

PHARMACY POLICY STATEMENT

Marketplace

DRUG NAME	Evkeeza (evinacumab-dgnb)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Evkeeza, approved by the FDA in 2021, is an ANGPTL3 (angiopoietin-like 3) inhibitor indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 5 years and older, with homozygous familial hypercholesterolemia (HoFH). Evkeeza is the first ANGPTL3 inhibitor to be approved. ANGPTL3 is a protein in the liver that has a role in regulating lipid metabolism. Its inhibition reduces LDL, HDL, and triglycerides.

Evkeeza (evinacumab-dgnb) will be considered for coverage when the following criteria are met:

Homozygous Familial Hypercholesterolemia (HoFH)

For **initial** authorization:

1. Member is at least 5 years of age; AND
2. Medication must be prescribed by or in consultation with a lipid specialist or cardiologist; AND
3. Member has a diagnosis of homozygous familial hypercholesterolemia (HoFH) confirmed by one of the following:
 - a) Genetic testing confirmation of two mutant alleles in the *LDLR*, *Apo-B*, *PCSK9*, or *LDLRAP1* gene locus; OR
 - b) LDL-C > 400 mg/dL before any lipid-lowering drug treatment AND at least one of the following:
 - i) Cutaneous or tendon xanthoma before 10 years of age; and/or
 - ii) Evidence of heterozygous FH in both parents; AND
4. Chart notes must include documentation of baseline LDL-C above goal within the past 90 days; AND
5. Member is unable to achieve LDL-C goal (see Note) after 8-week trials with ALL of the following:
 - a) High-intensity /max-tolerated statin in combination with ezetimibe (unless there is documentation of clearly established statin intolerance or statin contraindication—see note*); and
 - b) PCSK9 inhibitor (e.g., Repatha or Praluent; prior authorization required) unless there is evidence of no LDL receptor function (receptor-negative type HoFH) or the member does not meet the labeled age of PCSK9 inhibitors; AND
6. Evkeeza will be used as an adjunct to other lipid-lowering treatments (e.g., statin, ezetimibe, PCSK9 inhibitor, LDL apheresis), unless contraindicated or intolerant; AND
7. Prescriber attests that the member will adhere to a low-fat diet and exercise regimen; AND
8. Evkeeza is not being concomitantly initiated with Juxtapid.
9. **Dosage allowed/Quantity limit:** 15 mg/kg IV infusion once monthly.

NOTE: The LDL-C goals for adults are <70 mg/dL or <55 mg/dL with additional ASCVD risk factors, or <135 mg/dL for children.

**NOTE: If not on statin therapy, member must have documented contraindication to all statin drugs or documentation of intolerance to at least 2 different statins, including low/moderate intensity or alternate dosing such as every other day.*

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes/labs show at least 15% LDL-C reduction since starting Evkeeza.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Evkeeza (evinacumab-dgnb) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
03/23/2021	New policy for Evkeeza (evinacumab-dgnb) created.
02/21/2022	Updated J code.
05/19/2023	Updated age limit. Added references. Added receptor-negative and age as exceptions to PCSK9 requirement. Specified baseline LDL must be above goal. Simplified statement regarding Juxtapid (does not prohibit using them together, but they may not be started at the same time).
05/07/2024	Updated references. Changed LDL cutoffs of 500 or 300 to 400; specified at least 15% LDL reduction for reauth; changed LDL goals of 100 or 70 to 70 or 55; changed statin/ezetimibe and PCSK9 trials from 90 days to 8 weeks (EAS/Cuchel 2023). Added note about statin intolerance.

References:

1. Evkeeza [package insert]. Tarrytown, NY; Regeneron Pharmaceuticals, Inc.; 2023.
2. Raal FJ, Rosenson RS, Reeskamp LF, et al. Evinacumab for Homozygous Familial Hypercholesterolemia. *N Engl J Med.* 2020;383(8):711-720.
3. Doggrell SA. Will evinacumab become the standard treatment for homozygous familial hypercholesterolemia?. *Expert Opin Biol Ther.* 2021;21(3):299-302.
4. Wiegman A, Gidding SS, Watts GF, et al. Familial hypercholesterolaemia in children and adolescents: gaining decades of life by optimizing detection and treatment. *Eur Heart J.* 2015;36(36):2425-2437. doi:10.1093/eurheartj/ehv157
5. Horton AE, Martin AC, Srinivasan S, et al. Integrated guidance to enhance the care of children and adolescents with familial hypercholesterolaemia: Practical advice for the community clinician. *J Paediatr Child Health.* 2022;58(8):1297-1312. doi:10.1111/jpc.16096
6. Robinson JG. Management of familial hypercholesterolemia: a review of the recommendations from the National Lipid Association Expert Panel on Familial Hypercholesterolemia. *J Manag Care Pharm.* 2013;19(2):139-149. doi:10.18553/jmcp.2013.19.2.139
7. Gidding SS, Champagne MA, de Ferranti SD, et al. The Agenda for Familial Hypercholesterolemia: A Scientific Statement From the American Heart Association [published correction appears in *Circulation.* 2015 Dec 22;132(25):e397]. *Circulation.* 2015;132(22):2167-2192. doi:10.1161/CIR.0000000000000297
8. France M, Rees A, Datta D, et al. HEART UK statement on the management of homozygous familial hypercholesterolaemia in the United Kingdom. *Atherosclerosis.* 2016;255:128-139. doi:10.1016/j.atherosclerosis.2016.10.017
9. Mach F, Baigent C, Catapano AL, et al. 2019 ESC/EAS Guidelines for the management of dyslipidaemias: lipid modification to reduce cardiovascular risk [published correction appears in *Eur Heart J.* 2020 Nov 21;41(44):4255]. *Eur Heart J.* 2020;41(1):111-188. doi:10.1093/eurheartj/ehz455
10. Watts GF, Gidding SS, Hegele RA, et al. International Atherosclerosis Society guidance for implementing best practice in the care of familial hypercholesterolaemia. *Nat Rev Cardiol.* 2023;20(12):845-869. doi:10.1038/s41569-023-00892-0
11. 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. *Eur Heart J.* 2023;44(25):2277-2291. doi:10.1093/eurheartj/ehad197

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