**Appeal checklist and sample appeal letter**

If a health plan receives a prior authorization (PA) request and denies coverage for EVKEEZA for your patient, you may appeal the decision. You can use the checklist on page 3 to help ensure you have taken appropriate steps for a successful appeal, and you can use or adapt the sample appeal letter on page 4 if coverage for EVKEEZA is denied. The sample letter is provided for your guidance only.

Some health plans require an appeal letter along with additional documentation, such as:

* Appeal form, if provided by the plan
* Chart notes, including clinical cardiac history
* Lipid test results, including LDL-C
* Current or previous lipid therapies
* Family history of coronary disease, particularly if it occurred at an early age for the patient’s parents
* Supporting clinical studies
* Peer-reviewed literature
* Prescribing Information for EVKEEZA

It is important to note that supplying the information for the appeal does not guarantee the health plan will provide reimbursement for EVKEEZA. The information is not intended to substitute for or influence the physician’s independent medical judgment.

Visit **EVKEEZAhcp.com** for more information, including full Prescribing Information.

There are numerous reasons why health plans may deny a PA for EVKEEZA. Although the reasons vary by plan, some of the most common include:

* Errors in ICD-10-CM coding on the PA request
* Insufficient documentation on the PA request
* Health plan claims lack of medical necessity for EVKEEZA
* EVKEEZA is not covered by patient’s health plan because it is a rare disease medication, requiring escalation

Please keep in mind, just as reasons for denial vary, so do each health plan’s requirements for the appeal. It is important to check with the patient’s health plan to ensure you have all the information you need to proceed with the appeal.

ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*;LDL-C=low-density lipoprotein cholesterol.

**INDICATION**

EVKEEZA is an angiopoietin-like 3 (ANGPTL3) 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).

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 additional Important Safety Information on next page and** [**click here**](https://www.regeneron.com/sites/default/files/Evkeeza_PI.pdf) **for full Prescribing Information.**

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

**Warnings and Precautions**

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

**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 dosage.

**Adverse Reactions**

**Adults and Pediatric Patients (12 to 17 years):** Common adverse reactions (≥5%) were nasopharyngitis (16%), influenza-like illness (7%), dizziness (6%), rhinorrhea (5%), and nausea (5%).

**Pediatric Patients (5 to 11 years):** The safety profile was consistent with that observed in adults and pediatric patients aged 12 and older with the additional adverse reaction of fatigue in 3 (15%) patients.

**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.

**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 dosage of EVKEEZA.

**Pediatrics:** The safety and efficacy of EVKEEZA have not been established in pediatric patients with HoFH who are younger than 5 years old.

**Please** [**click here**](https://www.regeneron.com/sites/default/files/Evkeeza_PI.pdf) **for full Prescribing Information.**

For assistance, call us at **1-877-EVKEEZA** (1-877-385-3392) Monday–Friday, 9 am–9 pm Eastern time.



**Appeal chec****klist**

✔ **Determine** ifEVKEEZA is covered by the patient’s health plan for the appropriate diagnosis

✔ **Double check** the accuracy of the information provided on the initial PA request

* Patient information
* Coding—it is recommended to use the most specific applicable codes

✔ **Understand** the reason for the denial—it is often included in the Explanation of Benefits letter

✔ **Review** the plan’s appeal guidelines

* Deadline to submit appeal
* Timeline of review by health plan
* Number of appeals permitted
* Fax number or email address to be used to submit the appeal letter and any additional required information
* Required additional supporting documentation, such as:
	+ Appeal form, if provided by the plan
	+ Chart notes
	+ Test results
	+ Supporting clinical studies
	+ Prescribing Information for EVKEEZA

✔ **Clarify** any aspect of the appeal process with the health plan’s review department

✔ **Prepare** a written appeal. The appeal should be written by the physician (see sample letter on next page). In some cases, the patient can write the appeal

✔ **Gather** all required supporting documentation needed to help defend your rationale for coverage for EVKEEZA

✔ **Send** the written appeal, along with the supporting documentation, to the health plan for review

✔ **Follow up** with the plan on the status of the appeal

✔ **Save copies** of all appeal-related documentation, including:

* Documents submitted with the appeal letter
* Documents received from the patient’s health plan
* Health plan representative’s contact information

For assistance, call us at **1-877-EVKEEZA** (1-877-385-3392) Monday–Friday, 9 am–9 pm Eastern time.



