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.

<table>
<thead>
<tr>
<th>DATE</th>
<th>ACTION/DESCRIPTION</th>
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<tbody>
<tr>
<td>03/23/2021</td>
<td>New policy for Evkeeza (evinacumab-dgnb) created.</td>
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<tr>
<td>02/21/2022</td>
<td>Updated J code.</td>
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<tr>
<td>05/19/2023</td>
<td>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).</td>
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</table>

References:


Effective date: 01/01/2024
Revised date: 05/19/2023