Veopoz® (Pozelimab-Bbfg) (for Ohio Only)

Policy Number: CSOH2025D0128.C
Effective Date: July 1, 2025

➲ Instructions for Use

Table of Contents **Page**

Related Policies
None

Application

This Medical Benefit Drug Policy only applies to the state of Ohio. Any requests for services that are stated as unproven or services for which there is a coverage or quantity limit will be evaluated for medical necessity using Ohio Administrative Code 5160-1-01.

Coverage Rationale

Veopoz is proven for the treatment of CD55-deficient protein-losing enteropathy (PLE) (i.e., CHAPLE disease).

Veopoz is medically necessary for the treatment of CD55-deficient protein-losing enteropathy (PLE) (i.e., CHAPLE disease) when all of the following criteria are met:

- **Initial Therapy**
  - Diagnosis of CD55-deficient PLE or CD55 deficiency with complement hyperactivation, angiopathic thrombosis, protein-losing enteropathy (i.e., CHAPLE disease); **and**
  - Submission of medical records (e.g., chart notes, laboratory values) confirming a biallelic loss-of-function mutation in the *CD55* gene; **and**
  - Laboratory results, signs, and/or symptoms attributed to CHAPLE disease (e.g., hypoalbuminemia, peripheral or facial edema, abdominal pain, diarrhea, etc.); **and**
  - Patient is not receiving Veopoz in combination with another complement protein C5 inhibitor [e.g., Soliris (eculizumab), Ultomiris (ravulizumab)] for the treatment of CHAPLE disease; **and**
  - Dosing is in accordance with the United States Food and Drug Administration approved labeling; **and**
  - Prescribed by a hematologist or other specialist with expertise in the diagnosis of CHAPLE disease; **and**
  - Initial authorization will be for no more than 12 months
- **Continuation of Therapy**
  - Documentation demonstrating a positive clinical response from baseline (e.g., normalization of serum albumin, reduction of peripheral or facial edema, reduction in abdominal pain or diarrhea); **and**
  - Patient is not receiving Veopoz in combination with another complement protein C5 inhibitor [e.g., Soliris (eculizumab), Ultomiris (ravulizumab)] for the treatment of CHAPLE disease; **and**
  - Dosing is in accordance with the United States Food and Drug Administration approved labeling; **and**
  - Prescribed by a hematologist or other specialist with expertise in the diagnosis of CHAPLE disease; **and**
  - Reauthorization will be for no more than 12 months

Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Listing of a code in this policy does not imply that the service described by the code is a covered or non-covered health service. Benefit coverage for health services is determined by federal, state, or contractual requirements and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

HCPCS Code	Description
J9376	Injection, pozelimab-bbfg, 1 mg

Diagnosis Code	Description
D84.1	Defects in the complement system

Background

Pozelimab-bbfg is a human, monoclonal immunoglobulin G4P (IgG4P) antibody directed against the terminal complement protein C5 that inhibits terminal complement activation by blocking cleavage of C5 into C5a (anaphylatoxin) and C5b, thereby blocking the formation of the membrane-attack complex (C5b-C9, a structure mediating cell lysis).

Clinical Evidence

Proven

CD55-Deficient Protein-Losing Enteropathy (PLE)

