

## 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>

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 <sup>1,3,5,6,11,12</sup>

- 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 <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.; March 2023. Accessed March 2023.
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5. Jacobson et al. National Lipid Association recommendations for patient-centered management of dyslipidemia: Part 1 – executive summary. *Journal of Clinical Lipidology*.

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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-Statins 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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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.
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## 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/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

## Nondiscrimination & Language Access Policy

Aspirus Health Plan, Inc. complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability, sex, sexual orientation, or gender identity. *We* do not exclude people or treat them differently because of race, color, national origin, age, disability, sex, sexual orientation, or gender identity.

*We* will:

Provide free aids and services to people with disabilities to communicate effectively with us, such as:

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

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

- Qualified interpreters
- Information written in other languages

If *you* need these services, contact *us* at the phone number shown on the inside cover of this *contract*, *your* id card, or [aspirushealthplan.com](http://aspirushealthplan.com).

If *you* believe that *we* have failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, sex, sexual orientation, or gender identity, *you* can file a grievance with:

Nondiscrimination Grievance Coordinator  
Aspirus Health Plan, Inc.  
PO Box 1062  
Minneapolis, MN 55440  
Phone: 1.866.631.5404 (TTY: 711)  
Fax: 763.847.4010  
Email: [customerservice@aspirushealthplan.com](mailto:customerservice@aspirushealthplan.com)

*You* can file a grievance in person or by mail, fax, or email. If *you* need help filing a grievance, the Nondiscrimination Grievance Coordinator 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>.

## Language Assistance Services

**Albanian:** KUJDES: Nëse flitni shqip, për ju ka në dispozicion shërbime të asistencës gjuhësore, pa pagesë. Telefononi në 1.866.631.5404 (TTY: 711).

**Arabic:** تنبيه: إذا كنت تتحدث اللغة العربية، فإن خدمات المساعدة اللغوية متاحة لك مجاناً. اتصل بن اعلى رقم الهاتف 1.866.631.5404 (رقم هاتف الصم والبك : 711)

**French:** ATTENTION : Si vous parlez français, des services d'aide linguistique vous sont proposés gratuitement. Appelezle 1.866.631.5404 (ATS : 711).

**German:** ACHTUNG: Wenn Sie Deutsch sprechen, stehen Ihnen kostenlos sprachliche Hilfsdienstleistungen zur Verfügung. Rufnummer: 1.866.631.5404 (TTY: 711).

**Hindi:** \_यान द\_ : य\_द आप िहंदी बोलते ह\_ तो आपके िलए मु\_त म\_ भाषा सहायता सेवाएं उपल\_ध ह\_। 1.866.631.5404 (TTY: 711) पर कॉल कर\_।

**Hmong:** LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1.866.631.5404 (TTY: 711).

**Korean:** 주의: 한국어를 사용하시는 경우, 언어 지원 서비스를 무료로 이용하실 수 있습니다. 1.866.631.5404 (TTY: 711)번으로 전화해 주십시오.

**Polish:** UWAGA: Jeżeli mówisz po polsku, możesz skorzystać z bezpłatnej pomocy językowej. Zadzwoń pod numer 1.866.631.5404 (TTY: 711).

**Russian:** ВНИМАНИЕ: Если вы говорите на русском языке, то вам доступны бесплатные услуги перевода. Звоните 1.866.631.5404 (телетайп: 711).

**Spanish:** ATENCIÓN: si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al 1.866.631.5404 (TTY: 711).

**Tagalog:** PAUNAWA: Kung nagsasalita ka ng Tagalog, maaari kang gumamit ng mga serbisyo ng tulong sa wika nangwalang bayad. Tumawag sa 1.866.631.5404 (TTY: 711)

**Traditional Chinese:** 注意: 如果您使用繁體中文, 您可以免費獲得語言援助服務。請致電 1.866.631.5404 (TTY:711)。

**Vietnamese:** CHÚ Ý: Nếu bạn nói Tiếng Việt, có các dịch vụ hỗ trợ ngôn ngữ miễn phí dành cho bạn. Gọi số 1.866.631.5404 (TTY: 711).

**Pennsylvania Dutch:** Wann du Deitsch (Pennsylvania German / Dutch) schwetzsch, kannscht du mitaus Koschte ebbergricke, ass dihr helft mit die englisch Schprooch. Ruf selli Nummer uff: Call 1.866.631.5404 (TTY: 711).

**Lao:** ໄປ່ດຊາຍ: ຖ້າວ່າ ທ່ານເວົ້າພາສາ ລາວ, ການບໍລິການຊ່ວຍເຫຼືອດ້ານພາສາ, ໂດຍບໍ່ເສັຽຄ່າ, ແມ່ນມີພ້ອມໃຫ້ທ່ານ. ໂທ 1.866.631.5404 (TTY:711).