# Evkeeza™ (evinacumab-dgnb)
## (Intravenous)

**Effective Date: 07/01/2021**
**Dates Reviewed: 06/24/2021, 3/17/2022, 3/2/2023, 12/07/2023, 01/10/2024, 04/30/2025, 02/10/2026**
**Scope: Medicaid, Commercial, Medicare**

## I. Length of Authorization

Coverage is provided for three months for initial approval and may be renewed every 6 months.

## II. Dosing Limits

### A. Max Units (per dose and over time) [HCPCS Unit]:

- 1890 mg every 28 days
  *Vial strengths should be utilized to minimize drug waste based on calculated dose.

## III. Summary of Evidence

Evkeeza (evinacumab-dgnb) is an angiopoietin-like 3 (ANGPTL3) inhibitor indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients 1 years of age or older with homozygous familial hypercholesterolemia (HoFH). The ELIPSE-HoFH trial was a randomized, double-blind, placebo-controlled, 24-week clinical trial that evaluated percent change in LDL-C in 65 patients. Both groups continued to receive baseline background therapy, with 94% on statins, 75% on ezetimibe, 77% on a PCSK9 inhibitor, 22% on lomitapide, and 34% receiving lipoprotein apheresis. Baseline mean LDL-C was 255 mg/dL. The use of Evkeeza resulted in a 47% reduction in LDL-C from baseline compared with a 2% increase in the placebo group (mean treatment difference: -49%; p <0.0001). The most common adverse reactions of Evkeeza were nasopharyngitis, influenza-like illness, dizziness, rhinorrhea, and nausea.

## IV. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Medicare members who have previously received this medication within the past 365 days are not subject to Step Therapy requirements;
- Member is 1 year of age or older; **AND**
- Baseline low-density lipoprotein cholesterol (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high density lipoprotein cholesterol (non-HDL-C) labs must be obtained prior to initiating treatment (required for renewal); **AND**
- Member does not have heterozygous familial hypercholesterolemia (HeFH); **AND**

**Universal Criteria**

- Must be prescribed by, or in consultation with, a specialist in cardiology, lipidology, or endocrinology; **AND**

**Homozygous Familial Hypercholesterolemia (HoFH) † Φ** [1,2-11,12]

- Member has a confirmed diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) by any of the following:
  - Documented DNA test for functional mutation(s) in LDL receptor alleles or alleles known to affect LDL receptor functionality; **OR**
  - Untreated LDL-C > 500 mg/dL or treated LDL-C ≥ 300 mg/dL; **AND**
    - Cutaneous or tendon xanthoma before age 10 years; **OR**
    - Untreated LDL-C levels in both parents consistent with HeFH; **AND**
- Must be used as an adjunct to a low-fat or heart-healthy diet; **AND**
- Member has been receiving stable background lipid lowering therapy for at least 4 weeks; **AND**
- Therapy will be used in conjunction with diet and other LDL-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors, LDL apheresis); **AND**
- Member has tried and failed at least a 3-month trial of adherent therapy with: ezetimibe used in combination with the highest available (or maximally tolerated*****) dose of atorvastatin OR rosuvastatin, unless contraindicated; **AND**
- Member has tried and failed at least a 3-month trial of adherent therapy with: combination therapy consisting of the highest available (or maximally tolerated*****) dose of atorvastatin OR rosuvastatin, ezetimibe, AND a PSCK9 inhibitor indicated for HoFH (e.g., evolocumab, alirocumab), unless contraindicated; **AND**
- Despite pharmacological treatment with a PCSK9 inhibitor, statin, and ezetimibe, the member's LDL cholesterol ≥ 100 mg/dL [or ≥ 70 mg/dL for members with clinical atherosclerotic cardiovascular disease (ASCVD)]; **AND**
- Evkeeza (evinacumab) will not be used in combination with Juxtapid (lomitapide)

**†** FDA Approved Indication(s); **‡** Compendia recommended indication(s); Φ Orphan Drug

*****If the member is not able to use a maximum dose of atorvastatin or rosuvastatin due to muscle symptoms, documentation of a causal relationship must be established between statin use and muscle symptoms.

- Documentation must demonstrate that the member experienced pain, tenderness, stiffness, cramping, weakness, and/or fatigue and all of the following:
  - Muscle symptoms resolve after discontinuation of statin; **AND**
  - Muscle symptoms occurred when re-challenged at a lower dose of the same statin; **AND**
  - Muscle symptoms occurred after switching to an alternative statin; **AND**
  - Documentation ruling out non-statin causes of muscle symptoms (e.g., hypothyroidism, reduced renal function, reduced hepatic function, rheumatologic disorders, such as polymyalgia rheumatica, steroid myopathy, vitamin D deficiency, or primary muscle disease); **OR**
- The member has been diagnosed with rhabdomyolysis associated with statin use
  - The diagnosis should be supported by acute neuromuscular illness or dark urine **AND** an acute elevation in creatine kinase [usually > 5,000 IU/L or 5 times the upper limit of normal (ULN)]

## V. Renewal Criteria[1,12-15,21,23]

Coverage can be renewed based upon the following criteria:

- Member continues to meet universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section IV; **AND**
- Absence of unacceptable toxicity from therapy. Examples of unacceptable toxicity include the following: severe hypersensitivity, etc.; **AND**
- Member has had a reduction in LDL-C greater than 20% when compared to the initial baseline labs; **AND**
- Member continues to adhere to diet and background lipid lowering therapy (e.g., statin, ezetimibe, PCSK9 inhibitor, LDL apheresis); **AND**
- Evkeeza (evinacumab) will not be used in combination with Juxtapid (lomitapide).

