Harvard Pilgrim Health Care | TUFTS Health Plan

Medical Necessity Guidelines:
Medical Benefit Drugs
Evkeeza (evinacumab-dgnb)

Effective: September 1, 2026

Guideline Type	☒ Prior Authorization ☐ Non-Formulary ☐ Step-Therapy ☐ Administrative

Applies to:

Commercial Products

☒ Harvard Pilgrim Health Care Commercial products; Fax 617-673-0988

☒ Tufts Health Plan Commercial products; Fax 617-673-0988

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

☐ 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 an angiopoietin-like 3 (ANGPTL3) inhibitor indicated as an adjunct to diet and exercise and other low-density lipoprotein-cholesterol (LDL-C) lowering therapies to reduce LDL-C in adults and pediatric patients, aged 1 year and older, with homozygous familial hypercholesterolemia (HoFH).

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 1 year 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
   c. Ezetimibe

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:

1. Cuchel M, Meagher E, Theron H et al. Efficacy and safety of a microsomal triglyceride transfer protein inhibitor in patients with homozygous familial hypercholesterolemia: a single -arm, open label, phase 3 study. Lancet. 2013; 381(9860):40-6.
2. Evkeeza prescribing information. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; September 2025.
3. Food and Drug Administration (FDA). Designating an orphan product: drug and biological products. 2020 April. URL: https://www.fda.gov/industry/developing-products-rare-diseasesconditions/designating-orphan-product-drugs-andbiological-products. Available from Internet. accessed 2021 February 16.
4. Food and Drug Administration (FDA). Drugs@FDA. URL: http://www.accessdata.fda.gov/scripts/cder/drugsatfda. Available from Internet. Accessed 2021a February 16.
5. Food and Drug Administration (FDA). FDA approves add-on therapy for patients with genetic form of severely high cholesterol. 2021b February 11. URL: https://www.fda.gov/drugs/news-events-human-drugs/fda-approves-add-therapy-patients-genetic-form-severely-high-cholesterol-0.

6. Food and Drug Administration (FDA). Frequently asked questions: breakthrough therapies. 2021c February. URL: https://www.fda.gov/regulatory-information/food-and-drug-administration-safetyand-innovation-act-fdasia/frequentlyasked-questions-breakthrough-therapies. Available from Internet. Accessed 2021 February 16.
7. Institute for Clinical and Economic Review (ICER). URL: https://icer -review.org/. Available from Internet. Accessed 2021 February 17.
8. Juxtapid prescribing information. Cambridge, MA: Aegerion Pharmaceuticals, Inc.; 2019 December.
9. Lloyd-Jones D, Morris P, Ballantyne C et al. 2017 focused update of the 2016 ACC expert consensus decision pathway on the role of non-statin therapies for LDL-cholesterol lowering in the management of atherosclerotic cardiovascular disease risk: a report of the American College of Cardiology Task Force on Expert Consensus Decision Pathways. J Am Coll Cardiol. 2017; 70(14):1785-822.
10. National Institute for Health and Care Excellence (NICE). Evinacumab-dgnb for treating homozygous familial hypercholesterolemia in people aged 12 and over [ID2704]. URL: https://www.nice.org.uk/guidance/proposed/gidta10655. Available from Internet. Accessed 2021b February 17.
11. National Institute for Health and Care Excellence (NICE). Familial hypercholesterolemia: identification and management. 2019 October. URL: https://www.nice.org.uk/guidance/cg71. Available from Internet. Accessed 2021 February 17.
12. National Institute for Health and Care Excellence (NICE). URL: https://www.nice.org.uk/. Available from Internet. Accessed 2021a February 17.
13. National Organization for Rare Disorders (NORD). Familial Hypercholesterolemia. 2020. URL: https://rarediseases.org/rare-diseases/familial-hypercholesterolemia. Available from Internet. Accessed 2021 February 18.
14. Raal F, Honarpour N, Blom D et al. Inhibition of PCSK9 with Evolocumab in homozygous familial hypercholesterolemia (TESLA part b): a randomized, double-blind, placebo-controlled trial. Lancet. 2015; 385(9965):341-50.
15. Raal F, Rosenson R, Reeskamp L et al. Evinacumab-dgnb for homozygous familial hypercholesterolemia. N Engl J Med. 2020; 383(8):711-20.
16. RxPipeline. Available with subscription at https://www.caremark.com/wps/portal/client. Accessed 2021 February 19.
17. Sjouke B, Kusters D, Kindt I et al. Homozygous autosomal dominant hypercholesterolemia in the Netherlands: prevalence, genotype-phenotype relationship, and clinical outcome. Eur Heart J. 2015; 36(9):560-5.
18. Rosenson RS, Durrington P. Familial Hypercholesterolemia in Adults: Treatment. In: UpToDate, Parikh N (ed). Waltham, MA. September 2023.
19. Lloyd-Jones DM, Morris PB, Ballantyne CM, et al. 2022 ACC Expert Consensus Decision Pathway on the Role of Nonstatin Therapies for LDL-Cholesterol Lowering in the Management of Atherosclerotic Cardiovascular Disease Risk: A Report of the American College of Cardiology Solution Set Oversight Committee. J Am Coll Cardiol. 2022 Oct 4;80(14):1366-1418. doi: 10.1016/j.jacc.2022.07.006.
20. Ahmad Z, Agarwala A, Cuchel M, et al. Update on familial hypercholesterolemia: An expert clinical consensus from the National Lipid Association. J Clin Lipidol. 2026 Apr;20(4):708-737. doi: 10.1016/j.jacl.2026.01.011. Epub 2026 Jan 29. PMID: 41741298.
21. Blumenthal RS, Morris PB, Gaudino M, et al. 2026 ACC/AHA/AACVPR/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA Guideline on the Management of Dyslipidemia: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. J Am Coll Cardiol. 2026 May 19;87(19):2624-2757. doi: 10.1016/j.jacc.2025.11.016. Epub 2026 Mar 13. PMID: 41824590.

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:

- November 14, 2023: Expanded age requirements of Evkeeza based on updated package labeling. Administrative updates to add Medical Benefit Drugs to title and update MATogether and RITogether fax numbers to 617-673-0939 (effective 12/1/23).
- August 13, 2024: Effective January 1, 2025, added ezetimibe to list of drug classes for member to demonstrate an inadequate response, contraindication, or intolerance
- August 12, 2025: Administrative update to remove Harvard Pilgrim Health Care Stride Medicare Advantage from the Medical Necessity Guideline template. Administrative update to rebrand Tufts Health Unify to Tufts Health One Care.
- August 11, 2026: Updated overview and age requirement of Evkeeza to at least 1 year of age based on updated package labeling. Administrative updates: Template updated to remove CareLink from the Medical Necessity Guideline template. Template updated from “Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans” to “Tufts Health Together- MassHealth Accountable Care Partnership Plans.”

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.