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

The individual must meet **all** of the following criteria for 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, acrus 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, acrus 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 ≥ 1 year to < 10 years of age;

    OR

  - Is ≥ 10 years of age and meets both of the following:
    - Meets one of the following:
      - LDL-C level remains ≥ 70 mg/dL after a trial of one high-intensity statin therapy (i.e., atorvastatin ≥ 40 mg daily or rosuvastatin ≥ 20 mg daily) along with ezetimibe for ≥ 8 continuous weeks*;*

        OR

      - Has been determined to be statin intolerant by meeting ONE of the following:
        - 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
        - Meets all of the following:
          - Has experienced skeletal-related muscle symptoms (i.e., myopathy or myalgia) that occurred while receiving separate trials of both atorvastatin and rosuvastatin (as single-entity or combination products); AND
          - The skeletal-related muscle symptoms resolved upon discontinuation of each respective statin therapy (atorvastatin and rosuvastatin);
    - Meets ONE of the following:
      - LDL-C level remains ≥ 70 mg/dL after a trial of a PCSK9 inhibitor (i.e., Repatha or Praluent) for ≥ 8 continuous weeks;

        OR

      - Is known to have two LDL-receptor negative alleles;

The individual must meet the following criteria for reauthorization:

- Has experienced a response to 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: 12 months

Dosing Recommendations

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

References

1. Evkeeza™ injection for intravenous use [prescribing information]. Tarrytown, NY: Regeneron Pharmaceuticals; 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, Bruckert E, Ginsberg HN, et al, for the European Atherosclerosis Society Consensus Panel on Familial Hypercholesterolemia. Homozygous familial hypercholesterolemia: new insights and guidance for clinicians to improve detection and clinical management. A positive paper from the Consensus Panel on Familial Hypercholesterolaemia of the European Atherosclerosis Society. *Eur Heart J*. 2014;35:2146-2157.
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.