Evinacumab-dgnb (Evkeeza™)	Place of Service Office Administration Infusion Center Administration Home Infusion Administration Outpatient Facility Administration* [*Prior authorization required – see section (1)] HCPCS: **J1305** per 5 mg

Condition listed in policy (*see criteria for details*)

- Homozygous familial hypercholesterolemia

AHFS therapeutic class: Antilipemic agents, miscellaneous

Mechanism of action: ANGPTL3 (angiopoietin-like 3) inhibitor

(1) Special Instructions and pertinent Information

Covered under the medical benefit, please submit clinical information for prior authorization review via fax.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****

AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

Members with the following plans: **PPO, Direct Contract HMO, and when applicable, Medi-Cal, ASO/Shared Advantage/HMO (non-direct contract),** may be required to have their medication administered at a preferred site of service, including the home, a physician's office, or an independent infusion center not associated with a hospital.

For members that cannot receive infusions in the preferred home or ambulatory setting AND meet one of the following criteria points, drug administration may be performed at a hospital outpatient facility infusion center.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****

AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

ADMINISTRATION OF EVKEEZA IN THE HOSPITAL OUTPATIENT FACILITY SITE OF CARE REQUIRES ONE OF THE FOLLOWING: (*Supporting Documentation must be submitted)*

1. **Patient is receiving their first infusion of Evkeeza or is being re-initiated on Evkeeza after at least 6 months off therapy.** *Subsequent doses will require medical necessity for continued use in the hospital outpatient facility site of care.*

Additional clinical monitoring is required during administration as evidenced by one of the following:

2. **Patient has experienced a previous severe adverse event to Evkeeza based on documentation submitted.**

3. **Patient continues to experience moderate to severe adverse events to Evkeeza based on documentation submitted, despite receiving premedication such as acetaminophen, steroids, diphenhydramine, fluids, etc.**

4. **Patient is clinically unstable based on documentation submitted.**

5. **Patient is physically or cognitively unstable based on documentation submitted.**

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for evinacumab-dgnb (Evkeeza™) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Homozygous familial hypercholesterolemia (HoFH)

1. Recommended by a cardiologist or endocrinologist, **AND**
2. Confirmed HoFH by either positive genetic test for LDL-R genetic mutations confirming HoFH or clinical evidence supporting a diagnosis of HoFH, **AND**
3. Being used in combination with a standard lipid lowering combination regimen (e.g., a high potency statin and a non-statin lipid lowering agent), **AND**
4. One of the following:
    a. Inadequate response, intolerance, or contraindication to a PCSK9 inhibitor (e.g., Praluent, Repatha), or
    b. Provider attestation that patient has homozygous null-null variants

Covered Doses
Up to 15 mg/kg IV once monthly (every 4 weeks)

Coverage Period
Indefinitely

ICD-10:
E78.01

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for evinacumab-dgnb (Evkeeza™) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s)

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional Information

How supplied:

- 345 mg/2.3 mL (150 mg/mL) single-dose vial
- 1,200 mg/8 mL (150 mg/mL) single-dose vial

HoFH diagnostic criteria based on based on the following:

HoFH Genetic Diagnosis	HoFH Clinical Diagnosis
Based on the following genetic tests: o LDLR DNA Sequence Analysis o LDLR Deletion/Duplication Analysis for large gene rearrangement testing-only if the Sequence Analysis is Negative	A clinical diagnosis of familial hypercholesterolemia is best made with the following clinical features: o High levels of total cholesterol and LDL cholesterol (e.g. LDL-C > 500 in untreated patients or LDL-C > 300 in treated patients), **and** o Therapy resistant elevated LDL in both parents, **and** o At least **one** of the following: ▪ Xanthomas (waxy deposits of cholesterol in the skin or tendons), ▪ Xanthelasmas (cholesterol deposits in the eyelids), ▪ Corneal arcus (cholesterol deposit around the cornea of the eye). ▪ A strong family history of high levels of total and LDL cholesterol and/or family or personal history or early cardiovascular event (stroke or heart attack) ▪ Symptoms consistent with ischemic heart disease (chest pain, shortness of breath), peripheral vascular disease (pain and numbness in the legs), or aortic stenosis (fatigue, shortness of breath) in the member.

Reference: Raal FJ and Santos RD. Homozygous familial hypercholesterolemia: Current perspectives on diagnosis and treatment. Atherosclerosis 2012; 223: 262-68.

AACE 2017 Atherosclerotic CVD Risk Stratification		LDL goal
High risk	ASCVD equivalent including diabetes or stage 3 or 4 CKD with no other risk factors, or individuals with 2 or more risk factors and a 10-year risk of 10%- 20%)	<100 mg/dL
Very high risk	Established or recent hospitalization for acute coronary syndrome (ACS); coronary, carotid or peripheral vascular disease; diabetes or stage 3 or 4 CKD with 1 or more risk factors; a calculated 10-year risk greater than 20%; or heterozygous familial hypercholesterolemia [HeFH])	<70 mg/dL
Extreme risk	Progressive ASCVD, including unstable angina that persists after achieving an LDL-C <70 mg/dL, or established clinical ASCVD in individuals with diabetes, stage 3 or 4 CKD, and/or HeFH, or in individuals with a history of premature ASCVD (<55 years of age for males or <65 years of age for females)	<55 mg/dL

(6) References

- AHFS®. Available by subscription at http://www.lexi.com
- DrugDex®. Available by subscription at http://www.micromedexsolutions.com/home/dispatch
- Evkeeza™ (evinacumab-dgnb) [Prescribing information]. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; 2/2021.

(7) Policy Update

Date of last review: 3Q2022
Date of next review: 3Q2023
Changes from previous policy version:

- No clinical change to policy following routine annual review

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee