CODING & BILLING GUIDE for EVKEEZA™ (evinacumab-dgnb)

A concise reference of diagnostic, administration, product, and revenue codes that can help payers recognize, process, and pay claims for EVKEEZA.

INDICATION
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.

IMPORTANT SAFETY INFORMATION
Contraindication
EVKEEZA is contraindicated in patients with a history of serious hypersensitivity reactions to evinacumab-dgnb or to any of the excipients in EVKEEZA. Serious hypersensitivity reactions, including anaphylaxis, have occurred.

Please see Important Safety Information throughout and click here for full Prescribing Information.
A resource for coding, billing, and reimbursement of EVKEEZA™ (evinacumab-dgnb)

This guide provides coding and billing information to assist with understanding the reimbursement of EVKEEZA administered in the office or hospital outpatient setting.

This resource includes:

- **Diagnosis coding**: International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) codes
- **Administration coding**: Current Procedural Terminology (CPT) codes
- **Product coding**: Healthcare Common Procedure Coding System (HCPCS) Level II codes
- **Revenue codes**
- **Product information**

The following annotated samples of the 2 most common billing claim forms from the Centers for Medicare & Medicaid Services (CMS) are available at www.cms.gov/Medicare/CMS-Forms/CMS-Forms-List for quick reference:

- CMS-1500 (print) or 837P (electronic) for physician office reimbursement
- CMS-1450 (also known as UB-04) (print) or 837I (electronic) for hospital outpatient reimbursement

The coding information discussed in this guide is provided for informational purposes only, is subject to change and interpretation, and should not be construed as legal advice. The codes listed herein may not apply to all patients or to all health plans. Conversely, additional codes not listed in this guide may apply to some patients. Providers should follow payer-specific coding requirements and exercise independent clinical judgment when selecting codes and submitting claims to accurately reflect the services and products furnished to a specific patient. Providers must determine whether it is appropriate to submit any particular claim for reimbursement. Information provided in this guide is effective as of April 2021.

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**IMPORTANT SAFETY INFORMATION (cont’d)**

**Warnings and Precautions**

**Serious Hypersensitivity Reactions**: Serious hypersensitivity reactions have occurred with EVKEEZA. If signs or symptoms of serious allergic reactions occur, discontinue EVKEEZA infusion, treat according to the standard-of-care, and monitor until signs and symptoms resolve.

Please see Important Safety Information throughout and click here for full Prescribing Information.
**Diagnosis coding:**
**ICD-10-CM codes**

At least 1 ICD-10-CM diagnosis code is required on all claim forms. The diagnosis code selected should reflect the highest level of specificity available as documented in the patient’s medical record.

The following ICD-10-CM diagnosis code is the most specific code available for homozygous familial hypercholesterolemia (HoFH), although it does not differentiate the type of familial hypercholesterolemia (FH).

<table>
<thead>
<tr>
<th>ICD-10-CM code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>E78.01</td>
<td>Familial hypercholesterolemia</td>
</tr>
</tbody>
</table>

**Administration coding:**
**CPT codes for EVKEEZA™ (evinacumab-dgnb)**

EVKEEZA™ (evinacumab-dgnb) is an angiopoietin-like 3 inhibitor indicated as an adjunct to diet and other LDL-C lowering therapies for the treatment of adult and adolescent patients aged 12 years and older with homozygous familial hypercholesterolemia (HoFH). EVKEEZA is delivered by intravenous (IV) infusion.

CPT codes are assigned by physicians and by hospitals for outpatient services involving the administration of medications, including EVKEEZA.

The following CPT codes could be relevant to the administration of EVKEEZA.

<table>
<thead>
<tr>
<th>CPT code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>96365</td>
<td>Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour</td>
</tr>
<tr>
<td>96413</td>
<td>Chemotherapy administration, intravenous infusion technique, up to 1 hour, single or initial substance/drug</td>
</tr>
</tbody>
</table>

**IMPORTANT SAFETY INFORMATION (cont’d)**
**Warnings and Precautions (cont’d)**

**Embryo-Fetal Toxicity:** EVKEEZA may cause fetal harm when administered to pregnant patients. Advise patients who may become pregnant of the risk to a fetus. Consider obtaining a pregnancy test prior to initiating treatment with EVKEEZA. Advise patients who may become pregnant to use effective contraception during treatment and for at least 5 months following the last dose.

Please see Important Safety Information throughout and click here for full Prescribing Information.
Product coding: HCPCS Level II codes for EVKEEZA™ (evinacumab-dgnb)

HCPCS Level II codes help identify medications, including EVKEEZA, and are assigned in addition to the CPT code.

Because EVKEEZA does not currently have product-specific HCPCS codes, default NOC (Not Otherwise Classified) codes are used instead. When using default NOC codes, Medicare and most other payers require that additional details be provided on the CMS-1500 or CMS-1450 (UB-04) claim form, such as the name of the drug, amount given, National Drug Code (NDC) number, and invoice price.

<table>
<thead>
<tr>
<th>HCPCS code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>J-code</td>
<td></td>
</tr>
<tr>
<td>J3490</td>
<td>Unclassified drugs</td>
</tr>
<tr>
<td>J3590</td>
<td>Unclassified biologics</td>
</tr>
</tbody>
</table>

C-codes for Medicare reimbursement

Hospitals use C-codes for Medicare claims. These codes may also be accepted by other payers but must be verified with each payer.

Until a product-specific HCPCS Level II code is created, hospital outpatient departments will likely use miscellaneous C-code C9399 (unclassified drugs or biologicals) when submitting claims to Medicare.

<table>
<thead>
<tr>
<th>Medicare C-code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>C9399</td>
<td>Unclassified drugs or biologicals</td>
</tr>
</tbody>
</table>

When the drug is being reported with a NOC code, as is the case for EVKEEZA, it is appropriate to bill:

- 1 claim line with the NOC code, 1 unit for the amount administered, and the prorated price

IMPORTANT SAFETY INFORMATION (cont’d)

Adverse Reactions

Common adverse reactions (≥5%) were nasopharyngitis (16%), influenza-like illness (7%), dizziness (6%), rhinorrhea (5%), and nausea (5%).

Please see Important Safety Information throughout and click here for full Prescribing Information.
Revenue coding for hospital administration

Revenue codes allow hospitals to capture cost data for billing of services provided.

<table>
<thead>
<tr>
<th>Revenue code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Administration</strong></td>
<td></td>
</tr>
<tr>
<td>0510</td>
<td>Clinic</td>
</tr>
<tr>
<td>0500</td>
<td>Outpatient services</td>
</tr>
<tr>
<td><strong>Drug</strong></td>
<td></td>
</tr>
<tr>
<td>0636</td>
<td>Drugs requiring detailed coding</td>
</tr>
<tr>
<td>0250</td>
<td>Drugs and biologicals</td>
</tr>
<tr>
<td>0260</td>
<td>IV therapy</td>
</tr>
</tbody>
</table>

Product information for EVKEEZA™ (evinacumab-dgnb)

<table>
<thead>
<tr>
<th>Drug name/strength</th>
<th>10-digit NDC #</th>
<th>11-digit NDC #</th>
</tr>
</thead>
<tbody>
<tr>
<td>EVKEEZA 345 mg/2.3 mL (150 mg/mL)</td>
<td>61755-013-01</td>
<td>61755-0013-01</td>
</tr>
<tr>
<td>EVKEEZA 1200 mg/8 mL (150 mg/mL)</td>
<td>61755-010-01</td>
<td>61755-0010-01</td>
</tr>
</tbody>
</table>

IMPORTANT SAFETY INFORMATION (cont’d)

Use in Specific Populations

**Pregnancy:** EVKEEZA may cause fetal harm when administered to a pregnant woman. Advise pregnant women of the potential risk to a fetus. If a patient becomes pregnant while receiving EVKEEZA, healthcare providers should report EVKEEZA exposure by calling 1-833-385-3392.

**Lactation:** There are no data on the presence of evinacumab-dgnb in human milk or animal milk, the effects on the breastfed infant, or the effects on milk production. Maternal IgG is known to be present in human milk. The developmental and health benefits of breastfeeding should be considered along with the mother’s clinical need for EVKEEZA and any potential adverse effects on the breastfed infant from EVKEEZA or from the underlying maternal condition.

Please see Important Safety Information throughout and click here for full Prescribing Information.
If you have questions about coding and billing for EVKEEZA™ (evinacumab-dgnb), please call us at 

1-877-EVKEEZA (1-877-385-3392) 
Monday–Friday, 9 AM–7 PM Eastern time

For returns of product damaged in shipment and unused drugs appropriately discarded, please call 1-877-EVKEEZA (1-877-385-3392) Monday–Friday, 9 AM–7 PM Eastern time

IMPORTANT SAFETY INFORMATION (cont’d)

Use in Specific Populations (cont’d)

Females and Males of Reproductive Potential: Consider pregnancy testing in patients who may become pregnant prior to starting treatment with EVKEEZA. EVKEEZA may cause fetal harm when administered to a pregnant woman. Females of reproductive potential should use effective contraception during treatment with EVKEEZA and for at least 5 months following the last dose of EVKEEZA.

Pediatrics: The safety and efficacy of EVKEEZA have not been established in pediatric patients with HoFH who are younger than 12 years old.

Please click here for full Prescribing Information.