Evinacumab-dgnb (Evkeeza™)

IMPORTANT REMINDER

We develop Medical Policies to provide guidance to Members and Providers. This Medical Policy relates only to the services or supplies described in it. The existence of a Medical Policy is not an authorization, certification, explanation of benefits or a contract for the service (or supply) that is referenced in the Medical Policy. For a determination of the benefits that a Member is entitled to receive under his or her health plan, the Member's health plan must be reviewed. If there is a conflict between the medical policy and a health plan or government program (e.g., TennCare), the express terms of the health plan or government program will govern.

POLICY

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 Indications

Evkeeza is indicated as an adjunct to diet and exercise and other low-density lipoprotein-cholesterol (LDL-C) lowering therapies to reduce LDL-C in adult and pediatric patients, aged 1 year and older, with homozygous familial hypercholesterolemia (HoFH).

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

DOCUMENTATION

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

Initial and Continuation Requests:

- Genetic testing or medical records confirming the diagnosis of HoFH.
- LDL-C level dated within the six months preceding the authorization request.
- With clinical atherosclerotic cardiovascular disease (ASCVD): Chart notes confirming clinical ASCVD (if applicable) (see Appendix).
- For members 10 years of age and older: chart notes, medical record documentation, or claims history confirming the member is currently on maximally tolerated lipid-lowering therapy.
- For members 7 years of age to less than 10 years of age: chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.

COVERAGE CRITERIA

Homozygous Familial Hypercholesterolemia (HoFH)

Authorization of 6 months may be granted for members 1 year of age or older for treatment of homozygous familial hypercholesterolemia (HoFH) when all of the following criteria are met:

- Member has a documented diagnosis of HoFH confirmed by any of the following criteria:
  - Variant in two low-density lipoprotein receptor (LDLR) alleles.

  - Presence of homozygous or compound heterozygous variants in apolipoprotein B (APOB) or proprotein convertase subtilisin-kexin type 9 (PCSK9) gene.
  - Member has compound heterozygosity or homozygosity for variants in the gene encoding low-density lipoprotein receptor adaptor protein 1 (LDLRAP1).
  - Member has an untreated LDL-C of > 400 mg/dL and has either of the following:
    - Presence of cutaneous or tendinous xanthomas before the age of 10 years.
    - An untreated LDL-C level of ≥ 190 mg/dL in both parents.
- Prior to initiation of treatment with the requested medication, member meets/ has met either of the following criteria:
  - Member has a treated LDL-C level ≥ 70 mg/dL
  - Member has a treated LDL-C level ≥ 55 mg/dL and meets either of the following criteria:
    - Member has a history of a clinical ASCVD event (see Appendix).
    - Member has major ASCVD risk factors (e.g., 65 years of age or older, familial hypercholesterolemia, diabetes, chronic kidney disease, history of congestive heart failure).
- Prior to initiation of treatment with the requested medication, member meets/has met one of the following criteria:
  - Member is 10 years of age or older and meets both of the following criteria:
    - Member is receiving stable treatment with at least 3 lipid-lowering therapies (e.g., statins, ezetimibe, PCSK9 directed therapy) at the maximally tolerated dose.
    - Member will continue to receive concomitant lipid-lowering therapy at the maximally tolerated dose.
  - Member is 7 years of age to less than 10 years of age and meets either of the following criteria:
    - Member is receiving stable treatment with at least one lipid-lowering therapy (e.g., statins, LDL apheresis) at the maximally tolerated dose and will continue to receive concomitant lipid-lowering therapy at the maximally tolerated dose.
    - Member has an intolerance or contraindication to other lipid-lowering therapies.
  - Member is 1 year of age to less than 7 years of age.

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:

- Member meets all requirements in the coverage criteria.
- Member meets one of the following criteria:
  - Member is 10 years of age or older and is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
  - Member is 7 years of age to less than 10 years of age and meets either of the following criteria:
    - Member is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
    - Member has an intolerance or contraindication to other lipid-lowering therapies.
  - Member is 1 year of age to less than 7 years of age.
- The member is receiving benefit from therapy. Benefit is defined as either of the following:
  - LDL-C is now at goal.
  - Member has had at least 30% reduction of LDL-C from baseline.

APPENDIX

Clinical ASCVD

- Acute coronary syndromes
- Myocardial infarction
- Stable or unstable angina

- Coronary or other arterial revascularization procedure (e.g., percutaneous coronary intervention [PCI], coronary artery bypass graft [CABG] surgery)
- Stroke of presumed atherosclerotic origin
- Transient ischemic attack (TIA)
- Non-cardiac peripheral arterial disease (PAD) of presumed atherosclerotic origin (e.g., carotid artery stenosis, lower extremity PAD)
- Obstructive coronary artery disease (defined as ≥ 50% stenosis on cardiac computed tomography angiogram or catheterization)
- Coronary artery calcium (CAC) Score ≥ 300

APPLICABLE TENNESSEE STATE MANDATE REQUIREMENTS

BlueCross BlueShield of Tennessee's Medical Policy complies with Tennessee Code Annotated Section 56-7-2352 regarding coverage of off-label indications of Food and Drug Administration (FDA) approved drugs when the off-label use is recognized in one of the statutorily recognized standard reference compendia or in the published peer-reviewed medical literature.

ADDITIONAL INFORMATION

For appropriate chemotherapy regimens, dosage information, contraindications, precautions, warnings, and monitoring information, please refer to one of the standard reference compendia (e.g., the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) published by the National Comprehensive Cancer Network®, Drugdex Evaluations of Micromedex Solutions at Truven Health, or The American Hospital Formulary Service Drug Information).

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Policy

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