Veopoz® (pozelimab-bbfg)
(Intravenous/Subcutaneous)

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I. Length of Authorization

Coverage will be provided for twelve (12) months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Veopoz 400 mg/2 mL SDV – 8 vials, as a loading dose, on day 1 followed by 2 vials starting on day 8 and weekly thereafter

B. Max Units (per dose and over time) [HCPCS Unit]:

- Loading Dose: 3200 billable units on day 1
- Maintenance Dose: Beginning on day 8, up to 800 billable units weekly

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Patient is at least 1 year of age; **AND**

Universal Criteria [1]

- Patients must be administered a meningococcal vaccine (for serogroups A, C, W and Y, and serogroup B) at least two weeks prior to initiation of therapy and will continue to be revaccinated according to current medical guidelines for vaccine use *(If urgent Veopoz therapy is indicated in an unvaccinated patient, administer meningococcal vaccine(s) as soon as possible and provide patients with two weeks of antibacterial drug prophylaxis.)*; **AND**
- Patient must be administered vaccinations for the prevention of Streptococcus pneumoniae and Haemophilus influenzae type b (Hib) infections according to medical guidelines; **AND**
- Will not be used in combination with other complement therapies (i.e., sutimlimab, ravulizumab, avacopan, eculizumab, pegcetacoplan, etc.); **AND**
- Patient does not have an unresolved Neisseria meningitidis infection; **AND**

- Patient will avoid concomitant therapy with intravenous immunoglobulin, if therapy is unavoidable, the patient will be monitored closely for adverse reactions and/or worsening of disease symptoms; **AND**

Complement Hyperactivation, Angiopathic thrombosis, and Protein-Losing Enteropathy Disease (CHAPLE) † Φ [1]

- Patient has a confirmed clinical diagnosis of CD55-deficient protein-losing enteropathy (PLE) evidenced by biallelic CD55 loss-of-function mutation detected by genotype analysis; **AND**
- Patient has active disease as defined as hypoalbuminemia (serum albumin concentration of ≤3.2 g/dL) with one or more of the following signs or symptoms attributed to CD55-deficient PLE within the last six months:
  - abdominal pain
  - diarrhea
  - peripheral edema
  - facial edema

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria [1]

Coverage may be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: serious meningococcal infections (septicemia and/or meningitis), other serious bacterial infections, serious hypersensitivity reactions, etc.; **AND**
- Patient exhibits disease response compared to pretreatment baseline in ALL of the following:
  - Normalization/improvement in serum proteins (e.g., albumin, or immunoglobulin G, etc.); **AND**
  - Stabilization/improvement in signs and symptoms of disease; **AND**
  - Reduction in albumin transfusion requirements, exogenous immunoglobulin, and/or hospitalization (as applicable)

V. Dosage/Administration [1]

Indication	Dose
CHAPLE Disease	<u>Day 1 (Loading Dose):</u> Administer a single 30 mg/kg dose by intravenous infusion.

	Day 8 and Thereafter (Maintenance Dosage): **Inject 10 mg/kg as a subcutaneous injection* once weekly starting on Day 8** • *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 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.*

VI. Billing Code/Availability Information

HCPCS Code:

- J9376 – Injection, pozelimab-bbfg, 1 mg; 1 billable unit = 1 mg *(Effective 04/01/2024)*
- J3590 – Unclassified biologics *(Discontinue use on 04/01/2024)*

NDC(s):

- Veopoz 400 mg/2 mL single-dose vials for injection: 61755-0014-xx

VII. References

1. Veopoz [package insert]. Tarrytown, NY; Regeneron Pharmaceuticals, Inc; August 2023. Accessed August 2023.
2. 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). The Lancet PrePrint article, available at SSRN: https://ssrn.com/abstract=4485593 or http://dx.doi.org/10.2139/ssrn.4485593
3. Ozen A. CHAPLE syndrome uncovers the primary role of complement in a familial form of Waldmann's disease. Immunol Rev. 2019 Jan;287(1):20-32. doi: 10.1111/imr.12715. PMID: 30565236.
4. Ozen A, Comrie WA, Ardy RC, et al. CD55 Deficiency, Early-Onset Protein-Losing Enteropathy, and Thrombosis. N Engl J Med. 2017 Jul 6;377(1):52-61. doi: 10.1056/NEJMoa1615887. Epub 2017 Jun 28. PMID: 28657829; PMCID: PMC6690356.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D84.1	Defects in the complement system

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Articles (LCAs) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://wyww.cms.gov/medicare-coverage-database/search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC