Neighborhood Health Plan OF RHODE ISLAND™
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Evkeeza™ (evinacumab-dgnb)
(Intravenous)

Effective Date: 07/01/2021
Dates Reviewed: 06/24/2021, 3/17/2021
Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

I. Length of Authorization

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

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

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements;
- Patient age 12 years 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**
- 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) † Φ [1,2-11,12]

- Patient 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**
- Patient 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**
- 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 PSCK9 inhibitor indicated for HoFH (e.g., evolocumab, alirocumab), unless contraindicated; **AND**
- Despite pharmacological treatment 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)]; **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 patient 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 patient 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 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[1,12-15,21,23]

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 the following: severe hypersensitivity, etc.; **AND**
- Patient has had a reduction in LDL-C greater than 20% when compared to the initial baseline labs; **AND**

- Patient 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).

V. 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). • 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:

- C9079 – 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

VII. References

1. Evkeeza [package insert]. Tarrytown, NY; Regeneron, Inc.; February 2021. Accessed March 2022.
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.
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. Accessed July 29, 2015.
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. 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