Medical Policy

Healthcare Services Department

Policy Name	Policy Number	Scope
Pozelimab-bbfg (Veopoz®)	MP-RX-FP-136-24	☒ MMM MA ☒ MMM Multihealth

Service Category

☐ Anesthesia
☐ Surgery
☐ Radiology Procedures
☐ Pathology and Laboratory Procedures
☐ Medicine Services and Procedures
☐ Evaluation and Management Services
☐ DME/Prosthetics or Supplies
☒ Part B Drugs

Service Description

This document addresses the use of *Pozelimab-bbfg (Veopoz®),* a complement inhibitor approved by the Food and Drug Administration (FDA) 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.

Background Information

Pozelimab is a 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).

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. 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.

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Approved Indications

A. Treatment of adult and pediatric patients 1 year of age and older with CD55-deficient protein-losing enteropathy (PLE), also known as CHAPLE disease.

Other Uses

i. None.

Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Benefit coverage for health services is determined by the member specific benefit plan document 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	Description
J9376	Injection, pozelimab-bbfg, 1 mg

ICD-10	Description
D84.1	Defects in the Complement System

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Medical Necessity Guidelines

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.

Pozelimab-bbfg (Veopoz®)

A. Criteria For Initial Approval *(Provider must submit documentation [such as office chart notes, lab results, pathology reports, imaging studies, and any other pertinent clinical information] supporting the patient's diagnosis for the drug and confirming that the patient has met **all** approval criteria.)*

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 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) a 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.

B. Criteria For Continuation of Therapy

i. MMM considers continuation of Pozelimab-bbfg (Veopoz®) therapy medically necessary in members requesting reauthorization for an indication listed in Section A above (Criteria for Initial Approval) if the following information is provided:
    A. Individual has completed or updated meningococcal vaccination (for serogroups A, C, W, and Y, and serogroup B); **AND**

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B. 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).

C. Authorization Duration

i. Initial Approval Duration: Up to 12 months
ii. Reauthorization Approval Duration: Up to 12 months

D. Conditions Not Covered

Any other use is considered experimental, investigational, or unproven, including the following (this list may not be all inclusive):

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

Limits or Restrictions

A. Therapeutic Alternatives

The list below includes preferred alternative therapies recommended in the approval criteria and may be subject to prior authorization.

i. N/A

B. Quantity Limitations

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines. The chart below includes dosing recommendations as per the FDA-approved prescribing information.

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Pozelimab-bbfg (Veopoz®) 400 mg/2 mL (200 mg/mL) SDV

Dose	Recommended Dosage	Limit
Day 1 (loading dose)	30 mg/kg IV	Once
Day 8 and thereafter (maintenance dosage)	10 mg/kg sc once weekly starting on Day 8	Up to 2 vials once weekly.
Exceptions		
• The maintenance dosage may be increased to 12 mg/kg once weekly if there is inadequate clinical response after at least 3 weekly doses (i.e., starting from Week 4). • The maximum maintenance dosage is 800 mg once weekly. • Doses greater than 400 mg require 2 injections. • Veopoz for subcutaneous use must be prepared and administered by a healthcare provider.		

Reference Information

1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 7, 2023.
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 polices may take precedence over the application of this clinical criteria.

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Policy History

Revision Type	Summary of Changes	P&T Approval Date	UM/CMPC Approval Date
Annual Review 12/26/2024	Updated clinical criteria for initial use per published study; included meningococcal vaccination requirement in initial approval and continuation criteria. Updated quantity limit table to add dosage forms and strenght, and quantity limits. Minor wording and formatting changes. Updated references. Coding Reviewed: Added HCPCS J9376. Removed HCPCS J3590, C9399. Added ICD-10-CM D84.1.	3/20/2025	4/2/2025
Policy Inception 01/30/2024	Elevance Health's Medical Policy adoption	N/A	6/28/2024