Evinacumab-dgnb (EVKEEZA®)
Criteria for Use
September 2021

VA Pharmacy Benefits Management Services, Medical Advisory Panel, and VISN Pharmacist Executives

The following recommendations are based on medical evidence, clinician input, and expert opinion. The content of the document is dynamic and will be revised as new information becomes available. The purpose of this document is to assist practitioners in clinical decision-making, to standardize and improve the quality of patient care, and to promote cost-effective drug prescribing. THE CLINICIAN SHOULD USE THIS GUIDANCE AND INTERPRET IT IN THE CLINICAL CONTEXT OF THE INDIVIDUAL PATIENT. INDIVIDUAL CASES THAT ARE EXCEPTIONS TO THE EXCLUSION AND INCLUSION CRITERIA SHOULD BE ADJUDICATED AT THE LOCAL FACILITY ACCORDING TO THE POLICY AND PROCEDURES OF ITS P&T COMMITTEE AND PHARMACY SERVICES.

The Product Information should be consulted for detailed prescribing information.

See the VA National PBM-MAP-VPE Monograph on this drug at the *PBM INTERnet* or *PBM INTRAnet* site for further information.

Exclusion Criteria

If the answer to ANY item below is met, then the patient should NOT receive evinacumab-dgnb.

- ☐ History of serious hypersensitivity reactions to evinacumab-dgnb or any of its excipients
- ☐ Does not have a confirmed diagnosis of Homozygous Familial Hypercholesterolemia (HoFH)
- ☐ Patient is receiving lomitapide
- ☐ Patient is pregnant

Inclusion Criteria

The answers to all of the following must be fulfilled in order to meet criteria.

- ☐ Care provided by VA/VA Community Care provider specializing in Cardiology, Endocrinology or locally designed specialty provider with expertise in lipid management
- ☐ Genetic or clinical confirmation of HoFH diagnosis
- ☐ Receiving maximally tolerated low-density lipoprotein cholesterol (LDL-C) lowering therapies including statins, ezetimibe and PCSK9 inhibitors and need for further LDL-C lowering to reduce cardiovascular disease (CVD) risk *(e.g., LDL-C remains ≥ 70 mg/dL in patients with CVD, ≥ 100 mg/dL without CVD, LDL-C reduction <50% from baseline and/or patient having recurrent CVD events)*
- ☐ Receiving LDL-C apheresis if patient is a candidate for therapy and if therapy is accessible
- ☐ Patient is willing and able to travel to receive evinacumab-dgnb infusions every 4 weeks

For patients who can become pregnant:

- ☐ Evaluate pregnancy status prior to initiating treatment since evinacumab-dgnb may cause fetal harm. Contraceptive counseling on the potential risks vs. benefits of taking evinacumab-dgnb if a patient were to become pregnant is recommended. Use of effective contraception is advised during treatment and for at least 5 months after the last dose.

Supplemental Information

Patient meets one or more of the following genetic, clinical or laboratory diagnostic criteria for homozygous familial hypercholesterolemia (HoFH):

☐ Confirmation with genetic testing (mutation in LDL receptor: true homozygote or compound heterozygote),

OR

☐ Untreated LDL-C of >500 mg/dL

OR

☐ LDL-C remains ≥300 mg/dL despite maximally tolerated, clinically indicated lipid-lowering therapy (e.g., statins, ezetimibe, PCSK9 inhibitors) and adherence is confirmed, **AND**

- Presence of any of the following physical findings including tendon xanthomas at any age, arcus corneae in patients <45 years or tuberous xanthomas or xanthelasma in patients <20 years.

Raal FJ, Santos RD. Homozygous Familial Hypercholesterolemia: Current Perspectives on Diagnosis and Treatment. Atherosclerosis 2012;223:262-268

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