CareFirst Family of health care plans

CVS caremark™

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. CRITERIA FOR APPROVAL

Homozygous 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 a treated LDL-C of greater than or equal to 100 mg/dL.

III. 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.

IV. APPENDICES

APPENDIX A. Diagnosis of Homozygous FH
- Genetic confirmation
  - Mutations in both alleles at LDL receptor, ApoB, PCSK9 or LDL receptor adaptor protein gene locus
- Clinical diagnosis
  - Untreated LDL-C > 500 mg/dL OR unknown untreated LDL-C with treated LDL-C > 300 mg/dL **plus**
  - One of the following:
    - Tendon or cutaneous xanthomas at age 10 or younger
    - Diagnosis of FH by 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 in men and 60 years in women)
  - Tendon xanthoma
  - Sudden premature cardiac death

APPENDIX B: Diagnosis of familial hypercholesterolemia (FH)

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

- Genetic confirmation
  - An LDL-receptor mutation, familial defective apo B-100, or a PCSK9 gain-of-function mutation
- Simon-Broome Diagnostic Criteria for FH
  - Total cholesterol > 290 mg/dL or LDL-C > 190 mg/dL in patients over 16 years of age or total cholesterol > 260 mg/dl or LDL-C > 155 mg/dl in patients less than 16 years of age and one of the following
    - Tendon xanthomas in the patient, first (parent, sibling or child) or second degree relative (grandparent, uncle or aunt)
    - Family history of myocardial infarction in a first degree relative before the age of 60 or in a second degree relative before the age of 50
    - Total cholesterol greater than 290 mg/dl in an adult first or second degree relative
    - Total cholesterol greater than 260 mg/dl in a child, brother, or sister aged younger than 16 years
- Dutch Lipid Clinic Network Criteria for FH
  - Total score > 5 points

V. REFERENCES

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