

Evkeeza™ (evinacumab-dgnb) (Intravenous)

Document Number: IC-0591

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I. Length of Authorization

Coverage is provided for 3 months for initial approval and may be renewed every 12 months.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Evkeeza 345 mg/2.3 mL single-dose vial: 2 vials per 28 days
- Evkeeza 1200 mg/8 mL single-dose vial: 1 vial per 28 days

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

- 378 billable units (1890 mg) every 28 days

III. Initial Approval Criteria ¹

Coverage is provided in the following conditions:

- Patient is at least 5 years of age; **AND**
- Baseline low-density lipoprotein cholesterol (LDL-C) labs must be obtained prior to initiating treatment (required for renewal); **AND**
- Patient does not have a diagnosis of heterozygous familial hypercholesterolemia (HeFH); **AND**

Universal Criteria

- Must be prescribed by, or in consultation with, a specialist in cardiology, lipidology, or endocrinology; **AND**
- Will not be used in combination with lomitapide; **AND**

Homozygous Familial Hypercholesterolemia (HoFH) † Φ ^{1,3,5,6,11,12}

- Patient has a confirmed diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) by any of the following:
 - Confirmed 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**
- Patient has been receiving stable background lipid lowering therapy for at least 4 weeks; **AND**
- Therapy will be used in conjunction with other LDL-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors, LDL apheresis); **AND**
- Patient 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**
- Patient 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 PCSK9 inhibitor indicated for HoFH (e.g., evolocumab), unless contraindicated; **AND**
- Despite pharmacological treatment, unless contraindicated, with a PCSK9 inhibitor, statin, and ezetimibe, the patient's LDL cholesterol ≥ 100 mg/dL (or ≥ 70 mg/dL for patients with clinical atherosclerotic cardiovascular disease [ASCVD])

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

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

- Patient has evidence of 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**
 - 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) have been ruled out; **OR**
- The patient 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])

IV. Renewal Criteria ^{1,8}

Coverage can be renewed based upon the following criteria:

- Patient 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 III; **AND**
- Absence of unacceptable toxicity from therapy. Examples of unacceptable toxicity include: severe hypersensitivity reactions, etc.; **AND**

- Patient has had a reduction in LDL-C, when compared to the baseline labs (prior to initiating evinacumab); **AND**
- Patient continues to adhere to diet and background lipid lowering therapy (e.g., statin, ezetimibe, PCSK9-I, lomitapide, LDL apheresis)

V. Dosage/Administration ¹

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

VI. Billing Code/Availability Information

HCPCS code:

- J1305 – Injection, evinacumab-dgnb, 5 mg; 1 billable unit = 5 mg

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

VII. References

1. Evkeeza [package insert]. Tarrytown, NY; Regeneron, Inc.; March 2023. Accessed March 2023.
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, Durrington P. (2022) Familial hypercholesterolemia in adults: Overview. In: Freeman MW, Parikh N (Eds). UpToDate. Last updated: Oct 17, 2022. Accessed March 21, 2023. Available from: https://www.uptodate.com/contents/familial-hypercholesterolemia-in-adults-overview?search=Heterozygous%20Familial%20Hypercholesterolemia%20&source=search_result&selectedTitle=2~49&usage_type=default&display_rank=2.
4. Stone NJ, et al. 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*, 2014; 129(25 Suppl 2): S1–45.
5. Jacobson et al. National Lipid Association recommendations for patient-centered management of dyslipidemia: Part 1 – executive summary. *Journal of Clinical Lipidology*.

EVKEEZA™ (evinacumab) Prior Auth Criteria

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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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9. Jacobson TA, Ito MK, Maki KC, et al. National Lipid Association recommendations for patient-centered management of dyslipidemia: Part 1 – executive summary. *Journal of Clinical Lipidology*. 2014;8(5):473–488. DOI: 10.1016/j.jacl.2014.07.007.
10. Jacobson TA, Maki KC, Orringer C, et al. National Lipid Association Recommendations for Patient-Centered Management of Dyslipidemia: Part 2. *J Clin Lipidol*. 2015 Nov-Dec;9(6 Suppl):S1-122.e1. 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.
13. 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.

Appendix 1 – Covered Diagnosis Codes

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

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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: <https://www.cms.gov/medicare-coverage-database/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/LCA): 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

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- Qualified sign language interpreters
- Written information in other formats (large print, audio, accessible electronic formats, other formats)

Provides free language services to people whose primary language is not English, such as:

- Qualified interpreters
- Information written in other languages

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If you believe that PCHP has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, or sex, you can file a grievance with:

Grievance Specialist
PreferredOne Community Health Plan
PO Box 59052
Minneapolis, MN 55459-0052
Phone: 1.800.940.5049 (TTY: 763.847.4013)
Fax: 763.847.4010
customerservice@preferredone.com

You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, a Grievance Specialist is available to help you.

You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at <https://ocrportal.hhs.gov/ocr/portal/lobby.jsf>, or by mail or phone at:

U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1-800-368-1019, 800-537-7697 (TDD)

Complaint forms are available at <http://www.hhs.gov/ocr/office/file/index.html>.

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Fax: 763.847.4010
customerservice@preferredone.com

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You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at <https://ocrportal.hhs.gov/ocr/portal/lobby.jsf>, or by mail or phone at:

U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1-800-368-1019, 800-537-7697 (TDD)

Complaint forms are available at <http://www.hhs.gov/ocr/office/file/index.html>.

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