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**Sample appeal letter**

[Use physician’s letterhead]

[Date]

[Health Plan Contact Name]

[Title]

[Health Plan Organization Name]

[Address]

[City, State ZIP]

Re: [Subject line]

Patient: [Patient Name]

Date of Birth:[Patient DOB]

Insurance Policy ID Number: [Policy ID Number]

Group Number: [Group Number]

Claim Number: [Claim Number]

Dear [Health Plan Contact Name],

I am writing on behalf of my patient, [Patient full name], to appeal your denial of coverage for EVKEEZA® (evinacumab-dgnb). It is my understanding that EVKEEZA was denied because [state the specific reason the PA was denied].

I would like to explain why EVKEEZA should be covered for [patient name]. Along with this letter, I am providing information about the patient’s medical history and diagnosis of [diagnosis] (ICD-10-CM code: [code]), a statement summarizing my treatment rationale, and other documents that support the medical necessity of EVKEEZA for treatment of this patient.

EVKEEZA 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 5 years and older with homozygous familial hypercholesterolemia (HoFH).

**Overview of HoFH**

Homozygous familial hypercholesterolemia (HoFH) is an ultra-rare, inherited genetic disorder of lipid metabolism.1,2 HoFH is characterized by markedly elevated plasma levels of LDL-C, and the current estimated prevalence is 1 in 250,000 individuals.1-4 HoFH occurs when two copies of the alleles bearing the familial hypercholesterolemia (FH)-causing genes are inherited, one from each parent.1 Physical signs are generally severe and occur at an earlier age in patients with HoFH.2 Patients with HoFH generally have poor response to standard drug therapy and poor prognosis.3,5-9

HoFH represents a considerable burden for patients due to physical signs and limitations caused by the disease, as well as a number of psychosocial factors.1

**Summary of Patient’s Medical History** *Complete this section based on your clinical judgement when providing diagnosis and explanation of medical condition.*

[Patient name] was diagnosed with [diagnosis] on [date]. I believe EVKEEZA is needed for the treatment of this patient. The patient’s medical history includes:

* [Information from clinical diagnosis
* Information that summarizes the patient’s treatment history
* Response to past therapies
* Any recent symptoms and conditions, if applicable
* Opinion of the patient’s prognosis with and without treatment with EVKEEZA]

Given [patient name]’s clinical condition, along with the information included in the supporting documentation, I ask you to reconsider your previous decision and to approve coverage for EVKEEZA, which is indicated by the US Food and Drug Administration for this condition.

On behalf of [patient name], I appreciate your reconsideration. If you require additional information, please contact me at [phone number]. Thank you in advance for your immediate attention to this request.

Sincerely,

[Physician’s name, degree(s), participating provider ID number and signature]

Enclosures: [Attach EVKEEZA Prescribing Information and any additional documentation, as appropriate]

**References: 1.** Cuchel M et al. *Eur Heart J*. 2014;35(32):2146-2157. **2.** Raal FJ et al. *Atherosclerosis*. 2018;277:483-492. **3.** U.S. Food and Drug Administration. April 1, 2021. Accessed October 25, 2022. https://www.fda.gov/drugs/news-events-human-drugs/fda-approves-add-therapy-patients-genetic-form-severely-high-cholesterol **4.** de Ferranti SD et al. *Circulation*. 2016;133(11):1067-1072. **5.** Ito MK et al. *Drugs.* 2015;75:1715-1724. **6.** Bruckert E. *Atheroscler Suppl*. 2014;15(2):26-32. **7.** Thompson GR et al. *Eur Heart J*. 2018;39(14):1162-1168. **8.** Tromp TR et al. *Lancet.* 2022;399(10326):719-728. **9.** Bajaj A and Cuchel M. *J Atheroscler Thromb*. 2022;29(8):1125-1135.