

PHARMACY POLICY STATEMENT

North Carolina 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 > 500 mg/dL before any treatment or LDL-C > 300 mg/dL if treated with a lipid-lowering drug AND **one** of the following:
 - i) Cutaneous or tendon xanthoma before 10 years of age; OR
 - ii) Untreated elevated LDL-C levels consistent with heterozygous FH in both parents; AND
4. Chart notes must include documentation of baseline LDL-C level above goal within the past 90 days; AND
5. Member is unable to achieve LDL-C goal (see Note below) after trials with **both** of the following:
 - a) 90-day trial of a high-intensity statin plus ezetimibe. If intolerance occurs, a second attempt must be initiated with a moderate or low-intensity statin + ezetimibe;
 - b) 90-day trial with a 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, 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 administered by intravenous infusion once monthly.

Note: The LDL-C goals are <100 mg/dL for adults 18 years or older, < 135 mg/dL for children, and < 70 mg/dL for adults with clinical ASCVD.

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

For **reauthorization**:

1. Chart notes along with recent labs have been provided showing a meaningful reduction of LDL-C level from baseline OR LDL-C is at goal.

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).

References:

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3. 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;35(32):2146-2157.
4. Doggrell SA. Will evinacumab become the standard treatment for homozygous familial hypercholesterolemia?. *Expert Opin Biol Ther.* 2021;21(3):299-302.
5. 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
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. 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

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