greater than or equal to 160 mg/dL
   2. Treated LDL-C greater than or equal to 100 mg/dL with a history of any of the following:
      i. Clinically evident coronary heart disease or other atherosclerotic cardiovascular disease
      ii. Diabetes
      iii. A family history of very early coronary heart disease (less than 45 years of age in men and less than 55 years of age in women)
      iv. Current smoking
      v. Two or more coronary heart disease risk factors
      vi. Lipoprotein(a) levels of 50 mg/dl or greater

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members (including new members) who meet all initial authorization criteria and have achieved or maintained a LDL-C reduction greater than 20% from the levels immediately prior to initiation of treatment with Juxtapid after at least 12 months of treatment.
V. APPENDICES

APPENDIX A. Diagnosis of Homozygous FH

- Genetic diagnosis
  - Mutations in both alleles at LDL receptor, ApoB, PCSK9 or LDL receptor adaptor protein/ARH gene locus
- Clinical diagnosis
  - Untreated LDL-C > 500 mg/dL OR unknown untreated LDL-C with treated LDL-C > 300 mg/dL
  - One of the following:
    - Tendon or cutaneous xanthomas at age 10 or younger
    - Diagnosis of definite FH by genetic analysis, Simon-Broome Diagnostic Criteria or Dutch Lipid Clinic Network Criteria (See Appendix B) in both parents
    - Evidence of FH in both parents with a history including any of the following:
      - Total cholesterol ≥ 310 mg/dL
      - Premature ASCVD (before 55 years of age in men and 60 years of age in women)
      - Tendon xanthoma
      - Sudden premature cardiac death

APPENDIX B: Diagnosis of familial hypercholesterolemia (FH)

A definite diagnosis of FH is made when one of the following diagnostic criteria is met:

- Genetic diagnosis
  - An LDL-receptor mutation, familial defective apo B-100, or a PCSK9 gain-of-function mutation
- Simon-Broome Diagnostic Criteria for definite FH
  - Total cholesterol > 290 mg/dL or LDL-C > 190 mg/dL, plus tendon xanthomas in the patient, first (parent, sibling or child) or second degree relative (grandparent, uncle or aunt)
- Dutch Lipid Clinic Network Criteria for definite FH
  - Total score > 8 points

VI. REFERENCES