Harvard Pilgrim Health Care | TUFTS Health Plan

# Medical Necessity Guidelines:
# Evkeeza™ (evinacumab-dgnb)

*Effective: December 12, 2023*

| **Prior Authorization Required**<br>If REQUIRED, submit supporting clinical documentation pertinent to service request. | Yes ☒ No ☐ |
|---|---|

**Applies to:**

**Commercial Products**

☐ Harvard Pilgrim Health Care Commercial products; Fax 617-673-0988
☐ Tufts Health Plan Commercial products; Fax 617-673-0988
CareLink℠ – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

**Public Plans Products**

☐ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product); Fax 617-673-0988
☒ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans; Fax 617-673-0939
☐ Tufts Health RITogether – A Rhode Island Medicaid Plan; Fax 617-673-0939
☐ Tufts Health One Care* – A Medicare-Medicaid Plan (a dual eligible product); Fax 617-673-0956
*The MNG applies to Tufts Health One Care members unless a less restrictive LCD or NCD exists.

**Senior Products**

☐ Harvard Pilgrim Health Care Stride Medicare Advantage; Fax 617-673-0956
☐ Tufts Health Plan Senior Care Options (SCO), (a dual-eligible product); Fax 617-673-0956
☐ Tufts Medicare Preferred HMO, (a Medicare Advantage product); Fax 617-673-0956
☐ Tufts Medicare Preferred PPO, (a Medicare Advantage product); Fax 617-673-0956

**Note:** While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

## Overview

**Food and Drug Administration (FDA) Approved Indications:**

- Evkeeza (evinacumab-dgnb) is indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 12 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.

Familial hypercholesterolemia results mainly from autosomal dominant genetic defects in the LDL-C receptor, apo B, or proprotein convertase subtilisin kexin type 9 (PCSK9), all of which are involved in the normal processing and trafficking of LDL-C. It is estimated that one in every 500 individuals in the United States has heterozygous familial hypercholesterolemia, while one in one million individuals is affected by HoFH. Patients with HoFH carry two of the same defective genes, while patients with the heterozygous form of the condition carry one defective gene.

For patients with HoFH, plasma LDL-C levels are often five times greater than normal, and in a small sample of patients with HoFH, untreated TC levels were commonly between 700 mg/dL and 800 mg/dL. Other signs and symptoms of HoFH are a deposition of cholesterol (xanthomas) in the skin and tendons, especially the elbows, knees, Achilles' tendon, and hands. Patients may also present with cholesterol deposits in the cornea (corneal arcus). Xanthomas may become apparent during childhood in patients with HoFH, and severe coronary artery disease resulting in myocardial infarction or requiring interventions such as coronary artery bypass grafting is often present by the age of 20 years.

## Clinical Guideline Coverage Criteria

**Initial Coverage Criteria:**

The Plan may authorize coverage of Evkeeza (evinacumab-dgnb) for members when **ALL** of the following criteria is met:

1. Documentation of diagnosis of homozygous familial hypercholesterolemia (HoFH) received by, or in written consultation with (documentation required), a lipid specialist (e.g., cardiologist, endocrinologist, lipid specialist/lipidologist)

**AND**

2. Confirmed diagnosis of homozygous familial hypercholesterolemia (HoFH) based on both of the following (a and b):
    a. Documentation of one of the following:
        i. Documented genetic mutation at both alleles of at least one of the following genes by an FDA approved generic test: low-density lipoprotein receptor (LDLR), Apolipoprotein B (apo B), or Proprotein convertase subtilisin kexin type 9 (PCSK9)

**OR**

        ii. Documentation that genetic testing was completed, and results are negative, where the provider attests the member has a mutation that may cause FH but remains unknown.

**AND**

    b. Recognized diagnostic criteria is met (documentation provided):
        i. Dutch Lipid Clinic criteria

**OR**

        ii. Simon Broome criteria

**AND**

3. Member is concurrently taking other lipid-lowering medication(s), or the provider indicates clinical inappropriateness for concurrent use with other lipid-lowering medication(s)

**AND**

4. Member is 12 years of age or older

**AND**

5. The Member has demonstrated an inadequate response to an appropriate trial with or a contraindication and or intolerance to each of the following classes:
    a. Maximally tolerated statin
    b. PCSK9 Inhibitor

**AND**

6. A baseline LDL-C is provided

**Reauthorization Criteria:**

The plan may reauthorize coverage of Evkeeza (evinacumab-dgnb) for members when both of the following criteria is met:

1. The member continues to meet all initial authorization criteria

**AND**

2. The member has achieved or maintained an LDL-C reduction while on medication

## Limitations

- The plan does not cover Evkeeza (evinacumab-dgnb) for patients with hypercholesterolemia who do not have homozygous familial hypercholesterolemia (HoFH).
- Initial authorization duration is for a period of 6 months.
- Reauthorization period is for a period of 12 months.

## Codes

The following code(s) require prior authorization:

**Table 1: HCPCS Codes**

| HCPCS Codes | Description |
|---|---|
| J1305 | Injection, evinacumab-dgnb, 5 mg |

## References


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15. Raal F, Rosenson R, Reeskamp L et al. Evinacumab-dgnb for homozygous familial hypercholesterolemia. N Engl J Med. 2020; 383(8):711-20.
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18. UpToDate: https://www.uptodate.com/contents/familial-hypercholesterolemia-in-adultsoverview?sectionName=DEFINITION&search=homozygous%20familial%20hypercholesterolemia&topicRef=122013&anchor=H4212208012&source=see_link#H4212208012 accesses 5/27/21.


## Approval And Revision History

September 13, 2022: Reviewed by Pharmacy and Therapeutics Committee (P&T).

September 21, 2022: Reviewed by the Medical Policy Approval Committee (MPAC).

Subsequent endorsement date(s) and changes made:

- December 12, 2023: No changes. Retire Medical Necessity Guideline effective 1/31/24. Effective February 1, 2024, coverage falls to Unified Medical Policies Medical Necessity Guideline.
- December 2023: Administrative update to rebrand Tufts Health Unify to Tufts Health One Care for 2024.

## Background, Product and Disclaimer Information

Medical Necessity Guidelines are developed to determine coverage for benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. We make coverage decisions using these guidelines, along with the Member's benefit document, and in coordination with the Member's physician(s) on a case-by-case basis considering the individual Member's health care needs.

Medical Necessity Guidelines are developed for selected therapeutic or diagnostic services found to be safe and proven effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in our service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. We revise and update Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern. For Tufts Health Together (Medicaid), coverage may be available beyond these guidelines for pediatric members under age 21 under the Early and Periodic Screening, Diagnostic and Treatment (EPSDT) benefits of the plan in accordance with 130 CMR 450.140 and 130 CMR 447.000, and with prior authorization.

Treating providers are solely responsible for the medical advice and treatment of Members. The use of this guideline is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to eligibility and benefits on the date of service, coordination of benefits, referral/authorization, utilization management guidelines when applicable, and adherence to plan policies, plan procedures, and claims editing logic.