Enjaymo® (sutimlimab-jome)
(Intravenous)

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I. Length of Authorization

- Coverage will be provided for 6 months and may be renewed.

II. Dosing Limits

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

- Enjaymo 1,100 mg/22 mL (50 mg/mL) in a single-dose vial:
  - 7 vials on Days 1 and 8, then 7 vials every 14 days thereafter

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

- 770 billable units weekly for two doses, then 770 billable units every 2 weeks thereafter

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Patient is at least 18 years of age; **AND**

Universal Criteria [1-2]

- Patient must be vaccinated against encapsulated bacteria (e.g., Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis, etc.) at least two weeks prior to initiation of therapy in accordance with the most current Advisory Committee on Immunization Practices (ACIP) recommendations and will continue to be revaccinated *(Note: If urgent therapy is indicated in an unvaccinated patient, administer vaccine(s) as soon as possible and provide patients with two weeks of antibacterial drug prophylaxis)*; **AND**
- Patient does not have an active chronic systemic infection (i.e., hepatitis B, hepatitis C, or HIV, etc.); **AND**
- Will not be used in combination with another complement-inhibitor therapy (i.e., ravulizumab, eculizumab, pegcetacoplan, avacopan, etc.) or B-cell directed therapy (i.e.,

rituximab*) (Note: Not applicable when sutimlimab is used as bridge therapy to B-cell directed treatment)*; **AND**

- Patient does NOT have systemic lupus erythematosus (SLE) or other autoimmune disease with positive anti-nuclear antibody; **AND**
- Patient will avoid cold exposure where possible; **AND**

Cold-Agglutinin Disease (CAD) † Φ [1-3]

- Patient has a confirmed diagnosis of CAD based on ALL of the following:
  - chronic hemolysis
  - poly-specific direct antiglobulin test (DAT)
  - monospecific DAT specific for C3d
  - cold agglutinin titer ≥64 at 4°C
  - IgG DAT ≤1+; **AND**
- Patient has one of the following:
  - Recent blood transfusion within the previous 6 months (i.e., transfusion dependent); **OR**
  - No recent blood transfusion (i.e., within the previous 6 months or a history of >1 blood transfusion within the previous 12 months); **AND**
    - Patient had an inadequate response, or has a contraindication or intolerance, to rituximab with/without bendamustine *(Note: Excludes patients who require urgent use of sutimlimab due to acute hemolysis where transfusion is likely);* ***AND***
- Other causes of secondary CAD have been ruled out such as coexisting diseases or conditions (i.e., infection, rheumatologic disease, systemic lupus erythematosus, or overt hematologic malignancy, etc.) [***NOTE:*** *patients with a history of or concomitant low-grade lymphoproliferative disease are not subject to exclusion*]; **AND**
- Documented baseline values for ALL of the following (necessary for renewal):
  - Hemoglobin level ≤ 10 g/dL
  - Packed RBC transfusion requirement *(**NOTE:** Only applies to patients that are transfusion dependent)*
  - Markers of hemolysis (e.g., indirect bilirubin, reticulocyte count, lactate dehydrogenase [LDH], haptoglobin, etc.)

† 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 infections (viral and bacterial), severe infusion reactions, autoimmune disease (e.g., SLE), etc.; **AND**
- Patient has experienced a disease response compared to pretreatment baseline based on at least one of the following:
  - Patient was transfusion dependent prior to starting treatment; **AND**
    - Hemoglobin response defined as an increase from baseline in Hgb level ≥2 g/dL or a Hgb level ≥12 g/dL without transfusion over a four week or longer time period; **OR**
    - Absence of packed RBC transfusion; **OR**
    - Patient had an increase in Hgb and/or decrease in transfusion requirement, to a lesser extent than the above, AND also had an improvement in the signs and symptoms (e.g., fatigue, jaundice, shortness of breath) and/or markers of hemolysis (e.g., indirect bilirubin, reticulocyte count, LDH, haptoglobin, etc.); **OR**
  - Patient did not have a recent history of blood transfusion prior to starting treatment; **AND**
    - Hemoglobin response defined as an increase from baseline in Hgb level ≥1.5 g/dL without transfusion over a four week or longer time period; **OR**
    - Patient had an increase in Hgb, to a lesser extent than the above, AND also had an improvement in the signs and symptoms (e.g., fatigue, jaundice, shortness of breath) and/or markers of hemolysis (e.g., indirect bilirubin, reticulocyte count, LDH, haptoglobin, etc.)

V. Dosage/Administration [1]

Indication	Dose*
Cold-Agglutinin Disease (CAD)	Administer intravenously weekly for the first two weeks, with administration every two weeks thereafter based on the following weight-based dosing.: – 39 kg to less than 75 kg: 6,500 mg – 75 kg or more: 7,500 mg

**Doses should be administered at the above intervals, or within two days of these time points. Patients with cardiopulmonary disease should receive the infusion over 120 minutes.*

VI. Billing Code/Availability Information

HCPCS Code:

- J1302 – Injection, sutimlimab-jome, 10 mg; 1 billable unit = 10 mg

NDC:

- Enjaymo 1,100 mg/22 mL single-dose vials of solution for injection: 80203-0347-xx

VII. References

1. Enjaymo [package insert]. Waltham, MA; Bioverativ USA, Inc; January 2023. Accessed September 2023.
2. Röth A, Barcellini W, D'Sa S, et al. Inhibition of Complement C1s with Sutimlimab in Patients with Cold Agglutinin Disease (CAD): Results from the Phase 3 Cardinal Study. Blood 2019; 134 (Supplement_2): LBA-2. doi: https://doi.org/10.1182/blood-2019-132490.
3. Hill QA, Stamps R, Massey E, et al on behalf of the British Society of Haematology. The diagnosis and management of primary autoimmune haemolytic anaemia. BJH. Volume176, Issue3. February 2017. Pages 395-411. https://doi.org/10.1111/bjh.14478.
4. Röth A, Berentsen S, Barcellini W, et al. Sutimlimab in patients with cold agglutinin disease: results of the randomized placebo-controlled phase 3 CADENZA trial. Blood. 2022 Sep 1;140(9):980-991. Doi: 10.1182/blood.2021014955.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D59.12	Cold autoimmune hemolytic anemia

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://www.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