

Evkeeza® (evinacumab-dgnb) (Intravenous)

Document Number: IC-0591

Last Review Date: 08/04/2026

Date of Origin: 03/01/2021

Dates Reviewed: 03/2021, 07/2021, 10/2021, 04/2022, 04/2023, 04/2024, 08/2025, 11/2025, 08/2026

I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 6 months (180 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

II. Dosing Limits

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

- 345 billable units (1725 mg) every 28 days

III. Initial Approval Criteria ¹

Prior authorization validity is provided in the following conditions:

- Member is at least 1 year of age; **AND**
- Baseline low-density lipoprotein cholesterol (LDL-C) labs have been obtained prior to initiating treatment (required for renewal); **AND**
- Member 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**

Homozygous Familial Hypercholesterolemia (HoFH) † Φ ^{1,12,14-19}

- Member has a confirmed diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) by any of the following:
 - Confirmed DNA test for functional mutation(s) in low-density lipoprotein (LDL) receptor alleles or alleles known to affect LDL receptor functionality [i.e. *LDLR*, *LDLRAP1*, *ARH*, *PCSK9*, or *apoB* gene(s)]; **OR**
 - Untreated LDL-C > 400 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 other LDL-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors, lomitapide, bempedoic acid, LDL apheresis, etc.); **AND**
- Members prior therapy history includes one of the following:
 - A 6-month trial and failure of adherent therapy with maximally tolerated doses of lomitapide; **OR**
 - A 3-month trial and failure of adherent therapy with a combination regimen that includes ALL of the following, unless contraindicated:
 - Highest available (or maximally tolerated*) dose of atorvastatin OR rosuvastatin; **AND**
 - Ezetimibe; **AND**
 - A PCSK9 inhibitor indicated for HoFH (e.g., evolocumab, alirocumab); **AND**
- Despite receiving LDL-lowering therapy, the member's treated LDL-C remains ≥ 100 mg/dL (or remains above a lower specified LDL-C goal based on individual risk factors)

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

*If the member 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.

- Member 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 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])

IV. Renewal Criteria ^{1,17}

Prior authorization validity can be renewed based upon the following criteria:

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

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.; September 2025. Accessed July 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, Durrington P. (2025) Familial hypercholesterolemia in adults: Overview. In: Freeman MW, Botkin NF (Eds). UpToDate. Last updated: June 09, 2026. Accessed July 08, 2026. Available from: <https://www.uptodate.com/contents/familial-hypercholesterolemia-in-adults-overview>.
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*. 2014. Available at: <http://www.sciencedirect.com/science/article/pii/S1933287414002748>.
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.
8. Grundy SM, Stone NJ, Bailey AL, et al. AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA Guideline on the Management of Blood Cholesterol: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2018;000:e1–e120. DOI: 10.1161/CIR.0000000000000625.
 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.
 14. Wiegman A, Greber-Platzer S, Ali S, et al. Evinacumab for Pediatric Patients With Homozygous Familial Hypercholesterolemia. *Circulation*. 2024 Jan 30;149(5):343-353. doi: 10.1161/CIRCULATIONAHA.123.065529. Epub 2023 Oct 20. PMID: 37860863; PMCID: PMC10814999.
 15. Fornari E, Stefanutti C, Mancioppi V, et al. Safety and effectiveness of evinacumab in an infant with homozygous familial hypercholesterolemia: A new renaissance for the very young?. *J Clin Lipidol*. 2025;19(3):689-694. doi:10.1016/j.jacl.2025.02.012
 16. Cegla J, Walji S, Barton L, et al. Treatment of Homozygous Familial Hypercholesterolemia. *JACC Adv*. 2025 May;4(5):101708. doi: 10.1016/j.jacadv.2025.101708. Epub 2025 Apr 26. PMID: 40288084; PMCID: PMC12059681.
 17. Blumenthal, R, Morris, P, 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. *JACC*. 2026 May, 87 (19) 2624–2757. <https://doi.org/10.1016/j.jacc.2025.11.016>

18. Cuchel M, Raal FJ, Hegele RA, et al. 2023 Update on European Atherosclerosis Society Consensus Statement on Homozygous Familial Hypercholesterolaemia: new treatments and clinical guidance. *European Heart Journal*. 2023;44(25):2277-2291. doi:10.1093/eurheartj/ehad197
19. Warden B, Guyton J, Kovacs A, et.al. Assessment and management of statin-associated muscle symptoms (SAMS): A clinical perspective from the National Lipid Association. *Journal of Clinical Lipidology*, 2022; 17, 19-39. doi: [10.1016/j.jacl.2022.09.001](https://doi.org/10.1016/j.jacl.2022.09.001)

Appendix A – Non-Quantitative Treatment Limitations (NQL) Factor Checklist

Non-quantitative treatment limitations (NQLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	No: PA not a priority
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E78.010	Homozygous familial hypercholesterolemia [HoFH]

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

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents:

<https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications, including any preceding information, may be applied 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
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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