## VI. Dosage/Administration[1]

| Indication | Dose |
|---|---|
| Homozygous Familial Hypercholesterolemia (HoFH) | The recommended dose of Evkeeza is 15 mg/kg administered by intravenous (IV) infusion over 60 minutes once monthly (every 4 weeks).<br>• If a dose is missed, administer as soon as possible. Thereafter, Evkeeza should be scheduled monthly from the date of the last dose.<br>• Assess LDL-C when clinically appropriate. The LDL-lowering effect of may be measured as early as 2 weeks after initiation. |

## VII. Billing Code/Availability Information

HCPCS code:

- J1305 – Injection, Evinacumab-dgnb 5mg

NDC:

- Evkeeza 345 mg/2.3 mL (150 mg/mL) single-dose vial: 61755-0013-xx
- Evkeeza 1,200 mg/8 mL (150 mg/mL) single-dose vial: 61755-0010-xx

## VIII. References

1. Evkeeza [package insert]. Tarrytown, NY; Regeneron, Inc.; September 2025. Accessed January 2026.

2. Mozaffarian D, et al. Heart disease and stroke statistics--2015 update: a report from the American Heart Association. Circulation. 2015 Jan 27;131(4):e29-322. doi: 10.1161/CIR.0000000000000152. Epub 2014 Dec 17.
3. Rosenson RS, et al. Inherited disorders of LDL-cholesterol metabolism. UpToDate. Available at: http://www.uptodate.com/contents/inherited- disorders-of-ldl-cholesterol-metabolism?source=search_result&search=inherited+disorders+of+metabolism&selectedTitle=2%7E150#references. Accessed July 27, 2015.
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6. Gidding SS, Champagne MA, de Ferranti SD, et al. The Agenda for Familial Hypercholesterolemia: A Scientific Statement From the American Heart Association. Circulation. 2015 Dec 1;132(22):2167-92. doi: 10.1161/CIR.0000000000000297.
7. Lloyd-Jones DM, Morris PB, Ballantyne CM, 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 Oct 3;70(14):1785-1822.
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10. Jacobson TA, Maki KC, Orringer C, et al. National Lipid Association Recommendations for Patient-Centered Management of Dyslipidemia: Part 2. DOI: 10.1016/j.jacl.2015.09.002
11. Cuchel M, Bruckert E, Ginsberg HN, et al. Homozygous familial hypercholesterolaemia: new insights and guidance for clinicians to improve detection and clinical management. A position paper from the Consensus Panel on Familial Hypercholesterolaemia of the European Atherosclerosis Society. Eur Heart J. 2014 Aug 21;35(32):2146-57. doi: 10.1093/eurheartj/ehu274. Epub 2014 Jul 22
12. Raal FJ, Rosenson RS, Reeskamp et al; ELIPSE HoFH Investigators. Evinacumab for Homozygous Familial Hypercholesterolemia. N Engl J Med. 2020 Aug 20;383(8):711-720. doi: 10.1056/NEJMoa2004215.

## Appendix 1 – Covered Diagnosis Codes

| ICD-10 | ICD-10 Description |
|---|---|
| E78.00 | Pure Hypercholesterolemia, unspecified |
| E78.01 | Familial hypercholesterolemia |

## Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Articles may exist and compliance with these policies is required where applicable. They can be found at: http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/Article): N/A

| Medicare Part B Administrative Contractor (MAC) Jurisdictions | | |
|---|---|---|
| **Jurisdiction** | **Applicable State/US Territory** | **Contractor** |
| E (1) | CA, HI, NV, AS, GU, CNMI | Noridian Healthcare Solutions, LLC |
| F (2 & 3) | AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ | Noridian Healthcare Solutions, LLC |
| 5 | KS, NE, IA, MO | Wisconsin Physicians Service Insurance Corp (WPS) |
| 6 | MN, WI, IL | National Government Services, Inc. (NGS) |
| H (4 & 7) | LA, AR, MS, TX, OK, CO, NM | Novitas Solutions, Inc. |
| 8 | MI, IN | Wisconsin Physicians Service Insurance Corp (WPS) |
| N (9) | FL, PR, VI | First Coast Service Options, Inc. |
| J (10) | TN, GA, AL | Palmetto GBA, LLC |
| M (11) | NC, SC, WV, VA (excluding below) | Palmetto GBA, LLC |
| L (12) | DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA) | Novitas Solutions, Inc. |
| K (13 & 14) | NY, CT, MA, RI, VT, ME, NH | National Government Services, Inc. (NGS) |
| 15 | KY, OH | CGS Administrators, LLC |

**Policy Rationale:**

Evkeeza was reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use Evkeeza according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For Medicare members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.