Reference number(s)
4672-A

STANDARD MEDICARE PART B MANAGEMENT

EVKEEZA (evinacumab-dgnb)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Evkeeza is indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 12 years and older, with homozygous familial hypercholesterolemia (HoFH).

Limitations of Use:
- The safety and effectiveness of Evkeeza have not been established in patients with other causes of hypercholesterolemia, including those with heterozygous familial hypercholesterolemia (HeFH).
- The effects of Evkeeza on cardiovascular morbidity and mortality have not been determined

All other indications will be assessed on an individual basis. Submissions for indications other than those enumerated in this policy should be accompanied by supporting evidence from Medicare approved compendia.

II. DOCUMENTATION

The following documentation must be available, upon request, for all submissions:
A. Current LDL-C level for both initial requests and continuation requests. The level must be dated within the six months preceding the authorization request.
B. Genetic testing or medical records confirming the diagnosis of HoFH
C. Medical records confirming the member is currently on lipid lowering therapy for both initial requests and continuation requests

III. CRITERIA FOR INITIAL APPROVAL

Homozygous familial hypercholesterolemia (HoFH)
Authorization of 6 months may be granted for treatment of homozygous familial hypercholesterolemia when all of the following criteria are met:
A. Member has a documented diagnosis of homozygous familial hypercholesterolemia confirmed by any of the following criteria:
   1. Mutations in two alleles at the *LDLR*, *APOB*, *PCSK9* or *LDLRAP1* gene locus

2. An untreated LDL-C of greater than 500 mg/dL or treated LDL-C greater than or equal to 300 mg/dL and either of the following:
   i. Presence of cutaneous or tendinous xanthomas before the age of 10 years
   ii. An untreated LDL-C level of greater than or equal to 190 mg/dL in both parents

B. Prior to initiation of treatment with the requested medication, both of the following criteria are/were met:
   1. Member has a treated LDL-C of greater than or equal to 100 mg/dL (or greater than or equal to 70 mg/dL with clinical atherosclerotic cardiovascular disease [ASCVD])
   2. Member is receiving stable treatment with at least 2 lipid-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors)

C. Member will continue to receive concomitant lipid-lowering therapy

D. Member is 12 years of age or older

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

Authorization for 12 months may be granted when all of the following criteria are met:

A. The member is currently receiving therapy with Evkeeza

B. Evkeeza is being used to treat an indication enumerated in Section III

C. Member is currently receiving concomitant lipid-lowering therapy

D. The member is receiving benefit from therapy. Benefit is defined as either of the following:
   1. LDL-C is now at goal
   2. Member has had at least a 40% reduction of LDL-C from baseline

V. REFERENCES

1. Evkeeza [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals Inc.; February 2021.
2. Raal FJ, Rosenson RS, Reeskamp LF, et al. Evinacumab for homozygous familial hypercholesterolemia. *N Engl J Med.* 2020;383:711-20. DOI: 10.1056/NEJMoa2004215.
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;35:2146-2157.
4. Grundy SM, Stone NJ, Bailey, AL, Beam C, Birtcher KK, Blumenthal RS, Braun LT, de Ferranti S, Faiella-Tommasino J, Forman DE, Goldberg R, Heidenreich PA, Hlatky MA, Jones DW, Lloyd-Jones D, Lopez-Pajares N, Ndumele CE, Orringer CE, Peralta CA, Saseen JJ, Smith SC Jr, Sperling L, Virani SS, Yeboah J. 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. Circulation. 2019;139:e1082– e1143. DOI: 10.1161/CIR.0000000000000625.
5. McGowan MP, Dehkordi, SHH, Moriarty PM, Duell PB. Diagnosis and Treatment of Heterozygous Familial Hypercholesterolemia. *J Am Heart Assoc*. 2019;8:e013225.