Clinical Policy: Sutimlimab-jome (Enjaymo)

Reference Number: PA.CP.PHAR.503
Effective Date: 05/2024
Last Review Date: 04/2026

Description
Sutimlimab-jome (Enjaymo®) is a classical complement inhibitor.

FDA Approved Indication(s)
Enjaymo is indicated for the treatment of hemolysis in adults with cold agglutinin disease (CAD).

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of PA Health & Wellness® that Enjaymo is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Cold Agglutinin Disease (must meet all)**:**

1. Diagnosis of primary CAD;
2. Prescribed by or in consultation with a hematologist or oncologist;
3. Age ≥ 18 years;
4. Secondary CAD has been ruled out (i.e., cold agglutinin syndrome secondary to infection, rheumatologic disease, or active hematologic malignancy);
5. Member meets all of the following (a, b, c, and d):
   a. Active hemolysis as evidenced by elevated total bilirubin;
   b. Polyspecific direct antiglobulin test (DAT) (i.e., Coombs test) is positive;
   c. Monospecific DAT shows both of the following (i and ii):
      i. C3d DAT: strongly positive;
      ii. IgG DAT: negative or weakly positive;
   d. Cold agglutinin titer ≥ 64 at 4 degrees Celsius;
6. Hemoglobin ≤ 10 g/dL;
7. Enjaymo is not prescribed concurrently with rituximab or rituximab-based regimens (i.e., rituximab with bendamustine or fludarabine);
8. Dose does not exceed one of the following (a or b):
   a. For body weight 39 kg to < 75 kg: 6,500 mg (6 vials) on Day 0, Day 7, then every 2 weeks thereafter;
   b. For body weight ≥ 75 kg: 7,500 mg (7 vials) on Day 0, Day 7, then every 2 weeks thereafter.

Approval duration: 12 months

B. Other diagnoses/indications

1. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

II. Continued Therapy

A. Cold Agglutinin Disease (must meet all)**:**

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.PHARM.01) applies;
2. Member is responding positively to therapy as evidenced by one of the following since initiation of Enjaymo therapy (a or b):
   a. Increase in hemoglobin ≥ 1.5 g/dL or hemoglobin level ≥ 12 g/dL;
   b. Transfusion free or decreased number of transfusions/blood units;
3. If request is for a dose increase, new dose does not exceed one of the following (a or b):
   a. For body weight 39 kg to < 75 kg: 6,500 mg (6 vials) every 2 weeks;
   b. For body weight ≥ 75 kg: 7,500 mg (7 vials) every 2 weeks.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.PHARM.01) applies.
   Approval duration: Duration of request or 12 months (whichever is less); or
2. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

III. Diagnoses/Indications for which coverage is NOT authorized:

A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policies – PA.CP.PMN.53

IV. Appendices/General Information

Appendix A: Abbreviation/Acronym Key
CAD: cold agglutinin disease
DAT: direct antiglobulin test
FDA: Food and Drug Administration

Appendix B: Therapeutic Alternatives
Not applicable

Appendix C: Contraindications/Boxed Warnings

- Contraindication(s): known hypersensitivity to sutimlimab-jome or any inactive ingredients
- Boxed warning(s): none reported

Appendix D: Cold Agglutinins

- During passage through acral parts of the body, cooling of the blood allows cold agglutinins (CA) to bind to erythrocytes and cause agglutination.

- The antigen-IgM complex binds complement protein 1q (C1q) on the cell surface and initiates the classical complement pathway.
- C1 esterase activates C2 and C4, generating C3 convertase which results in the cleavage of C3 to C3a and C3b.
- Upon warming to 37°C in the central circulation, the CA detach from the cells, allowing agglutinated erythrocytes to separate, while C3b remains bound.
- C3b-opsonized cells are prone to phagocytosis by the mononuclear phagocytic system, mainly in the liver, a process known as extravascular hemolysis.
- On the surface of the surviving erythrocytes, C3b is cleaved, leaving high numbers of C3d molecules that can be detected by the DAT.

Berentsen S. How I manage patients with cold agglutinin disease. British Journal of Haematology. 2018;181:320–330.

V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
CAD	Weight-based dose IV weekly for 2 weeks then every 2 weeks thereafter: • 39 kg to < 75 kg: 6,500 mg (6 vials) • ≥ 75 kg: 7,500 mg (7 vials) Must be administered at the recommended dosage regimen time points or within 2 days of these time points	• 39 kg to < 75 kg: 6,500 mg/dose • ≥ 75 kg: 7,500 mg/dose

VI. Product Availability

Solution for injection in single-dose vial: 1,100 mg/22 mL (50 mg/mL)

VII. References

1. Enjaymo Prescribing Information. Bridgewater, NJ: Recordati Rare Diseases Inc.; November 2024. Available at https://www.enjaymohcp.com/. Accessed January 23, 2026.
2. A study to assess the efficacy and safety of BIVV009 (sutimlimab) in participants with primary cold agglutinin disease who have a recent history of blood transfusion (Cardinal Study). NCT03347396. ClinicalTrials.gov. Available at https://www.clinicaltrials.gov/ct2/show/NCT03347396. Accessed January 23, 2026.
3. A study to assess the efficacy and safety of BIVV009 (sutimlimab) in participants with primary cold agglutinin disease without a recent history of blood transfusion (Cadenza). NCT03347422. ClinicalTrials.gov. Available at https://clinicaltrials.gov/ct2/show/NCT03347422. Accessed January 23, 2026.
4. Ulrich J, D'Sa S, Schorgenhofer C et al. Inhibition of complement C1s improves severe hemolytic anemia in cold agglutinin disease: a first-in-human trial. Blood. February 28, 2019;133(9):893-901.
5. Hill QA, Stamps R, Massey E, et al. The diagnosis and management of primary autoimmune haemolytic anaemia. British Journal of Haematology. 2017;176:395-411. https://doi.org/10.1111/bjh.14478.

6. Bylsma LC, Ording AG, Rosenthal A, et al. Occurrence, thromboembolic risk, and mortality in Danish patients with cold agglutinin disease. Blood Adv. 2019 Oct 22;3(20):2980-2985. DOI:10.1182/bloodadvances.2019000476.
7. Berentsen S. How I manage patients with cold agglutinin disease. British Journal of Haematology. 2018;181:320-330.
8. Berentsen S, Ulvestad E, Langholm R, et al. Primary chronic cold agglutinin disease: a population based clinical study of 86 patients. Haematologica. 2006;91:460-466.
9. Röth A, Barcellini W, D'Sa S, et al. Sutimlimab in cold agglutinin disease. N Engl J Med. 2021;384(14):1323-1334. doi:10.1056/NEJMoa2027760.
10. Roth 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;140(9):980-991. doi: 10.1182/blood.2021014955.

Coding Implications
Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J1302	Injection, sutimlimab-jome, 10 mg

Reviews, Revisions, and Approvals	Date
Policy created	04/2024
2Q 2025 annual review: no significant changes; references reviewed and updated.	04/2025
2Q 2026 annual review: no significant changes; updated initial approval duration from 6 months to 12 months references reviewed and updated.	04/2026