# Medical Drug Clinical Criteria

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| **Subject:** | Veopoz (pozelimab-bbfg) | | |
| **Document #:** | CC-0250 | **Publish Date:** | ~~10/23/2024~~10/24/2025 |
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## Table of Contents



## Overview

This document addresses the use of Veopoz (pozelimab-bbfg), a monoclonal antibody directed against the terminal complement protein C5. Veopoz is indicated for the treatment of adult and pediatric patients 1 year of age and older with CD55-deficient protein-losing enteropathy (PLE), also known as CHAPLE disease.

CHAPLE disease is an ultra-rare disorder affecting less than 100 individuals worldwide. Caused by a biallelic CD55 loss-of-function mutation, individuals with the disorder experience an overactive complement system, leading to damage to blood and lymph vessels in the upper digestive tract. Affected individuals are also susceptible to large-vein thrombosis and severe hypoalbuminemia leading to edema, abdominal pain, nausea, vomiting, diarrhea, and malnutrition. The disease typically manifests in childhood and can be life-threatening. Prior to the approval of Veopoz, treatment of CHAPLE disease included supportive therapy according to the individual's clinical condition. Veopoz works by preventing cleavage of C5, thereby stopping formation of the membrane attack complex and preventing over activation of the complement system.

The efficacy and safety of Veopoz was established based on a single-arm study where 10 diagnosed individuals were treated weekly with weight-based dosing of Veopoz. All 10 patients achieved normalization of serum albumin levels by week 12 of the study through at least 72 weeks of treatment. Additionally, all 10 patients experienced improvement in 4 clinical outcomes (including frequency of problematic abdominal pain, bowel movement frequency, facial edema severity and peripheral edema severity) with no worsening at week 24. Veopoz is given as a single weight-based intravenous loading dose followed by subcutaneous administration by a healthcare provider weekly.

As with other agents inhibiting the complement system, Veopoz carries a black box warning for serious meningococcal infections. Life threatening and fatal meningococcal infections have occurred in patients treated with complement inhibitors. At least 2 weeks prior to the first administration, patients should complete or update meningococcal vaccination (for serogroups A, C, W, and Y, and serogroup B), unless the risks of delaying therapy outweigh the risk of developing a meningococcal infection. Antibacterial drug prophylaxis should be provided for individuals indicated for urgent Veopoz treatment. Individuals are also at increased risk for invasive disease caused by *N. meningitidis*, even if they develop antibodies following vaccination. Monitor patients for early signs of meningococcal infections and evaluate immediately if infection is suspected.

## Clinical Criteria

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

**Veopoz (pozelimab-bbfg)**

Initial requests for Veopoz (pozelimab-bbfg) may be approved if the following criteria are met:

I. Individual is 1 year of age or older; **AND**

II. Individual has a diagnosis of CD55-deficient protein-losing enteropathy (PLE), also known as CHAPLE disease; **AND**
III. Diagnosis has been verified by all of the following (Ozen 2024):
    A. Documentation is provided showing biallelic CD55 loss-of-function mutation; **AND**
    B. Documentation is provided showing hypoalbuminemia (defined as serum albumin concentration of ≤3.2 g/dL); **AND**
    C. One or more of the following signs or symptoms within the last six months:
        1. Abdominal pain; **OR**
        2. Diarrhea; **OR**
        3. Peripheral edema; **OR**
        4. Facial edema; **OR**
        5. Infection with concomitant hypogammaglobulinemia; **OR**
        6. New thromboembolic event;

**AND**
IV. Individual has completed or updated meningococcal vaccination (for serogroups A, C, W, and Y, and serogroup B) at least 2 weeks prior to administration of the first dose of Veopoz (pozelimab-bbfg), unless the risks of delaying Veopoz (pozelimab-bbfg) outweigh the risk of meningococcal infection.

Requests for continued use of Veopoz (pozelimab-bbfg) may be approved if the following criteria are met:

I. Individual has a diagnosis of CD55-deficient protein-losing enteropathy (PLE), also known as CHAPLE disease; **AND**
II. At initiation of therapy, diagnosis was verified by all of the following (Ozen 2024):
    A. Documentation is provided showing biallelic CD55 loss-of-function mutation; **AND**
    B. Documentation is provided showing hypoalbuminemia (defined as serum albumin concentration of ≤3.2 g/dL); **AND**
    C. One or more of the following signs or symptoms within the six months prior to initiation:
        1. Abdominal pain; **OR**
        2. Diarrhea; **OR**
        3. Peripheral edema; **OR**
        4. Facial edema; **OR**
        5. Infection with concomitant hypogammaglobulinemia; **OR**
        6. New thromboembolic event;

    **AND**
~~I.~~III. Individual has completed or updated meningococcal vaccination (for serogroups A, C, W, and Y, and serogroup B); **AND**
~~II.~~IV. There is clinically significant improvement or stabilization in clinical signs and symptoms of the disease (including but not limited to normalization of serum albumin levels; improvement of abdominal pain, diarrhea, and/or edema).

Requests for Veopoz (pozelimab-bbfg) may not be approved for the following:

I. Individual has evidence of an active meningococcal infection; **OR**
II. When the above criteria are not met and for all other indications.

## Quantity Limits

**Veopoz (pozelimab-bbfg) Quantity Limits**

| Drug | Limit |
|---|---|
| Veopoz (pozelimab-bbfg) 400mg/ 2mL (200 mg/mL) single-dose vial | 10 mg/kg, up to a maximum of 800 mg [2 vials], once weekly |
| **Override Criteria** | |
| May approve a single 30 mg/kg loading dose at the initiation of therapy.<br><br>May approve up to 12 mg/kg, up to a maximum of 800 mg [2 vials], once weekly if there is inadequate clinical response after at least 3 weekly doses of the 10 mg/kg dosing. | |

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## Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

**HCPCS**

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| J9376 | Injection, pozelimab-bbfg, 1 mg [Veopoz] |

**ICD-10 Diagnosis**

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| D84.1 | Defects in the Complement System |

## Document History

Revised: 09/08/2025
Document History:

- 09/08/2025 – Annual Review: Add diagnosis confirmation to continuation criteria; wording and formatting updates. Administrative update to add documentation. Coding Reviewed: Updated description for HCPCS J9376.
- 09/09/2024 – Annual Review: Update references; update inclusion criteria per published study; include meningococcal vaccination requirement in continuation criteria; wording and formatting updates. Coding Reviewed: No changes.
- 09/11/2023 – Annual Review: Add new clinical criteria and quantity limit for Veopoz. Coding Reviewed: Added HCPCS code J3590, C9399. All diagnoses pend. 4/1/24 Added HCPCS J9376. Removed HCPCS J3590, C9399. Added ICD-10-CM D84.1.

## References


1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: August 25, 2025.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
4. Ozen A, Chongsrisawat V, Sefer AP, et al. Evaluating the efficacy and safety of pozelimab in patients with CD55 deficiency with hyperactivation of complement, angiopathic thrombosis, and protein-losing enteropathy disease: an open-label phase 2 and 3 study. *Lancet*. 2024;403(10427):645-656. doi:10.1016/S0140-6736(23)02358-9.


Federal and state laws or requirements, contract language, and Plan utilization management programs or policies may take precedence over the application of this clinical criteria.