

Evkeeza® (evinacumab-dgnb)

When requesting Evkeeza® (evinacumab-dgnb), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-Approved Indication

Evkeeza is indicated as an adjunct to diet and exercise and other low-density lipoprotein cholesterol (LDL-C) lowering therapies to reduce LDL-C in adults and pediatric patients, aged 1 year and older, with homozygous familial hypercholesterolemia (HoFH).

Coverage Guidelines

Homozygous Familial Hypercholesterolemia (HoFH)

The individual must meet all of the following criteria for initial approval:

- Meets one of the following:
 - The diagnosis has been confirmed by genetic testing (e.g., pathogenic variants at the low-density lipoprotein receptor (LDLR), apolipoprotein B (APOB), proprotein convertase subtilisin kexin type 9 (PCSK9) or low-density lipoprotein receptor adaptor protein 1 (LDLRAP1) gene locus); OR
 - Has an untreated LDL-C level > 400 mg/dL AND meets one of the following:
 - Had clinical manifestations of HoFH before the age of 10 years (i.e., cutaneous xanthomas, tendon xanthomas, arcus cornea, tuberous xanthomas, or xanthelasma); OR
 - At least one parent of the patient had untreated LDL-C levels or total cholesterol levels consistent with familial hypercholesterolemia (e.g., untreated LDL-C level ≥ 190 mg/dL and/or untreated total cholesterol level > 250 mg/dL); OR
 - Has a treated LDL-C level ≥ 300 mg/dL AND meets one of the following:
 - Had clinical manifestations of HoFH before the age of 10 years (i.e., cutaneous xanthomas, tendon xanthomas, arcus cornea, tuberous xanthomas, or xanthelasma); OR
 - At least one parent of the patient had untreated LDL-C levels or total cholesterol levels consistent with familial hypercholesterolemia (e.g., untreated LDL-C level ≥ 190 mg/dL and/or untreated total cholesterol level > 250 mg/dL);
- Meets ONE of the following:
 - Is one year to less than 10 years of age; OR
 - Is 10 years of age or older and meets both of the following:
 - Has tried one proprotein convertase subtilisin kexin type 9 (PCSK9) inhibitor for ≥ 8 continuous weeks (e.g., Repatha [evolocumab],

Praluent [alirocumab] and the LDL-C level still remains ≥ 70 mg/dL OR is known to have two LDL-receptor negative alleles;

- Meets one of the following:
 - ❖ Has tried one high-intensity statin therapy (i.e., atorvastatin ≥ 40 mg daily; rosuvastatin ≥ 20 g daily [as a single entity or as a combination product]) along with ezetimibe (as a single entity or as a combination product) for ≥ 8 continuous weeks AND LDL-C level still remains ≥ 70 mg/dL; OR
 - ❖ Has experienced statin-related rhabdomyolysis
Note: rhabdomyolysis is associated with elevated creatine kinase levels at least 10 times the upper limit of normal, along with evidence of end organ damage which can include signs of acute renal injury (a ≥ 0.5 mg/dL increase in serum creatinine [SCr] levels or doubling of the SCr) and/or myoglobinuria [myoglobin present in urine]); OR
 - ❖ Has experienced skeletal-related muscle symptoms (i.e., myopathy or myalgia) that occurred while receiving separate trials of both atorvastatin and rosuvastatin (as a single entity or combination products) AND these muscle symptoms resolved upon discontinuation of each respective statin therapy (atorvastatin and rosuvastatin).

The individual must meet the following criterion for reauthorization:

- Has experienced a response to Evkeeza therapy. (e.g. decreasing LDL-C, total cholesterol, non-high-density lipoprotein cholesterol, or apolipoprotein B levels)
Note: An individual who is currently on therapy but has not been previously approved via EviCore coverage determination or is restarting therapy is reviewed under criteria for initial therapy.

Approval duration (initial and renewal): 12 months

Dosing Recommendation

The recommended dose is 15 mg/kg administered by intravenous infusion once every 4 weeks.

References

1. Evkeeza® intravenous infusion [prescribing information]. Tarrytown, NY: Regeneron; September 2025.
2. Raal FJ, Rosenson RS, Reeskamp LF, et al, for the ELIPSE HoFH investigators. Evkeeza for homozygous familial hypercholesterolemia. *N Engl J Med.* 2020;383(8):711-720.
3. Rosenson RS, Burgess LJ, Ebenbichler CF, et al. Evkeeza in patients with refractory hypercholesterolemia. *N Engl J Med.* 2020;383(24):2307-2319.
4. Raal FJ, Hovingh GK Catapano AL. Familial hypercholesterolemia treatments: guidelines and new therapies. *Atherosclerosis.* 2018;277:483-492.
5. 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;44:2277-2291.
6. Lloyd-Jones DM, Morris PB, Ballantyne CM, et al. 2022 ACC Expert Consensus Decision Pathway on the Role of Non-Statin Therapies for LDL-Cholesterol Lowering in the Management of Atherosclerotic Cardiovascular Disease Risk. *J Am Coll.* 2022;80(14):1366-1418.