

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

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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 <sup>1</sup>

Submission of medical records (chart notes) related to the medical necessity criteria is **REQUIRED** on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Medical records may be submitted via direct upload through the PA web portal or by fax.

Coverage is provided in the following conditions:

- Patient is at least 12 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 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) † $\Phi$ 1,5,6,11,12

- 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  $\geq$  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, lomitapide, 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  $\geq$  100 mg/dL (or  $\geq$  70 mg/dL for patients with clinical atherosclerotic cardiovascular disease [ASCVD])

† FDA Approved Indication(s); ‡ Compendia recommended indication(s);  $\Phi$  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 <sup>1,8</sup>

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 <sup>1</sup>

Indication	Dose
Homozygous Familial Hypercholesterolemia (HoFH)	Administer 15 mg/kg as an intravenous (IV) infusion once monthly (every 4 weeks). <ul style="list-style-type: none"> <li>• If a dose is missed, administer as soon as possible. Thereafter, Evkeeza should be scheduled monthly from the date of the last dose.</li> <li>• Assess LDL-C when clinically appropriate. The LDL-lowering effect of may be measured as early as 2 weeks after initiation.</li> </ul>

## 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.; February 2021. Accessed March 2022.
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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>. Accessed July 29, 2015.
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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.
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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: <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/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