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

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
Evkeeza is indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 5 years and older, with homozygous familial hypercholesterolemia (HoFH).

Limitations of Use:
- The safety and effectiveness of Evkeeza have not been established in patients with other causes of hypercholesterolemia, including those with heterozygous familial hypercholesterolemia (HeFH).
- The effects of Evkeeza on cardiovascular morbidity and mortality have not been determined.

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

II. 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. 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.
D. 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.

III. CRITERIA FOR INITIAL APPROVAL

Homozygous familial hypercholesterolemia (HoFH)
Authorization of 6 months may be granted for members 5 years of age and older for the treatment of homozygous familial hypercholesterolemia when both 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:
   i. Presence of cutaneous or tendinous xanthomas before the age of 10 years.
   ii. 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 has 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]).
2. Member meets one of the following:
   i. Member is 10 years of age or older and meets both of the following:
      a. Member is receiving stable treatment with at least 3 lipid-lowering therapies (e.g., statins, ezetimibe, proprotein convertase subtilisin/kexin type 9 (PCSK9) directed therapy) at the maximally tolerated dose.
      b. Member will continue to receive concomitant lipid-lowering therapy at the maximally tolerated dose.
   ii. Member is 7 years of age to less than 10 years of age and meets one of the following:
      a. 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.
      b. Member has an intolerance or contraindication to other lipid-lowering therapies.
   iii. Member is 5 years of age to less than 7 years of age.

IV. 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 meets one of the following:
1. Member is 10 years of age or older and is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
2. Member is 7 years of age to less than 10 years of age and meets either of the following:
   a. Member is currently receiving concomitant lipid-lowering therapy at the maximally tolerated dose.
   b. Member has an intolerance or contraindication to other lipid-lowering therapies.
3. Member is 5 years of age to less than 7 years of age.

C. The member is receiving benefit from therapy. Benefit is defined as either of the following:
1. LDL-C is now at goal.
2. Member has had at least 30% reduction of LDL-C from baseline.

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).

REFERENCES

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6. ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). Evaluate the efficacy and safety of evinacumab in pediatric patients With homozygous familial hypercholesterolemia. Identifier: NCT04233918. Updated June 7, 2023. Accessed November 13, 2023. https://clinicaltrials.gov/ct2/show/record/NCT04233918.
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EFFECTIVE DATE 7/31/2024

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