The efficacy and safety of Veopoz were evaluated in a single-arm study (NCT04209634) where outcomes were compared to pre-treatment data in patients with active CD55-deficient protein-losing enteropathy (PLE) who had hypoalbuminemia. Diagnosis was based on a clinical history of PLE and with a confirmed genotype of biallelic CD55 loss-of-function mutation. Active CD55-deficient PLE was defined as hypoalbuminemia (serum albumin concentration of ≤ 3.2 g/dL) with one or more of the following signs or symptoms within the last six months: abdominal pain, diarrhea, peripheral edema, or facial edema. Patients received a single 30 mg/kg loading dose of Veopoz administered by intravenous infusion over approximately one hour, followed by a once weekly weight-tiered maintenance dosage, administered as a subcutaneous injection starting one week after the loading dose. All patients received meningococcal vaccination prior to treatment with Veopoz and antibacterials for prophylaxis of meningococcal infection. Patients were permitted to receive additional therapies as part of standard of care. Use of other complement inhibitors was prohibited. Ten patients ranging from 3 to 19 years of age (median of 8.5 years) were assessed for efficacy. The mean baseline serum albumin concentration was 2.2 g/dL with a range of 1.1 to 2.9 g/dL. The median time for serum albumin to reach at least 3.5 g/dL was 15.5 days (n = 10; 95% CI: 8 to 28). All 10 patients achieved normalization by Week 12 and maintained serum albumin concentrations within the normal range through at least 72 weeks of treatment. At baseline, seven patients (70%) were evaluable for improvement in frequency of problematic abdominal pain (Figure S7), one patient (10%) was evaluable for improvement in the number of bowel movements per day, four patients (40%) were evaluable for improvement in facial edema severity, and five patients (50%) were evaluable for improvement in peripheral edema severity. Following 24 weeks of treatment with pozelimab, all patients that were evaluable at baseline met the criteria for improvement in these four prespecified clinical outcomes (frequency of problematic abdominal pain, bowel movement frequency, facial edema severity, and peripheral edema severity), with other patients experiencing no worsening. Five of the 10 patients received a total of 60 transfusions in the 48 weeks prior to treatment. In the 48 weeks after starting treatment, one patient received one albumin transfusion. Nine of the 10 patients were hospitalized for a total of 268 days in the 48 weeks prior to treatment. In the 48 weeks after starting treatment, two patients were hospitalized for a total of 7 days. Serum IgG concentrations reached normal values for age in all patients within the first 12 weeks of treatment; improvement was maintained through at least 72 weeks of treatment.

U.S. Food and Drug Administration (FDA)

This section is to be used for informational purposes only. FDA approval alone is not a basis for coverage.

Veopoz (pozelimab-bbfg) is a complement inhibitor 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.

References

1. Veopoz™ [prescribing information]. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; March 2024.
2. Ozen A, Comrie WA, Ardy RC, et al. CD55 Deficiency, Early-Onset Protein-Losing Enteropathy, and Thrombosis. N Engl J Med. 2017;377(1):52-61. doi:10.1056/NEJMoa1615887.
3. Open-Label Efficacy and Safety Study of Pozelimab in Patients With CD55-Deficient Protein-Losing Enteropathy (CHAPLE Disease). Clinicaltrials.gov website https://classic.clinicaltrials.gov/ct2/show/NCT04209634. Accessed March 12, 2025.
4. Ozen A, Chongsrisawat V, Sefer AP, et al., A Phase 2/3 Study Evaluating the Efficacy and Safety of Pozelimab in Patients with CD55 Deficiency with Hyperactivation of Complement, Angiopathic Thrombosis, and Protein-Losing Enteropathy (CHAPLE Disease). Available at SSRN: https://ssrn.com/abstract=4485593.
5. Yu CY, Ardoin SP. Complement inhibitor for therapy of CHAPLE. Nat Immunol. 2021;22(2):106-108. Doi:10.1038/s41590-020-00842-9.

Policy History/Revision Information

Date	Summary of Changes
07/01/2025	**Coverage Rationale** • Revised coverage criteria; replaced criterion requiring "the patient is not receiving Veopoz in combination with Soliris (eculizumab) or Ultomiris (ravulizumab)" with "the patient is not receiving Veopoz in combination with *another complement protein C5 inhibitor [e.g.,* Soliris (eculizumab), Ultomiris (ravulizumab)] *for the treatment of CD55-deficient protein-losing enteropathy (CHAPLE) disease*" **Supporting Information** • Updated *References* section to reflect the most current information • Archived previous policy version CSOH2024D0128.B

Instructions for Use

This Medical Benefit Drug Policy provides assistance in interpreting UnitedHealthcare standard benefit plans. When deciding coverage, the federal, state (Ohio Administrative Code [OAC]), or contractual requirements for benefit plan coverage must be referenced as the terms of the federal, state (OAC), or contractual requirements for benefit plan coverage may differ from the standard benefit plan. In the event of a conflict, the federal, state (OAC), or contractual requirements for benefit plan coverage govern. Before using this policy, please check the federal, state (OAC), or contractual requirements for benefit plan coverage. UnitedHealthcare reserves the right to modify its Policies and Guidelines as necessary. This Medical Benefit Drug Policy is provided for informational purposes. It does not constitute medical advice.

UnitedHealthcare may also use tools developed by third parties, such as the InterQual® criteria, to assist us in administering health benefits. The UnitedHealthcare Medical Benefit Drug Policies are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.