Evkeeza® (evinacumab-dgnb) (Intravenous)

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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]

For groups in scope for the Site of Care (SOC) specialty infusion program, all program requirements are met.

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); Φ 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

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

Non-quantitative treatment limitations (NQTLs) 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 NQTL 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