Evkeeza (evinacumab)

Override(s)	Approval Duration
Prior Authorization	Initial Approval: 6 months Continuation Approval: 1 year

Medications	Dosing Limit
Evkeeza (evinacumab) 345 mg/2.3 mL, 1200 mg/8 mL	15 mg/kg every 4 weeks

APPROVAL CRITERIA

Initial requests for Evkeeza (evinacumab) may be approved when the following criteria are met:

I. Individual is 5 years of age or older; **AND**
II. Documentation is provided that individual has Homozygous Familial Hypercholesterolemia (HoFH) verified by (Cuchel 2023):
   A. Presence of two mutant alleles at the LDLR, apoB, PCSK9 or ARH adaptor protein (LDLRAP1) gene locus; **OR**
   B. An untreated LDL-C concentration greater than 400 mg/dL (10 mmol/L) **AND one** of the following:
      1. Cutaneous or tendonous xanthoma before age of 10 years; **OR**
      2. Untreated LDL-C levels consistent with heterozygous familial hypercholesterolemia in both parents (greater than 190 mg/dL);

AND

III. Individual meets **one** of the following:
   A. Individual is on high intensity statin therapy or statin therapy at the maximum tolerated dose (high intensity statin is defined as atorvastatin 40 mg or higher **or** rosuvastatin 20 mg or higher) (AHA/ACC 2018); **OR**
   B. Individual is statin intolerant based on one of the following:
      1. Inability to tolerate at least two statins, with at least one started at the lowest starting daily dose, demonstrated by adverse effects associated with statin therapy that resolve or improve with dose reduction or discontinuation (NLA 2022); **OR**
      2. Statin associated rhabdomyolysis or immune-mediated necrotizing myopathy (IMNM) after a trial of one statin; **OR**
   C. Individual has a contraindication for statin therapy including but not limited to active liver disease, unexplained persistent elevation of hepatic transaminases or pregnancy;

AND

IV. Individual has had a trial (medication samples/coupons/discount cards are excluded from consideration as a trial) and inadequate response or intolerance to ezetimibe (AHA/ACC 2018);

AND

V. Documentation is provided that individual has had a trial and inadequate response or intolerance to proprotein convertase subtilisin kexin type 9 (PCSK9) inhibitor therapy (AHA/ACC 2018);

OR

VI. Documentation is provided that genetic testing has verified the individual is LDLR negative (NLA 2017);

OR

VII. Individuals 5 - 9 years of age who do not meet RN III, IV, V or RN III, IV, VI above, may be approved if the following criteria are met:

VIII. Documentation is provided that individual has Homozygous Familial Hypercholesterolemia (HoFH) verified by (Cuchel 2014; Singh 2015):

A. Presence of two mutant alleles at the LDLR, apoB, PCSK9 or ARH adaptor protein (LDLRAP1) gene locus; **OR**

B. An untreated LDL-C concentration greater than 400 mg/dL (10 mmol/L) **AND one** of the following:

1. Cutaneous or tendonous xanthoma before age of 10 years; **OR**
2. Untreated LDL-C levels consistent with heterozygous familial hypercholesterolemia in both parents (greater than 190 mg/dL);

AND

IX. Documentation is provided that individual has had a trial and inadequate LDL reduction with at least one other lipid lowering therapy (including but not limited to statin therapy, ezetimibe).

Continuation requests for Evkeeza (evinacumab) may be approved when the following criteria are met:

I. Individual continues to use in combination with lipid lowering therapy (including but not limited to maximally tolerated statin therapy, ezetimibe, PCSK9 inhibitor therapy and/or LDL-C apheresis); **AND**

II. Documentation is provided that individual has achieved LDL-C reduction.

Evkeeza (evinacumab) may not be approved when the above criteria are not met and for all other indications.

Key References:

1. Cheeley MK, Saseen JJ, Agarwala A, et. al. NLA scientific statement on statin intolerance: a new definition and key considerations for ASCVD risk reduction in the statin intolerant patient. *J Clin Lipidol*. 2022. https://doi.org/10.1016/j.jacl.2022.05.068.
2. 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: 2277-2291.

3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: July 24, 2024.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. Grundy SM, Stone NJ, Bailey AL, et al. 2018 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. J Am Coll Cardiol 2019;73:e285–350.
6. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
7. Orringer CE, Jacobson TA, Saseen JJ, et. al. Update on the use of PCSK9 inhibitors in adults: Recommendations from an Expert Panel of the National Lipid Association (NLA). *J Clin Lipidol*. 2017 Jul-Aug;11(4):880-890.
8. Rosenson RS, Durrington P. Familial hypercholesterolemia in adults: Overview. Last updated: December 10, 2023. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: July 24, 2024.
9. Rosenson RS, Durrington P. Familial hypercholesterolemia in adults: Treatment. Last updated: December 7, 2023. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: July 24, 2024.

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.